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Valerie Newton

Paediatric Audiological Medicine

Second Edition

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SECOND EDITION

Edited by

Valerie E. Newton
Emerita Professor in Audiological Medicine, University of Manchester

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Contents

Contributors

P. Axon
Department of Otolaryngology
Addenbrooke's Hospital
Cambridge, UK

G. Baird
Newcomen Centre
Guy's and St Thomas' NHS Foundation Trust
London, UK

D. Bamiou
Academic Unit of Audiological Medicine
Institute of Child Health
London, UK

F.H. Bess
Department of Hearing and Speech Sciences
Vanderbilt Bill Wilkerson Center for Otolaryngology and Communication Sciences
Vanderbilt Medical Center
Nashville, Tennessee, USA

L.M. Bondurant
Department of Communicative Disorders
School of Behavioral and Brain Sciences
University of Texas at Dallas
Dallas, Texas, USA

R. Booth
University Department of Otolaryngology Head and Neck Surgery
Manchester Royal Infirmary
Manchester, UK

F. Calzolari
Department of Neuroradiology
University Hospital of Ferrara
Ferrara, Italy

C.B. Coelho
Department of Otolaryngology – Head and Neck Surgery
University of Iowa
Iowa City, USA

A. Davis
MRC Hearing and Communication Group
School of Psychological Sciences
University of Manchester
Manchester, UK

K. Davis
Maudsley Hospital
London, UK

J.W. Hall III
Department of Communicative Disorders
College of Public Health and Health Professions
University of Florida
Gainesville, Florida, USA

K. Harrop-Griffiths
Nuffield Speech and Hearing Centre
Royal National Throat, Nose and Ear Hospital
London, UK

R.W. Keith
Department of Communication Sciences and Disorders
College of Allied Health Sciences
University of Cincinnati
Cincinnati, Ohio, USA

R. Kentish
Nuffield Hearing and Speech Centre, Royal National Throat, Nose and Ear Hospital
London, UK

P.E. Klapper
Department of Clinical Virology
Clinical Sciences Building
Manchester Royal Infirmary
Manchester, UK

T. Loucas
School of Psychology and Clinical
Language Sciences
University of Reading
Reading, UK

D. Lucas
Nuffield Hearing and Speech Centre
Royal National Throat, Nose and Ear Hospital
London, UK

B. Mac Ardle
Nuffield Hearing and Speech Centre
Royal National Throat, Nose and Ear Hospital
London, UK

J. Mance
Nuffield Hearing and Speech Centre
Royal National Throat, Nose and Ear Hospital
London, UK

J.S. Martin
Callier Advanced Hearing Research Center
School of Behavioral and Brain Sciences
University of Texas at Dallas
Dallas, Texas, USA

A. Martini
Department of Audiology and
Otolaryngology
University of Ferrara
Ferrara, Italy

G. Mencher
MRC Hearing and Communication Group
School of Psychological Science
University of Manchester
Manchester, UK

C. Möller
Department of Audiology and Institute of
Disability Research
Örebro University Hospital
Örebro, Sweden

C. Munro
Nuffield Hearing and Speech Centre
Royal National Throat, Nose and Ear Hospital
London, UK

V.E. Newton
Emerita Professor of Audiological
Medicine
University of Manchester
Manchester, UK

G. Parker
Sheffield Teaching Hospitals and Sheffield
Children's Hospital
Sheffield, UK

E. Raglan
Department of Audiology and Audiological
Medicine
Great Ormond Street Children's Hospital
Bloomsbury, London, UK

R.T. Ramsden
University Dept of Otolaryngology Head
and Neck Surgery
University of Manchester
Manchester, UK

A.P. Read
Department of Medical Genetics
University of Manchester
Manchester, UK

A. Rothpletz
Department of Psychological and Brain
Sciences
Heuser Hearing Institute
University of Lousville
Louisville, Kentucky, USA

T. Sirimanna
Department of Audiology
Great Ormond Street Children's Hospital
Bloomsbury, London, UK

D.P. Sladen
Communication Sciences and Disorders
The University of Texas at Austin
Austin, Texas, USA

J. Stevens
Department of Medical Physics and Clinical Engineering
The Royal Hallamshire Hospital
Sheffield, UK

E.A. Tobey
Callier Advanced Hearing Research Center
University of Texas at Dallas
Dallas, Texas, USA

D. Toe
Early Learning, Development and Inclusion
Melbourne Graduate Faculty of Education
The University of Melbourne
Victoria, Australia

I. Tucker
Mary Hare Grammar School
Arlington Manor
Newbury, Berkshire, UK

F. Tweedy
University Department of Otolaryngology
Head and Neck Surgery
Manchester Royal Infirmary
Manchester, UK

R.S. Tyler
Department of Otolaryngology – Head and Neck Surgery
University of Iowa
Iowa City, Iowa, USA

P.J. Vallely
Virology, Genomic Epidemiology
School of Translational Medicine
University of Manchester
Manchester, UK

A.D. Warner-Czyz
Callier Advanced Hearing Research Center
University of Texas at Dallas
Dallas, Texas, USA

L. Watson
School of Education
University of Birmingham
Edgbaston, Birmingham, UK

Foreword

It is a great pleasure for both academic and personal reasons to support the publication of the second edition of **Paediatric Audiological Medicine**. Sophisticated new techniques have led to the acquisition of a vast body of knowledge, which has extended the boundaries of Audiological Medicine in the basic sciences underpinning this clinical discipline and allowing clearer understanding of the pathophysiology and thus management options. In addition, technology has enabled the development of new diagnostic and rehabilitation tools. The range and diversity of information frequently limits the individual clinician in accessing new developments in a manageable and understandable format. As in the first edition, emphasis has been placed on clarity of presentation across all aspects of hearing and balance, including the allied clinical disciplines. The text is a courageous attempt to synthesise what is valuable and necessary in all areas of diagnosis and management of both hearing and balance disorders in children, and in this regard provides a unique resource to promote the care of children with symptoms that have a major impact not only on their social and educational development, but also upon the whole family.

Professor Newton has striven to maintain and indeed develop the outstanding reputation established at the University of Manchester for the care of hearing-impaired children. She is internationally one of the foremost paediatric audiovestibular physicians and has contributed significantly to service development and improvements worldwide. In the United Kingdom her postgraduate teaching activity is renowned, but internationally she has contributed clinically, and in terms of teaching, in both the developed and developing world. Drawing on this broad expertise, the second edition of **Paediatric Audiological Medicine** builds on the outstanding first edition, and fills a critical need for a comprehensive resource on hearing and balance disorders in children. The book will be welcomed by scientists, physicians and surgeons managing these disorders, not least as a result of the outstandingly able multidisciplinary authorship that Professor Newton has attracted to contribute to this volume. In addition, the text will enthuse the reader to pursue and seek continuing developments to alleviate the suffering of children with inner ear disorders.

On a personal note, Professor Newton's outstanding contribution to teaching, research and clinical vision in Audiovestibular Medicine over many years should be acknowledged and I have no doubt that elements of these attributes will be apparent to every reader, who is fortunate enough to acquire a copy of this scholarly text. I wholeheartedly congratulate Professor Newton and her authors on their work.

Professor Linda Maitland Luxon
University College London Ear Institute

Preface

The First Edition of *Paediatric Audiological Medicine* was well received. Written by experienced academics and clinicians, it was the only book to provide the clinician with a comprehensive text on the range of topics pertinent to clinical practice. Since its publication there have been a number of significant developments which have improved diagnosis and management options, and the need for a Second Edition has become apparent.

In this Second Edition, there are 26 chapters, three of which are new to this book. Although covered within the previous edition, the increased knowledge accumulated and/or more emphasis on these subject areas have warranted separate chapters. To make room for these, the chapters on developmental anatomy and physiology have regretfully had to be omitted.

The first chapter on epidemiology provides information on the prevalence of disabling hearing impairment in children and the causative factors, many of which are potentially preventable. Early detection of a hearing impairment is vital for maximising habilitative potential. The next few chapters cover screening and detail the various methods currently available for identification of the type and degree of impairment.

Developments in the radiological field are comprehensively described in Chapter 5, which includes an ample number of supporting images. Radiology is increasingly important in helping to establish a diagnosis and in assisting clinicians in making management decisions.

The next few chapters relate to the main causes of a hearing impairment. Ascertaining that a hearing impairment is due to defective genes has implications for families, and in Chapter 6, the advances that have been made in reaching this diagnosis are recorded. Some of the syndromes which are inherited and which feature craniofacial abnormalities are described in the chapter that follows. Infections are particularly important causes of hearing impairment in developing countries and are potentially preventable. Chapter 8 gives an account of the range of congenital and acquired infections which can result in a hearing impairment, the pathology, methods of diagnosis and the means used for prevention. Perinatal factors have been associated with hearing loss and these factors are considered in Chapter 9. The main post-natal cause of hearing loss is otitis media, and the risk factors, diagnosis and management of this condition are among the aspects covered in Chapter 10.

There has been an increasing awareness in recent years of the number of children with central auditory processing disorders and of those with auditory neuropathy. Knowledge regarding the diagnosis and management of these conditions is still comparatively limited but information currently available is clearly outlined in separate chapters for each of these conditions.

Progressive hearing is of great concern to parents and professionals, and can be a devastating experience for the children concerned. In a chapter devoted to this topic, the causes of progressive hearing loss, management in the event of this occurring and some possibilities for prevention are among the areas discussed.

The management of children with a hearing impairment involves a number of different professionals and, to be most effective, needs to be in partnership with parents. The principles

underlying good management strategies and the hearing aid systems and assistive devices currently available are among the topics covered in three separate chapters. Appropriate selection of hearing aids and effective fitting are essential for maximum benefit to be obtained, and the methods of achieving this goal are included in Chapter 16. A new chapter in this edition stresses the importance of the acoustic environment for listening and the various assistive devices available to hearing-impaired children. The value of cochlear implantation for those children who cannot benefit adequately from hearing aids is now accepted and the chapter on this topic includes information on the impact of the communication prospects for children with very severe/profound hearing impairment.

Increasingly, clinicians have become aware of the need to be more informed regarding balance disorders in children and methods of assessing balance in this age group. An extra chapter has been included in this edition to give better coverage of this important field.

Tinnitus in children is often not recognised but can cause as many problems for children as for adults, so this topic is represented and its causes and management discussed. Unilateral hearing loss can also present children with difficulty and can be detrimental to their progress in an educational setting. The chapter on this topic explains the physical basis for these problems and suggests remedial solutions.

The need to communicate is a fundamental human need, and so three chapters are devoted to the development of speech and language in normal hearing and hearing-impaired children, delay and disorder in speech and language and the different communication options for hearing-impaired children. The chapter on the psychological effects of a hearing impairment shows how hearing impairment impacts upon the family as well as the child.

The chapter on educational provision for hearing-impaired children includes the changes that have taken place as a result of medical and technical advances and changes in UK legislation. This chapter completes the book.

The clinician working in the field of paediatric audiological medicine needs to have knowledge in many areas. In this one volume, the reader has been able to access a wide range of these essential topics. A comprehensive list of references has been included which can be used for further study.

Valerie Elizabeth Newton
Emerita Professor in Audiological Medicine
University of Manchester

1 Epidemiology of permanent childhood hearing impairment

A. Davis, K. Davis and G. Mencher

INTRODUCTION

Hearing impairment is the most frequent sensory impairment in humans, with significant social and psychological implications. The effect of the impairment will vary from individual to individual due to factors such as severity, age of onset, treatment / management options and the hearing status of their parents. It is likely that the greatest impact of hearing impairment upon a child is on the acquisition of language and development of communication, which in turn can lead to poor literacy skills[1,2] and altered long-term employment opportunities.[3,4] It is likely that other areas of development will also be affected, for example, mental health,[5,6] with one study finding 50% of a sample of hearing-impaired 11- to 16-year-olds met diagnostic criteria for a mental illness.[7]

Despite these difficulties, it is possible that, given adequate support, their impact may be reduced. For example, language development may be enhanced through the use of language support programmes, and residual hearing may be used effectively through adequate amplification from hearing aids or cochlear implants.[4,8] It has long been suspected that earlier diagnosis leads to better adjustment,[9] and evidence increasingly shows that a support programme starting in the first few months of life, used in tandem with early identification procedures, is beneficial for hearing-impaired children and their families (see Davis et al.[10] for an overview or Barton et al.[11] for an example). Support may be available through educational services, audiology services, social services and mental health services – and this support should be individualised, family-friendly and culturally sensitive.[10,12,13]

In the light of the impact that permanent childhood hearing impairment (PCHI) can have on children and their families, the importance of epidemiological studies cannot be underestimated. Epidemiological studies can provide information concerning the aetiology of hearing impairment and the groups within a population who are most at risk, which can be used to plan primary prevention by modifying relevant risk factors; it can be used to target those most likely to become hearing impaired and help detect them. They can also provide information on the overall prevalence of hearing impairment that can help estimate how many children have PCHI in different areas, helping plan secondary prevention of complications. Demographic and follow-up data can be used to make sure the services on offer are appropriate for users. For meaningful epidemiological studies, hearing impairment needs to be classified. Definitions may take into account not only the severity of the hearing impairment, but also the pathology and ontogenesis of the impairment, hence these factors are a major focus of this chapter.

DEFINITIONS USED IN EPIDEMIOLOGICAL STUDIES

Epidemiology is the study of how often diseases occur in different groups of people and why. When talking about research into hearing impairment, Sancho et al.[14] use the term 'epidemiology' to refer to 'the study of the distribution and determinants of hearing disorders in a population, and the application of the knowledge obtained to the prevention and amelioration of hearing problems'. A population study is the primary methodology for gathering information. The word 'population' in this case refers to the whole collection of units from which a sample may be drawn, but not necessarily to a population of people. For example, it may be a collection of hearing aid clinics or schools for the deaf. The sample is intended to give results that are representative of the population as a whole. A *cohort* is that component of a population born during a particular period and identified by period of birth, so that its characteristics (such as prevalence of childhood hearing impairment or age at first hearing aid fitting) can be ascertained as it enters successive time and age periods. If an epidemiological study follows a cohort and studies the group at several different intervals, the project is called a *cohort study*. A cohort study can be a follow-up study, a prospective study or a longitudinal study. It is essential for understanding change over time and the impact of services.

Another key term associated with epidemiology is *incidence.* This refers to the number of new instances of a specific condition (such as hearing impairment from meningitis) occurring during a certain period in a specified population. The incidence rate is the rate at which this occurs per standard population, for example 10 new cases per year per 100,000 children. The term *prevalence* is often confused with incidence. However, these are not the same thing. Prevalence is the total number of instances within a given population at a specific time in which a specific condition (for example, Pendred syndrome) is present. In the case of hearing impairment, prevalence may be described as 'the proportion of individuals with a defined type of hearing impairment in a specified population cohort'.[14] Accordingly, the prevalence rate is the number of individuals who have the condition or attribute divided by the population at risk at a point in time.

When attempting a prevalence study, if there are n children with hearing impairment in the study and the whole population is N, then the prevalence rate is $(n \times 100/N)\%$. In this case we must be sure that the n hearing-impaired children really come from all the birth cohorts of children represented by the population of N and that there is a coterminosity of n and N in terms of geographical boundaries. It is quite common to either underestimate n (because not all children with a given condition have been found) or to confuse populations (often because of migration of children into or out of particular districts).

THE DIFFICULTIES IN ESTIMATING PREVALENCE

Accurate estimations for the prevalence of childhood hearing impairment worldwide are hindered by the great difficulty in interpreting the data; perhaps leading to the variability in prevalence rates seen from study to study. These variations may be thought of as arising from three factors: how cases of hearing impairment are defined; how cases of hearing impairment are found; and the population from which the cases come. The importance of having agreed definitions for epidemiological studies, such as the ones outlined in the previous section, can be seen to be of paramount importance. The lack of agreed prevalence rates hinders investigation of possible risk factors and aetiologies, in turn, having implications for the planning of

service provision. Boxes 1.1 and 1.2 present the commonly used definitions for the various types of hearing impairment.

The term 'deaf' is generally associated with the most extreme form of hearing impairment, in which there is no response to auditory stimuli in excess of 120–125 dB at any frequency. This condition is practically never seen and is considered very rare. Hearing impairment, on the other hand, primarily refers to a series of descriptive terms that define the decibel level at which an individual responds to sound (see Box 1.2). Hearing impairment is also defined by the frequency range the person can hear. That is, a low-frequency range is <500 kHz; a mid-frequency range is 500 to 2,000 kHz; a high-frequency range is 2,000 to 8,000 kHz; and an extended high-frequency range is >8,000 kHz. The pattern of the frequencies is also important with some fairly self-explanatory terms, such as u-shaped, low-frequency ascending, flat and high-frequency sloping, used as descriptors of the responses plotted on an audiogram.

Box 1.1 Definitions of the various types of hearing impairment.

Type of impairment	Definition
Sensorineural	Related to disease/deformity of the inner ear/cochlear nerve with an air–bone gap less than 15 dB averaged over 0.5, 1 and 2 kHz
Conductive	Related to disease or deformity of the outer/middle ears. Audiometrically there are normal bone conduction thresholds (less than 20 dB) and an air–bone gap greater than 15 dB averaged over 0.5, 1 and 2 kHz
Mixed	Related to combined involvement of the outer/middle ears and the inner ear/cochlear nerve. Audiometrically greater than 20 dB HL in the bone conduction threshold together with greater than or equal to 15 dB air–bone gap averaged over 0.5, 1 and 2 kHz
Sensory	A subdivision of sensorineural related to disease or deformity in the cochlea
Neural	A subdivision of sensorineural related to a disease or deformity in the cochlear nerve
Central	Sensorineural hearing loss related to a disease or deformity of the central nervous system rostral to the cochlear nerve

Box 1.2 Definitions of hearing impairment in dB levels.

Type of impairment	Definition
Average hearing level	The level of the thresholds (in dB HL) measured in the better hearing ear at 0.5, 1, 2, 4 kHz
Mild	Average hearing level 20–39 dB HL
Moderate	Average hearing level 40–69 dB HL
Severe	Average hearing level 70–94 dB HL
Profound	Average hearing level +95 dB HL

Given the variety of types of hearing impairments presented in Box 1.1, it can easily be understood why there may be some confusion when attempting to define prevalence and/or incidence. However, the problem is compounded even further when various generalised categories for the course of the hearing impairment and the pattern of the hearing impairment are taken into consideration. Hearing impairment can be *congenital*, meaning to be present and detectable using appropriate tests at or very soon after birth, or *acquired*. However, there can be a difference in the meanings of these terms when considering aetiology as well as prevalence – as the cause of hearing impairment may be present at birth, but problems in hearing appear later in life. *Temporary* hearing impairment (usually, but not always, a conductive hearing impairment) can be treated and corrected by medical or surgical intervention. Such an impairment is often short-lived and of a mild nature. On the other hand, *permanent* hearing impairment cannot be readily treated by surgical or medical intervention. Both temporary and permanent hearing impairments can be *unilateral* (one ear only has either a greater than 20 dB hearing impairment through 500, 1,000 and 2,000 kHz or one frequency exceeding 50 dB, with the other ear normal) or *bilateral* (a greater than 20 dB hearing impairment through 500, 1,000 and 2,000 kHz or one frequency exceeding 50 dB in both ears). A unilateral situation is, of course, asymmetrical. However, in studies of hearing, the term *asymmetrical* hearing impairment specifically refers to a greater than 10 dB difference between the ears in at least two frequencies, with the pure-tone average in the better ear exceeding 20 dB HL. Finally, both temporary and permanent hearing impairments can be *progressive* – that is, there is a deterioration greater than or equal to 15 dB in the pure-tone average within a 10-year period.

Traditionally, studies have tended to be cross-sectional and based on retrospective ascertainment. A selection of these studies is shown in Table 1.1. It can be seen that estimates for prevalence of PCHI vary up to 10-fold (0.58 per 1,000, Baille et al.,[15] to 6.59 per 1,000, Parving and Hauch[16]) depending on definition, but most found levels of between 1.1 per 1,000 and 1.7 per 1,000 for their broadest definition.

Another method of cross-sectional study uses results from screening. This has the advantage of including cases that have not yet been diagnosed and works best for generating epidemiological data when the impairment is mild (and common) or where the whole population is screened. There are three common types of screen for hearing impairment: newborn; infant distraction test; sweep test.[23] A 'sweep' test asks a child to respond to low-intensity pure tones at three or four set frequencies, and has been done by school nurses and others. It has been used routinely on school entry in the UK since 1955, but protocol and implementation vary around the country, and there have been few attempts at measuring outcomes until recently. The infant distraction test assesses children as young as seven months by testing their behavioural response to noise, but it has the potential to miss serious cases, and refer many infants with no hearing problem. Again, despite being routine in the UK, sensitivity, specificity and outcome were not monitored. In the 1990s, technology became available to provide proxy measures of hearing in even newborn babies. Transient evoked otoacoustic emissions identify the presence or absence of outer hair cell activity from the inner ear, and auditory brainstem response (ABR) identifies the presence or absence of electrophysiological activity from the early auditory pathway. Automated equipment is now available for each of these tests, which can be used by trained screeners in the NICU, at the mother's bedside or in the community soon after birth. This has led to programmes of screening either high-risk babies or offering the test universally to every newborn baby. Universal newborn hearing screening (UNHS) has been has been recommended by the Joint Committee on Infant Hearing since 2000,[24] and by

Table 1.1 Selection of studies showing how population, definition of case and method of ascertainment can affect prevalence.

Study	Population	Definition of case			Method of detection	Prevalence per 1,000 children
		Threshold*	Congenital or acquired	Cause		
Russ et al. 2002[17]	Aged six living in Victoria, Australia born 1989	≥40 dB all with hearing aids (including unilateral)	Congenital	All	Hearing-aid clinic	1.24 2.09
Mytton and Mackenzie 2005[18]	Variable age living in Oldham, UK born 1991–2002 All racial origins	≥40 dB	Both	All	Audiology and educational sources	2.39
	Asian origin					4.64
Parving and Hauch 2001[16]	Aged <1–10 living in Copenhagen, Denmark born 1990–1999	>20 dB either ear at any frequency	Both	All	Surveillance programme of all hearing impaired	2.91
	Aged 11–20 living in Copenhagen, Denmark born 1980–1989					6.59
Fortnum and Davis 1997[19] and	Aged 5–10 living in Trent, UK, born 1985–1990	≥40 dB	Both	All	Audiology services	1.33
Fortnum et al. 2001[20]	Aged 5–10 living in UK, born 1988–1993	>40 dB			Audiology and education services	1.63
	Aged 3–8 living in UK, born 1985–1990				with adjustment for under-ascertainment	1.44
Baille et al. 1996[15]	Up to age nine, living in France, born 1976–1985	>70 dB	Both	All	Administrative departments	0.58
MacAndie et al. 2003[21]	Variable age living in Glasgow, UK born 1985–1994	≥40 dB	Both Congenital	All	Educational audiology database	1.23 1.09
Nekahm et al. 2001[22]	Variable age living in Tyrol, Austria born 1980–1994	≥40 dB	Both	All Sensorineural	Medical records	1.23 1.11

Threshold: Pure tone thresholds measured in dB HL (hearing level). Most studies averaged across thresholds at 0.5, 1, 2 and 4 kHz.
Better ear unless stated.

2005, approximately 95% of newborn infants in the United States were screened for hearing loss before they were 1 month old.[12] UNHS has been piloted in the UK since 2001 and became standard in England in 2006. Unlike its predecessors, it was accompanied by an IT system and rigorous quality control (see Bamford et al.[23] and the programme website http://hearing.screening.nhs.uk).

With the implementation of UNHS in some areas, more data are becoming available for estimating prevalence. Results from studies of UNHS express a 'rate' of hearing impairment detected per baby screened. Depending on the coverage the sensitivity of the test and the confidence of the diagnosis, prevalence of congenital hearing impairment can be made with increasing confidence. Uus and Bamford[25] gave the rates for the newborn hearing screening programme (NHSP) in England based on the 21 pilot sites around the UK between February 2002 and June 2004. A total of 169,487 babies were screened and amongst the children referred from the screen, a confirmed permanent bilateral hearing loss of moderate or greater severity was found in 169. This leads to a rate of 1.00 (95% confidence interval 0.78–1.22) per 1,000 babies screened. The programme achieved 96% coverage, and 90% of those babies who needed further tests were followed up. Yields of PCHI outside the UK have ranged from 0.68 per 1,000 in Western Australia[26] to 4.4 per 1,000 in Jackson, Mississippi, USA.[27] Trying to look for true geographical differences is once again difficult due to definition of a case, which sometimes includes unilateral and mild impairments, and other differences between studies. Some hospitals excluded results from NICU babies when reporting and other hospitals were tertiary referral centres handling very ill babies. Some programmes / studies found that effectiveness was limited by poor rates of screening or attending for follow-up. Vohr et al.[28] in Rhode Island, US, found that those with traditional Medicaid insurance were less likely to be screened or re-screened, whilst Prince et al.[29] in Hawaii found that low birth weight babies and those born to women who had not completed high school were twice as likely not to complete follow-up.

Prevalence of hearing impairment in the UK

Various studies have been carried out in the UK to ascertain accurate prevalence rates; however, there has been considerable disagreement between the rates established. Variation in sample populations, hearing levels included in the study, the fluctuating numbers of children with hearing impairment, and no easy way of ensuring complete ascertainment of cases were all factors that led to such variation in prevalence figures. With no agreement on numbers, there was uncertainty about the extent of the problem that extended into the 1990s. For example, in Nottingham, prevalence of PCHI was estimated at 0.55/1,000 by Pabla et al.[30] but 1.2/1,000 by Davis and Wood.[31]

An extensive study of epidemiology of PCHI was carried out for the Trent Regional Health Authority by Fortnum and Davis.[19] The aim was to include all children with a permanent hearing impairment of 40 dB HL average or greater in their better ear, who had been born between 1 January 1985 and 31 December 1993 and were living within the boundary of Trent Regional Health Authority at the time of data collection (June–September 1995). Sources of information included the Education Database, the Community Audiology and Child Health Database, the Neonatal Screening Database, audiology, medical records and hearing aid records. The data collected were divided into two main groups: congenital hearing impairment and acquired hearing impairment. The congenital group consisted of those children presumed to have had a prenatal or perinatal hearing impairment. The acquired

group included those whose hearing impairment came later in life due to disease, progressive hearing impairment or late-onset hearing impairment where there was evidence that the child may have been able to hear at an earlier stage. Prevalence rates of 1.3/1,000 for both acquired and congenital permanent hearing impairment were reported. For congenital hearing impairment alone, the prevalence rate was 1.1/1,000. Taking the prevalence estimates derived from the Trent region, it was possible to estimate that there would be approximately 1,000 children with a hearing impairment of at least moderate severity in the UK per annual birth cohort, around 84% having a congenital hearing impairment. These numbers undoubtedly contributed to a decision by the government of the UK to develop a newborn hearing screening programme.

For a more accurate calculation of prevalence across the whole of the United Kingdom, Fortnum et al.[20] approached the health professionals and the education professionals responsible for hearing-impaired children around the country, requesting details on every child with PCHI under their care. A total of 486 professionals replied, with over 26,000 sets of details. Many of these overlapped if the child was known to education and health services and the child's details were provided by both. The fact that there was no total overlap implies that there was some under-ascertainment. This can be adjusted for with a capture–recapture method – thus records for 17,160 children suggested there were around 21,500 children aged 3 to 18 in the UK with a permanent bilateral hearing impairment more than 40 dB. The inclusion of such a large number in the study allowed a more accurate breakdown into subgroups. It was shown that the observed prevalence increased with age until reaching a plateau at age 9, and that this was present at all three severities studied (41–70, 71–95, >95 dB HL). The adjusted prevalence at age 3 was around 1.1 per 1,000, rising to 2.1 per thousand at ages 9–16, a rise of 92%. This significant rise in prevalence during early childhood could be highly relevant for the planning of audiology and support services for secondary prevention of complications of hearing impairment, but this cross-sectional study is not ideal to confirm changes over time – because the change could arise either from the age of the cohort or the year in which the cohort was born.

Better ideas of change over time come from longitudinal studies, such as those carried out in the East London borough of Waltham Forest.[32] The relevant cohorts were born between 1992 and 2000 and numbered around 33,000. The numbers of children with PCHI, the method of identification and audiological data were collected from educational and audiology services. These children had UNHS, some had the infant distraction test, and they all had a school-entry 'sweep' screen. Newborn screening identified 1.58 per 1,000 children as having PCHI. More babies with PCHI were later identified due to concerns raised by parents or health visitors before the children were 12 months old in a further 0.24 cases per 1,000. A further 1.30 per 1,000 children were identified as having permanent hearing loss before they entered school at age 5, mainly due to parental concern. Finally, 0.34 per 1,000 were identified by the school-entry screen. This gives a combined total prevalence of 3.47 per 1,000 children by primary school age identified as having PCHI – of which 43% were of a moderate or greater severity bilateral hearing impairment, 35% mild bilateral and 22% unilateral (mild or above). This increase came partly from people moving into the area, but also from children who had not been offered, who had declined or who had failed to complete the screening process. Around 10% of the later identified children had a history of meningitis, 15% a family history of hearing impairment, and 30% had some other developmental abnormality (especially craniofacial). Thus, it seems there is a real increase in prevalence of PCHI as a cohort ages, and therefore a need for services to identify and manage this impairment.

Prevalence of hearing impairment in the United States

Early detection and hearing intervention (EDHI) programmes were legally required in 41 states by 2007[33] to help improve outcomes for children with hearing impairments. In order to design these programmes and their predecessors, an estimate of prevalence is essential. The Metropolitan Atlanta Developmental Disabilities study was partly set up to help develop methods of surveillance of children with special needs such as hearing impairment. The study collected data on the prevalence of mental retardation, cerebral palsy, hearing loss, vision impairment and epilepsy in children aged 10 years living in five counties in metropolitan Atlanta. Cases were actively sought from records at a number of sources, educational and medical, public and private, to maximise ascertainment. A hearing loss was defined as a permanent impairment of 40 dB HL averaged across thresholds at 0.5, 1 and 2 kHz in the better ear. Drews et al.[34] report on the prevalence of hearing loss in the cohort born during 1975, 1976 and 1977; who were age 10 in 1985–87. One hundred of the 10-year-old children had been identified and confirmed with PCHI out of a population of 89,534. This gives a prevalence of 1.1 (CI 0.9–1.4) per 1,000. Van Naarden et al.[35] looked at children aged 3 to 10 in 1991–1993, finding 411 cases, giving a prevalence of 1.10 (CI 1.00–1.20) per 1,000 children aged 3 to 10. The prevalence varied with age, being lowest at 0.67 per 1,000 3-year-olds and rising steadily to 1.38 per 1,000 10-year-olds. The latter number shows an apparent increase in prevalence from the previous study of 10-year-olds. Both studies found that about 30% of the children had another disability, the most common being mental retardation. They also found that the prevalence was around 20% higher amongst black residents than white. An accompanying paper[36] gives evidence for much of this difference being due to differing birth-weights: more babies with low birth weight are born to black mothers, and their outcome is less favourable than babies of low birth weight born to white mothers.

Another large population study to include childhood hearing impairment was the Third National Health and Nutrition Examination Survey (NHANES III), which used a 40,000-person sample with characteristics representative of the US population as a whole over 1988–1994. Niskar et al.[37] report on children aged 6 to 19 who were asked about their hearing status and screened using pure-tone audiometry in a mobile examination centre. Self-report of 'hearing difficulties' (not necessarily permanent) was 34 per 1,000 children. They screened using frequencies representing speech (0.5, 1 and 2 kHz) and at higher frequencies (3, 4 and 6 kHz), and defined hearing loss as average thresholds for either frequency above 15 dB HL. A hearing loss in at least one ear was present in 149 per 1,000 children (14.9%), most of which was unilateral and slight in severity, and some of which was likely to have been temporary or fluctuating. However, by extrapolating from their data, the authors estimate that there are 7 million US children at any one time that may need extra help in the classroom due to a hearing impairment. An accurate count of how many children nationwide are getting extra funding for special education can be obtained from the reports of the Individuals with Disabilities Education Act Program (IDEA-B). Data for the 2005 report are broken down by disability and age for children aged 3 to 16.[38] The number of children aged 3 to 16 for whom special education is funded due to a hearing impairment alone (there will also be children with PCHI in 'deaf-blind' or 'multiple disabilities' categories) was 70,702 – ranging from 2,174 aged 3 to 6,269 aged 12.

Universal hearing screening was taken up at differing rates across the United States, and a number of groups have published results from the early years of screening in individual hospitals and states – a selection of which is shown in Table 1.2. Once again, there is great diffi-

Table 1.2 Selected results from US screening programmes showing large variation in definition and rates.

Citation	Site	Definition	Rate per 1,000 screened
Barsky-Firsker and Sun 1997[41]	Tertiary referral centre, New Jersey	Sensorineural. Bilateral or unilateral. Not including babies from NICU.	2.1
Vohr et al. 1998[42]	Rhode Island (statewide)	Sensorineural and permanent conductive	2.0
Mason and Herrmann 1998[43]	University hospital, Honolulu, Hawaii	Bilateral loss requiring amplification	1.4
Finitzo et al. 1998[44]	Texas (multi-site)	Detectable permanent. Bilateral or unilateral	3.14
Dalzell et al. 2000[45]	New York (statewide)	Bilateral or unilateral	2.0
Stewart et al. 2000[46]	Kentucky (multi-site)	Sensorineural	2.7
Mehl and Thomson 2002[47]	Colorado (multi-site)	Sensorineural or permanent conductive. Bilateral	1.39
Connolly et al. 2005[27]	Tertiary referral centre, Jackson, Mississippi	Detectable permanent. Bilateral or unilateral	4.4

culty comparing between studies, and further difficulty in generalising from these yields to prevalence across the country since these pioneering programmes were likely to have taken place in large, well-resourced or well-motivated hospitals, many of whom had a large proportion of babies in NICU.[39] As part of EHDI, data are being collected by the Centers for Disease Control and Prevention (CDC). The annual data for 2005[40] show that of nearly 4 million births in states who were screening, 91.5% of babies were screened. Unfortunately, of the 64,421 babies referred for further screening or investigation, outcomes are only known for 43.5%, the majority of the rest being lost to follow-up or lost to documentation. The incidence of unilateral or bilateral PCHI of any severity reported to CDC is 0.92 per 1,000 babies screened, but this seems likely to be a vast underestimate of the actual rate.

Norton et al.[48] and Johnson et al.[49] have both re-screened babies at high risk of hearing impairment to test the sensitivity of newborn screening protocols. They found that although they are very good at detecting hearing loss of moderate severity or above, they miss a large proportion or slight and mild impairments. This must be clear to parents, professionals and those who plan provision for children with hearing loss.

Other 'developed' countries

Martin et al.[50] performed an ascertainment study of hearing-impaired children in the European Community (nine countries) who were born in 1969, were eight years old at the time of the study and who had a hearing impairment of at least 50 dB HL. They found a prevalence rate of 0.9 per 1,000. They also reported that 29% of the children had additional disabilities. These figures agree well with similar studies in the United States.[34] More recently, epidemiologists from European countries have compared prevalence of hearing loss from cohorts of children in the 1980s with the data from the Trent study in the UK.[19] In Denmark, Davis and Parving[51] reported on the prevalence of bilateral sensorineural or mixed PCHI of at least moderate severity (average threshold >40 dB HL), and the prevalence is shown by severity profile in

Table 1.3 Prevalence of all (congenital and acquired) permanent sensorineural and mixed childhood hearing impairments per 1,000 children in the birth cohort 1982–1988 for Denmark and children living in Trent Region, England, 1985–1993.

Country	40–130 dB HL	70–130 dB HL	95–130 dB HL
Denmark[51]	1.45	0.86	0.54
England[19]	1.32	0.59	0.31

Table 1.3 where it is compared with figures generated from the Trent study.[19] Approximately 90% of hearing impairment reported was congenital in both studies. It can be seen that there were significantly more severely and profoundly hearing-impaired children in Denmark than in England. When risk factors were investigated it was found that significantly more congenitally hearing-impaired children had an NICU history in England (33%) than in Denmark (17%), whereas more hearing-impaired children had a family history of hearing impairment in Denmark (40%) than in England (27%). Further work by Uus and Davis,[52] centred around the same issues in Estonia, reported that the prevalence of hearing impairment in Estonia (1.72 per 1,000) was higher than that of England (1.32 per 1,000) and Denmark (1.45 per 1,000).

Data from universal screening in parts of some countries are also available. Results from groups in Paris[53] and Siena[54] show rates of bilateral permanent hearing impairment >40 dB of 1.4 and 1.42 per 1,000 babies screened, respectively, which are slightly higher than the rate of 1.0 per 1,000 from the pilot sites in England, despite the fact the Paris study did not include babies from NICU. In Western Australia, the yield was found to be 0.68 per 1,000.[26] Out of 28,708 babies screened over 7 months, only nine babies were diagnosed with permanent bilateral hearing loss and eight of these had known risk factors for PCHI. This was seen to represent a poor detection rate, and universal screening was subsequently stopped, with a return to targeted screening. In Asia, the yield of bilateral PCHI detected per 1,000 children screened was 2.8 in a university hospital in Hong Kong[55] and 2.0 in a university hospital in Japan.[56] It is not immediately clear whether the difference in yields between countries, has to do with the performance of the screening programmes, the prevalence of risk factors or some specific environmental / genetic influence on the population. In Taiwan, two studies looked at the feasibility of screening in two environments: a hospital in Taipai[57]and a community-based screen in Tainan.[58] They achieved similar yields of 1.3 and 1.5 confirmed bilateral cases of hearing loss per 1,000 babies screened, but the lack of babies from NICU and the need for parents to choose to pay for the test makes the figures difficult to compare with those from England.

It thus may not be possible to generalise the findings of one study regarding prevalence to other geographical areas or, for that matter, to other birth cohorts. For that reason, epidemiological studies at the local level should be considered necessary to determine needs when planning for service provision.

Prevalence rates in disadvantaged countries

In order to obtain local data from developing countries, there have been some attempts to screen children, mainly of school age, in their communities, see Table 1.4. These have generally taken the form of pure-tone audiometry with or without otoscopy and tympanometry. Once again, the inconsistencies between the studies makes it difficult to compare them, and difficult to compare estimated prevalence with estimates from studies in developed countries. Neverthe-

Table 1.4 Selected results from child hearing screening studies in disadvantaged countries showing the differences in definitions and results.

Study and location	Population	Definition of case	Cases per 1,000
Abdel–Rahman et al. 2007[61] Ismailia, Egypt	Secondary school children	Sensorineural hearing loss ascertained by Rinne and Weber tests	222
Sobhy 1998[62] Alexandria, Egypt	School children	Excluding wax occlusion and OME Bilateral Average thresholds >25 dB	1.17–2.59
Seely et al. 1995[63] Pangama, Sierra Leone	Children	Bilateral Average thresholds >40 dB	6.5
Olusanya et al. 2000[64] Lagos, Nigeria	School children (mainstream school)	Conductive and sensorineural Unilateral or bilateral	139
Hatcher et al. 1995[65] Kiambu, Kenya	School children	Bilateral Average thresholds >30 dB	22
Westerberg et al. 2005[66] Zimbabwe	Primary school children	Sensorineural only Unilateral or bilateral Average thresholds >30 dB	10
Swart et al. 1995[67] Swaziland	First year school children	Sensorineural only Bilateral	2.1
Swart et al. 1995[67] Swaziland	First year school children	Middle-ear disease with hearing loss (conductive, mixed or sensorineural) Unilateral or bilateral	22
Minja et al. 1996[68] Rural Dar es Salaam, Tanzania	Primary school children	Sensorineural only	141
Minja et al. 1996[68] Urban Dar es Salaam, Tanzania	Primary school children	Sensorineural only	77
Elahi et al. 1998[69] Rural areas, Pakistan	Children	Sensorineural or permanent conductive Bilateral Average thresholds >30 dB	39
Rao et al. 2002[70] Rural south, India	First year school children	Sensorineural or mixed Unilateral or bilateral Average thresholds >30 dB	32
Liu et al. 2001[71] Sichuan, China	Children <15 y	Unilateral or bilateral Average thresholds >30 dB	2.6
Mencher and Madriz Alfaro 2000[72] Costa Rica	School children	Bilateral Permanent	1.50–1.63

less, there is a consensus that the levels of PCHI are greater in underdeveloped countries, with Davidson et al.[59] estimating that sensorineural loss is twice as common. Evidence also seems to point towards a higher rate of hearing impairment amongst disadvantaged communities in richer countries,[18,34] with Niskar et al.[37] finding that children from families with incomes at or below the national poverty line were significantly more likely to have a hearing impairment when screened. The World Health Organization in its report on chronic diseases[60] views the

process from poverty to chronic diseases as 'interconnected in a vicious cycle', as poor people have greater exposure to risks and decreased access to health services.

Alberti[73] estimated that half of all disabling hearing loss worldwide was preventable by primary means, from vaccination to better protection from noise exposure. Consanguinity is a common risk factor in some communities.[69] There also seems to be an increased prevalence of middle-ear disease in disadvantaged communities and this can be aggressive, becoming chronic suppurative otitis media (CSOM) or leading to cholesteatoma.[74] The presence of recurrent or chronic middle-ear disease is highly correlated with a permanent hearing loss in this population because of the reduced access to effective treatment.[69,75]

There is some impetus for an increased effort of identification of PCHI in developing countries, and this seems to be backed by the opinion of mothers.[76] Trials of UNHS at immunisation clinics have been undertaken in Nigeria and South Africa and were successful in terms of coverage, but the attendance at follow-up was poor. Swanepoel et al.[77] report that out of 68 subjects (14% of screened sample), only 40% returned for the second follow-up and 44% for the third follow-up. Some argue that primary prevention strategies should take priority, as the current high prevalence would overwhelm the capacity for early intervention.[78] Others would argue that with facilities available for deaf and hearing-impaired children throughout the world, children worldwide should be identified to take advantage of those facilities.[79]

RISK FACTORS

Risk in this context refers to an increased probability that an event will occur; in this case, that a child will have a hearing impairment. Factors that increase the likelihood can be non-specific, i.e. affecting a whole population but not by much, or can be specific to the child. The former is important to know for planning services, and examples might be poverty or being aged less than 9 years old. This section will concentrate more on the latter. Specific risk factors – the most notable being a family history of permanent hearing impairment present since childhood in a parent, sibling, grandparent, great-grandparent, aunt, uncle, nephew, niece or cousin or a lengthy stay in NICU – can be used for targeted screening during universal screening. The practice of targeted screening in babies was common between the invention of the technology for newborn screening and the infrastructure being put into place for UNHS. An example of this was in the Redbridge District of London, where there was a targeted newborn hearing screen for 10 years between 1990 and 2000.[80] From the 32,890 babies born, 3.5% were identified before discharge from hospital as high-risk using the appropriate JCIH guidelines at the time, and screened using ABR. The yield was 1.6 per 1,000 babies screened for bilateral impairment 40 dB HL (17 children, or 0.52 per 1,000 live births). By the time these children and their peers started primary school, they made up only 40% of all cases of bilateral PCHI 40 dB HL; 18% had risk factors at birth but had not been screened and 42% had no obvious risk factors. This is a compelling argument for universal over-targeted screening.

Some risk factors have come from understanding the aetiology of PCHI, conversely the aetiology has sometimes been worked out after observational studies showed something as a risk factor. This is a continuing reason for studying risk factors – in order to understand more about what might be causing hearing loss. Other reasons are that the high risk may extend beyond the neonatal period, indicating the need for further observation of a child as he or she develops, and to help parents who encounter one of these risk factors understand the increased risk their children may face.

A frequently quoted list of risk factors is published by the Joint Committee on Infant Hearing.[12] Some are highlighted as particularly relevant when thinking about progressive or delayed-onset cases, and they recommend that any child who has these risk factors is seen by an audiologist before 30 months old if the newborn screen is clear. The Newborn Hearing Programme (NHSP) in the UK publish their own guidelines on the management / surveillance of high-risk individuals.[81] It is recommended that any neonates with meningitis are referred straight to audiology without a screen, and children who recover from meningitis be offered an audiology appointment within 4 weeks of discharge from hospital. Babies born with cranio-facial abnormalities (including cleft palate) or Down syndrome should be screened again at eight months. Other babies who should be offered an assessment at eight months and at intervals throughout their childhood are those with: a family history of PCHI; assisted ventilation in NICU for >5 days; neonatal jaundice to a level needing exchange transfusion; congenital infection with toxoplasmosis, rubella, cytomegalovirus (CMV) or herpes; and developmental delay associated with a neurological disorder. They recommend audiological testing for babies who have had high levels of ototoxic drugs and caution strongly against their use if there is a family history of hearing loss after antibiotics.

Weichbold et al.[82] examined the histories for 23 9-year-old children who had developed bilateral PCHI after a clear newborn hearing screen. Eleven children had risk factors (as defined by JCIH 2000): three had a family history of hearing loss; two had recovered from meningitis; two had a cranio-facial malformation; one had persistent pulmonary hypertension; one had a congenital CMV infection; one received extracorporeal membrane oxygenation; and one had recurrent otitis media with effusion. They also found that five children had received ototoxic therapy (not on the list of risk factors at the time) and two had been born before the 33rd gestational week (one child had a combination of the last two). Six children (26%) showed no risk indicators for post-natal hearing loss.

AETIOLOGY

The major aetiological classification system suggested by Davidson et al.[59] has been used in most recent studies. The categories are:

- genetic;
- prenatally acquired;
- perinatally acquired;
- post-natally acquired;
- cranio-facial anomalies; and
- other.

Unfortunately, it is common for a large percentage of children in epidemiological studies to have an unknown aetiology, referred to as ‘missing’. In a selection of recent studies stating the aetiology of PCHI for different populations, there are reports of 16 to 55% of unknown origin (see Table 1.5).[19,21,34,83–85]

Several studies have looked at ways of finding the underlying aetiology in missing cases, both as a way of improving epidemiological data and for clinical reasons. In the Trent study,[19] 41% of children did not have an identifiable aetiology. Nevertheless, it was possible to impute aetiology from other data such as medical notes, and this reduced the percentage of people who

Table 1.5 Selection of cross-sectional cohort studies with percentage of cases from each aetiological category, showing differences across time and study.

First author (year published)	**Drews (1994)[34]**	**Parving (1993)[83]**	**Fortnum (1997)[19]**	**MacAndie (2003)[21]**	**Fortnum (2002)[85]**	**Billings (1999)[84]**
Cohort (N)	**Atlanta, born 1977–1979 (100)**	**Copenhagen, born 1980–1990 (228)**	**Trent, born 1985–1993 (653)**	**Glasgow, born 1985–1994 (130)**	**UK, born 1980–1995 (17160)**	**Boston, diagnosis 1993–1996 (301)**
Category						
Genetic	13	36	45	43	30	23*
Prenatal	7	16	4	3	4	2
Perinatal	–	14	17	15	8	18
Post-natal	24†	5	6	7	7	9
CFA	1	–	3	12	–	7
Other	–	–	2	–	2	9**
Missing	55	27	25	16	49	32

*Including 'known syndrome' (12) 'family history' (11) **'congenital abnormality, other'
†Including 13 cases of Hib meningitis.

had no aetiological information to approximately 25%. Taking this one step further, Parker et al.[86] report investigating 82 children from the Trent study using a questionnaire, home visit and genetic test for the most common genetic mutation causing hearing impairment (Connexin 26 35delG, see Genetic hearing impairment–non-syndromic below). They found eight children had a genetic syndrome not previously assigned and seven further cases had the Connexin 26 35delG mutation. Parving[87] found that aetiology was significantly more likely to be found if a child with PCHI had a non-audiological examination in addition to a standard audiological exam (37/61 vs. 61/117). Peckham et al.[88] suggested that congenitally acquired CMV might be responsible for a large proportion of children for whom no other obvious cause is found for PCHI. Their study found that such children were twice as likely to have CMV excreted in their urine than children with normal hearing (13% vs. 7%). There are a number of guidelines now available for clinicians investigating the cause of hearing loss in individual children[89] with core investigations that all children with PCHI should receive; additional tests are suggested depending on the circumstances (Table 1.6).

The BAAP give several reasons for investigating the cause of hearing loss:

- To try to answer parents who ask 'why is my child deaf?'
- To help identify, monitor, treat or prevent associated medical complications in some patients.
- To help prevent further deterioration of hearing loss in some patients.
- To enable better-informed genetic counselling.
- To inform epidemiological research.
- If the diagnosis is known, then the doctor can provide better advice to parents, such as assisting the family in making decisions about the most appropriate communication mode, about educational placement and about cochlear implantation.

Table 1.6 The recommended history and examination for a child with PCHI – this may help discover aetiology.

Core	Additional
Personal history: pre- and perinatal problems, general development, general health and head injury.	Genetic tests: Chromosomal examination (karyotyping) if developmental delay or dysmorphic features; Connexin 26 and 30 gene testing for common mutations if PCHI severe or greater; testing for other mutation, including mitochondrial, as suggested by the history.
Family history: looking back three generations, including congenital and acquired hearing loss.	Renal ultrasound: If syndrome with multi-system abnormalities suspected, or if family history of renal problems.
Imaging of head and neck: Computerised Tomography (CT) or Magnetic Resonance Imaging (MRI) scans.	ECG, as some syndromes are associated with dangerous cardiac conduction abnormalities.
Infection screen: for CMV and rubella	
Ophthalmology: may show changes due to a syndrome such as Usher or congenital rubella. Also important for ascertaining extra needs for children with hearing impairment.	Infection screen: toxoplasmosis and syphilis tests if indicated.
Thyroid function: usually done at birth.	Blood tests and urine examination: if syndromes involving kidneys are suspected, such as Alport or Alstrom syndromes.

The presence of one possible aetiology does not exclude other causes. For example, it is increasingly recognised that some mutations do not in themselves cause hearing loss, but lower than the threshold for environmental insults pre, peri and post-natally.[90,91] Such mutations include the A1555G mitochondrial gene mutation, which predisposes to hearing loss when a child takes aminoglycoside antibiotics, such as gentamicin. There is also a controversy over whether perinatal problems, often cited as the cause of congenital defects, are actually the effect of pre-existing developmental anomalies.[92] Children with a sensorineural hearing loss can be more at risk of conductive problems such as chronic otitis media,[93] something that can potentially be treated to improve hearing.

As might be expected for a condition with such a variety of causes, the frequency of occurrence of some causes of PCHI varies over time and geographical areas. Parving and Hauch[94] looked at the ascribed causes of hearing loss in children attending the School for the Deaf in Copenhagen in 1993–1994 in comparison to causes evaluated 10 and 40 years previously. They found that the frequency of congenital inherited hearing impairment increased steadily with time, whilst between 1953 and 1983 there had been a significant increase in prenatal infections, which then declined between 1983 and 1993. Admiraal and Huygen[95] in the Netherlands found a similar decrease in prenatal infectious causes from 1988 to 1998, whilst the proportion of PCHI thought to have a perinatal cause had increased.

The changes in the developed world over the last few decades have shown the success of primary prevention. Measles, mumps, rubella and meningitis are all implicated in PCHI, and all have been the subject of immunisation programmes. Secondary prevention has also helped, with better nutrition and treatment leading to better outcomes from infections such as measles and meningitis. Meanwhile there has been a rise not just in the proportion of genetic cases but in the actual numbers. In some cases this is due to better neonatal care leading to the survival of babies with life-threatening syndromes, in others it is due to the increase in prevalence of particular mutations. Nance and Kearsey[96] suggest that the frequency of PCHI caused by Connexin 26 or 30 mutations may have doubled in the last 200 years due to the establishment of a 'Deaf community'* leading to healthier hearing-impaired adults. These adults go on to have children and this decreases genetic selection for the unmutated forms of the Connexin gene.

In contrast, Dunmade et al.[97] looked at the aetiologies of sensorineural hearing loss in children in Nigeria, comparing aetiologies of hearing loss in 1980 and 2000, and found there had been no significant decrease in infectious causes. The figures for the 115 children studied in 2000 showing some common causes were febrile illness (18.3%), measles (13.9%), meningitis (8.7%) and mumps (6.9%). Saunders et al.[98] offered the Connexin 26 35delG genetic test to children with PCHI in an audiology clinic in Jinotega in Nicaragua and found that despite a family history of hearing loss in 33%, this mutation, so common in the UK, was not present in any of their children. Another difference he found was in the unmonitored use of ototoxic antibiotics, which are cheaper than their alternatives.[99]

GENETIC HEARING IMPAIRMENT

At least half of all cases of PCHI are known to have a genetic cause.[100,101] However, despite significant advances in the understanding of the molecular basis of hearing loss, identifying

*The use of the capital D indicates the community of deaf people who use BSL as their language and identify with other deaf people who share their language, culture and history.

the precise genetic cause in an individual remains difficult. Using systematic investigation, such as that described in Table 1.6, will increase the chances of finding the aetiology, but it is estimated that a mutation in one of between 300 and 500 genes (around 1% of the total number of genes) can cause hearing loss.[102] Approximately 120 of these genes have been identified so far – around 80 causing syndromes that include hearing loss and over 40 responsible for 'non-syndromic' hearing loss. Most of these genes are located on the autosomal chromosomes, up to 20% on the X-chromosome and up to 20% in the maternally inherited mitochondrial DNA. This confirms the findings from the questionnaire section of the Parker et al.[86] study based on the Trent cohort: the families of 526 hearing-impaired children (aged 4–13) were sent questionnaires asking about any family history of hearing loss, the results pointing towards different genetic disorders with autosomal dominant, autosomal recessive and sporadic inheritance.

Syndromic PCHI

If hearing loss is one of several clinical findings, the disorder is described as a syndrome. Approximately 30% of genetic hearing impairment is syndromal.[100,101] Over 400 syndromes featuring PCHI have been described and many of the genetic abnormalities responsible identified. Syndromal hearing impairment can be sensorineural or conductive, due to structural anomalies of the auditory system. McClay et al.[103] report that the presence of any congenital syndrome significantly increased risk of an abnormality of the temporal bone involving the cochlear or vestibular system visible on a CT scan. This risk was found to be elevated regardless of the presence of PCHI, but higher still if PCHI was present. The presence of a genetic syndrome in children with PCHI should not be overlooked as it can be important in determining prognosis and intervention measures – as well as for estimating the recurrence risks in the family.[104]

Chromosomal syndromes may occur either during meosis or mitosis, resulting in too much or too little genetic material, and many increase the risk of PCHI. Two of the most common syndromes caused by chromosomal abnormalities are Down and Turner syndromes. Maatta et al.[105] studied 129 individuals (mainly children) with Down syndrome, and found that one-third of the sample had hearing impairment or recurrent ear infections. Overall, the risk of sensory impairments increased with increasing levels of intellectual disability.

Genetic syndromes caused by mutations, deletions or additions on the autosomal chromosomes can be inherited in a recessive or dominant manner. The majority of syndromal genetic hearing impairments are inherited in an autosomal recessive way and are detectable at birth. Recessive inheritance occurs when both parents – who may not necessarily exhibit the trait – carry a mutated gene that may cause a genetic syndrome. If both parents carry one normal copy of the gene and one mutated copy of the gene, there is a 25% chance of the child inheriting both of the mutated genes (one from each parent) and manifesting the genetic disorder. There is also a 50% chance that the child will inherit one of the mutated genes and become a carrier for that disorder but not manifest the syndrome. Such disorders include Usher syndrome, Cockayne syndrome, Pendred syndrome, Jervell and Lange-Nielsen syndrome, Hurler syndrome and Alstrom syndrome.[106] Usher syndrome is one of the most studied of these syndromes. It was formally classified into three clinical types and was expected to be caused by three corresponding mutations. However, recent work reported by Cohen et al.[107] suggests that there are more than three genetic causes of Usher syndrome, each having different potential effects in different individuals with very little evidence for phenotypic–genotypic correlations.

For dominant inheritance, only one mutated copy of the gene is required for a syndrome to be manifest. Usually, one parent will have the syndrome, and there is at least a 50% chance of the child inheriting the gene and manifesting the genetic disorder. If both parents exhibit the trait, there is a 75% chance of the child manifesting the disorder. Hearing impairment inherited in this way usually manifests itself after the neonatal period, either because it is congenital and progressive or because it is late-onset. Examples of autosomal dominant syndromes include Marshall-Stickler syndrome, Waardenburg syndrome and Treacher Collins syndrome.[106]

Syndromes carried on the X-chromosome affect males predominantly because they have only one X-chromosome. Females 'carry' the mutated syndrome-causing gene but are unaffected if they have a normal copy on their other X-chromosome. Any male children of a carrier will inherit the genetic material for their X-chromosome from their mother (their father contributing the Y-chromosome instead), with a 50% chance that this will include the mutated gene. If this occurs, since there is no copy of the gene on the Y-chromosome, the syndrome will be manifest. Examples of X-linked syndromes include Hunter syndrome, Alport syndrome and Norrie syndrome, all of which do not manifest at birth but develop in early infancy.[106] It is a mutation in an X-linked gene that is responsible for 'deafness with fixation of the stapes', which gives a progressive hearing loss of sensory and conductive types. Although this mutation is very rare, diagnosis is important because if this is not recognised, there can be further damage to hearing if surgical methods to release the stapes are not attempted.[108]

Mitochondria are small organelles located within the cytoplasm of the cell and have their own DNA (mDNA), which is independent of the nuclear DNA. Mitochondria are inherited from the mother only. Thus, a mother who has hearing loss from a mutation on her mDNA will pass this mutation onto her children of whatever sex, but a father with the same mutation will not pass it on. There are multiple copies of mDNA in each mitochondrion, and, therefore, expression of a syndrome-causing gene is not inevitable. Thus the clinical phenotype is extremely variable. An important syndrome to recognise is the MELAS syndrome, where permanent hearing loss may be the first manifestation; and recognition allows better management of subsequent complications.[109]

Non-syndromic

Autosomal recessive non-syndromic hearing impairment is the most common form of genetic deafness, accounting for around 80% of all cases.[110] Thus, it can be estimated to account for around 40% of all profound PCHI. Numerous non-syndromal recessive hearing impairment genes have been localised, with Petersen and Willems[110] reporting 85 loci on 39 different genes. Autosomal dominant inheritance is thought to account for approximately 15% of the cases. X-linked inheritance accounts for approximately 2–3% of the inherited hearing impairments (but 5% of those affecting males).

Mutations in the GJB2 gene are responsible for as much as 50% of autosomal recessive non-syndromic PCHI. This gene codes for a protein called Connexin 26, a gap junction protein regulating the passage of ions in and out of the cell, and was identified in 1997. As with the mutations responsible for Usher syndrome, it has become obvious that genotype–phenotype relationships are more complex than once thought.[111,112] Green et al.[113] studied the prevalence of mutations in the GJB2 gene in 52 people with congenital sensorineural hearing loss at a clinic in Iowa. Twenty-two were found to have GJB2 mutations, 19 of whom had a mutation on both chromosomes. Of the 41 abnormal copies of GJB2, 29 had the same mutation – 35delG.

The siblings of these 52 people were also screened, and it was found that all those who had two abnormal copies of the gene also had PCHI. A total of 560 unrelated children were also screened and there were 14 in whom one copy of the GJB2 gene had a mutation. This gives a carrier rate of 3.0% (probable range 2.5–3.6%). It is important to remember that this carrier rate will be specific for this particular population – mid-western United States. Pandya et al.[114] searched the DNA of children from the Annual Survey of Deaf and Hard of Hearing Children and Youth, conducted at the Research Institute of Gallaudet University, and found that GJB2 mutations accounted for 22.2% of deafness in the overall sample but differed significantly amongst Asians, African Americans and Hispanics. Ethnic differences are particularly marked where there is a small founder population, such as in some Jewish communities.[115]

PRENATAL FACTORS

Infections

Infections are considered to be the main cause of prenatally acquired hearing impairment. In the 1970s–1980s, congenital rubella was the single most common reported cause of sensorineural hearing impairment in childhood, accounting for 16–22% of cases of hearing impairment in babies.[94] If infected during the first month, there is a 50% chance of the developing fetus being affected such that congenital rubella defects are detectable. This risk declines throughout pregnancy to an approximate 6% chance in the fifth month and beyond. Problems associated with congenital rubella (CRS) include learning disability, heart disease, cataracts, microcephaly, hepatomegaly, splenomegaly, bone lesions, purpura, glaucoma and hearing impairment. Hearing impairment is the most common permanent manifestation and affects 68 to 93% of children with congenital rubella.[116] The hearing impairment is usually severe to profound sensorineural hearing impairment and can be progressive.[117]

Congenital rubella was a devastating syndrome that became a major public health issue. A rubella vaccine was first licensed in 1969. By 1999, 105 (49%) of the 214 countries and territories reporting to WHO had introduced the rubella vaccine in their national immunisation programme.[118] In the UK, the rubella vaccine was offered to schoolgirls in the United Kingdom from 1970, and post-partum susceptible women shortly after. Mass vaccination with MMR (measles–mumps–rubella vaccine) of babies was introduced in 1988. Schoolgirl vaccination was discontinued in 1996, although post-partum vaccination of susceptible women identified through antenatal testing continues. Reported cases of CRS declined from about 50 a year in 1971–1975 to just over 20 a year in 1986–1990.[119] About 40 infants with CRS were reported over all of the next 12 years. Women living in the UK who were born abroad have much higher rubella susceptibility rates than UK-born women, and two-thirds of the CRS cases since 1991 have been to mothers born outside the UK. The previously high coverage of children interrupted the epidemic transmission (which was mainly in children), but concerns over the safety of MMR have led to a decrease in immunity amongst children. If an epidemic of rubella occurred in the UK, women born in places without vaccination will be at increased risk of acquiring infection in pregnancy. The likelihood of importation of infection is high, as the developing world still has endemic rubella. Rittler et al.[120] found 43 cases of CRS recorded from the records of 3,883,165 live births collected by the Latin-American Collaborative Study of Congenital Malformations, World Health Organization (WHO) Collaborating Centre for

the Prevention of Birth Defects (ECLAMC), which suggests a prevalence of CRS in Latin America of around 1 : 100,000 live births.

Another prenatal infection that causes congenital abnormalities is toxoplasmosis. Sever et al.[121] studied 23,000 mothers and children from around 20 weeks gestation until 7 years old. Of these mothers, 38.7% had antibodies to toxoplasmosis during pregnancy, and children born to these mothers had double the risk of developing PCHI by age 7 (0.4% vs. 0.2%, $p = 0.01$).

Cytomegalovirus CMV is a common chronic asymptomatic infection in adults, which can cross over the placenta to affect the developing fetus and child. Roizen[117] has observed that CMV infection occurs in 2.2% of all newborns, making it the most common intrauterine infection. Lipitz et al.[122] report that from their sample of 18 babies with confirmed CMV, four (22%) had neurological problems at birth. Fowler and Boppana[123] summarised seven studies between 1982 and 2004, and found that the risk of PCHI was 22–65% in those babies symptomatic at birth and 6–23% in those asymptomatic at birth. Amongst those affected by PCHI, there were progressive, fluctuating and delayed-onset cases. They were unable to identify any way of predicting which babies were more at risk of PCHI from asymptomatic CMV infection, and they flag up the fact that UNHS may miss many babies with PCHI due to CMV because of its variable course. It is not yet established how much CMV infection contributes to the overall prevalence of PCHI, as studies vary in their method of investigating infection. In the meta-analysis by Morzaria et al.[124], the mean of cases reportedly due to CMV in the studies from 1990 to 2002 was 0.92% (s.d. 1.07) of the total, but Peckham et al.[88] reported that 14% of those diagnosed with PCHI of unknown aetiology excreted CMV in their urine (compared with a base rate of 7%), and Barbi et al.[125] reported that of 130 children with PCHI, 24.7% had CMV in blood retained from a sample at birth (base rate not given). Given the availability of an antiviral treatment for CMV,[126] there is an argument for screening newborn babies for CMV.[125,127]

Maternal drug therapy

Maternal drug therapy during pregnancy can also contribute to congenital hearing impairment. Some substances may permanently injure or destroy the hair cells of the cochlea resulting in a sensorineural PCHI. For example, alcohol, streptomycin, quinine and chloroquine phosphate may destroy neural elements of the inner ear.[128] The loss is usually triggered by the ingestion of ototoxic drugs during the first trimester, with damage to the auditory system occurring especially in the sixth or seventh week after conception. Conductive PCHI can also result from ototoxicity, primarily as a result of ossicular malformations of the middle ear. Brent[91] emphasises the gene–environment interaction involved in teratogenic drugs.

Perinatal factors

Perinatal factors which may predispose to PCHI include prematurity, hyperbilirubinaemia (kernicterus), anoxia (including apnoea and cyanosis), severe neonatal sepsis, rhesus incompatibility, low birth weight and trauma.[129] Some perinatal problems that were known to cause neurological damage have been much diminished in the modern maternity hospital, for example, the introduction of photosynthetic lights to reduce jaundice (hyperbilirubinaemia) to non-toxic levels and rhesus inoculation to prevent rhesus incompatibility in future pregnancies. On the

other hand, medical advances have ensured that more premature, anoxic and low-birth-weight (LBW) babies survive, leading to more babies graduating from NICU with a hearing impairment. Davis and Wood[31]showed that a baby admitted to NICU for any reason had a risk of developing PCHI by 3 years old that was seven times higher than those who had not. Razi and Das[129] showed that even in children who had a hearing threshold within the normal range, the mean high-frequency threshold was higher in children who had experienced an adverse perinatal event.

Prematurity is a risk factor for PCHI, but it is not clear whether this itself is the causal factor or whether the causes are factors associated with prematurity such as anoxia, hyperbilirubinaemia, increased bacterial and viral infections, treatment with ototoxic drugs and/or LBW. Veen et al.[130] concluded that in their study of 890 5-year-olds who had very LBW or had been very premature, the prevalence of sensorineural hearing impairment was 15 times higher than the average Dutch population of 5- to 7-year-olds. Van Naarden and Decoufle[131] studied 320,000 children born in Georgia US in 1991–1993 and found that by age 3, 169 children had developed PCHI, of which 17 had been amongst the 3,362 children who had been born weighing less that 1,500 g. This gives a relative risk of 13.9 (95% CI 8.2–23.4). In this group of very low weight babies, it is hard to identify the precise cause of hearing impairment due to the sheer numbers of possible complications those infants may experience.

Children with adverse perinatal events are also at risk of having other developmental disabilities, making them particularly high need. Van Naarden and Decoufle[131] estimated that for a child who was born weighing less than 1,500 g, the risk of developing a hearing impairment *plus* another disability was 27.8 times (95% CI 11.6–66.5) that of a child born weighing over 3,000 g. Davis and Wood[31] found NICU babies with a hearing impairment were considerably more likely to have another disability (odds ratio 8.7 to 1). Yoon et al.[132] suggest that UNHS may not pick up all of the NICU graduates who develop PCHI due to the high incidence of middle ear problems and delayed-onset sensorineural hearing loss.

Post-natal factors

It is possible for post-natal causes of acquired PCHI to be genetic due to delayed onset of hearing impairment, but most acquired cases are caused post-natally by infection, ototoxic agents or trauma.[59] Otitis media should be included even though permanent hearing impairment secondary to otitis media is uncommon in the developed world, because it may delay the detection of permanent hearing impairment. Systemic and neurological infections that have been linked with PCHI are bacterial meningitis, measles, mumps, HIV[133]and CJD.[134] Thanks to a successful vaccination programme and better general health, new-onset measles and mumps-related hearing loss is now rare in the developed world.

Bacterial meningitis is a serious infectious disease both in the neonatal period and throughout childhood. It can be caused by a variety of pathogens, including *Haemophilus influenzae* type b (Hib), *Streptococcus pneumoniae* (pneumococcus) and tuberculosis (TB). For children who survive meningitis, there are often sequelae, which include learning disabilities, hydrocephalus, motor abnormalities, vestibular deficits, psychosis, hyperactivity and visual and sensorineural hearing impairments. Reports have indicated that acquired hearing impairment represents 9.5% of total PCHI, with 6.5% of these cases being caused by meningitis.[135] Meningitis-induced hearing impairment is often bilateral, severe or profound and rapid in onset. Clinical and experimental studies have shown that the loss results from direct damage to the

cochlea by the infection, but it may be exacerbated by additional cochlear damage resulting from any ototoxic drugs used to treat the disease.[136] Children who have lost their hearing to meningitis are often considered to be amongst the best candidates for cochlear implants due to their previous experience with language and their total loss of any auditory neural function. The incidence of post-meningitic hearing impairment varies from 7 to 31%, depending on the type of meningitis and type of hearing impairment included.[137–141] Wellman et al.[140] and Kutz et al.[141] also compared the complication rate between Hib and pneumococcus, finding the latter significantly more likely to lead to hearing impairment. In 2004, a Hib vaccine was added to the routine childhood vaccination schedule in England, and from 2006, a vaccine against invasive pneumococcal disease was also added. It is hoped that this will reduce the incidence of acquired PCHI.

Children may be given a number of ototoxic treatments, for example, aminoglycosides (such as gentamicin) for severe infections or those resistant to penicillin; platinum-containing chemotherapy such as carboplatin for retinoblastoma a childhood cancer of the eye, and radiotherapy for tumours in the glands of the neck. Many of these treatments are the best available[142] but often the adverse effects can be minimised by action such as co-administering aspirin with gentamicin,[143] careful dosing of carboplatin[144] and well-placed radio-opaque shields.[145]

CONCLUSION

Hearing impairment is the most frequent sensory impairment in humans, with significant social and psychological implications. In the light of the impact that PCHI can have on children and their families, the importance of epidemiological studies cannot be underestimated. In developed countries, around 1 in 1,000 babies is born with at a serious permanent bilateral hearing loss, and permanent hearing loss becomes more common as children grow older.

In developing countries, the prevalence may be higher and in some countries it may be considerably higher, but there is a lack of large-scale, robust epidemiological studies.

Epidemiologal data were at the forefront of public health and audiological arguments for universal hearing screening, and have also been used to plan and monitor primary prevention such as vaccination. This chapter explained the difficulties in collecting data on incidence, prevalence and aetiology. Recent results of studies from throughout the world on the prevalence and aetiology of deafness have been presented that show the changing nature of deafness throughout the world. Clearly, a greater emphasis on collecting routine data on the pattern, degree, aetiology and natural history of children with deafness is needed throughout the world. It is only by recording these data that we will understand the extent and nature of childhood deafness and propose realistic public health plans to provide support for these children and their families.

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2 Screening and surveillance

J. Stevens and G. Parker

INTRODUCTION

In the absence of an effective hearing screen, the detection of permanent childhood hearing impairment (PCHI; average hearing levels (0.5, 1, 2 and 4 kHz) measured at ≥40 dB nHL in the better ear) can be delayed by as much as 2 to 3 years. Even with screening programmes in place, considerable delays can occur. This is illustrated in Figure 2.1 where the cumulative age of confirmation of PCHI from a major UK study is shown.[1] At the time of this study, the UK had a universal screen at the age of 8 months using the infant distraction test (IDT) and partial targeted newborn hearing screening (TNHS).

The lack of an effective screen has limited our knowledge of the age of onset of hearing impairment in early life, as most studies have had to rely mainly on retrospective analysis of case studies.[1,2] These studies have indicated that the majority of PCHI is probably present at birth.

There is also the question of why is it important to detect PCHI at an early age. There is evidence that early diagnosis with effective management can improve the outcome for language development for those children with PCHI.[3–6] Effective early hearing screening would therefore appear to be an essential part of any child health programme. Until the introduction of universal newborn hearing screening (UNHS) the only universal method used in the first year of life was the IDT. Its implementation was variable between countries and, as will be discussed later, its effectiveness in practice has been brought into question. Most countries that carry out an early screen for PCHI are now adopting UNHS as the preferred universal screen before school entry. However, as previously noted, not all PCHI is present at the time of a newborn screen, and systems to detect acquired hearing impairment need to be part of an overall screening and surveillance programme.

It is important to determine which types of hearing losses a screening programme should aim to detect. Whereas there is agreement on the detection of PCHI of moderate or greater degree, there is less agreement on the need to detect unilateral hearing impairment, temporary hearing impairment or mild hearing impairment.

This chapter will start by considering the principles of screening (see Chapters 1, 13, 15, 26). It will then consider the implications of the prevalence of different types of hearing loss and the rationale for different types of newborn screening. UNHS will be covered in detail as this is the most important age to detect PCHI. Later screens carried out at 2 years and school entry will then be considered.

PRINCIPLES OF SCREENING

Ten principles of screening were laid out by Wilson and Jungner.[7] These have been considered by Haggard and Hughes[8] in a review of screening children's hearing and by Davis et al. in the

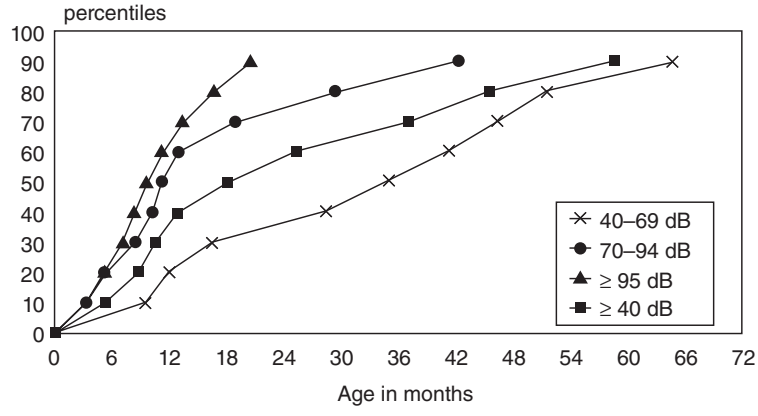

Figure 2.1 Age of confirmation of permanent hearing impairment (N = 309). *Source*: Fortnum and Davis 1997.

critical review of the role of newborn hearing screening in the detection of congenital hearing impairment.[9]

Haggard suggested four further principles which were also included in the review by Davis et al.[9] In summary, these principles, adapted for hearing impairment in a similar manner to that by Davis et al.,[9] can be stated as follows:

Principles

1. The condition (hearing impairment) should be an important health problem.
2. There should be an accepted treatment, i.e. an acceptable means of habilitation for those identified by the screen.
3. Facilities for assessment, diagnosis and treatment should be available.
4. The hearing impairment should be recognisable at an early stage.
5. There should be a suitable test for use as the screen.
6. The test should be acceptable to the parents and to the child.
7. The natural history of the condition should be known and understood.
8. There should be an agreed policy on whom to treat.
9. The cost of case finding (including all consequential costs of the screening programme) should not be disproportionate to overall healthcare costs for the hearing-impaired child.
10. Case finding should be seen as a continuous process.
11. The incidental harm should be small compared with the overall benefits.
12. There should be guidelines on how to explain results to parents with appropriate support.
13. All hearing screening arrangements should be reviewed in the light of changes in demography, epidemiology and other factors.
14. Costs and effectiveness of hearing screening should be examined on a case type basis to maximise the effectiveness and benefit for each type before considering overall costs, effectiveness and benefit.

Current knowledge of principles 1, 2 and 7 is presented elsewhere in this book. In summary, hearing impairment is an important health problem as it can affect quality of life in several ways. Language development and the ability to communicate are affected with subsequent effects on educational achievement, social development and employment prospects. Condition

2 is satisfied for bilateral PCHI of moderate or greater level as there is an effective treatment in terms of amplification by using a hearing aid or, in some cases of severe and profound hearing impairment, cochlear implantation. For unilateral PCHI, mild PCHI and temporary hearing impairment, there is less evidence of the effectiveness of treatment, and there are a range of views on whether or not a screen should aim to detect these cases. Evidence so far published indicates that the majority of significant PCHI is present at birth. Under condition 7, knowledge of the age of onset of hearing impairment is one of the most important factors when selecting which screen to implement. As more UNHS is implemented, knowledge, not only about the age of onset, but also about the rate of progression of hearing impairment, will improve.

Principles 3, 8,10, 12 and 13 relate to the quality of implementation, which is outside the scope of this chapter. Clearly, good implementation of these principles is needed if any screen is to be successful. The chapter will therefore focus on principles 4, 5, 6, which relate to the test method, and principles 9, 11 and 14, which relate to cost, effectiveness and benefit.

PREVALENCE OF HEARING IMPAIRMENT

The prevalence of significant PCHI is discussed at length in Chapter 1. A prevalence of 1–2 per 1,000 is generally reported in European studies, depending on population selection, definitions and methodology.[10,11] Using results drawn from the Trent Ascertainment Study,[1] the overall prevalence for permanent bilateral childhood hearing impairment ≥40 dB HL was estimated to be 1.33 per 1,000 live births (1 in 750 children), with equivalent figures of 1.10 per 1,000 (1 in 900) for hearing impairment ≥50 dB HL, 0.59 per 1,000 (1 in 1,700) for hearing impairment ≥70 dB HL and 0.31 per 1,000 (1 in 3,200) for hearing impairment ≥95 dB HL.

Davis et al.,[9] reviewing the literature, reported yields of 1–1.5/1,000 from UNHS.[12–14] This has been supported by similar yields reported from subsequent large-scale programmes, including the national screening programme (NHSP) in England.[15] This is consistent with a high proportion of PCHI being present at the time of UNHS.

Figure 2.2 compares the prevalence of PCHI with that of other conditions in the UK for which screening programmes are currently available.

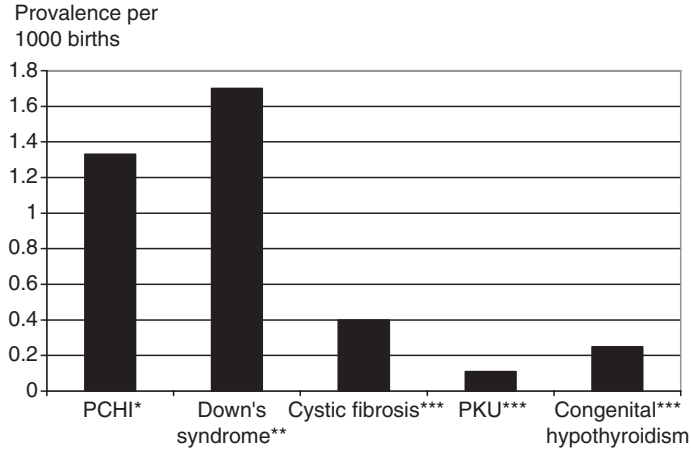

Figure 2.2 Graph to show relative prevalences of PCHI; Down syndrome; cystic fibrosis; PKU; primary persistent congenital hypothyroidism. * Davis et al. 1997; ** Howe et al. 2000; *** Pollitt et al. 1997.

RATIONALE FOR TARGETED SCREENING

TNHS has largely been superseded by the introduction of UNHS, but in some situations where funding or access to the screen is limited, it offers the opportunity of testing a relatively small high-risk group, which is likely to provide a high yield of cases.

The awareness of an increased risk of hearing impairment in graduates from neonatal intensive care units (NICUs) first led some centres to introduce screening of these babies.[16,17] Davis and Wood[18] provided epidemiological evidence which has further led to the concept of key factors for permanent hearing impairment, which might be suitable for defining an 'at risk' population. One of their most significant findings was that babies who were admitted to an NICU for more than 48 hours were 10.2 (95% CI 4.4–23.7) times more likely to have a permanent hearing impairment (≥50 dB HL) than those who did not undergo intensive care, assuming no other predisposing factors.

Three key risk factors were described:

- a history of admission to NICU ≥48 hours;
- a family history of early childhood deafness; and
- a syndrome associated with hearing loss, e.g. craniofacial anomaly such as cleft palate.

It was estimated that 50% of all children with bilateral hearing impairment ≥50 dB HL had one or more of these three factors.

This concept has been supported by subsequent epidemiological studies[1,19] and from UNHS programmes. The Wessex UNHS study,[14] for example, reported that of over 25,000 neonates screened, 8.1% (5.1% from the post-natal wards and 3.0% from the special care units) fulfilled high-risk criteria for PCHI.

The feasibility of targeting the 'at risk' group for hearing screening, however, also depends on the ease of identification of each factor. Whilst admission to NICU and the presence of a craniofacial anomaly can be readily highlighted for inclusion in a screening programme, accurate identification of babies with a relevant family history is likely to prove more difficult.[20]

The cost of a targeted newborn screening programme would depend, in part, on the proportion of the population fulfilling the screen selection criteria. Davis and Wood[18] found that 5.9% of all births in the Nottingham Health District were admitted to NICU >48 hours, although the percentage may be as high as 12% in other districts. Fortnum and Davis[1] estimated that, in practice, the yield from a targeted screen using the three key factors noted above would be 50% or less.

SCREENING IN THE FIRST YEAR OF LIFE

Potential methods and opportunities

In order to produce a satisfactory method of screening, the test must be acceptable, non-invasive, reliable, simple and quick to perform and have a high sensitivity and specificity (relating to principles 5, 6, 11 and 14). There are many physiological responses to sound which might be used for a hearing screen in the first year of life. They originate at different levels of the auditory pathway, from responses of the cochlea up to responses involving the central nervous system. It is important to note that all of these tests check the function of only part

of the auditory pathway. None are actually a complete test of hearing. However, as most pathology of the auditory pathway occurs at the cochlear level, it is possible to use most of these physiological responses as potential methods for screening.

Any screening method must determine that there is sufficient hearing present for the child to learn normal language. This means that responses should be obtained to quiet levels of sound (around 40 dB nHL or below) or it must be shown that the test can demonstrate that hearing is present at this level. Many of the physiological responses only occur at high sound levels and so are not useful as a screen (such as heart rate and respiration rate changes). Those that have proved able to meet the above criteria at some point in the first year of life can be grouped under:

(a) oto-acoustic emissions (OAEs) from the cochlea;
(b) electrical responses of the early auditory pathway; and
(c) behavioural responses.

The actual use of these methods in practice depends on such practicalities as the baby being in a suitable state for the screen to be carried out efficiently and effectively. Methods (a) and (b) require that the baby is quiet and still. The best opportunity for this is in the first 2–3 months of life, which has the added benefit of the earlier detection of a PCHI. Of the electrical responses from the early part of the auditory pathway, the click-evoked auditory brainstem response (ABR) has proved to be the most effective for use as a screen to date. With the need to carry out follow-up tests, the screen needs to be done soon after birth and at the latest by about 4 weeks corrected age.

Behavioural responses can be used at birth, but the levels of sound required to elicit them are high, and it is not until around 8 months of age that responses can be readily obtained to the required low levels of sound. At birth, reflex movement of head and body has generally been used with the baby supine, whereas at 8 months, the reflex turning of the head towards a sound source is used (the infant distraction test (IDT)).

In summary, the practical screening options in the first year of life are OAEs or the ABR soon after birth (or a combination of the two) and the IDT at 8 months of age. Behavioural responses soon after birth are a further possible method but a high stimulus level has to be used. Each method will now be considered in detail.

NEWBORN SCREENING

In the 1980s and 1990s, many studies were carried out on the use of the aforementioned methods (e.g. OAEs[12–14, 21–23]; ABR[17,22,24–27]; and auditory response cradle (ARC)[28,29]). Studies are difficult to carry out due to the large cohorts required and the follow-up of children to the point at which their hearing levels are confirmed. If the number of missed or late-onset cases is to be determined, then all children need to be followed up. A review of these studies was compiled by Davis et al.[9] and the reader is referred to this document for more detail.

Otoacoustic emissions

In 1978, Kemp[30] recorded the presence of acoustic energy emitted from the ear in response to sound. Studies have shown[31] that these emissions originate in the cochlea and relate to some biomechanical process associated with normal hearing. The important property for screening is that OAEs can be recorded only when a region of normal cochlear function is present. All

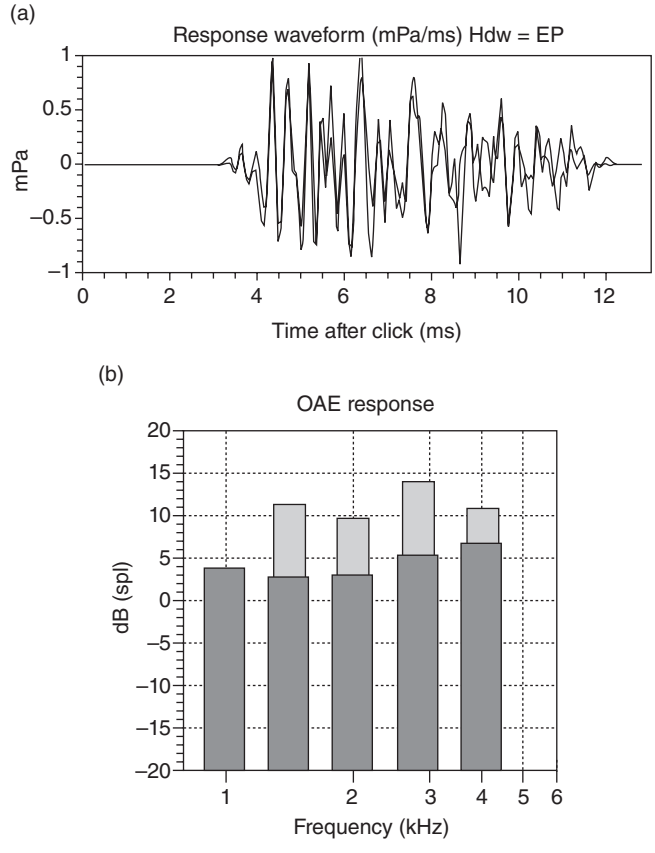

Figure 2.3 Typical neonatal TEOAE response displayed in the time domain (a) and the frequency domain (b). The response shown is the non-linear component between 2.5 and 12.5 ms post stimulus. For (b) the TEOAE sound pressure level is shown in half octave bands; the darker shaded area is the noise component.

types of OAE are recorded by the placement of a probe into the ear canal. The probe contains one or more miniature loudspeakers to generate the stimulus and a microphone to record the sound in the ear canal.

Classification

OAEs are normally classified by the method of recording. The transient evoked oto-acoustic emission (TEOAE) is the response to a short transient of sound. A typical stimulus and response in a baby is shown in Figure 2.3a. The response is also commonly viewed in the frequency domain as shown in Figure 2.3b. The response is small compared with the acoustic stimulus presented to the ear and has a sound level around the threshold of hearing.

Recording method for TEOAE

As it is below the background noise levels, a technique known as averaging is used to detect the response. This is achieved by adding many hundreds of responses together. The response

is also filtered to remove unwanted low- and high-frequency components. To minimise the chance of the response being an artefact (e.g. a mechanical echo of the stimulus), it is normal to only consider the part of the response which is not proportional to the stimulus (the non-linear component). High continuous background noise levels (e.g. incubator fan, air conditioning, computer cooling fan) will prolong the test time or can make it impossible for the test to be carried out. A test room with noise levels below 35 dBA SPL should be used.

Properties of TEOAE and screen pass criteria

The TEOAE is unique to the individual ear, although the recording is dependent on the probe characteristics. There is a large variation in both amplitude and waveform among individuals. As noted, the presence of a TEOAE indicates a region of normal cochlear function. Its absence could have many causes, such as poor recording conditions, too small an amplitude to record, or the presence of outer-ear or middle-ear disease. The frequency spectrum of a typical newborn TEOAE is shown in Figure 2.3b. This frequency range can vary among babies with normally hearing ears; some will only give a narrow range of emission frequencies, whilst others will produce a broad range. These factors lead to the following typical choice for a pass criterion when using TEOAE as a newborn screen.

- The response is present in a limited number of frequency bands, e.g. in two half-octave bands from half-octave bands with centre frequencies at 1.5, 2, 3 and 4 kHz.
- There is a high response to background noise ratio, e.g. the response is 6 dB or more above the background level.
- The amplitude is in the physiological range.
- There is a low chance of artefact from the stimulus.

TEOAE recorded in babies are generally larger and contain higher frequencies compared with those recorded in adults.[32,33] OAEs are not normally present when the hearing loss is greater than about 30 dB HL.[34]

The distortion product otoacoustic emission (DPOAE) is the other type that has found application in screening. The stimulus consists of two tones at different frequencies. Sounds at other frequencies (called distortion products) at very low sound levels can be recorded. There is a wealth of evidence[31] that these sounds reflect the properties of a non-linear process associated with outer hair cell motility. For clinical measurements the distortion product $2f_1$-f_2 is normally recorded at a number of frequency pairs f_1, f_2. The correlation between audiometric threshold and DPOAE amplitude is weak, and it is not possible to use the DPOAE amplitude to predict hearing threshold with any accuracy except to say that it is inside or outside normal limits.[31]

Auditory brainstem response

Choice of ABR

It is possible in adults to record electrical responses from the auditory pathway from the cochlea to the cortex. In babies, at low stimulus levels used for screening, the cortical responses are difficult to record and early potentials have been used. The non-invasive requirement

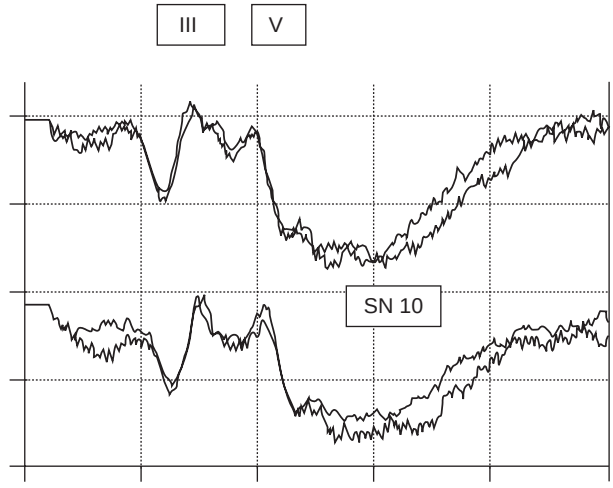

Figure 2.4 Typical ABR waveforms at 50 dBnHL (upper traces) and 40 dBnHL (lower traces) stimulus levels from neonatal screening, showing waves III, V and SN10. The small divisions on the axes are 0.25 μV (vertical) and 4 ms (horizontal).

restricts measurement to potentials which can be recorded on the skin surface. The result is that the ABR has become the method of choice in babies. The ABR records the electrical activity occurring in the first 15–20 ms after the stimulus. The so-called wave V, which is recorded at around 8 ms to click stimuli in neonates, is the most prominent wave at low sound levels. A typical newborn ABR response at 40 dB nHL and 50 dB nHL stimulus levels is shown in Figure 2.4. The wave III and V complex (Figure 2.4) is normally used together with the later slow wave (SN10) to determine whether a response is present.

Choice of stimulus and limitations of ABR for screening

For screening, a click stimulus is normally used as it gives the maximum response amplitude. However, it stimulates the whole of the cochlea. This limits the test, as it is only possible to infer from the result that there is a region of normal hearing function up to the brainstem level. The relationship between the click ABR threshold and pure tone threshold in adults with different degrees of hearing loss is shown in Figure 2.5.[35] The click ABR is also therefore only a limited measure of hearing. However, as with OAE, it is sufficient to detect the majority of clinically significant hearing impairments.

Absence of a response can be due to several factors apart from a raised hearing threshold. Poor recording conditions may affect the detection of the response, and any factor which affects the nature and amplitude of the ABR response (such as delayed maturation) should be taken into account.

The choice of stimulus level for the ABR screen needs to be carefully chosen so as to detect the type of hearing loss targeted by the screen. Johnson et al.[36] note that much of the current automatic ABR (AABR) screening equipment was designed to identify infants with moderate or greater hearing impairment. They found that in a two-stage OAE /AABR protocol screen, approximately 23% of babies with PCHI at 9 months of age would have passed automatic

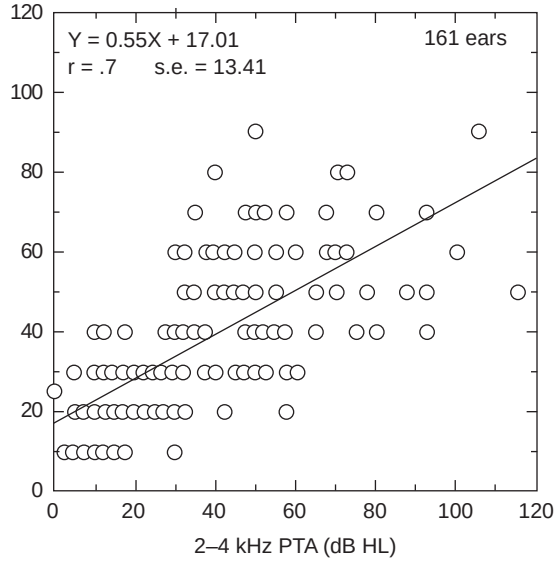

Figure 2.5 Relationship between the click ABR threshold (vertical axis dBnHL) and pure tone threshold in adults (161 ears with sensori-neural hearing impairment). *Source*: Stapells and Oats 1997.

ABR screening. They concluded that this happens in part due to the choice of AABR screening level to detect moderate or greater hearing impairments.

Recording method

The ABR response is less than 1 microvolt in amplitude. Like the OAE, the averaging of many responses is required to detect the response above the background electroencephalogram. A good electrical and acoustic environment is required. Protocols for good practice should be followed to avoid errors being made (e.g. National Newborn Hearing Screening Programme (England) 2007[37]). As with TEOAE, pass criteria should include the conditions of a recognisable physiological response, high repeatability and the absence of artefact.

An electrical response from the post-auricular muscle (PAM) can also be recorded around 15 ms after the stimulus. The response is very dependent on muscle state and is not always present. However, if the response is present, it can be used in place or in addition to the ABR response in screening. On occasions, when a baby is too restless for an ABR to be recorded, it may be possible to obtain a PAM response.

Automated OAE and ABR for screening

Due to the fact that a waveform is recorded for both OAE and ABR, it is possible to include in the equipment a mathematical algorithm, which can be used to give a measure of the confidence of the presence of a response. As well as determining a measure of the confidence in the response, the algorithms can also check for potential artefacts and that the response is within the physiological range. Several examples from early work have been incorporated into clinical instruments. Their efficacy has been demonstrated in a number of trials.[25,38,39] Nearly

all OAE and most ABR screening is now carried out with automated equipment making use of a range of different mathematical algorithms. It is important that all automated equipment is properly trialled to determine that it meets screening programme specifications for false positive and false negative outcomes.

There is variation amongst the automated equipment being currently manufactured as to whether the complete ABR/OAE response is stored or not. One of the advantages of both ABR and OAE over the IDT for screening is that a record of the response is available which can be reviewed at a later stage. If screening equipment does not record the full response this advantage is lost.

Head and body movement

The third method that has been tried in practice involves the measurement of chest wall movement, head movement and general body movement to sound. The Cribogram[40] and the ARC[41] were two attempts to use this method. Both require the use of high sound levels to evoke responses. Evaluation studies for the ARC have been reported by Tucker and Bhattacharya[28] and by McCormick et al.[42] Although these methods were used in some centres in the 1980s and 1990s, they have been largely superseded by screens using AOAE and AABR.

NEWBORN SCREENING PROTOCOLS

As will be discussed later, universal newborn hearing screening has been recommended in consensus statements in the USA and Europe.[43,44] The statements also recommend using a hospital-based system as it offers unique accessibility to a high proportion of babies with the potential to achieve high coverage. This is the model that will be described here. However, in communities where the majority of babies are not born in large maternity units, a different model may be more appropriate.

Universal newborn hearing screening

TEOAE, DPOAE and AABR are all being used for UNHS. The TEOAE or DPOAE methods do not require the use of electrodes, resulting in lower disposable costs and a less-invasive procedure. However, the pass rate for OAE methods is reduced in the period up to 24 hours following birth. The NIH document recommends that a two-stage process be adopted with TEOAE being used on all babies and ABR (or AABR) being used on those babies who do not pass OAE to increase the specificity. The document also notes that some centres are using ABR screening alone and encourages sites to continue these programmes. There are also centres that have successfully used TEOAE only, achieving a high screening programme specificity by retesting those babies not passing in hospital in a screening clinic at around 4 weeks of age. A typical hospital-based screening model is shown in Figure 2.6.

Method for NICU

A separate protocol is also adopted in many centres for the testing of babies admitted to NICU or those admitted to NICU for significant periods. There is evidence that a small proportion of NICU babies with PCHI will pass OAE, although they would fail an ABR screen.[45] Babies

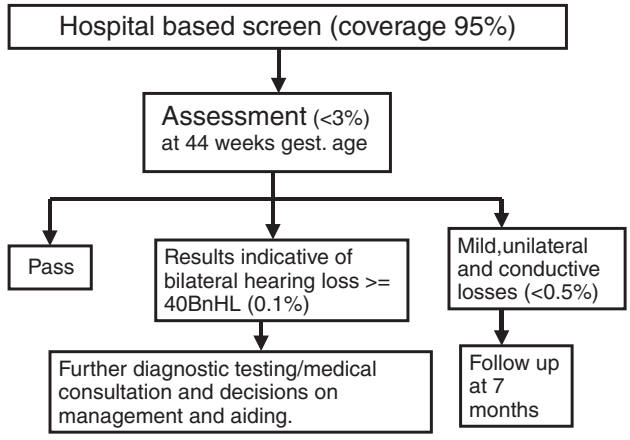

Figure 2.6 Outline of a typical hospital-based UNHS screening programme.

developing hyperbilirubinaemia requiring exchange transfusion and infants requiring prolonged assisted ventilation are particularly at risk. Rance et al.[46] have also reported a series of babies with the similar anomalous combination of raised ABR thresholds in conjunction with recordable OAEs. The pathology is assumed to be 'central' (proximal to the cochlea) and categorised as 'auditory dys-synchrony/auditory neuropathy'. Audiological outcome in such cases has been found to be variable[45,47] and presents a dilemma with regard to aiding decisions or decisions on cochlear implantation. Many programmes have therefore adopted the practice of carrying out ABR or AABR on all long-stay NICU babies. Current data suggest that this mainly occurs in babies admitted to NICU, but there may be babies in the well-baby population where this is the case.[48]

Targeted newborn hearing screening

Although most centres that carry out newborn hearing screening have adopted a universal approach, targeted screening, as noted earlier, still offers a lower-cost method to detect a proportion of the hearing-impaired babies where universal screening cannot be funded. A protocol for a targeted newborn hearing screen (TNS) requires the implementation of a well-controlled questionnaire. Typically, midwives will be asked to check for family history and refer cases to the programme. The medical staff will be asked to refer on any baby that fits the high-risk criteria. In some centres, a simple criteria of >48 hours in NICU is used for this risk group. TNS involves much smaller numbers. Given the preferred use of ABR in long-stay NICU babies and the higher specificity of ABR in the NICU group, most TNS programmes use ABR on all babies.

Follow-up of newborn hearing screening

Assessment of hearing following failure to pass a newborn screen involves a range of tests. The click ABR test is extended to include the use of tone pip stimulation to give more frequency-specific thresholds.[35] An alternative/complementary method is to use modulated test tones (auditory steady state response).[49,50] The conductive component of any hearing loss can

be estimated by using bone conduction ABR.[51–53] This may be supported by the use of the ABR latency-stimulus intensity function and high-frequency tympanometry.[54] OAE testing is important on all babies who fail the ABR screen to check for auditory dys-synchrony/neuropathy. Information may also be obtained from behavioural observation and parental observation. The US Joint Committee on Infant Hearing 2000 position statement[55] provides specific recommendations for diagnostic testing following the newborn hearing screen. Major screening programmes have also produced guidelines on the assessment of hearing in babies that are referred from the newborn hearing screen.[56,57]

SCREENING USING THE IDT OR SURVEILLANCE QUESTIONNAIRE

Ewing and Ewing[58] first described the IDT, a behavioural test suitable for use in babies over 6 months up to a developmental age of around 2 years. Its introduction as a universal screen varied from country to country. By the 1960s, this test had been adopted throughout most of the UK as a universal hearing screen, usually performed by health visitors around 6–9 months, the so-called Health Visitor Distraction Test (HVDT). It was also adopted in many other European countries but was not introduced, for example, in the United States. The methodology is described in Chapter 3.

Unfortunately, almost as soon as the HVDT was introduced, concerns began to emerge regarding its effectiveness at detecting PCHI.[59] Inaccuracy of the test became the prime suspect when it was realised that fewer than 50% of children with bilateral PCHI >50 dB HL were being diagnosed by the age of 3 years.[10] It emerged that only 10–20% of cases with apparently congenital hearing loss were generally being identified by the screen.[60] In fact, age at diagnosis was not dissimilar to that in the USA, where no screen equivalent to the HVDT was employed.[43] It was felt that the screen might even have delayed diagnosis in some cases due to false reassurance.

During the 1980s, efforts were made to improve the performance of the HVDT in the UK, by use of calibrated warblers and emphasis on training, particularly headed by McCormick (1983) in Nottingham.[60] An improved sensitivity of 86% was subsequently reported in this area,[61] but Wood et al.[62] also provide evidence of the difficulty of maintaining this quality of service over the time span of a decade. Fonseca et al.[63] surveyed the routes of identification for 104 children with congenital PCHI in nine UK centres. Whilst the HVDT correctly identified 23 cases, a further 20 remained undetected. It was concluded that unreliability of the HVDT was a significant factor in failure of the service to meet NDCS[64] targets for early identification. Data from the more extensive Trent ascertainment study[9] indicated an overall test sensitivity of only 65% (PCHI ≥40 dB HL), ranging from 54% for moderate to 80% for profound impairments. Coverage was estimated to be in the range of 80–95%, but fell to around 60% in urban areas. The relatively high referral rate, generally around 5–10%, also had resource implications. In summary, for a typical district of 4,000 births per year, one would expect 4–5 cases of PCHI >40 dB HL of which 1–2 might be identified by the HVDT, despite generating 160–280 referrals.

One of the reasons for the poor specificity of the HVDT is likely to be the relatively high incidence of fluctuating hearing loss due to OME in the population at the time of screening.[8] Whilst there is evidence of the impact of persisting OME on language development and behaviour in older children,[65,66] it is hard to justify a single screen in the first year of life for the identification of such a fluctuant condition. Moreover, the benefits of intervention in such a young age group have not yet been established.[8,67–69]

Although the HVDT has largely been withdrawn following the introduction of UNHS programmes, health visitor surveillance (HVS) continues to be practised, either in the form of unstructured enquiry, language assessments or using a parental questionnaire, most often based on the 'Hints for Parents' sheet.[60] A survey of HVS programmes prior to the introduction of UNHS indicated that they performed as well but no better than the HVDT.[9,70] Vigilance by health visitors, speech therapists and other professionals may be valuable however in the identification of late-onset or progressive hearing impairment following UNHS.

TARGETED SURVEILLANCE PROGRAMMES

Even with the introduction of a well-implemented UNHS programme, some children will present subsequently with significant permanent hearing loss.[71]Although a few such cases may be false negatives, UNHS sensitivity is high and most cases will be due to failure to complete the screen or due to late-onset or progressive hearing losses.[72,73] There is some evidence to suggest that there may be identifiable risk factors for progressive hearing loss, which has led to the introduction of targeted surveillance programmes following UNHS. The 2007 position statement of the US Joint Committee on Infant Hearing[74] recommends at least one diagnostic hearing re-evaluation by 24 to 30 months in high-risk children. In England, the national programme (www.hearing.screening.nhs.uk) has introduced the following guidelines for hearing assessments to be offered by the audiology service immediately or as soon as possible to:

- babies excluded from the screen, e.g. neonatal meningitis, microtia/atresia;
- babies referred from the screen;
- meningitis or temporal bone fracture occurring any time after the screen[75–77]; and
- parental or professional concern at any age.

In addition, the following babies should be offered behavioural hearing assessments generally around the age of 7–12 months:

- babies who missed the screen or audiological follow-up[72];
- babies with craniofacial abnormalities/Down syndrome (DSMIG, 2000[78], /cleft palate[79];
- babies with specific risk factors for late-onset deafness[80]
  - family history of PCHI in parents or siblings[79,81,82]
  - NICU/SCBU >48 hrs with no clear responses on OAEs on both ears despite clear responses on AABR[36]
  - NICU/SCBU >48 hrs who have required assisted ventilation (IPPV) for more than 5 days
  - jaundice/hyperbilirubinaemia where bilirubin reached (normally unconjugated) a level indicating the need for exchange transfusion
  - proven or suspected congenital infection due to toxoplasmosis, cytomegalovirus, rubella or herpes[83–86]
  - neurodegenerative or neurodevelopmental disorders[87]
- babies who have high levels of ototoxic drugs.

Of these criteria, the most debatable would seem to be those relating to admission to NICU and the use of ototoxic drugs. The evidence for increased risk of progressive hearing loss in NICU graduates is based on relatively small numbers of cases in the absence of any large-scale

follow-up studies. Borradori et al.[88] followed up 547 infants <34 weeks gestation who had undergone newborn ABR screening. Eight (1.4%) of these infants developed severe progressive bilateral sensorineural hearing loss. All had required prolonged assisted ventilation complicated by pneumothoraces and all had been administered aminoglycosides. Nield et al.[89] described nine children with satisfactory responses on newborn ABR testing who were subsequently identified with late-onset severe losses and again the aetiology appeared to be linked to a combination of mechanical ventilation and potentially ototoxic medication. Further studies by Konkle and Knightly,[90] Borg[91] and Robertson et al.[92] report that preterm babies with evidence of severe respiratory failure, hypoxic ischemic encephalopathy and persistent pulmonary hypertension may be particularly at risk; several reports have emerged indicating an association between extracorporeal membrane oxygenation for severe cardiorespiratory failure and subsequent progressive hearing impairment and is particularly highlighted by the JCIH.[74,93–95]

Whilst a number of drugs are known to be potentially ototoxic, those of most concern in neonates are aminoglycosides (e.g. gentamicin) and frusemide.[96,97] Although there are case reports of late-onset hearing losses following use of ototoxics in neonates, most late-onset hearing losses occur in infants with additional factors as discussed above. A small number of babies are abnormally susceptible to aminoglycoside toxicity associated with the A1555G mitochondrial mutation and may have a relevant family history. There is limited evidence that late-onset effects are dose-related and the inclusion of criteria relating to exposure to high levels of ototoxic drugs is described as ‘pragmatic’.

The effectiveness of targeted surveillance programmes is yet to be established and the outcomes of large-scale follow-up studies are awaited.

COST AND EFFECTIVENESS OF SCREENS IN THE FIRST YEAR OF LIFE

The chapter has so far focused on the methodology of each of the screening options in the first year of life and how they relate to screening principles 4, 5 and 6. Screening principles 9, 11 and 14 will now be considered, which relate to the cost, effectiveness and benefit of the screening programme.

In deciding which policy to adopt in the first year of life the cost, effect and benefit (or disbenefit) of each option of screening policy would ideally be known. The costs should include health service costs, costs to the family and costs to society.

Costs

Grill et al.[98] report on the costs of the English newborn hearing screening programme. Their data were taken from the first phase of this programme. They compared hospital and community-based sites. The cost per infant screened was found to be similar, £36.9 in hospital and £33.4 for the community. The cost of UNHS in the UK to the health service had been reported earlier[99] using data from three sites carrying out UNHS before the English programme started. The costs were around £14 per infant screened (at 1994 prices). This figure represented the cost difference between no screen and UNHS, including the follow-up of false positives. Davis et al.[9] compared this figure with those from programmes in the USA, noting that it fell in the middle of the range. Using the change in average earnings index (Office of National

Statistics, UK[100]) between 1994 and 2006, the figure of £14 per infant screened becomes £23 per infant screened for 2006. This value is less than that reported by Grill et al.[98] Factors such as the introduction of a national database and differences in protocol could explain the difference in values.

Effectiveness of screens

The effect of UNHS can be measured in terms of yield. The yield of any screen is affected by the coverage and the sensitivity of the screen. Studies on the performance of UNHS have necessarily been limited due to the size of the study required (typically 1,000 babies required per case identified). The majority of studies have therefore been carried out on an at-risk (AR) population. Obtaining follow-up data has also proved difficult, and very few studies have complete follow-up data as a result. Davis et al.[9] summarised the results from such studies. The Wessex trial[14] represents the only randomised controlled trial amongst these studies. The protocol used TEOAE followed by AABR. The yield for PCHI for the newborn screen was 1.2/1,000 (confidence intervals 0.8–1.7), which is close to the expected prevalence of 1.12/1,000.[9] Davis et al. also report the other large source of data on UNHS in the UK where the yield was 1.5–2/1000.[13] Results from the USA (e.g. White et al.[12]) generally give higher yields but often include cases of milder hearing loss. Results from studies on targeted populations, e.g. Lutman et al.[23] and Mason et al.,[38] suggest that the field sensitivity of both the OAE and ABR should be high, although there is evidence of a significant number of cases of acquired hearing impairment as noted earlier, e.g. Stevens and Webb.[22] Davis et al.,[9] reviewing the available data, suggest a value nearer to 80% for the expected programme sensitivity when using OAE, ABR or a combination of the two.

Cost per case

The cost per case is very sensitive to the definition of the hearing threshold of the target population. In the English programme, the target population is cases where the average hearing threshold is estimated to be >40 dB HL. Uus et al.[101] reported on a study carried out within the English newborn hearing screening program (NHSP), which aimed to assess the full economic costs of implementing the programme. Average costs per case detected across sites was £31,410 for the NHSP, which was approximately half that of the IDT at 8 months of age which was being phased out. The NHSP had a higher yield. The difference was even greater when family costs were included. They concluded that the findings supported the policy of implementation of NHSP and the phasing out of the IDT at 8 months of age.

In addition to being the most cost-effective hearing screen in the first year of life, UNHS also has the benefit of an earlier diagnosis compared with later screens. The published evidence from UNHS programmes has reinforced the conclusion in the NIH consensus document, which recommended that universal screening for hearing impairment be carried out before 3 months of age and which was the basis for similar statements made in the European consensus statement on newborn hearing screening.

Finally, it is important to consider the harm that a screen can produce (principle 11). Reports to date indicate that the degree of anxiety raised by UNHS is very limited.[102,103] However, there remains the potential to cause concern to parents particularly with false positive results. Programmes should ensure that follow-up of false positives is quick and efficient.

OUTCOMES OF UNHS PROGRAMMES

As more UNHS programmes are established, data are beginning to be published on the outcome of these programmes. Weichbold et al.[73] report on a 10-year outcome of the Austrian hospital-based UNHS programme. Children who had undergone screening were compared with those who had not. At age 6 months, 69% of screened children were diagnosed with a hearing impairment compared with 6% in the unscreened group. The figures for intervention by 6 months for the screened and unscreened groups were 61% and 4%, respectively.

Uus and Bamford.[104] report on the outcomes from the first phase of the English UNHS programme. A total of 169 infants had been detected with permanent bilateral hearing impairment from 169,487 screened. The median age of identification and early enrolment in the support programme was 10 weeks and the median age of hearing aid fitting was 16 weeks. Infants with moderate hearing impairments were fitted significantly later than those with a severe or profound impairment,

SCREENING AFTER THE FIRST YEAR OF LIFE

Intermediate screens (12 months to school entry)

Fortnum et al.[71] ascertained 17,160 children with PCHI ≥40 dB HL and demonstrated that the prevalence increased from an estimated 1.07 per 1,000 (1.03–1.12) at 3 years to 2.05 (2.02–2.08) per 1,000 for children aged 9–16 years. In the past, a variety of methods were often employed by health visitors or clinic doctors in an attempt to screen hearing in children over 12 months. These included speech discrimination tests, e.g. Kendal or McCormick toy tests, or performance tests using warble tones or voiced sounds ('go/ss').

Haggard and Hughes[8] reviewed the evidence relating to intermediate screens and found that practice was highly variable, often with a lack of clear objectives and quality control. There was little usable data on sensitivity, specificity or incremental yield and no comparison between alternative arrangements. Whilst identification of children with previously undetected, acquired or progressive permanent hearing impairment is obviously desirable, the number of such cases would be relatively very small compared with the large number of children with conductive hearing loss in this age group. A single screen could not determine which cases were transient, so that specificity would inevitably be low. It was concluded that there was no justification for an intermediate screen.

Most districts now operate a reactive system for this age group, referring for an audiological assessment in cases where there is parental or professional concern regarding hearing or speech development, a history of repeated ear infections, upper airway obstruction or developmental or behavioural problems. There is, however, compelling evidence to support testing of the hearing of all children following bacterial meningitis[105] and temporal bone fracture.[76,77]

Local guidelines for continued surveillance of children with Down syndrome or congenital cleft palate generally include hearing assessments at least annually until the age of 5 years (Cone-Wesson et al.[79] (www.dsmig.org.uk)).

School-entry screening

The first attempts to test hearing in school in the UK date back to the 1920s when the 'whispered voice' test was introduced. In the 1940s, screening pure-tone audiometry became avail-

able and was introduced as a universal screen throughout virtually all districts over the next 20 years. In 1976, the Court report in the UK recommended that hearing screens be carried out at least twice at school, the initial screen including a test on school entry.[105]

Previous reviews of hearing screening of school-aged children have commented on the variation in practice, standards and lack of audit of outcome measures.[9,106–108] In 2007, a Health Technology assessment reviewed the current practice, accuracy, effectiveness and cost-effectiveness of the school-entry screen (SES) in the UK.[109] This included the results of an extensive postal questionnaire sent to lead clinicians responsible for the SES.

It was found that the initial screen is predominantly accomplished by an adaptation of pure tone audiometry at specific frequencies, generally 0.5, 1, 2 and 4 kHz often performed at a fixed level (the sweep test). The pass threshold is generally set at 20 or 25 dB HL, with some districts accepting low-frequency thresholds up to 30 dB HL. One or two tests may be performed, usually during the year of school entry, with test intervals ranging from the same day to in excess of 12 weeks. The test is most often performed by the school nurse or their assistants (85%), but some districts utilise audiometricians or health visitors. Testing is generally carried out within the school, where excessive ambient noise may limit specificity. Guidelines for training and testing by non-audiology professionals have been compiled[110] but there has been little monitoring or evaluation of quality standards.

The survey as reported in the HTA indicated that the SES is used in 90% of state schools achieving over 95% coverage for those eligible. Referral rates are variable with a median of 8%. Whilst the yield from UNHS is estimated to be approximately 1.0 per 1,000[9,15] the prevalence of bilateral PCHI ≥40 dB HL has been shown to increase through childhood to approximately 2 per 1,000 by the age of 9 years.[71] Obviously, post-newborn routes of referral including IDT and professional/parental concern are important, but Davis et al.[9] estimated the yield from SES for bilateral PCHI ≥40 dB HL to be 0.4 per 1,000 children screened. More recently, valuable cohort comparisons provided by the Waltham Forest group were reported in the HTA survey and indicate that prior to the introduction of UNHS, SES provided a yield of 0.48 per 1,000, compared with a yield of 0.27 per 1,000 following UNHS.[109] The report argues that although there is no RCT evidence, it is reasonable to assume that the effects of moderate or greater bilateral hearing loss identified at school entry are marked and there is a long-standing belief in the necessity for intervention.

Unilateral or mild sensorineural impairments may also be identified. In the Waltham Forest study, 0.63 per 1,000 unilateral cases were newly identified in the pre-UNHS group compared with 0.07 per 1,000 following the introduction of UNHS. The clinical implications and benefits of intervention in this group, however, are not established.[8]

OME is highly prevalent at the age of school entry and can affect long-term cognitive development and behaviour and ultimately educational achievement.[65,66,111] (The effectiveness of referral and intervention, however, remains under scrutiny.) A recent meta-analysis confirmed that for well-defined cases, ventilation tubes do improve hearing for as long as they are in place, but that recurrence in persistent cases may require re-insertion.[112] Evidence from the comprehensive UK TARGET study (Trial of Alternative Regimens for Glue Ear Treatment) supports consistent if modest benefits to physical health and development from intervention for children of school-entry age.[109]

The value of a single screen for a fluctuating condition such as OME is questionable. Specificity for persisting cases will be low, leading to costly, unnecessary referrals, whilst sensitivity for OME may be affected by transient resolution, depending, for example, on seasonal variation.

The use of screening tympanometry has been considered[113,114,115] but a range of sensitivities relative to PTA from 40 to 90% has been reported, and tympanometry would not, of course, identify sensorineural impairment.[8] The use of a parental questionnaire relating mainly to a previous or family history of ear symptoms or current difficulties has also been tried as an alternative to screening audiometry, but sensitivities similarly ranged from 34 to 71%.[116,117] Other methods including spoken word tests and use of OAEs have produced variable results, although notably Lyons[118] reported a 98% sensitivity using a combination of DPOAE and tympanometry.

There is no evidence to justify further screens after school entry. However, professional vigilance should continue and a hearing test performed on any child experiencing learning or behavioural difficulties or following specific conditions such as meningitis or significant head injury.

CONCLUSIONS

The chapter has briefly reviewed the options for early identification of hearing impairment with reference to principles of effective screening. In the first year of life, UNHS has been shown on current evidence to offer the most cost-effective method of detecting PCHI. Published reports indicate that with UNHS it is possible to achieve the screening goals of high coverage, low-refer rate and a high yield. The application of electrophysiological techniques, supported by OAE and tympanometry, allows for initial diagnostic assessment to be rapidly completed. However, a significant minority of PCHI is not present at birth and there is a need for a continuing hearing surveillance programme in infancy and early childhood.

There remain many questions. For example, screening for unilateral and mild hearing impairment is still a subject of debate as, unlike moderate or greater bilateral PCHI, this type of impairment does not fit the screening principles so well. There is little knowledge on the precise degree of impairment at which a hearing aid should be fitted and the age at which this should be carried out following failure to pass a UNHS. The proportion of PCHI present at birth and the onset and time course of acquired hearing losses is not known with precision. To answer these and other questions on screening and surveillance will require major research programmes conducted on large birth populations.

After the first year of life, none of the potential methods for screening are able to meet fully the screening principles set out at the beginning of the chapter. Universal school-entry screening is a practical screen and is still implemented in many countries. As UNHS is introduced, further studies are needed to determine whether universal school-entry screening is cost-effective as part of the overall programme to detect permanent hearing impairment in childhood.

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3 Behavioural tests of hearing

F. Tweedy and R. Booth

INTRODUCTION

Behavioural tests of hearing assessment have been in clinical use well before electrophysiological techniques. However, with the introduction of newborn hearing screening in some countries, the identification of hearing loss for many children is through the application of electrophysiological techniques. Nevertheless, behavioural confirmation of the frequency-specific and ear-specific hearing thresholds as early as possible is essential to confirm the results of electrophysiological testing, for effective ongoing management and optimisation of any amplification. In addition, behavioural testing is appropriate for the identification of hearing loss when onset occurs after screening and progressive losses which are too mild at birth to be detected.

Strategies and methods for behavioural testing have been developed and refined over many years to structure the conditions which optimise the elicitation of responses appropriate to the child's developmental level. Such responses can be to stimuli with specific acoustic parameters, thus giving detailed information about a child's complex auditory function. Not only do behavioural tests demonstrate that the sound is being processed by the ears and auditory pathways, but the child can be observed to respond to sound due to high-level cortical processing.

The results of any behavioural hearing assessment should allow the clinician to comment on hearing thresholds, the extent and nature of any hearing loss and the inter-aural symmetry of hearing. This requires the clinician to routinely use sound-field, air and bone conduction and ear-specific behavioural test techniques. This chapter discusses the behavioural techniques applicable to routine clinical practice.

BEHAVIOURAL OBSERVATION AUDIOMETRY

Behavioural Observation Audiometry refers to a technique formerly used in the assessment of very young babies (0–6 months) whereby changes in the state of activity of the child were observed and judged as to whether they were in response to sound stimuli. Stimuli most likely to produce behavioural responses in such young children were usually broad-band and high-intensity. Unfortunately, this meant that the assessment was not frequency-specific, and prediction of thresholds of hearing from the results was unreliable. Furthermore, there was variability in judgement of responses among testers, and testers could have a high rate of misinterpreting random movements as responses in control trials with no auditory presentations.[1] Thus Behavioural Observation Audiometry is a very questionable technique, particularly since the advent of electrophysiological techniques. Behavioural Observation Audiometry

should not, however, be considered a low-technology option for hearing assessments: Gans[2] recommends that systematic scoring, without observer bias, is obtained using video recordings of the procedure with presentation of sound and no-sound trials. On playback, the observers score the behaviour of the child but are denied knowledge of the details of the stimulus or whether a stimulus was presented. Automated systems for assessing the motor responses of young babies have, however, been found to be much less reliable than electrophysiological techniques for screening the hearing. Fortunately, as most babies mature, their behavioural responses can be reliably assessed and normally be used to test hearing in a structured clinical setting such as in the Distraction Test.

THE DISTRACTION TEST

This test has been developed from the 'Distracting Test' described by Ewing and Ewing in Manchester in 1944. It involves the attracting and releasing of the attention in front of the child with a play activity, the presentation of a series of frequency-specific auditory stimuli outside the child's visual field and observing the child's response of turning towards and localising the sound source.

Distraction testing was traditionally used in the screening of infants at 8 months of age by health visitors in the UK. Its role has diminished, although it is still used to quantify hearing loss in children from around 6 or 7 months of age. Its reliability as a test decreases for older children as discussed subsequently in the section Use of the Distraction Test.

The test arrangement

The testing is performed in a quiet, sound-treated room arranged as shown in Figure 3.1. Care is taken to ensure that there are no reflective surfaces or shadows cast which might cause a child to turn for reasons other than hearing the auditory stimulus. Background noise should not exceed 30 dB (A).

The test involves two trained professionals, the distractor and the assistant, and the child and one parent (or other adult). The child sits halfway along the parent's lap supported at the waist. The parent and child sit facing the distractor.

Role of the distractor

The distractor usually kneels or sits behind a low table facing the child. Examination of the child should be made to see if the child can physically make a head turn sufficient to show a response and can be visually distracted. Usually, this can be observed while the history is being taken but with some handicapped children it will be necessary for preliminary tests to be carried out, e.g. testing the child's ability to visually follow a moving object in a 180-degree arc from side to side.

The distractor is in charge of the test and is the person who controls the child's attention, indicates which stimuli are to be presented and judges whether there has been a true response to the sound. Toys are used to engage the child's attention forward and then, by reducing the play activity, releasing the child's attention prior to introduction of the stimulus. The common technique is to cover the toy being used whilst continuing minimal movement of fingers or toy. This 'phasing' of the child's attention is an important aspect of the test. The developmental

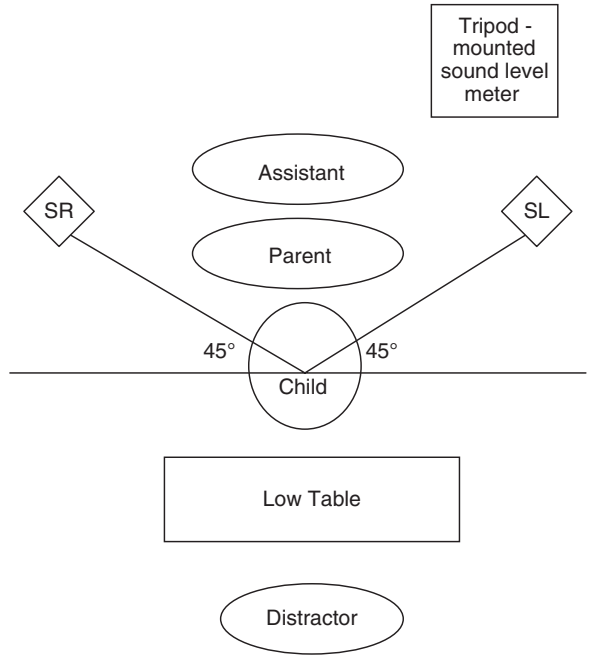

Figure 3.1 Test arrangement for the Distraction Test.

stage of a young infant's attention and listening skills means that his or her attention will not be oriented towards the sound if he or she is too engrossed in the activity in front. Pauses in distracting should not be so long that they allow the child's gaze to wander and care must be taken not to distract in a manner which is so interesting that the child does not turn even though the sound has been heard. The distraction activity can be modified to suit the child's abilities, and it may be that for an older child, the Distraction Test level of play activity can be modified to prevent false positives occurring. Visually impaired children can be distracted using tactile (stroking) stimuli, or the room lights can be dimmed and a light source, such as an otoscope, can be used to control the child's attention.

It is the responsibility of the distractor to indicate to the assistant which stimuli should be presented, on which side and at which time (by phasing attention). It is the distractor's role to assess whether or not a turn was in response to the stimulus or for other reasons, such as visual cues, competing auditory stimuli, or a random check, and whether or not there was correct localisation of the sound source. The distractor needs to be able to observe the child's face as this may give clues that a stimulus has been heard prior to localisation, e.g. a look of recognition such as widening of the eyes or a smile prior to turning. Care must be taken not to maintain eye contact with the child, which may fix his/her attention forward, and so it is better to concentrate the gaze a little below the eyes. The distractor must be careful not to glance towards the stimulus and give cues of its presence and location. In some cases, where the child has poor head control or is visually impaired, the test may be modified so that repeatable responses, such as eye turns or reaching for the stimulus, may be accepted.

Role of the assistant

The assistant introduces the auditory stimuli and gives the social reward as directed by the distractor. Stimuli are presented on either side as follows:

- One metre from the child's ear to enable the sounds to be presented at the ear at 30 dB (A). (If a child has significant developmental delay, then sometimes the test is started at a distance of about 15 cm from the ear in order to stimulate the child within a distance in which they might show interest.)
- At an angle of 45 degrees on either side behind the child to avoid visual cues being given.
- On a level with the child's ear, as this makes it easier for the child to localise.
- For up to 5 seconds at the initial intensity levels, with the intensity then raised if the child does not respond.

Historically, distraction testing involved the use of noise-makers with varying bandwidth and frequency specificity. Since the advent of electronically produced stimuli, both bandwidth and specificity have been optimised by the use of warble tones. However, the high-frequency rattle was developed for the purpose of this test and still has high credibility in testing.

The stimuli should be presented at the minimum intensity level. If the child does not turn, then the sound is made at a higher intensity level. This can be done in two ways, either by coming closer to the child's ear or by raising the intensity of a sound from the same distance of 1 metre. The advantage of the latter method is that one is much less likely to give unintentional clues as to the presence of the stimulus or assistant. In addition, errors in replicating the distance when measuring sound intensity have smaller effects at this larger distance. However, the disadvantages are that one might not be able to produce sufficiently high intensity for some hearing-impaired children to hear at that distance and some sounds will lose their frequency specificity at higher intensities (e.g. sibilant /s/). If there is no response at maximum intensity, the child is given a tactile or visual clue to see if this elicits a turning response. If so, this suggests that the child's state of arousal is appropriate for the test but that he or she has not heard the auditory stimuli. The introduction of a visual or tactile stimulus is helpful in differentiating the child who has not heard from the one who has not responded for other reasons.

When presenting the stimuli, care has to be taken to avoid presentation in a predictable alternating manner and to avoid inadvertent sensory information, which may cue the child as to the presence of the stimulus. Examples of such information include shadows or reflections and the sight or sound of the assistant moving. If the child turns and localises the sound source accurately, this is recorded as a response.

If a response is obtained, the intensity is measured using the dB (A) weighted scale on a tripod-mounted sound-level meter with the 'fast' response of the meter in order to observe and record the peaks in intensity of the stimulus. The intensity, type and side of the stimulus producing the response are recorded. The maximum level at which the child failed to respond to the auditory stimulus is measured and recorded (e.g. NR (no response) at 90 dB (A)).

Control trials in which all conditions of testing are met other than making the sound should help the distractor in deciding if the child is turning genuinely to the stimuli or is turning for some other reason. These should be performed periodically to ensure that the responses are true. If there is random 'checking' behaviour by the child, this may be stopped by one of these ploys:

- phasing the child's attention without presenting a stimulus until the checking ceases;
- keeping a toy in view to attract the child's attention forward, i.e. a more interesting distraction in front; and
- the distractor and the assistant changing places.

The Distraction Test tests both ears together, and assessment of the responses from presentation of sounds on the right and left does not mean that ear-specific testing has been carried out. Care must therefore be taken in the way in which results are reported to parents and professionals. Presentation of sounds on the right and left can provide information about the child's ability to localise.

Any localisation difficulties should be noted, as these difficulties may indicate a difference in hearing level between the two ears, though children with severe and profound bilateral hearing loss may have difficulty localising generally.[3] Most importantly, one should always be vigilant that the apparent responses were not, in fact, random turns or in response to other non-test stimuli. Certainly, mislocalisation is a criterion for failure of the baby in any screening programme.

Test stimuli

Frequency-specific auditory stimuli are used to test high, middle and low frequencies separately. This is necessary to measure hearing loss in those instances where auditory sensitivity differs over the speech frequency range, e.g. those with a 'ski-slope' or 'U' shaped hearing loss. Specification of spectra of frequency-modulated (FM) warble tones is variable amongst the different manufacturers of signal generators but are generally preferable to mechanically produced sounds, because of increased frequency specificity (Figure 3.2).

High-frequency stimuli

The Manchester high-frequency rattle (available from the Ewing Foundation, c/o Human Communication and Deafness, University of Manchester)

The high-frequency rattle is often presented first. Its spectrum contains a broad band of high frequencies, from about 6 kHz to above 20 kHz, which usually attracts the attention of babies.

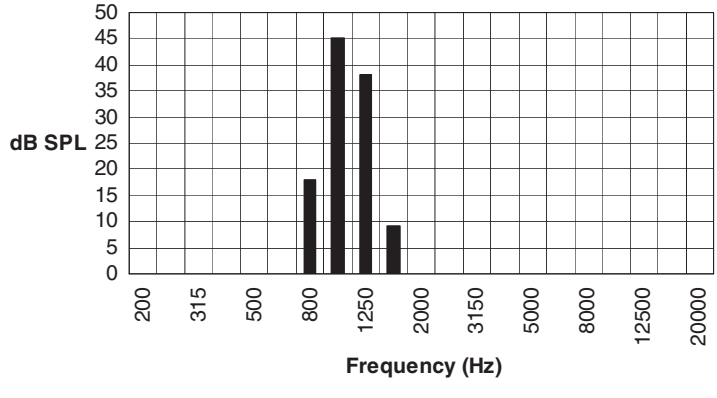

Figure 3.2 Spectrum of a frequency-modulated warble tone, centred at 1 kHz.

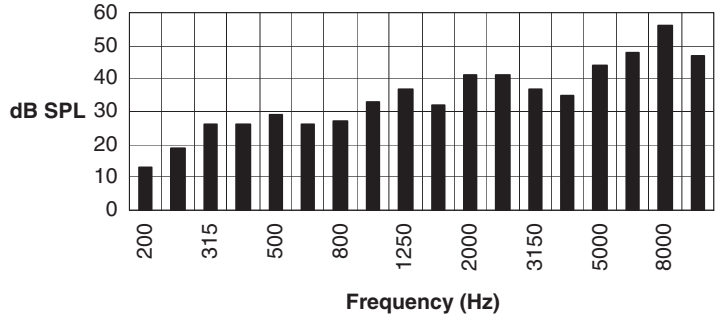

Figure 3.3 Spectrum of sibilant /s/ at raised intensity.

Repetitive sibilant /s/

When correctly produced, this contains audible frequencies from about 3–10 kHz. It loses frequency specificity when the sound is raised due to the noise of the increased airstream (Figure 3.3).

Warble tones centred at 3 or 4 kHz

This gives more frequency specificity than the two previous stimuli, but young babies might be less responsive to warbles because of their narrow bandwidth.

Middle-frequency stimuli

Warble tones centred at 1 or 2 kHz

These may be used and are more frequency-specific than other stimuli (Figure 3.2).

'G' chime bar (about 10 or 11 cm in length)

This produces frequencies around 1,600 Hz when struck with the knuckle or soft hammer (Figure 3.4). Impact energy is problematic if the chime is hit with a hard striker such as a hammer or fingernail, as the signal then becomes broad-band (Figure 3.5).

Low-frequency stimuli

Warble tones centred at 500 or 250 Hz

Warble tones are more frequency-specific than other low-frequency stimuli.

Humming sound

This is a continuous voiced low-frequency sound produced with the lips closed. A little intonation in the voice makes a more attractive sound but care is needed that this does not produce intensity fluctuations. The continuous nature of the voicing in this stimulus reduces intensity

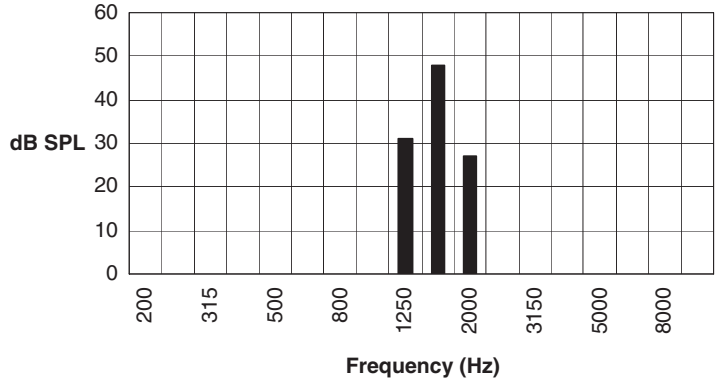

Figure 3.4 Spectrum of 'G' chime bar (struck with knuckle).

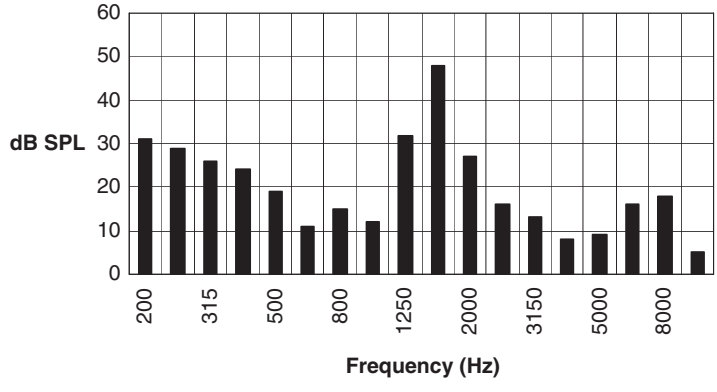

Figure 3.5 Spectrum of 'G' chime bar (struck with hard hammer).

fluctuations which can be present at the initiation of each voicing in other voiced stimuli, such as in a repetitive /oo/ stimulus or mumbled speech.

'C' chime bar (about 17 or 18 cm in length)

This produces frequencies around 512 Hz when struck with the knuckle or soft hammer. The problems with this stimulus are the same as for the middle-frequency 'G' chime.

Additional stimuli

/baba/

This can be used if a high-intensity stimulus is required though it does contain low, middle and high frequencies so does not facilitate testing at any particular frequency range.

A drum may be used if there is no response to the stimuli described earlier and may be used to test for an auro-palpebral reflex (see below, The Auro-palpebral Reflex). Such stimuli contain broad bands of frequencies.

Use of the Distraction Test

Prior to newborn screening, the Distraction Test was the basis of health visitor screening of infants in the UK at the ages of 7–8 months. Some babies with a hearing impairment were found to pass the screen, and in some instances, this was attributable to poor test techniques. McCormick[4] was able to show an improvement in test reliability when the testers were given refresher courses. With the onset of newborn (neonatal) screening, using electrophysiological techniques, fewer babies have their hearing loss detected using the Distraction Test.[5] However, not all pre-lingual hearing losses are present in the newborn (neonatal) period, or a hearing loss may progress from a mild loss, facilitating a pass in newborn screening, to a more significant loss in infancy. Typical surveillance programmes also require children with known risk factors for progressive hearing loss to be seen again around 8 months of age, regardless of the outcome of the screen.

The Distraction Test works well for the 6- to 8-month age group due to its use of a 'social reward' to an infant's response to a sound. As children become older, this reward becomes less worthwhile to them. It is not sufficient as a reward for continued response to the sounds and they quickly inhibit their response to the sounds. Further difficulties in the older infant arise as they will be more able to pick up cues and to learn when the sounds will be presented because of the phasing of attention prior to presentation of sounds. The Distraction Test can sometimes remain the test of choice in a slightly older infant in the presence of developmental delay or other needs. In general, it should be used in older children with caution. The Distraction Test should be carried out only by experienced testers because of the high risk of children passing the test while having a hearing loss. In addition, the format of the test, the stimuli used to test the hearing and the methods for measuring the sound levels means that the hearing can be screened only at 30 to 35 dB (A). With the advent of newborn hearing screening, behavioural tests must be more able to identify the mild and unilateral hearing losses which are not targeted by the newborn hearing screen, but these can be missed by the Distraction Test. The Distraction Test can be a tool in identification of hearing loss though vigilance is necessary to ensure that the standard of test technique using this tool is high. The test can be a useful tool when carried out well, but changes in the focus of paediatric audiology mean that the Distraction Test no longer provides all the audiological information required by current audiological services.

The Auro-Palpebral (blink) Reflex (APR)

Examination is often made for the presence or absence of an APR whenever the responses in the Distraction Test are at raised intensities. This is usually done by introducing a /ba/ sound near the ear at high intensities. A small screen or the assistant's hand should be placed over the assistant's mouth to prevent tactile elicitation of the reflex. Usually the sound is first introduced at about 80 dB (A). If there is no corresponding blink, the sound is made at a higher intensity: if there is a reflex, it may be useful to find the APR threshold by testing at lower intensities, though habituation may soon occur. The reflex threshold is thus obtained. The test is carried out on each side separately. It provides limited information about hearing levels. A reflex present at 'normal' levels of about 80–100 dB (A) does not exclude a hearing impairment because the child may have a cochlear hearing loss with accompanying loudness recruitment and/or hearing, which varies in sensitivity across frequencies.

VISUAL REINFORCEMENT AUDIOMETRY (VRA)

This procedure enables hearing thresholds to be measured in young children whereby a head turn, in response to hearing a sound stimulus, or in some cases a tactile stimulus, is reinforced by a visual reward. Localisation of the sound stimulus is not required, and, indeed, the stimulus and the reinforcer may be spatially separated. The ability of the child to be conditioned to turn is a prerequisite for the test. Since conditioning can be to a tactile stimulus in difficult cases, the test does not depend on the auditory skill of localisation and, thus, is not confounded by poor localisation ability, unlike the Distraction Test or Conditioned Orientation Audiometry.

VRA is usually possible from the age of 6 months. The test is applicable up to an age of 2 or 3 years at which age children quickly inhibit responses and become less motivated by the visual reward. Habituation reduces the number of signals which can be used to determine threshold levels, and the older the child, the more quickly he or she tends to habituate.[6,7]

The test arrangement

Testing is usually carried out by two testers using two rooms arranged as depicted in Figure 3.6. (If necessary, the test can be performed in one room only and with a single tester.) The visual reinforcer is placed next to or on top of the loudspeaker but they can be separated. When they are close to each other, localisation of the sound may facilitate the turn to the reinforcer. Primus[8] has shown that the VRA response is not contingent upon localisation but that the performance in the test was affected by localisation of the sound. More infants were conditioned and the number of trials needed for conditioning were fewer when the visual reinforcer and sound source were adjacent than when they were separated.

The loudspeakers may be placed at angles of 45, 60 or 90 degrees from the child. The distance of the loudspeakers from the child's ears was investigated by Magnussonet et al.[9] In one arrangement, the loudspeakers were placed upon movable arms and positioned 15 cm from

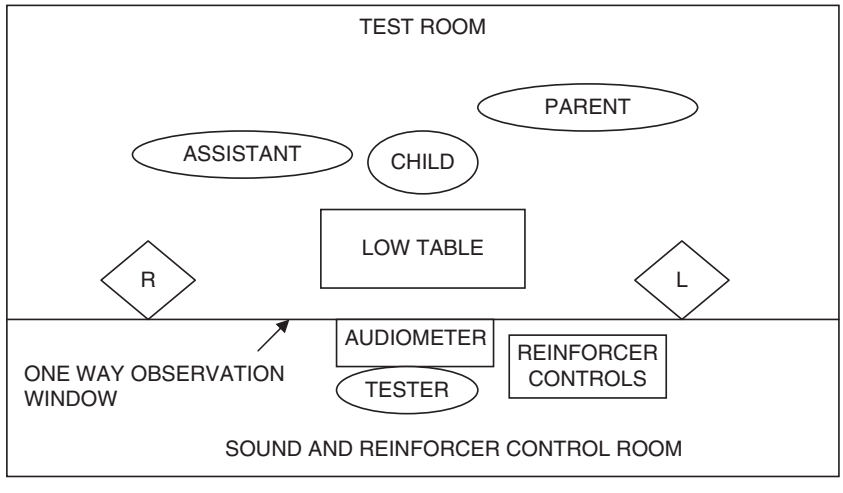

Figure 3.6 Test arrangement for Visual Reinforcement Audiometry. R: Right loudspeaker and adjacent reinforcer; L: left loudspeaker and adjacent reinforcer.

each ear. In the other arrangement, the loudspeakers were placed 50–70 cm from the child. Real ear sound pressure measurements were made and compared between the two positions. Predictably, it was found that in the 15-cm position the measurements were very variable due to slight head movement, whereas the more distant positioning of 50–70 cm was found to be more consistent; variations increased again at a distance of 1 metre. However, the International Standard ISO 8253 requires that the loudspeaker distance is at least 1 metre. This suggests that clinicians should carefully calibrate their VRA clinic and should be aware of the magnitude of fluctuations for their own particular acoustic conditions. The sound-field should be calibrated in dB HL to allow plotting on an audiogram for comparison with future ear-specific results and use in hearing aid fitting procedures.

Reinforcers which are commercially available include revolving lights, toys with eyes which light up, and puppets which illuminate; there is increasing use of video displays.

Test method

The test involves conditioning a child to respond, with a turn to a visual reinforcer, to a sound introduced from a loudspeaker in the sound-field or through earphones or a bone vibrator. The sound and visual reinforcer are initially introduced together with the assistant pointing out the reinforcer, if necessary, at this stage. When the child begins to anticipate the visual reinforcement, the sound is introduced alone and an appropriate turning response is rewarded subsequently with the visual reinforcer. Children unable to turn their head or unable to see the visual reinforcer will need to have their hearing assessed using other methods or modifications made to the standard VRA technique and set-up.

Before introduction of the test signal, the child's attention is kept in a forward position by the use of play activity. This should be maintained throughout, although the level of the play activity should be modified according to the child's developmental level. If there are two testers, one in the test room and one in the adjacent room, then agreement between the two as to whether or not a response was a true one may increase the validity of the testing procedure. Computer control of signal level and time period for scoring a response can add objectivity to the test execution, and storage of false positive responses (in control trials) can be used in computer decision-making algorithms.[10]

Test signals

VRA may be performed using frequency-modulated warble tones usually centred at frequencies of 0.5, 1.0, 2.0, 3.0 and 4.0 kHz. Narrow-band noise can be used if the tones fail to elicit a response. The filter roll-off of most audiometric narrow bands is, however, usually very gradual giving a much wider bandwidth than desirable. The duration of the signal is usually about 2 seconds. The intensity of the sound in a particular clinic, at a particular location where a child is normally seated for testing, may be calibrated using a sound level metre fitted with octave or one-third octave band filters and the dial reading on the audiometer pre-calibrated to read in dB HL.

The signal may be introduced through insert earphones, as shown in Figure 3.7. The earphones are placed in the ear canal using a foam tip or, if the child wears hearing aids, they may be attached to the earmould. Insert earphones enable more precise information to be obtained about individual ear thresholds. Masking may be used, the narrow-band noise being introduced at specific sensation levels in one earphone whilst the signal is applied through the

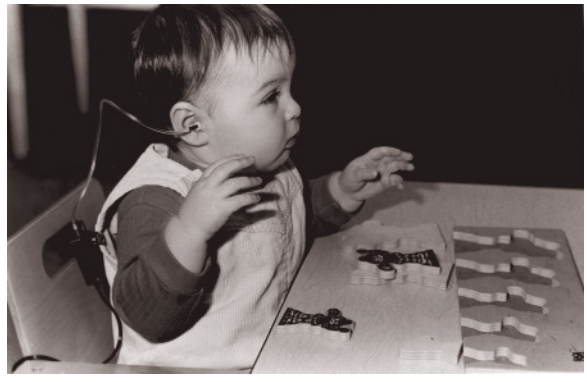

Figure 3.7 Use of insert earphones.

other and conditioning re-established to ensure responses to the signal and not the masker. Bone conduction testing can also be carried out using the same principles of testing as air conduction.

Habituation of the response

Habituation of the response was studied by Primus and Thompson[6] in relation to the number of reinforcers available and breaks in the test sessions. Two reinforcers led to more responses than one before habituation and a 10-minute break in the session led to a minimum of five additional responses in the rest of the session for 1-year-old but not 2-year-old children. Culpepper and Thompson[11] investigated the effects of reinforcer duration of 0.5, 1.5 and 4.0 seconds on the response behaviour of 60 pre-term 2-year-olds using a 50 dB HL bandpass noise stimulus. They found that significantly slower rates of habituation were obtained when the duration of the reinforcer was 0.5 seconds compared with 4.0 seconds. It was thought that decreasing the duration would be particularly helpful in increasing the duration of testing opportunities in children at the older end of the VRA age range.

CONDITIONED ORIENTATION AUDIOMETRY (COA)

This technique has some similarities to both Distraction Testing and VRA, but is sometimes confused with the latter as it involves visual reinforcement of a head turn. However, it requires the child to be able to localise auditory stimuli on either side and thus is dependent on this ability. COA uses stimuli presented further away from the child than in the Distraction Test and thus localisation is more challenging. As such, it is important that the acoustic testing environment, the acoustic stimuli and the angle of presentation are selected to facilitate this ability in young children. For a child who is unable to localise, the test results may be compromised and the technique of VRA may be appropriate, i.e. the child is conditioned to turn to one side only.

Localisation usually involves complex binaural processing of interaural cues at both brainstem and cortical levels, so that failure to exhibit such behaviour implies audiological impair-

ment, which could be peripheral or central in origin. Since poor localisation is a recognised impairment in auditory processing disorder, this could be an early indicator of this disorder, though far more commonly the cause will be associated with a peripheral, and particularly asymmetrical, hearing impairment.[12] However, localisation is possible with hearing impairment, whether symmetrical or not, so the ability to localise cannot be taken to imply binaural hearing.

In COA, stimuli are presented from either side, and if the child localises the source correctly, he or she is rewarded by visual reinforcement. Localisation may, however, be impeded by reverberation if the room is not well sound-treated and by the narrow bandwidth of frequency-specific stimuli, such as frequency-modulated warble tones. Furthermore, some frequencies are more difficult to localise than others; these are different in babies and young children compared with adults due to the difference in head size impacting on head shadow and interaural intensity cues. As a consequence of smaller head size, localisation of mid-frequencies is more difficult for babies, although there is very little experimental data on the developmental aspect of this function. However, the laws of physics suggest that for a child of, say, 6 months, with an average head circumference of 42 cm, head shadow will not be optimal for frequencies below 1.3 kHz. Thus attempting to condition a child to turn to a sound of 1.0 kHz may compromise the child's ability to carry out the test.. For low-frequency sounds of 0.5 kHz and below, utilisation of interaural time cues would be predicted, regardless of head size. Alternatively, narrow-band noise may be used, but this is less frequency-specific than frequency-modulated tones.

When successful, COA demonstrates hearing and localisation ability but, in the absence of the latter, VRA should be carried out if the child is able to maintain cooperation.

THE PERFORMANCE TEST AND PLAY AUDIOMETRY

The terms Performance Test and Play Audiometry are often used interchangeably. Both are appropriate for the assessment of hearing of children of a developmental age of 2 ½ years and older until cooperation with pure-tone audiometry is achieved.

The Performance Test was described by Ewing and Ewing[13] and in its earliest form used a voiced /go/, a sibilant /s/ and other consonants presented in the sound-field to assess the child's hearing. Now warble tones are more commonly used and clinicians more readily progress on to ear-specific and bone-conduction testing. It is the latter that is sometimes referred to as play audiometry.

The term is not particularly important; what is important is that both adopt a conditioned play response to a sound stimulus to assess the hearing sensitivity. The response required is often to put a man in a boat, a peg in a board or a ring on a stick or a similar simple action each time the sound is heard. The conditioning process involves showing the child what to do and does not depend on receptive language for cooperation in the procedure. Success of the test does depend on the child's ability to inhibit his or her response until the stimulus has been detected. Often it is the younger child's inability to wait for the stimulus that prevents the test being used rather than his or her inability to respond when the sound is heard.

Toys which demand more advanced skills, e.g. those with assorted shapes which have to be put into a particular hole, are generally unsuitable as they tend to distract the child's attention from listening to the sound signal. Frequent changing of the play activity can be important to maintain the child's interest and active listening.

Test method

The child is conditioned from in front to respond with the required play action to presentation of the sound stimulus at a moderate level of intensity. Typically, conditioning is achieved by the clinician and child doing the play action together. Once the clinician can sense the child initiating the action, the clinician removes him- or herself from the activity, allowing the child to respond on his or her own. The stimulus is presented at progressively lower intensities after each positive response with variable inter-stimulus intervals. If the child fails to respond at a reduced intensity, the tester progressively raises the intensity of the stimulus. A repeatable response should be obtained at the level taken as the minimum response level. As for any behavioural hearing test, it is important to establish from behavioural cues that the child is hearing the sound during the conditioning procedure. As for VRA, a tactile stimulus can be used to establish whether the child is developmentally ready for the performance test, in order to distinguish between the child not hearing the sound and the child not responding for other reasons. An alternative is to use visual cues in addition to the sound stimulus in the initial stages, removing the visual cue once the child appears conditioned. If either of these leads to conditioned responses not being achieved when using sound alone, concerns should be raised about the hearing thresholds. Higher intensity levels can be achieved using insert earphones and headphones than by presenting stimuli in the sound-field. For children who wear hearing aids, conditioning may be carried out in the sound-field with the hearing aids *in situ* and then taken out for threshold testing.

Presenting sounds in the sound-field either from a loudspeaker or hand-held warbler tests the better-hearing ear or both ears, if equivalent. Without insert or headphone presentation, it is not possible to ascertain ear-specific information due to transmission and diffraction of the stimuli from one side of the child to the other with little or no attenuation. Even if a handheld warbler is held close to each ear, the hearing levels within each ear are not tested. An apparent interaural difference of 10 dB at the high frequencies (4 kHz) may suggest an asymmetry but does not provide any ear-specific information. The transition from sound-field testing to use of other transducers is possible even in children aged 2 ½ years and maximises information about the symmetry of the hearing and nature of any hearing loss.

Neilsen and Olsen[14] found that it was possible to obtain six thresholds from nearly 75% of children from the age of 3 years. This can be achieved or even exceeded with changes in play activity as the child gets bored, with praise and encouragement and with involvement of the family. It is also important to realise that a child will concentrate only for a limited amount of time, typically shorter for the younger child. It is therefore important to maximise the amount of threshold information obtained by using larger step sizes and not wasting responses on sounds that can easily be heard. The amount of time the child will sit and concentrate can be optimised by using just one tester to control the child and his or her attention, to condition the child, to control the play activity and to present the stimuli.

Thus one might obtain air-conduction information at three frequencies for each ear unless the minimum values of transcranial attenuation are exceeded and the results confounded by cross-hearing. Alternatively, or in addition, one or two unmasked bone-conduction thresholds may be attempted at this age if the child has a bilateral hearing loss, in order to determine if there is a sensorineural element present. However, the possibility of the thresholds being perceived by tactile rather than auditory sensations increases at the higher intensity levels, especially the low frequencies.[15] Masking can be attempted for the reliable responder. Even masking just one frequency can provide important information about the true nature and extent of any hearing loss.

Test signals

Frequency-modulated warble tones are the preferred option as they have good frequency specificity and can be used to test low-, mid- and high-frequency hearing in the sound-field or with insert-phones. Also the transition from warble tones to pure-tone audiometry may be easier than with other sounds. If warble tones are used, low-, mid- and high-frequency information is sought, usually from stimuli centred at 0.5, 1, 2 or 4 kHz.

Stimulus durations should be of around 1 to 3 seconds, as younger children will typically respond when the stimulus goes off, regardless of how they have been conditioned. It is important to have variable inter-stimulus intervals. Use of variable inter-stimulus intervals is essential to ensure that the child is not merely performing the task independent of hearing.

If a less abstract signal is needed by the child, the low-frequency live voiced /go/ and high-frequency sibilant /s/ may be used. This can be used to get the child's initial interest in the game, allowing the clinician to proceed to frequency testing as the child becomes more confident. Some threshold can be obtained by using lower voice intensities but is no substitute for frequency-specific results. It is also important that, when using the /go/ or /s/ in testing any child with the performance test, thresholds of hearing may be obtained only when the whole face is out of vision of the child. Even if the child sees only part of the face, a very small muscle movement may be sufficient to indicate that the stimulus has been uttered, and thus the procedure is no longer valid.

The minimum information to inform a management decision should be for repeatable responses at three frequencies across the speech range in the sound-field to be obtained provided there are no risk factors indicating the need for ear-specific testing. Criteria for classification as normally hearing for ear-specific testing and bone conduction should be the same as for adults, that is, 20 dB HL. There can be arguments for screening at this level and also for testing a child's hearing sensitivity to threshold depending on the clinical question being asked.

PURE-TONE AUDIOMETRY

The conventional technique for obtaining frequency-specific and ear-specific information about a person's hearing is pure-tone audiometry using supra-aural headphones or bone conduction and masking. There are recommended procedures such as the British Society of Audiology Recommended Procedures[16] and results relate to an international baseline of average normal hearing for adults.[17] This technique may be adapted for use with children, and results may be interpreted from some children as young as 2 ½ years old where it is generally referred to as play audiometry as discussed in the previous section.

The recommended procedure is generally followed for children 7 years and older, although this can be very dependent on the child and his or her attention, listening skills and behaviour. The procedure must be adaptable and changes made where appropriate as testing progresses and the child's attention reduces, while maintaining the principle of repeatable responses at thresholds.

Insert earphones (Figure 3.8) are less cumbersome and may be accepted more readily than headphones. Furthermore, they facilitate more transcranial attenuation of the sound source to the non-test ear than headphones and prevent the collapse of the ear canal, which may occur with supra-aural headphones.

Where minimum transcranial attenuation figures for headphone or insert earphone thresholds have been exceeded or where ear-specific bone conduction thresholds are desired, masking

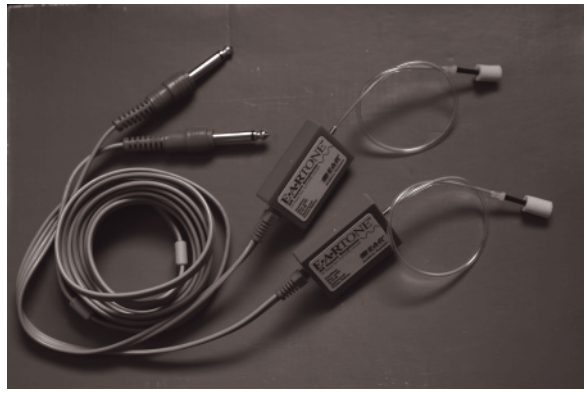

Figure 3.8 Insert earphones.

of the non-test ear is required, but cooperation with this technique will depend partly on the cognitive developmental level of the child. The technique used for masking should be a plateau technique[18] or a formula method,[19] the former being practised in Britain with the Recommended Procedure of the British Society of Audiology.[16] Calibration of narrow-band noise to ISO 389–4 now allows masking to be performed with very young children as there is no need for verbal explanation of the task; rather, the noise is introduced gradually and any superfluous responses to changes in masking level can be ignored.

NON-ORGANIC HEARING LOSS

The elevation of thresholds above organic thresholds may occur in testing some children, either subconsciously or consciously on their part. In most cases, this presents as a moderate–severe bilateral sensorineural hearing loss with no relevant medical history.[20] Occasionally, it may present as a unilateral hearing loss, often severe or total with no shadow results from the non-test ear for unmasked thresholds. In both cases, it is the role of the audiologist to try to determine the true organic thresholds in order to manage any underlying hearing loss appropriately.

There are several methods of determining organic hearing thresholds in the presence of a non-organic pathology, most of which entail distracting the child's attention away from the loudness of the signal to some other task such as in an ear-pointing technique for bilateral cases where the child concentrates on lateralising the sound, as described by Nolan and Tucker.[21] Other successful techniques are to ask the child to say 'yes' or 'no' when the sound is heard; with a 'no' response revealing to the clinician that the sound must have been heard. Speech audiometry, using pre-recorded word lists such as the AB wordlist,[22] may also be helpful if a child is asked 'if he or she knows' the words to be presented and repeated, and the intensity of which is surreptitiously reduced in order that the level required for discrimination of speech may be compared with normal values. For unilateral non-organic hearing losses, the Stenger Test should give organic thresholds for pure tones with relative ease due to the listener being unaware of sound being present in the admitted normal ear if louder sounds are perceived in the opposite ear.[23]

CHILDREN WITH COMPLEX NEEDS

Behavioural testing of children with complex needs is preferable to electrophysiological assessment, where possible, as results can often be more frequency-specific and there is no need for sedation or anaesthetisation. Testing children with complex needs should follow the same principles as testing children without additional needs, but some adaptations may be needed.

Repeatable motor responses are required to verify true responses with the usual requirement of control trials as part of the procedure. For some children with poor head control this can be as simple as a repeatable eye deviation. Often the response is much more delayed because of the time needed to process the sound and to initiate the response. In addition, a longer duration stimulus can be helpful to allow the child to recognise the stimulus and respond to it.

Carers may be able to help suggest appropriate motor responses that can be used in performance or play audiometry when a child has difficulty with fine motor control. For children being tested with VRA, modifications may be needed such as re-positioning or changing types of reinforcers. Dimming of room light may make the reinforcers more effective in some cases while tangible rewards may be more effective for others.

For a child who cannot be conditioned to respond, has limited movement towards a visual reinforcer, or has severe visual problems, Distraction Testing is the most appropriate test. When responses to acoustically specific stimuli cannot be obtained, one might resort to broadband noise-makers and sounds which carers know arouse the child, but responses to these do not ensure auditory access to all the necessary speech frequencies required for language development. Changes in the stimulus may also be required. For example, a longer duration stimulus to help recognition may be helpful.

When testing children with complex needs, it is important to involve the carers in planning for the behavioural hearing tests and in the interpretation of the child's responses. Even though there are difficulties obtaining behavioural responses in this group, it is important to work towards testing which gives repeatable responses and not to rely on subtle behavioural changes.

AUDITORY DISCRIMINATION OF SPEECH

The testing of auditory discrimination of speech has high face validity in the assessment of young children. Although speech discrimination is not a frequency-specific function, it illustrates a prime function of hearing in the developing young child and can correlate highly with hearing sensitivity in normally hearing and mildly or moderately hearing-impaired children. Increasingly, there has been an interest in testing children's hearing of speech since the use of cochlear implants in young children[24]. As for severely and profoundly hearing-impaired, conventionally aided children, threshold testing is a poor predictor of a child's ability to discriminate speech. Similarly, since the introduction of compression hearing aids, speech discrimination scores rather than aided thresholds have become more useful. Marriage and Moore[25] demonstrated the benefit of fast-acting compression for consonant discrimination in a group of children with hearing aids.

Developments in the field of speech discrimination include the use of technological devices for the presentation and response mode of tests to improve signal reproducibility and facilitate

compliance appropriate to the child. In particular, spoken responses are generally avoided when assessing auditory discrimination to eliminate the confounding variability of children's articulation.

THE COOPERATIVE TEST

The Cooperative Test, first described by Ewing and Ewing,[13] is suitable for children with the normal linguistic development level of about an 18-month-old child or older. Usually, other tests requiring a greater receptive vocabulary are more appropriate to the normally linguistically developing child by the age of 2 ½ years.

In this test, the child is required to carry out instructions in response to simple verbal commands. The object is to record the minimum intensity required for the comprehension of simple verbal instructions.

The stimuli for the Cooperative Test are voiced instructions and, thus, contain a wide range of frequencies. Results of the test are not frequency-specific. Usually, three different instructions are used. For example, the instruction may be 'Give it to Mummy' or 'Give it to Daddy' or 'Give it to Teddy' or 'Put it in the box'.

A peg or similar small toy is handed to the child and the instruction is given. It is often important for the tester to hold on to the peg until the command has been given, otherwise the child may pre-empt the command and deposit the peg elsewhere. Further pegs and instructions are given at conversational voice levels, in random order and initially in front of the child to ascertain his or her comprehension and cooperation in the procedure.

If the child fails to respond, the tester should ensure that the child is watching the tester's face and help the child carry out the commands in the first few instances. If the child seems uncertain, it may also be appropriate to raise the intensity of the voice slightly to about 60 dB (A). Many children with temporary conductive losses can do this test if the commands are given at a sufficient intensity level.

For a child who cooperates, the test should proceed by preventing lip-reading by covering the tester's mouth, or going behind the child and reducing the intensity of commands until the child fails to respond and then increasing them until he or she responds again. The lowest intensity at which the child consistently discriminates the commands correctly without visual cues is then measured. For a child with normal hearing and speech discrimination ability, the threshold for the Cooperative Test will be at 35–40 dB (A). Failure to discriminate without lip-reading or a tendency to look for visual information should be noted as indicators of hearing loss.

For some children who, owing to shyness, inhibit their responses, other methods within the child's limited receptive vocabulary may be necessary, such as in McCormick's Four Toy Eye Pointing Test.

The Toy Discrimination Test (McCormick, 1977)[26]

By the age of 2 ½ years, the receptive vocabulary of children has grown to include many nouns which can be represented in toy form and used in a test involving discrimination of pairs of similar sounding words such as /tree/ and /key/. The Toy Test uses seven such pairs of words so that consonant discrimination of young children can be tested. Administration of the test, using live voice, has been detailed by McCormick.[26] Presentation of a digitised recording of

the speech stimuli via a loudspeaker can also be used in the IHR-McCormick Automated Toy Discrimination Test and more recently in the Phoenix version of the test whereby predictions of average better ear thresholds can be made from the results of this test.[27] This version of the test uses an algorithm to vary the intensity of presentation according to the child's responses. A portable digital version of the test, the 'Parrot', allows the tester to choose the presentation level.[28] A derived version of this test for children with limited knowledge of English, the E2L test, is also available in this format.[29] The Phoenix version of the test also has a speech-in-noise format of the test, though published normative data for this are not yet available. For children with less developed language or those who inhibit their finger-pointing response, the McCormick Four Toy Eye Pointing Test may be utilised.

McCORMICK'S FOUR TOY EYE POINTING TEST (1988)

In this test, two pairs of the McCormick Toy Discrimination Test items are spaced apart on a low table in front of the child who is asked where the items are, in random order, initially at conversational levels of intensity. Observation of the child's eye movements by the tester is made and the intensity of the voice reduced, visual cues are obliterated by covering the tester's mouth, until threshold levels of 80% correct discrimination are achieved. Details of this test, to ensure avoidance of pitfalls, are given by McCormick.[30]

The Manchester Picture Vocabulary Test

This test, developed by Watson[31] using language found to be familiar to 5-year-olds, involves a pointing response to pictures consisting of vowel and consonant confusion matrices. This test has had several updated versions,[32] the latest being in a recorded digitised format.

The Consonant Confusion Test (CCT) and Auditory Performance Task 1 (APT1)

These are two monosyllable, consonant confusion tests with picture matrices and the target words recorded on CD along with optional speech-shaped noise on the second recording channel. The difficulty of the CCT is lower than that of the APT1 so that the appropriate test may be selected dependent on the discrimination ability of the child; these have been used to demonstrate the benefits of wide-dynamic, fast-acting compression hearing aids.[25]

The Three-Interval, Forced Choice Test of speech pattern contrast perception (the THRIFT test)

For children with cognitive skills of 8 years of age or more, Boothroyd's THRIFT test gives a detailed examination of discrimination ability.[32] This tests abilities such as auditory discrimination of place of articulation, intonation pattern and consonant voicing, which is useful in the assessment of hearing-impaired children. The test is computer-controlled, with the option of a visual presentation of the speaker and with a touch screen for the child to select his or her response. Age-related normative data for this test have been published.[33]

Such technological devices enable controlled replication of the speech stimulus at known intensities and thereby increase the reliability of the test. Use of pre-recorded or computer-

generated material can also be presented using headphones or insert earphones to give ear-specific information.[34]

CONCLUSION

Hearing assessment, using the methods described in this chapter, can be used to determine frequency-specific hearing thresholds and speech discrimination ability of young children by observation of the child's behaviour in response to sound. Such behaviour is observed within a structured clinical context and in response to defined acoustic stimuli from which a child's auditory function for the real world might be predicted.

With the introduction of newborn hearing screening, the demands for behavioural testing have become more challenging. There is still the need to identify those children with transient hearing losses related to middle-ear effusion and those children identified through the newborn screen who need to be monitored and managed. In addition, the behavioural testing adopted in all clinics needs to identify those children with mild hearing losses or sloping hearing losses who are likely to have passed the newborn hearing screen. Furthermore, whilst carrying out behavioural testing, the clinician should not be blinded to an expected result based on the results of any newborn screen because there remain the possibilities of late-onset or progressive hearing losses and the possibility of false negatives on the screen.

Thus, the development of newborn screening places demands on the behavioural assessment to use frequency-specific stimuli and obtain reliable ear-specific information. This allows the clinician to test children using criteria of normality of hearing as close as possible to criteria used in adult assessment.

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4 Neuro-diagnostic paediatric audiology

J.W. Hall III and L.M. Bondurant

INTRODUCTION

Historical overview

Audiology as a profession, and formal audiological assessment as a clinical service, dates back only to the middle of the twentieth century. About 60 years later, however, the profession and the techniques available to evaluate auditory function have advanced remarkably. Our knowledge of the auditory system – from the cochlea to the cortex – has also expanded exponentially, especially within the past 20 years. Application of computer-based technology for assessment, coupled with the newly identified principles of auditory neuroscience, are perhaps most obvious in neuro-diagnostic paediatric audiology, that is, the thorough description of type, configuration and degree of hearing impairment in children from the perinatal period onwards. Our audiological armamentarium extends far beyond the simple audiogram. Hearing screening of newborn infants within hours after birth is rapidly becoming the universal standard. The application of two techniques – otoacoustic emissions (OAEs) and auditory brainstem response (ABR) – in combination for newborn hearing screening yields multiple important benefits including refer rates less than 2%, information on the site of auditory dysfunction for screening 'failures', and earlier identification of communicatively significant hearing loss. With a test battery of electro-acoustic and electrophysiological measures, it is possible to diagnose auditory dysfunction during infancy and to differentiate amongst middle ear, cochlear, neural and central auditory system sites of lesions. Fortunately, paediatric audiology is by no means limited to the diagnosis of peripheral hearing impairment, as we really hear with our brain as much as with our ears. Behavioural and electrophysiological measures are available for evaluating central auditory pathways and processes. Indeed, there is unprecedented research and clinical interest in the assessment of central auditory nervous system function with sophisticated techniques and technology, such as the mismatch negativity (MMN) response and functional magnetic resonance imaging (fMRI). In this chapter, we provide a summary of exciting trends in neuro-diagnostic paediatric audiology.

AN UPDATED CROSS-CHECK PRINCIPLE

Over 30 years ago, Jerger and Hayes[1] (1976) defined fundamental clinical guidelines for audiological assessment of children – 'the cross-check principle'. In an era when behavioral audiometry was relied on almost exclusively in paediatric hearing assessment, the authors presented a compelling argument for the evaluation of children with a test battery consisting

of behavioral audiometry, aural impedance measurements and the ABR. Jerger and Hayes[1] (1976) supported their argument with ample clinical evidence. The cross-check principle is now updated and expanded with the inclusion of such techniques as frequency-specific ABRs elicited with tone-burst stimuli, the auditory steady state response (ASSR) and, of course, OAEs. As noted in Table 4.1, OAEs have earned a unique and valued complementary clinical role in the 'standard-of-care' paediatric audiologic test battery.

DIAGNOSTIC PAEDIATRIC AUDIOLOGY IN THE ERA OF UNIVERSAL NEWBORN HEARING SCREENING (UNHS)

Introduction

The importance of hearing integrity within the first 3 to 4 years after birth for normal acquisition of speech and language has long been appreciated (see, e.g. Lenneberg et al.[2]). During this sensitive period, speech and language will almost always develop rapidly and normally if the auditory and language regions of the brain are adequately stimulated by sound and, especially, the sounds of communication. Unfortunately, by the time hearing impairment in infancy and early childhood is suspected, audiologically evaluated and appropriately managed, two or more of these communicatively important years have elapsed and the child has lost an enormous developmental advantage.

The trend towards UNHS has led predictably to a demand for paediatric diagnostic audiological assessment of infants within months after birth. Infants who do not pass the hearing screening at birth must, within 1 or 2 months, undergo diagnostic audiological testing to confirm and to define hearing impairment so that intervention can be initiated no later than 6 months. Yoshinaga-Itano and colleagues[3] at the University of Colorado have provided definitive evidence of the benefits of early intervention on language abilities of children with hearing impairment. An important message taken from this investigation is the definition of 'early' intervention for hearing impairment, namely, the intervention for hearing impairment by 6 months of age. Secondary to the marked positive influence of early intervention on language acquisition are clear academic, cognitive, social and economic benefits.[4] Early intervention is entirely dependent, however, on prompt and accurate definition of hearing impairment as soon as possible after birth. A detailed discussion of neonatal hearing screening is presented in Chapter 2.

The rationale for early identification of and intervention for hearing impairment in infants, of course, is to optimise language and communication development. During the 1960s, an international collection of papers revealed increasing interest in hearing screening of young children.[5–7] At the time, however, screening was conducted with behavioural techniques that lacked adequate sensitivity and specificity. Early identification of infant hearing impairment was altered dramatically by the discovery of the ABR by Jewett and Williston[8] and a subsequent paper by Hecox and Galambos[9] describing the clinical application of ABR in auditory assessment of infants and young children. As clinical experience accumulated and screening equipment, techniques and strategies were modified, test performance improved steadily and automated techniques were introduced.[10]

OAEs were discovered by Kemp in 1978[11] and, within several years, were used in newborn hearing screening (see Hall[12] for review). Experience with OAEs in newborn hearing screening has led to major modifications in equipment design and, more recently, lower failure rates.

Table 4.1 Summary of diagnostic audiological techniques and strategies for children as a function of age (ages are approximate). Techniques are arranged with the most important first.

Birth to 4 Months

Auditory brainstem response (ABR)
- Latency-intensity functions for click stimuli
- Analysis of inter-wave latencies (cochlear vs. retrocochlear status)
- Threshold estimation for frequency-specific stimuli (tone bursts)
- Threshold estimation for bone-conduction stimuli (as indicated)

Otoacoustic emissions (OAEs)
- Distortion product OAEs or transient OAEs
- Verify cochlear hearing impairment
- Rule out auditory neuropathy

Aural immittance measurement
- Tympanometry to assess middle-ear status
- Acoustic reflexes (pure-tone vs. broad-band noise thresholds) to estimate hearing impairment

Behavioural audiometry (if feasible)
- Behavioural observation audiometry (BOA) in the sound-field (not ear-specific)
- Evaluate responses to pure tone and speech signals

5 to 24 Months

Behavioural audiometry
- Visual reinforcement audiometry (VRA)
- Evaluate ear-specific responses (with earphones)
- Estimate pure-tone thresholds for speech frequencies
- Estimate speech reception thresholds

Otoacoustic emissions (OAEs)
- Distortion product OAEs or transient OAEs
- Verify cochlear hearing impairment
- Rule out auditory neuropathy

Aural immittance measurement
- Tympanometry to assess middle-ear status
- Acoustic reflexes

Auditory brainstem response (ABR)
- Essential if behavioural audiometry findings are inconsistent, incomplete or inconclusive
- Latency-intensity functions for click stimuli
- Analysis of inter-wave latencies (cochlear vs. retrocochlear status)
- Threshold estimation for frequency-specific stimuli (tone bursts)
- Threshold estimation for bone-conduction stimuli (as indicated)
- Sedation usually required

24 to 48 Months

Behavioural audiometry
- Visual reinforcement audiometry (VRA), tangible reinforcement audiometry (TROCA), visual reinforcement conditioned audiometry (VROCA), or conditioned play audiometry
- Evaluate ear-specific responses (with earphones)
- Estimate pure-tone thresholds for speech frequencies
- Estimate speech reception thresholds
- Measure word recognition scores (e.g. speech discrimination)

Otoacoustic emissions (OAEs)
- Distortion product OAEs or transient OAEs
- Verify cochlear hearing impairment
- Rule out auditory neuropathy

Aural immittance measurement
- Tympanometry to assess middle-ear status
- Acoustic reflexes to confirm hearing impairment

Auditory brainstem response (ABR)
- Only if behavioural audiometry findings are inconsistent, incomplete or inconclusive
- Latency-intensity functions for click stimuli
- Analysis of inter-wave latencies (cochlear vs. retrocochlear status)
- Threshold estimation for frequency-specific stimuli (tone bursts)
- Threshold estimation for bone-conduction stimuli (as indicated)
- Sedation required

Both OAEs and AABR techniques are now endorsed for newborn hearing screening.[13–15] Indeed, the application of both OAEs and AABR in combination, a recent development in newborn hearing screening, results in lower refer rates, differentiation amongst types of hearing impairment even in the neonatal period, lower overall costs associated with hearing screening and follow-up assessments and, most important, earlier and more precise intervention strategies.[16]

AUDITORY ELECTRO-ACOUSTICAL MEASURES

Aural immittance measures

Aural immittance (impedance) measures are an important part of the basic paediatric audiometric test battery. Immittance measures are, in fact, standard-of-care for audiological assessment of infants and young children, as defined by the Joint Committee on Infant Hearing[13] (see Table 4.1). Immittance is a term derived from the terms for two related techniques for assessing middle-ear function (*im*pedance and ad*mittance*) – techniques that have been applied clinically since 1970. Briefly, the external ear canal is sealed with a soft rubber probe tip. The probe tip is connected to a device that produces a tone, which is delivered towards the eardrum. Middle-ear impedance or admittance is calculated from the intensity, and other physical properties (such as phase) of the tone in the ear canal. A middle ear (tympanic membrane and ossicular system) with low impedance (higher admittance) more readily accepts the acoustic energy of the probe tone, whereas a middle ear with abnormally high impedance (lower admittance) due, for example, to fluid within the middle-ear space tends to reject energy flow. Thus, impedance (admittance) characteristics of the middle-ear system can be inferred objectively with this technique and related to well-known patterns of findings for various types of middle-ear pathology.

Tympanometry is the dynamic recording of middle-ear impedance as air pressure in the ear canal is systematically increased or decreased. The technique is a sensitive measure of tympanic membrane integrity and middle-ear function. Compliance (the reciprocal of stiffness) of the middle ear, the dominant component of immittance, is the vertical dimension of a tympanogram. Tympanometry is very popular clinically, in part because it requires little technical skill and only several seconds of time; it is an objective (as opposed to behavioural) method that does not depend on the cooperation of the patient and is a very sensitive index of middle-ear function. Tympanometric patterns, in combination with audiogram patterns, permit differentiation amongst and classification of middle-ear disorders. The most clinically widespread approach for describing tympanograms was first reported in 1970 by James Jerger.[17]

The stapedius muscle within the middle ear is the smallest muscle in the body. Measurement of contractions of the middle-ear stapedius muscle to high sound intensity levels (usually 80 dB or greater) is the basis of the acoustic reflex. Acoustic reflex measurement is clinically useful for estimating hearing sensitivity and for differentiating amongst sites of auditory disorders, including the middle ear, inner ear, eighth cranial nerve and auditory brainstem. The afferent portion of the acoustic reflex arc is the eighth cranial nerve. There are complex brainstem pathways leading from the cochlear nucleus on the stimulated side to the region of the motor nucleus of the seventh (facial) nerve on both sides (ipsilateral and contralateral to the stimulus) of the brainstem. The efferent portion of the arc is the seventh cranial nerve, which innervates the stapedius muscle. The muscle then contracts, causing increased stiffness

(decreased compliance) of the middle-ear system. The small change in compliance that follows stapedius muscle contraction within 10 ms is detected by the probe and immittance device, much as compliance changes are detected during tympanometry. Acoustic reflex measurement is very useful clinically because it can quickly provide objective information on the status of the auditory system from the middle ear to the brainstem. Distinctive acoustic reflex patterns for ipsilateral and contralateral stimulation and measurement conditions characterise middle ear, cochlea, eighth nerve, brainstem and even facial nerve dysfunction.

Although the essentials of aural immittance measurement remain constant for most paediatric applications, different strategies should be followed in selected populations to achieve specific clinical objectives. For example, tympanometry measurement in infants younger than 6 months old should be performed with a higher frequency (e.g. 1,000 Hz) probe tone than the 226 Hz typically employed clinically. Due to anatomical characteristics of the infant ear canal, principally a highly compliant cartilaginous ear canal wall, systematic change in ear canal pressure during tympanometry may produce a corresponding apparent change in compliance mimicking changes in middle-ear compliance. The practical consequence of this phenomenon is the possibility that infants with abnormal middle-ear function, e.g. restricted mobility secondary to otitis media as reflected by a type B (flat) tympanogram, may, instead, yield a normally appearing type A tympanogram. The end result is a false-negative immittance finding – the suggestion of normal middle-ear function in an infant with, in fact, significant middle-ear dysfunction.

The specific approach taken for measurement of the acoustic reflex may also vary within some patient groups, depending on clinical goals. With infants under the age of 6 months, the high-frequency probe tone noted above should be used also in acoustic reflex measurement. Depending on how it is performed, acoustic reflex measurement can be applied in estimation of hearing sensitivity or as an objective neuro-diagnostic index of retrocochlear and central auditory functioning (at the brainstem level), as well as facial (7th cranial nerve) status.

In the early 1970s, when acoustic reflex measurement was introduced as a clinical test procedure, papers appeared describing estimation of auditory sensitivity by comparison of acoustic reflex thresholds for pure-tone versus noise signals.[18,19] The most popular of such approaches was the SPAR (Sensitivity Prediction by Acoustic Reflex). Later clinical investigation in a large series of children with varying degrees of hearing loss[20] showed that analyses of acoustic reflex thresholds for a broadband noise (BBN) signal alone provided a quick and reasonable accurate technique for identification and estimation of the degree of sensory hearing loss. Specifically, in normal hearing persons the acoustic reflex threshold for a BBN signal was typically recorded at intensity levels of less than 80 dB HL and often for levels as low as 65 to 70 dB HL. Then, the acoustic reflex threshold systematically increased directly with increased sensory hearing loss. As a rule of thumb, acoustic reflex thresholds for BBN signals of less than 85 dB HL were associated with generally normal hearing sensitivity, whereas acoustic reflex thresholds for BBN signals of greater than 90 dB HL almost always reflected a communicatively important sensory hearing loss (greater than 30 dB HL). In young children with limited cooperation and who must be assessed quickly, acoustic reflexes elicited by a BBN stimulus alone can be useful for objective estimation of auditory thresholds[20] using the guidelines summarised above.

Finally, comparison of acoustic reflex thresholds amongst four different measurement conditions – right and left ear stimulation and ipsilateral and contralateral stimulation – offers a quick and objective means of differentiating amongst sites of lesions affecting the peripheral auditory system, portions of the auditory brainstem and the facial nerve.[21,22] In the assessment

of children for auditory processing disorders (APD), measurement of acoustic reflex thresholds for each ear in the ipsilateral versus contralateral conditions is a clinically feasible initial step in the identification of central auditory dysfunction, even for very young or difficult-to-test children.[22]

Aural immittance measures in combination with other electro-acoustical, electrophysiological and behavioural auditory procedures contribute to the confident differentiation amongst a wide variety of disorders affecting the auditory system. It is appropriate to note at this juncture the recent emergence of more sophisticated techniques for objective measurement of middle-ear mechanical status and acoustic reflex functions. Keefe and colleagues[23–25] and Feeney and Sanford[26] describe clinical investigation of what is referred to as a 'wideband acoustic transfer function (WATF) system' to measure middle-ear function and acoustic reflexes in children, including newborn infants, and adults. In part, the approach involves the use of a broadband (click) probe sound, rather than a conventional 226 or 1,000 Hz probe tone. The system, which is based on detection of either admittance magnitude or energy reflectance, appears to result in acoustic reflex thresholds lower than expected for typical aural immittance measurements. The WATF system also helps to explain curious DPOAE findings in infancy, e.g. amplitudes larger in neonates than adults, and may contribute in other ways to the analysis and interpretation of clinically recorded OAEs.

Otoacoustic emissions

OAES are low-intensity sounds produced by the cochlea in response to an acoustic stimulus (see Hall[12] for review). A moderate intensity click, or an appropriate combination of two tones, can evoke outer hair cell movement or motility. Outer hair cell motility affects basilar membrane biomechanics, resulting in a form of intra-cochlear energy amplification, as well as cochlear tuning for more precise frequency resolution. The outer hair cell motility generates mechanical energy within the cochlea that is propagated outwards, via the middle-ear system and the tympanic membrane, to the ear canal. Vibration of the tympanic membrane then produces an acoustic signal (the OAE), which a sensitive microphone can measure. According to conventional taxonomy, there are two broad classes of OAEs: spontaneous and evoked. Spontaneous otoacoustic emissions (SOAEs), present in only about 70% of persons with normal hearing, are measured in the external ear canal when there is no external sound stimulation. A significant gender effect characterises SOAEs, with females demonstrating SOAEs at twice the rate of males. Evoked OAEs, elicited by moderate levels (50 to 80 dB SPL) of acoustic stimulation in the external ear canal, are generally classified according to characteristics of the stimuli used to elicit them or characteristics of the cochlear events that generate them.

Distortion-product otoacoustic emissions (DPOAEs) are produced when two pure-tone stimuli at frequencies f1 and f2 are presented to the ear simultaneously. The most robust DPOAEs occur at the frequency determined by the equation 2f1-f2, whereas the actual cochlear frequency region that is assessed with DPOAE is between these two frequencies, and probably close to the f2 stimulus for recommended test protocols.[12] Transient-evoked otoacoustic emissions (TEOAEs) are elicited by brief acoustic stimuli such as clicks or tone bursts. Although there are distinct differences in the methodology for recording DPOAEs versus TEOAEs, and the exact cochlear mechanisms responsible for their generation are also different, each type of evoked OAE is now being incorporated into routine auditory assessment of children and adults, including newborn hearing screening.[12] As with ABR, devices permitting automated OAE measurement and analysis, and designed primarily for newborn hearing screening, are now

available from a variety of manufacturers. Most of these devices are hand-held and very simple to operate. A recent report of accumulated experience with OAE screening of over 50,000 babies confirmed a failure rate of approximately 10%, with a final false-positive failure rate of less than 2%.[27] The use of OAEs in paediatric diagnostic assessment is, perhaps, more valuable and more powerful than any other application in children or adults.

Why are OAEs a necessary component of the modern paediatric test battery? The answer is their remarkable sensitivity and specificity. OAEs are the product of the highly metabolic activity of outer hair cells. Virtually any insult to the cochlea, including even subtle disruptions in blood flow to the stria vascularis, will be reflected by OAE changes. There is no more sensitive measure of cochlear function. OAEs are almost entirely sensory and 'pre-neural'. Their measurement does not depend on the functional status of any synapses, nor on the rest of the auditory system, i.e. retro-cochlear pathways. This site specificity is a distinct clinical advantage for a component of a diagnostic test battery. In addition to these two essential features – sensitivity and specificity – OAEs are electro-acoustical, requiring no behavioural response from the paediatric patient. These fundamental features of OAEs take on very practical everyday importance in paediatric audiology. Most audiological management of children is predicated on the premise that the hearing impairment is sensory, affecting the cochlea. By definition, audiologists are responsible for evaluation and diagnosis of all types of auditory impairment. Conductive hearing impairment, however, is traditionally treated medically or surgically by physicians. Although the audiologist is integrally involved in the detection of eighth nerve (retrocochlear) and central auditory nervous system dysfunction, treatment (if available) is most often a team effort, which may or may not include the audiologist. Determining whether the hearing impairment is sensory or neural (or some combination) depends very much on results of OAE measurement. If the hearing impairment is sensory, then the audiologist is the professional with primary responsibility for implementing and coordinating management with amplification and a complement of habilitation or rehabilitation strategies and techniques. For auditory assessment of children, the measurement of OAEs is now considered standard-of-care. This serious clinical conclusion is amply supported by diverse OAE applications that are reviewed in this chapter. In short, OAEs are not simply a handy or convenient procedure for assessing auditory function but, rather, an essential component of the paediatric test battery. They can play a pivotal and critical role in decisions regarding audiological or medical management of auditory impairment. The clearest example of this role is in patients with suspected 'auditory neuropathy'.[12]

AUDITORY ELECTROPHYSIOLOGICAL MEASURES

Auditory brainstem response (ABR)

Auditory-evoked responses are electrophysiological recordings of responses to sounds. With proper test protocols, the responses can be recorded clinically from activation of all levels of the auditory system, from the cochlea to the cortex (see Hall[28] for review) (Table 4.2). Amongst these responses, the ABR (often referred by neurologists as the brainstem auditory evoked response, or BAER) is applied most often clinically. The ABR is generated with transient acoustic stimuli (clicks or tone bursts) and detected with surface electrodes (discs) placed on the forehead and near the ears (earlobe or within external ear canal). Using a commercially available, computer-based device, it is possible to present rapidly (e.g. at rates of 20 to 30 per

Table 4.2 A protocol for measurement of frequency-specific auditory brainstem response (ABR). The conventional ABR protocol for air conduction click signals must be modified to successfully record ABRs for tone burst signals. The main differences between protocols for click versus tone burst ABRs are noted under comments.

Parameters	Suggestions	Comments
Stimulus Parameters		
Transducer	Insert	Insert earphones offer many advantages in clinical ABR measurement, especially with infants and young children.
Polarity	Alternating	Instead of the usual rarefaction polarity, alternating polarity stimuli can be used to minimise the possibility of a frequency-following type response.
Ramping	Blackman	Ramping refers to how the rise/fall portions of the tone burst are shaped. Some non-linear ramping or windowing techniques reduce spectral splatter and increase frequency specificity of tone burst stimulation. Blackman windowing is the best, and most current AER systems include it in their stimulus package.
Duration	Variable	The rise/fall, and plateau, times for the tone burst stimuli vary depending on the frequency. As a rule, it is desirable to use longer times for lower frequencies so as to include more cycles to increase the chance that the stimulus sounds like the desired frequency, and not a click. However, as discussed in this section of the chapter, the use of a very brief (0.5 cycles or 2 ms) 250 Hz tone burst will generate a more well formed and distinct ABR, albeit not quite as frequency-specific (an energy band within the frequency range of 100 to 600 Hz). The most common approach for signal duration is to use 2 cycles rise time, 0 cycle plateau, and 2 cycles fall time or, in milliseconds (ms): 500 Hz: 4 ms rise/fall and 0 ms plateau 1,000 Hz: 2 ms rise/fall and 0 ms plateau 2,000 Hz: 1 ms rise/fall and 0 ms plateau 4,000 Hz: 0.5 ms rise/fall and 0 ms plateau
Intensity	Variable	Keep in mind that the intensity levels on the screen for your ABR system will usually not be defined in dB nHL, as they are for a click. More often, the values are in dB SPL. That is, 95 dB may be selected, but the intensity range for the tone burst frequency may go as high as 115 dB. Always obtain behavioural threshold data for each tone burst stimulus to be used for ABR recording (with the earphones specific to the evoked response system and in the room where ABRs will be recorded), and then develop a biologic normative data for tone burst intensity. For example, if the maximum dial setting for a 500 Hz tone burst is 115 dB, but normal subjects have an average threshold of 30 dB for this stimulus, then at 115 dB on the dial the intensity level is really 85 dB nHL (referenced to the normal behavioural threshold for the stimulus). With most evoked response systems, these 'correction factors' can be incorporated into the intensities displayed on the screen so that all intensity values are in dB nHL according to clinic normative data. It is then advisable to actually record ABRs for this 500 Hz stimulus from a few of these normal hearing subjects to estimate the lowest intensity level that produces an observable and reliable ABR wave V.

Table 4.2 *Continued*

Parameters	Suggestions	Comments
Acquisition Parameters		
Electrode sites	Fz–Ai	Non-inverting (positive) electrode is located in the midline on the high forehead (Fz) and the inverting electrode is located on the earlobe ipsilateral to the stimulus ear (Ai). With an ear clip electrode design, the earlobe electrode is easily applied, impedance is low, and the electrode is removed from the mastoid region. The earlobe electrode records a larger wave I than the mastoid electrode, and is associated with less stimulus artefact in bone-conduction ABR recordings. The ground electrode can be located on the low forehead (Fpz) or the contralateral earlobe (limits recordings to a single channel).
Filter settings	30 to 3,000 Hz	A low-frequency cut-off for the high pass filter (e.g. 30 Hz) is very important because the tone burst ABR is dominated by low-frequency energy, especially in infants.
Analysis time	15 to 20 ms	For click signals and higher frequency tone burst signals, an analysis time of 15 ms is adequate to encompass the wave V component even under conditions associated with delayed wave V latency, e.g. low signal intensity level, hearing loss, very young age (immaturity of the auditory pathways). For tone burst signals of 1,000 Hz and below, a 20 ms analysis time is recommended.
Sweeps	Variable	The number of sweeps (stimulus repetitions or number of signal averages for an ABR recording) is dependent on the signal-to-noise ratio. When the signal (ABR amplitude) is larger (e.g. at a high intensity level with a normal hearing patient) and/or when background noise is low (e.g. the patient is sedated or anaesthetised), then relatively fewer stimulus repetitions are needed. On the other hand, when ABR amplitude is smaller (e.g. at lower signal intensity levels and/or in a patient with hearing loss) and noise is greater (a restless un-sedated child), more signal averaging (more stimulus repetitions) will be needed. As a rule, fewer stimulus repetitions are required for the second (replication) ABR run when the goal is to simply verify that the response is reliable (and not just artefact).

second) thousands of sound stimuli and to average reliable ABR waveforms in a matter of minutes. Extensive research confirms that the ABR wave components arise from the eighth cranial nerve and auditory regions in the caudal and rostral brainstem. Wave I unquestionably represents the synchronously stimulated compound action potentials from the distal (cochlear end) of the eighth cranial nerve. Wave II may also arise from the eighth nerve, but near the brainstem (the proximal end). Waves I and II are generated by structures ipsilateral to the ear stimulated. All later ABR waves have multiple generators within the auditory brainstem. Wave III, which is usually prominent, is generated within the caudal pons, with likely contributions from the cochlear nuclei, the trapezoid body and the superior olivary complex.[28] The most prominent and rostral component of the ABR – wave V – is thought to arise in the region of the lateral lemniscus as it approaches the inferior colliculus, probably on the side contralateral to the ear stimulated.

In ABR waveform analysis, the first objective is to assure that the response is reliably recorded. Minimally, two replicated waveforms should be averaged until the presence of an

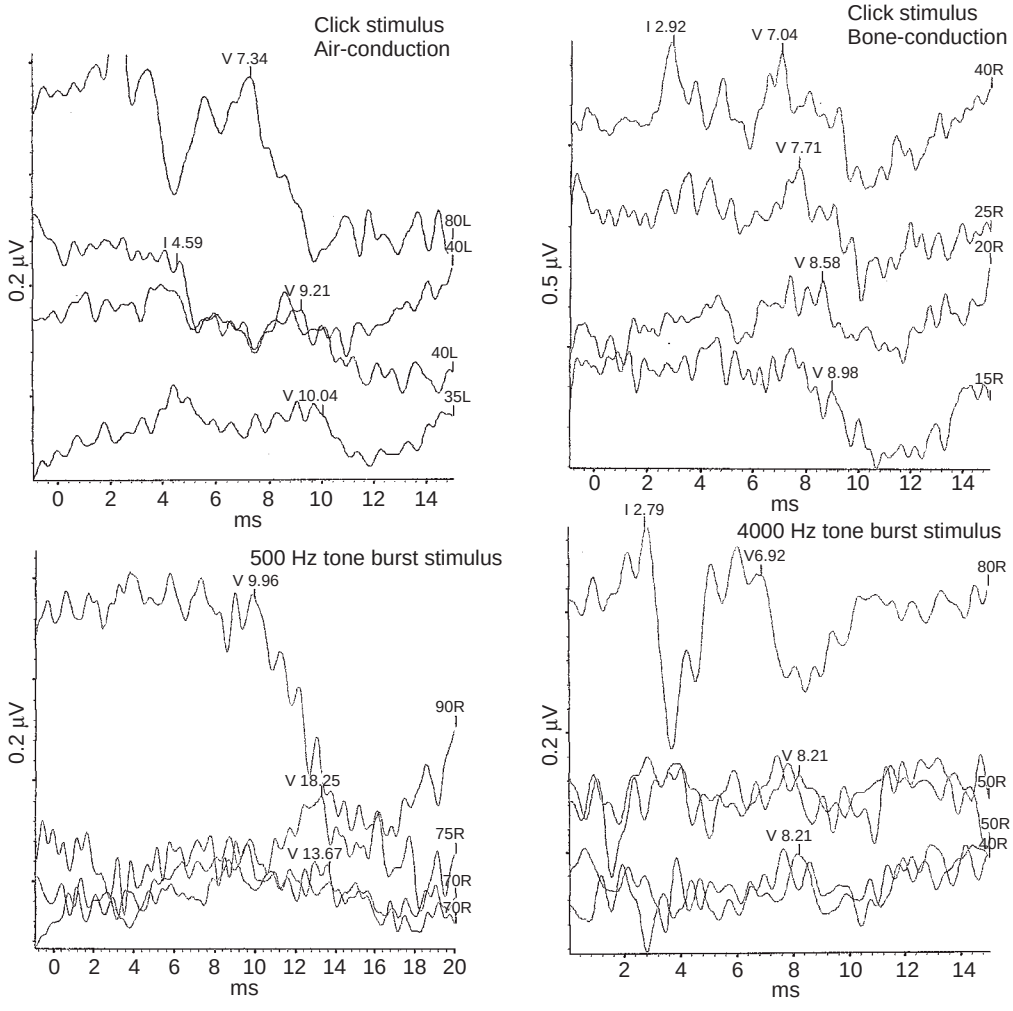

Figure 4.1 Example of auditory brainstem response (ABR) waveforms elicited with click and tone-burst stimulation delivered via air conduction, and click stimuli delivered via bone conduction.

ABR is confirmed. To permit maximum use of test time, ABR averaging is not always repeated for each intensity level, particularly if the waveform morphology is good and the latency for wave V occurs within an expected latency region, i.e. slightly longer than for the next highest intensity level. Examples of ABR waveforms for click and tone burst air conduction stimulation, and for bone-conduction click stimulation, at progressively lower intensity levels are illustrated in Figure 4.1.

If the response is not highly replicable, however, modifications in the test protocol must be made, and potential technical problems must be considered and systematically ruled out. Remember, 'when the ABR does not repeat, the assessment is not complete'. When a replicable response is confirmed, absolute latencies for each replicable wave component, and relative (inter-wave) latencies between components, are calculated in milliseconds, and usually com-

pared with appropriate normative data. Stimulus intensity is then decreased in an attempt to determine the lowest intensity level that still produces a reliable ABR wave V component. ABR minimum response level is then used to estimate behavioural auditory threshold for click and tone burst stimuli.

When applying the ABR in newborn hearing screening, waveform analysis is typically limited to the identification of a reliable wave V component within the expected latency region for a single stimulus intensity level (e.g. 35 dB nHL). There are now automated ABR (AABR) devices on the market designed specifically for newborn hearing screening by non-professional testers.[28] With automated ABR devices, stimulus presentation and response analysis is under the control of computer-based algorithms and statistical criteria. Data for one automated ABR system (the ALGO-2 device from Natus, Inc.) confirmed failure rates as low as 2% in a healthy baby population and only 4% for an intensive care nursery infant population.[29]

One of the newest, and exciting, advances in clinical ABR application in children is the elicitation of the response with speech stimuli. Kraus and colleagues concluded from clever clinical investigations in children with psycho-acoustical evidence of temporal processing deficits that 'speech-evoked brainstem responses are a biological marker for auditory temporal processing ability'.[30]

Auditory steady state response (ASSR)

The ASSR is an electrophysiological technique that is very useful for estimation of auditory thresholds in infants and young children.[28,31] The ASSR is elicited with pure-tone (steady-state) signals that are rapidly modulated or changed in amplitude and, usually, frequency. Fast stimulus modulation rates (e.g. >80 Hz) are utilised when the ASSR is recorded from children who are sedated or lightly anaesthetised. As with ABR measurement, sedation or anaesthesia is necessary in recording the ASSR from children to eliminate the deleterious effects on response detection of muscle or movement-related measurement artefact. ASSR measurement is now possible with commercially available evoked response systems used also for ABR assessment.

The ASSR offers three potential advantages over the ABR for auditory assessment of young children. First, because the response is elicited with pure-tone (steady-state), rather than transient (very brief) stimuli, it is possible to present stimuli with intensity levels up to 120 to 125 dB HL. Second, depending on the extent of the modulation, especially the frequency modulation, the stimuli used to elicit the ASSR can be rather frequency-specific, and valuable for electrophysiological estimation of the audiogram. Finally, the analysis of either the phase and/or the frequency content of brain activity elicited by the modulated pure-tone signals is fully automated, and independent of the skills and experience of the tester. Clinical experience confirms that the ASSR complements the ABR in the electrophysiological assessment of auditory function of infants, in particular the estimation of auditory thresholds for those with severe and profound hearing loss. Application of the ASSR in hearing assessment of infants and young children in isolation is not advised. Without valuable information provided by the ABR, and other audiological procedures appropriate for infant hearing assessment, e.g. aural immittance measures, it is not possible to differentiate confidently amongst very diverse types of auditory dysfunction, such as conductive hearing loss, sensory hearing loss, auditory neuropathy and brainstem auditory abnormalities. However, in combination, the ABR and the ASSR offer a powerful diagnostic duo for early assessment of hearing in children in the era of UNHS.

Electrocochleography (ECochG)

Since the 1960s, electrocochleography (ECochG) has been applied in the assessment of peripheral auditory function in paediatric populations, in addition to intra-operative monitoring of cochlear and eighth nerve status and in the diagnosis of Ménière's disease. Over the past three decades, stimulus and acquisition parameters for recording ECochG have undergone considerable refinement.[28] Optimal ECochG waveforms are recorded from a small needle electrode placed through the tympanic membrane onto the promontory, although tympanic membrane and, to a lesser extent, ear canal electrode locations are also clinically useful. The three major components of the ECochG are the cochlear microphonic (CM), the summating potential (SP) and the action potential (AP). The CM and SP reflect cochlear bioelectric activity, whereas the AP is generated by synchronous firing of distal afferent eighth nerve fibres, and is equivalent to ABR wave I. For this reason, the application of ECochG techniques and principles in paediatric audiological assessment has reinforced their value in diagnosis of infants with suspected 'auditory neuropathy'.

CORTICAL AUDITORY EVOKED RESPONSES

More than a dozen subtypes of auditory evoked responses can be recorded beyond the brainstem, from auditory regions of the thalamus, hippocampus, internal capsule and cortex. Prominent amongst them in clinical audiology are the auditory middle latency response (AMLR), the auditory late response (ALR), the P300 response and the mismatch negativity response.[28] In fact, cortical auditory evoked responses were reported as early as the 1930s, and, with the exception of the MMN, all of the above responses were well described before the ABR was even discovered. Cortical auditory evoked responses are characterised by longer latencies (100 to 300 ms) than ECochG and ABR because they arise from more rostral regions of the auditory CNS and are dependent on multi-synaptic pathways. Amplitudes of the cortical responses are considerably larger (two to 20 times larger) than those of the earlier responses because they reflect activity evoked from a greater number of neurons. Measurement parameters are distinctly different for the cortical versus cochlear or brainstem responses. For example, stimulus rate must be slower and physiological filter settings lower. As a rule, stimulus intensities are moderate, rather than high. Cortical evoked responses are best elicited with longer duration, and therefore frequency-specific, tonal stimuli, rather than the click stimuli that are optimal for evoking the ECochG and ABR. The analysis time must, of course, extend beyond the expected latency of the response (>300 ms) for the cortical responses. Recording electrode sites also are different for the cortical responses, with more emphasis on scalp sites over the hemispheres and less concern about electrode sites near the ears.

The AMLR consists of a prominent positive voltage (labelled Pa) component in the 25 to 30 ms region. When recorded with electrodes located over the temporal-parietal region, the AMLR is generated by pathways leading to the primary cortex and from this region of the temporal lobe. The AMLR is reasonably reliable in children, as well as adults. It is thus a good selection for electrophysiological assessment of higher-level auditory CNS function in patients at risk for or undergoing evaluation of neurological disease or dysfunction involving the thalamus or primary auditory cortex. The P300 response is recorded using what is typically referred to as the 'oddball paradigm'. Two types of stimuli are used. One – the frequent stimulus – is presented frequently in a very predictable manner. The other – the rare or deviant stimulus – is

presented infrequently and pseudo-randomly. The rare stimuli account for less than 20% of the total stimuli presented. The patient is instructed to ignore the frequent stimuli and to attend to the rare stimuli. The waveform for the frequent stimulus is essentially an auditory late response consisting of a positive peak of 5 to 10 mV within the 150 to 200 ms region. In contrast, the waveform averaged from the attended rare stimuli is characterised normally by a large positive peak in the 300 ms region, hence the term 'P300 response'. Presumed generators of the P300 response include regions of the medial temporal lobe (hippocampus) that are important in auditory attention.

One limitation of the conventional P300 response paradigm is the requirement for patient's conscious attention to the rare stimulus. This requirement may preclude measurement of the P300 response in patients for whom objective, electrophysiological information on higher-level auditory CNS function is most desired, such as infants, children with language-learning disorders, children with attention deficit disorder and brain-injured adults. However, the P3a auditory evoked response, recorded with a *passive test paradigm* and occurring with a latency of about 250 ms, is 'automatic' and not dependent on active subject attention.[32] The passive P300 response appears to be a reflection of automatic detection of a different signal, e.g. signal novelty.

Another cortical response – the MMN response – is an automatic and unconscious index of differences between acoustic stimuli. The MMN is also recorded to frequent and rare (deviant) stimuli, although the distinction between the two types of stimulus is very small. For example, if the two types of stimulus differ along the frequency domain, the P300 response might be elicited by 1,000 Hz (frequent) versus 2,000 Hz (rare) tones, whereas the MMN might be elicited by 1,000 Hz versus 1,200 Hz tones, or even speech sounds, such as /da/ versus /ga/. The MMN is thought to be generated before conscious perception by the neuronal mismatch in the brain created when the repetitive frequent stimuli are followed by an acoustically different deviant stimulus. Importantly, the MMN does not require attention to the stimuli. Rather, the patient can be sleeping or involved in some non-auditory task (such as watching a silent movie). Another clinical advantage of the MMN is the wide range of stimulus possibilities, including rather complex speech signals. Whether the MMN will someday be applied in clinical assessment of auditory CNS function remains to be seen. The MMN response, however, is already a powerful research tool for uncovering fundamental auditory processes and mechanisms in normal-hearing persons and identifying the nature of APD in clinical populations (see Hall[28] for review).

BEHAVIOURAL MEASURES: PERIPHERAL AUDITORY ASSESSMENT

Whilst electro-acoustic and electrophysiological evaluation of auditory sensitivity has proved to be a fundamental tool for achieving the goal of early detection of hearing loss, it is essential to remember that the diagnosis of hearing impairment in children does not end with the abnormal results of these procedures, e.g. aural immittance measures, OAEs, ABR and ASSR. A complete behavioural hearing evaluation is crucial for providing information about how the auditory system functions from the peripheral auditory pathway through the auditory cortex. Even in infants, accurate behavioural evaluations can yield ear- and frequency-specific information beyond that obtained by ABRs and OAEs. Description of behavioural audiometric techniques and clinical strategies is reviewed in detail in Chapter 3. There is a substantial

literature on the strategies and protocols for diagnostic paediatric assessment (e.g. Hall and Mueller[21]). However, there is no 'one-size-fits-all' set of instructions for obtaining the necessary information to diagnose and manage hearing loss in children, so in order to achieve this goal, the audiologist must be efficient, accurate and, above all, flexible. A general outline of age-appropriate approaches for paediatric audiometry was summarised in Table 4.1. Today's basic paediatric audiological test battery must include, minimally, aural immittance measures, otoacoustic emissions, pure-tone audiometry and speech audiometry. Furthermore, in the clinical assessment of auditory function of young children, these procedures are best performed in the order just listed, of course after obtaining a thorough case history.

Beginning an evaluation with aural immittance measures and otoacoustic emissions allows the clinician not only the chance to obtain a wealth of information in a very short time, but also the invaluable opportunity to observe the behaviour of the child. Armed with this information, the audiologist has a better idea of how to approach the behavioural portion of the evaluation; for example, if the child had flat tympanograms or normal tympanograms with absent OAEs or acoustic reflexes, the audiologist should consider increasing the initial level at which they present speech stimuli to obtain the speech awareness or speech recognition threshold. Doing so may save valuable time and decrease the likelihood of premature habituation or fatigue. Additionally, by noting the child's behaviour during immittance and OAEs, the audiologist should be able to adapt the test paradigm appropriately for the needs of that particular child.

During the past 60 years, a number of procedures have been added to the paediatric audiology test battery and virtually none have been abandoned along the way. In today's healthcare environment, however, the clinician must strive for efficiency in constructing the test battery to be used for the hearing assessment of a particular child. Although it is inappropriate to eliminate certain test procedures simply to 'save time', there is on the other hand no logical reason to perform any and all procedures that have, traditionally, been a part of the test battery. To be incorporated into the test battery for paediatric hearing assessment, each procedure must be selected based on the likely value it will add to the diagnosis and management of the child.[33] That is, procedures must pass the 'value added test' criterion before they are included within the test battery to be used for a specific patient. With the addition of relatively new procedures, such as OAEs, some of the older and time-tested procedures, e.g. bone-conduction pure-tone threshold measurement, may not always be necessary for thorough assessment of peripheral auditory function in children.

BEHAVIOURAL MEASURES: CENTRAL AUDITORY ASSESSMENT

In the early years of the profession of audiology, Mylkebust[34] noted that 'hearing is a receptive sense . . . and essential for normal language behaviour' (p. 11), and he noted that 'the diagnostician of auditory problems in children has traditionally emphasized peripheral damage. It is desirable that he also include considerations of central damage' (p. 54). He also explained that 'central deafness [central auditory processing disorder] is a deficiency in transmitting auditory impulses to the higher brain centres while receptive aphasia [language disorder] is a deficiency in the interpretation of these impulses after they have been delivered' (p. 153). During this era, Bocca et al.[35] reported that surgically confirmed central auditory system pathology could be detected with sufficiently sensitive audiological procedures. These pioneering observations and studies have since been validated by many clinical investigations. There are now a variety

of behavioural, electro-acoustic and electrophysiological techniques for the assessment of peripheral and central auditory system function, including APD. The term APD is used to describe a deficit in the perception or complete analysis of auditory information due to dysfunction anywhere within the auditory nervous system, usually but not invariably or exclusively within the central auditory nervous system.[22,36,37] APD may be found for very simple auditory tasks (e.g. detection of the presence of sound, as in pure-tone threshold measures) to complex tasks (e.g. dichotic listening tests). Auditory processing takes place before language processing or comprehension. The evaluation and management of APD is well within the scope of audiological practice and an accepted clinical activity within the field of communicative disorders as defined by the professional organisations representing audiology. The topic of audiological assessment and management of APD is addressed in Chapter 11 of this textbook.

NEURO-DIAGNOSIS OF AUDITORY DYSFUNCTION

Electrophysiological estimation of auditory sensitivity in infants and young children

The first and essential step in confirmation of hearing impairment is the estimation of hearing sensitivity. A sizable proportion of newborn infants who do not pass hearing screening, and also older children suspected of hearing impairment, will have hearing sensitivity within normal limits bilaterally throughout the frequencies important for speech perception and acquisition. For children whose developmental age is greater than about 6 months, definition of hearing sensitivity may be made with behavioural audiometry, at least for the better-hearing ear (Table 4.1). Electro-acoustical and electrophysiological techniques (e.g. OAEs and ABR) should be considered routinely, however, for ear-specific hearing assessment and to confirm incomplete, inconclusive or inconsistent behavioral findings. Applied in combination, these electrophysiological techniques permit reasonably accurate frequency-specific and ear-specific estimation of the configuration and degree of hearing sensitivity loss, and permit the early differentiation amongst sites of auditory dysfunction (e.g., middle ear, cochlea, neural, and central auditory pathways).

The clinical demand for reasonably accurate information on auditory thresholds of infants under the age of 6 months prior to initial management steps, such as hearing aid fitting, has generated unprecedented interest and investigation of auditory electrophysiological techniques and strategies. ABRs elicited by tone burst stimuli are now regularly applied for frequency-specific estimation of auditory thresholds within the speech frequency region (500 to 4,000 Hz). Within the past 30 years, accumulated clinical experience and formal clinical research have led to a proven protocol for ABR measurement with tone-burst stimuli. An example of a tone-burst ABR protocol was displayed in Table 4.2. Successful and reasonably accurate electrophysiological estimation of auditory thresholds for selected audiometric frequencies is greatly enhanced by the use of the proper test protocol and, of course, a sleeping, sedated or lightly anaesthetised child.

Auditory thresholds at frequencies important in speech perception, e.g. 500, 1,000 and 4,000 Hz, are estimated by analysis of ABR waveforms, specifically the presence of a reliable wave V, at the intensity levels within 10 dB of actual behavioural auditory thresholds. A clear and reliable ABR waveform is first recorded for a high intensity level (e.g. 80 to 85 dB nHL).

The ASSR is also available as a clinical tool for frequency-specific threshold estimation, particularly for children with severe and profound hearing loss. A full explanation of the protocols and procedures for tone-burst ABR measurement and ASSR recording is beyond the scope of this chapter. The reader is referred to recent resources on the topic.[28] The temptation to critically compare the two procedures and to ask questions such as 'Which technique is the best?' should be avoided. Rather, clinical use of frequency-specific ABR and ASSR techniques should be guided by the question 'Which procedure is likely to provide the information I require to properly and promptly assess and manage this child?' In many cases, the ABR and ASSR both should be included in the paediatric test battery.

Differentiating amongst sites of auditory dysfunction

There are at least three principles of paediatric hearing assessment today that contribute to accurate description of auditory status and, therefore, lead to a rational and evidence-based strategy for effective management. First, a test-battery approach is essential in the evaluation of hearing in children of any age. With the advent of newborn hearing screening there is increased demand for diagnostic audiological assessment of infants under the age of 4 months. That is, infants who do not pass a hearing screening at birth require follow-up evaluation within months to confirm and define the hearing impairment so that intervention can begin before 6 months after birth. This initial description of the type and degree of hearing impairment for each ear is based typically on electrophysiological procedures (see Table 4.1). This information is essential for determining whether hearing aids are indicated and, if so, the specifications of the hearing aid selection and fitting. Reasonably accurate electrophysiological estimations of auditory thresholds for three or four data points within the speech frequency region permit precise 'prescriptive' hearing aid selection and fitting,[38] even in infants as young as 2 to 3 months. The hearing aid fitting will later be adjusted and refined as the hearing impairment is better defined with behavioural audiometry (Table 4.1). Although electro-acoustic and electrophysiological measures of auditory function are invaluable during infancy, only behavioural measures truly reflect a child's hearing status.

Second, the audiological evaluation should lead to a differentiation of the type of hearing impairment, the general site of lesion and, along with other medical studies, a diagnosis. Examples of types of hearing impairment include conductive (middle-ear dysfunction), sensory (cochlear dysfunction), neural (eighth cranial nerve and/or central auditory nervous system) or combinations of these types. Thus, in addition to estimation of hearing thresholds, the objective of audiometry in infants must provide accurate information on the site of dysfunction within the auditory system. The importance of diagnostic paediatric assessment is easily appreciated by considering the distinctly divergent management approaches taken with three clinical entities all presenting with elevated (abnormal) auditory thresholds for air-conducted signals, but otherwise very different patterns of auditory findings. Middle-ear disease or malformation can be identified by abnormal immitance findings, and better auditory thresholds for bone- versus air-conduction stimulation with either ABR or pure-tone audiometry. Prompt identification of middle-ear disease with proper referral can lead to successful medical management and eliminate the need for amplification. A variety of paediatric diseases are associated with cochlear dysfunction. Some are amongst the Joint Committee on Infant Hearing[39] risk indicators. Diagnosis of the cause for sensory hearing impairment may require radiological studies and laboratory tests. Audiological findings for OAEs and ABR and pure-tone audiometry for older children can almost always confirm a sensory hearing impairment and even

differentiate between outer and inner hair-cell dysfunction. Amplification, rather than medical therapy, is the most common management strategy for a pure sensory deficit. Management is radically different for a third clinical entity – 'auditory neuropathy' – described in more detail in Chapter 12. The term auditory neuropathy is rather misleading, as some children falling into this category of auditory dysfunction may actually have inner hair-cell abnormalities. Gravel and Rapin[40] recently reviewed the disorder and offered guidelines for more accurate terminology. Comprehensive diagnostic audiometry for a small proportion of infants with hearing impairment show normal cochleae, or at least outer hair-cell, function. Based on ABR or behavioural measures, the hearing impairment may initially appear to be sensory in nature. Yet OAE recordings are normal, confirming cochlear outer hair-cell integrity.

Complete medical diagnostic work-up typically yields the diagnosis of a disorder secondary to neurological dysfunction, such as cerebral palsy, developmental delay or even neurodegenerative diseases. Although children with auditory neuropathy are most often graduates of the intensive care nursery, there are reports of auditory neuropathy in the well-baby population. Management of auditory neuropathy is not straightforward, and varies substantially depending on the pattern of auditory findings (e.g. pure-tone hearing thresholds) and the site of the dysfunction. Cochlear implants are now recognised as an effective management option for some children with auditory neuropathy.

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5 Radiological abnormalities of the ear

F. Calzolari and A. Martini

INTRODUCTION

In addition to clinical, genetic and audiological examination, diagnostic imaging is essential to precisely define ear abnormalities in newborns and children, in order to plan a complete therapeutic/habilitative programme as soon as possible and to select the eventual candidates for aesthetic and functional surgery.

The vortical and continuous development of new diagnostic imaging techniques during recent years has presented several ways of diagnosing and representing ear malformations. The radiologist plays an important role in suggesting and employing modern diagnostic techniques appropriately. Ear abnormalities are often associated with other malformations or may be an expression of inherited syndromes. The knowledge of up-to-date embryological and genetic concepts enables the production of a radiological report, which is not only descriptive but also interpretative, suggesting further investigations or genetic counselling.

IMAGING TECHNIQUES

Computed tomography (CT) and magnetic resonance (MR) represent the imaging techniques of choice for a complete description of congenital abnormalities of the external, middle and inner ear. Often CT and MR are both necessary to obtain a precise characterisation of extremely small structures of the ear. In the temporal bone region, airy spaces, bone and a wide range of soft tissues are present. CT accurately depicts the osseous portion of the external auditory canal (EAC), the tympanic membrane, the tympanic cavity, the ossicular chain, other petrous and mastoid cavities as well the inner-ear bone structures. On the other hand, MR well demonstrates soft tissues of the external ear, the liquids of the inner ear and the nerves within the internal auditory canal (IAC) and cerebellopontine angle. CT and MR are complementary for representation of the main vessels close to the ear.[1–3] Nowadays, many CT and MR techniques of image acquisition are available.

CT images should be acquired in high resolution with a bone algorithm using a single-detector or multi-detector technique (multislice CT), and displayed with a wide window of the grey scale for correct bone representation. Images must be thin, 1 mm or less. Both axial (Figure 5.1) and coronal images (Figure 5.2) should be obtained for an exhaustive CT examination of the petrous bone.

Direct axial images are easily obtained with the patient supine, with the canthomeatal line perpendicular to the tabletop.[4] The axial plane can be differently inclined. Usually, sections are made in a plane rotated 30° superior to the anthropological baseline, a line intersecting the

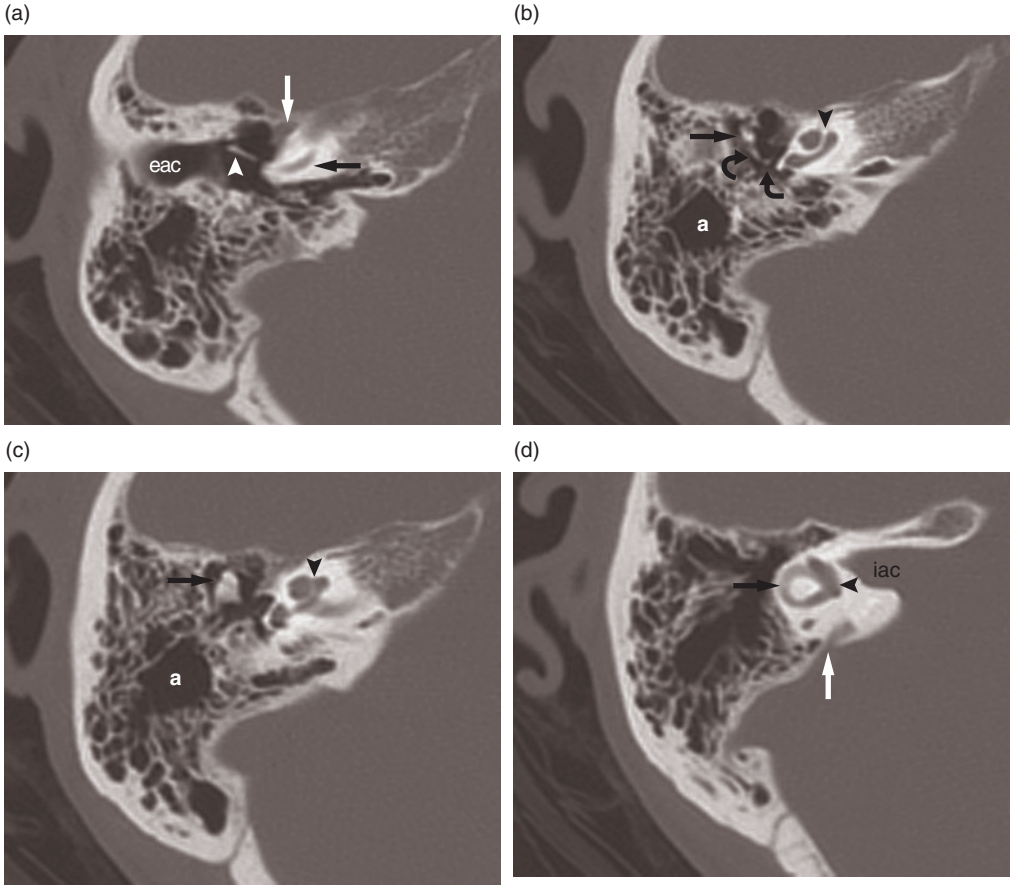

Figure 5.1 High-resolution CT of the right petrous bone: axial images. (a) External auditory canal (eac), malleus handle (arrowhead), tensor tympani (white arrow), basal turn of the cochlea (black arrow); (b) malleus head (straight arrow), incus (curved arrow), stapes (angled arrow), cochlea (arrowhead), mastoid antrum (a); (c) malleoincudal joint (arrow), cochlea (arrowhead), mastoid antrum (a); (d) vestibule (arrowhead), lateral semicircular canal (black arrow), vestibular aqueduct (white arrow), internal auditory canal (iac) ('b' and 'c' reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

inferior orbital rim and the external auditory meatus. This plane allows a good separation of the main petrous structures, so that they are better visualised, with less overlap and fewer partial volume artefacts.[5] For instance, this plane is particularly useful for incudo-malleolar joint and foramen lacerum representation. Moreover, using this plane avoids direct exposure of the lens, particularly important in paediatric patients.[6] Axial images are usually acquired and displayed from bottom to top, in a range between the temporomandibular joint and the superior edge of the petrous bone.[6] Visualisation of axial images only can lead to misinterpretation of structures parallel to the plane of section, such as the floor of the EAC, the tegmen or the convexity of the superior semicircular canal; for this reason, an exhaustive study of the ear implies also obtaining the coronal view.

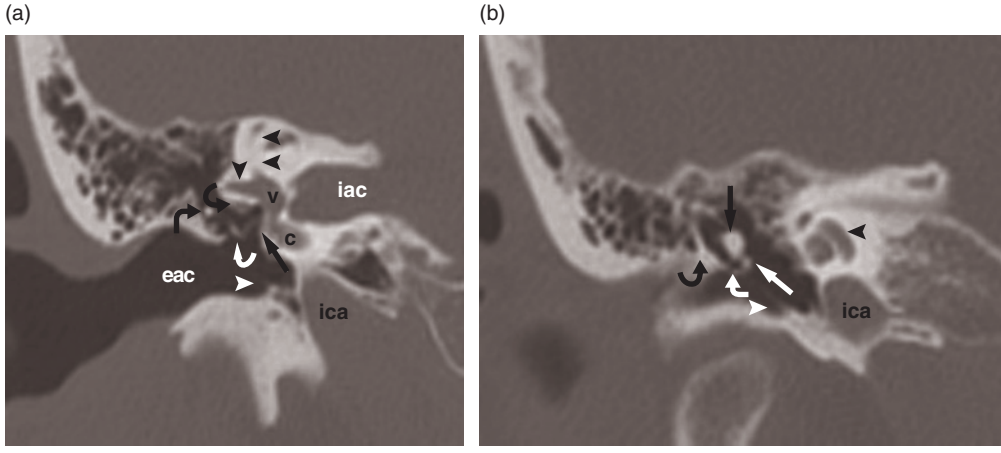

Figure 5.2 High-resolution CT of the right petrous bone: coronal images. (a) External auditory canal (eac), tympanic membrane (white arrowhead), malleus head (angled arrow), incus (curved white arrow), stapes (straight arrow), facial nerve (curved black arrow), cochlea (c), vestibule (v), lateral semicircular canal (arrowhead), superior semicircular canal (double arrowhead), internal auditory canal (iac), internal carotid artery (ica); (b) Tympanic membrane (white arrowhead), Prussak pouch (angled arrow), lateral tympanic wall (curved arrow), malleus head (black straight arrow), malleus short process (white straight arrow), cochlea (black arrowhead), internal carotid artery (ica) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

Coronal images can be obtained directly or through data reconstruction. Direct coronal images are acquired with discomfort for the patient, who lies on the cradle supine or prone, with the head overextended. Overextension of the head is not tolerated in patients with a short neck, particularly in the supine position in patients with vertigo and cervical spine diseases, and in the prone position in dyspnoeic and obese subjects; direct coronal scans are difficult to acquire in unsedated paediatric patients.[6] The coronal plane should be, as much as possible, parallel to the ascending branch of the mandible.[6] Coronal sections are taken and displayed from the loop of the posterior semicircular canal to the bony eustachian tube.[4]

Today, multi-slice CT scanners not only allow fast imaging but also enable us to reconstruct very thin images every 0.5 or even every 0.1 mm.[3] This obviates the need to scan in a second plane, making an excellent reconstruction possible, not only in coronal, but also in other planes.[7] The possibility of acquiring a single volume of data and reconstructing images in different planes allows X-ray exposure to be reduced. Moreover, through post-processing of CT multislice datasets three dimensional (3D) imaging and virtual endoscopy of the middle ear can be obtained, particularly useful for surgical planning.[8,9]

MR imaging of the ear is achieved through a wide range of techniques, usually using a head coil. During MR examination, the patient remains still, but in a comfortable position. For analysis of inner-ear malformations axial, coronal and 3D images are more frequently displayed; oblique images can be useful for representation of the nerves in the IAC.[1,3,5] The membranous labyrinth is only 12 mm high, so it should be examined in detail using submillimetric (≤0.7 mm thickness) images.[3]

T2-weighted images allow a high contrast between intralabyrinthine/cerebrospinal fluid and nerves and bone. In these images, intralabyrinthine/cerebrospinal fluid appears white in the

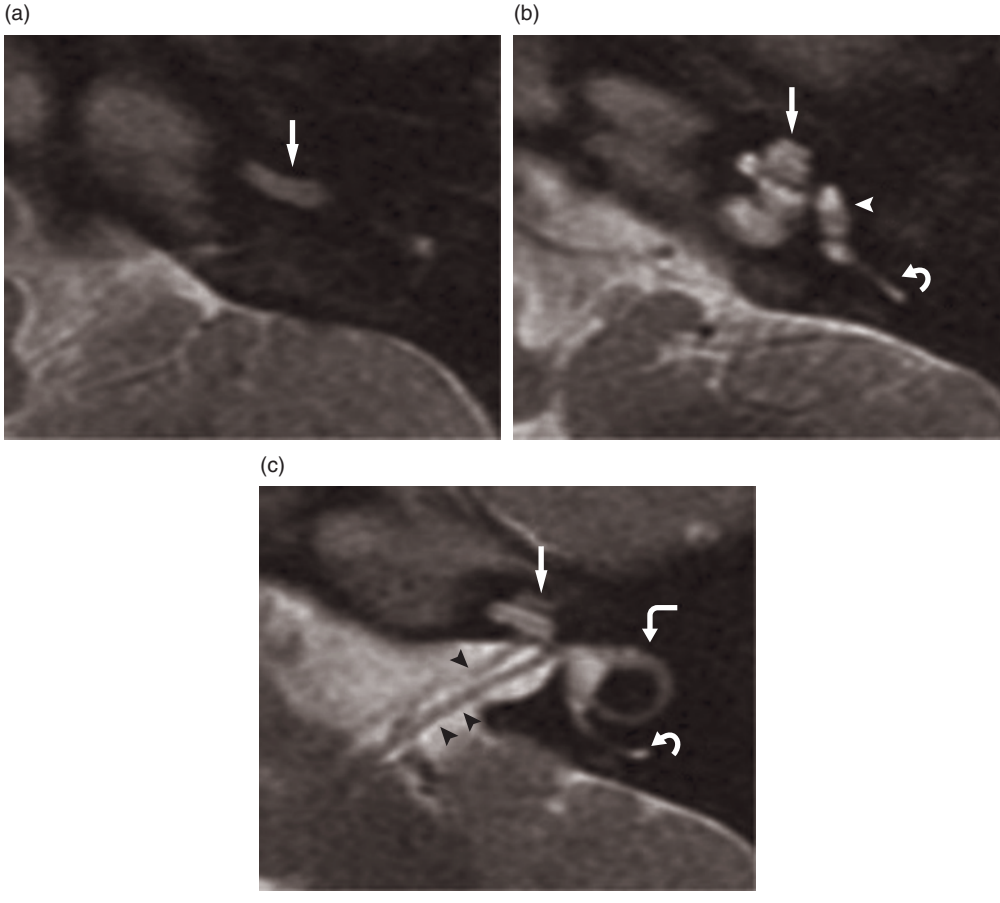

Figure 5.3 Axial T2-weighted MR images. (a) basal turn of the cochlea (arrow); b) cochlea (straight arrow), vestibule (arrowhead), posterior semicircular canal (curved arrow); c) cochlea (straight arrow), lateral semicircular canal (angled arrow), posterior semicircular canal (curved arrow), cochlear nerve (arrowhead), inferior vestibular nerve (double arrowhead).

grey scale (Figures 5.3 and 5.4). 3D-T2 weighted sequences such as constructive interference in the steady state (CISS) or fast imaging employing steady-state acquisition (FIESTA) (Figure 5.5) are frequently used for high-resolution imaging of the labyrinth and the IAC.[1,3,5,10]

Using high-resolution T2-weighted MR imaging, the scala vestibuli and scala tympani can be differentiated through an optimised combination of an imaging protocol and a 3D visualisation technique.[11]

In MR imaging, the signal-to-noise ratio (SNR) can be improved with high magnetic field machines: MR machines characterised by power of the magnet: or >3.0 Tesla. A suitable technique permits high-resolution representation of the inner ear with a clinically acceptable imaging time even at 3.0 (T).[12] The MR representation of the inner ear is clearly improved at 3.0 T (Figure 5.6); to obtain the same SNR at 1.5 T, approximately, double the measuring time would be required, with a reduction of patient comfort and increased risk of movement of the

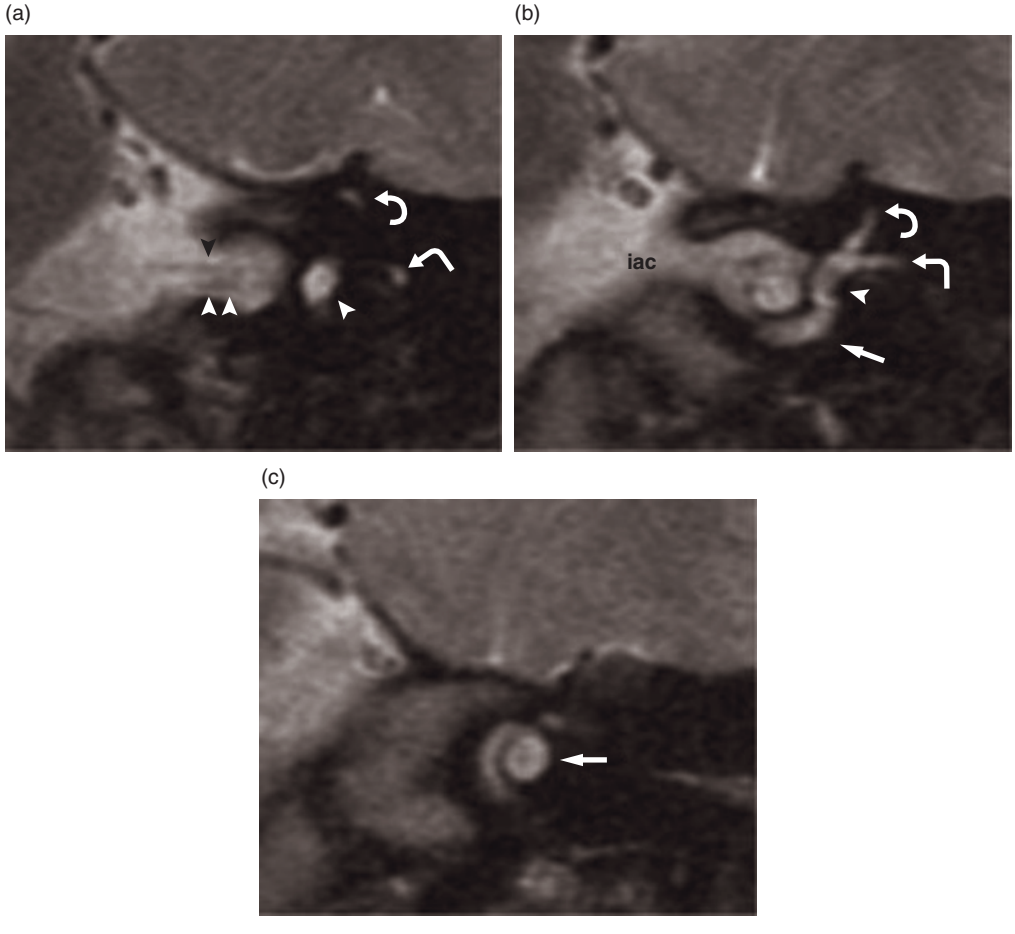

Figure 5.4 Coronal T2-weighted MR images. (a) vestibule (white arrowhead), superior semicircular canal (curved arrow), lateral semicircular canal (angled arrow), superior vestibular nerve (black arrowhead), inferior vestibular nerve (double arrowhead); (b) cochlea (straight arrow), vestibule (white arrowhead), superior semicircular canal (curved arrow), lateral semicircular canal (angled arrow), internal auditory canal (iac); (c) cochlea (arrow).

head.[13] Both 3D fast recovery fast spin-echo (FRFSE) and 3D CISS techniques provide high-resolution images of the labyrinth and cochlear nerve at 3.0 T. Moreover, excellent 3D reconstructions of the inner ear can be obtained (Figure 5.7). Unfortunately, differentiation of endolymph and perilymph fluid cannot be achieved using a high-magnetic field.[14]

Imaging for ear abnormalities is often requested for paediatric patients. Newborn babies, infants and young children are not cooperative, so sedation or general anaesthesia may be necessary. Sometimes newborns can be examined during the physiological sleep, after a regular breast or bottle feed; a sound sleep is usually more easily obtained if sleep deprivation occurs during feeding.[15] Actually, 'fast imaging' on both CT and MR has reduced the need for sedation and the time of anaesthesia. In particular, the short time taken for data

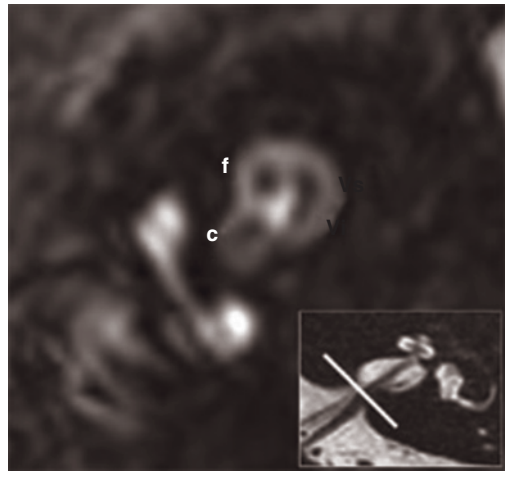

Figure 5.5 3D-T2 weighted MR sequence (FIESTA): image parallel to the internal auditory meatus: facial nerve (f), coclear nerve (c), superior vestibular nerve (vs), inferior vestibular nerve (vi) (courtesy of S. Battaglia and M. Leonardi, Neuroradiology Bellaria Hospital, Bologna, Italy).

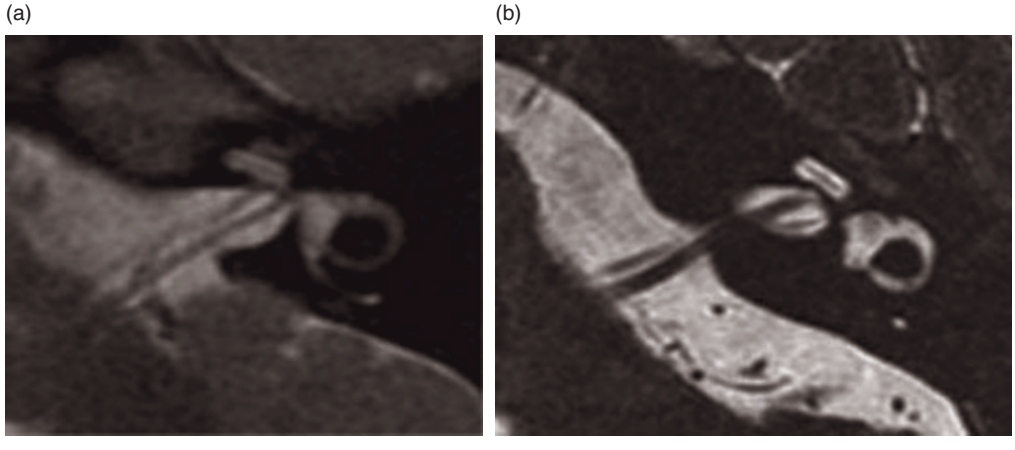

Figure 5.6 T2-weighted MR axial images at 1.0 T (a) and 3.0 T (b): the representation of the inner ear and nerves is clearly improved al 3.0 T ('b' courtesy of S. Battaglia and M. Leonardi, Neuroradiology Bellaria Hospital, Bologna, Italy).

acquisition using multi-slice CT reduces the probability of motion artefacts and the need for sedation.

CT and MR are both invasive, not only for the need of anaesthetic drugs. Invasiveness of CT examination may be reduced by use of low-dose techniques and multiplanar reconstructions as an alternative to direct scans.[16] There are also potential risks related to MR examination, so that precise safe practice guidelines must be followed.[17,18] Many MR sequences are

Figure 5.7 3D-FRFSE T2-weighted MR sequence: three-dimensional representation of the inner ear (courtesy of S. Battaglia and M. Leonardi, Neuroradiology Bellaria Hospital, Bologna, Italy).

noisy and particular care must be taken to protect the ear, especially with babies.[19] Thermal injury from excessive radio frequency power deposition represents another possible risk. The probability of this injury is especially greater on higher field scanners (e.g. 1 T and above); sedated, anaesthetised, unconscious or young patients may not be able to express related symptoms.[17] In the neonatal and paediatric population, special attention is needed in monitoring body temperature in addition to other vital signs.[18] Newborns and young infants are more sensitive to tissue heating and acoustic noise, present in particular using MRI scanners operating at ≥3 T.[20]

Nowadays, ear abnormalities may be diagnosed even during fetal life. The advent of 3D ultrasound has improved the possibility of prenatal diagnosis of anomalies of the auricle.[21] On the other hand, fetal MR may recognise inner-ear abnormalities.[22] Ear malformations are often associated with more complex fetal malformations, so that fetal ultrasound and MR are very important for prenatal diagnosis and genetic counselling.

ABNORMALITIES OF THE EXTERNAL EAR

The auricle develops from six mesenchymal swellings, called auricular hillocks, which arise at 6 weeks' gestation around the margins of the first branchial groove. The mesenchyme in these hillocks is derived from mesoderm in the first and second branchial arches.[23] All the auricle except the tragus develops from the second (hyoid) arch, whereas the tragus derives from the first (mandibular) arch.[24] The auricle begins to develop in the upper part of the future neck region; as the mandible develops, the auricle moves to the side of the head and ascends to the level of the eyes.[23]

The EAC arises from deepening of the first branchial groove in the 9th week, but opening of the bony part of the EAC starts only in the 30th week, after complete differentiation of the external, middle and inner ear.[2] The ectodermal lining cells of the developing external meatus proliferate to form a meatal plug that subsequently recanalises.[25]

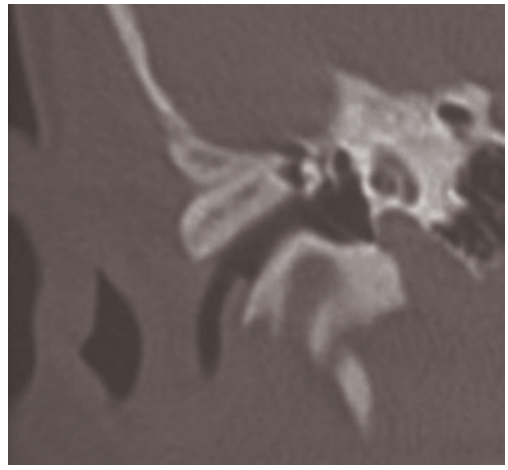

Figure 5.8 Coronal CT scan. Abnormal angulation of the external auditory canal.

Mild errors of morphogenesis of the auricle are predictive of the identification of major malformations, genetic syndromes and metabolic and psychiatric diseases.[26] Microtia is a congenital malformation characterised by an underdeveloped auricle. It can be classified in three degrees, varying from a small auricle to its total absence. In the most serious cases, there are only rudimentary remnants; anotia is the complete lack of the auricle.[27] Microtia is easily diagnosed at birth by simple clinical examination; otherwise imaging should be utilised in order to depict other ear malformations present. Microtia is often associated with external, middle and, less frequently, inner-ear malformations.[2,16,24,28–30]

Because of abnormal development of the first branchial groove, microtia is often associated with abnormalities of the EAC.[2,6,16,24] The failure of auricle development and its caudocranial migration in neck region may allow abnormal angulation of the EAC (Figure 5.8). The failure of groove deepening or of recanalisation of the meatal plug results in various degrees of soft-tissue and bony EAC stenosis or atresia[25] (Figure 5.9). Congenital stenosis and atresia of the EAC may involve the cartilaginous portion, bone portion, or both (Figures 5.10 and 5.11).

Duplication of the EAC is another rare malformation where a second, more or less rudimentary EAC coexists with a usually normal EAC.[31] Generally, congenital abnormalities of the EAC are bilateral in 30% of cases and occur more often in males and in the right ear.[16,24]

Moreover, dysplasia of the EACs often associated with other craniofacial malformations, regarding anatomical structures which develop from the same branchial derivative.[25,32] On the side of the atresia, frequently there is hemifacial microsomia, mandible hypoplasia and temporomandibular joint dysplasia (Figure 5.12), mastoid hypoplasia and small middle cranial fossa (Figure 5.13).

The position of the mandibular condyles may be highly variable. In the absence of a normal tympanic bone, the condyle articulates with the mastoid directly and often assumes a more posterior and superior position.[33]

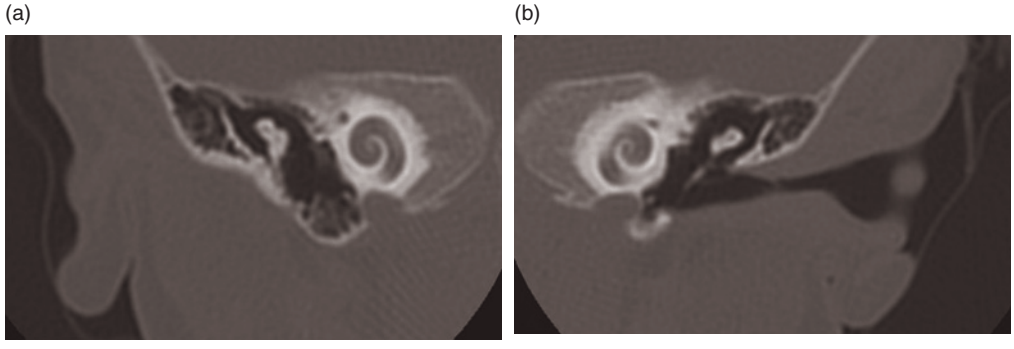

Figure 5.9 Coronal CT scans. Right atresia (a); normal left external auditory canal (b) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

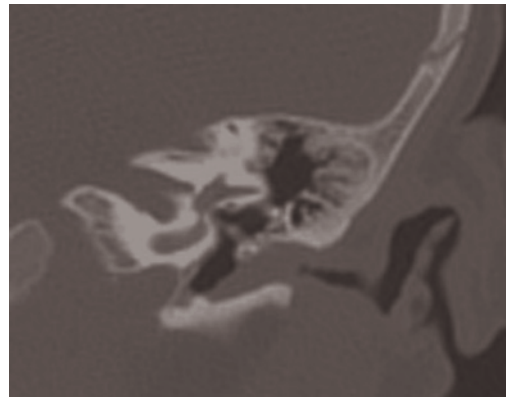

Figure 5.10 Coronal CT scan. Stenosis of the left external auditory canal, involving the soft tissue only.

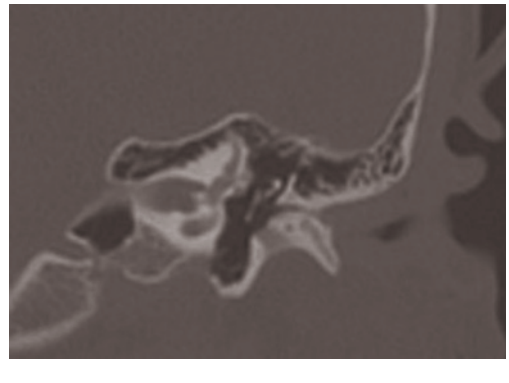

Figure 5.11 Coronal CT scan. Stenosis of the bone portion of the left external auditory canal.

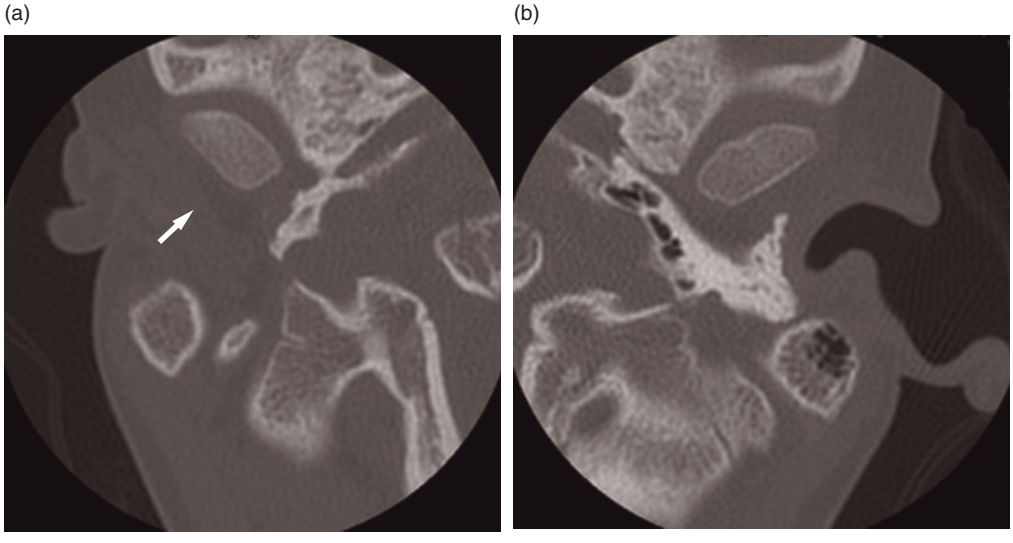

Figure 5.12 Axial CT scan. Microtia and absence of the posterior wall of the glenoid cavity (arrow) on the right (a), normal EAC and glenoid cavity on the left (b).

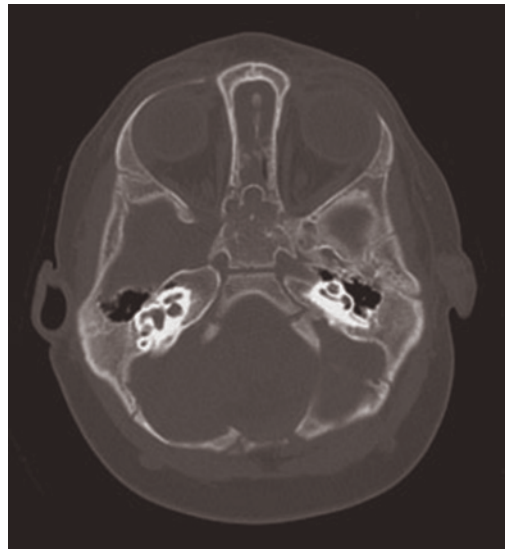

Figure 5.13 Axial CT scan. Hypoplasia of the middle and posterior fossa on the left side in patient with left microtia and atresia.

ABNORMALITIES OF THE MIDDLE EAR

Microtia and congenital malformations of the EAC are frequently associated with middle-ear abnormalities, probably because of the common embryological origin of the external and

middle-ear structures.[2,16,23,24] The eustachian tube and tympanic cavity are formed from the first pharyngeal pouch, so that they are of entodermal origin; the medial constricted portion becomes the eustachian tube, whilst the remainder becomes the tympanic cavity. The terminal end of the first pharyngeal pouch lies against the epithelium of the infolded first branchial groove at the site of the future tympanic membrane.

However, this close relationship is of short duration because tissue soon grows between them.[24] Here, the ossicular chain develops from the mesodermal first and second branchial arches.[2,23] In particular, failure of differentiation of the first branchial arch leads to malformations of the tensor tympani muscle and incudomalleal joint, whilst failure of the second branchial arch affects the stapedius muscle, the facial canal, the styloid process and the lower part of the ossicular chain, with the exception of the vestibular portion of the stapes footplate.[2,24] The vestibular portion of the stapedial footplate develops from the otic capsule, so that the stapes has a dual origin.[24]

The ossicular chain develops by condensation of mesenchyme; the development of the stapes starts during the 4th week, the malleus and incus in the 7th week. Concerning the stapes, chondrification begins at the 8th week, ossification around the 18th week; the final remodelling of the stapes is complete by the 38th week. Chondrification of the malleus and incus begins at the 8th week; ossification starts first in the incus at the 16th week, followed shortly by the malleus at week 16.5. Malleus and incus are largerly completed by week 30.[24]

The normal condition of the middle ear at birth should not mistaken for congenital malformations. At birth there is still a remnant of unresorbed embryonal tissue that fills the tympanic cavity. Mesenchyme occupies 20% of the middle ear at birth and disappears by 1 year of age. On the other hand, in middle ears with congenital anomalies, mesenchyme occupies about 30% of the middle ear at birth and does not resolve until 3 years of age (Figure 5.14). Mesenchyme is found most frequently in the mesotympanum, followed by epitympanum, aditus ad antrum and mastoid antrum. Amniotic fluid can be also detected in the middle ear of newborns.[24]

Ossification of the ossicles seems to occur steadily throughout fetal life and after birth. Bone marrow was observed in both the malleus and incus in children until 25 months of age, whilst after the age of 25 months, no bone marrow tissue was present in either of the ossicles.[34]

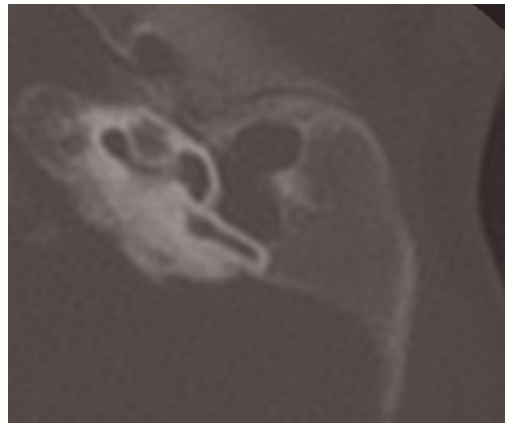

Figure 5.14 Axial CT scan. Two-month-old female with left atresia and absence of the ossicles. Mesenchyme occupies the middle ear.

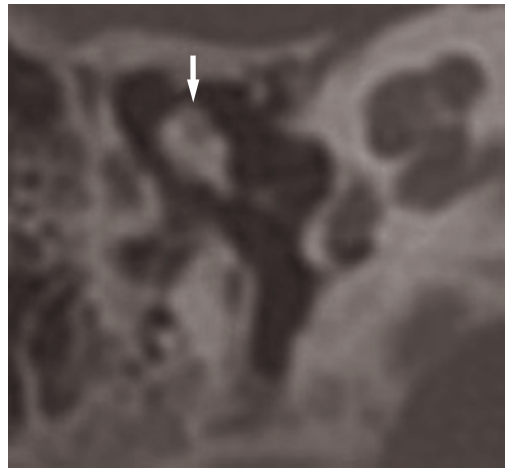

Figure 5.15 Axial CT scan. Seven-month-old male. Abnormal fusion of the head of the malleus with the body of the incus. Hypodensity due to incomplete ossification of the head of the malleus (arrow), normal at this age.

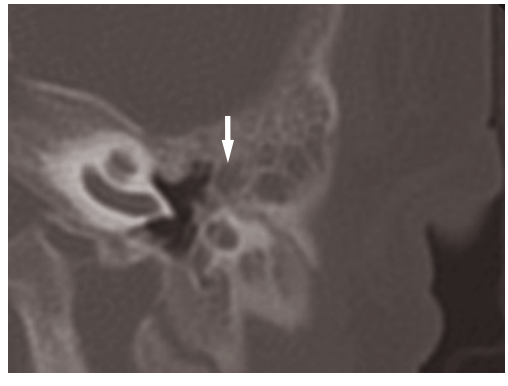

Figure 5.16 Axial CT scan. Three-month-old male. Small tympanic cavity and atresia. Mesenchyme occupies the middle ear (arrow).

So, on CT examination performed before 25 months of age, hypodensities of the ossicular chain should not be misinterpreted as congenital abnormalities or acquired lesions (Fig. 5.15).

The most frequent abnormalities of the middle ear are the reduction of the tympanic cavity and dysplasia of the ossicles; these anomalies are often associated. The tympanic cavity may be completely absent or very small. Hypoplasia of the tympanic cavity is frequently associated with atresia of the EAC (Figure 5.16). Moreover, volumetric reduction of the middle ear is often correlated with the degree of microtia. In general, there is a high correlation between the degree of microtia and the frequency of EAC and middle-ear malformations.[2,16] In other words, 'the better developed the auricle, the better developed the middle ear'; thus, the degree of microtia can be used as an indicator of middle-ear development.[28,35]

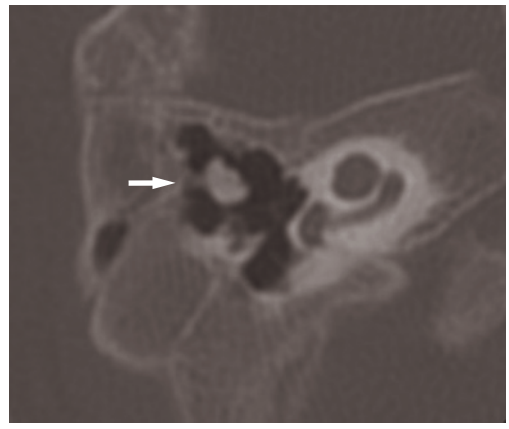

Figure 5.17 Axial CT scan. Abnormal fusion of the head of the malleus with the body of the incus; fixation of the head of the malleus to the tympanic wall (arrow).

A close relationship between the formation of the EAC and that of the malleal manubrium has been demonstrated in humans. The manubrium was identified in all ears with EAC stenosis, whereas it was absent in all ears with EAC atresia.[36] Nevertheless, congenital middle-ear defects may coexist with an intact external ear.[37]

Amongst the anomalies of the ossicles, dysplasia or absence of the malleoincudal joint and incudostapedial joint are relatively frequent.[2,6,16] Malleoincudal joint abnormalities are highly correlated with third-degree microtia and atresia.[16] CT enables representation of the characteristic appearance of incudomalleal joint dysplasia, with fusion of the head of the malleus with the body of the incus (Figure 5.15). The ossicular chain may be completely absent (Figure 5.14) or dysplastic. Other frequent ossicle malformations are fixation of the head of the malleus to the epitympanic wall (Figure 5.17), fixation of the short process of the incus to the wall of the incudal fossa, malleoincudal fixation, absence of the long process of the incus and incudostapedial disconnection.

Incudostapedial disconnection is supposed to be the most common isolated ossicular abnormality; it is thought to be secondary to a lack of development of the long process of the incus rather than underdevelopment of the head of the stapes.[24] The stapes is not involved as frequently as the rest of the ossicular chain.[2,16] Even in severe microtia, about 30% of the stapes remains normal. In minor microtia, the results are similar, so stapes anomalies have no significant correlation with the degree of auricular anomalies. The most commonly observed abnormality is a missing stapes.[2] Congenital absence of the stapes and the oval window is an anomaly reported in only sporadic cases.[38] However, this anomaly was found in two relatives and it may be related to inheritance.[39]

Anomalies of the stapes and the oval window are frequently associated with abnormal development and malposition of the horizontal facial nerve canal.[40,41] If the stapes is absent, the horizontal tympanic portion of the nerve may run just in front of the oval window, as well demonstrated on both axial and coronal CT images (Figure 5.18).

Displacement of the nerve and lack of a bony cover are two conditions that place the facial nerve at risk of being injured by the unwary surgeon.[41] In general, paediatric otorhinolaryngologists should be cautious when exploring patients with ear malformations because associ-

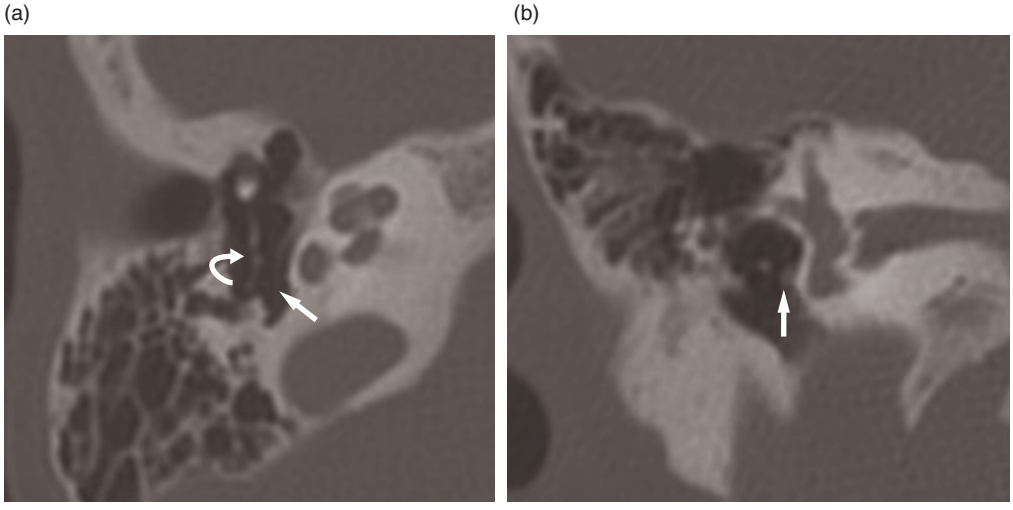

Figure 5.18 Axial (a) and coronal (b) CT scans. Tympanic dehiscence of the facial nerve (arrow); incudostapedial dysplasia (curved arrow) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

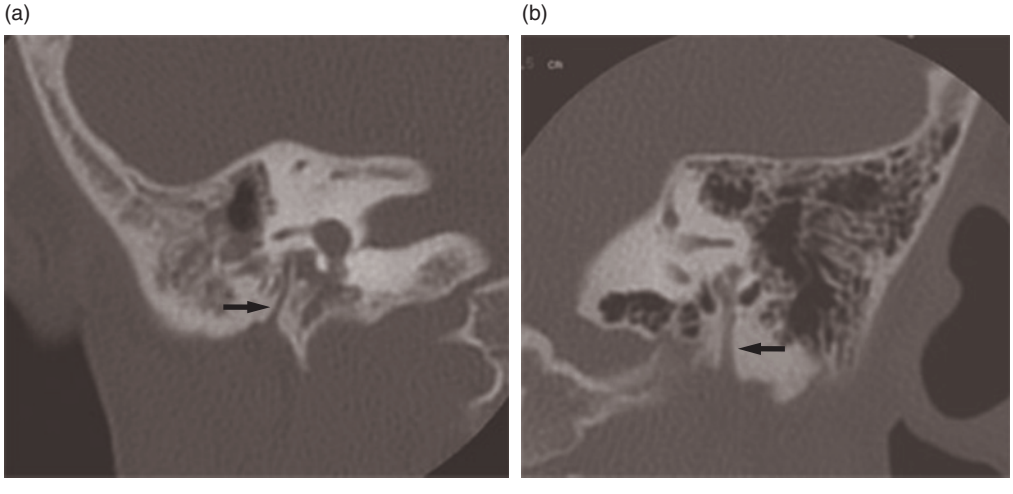

Figure 5.19 Coronal CT images. (a) Right atresia and anterior dislocation of the mastoid segment of the facial nerve (arrow); (b) normal situation of the nerve on the left (arrow) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

ated facial nerve anomalies may be present.[42] Abnormalities of the vertical portion of the facial nerve coexist very often with external and middle-ear malformations.[2,16,30] In the case of EAC atresia, tympanic cavity reduction and mastoid hypoplasia, coronal CT images show precisely anterior dislocation of the mastoid segment of the nerve, which runs obliquely, medial to lateral, in the frontal plane (Figure 5.19).

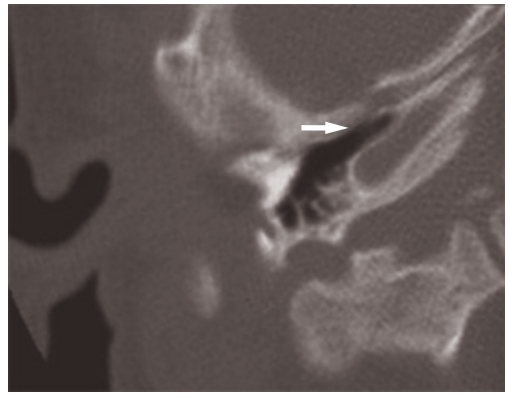

Figure 5.20 Axial CT scan. Dilation of the eustachian tube (arrow) and atresia (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

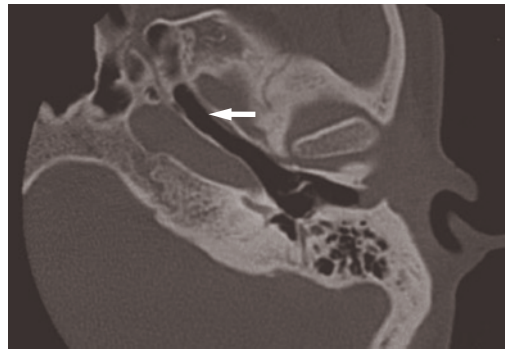

Figure 5.21 Axial CT scan. Persistence of the tubotympanic recess (arrow) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

The mastoid segment of the facial nerve has been described as being 3 mm more anteriorly displaced in patients with 2nd and 3rd grade microtia than in those with 1st grade microtia.[30] Bifurcation of the intratemporal facial nerve has been also reported.[43]

Direct axial CT images and oblique 'reconstructions' are useful to represent eustachian tube abnormalities. Dilation of the tube can be found in cases of atresia (Figure 5.20) or other skull base anomalies characterised by persistence of the first branchial pouch or the tubotympanic recess (Figure 5.21). A eustachian tube of reduced volume has been described in patients with chromosomal aberrations, such as trisomies 13, 18, 21 and 22 and inversion of chromosome 1.[44]

ABNORMALITIES OF THE INNER EAR

The inner ear has an embryological origin completely different from that of the external and middle ear. The inner ear develops from neuroectoderm between the 4th and 8th week of gestation. The otic placode, an ectodermal thickening in the neighbourhood of the myelencephalon, invaginates and becomes the otic vesicle (or otocyst) by week 5. The otic vesicle subdivides

into two pouches: a ventral (cochlear) pouch, which is the precursor of the cochlear duct and saccule, and the dorsal (vestibular) pouch, the precursor of the endolymphatic duct, utricle and semicircular canals. Cochlear development is complete at the 8th week. The saccule, endolymphatic duct and utricle are completed at 11 weeks, the semicircular canals between the 19th and 22nd weeks. The superior semicircular canal is completed first, followed by the posterior and finally by the lateral semicircular canal. Ossification of the inner-ear structures begins at week 15 or 16 and is complete by week 23; 14 ossification centres have been demonstrated for ossification of the periotic capsule of human fetuses.[3,24]

Development of the inner ear requires intrinsic and extrinsic factors that regulate proliferation. Most inner-ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy. The cause of interruption may be the result of an inborn genetic error or a consequence of teratogenic exposure (i.e. virus or radiation) during the period of inner-ear organogenesis.[45]

A universally accepted classification of inner-ear abnormalities does not exist yet.[46] Anomalies may involve all inner-ear structures: cochlea, vestibule, semicircular canals, IAC, vestibular aqueduct and cochlear aqueduct. Generally, two types of malformations may be present: single-branch and multi-branch abnormalities; single-branch abnormalities involve only one anatomical structure, whilst in multi-branch abnormalities, two or more structures are involved. The abnormalities may be symmetrical or asymmetrical. A genetic defect would be expected to cause identical anomalies on both sides; on the other hand, in cases of asymmetrical deformities, the cause is more probably represented by an external factor.[47]

In 1987, Jackler et al.[48] proposed a widely accepted classification of inner-ear malformations, which were categorised as complete labyrinthine aplasia (Michel deformity), cochlear aplasia, cochlear hypoplasia, incomplete partition (Mondini deformity) and common cavity; incomplete partition cases were classified as mild and severe. The Jackler classification was quoted by the European Congenital Ear Anomaly Inventory.[49] In 2002, Sennaroglu and Saatci[47] outlined the limits of the Jackler classification in cases of incomplete partition and proposed a similar classification, which distinguished incomplete partition type I (IP-I) and type II (IP-II).[47] Moreover, the same authors asserted that it is more appropriate to classify these malformations as 'cochleovestibular malformations' than as 'cochlear malformations'.[47]

Michel deformity is a malformation characterised by absence of all cochlear and vestibular structures (complete labyrinthine aplasia); it results from developmental arrest at the 3rd week.[47] This deformity is very rare; the diagnosis is made on CT and MR when the complete inner ear is absent or when a single fluid-filled cavity (otocyst-like cavity) replaces the normal labyrinthine structures.[3,24]

Cochlear aplasia is the anomaly characterised by a completely absent cochlea; it may coexist with a normal, dilated or hypoplastic vestibule and semicircular canals. This abnormality is rare and results from arrest of development late in the 3rd week.[47] The region of the cochlea is replaced by dense labyrinthine bone; because of the absence of the cochlea, the labyrinthine segment of the facial canal is more anterior than its usual location in the cochlea.[24,47] It is important to differentiate cochlear aplasia from cochlear ossification. In complete ossification of the cochlea, the basal turn of the cochlea produces the characteristic bulging in the middle ear, the so-called promontory; conversely, in the aplastic cochlea bulging of the promontory is absent.[47]

The common cavity represents a further step of failure of development, at the 4th week of gestation. The cochlea and vestibule form a common cavity, without any differentiation (Figure 5.22); a small common cavity probably represents earlier arrest than a large common cavity.[47] One-fourth of all cochlear malformations are common cavity; in this malformation, the IAC can be recognised, in contrast to complete labyrinthine aplasia.[3]

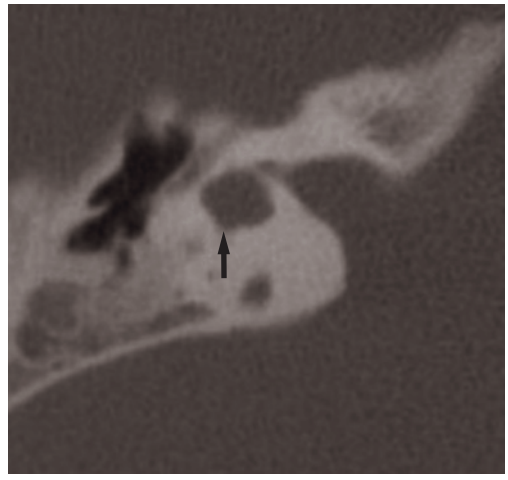

Figure 5.22 Axial CT scan. Inner ear malformation: the cochlea and vestibule form a 'common cavity' (arrow). Patient with BOR syndrome.

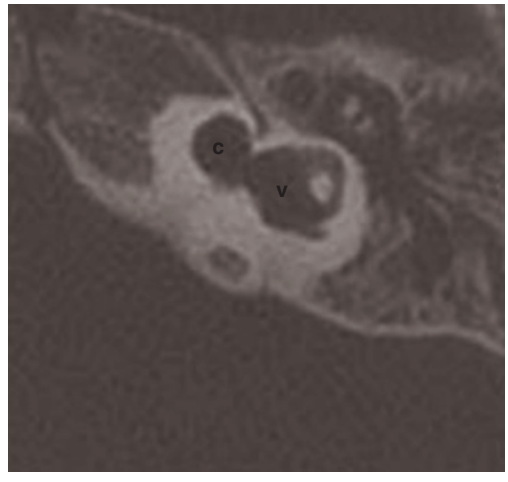

Figure 5.23 Axial CT scan. Unpartitioned completely empty cochlea (c) and dilated vestibule (v): 'incomplete partition type I'.

IP-I is a cystic cochleovestibular malformation: an unpartitioned completely empty cochlea, without interscalar septum and modiolus and associated with a grossly dilated vestibule (Figures 5.23 and 5.24). In this case, the arrest of development is thought to occur at the 5th week.[47,50]

Cochleovestibular hypoplasia is a further differentiated malformation so that the cochlea and vestibule are separate from each other, but their dimensions are smaller than normal (Figures 5.25 and 5.26): this probably represents failure of development at the 6th week. The vestibule may be hypoplastic or absent, no vestibular aqueduct malformation is observed, and the IAC is usually normal or smaller.[47]

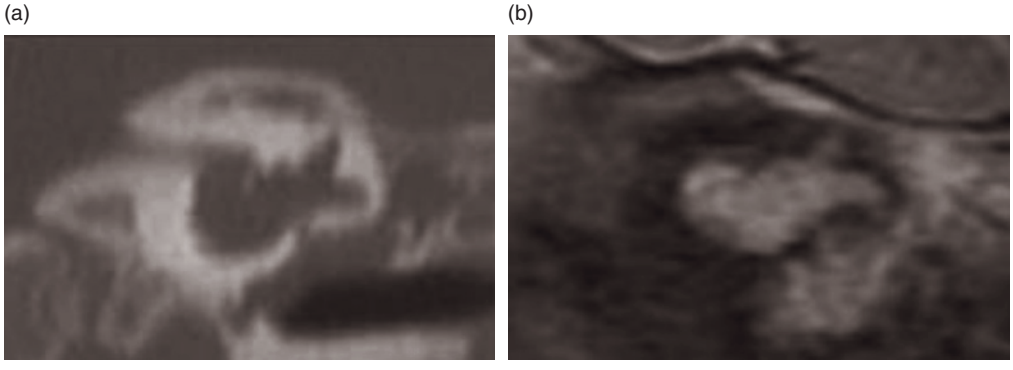

Figure 5.24 'Incomplete partition type I': coronal CT (a) and T2-weighted MR (b) images.

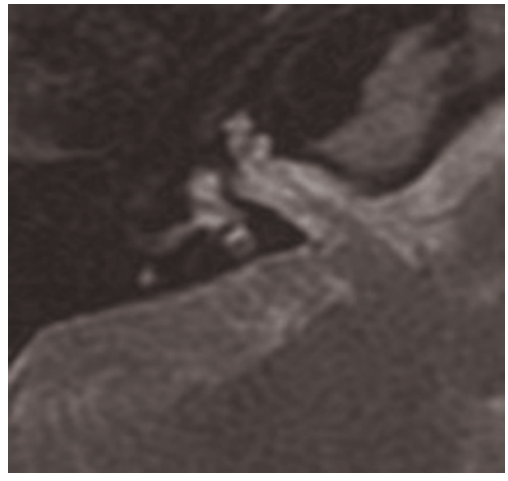

Figure 5.25 T2-weighted axial MR image: 'cochleovestibular hypoplasia'.

IP-II represents later developmental arrest at the 7th week of gestation. The cochlea has 1.5 turns and its internal organisation is more developed. There is a normal basal turn, whilst the middle and apical turns form a cystic cavity (cystic apex); the basal part of the modiolus is present. Vestibular dilation is minimal in IP-II compared with IP-I; the vestibular aqueduct is always enlarged[47,50] (Figure 5.27).

Recently, a previously undescribed anomaly of human cochleae with three turns has been reported; histological sections showed the basilar membranes to be longer than normal. This finding of an extra apical turn of the cochlea is different from those deformities produced by an interruption in development because it represents a hyperplasia of the cochlea; so, it should be considered a new category of anomaly.[45]

Cochleovestibular malformations are well demonstrated on both CT and MR imaging. The interscalar septal defect and absence of the osseous spiral lamina of the middle and apical turns can be best demonstrated on heavily T2-weighted MR images. Sometimes defects between the scala tympani and scala vestibuli can be found as the only detectable malformation in an oth-

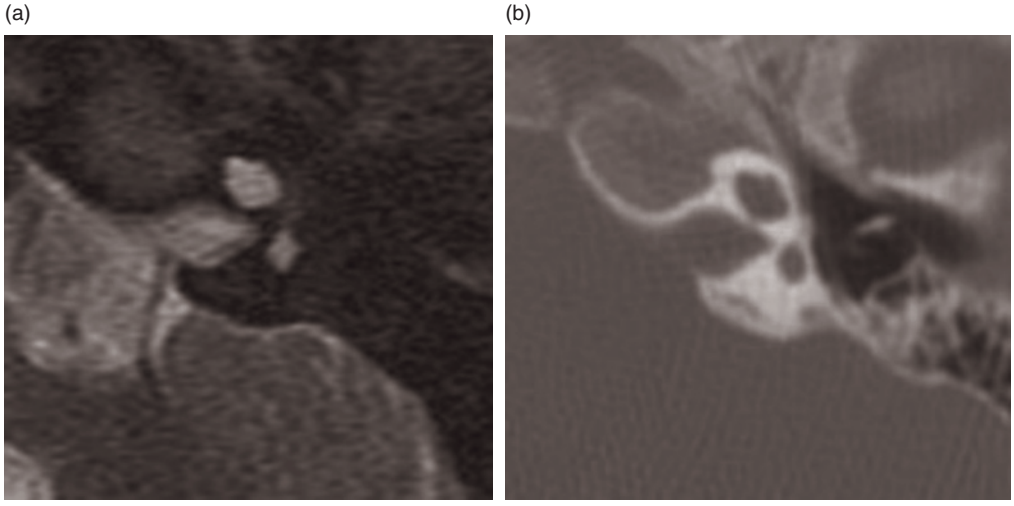

Figure 5.26 'Cochleovestibular hypoplasia': T2-weighted MR (a) and CT (b) axial images.

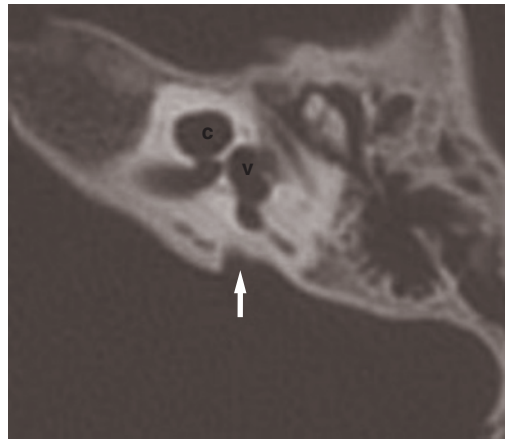

Figure 5.27 Axial CT scan. 'Incomplete partition type II': cystic apex of the cochlea (c), minimal vestibular dilation (v), enlargement of the vestibular aqueduct (arrow).

erwise normal cochlea. These anomalies can be diagnosed only on high-resolution T2-weighted MR images and remain invisible on CT.[3]

The saccule and utricle are completely formed at the 11th week of gestation. Absence, hypoplasia and dilation of the vestibule alone are rare, and most often these malformations are associated with other inner-ear malformations such as semicircular canal anomalies or cochlear anomalies. Vestibular malformations may also be isolated.[3,24]

The semicircular canals develop between the 6th and 8th week together with other inner-ear structures, and their embryology is completed between the 19th and 22nd week of gestation. The superior semicircular canal develops first, whilst the lateral semicircular canal last. A short broad cystic lateral semicircular canal confluent with the vestibule, the so-called lateral semicircular canal / vestibule dysplasia, is one of the most frequent inner-ear malformations

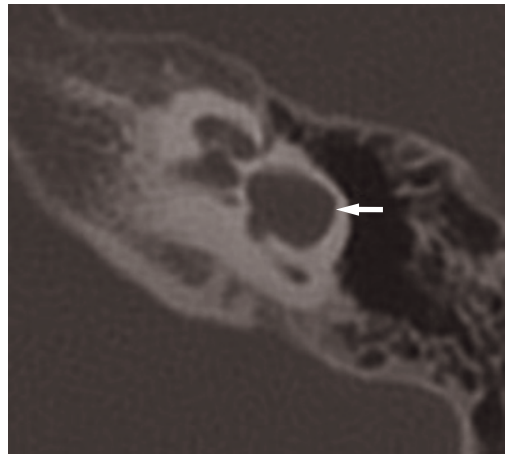

Figure 5.28 Axial CT scan. Cystic lateral semicircular canal confluent with the vestibule (arrow).

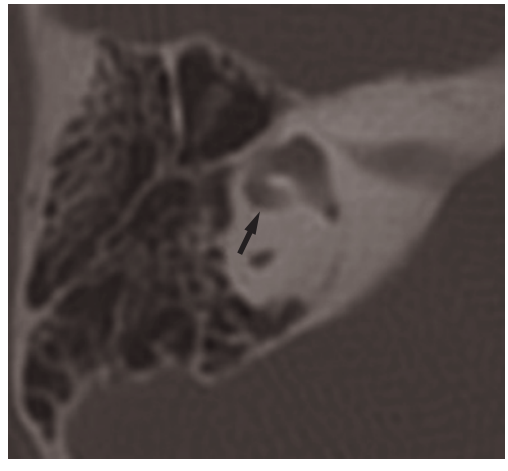

Figure 5.29 Axial CT scan. Short lateral semicircular canal (arrow).

(Figure 5.28). It may occur in isolation or bilaterally and can be associated with other inner-ear anomalies. Other more subtle malformations such as short (Figure 5.29), wide, narrow, partially or totally absent and ectatic or narrowed semicircular canals can also be easily diagnosed on CT and MR.[3]

Semicircular canal dehiscence consists of a defect of the bone over the superior or posterior semicircular canals, which normally separates the canal from the intracranial subarachnoid spaces. This anomaly should be sought for in patients with vertigo induced by loud noise and/or pressure changes in the middle ear or intracranial spaces. Dehiscence of the superior and posterior semicircular canal is readily detected by CT on coronal (Figure 5.30) and axial images. Heavily T2-weighted MR images can diagnose the dehiscence with a sensitivity of 96% and specificity of 98% compared with CT; the subarachnoid space gives a high signal similar to that of the fluid-filled semicircular canals: if the former is narrow, connection of the semicircular canal to the subarachnoid space might difficult to identify.[51]

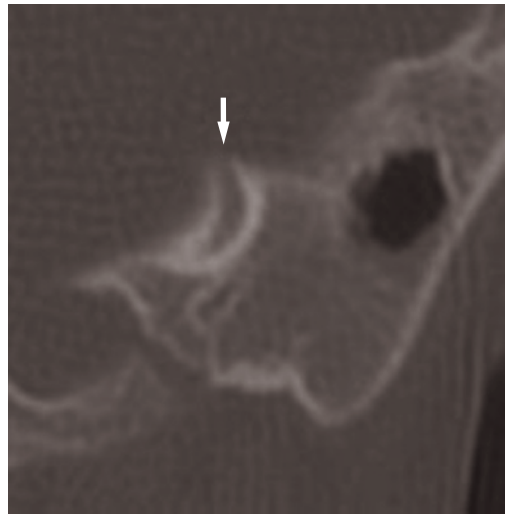

Figure 5.30 Coronal CT scan. Superior semicircular canal dehiscence: defect of bony wall (arrow).

An enlarged vestibular aqueduct (EVA) with a large endolymphatic duct and sac (LEDS) is one the most common malformations associated with congenital and progressive sensorineural hearing loss (SNHL). The pathogenetic mechanism by which LEDS causes progressive SNHL remains speculative. Cerebrospinal fluid pressure waves transmitted through the enlarged endolymphatic structures to the cochlea or reflux of hyperosmolar fluid of the endolymphatic sac into the cochlea may damage hair cells.[52,53] LEDS is commonly associated with other cochlear and vestibular anomalies, such as an enlarged and rounded vestibule, hypoplastic cochlea and abnormal appearance of the semicircular canals.[54,55] This malformation is probably due to an arrest of the normal development of the endolymphatic duct and sac: instead of the normal inverted J shape, the duct and sac remain enlarged in their early embryological form.[54] The vestibular aqueduct is considered enlarged when its anterior-posterior diameter or lateral-medial dimension, measured at its mid-portion, is 1.5 mm or greater.[54,55] Sometimes, the transverse midpoint diameter on MR images may be minimally larger than that on CT scans; a 'blooming effect' of the bright endolymph on MR images may explain this small discrepancy.[54] However, CT and MR both should be performed in order to evaluate a large vestibular aqueduct and large endolymphatic duct and sac: in fact, CT shows the bony vestibular aqueduct very well, whilst the extraosseous endolymphatic sac is demonstrated only by MR[54] (Figure 5.31). Some patients with SNHL have vestibular aqueducts not dilated on CT, because only the extraosseous sac is enlarged, so visible on MR images, but not on CT.[54,56] Axial T2-weighted MR images are the best to visualise the endolymphatic duct and sac and to differentiate them from subarachnoid spaces because of visualisation of the dura between the sac and cerebrospinal fluid (Figure 5.31). Usually, the signal intensity of the fluid within the enlarged endolymphatic duct and sac is similar to that of cerebrospinal fluid; an abnormal hyperintense signal on T1- and T2-weighted MR images could be due to protein-rich and hyperosmolar endolymph.[56,57] (Figure 5.32).

Enlarged cochlear aqueduct is a distinct entity that has been described with other inner-ear malformations. This abnormality is easily detected by CT thin scans (Figure 5.33). This malformation is extremely rare.[47] Isolated enlargement of the cochlear aqueduct has still not been

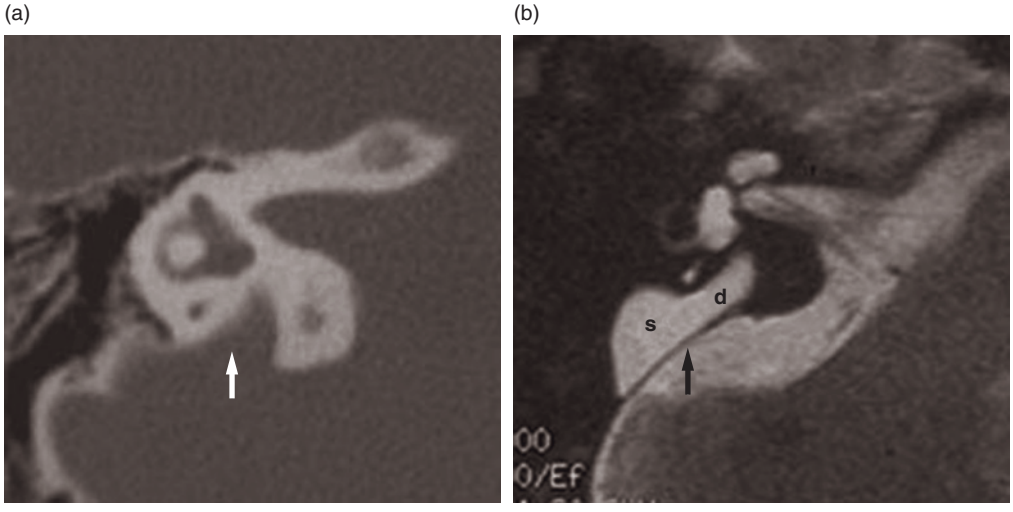

Figure 5.31 Axial CT (a) and T2-weighted MR (b) images. CT shows dilation of the vestibular aqueduct (white arrow), while enlargement of the endolymphatic duct (d) and sac (s) is demonstrated by MR. The dura (black arrow) separates the sac and subarachnoid spaces. Patient with Pendred syndrome.

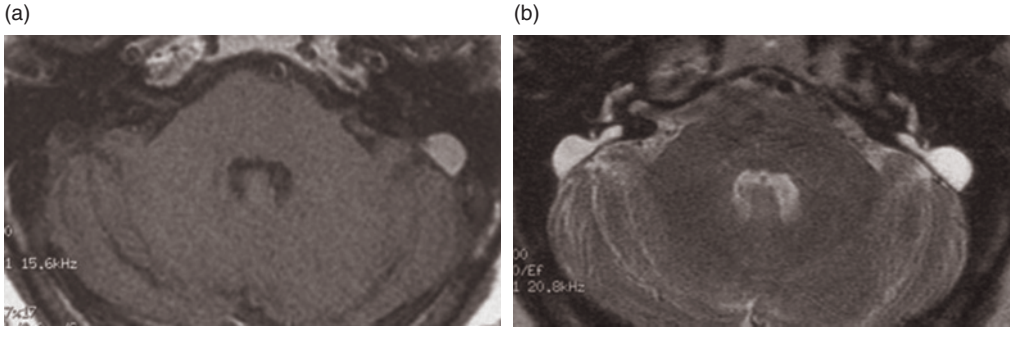

Figure 5.32 T1-weighted (a) and T2-weighted (b) axial MR images. Bilateral dilation of the endolymphatic duct and sac. The high hyperintensity of the left sac on both T1 and T2-weighted images could be due to protein-rich and hyperosmolar endolymph.

documented by imaging, nor it has been implicated as an actual cause of perilymph/cerebrospinal fluid fistula.[58]

IAC malformations are described as absent, hypoplastic (Figures 5.34 and 5.35) or enlarged[47] (Figure 5.36). The IAC can be dilated and bifid (Figure 5.37). When the diameter of the IAC meatus is smaller than 2–2.5 mm, congenital absence of the cochlear nerve should be suspected.[59] However, absence of the 8th nerve has been demonstrated in patients with normal-sized IAC.[60] Absence of the cochlear nerve occurs at the 5th week of gestation; it is due to a failure of development of the cochleo-vestibular nerve and connection with the inner ear, meanwhile the facial nerve is normally developed.[60] It can be very difficult to distinguish the facial nerve and the three branches of the vestibulocochlear nerve inside a stenotic IAC, so that the nerves should be demonstrated in the cerebellopontine angle. The best way to visualise

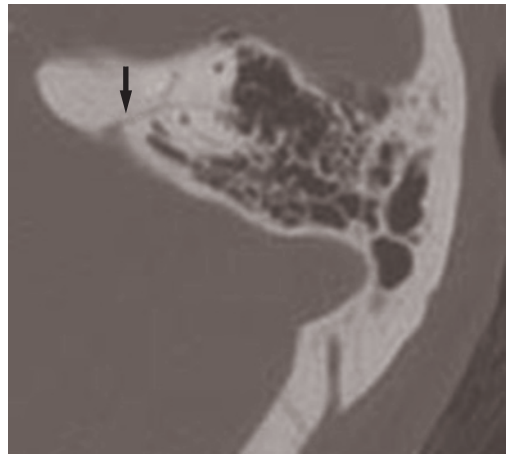

Figure 5.33 Axial CT scan. Slight dilation of the cochlear aqueduct (arrow).

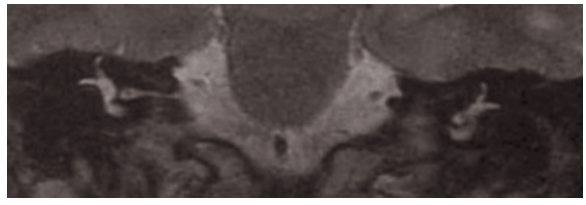

Figure 5.34 Coronal T2-weighted MR image. Malformations of both internal auditory canals: stenosis on the right and absence on the left.

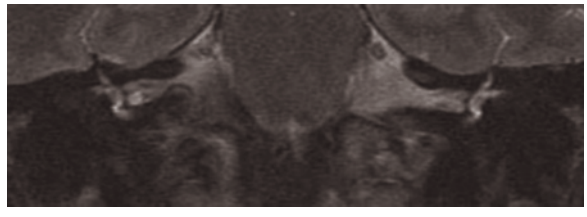

Figure 5.35 Coronal T2-weighted MR image. Stenosis of the right internal auditory canal.

the nerves in the cerebellopontine angle is to evaluate T2-weighted images perpendicular to the nerves and IAC meatus acquired through sequences such as 3D CISS or FIESTA.[3] (Figure 5.5). Both the narrowing and the dilation of the IAC have been described in association with other inner-ear abnormalities; in particular, dilation has been found in cases of common cavity, IP-I and IP-II.[47]

Accurate imaging of inner-ear malformations must be performed in candidates for cochlear implants. Clinical studies have found that patients with minor anomalies (such as an EVA or IP-II) may be implanted with standard techniques; hearing results are approximately equal to those with normal imaging.[61] Ganglion cells in humans are found in the lower 1.5 turns of the cochlea; as a result, if the basal part of the modiolus is present, the likelihood of the presence of the spiral ganglions and nerve endings is much greater in IP-II than in IP-I.[47] On the other hand, patients with major anomalies (such as common cavity) require specialised surgical

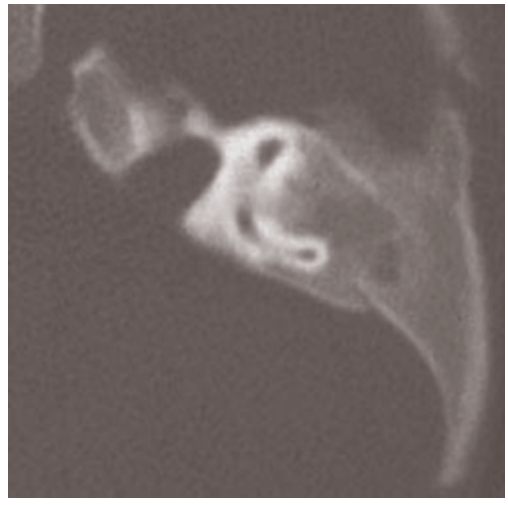

Figure 5.36 Axial CT scan. Dilation of the internal auditory canal (reprinted from Calzolari F. et al. Malformazioni dell' orecchio nelle anomalie congenite cranio-facciali. Rivista di Neuroradiologia 2003; 16: 411–420 [36]. With permission of Edizioni del Centauro).

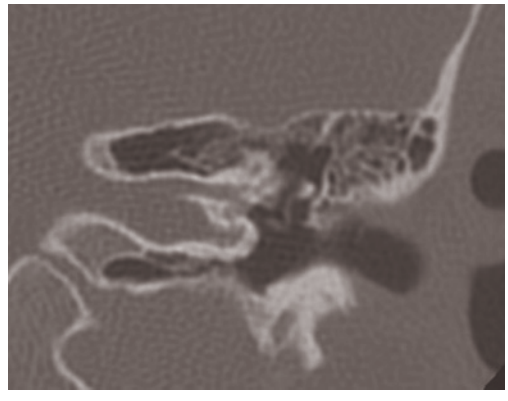

Figure 5.37 Coronal CT scan. Left internal auditory canal enlarged and bifid.

techniques, and the outcome can be variable. Otherwise, bilateral agenesis of the inner ear (Michel deformity) and bilateral absence of the cochlear nerve represent absolute contraindications to an implant.[3,59,61]

Measurement techniques for inner-ear structures using CT have recently been suggested for precise diagnosis of inner-ear anomalies. For instance, quantitative measurements of the cochlea may increase the detection of cochlear hypoplasia compared with relying on simple visual inspection.[46]

Congenital abnormalities of the inner ear have also been described in connection with cerebrospinal fluid leaks and/or recurrent meningitis. The possible route of infection in patients with dysplastic inner-ear structures is an abnormal pathway connecting the cerebrospinal fluid with the middle ear via the perilymphatic spaces of the inner ear. Patent cochlear aqueduct or small defects in the fundus of the IAC are possible explanations for oozing. Moreover, when the basal turn of the cochlea is dilated and communicates with the vestibule, there is a risk of

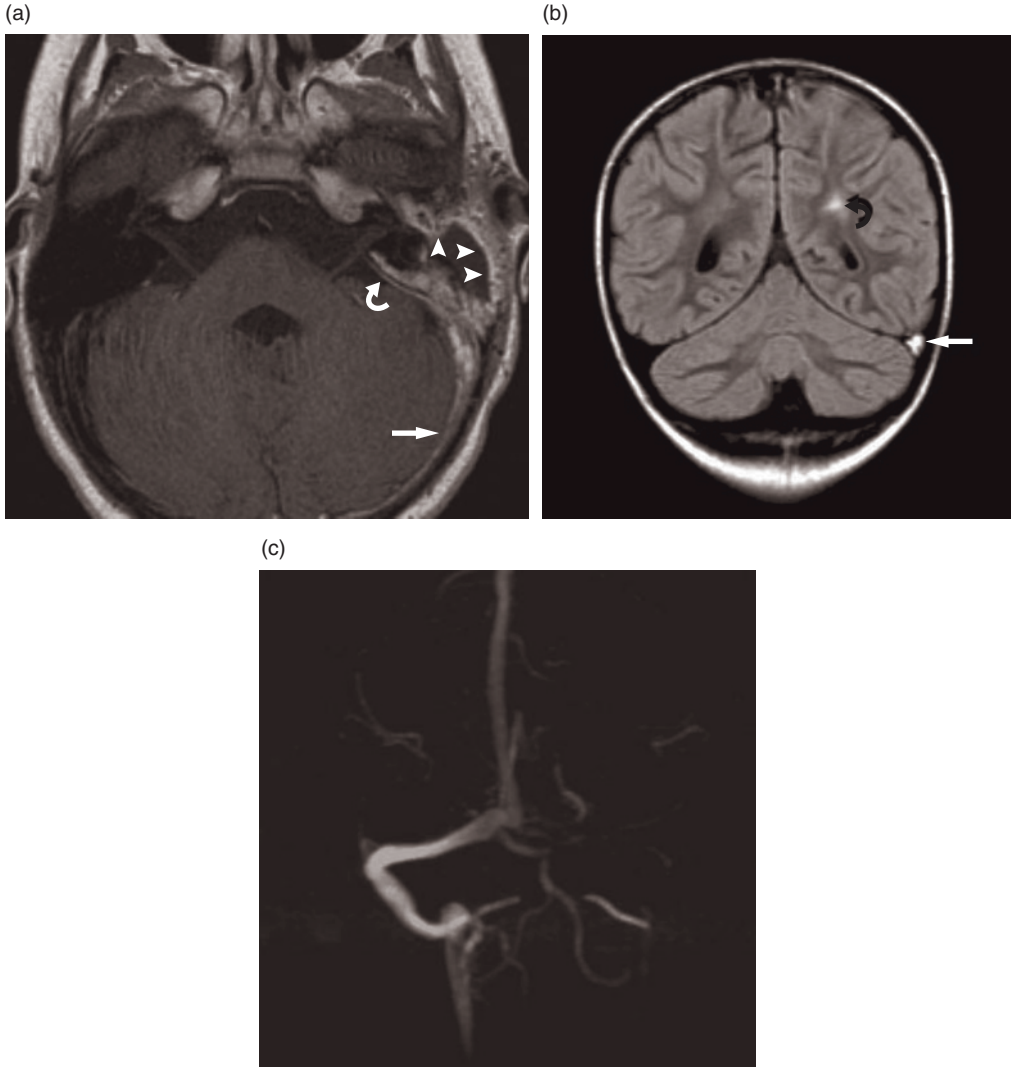

Figure 5.38 Intracranial complications of oto-mastoiditis. (a) Axial T1-weighted MR image with contrast. Enhancement of the wall of the petrous bone (arrowheads) and leptomeninges of the posterior fossa (curved arrow), suggesting inflammatory tissue and meningitis. Hyperintensity of the transverse sinus suggesting thrombosis (straight arrow). (b) Coronal FLAIR T2-weighted image. Sinus thrombosis (straight arrow) and cerebritis (curved arrow). (c) 2D-TOF MR angiography. Absence of blood flow into the left transverse sinus due to thrombosis.

recurrent meningitis as well as spontaneous cerebrospinal fluid leak because of possible communication with the subarachnoid spaces in the IAC.[24]

Cerebrospinal fluid leakage occurs more often during otological surgery: congenital anomalies discovered during preoperative imaging studies cannot only be the cause of SNHL but also can increase the surgical risk for having a 'gusher-ear', e.g. during electrode insertion in implant surgery.[59] MR represents the technique of choice in order to diagnose meningitis or other intracranial complication such as sinus thrombosis (Figure 5.38).

VASCULAR ABNORMALITIES

Correct imaging representation and analysis of normal and abnormal vascular structures close to the ear may explain some clinical pictures such as tinnitus, pulsation, SNHL or other symptoms related to neuro-vascular conflict. Moreover, it is mandatory to plan surgery and to avoid serious or even lethal haemorrhages during ear surgery. Furthermore, description of anomalies of calibre and course of the arterial and venous structures may influence, together with other factors, the choice of the side for a cochlear implant.[6]

For imaging of the main petrous vessels, CT and MR are the techniques usually employed. High-resolution CT shows in detail skull base structures, for instance the carotid canal or the jugular fossa. MR imaging can easily demonstrate the relationships between arterial or venous vessels and adjacent anatomical structures. MR angiography (MRA) is a non-invasive technique: through flow-dependent 3D or 2D time-of-flight sequences the main arteries and veins may be displayed, without need of contrast enhancement. Actually, the use of digital subtraction angiography is limited to clarify dubious cases or for endovascular therapeutic procedures.

An aberrant internal carotid artery (ICA) in the middle ear is a congenital finding easily diagnosed by CT (Figure 5.39). If the diagnosis is not made before middle-ear surgery, haemorrhage, stroke or death may occur.[62,63] MRA may be useful to confirm the presence of an anomalous vessel in the middle ear.[64,65]

The interval between the cochlea and the ICA varies widely amongst subjects. Mid-tone SNHL at audiometric examination may be a characteristic finding of absence of bone between the petrous ICA and the basal turn of the cochlea or the thin cochlea–carotid interval. Detailed CT evaluation of these structures may help to prevent inadvertent carotid canal penetration, in particular during cochlear implant surgery.[66]

A duplicated ICA is a rare congenital variant; the clinical symptoms and signs are often non-specific or absent. If ear, neck or tonsil surgery is planned, the knowledge of this variant is very important because a misdiagnosis could have disastrous consequences.[63,67] However, MRA may be useful to confirm or to exclude the carotid canal duplication suspected by CT.[6]

Other rare congenital anomalies are agenesis and hypoplasia of the ICA. MR and MRA may not be able to distinguish between agenesis and hypoplasia and may suggest acquired

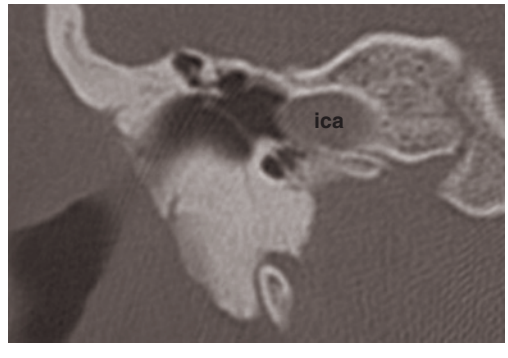

Figure 5.39 Coronal CT scan. Protrusion of the internal carotid artery (ica) into the middle ear (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

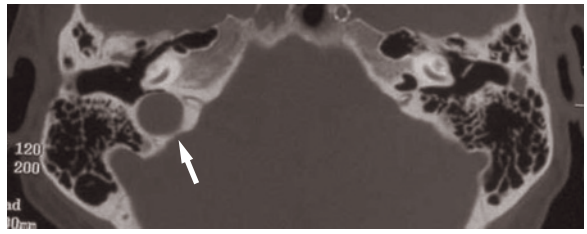

Figure 5.40 Axial CT scan. Large jugular bulb (arrow) on the right side (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

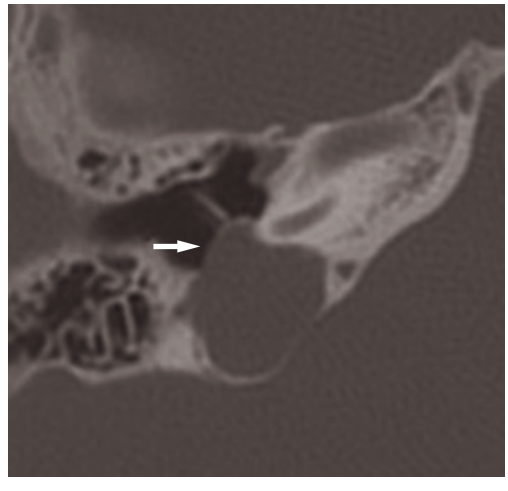

Figure 5.41 Axial CT scan. Tympanic dehiscence of the jugular bulb (arrow).

stenosis or occlusion. CT of the base of the skull provides the final proof for the diagnosis of ICA agenesis or hypoplasia if the carotid canal is absent or thin.[68,69]

A persistent stapedial artery is another vascular anomaly due to failure of regression of a vessel transiently present in normal fetal life and connecting the future external carotid artery to the ICA; it may be isolated or associated with an aberrant ICA. If the stapedial artery persists, the middle meningeal artery arises from it and the foramen spinosum is consequently absent. CT findings, such as a small canaliculus exiting the carotid canal (in cases in which aberrant ICA is not present), the presence of a linear soft tissue crossing the middle ear over the promontory, an enlarged facial nerve canal or a separate parallel canal, may suggest the presence of persistent stapedial artery. It can cause tinnitus and hearing loss. In the presence of this anomaly, stapes surgery is complicated because the stapedial artery passes through the obturator foramen of the stapes.[63,70]

Amongst the venous anomalies, a large jugular bulb is undoubtedly the most frequent (Figure 5.40). Abnormal dilation of the jugular vein and its eventual tympanic dehiscence (Figure 5.41) may influence the choice of the side for cochlear implantation.

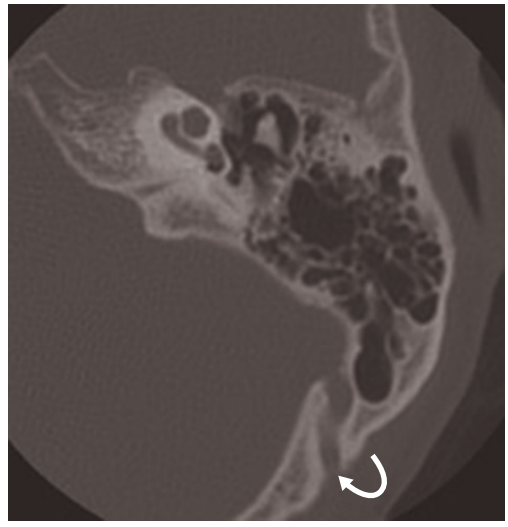

Figure 5.42 Axial CT scan. Large mastoid emissary vein (arrow).

The mastoid emissary vein is a venous variant which should not be mistaken for the lambdoid suture (Figure 5.42). Its presence can make difficult a retro-mastoid surgical approach or the placing of the external component of the cochlear implant.[59]

The petrosquamosal sinus is another variant characterised by an embryonic venous remnant, which usually regresses during fetal life. It courses over the extreme lateral part of the petrous bone and connects the lateral sinus with the retromandibular vein, e.g. the internal jugular vein with the external jugular vein. Both CT and MR can demonstrate the presence of this vein. Venous drainage through the petrosquamosal sinus could have a potential role in the spread of infection of the EAC and in complications of middle-ear surgery, such as bleeding or thrombophlebitis.[71]

EAR ABNORMALITIES IN CRANIOFACIAL DEFORMITIES AND GENETIC SYNDROMES

Many syndromes with associated ear malformations detectable on CT and MR have been reported. Craniofacial syndromes are described in detail in Chapter 7. In these cases, diagnostic imaging is mandatory for the coordination of treatment, both aesthetic and functional, by the otorhinolaryngologist, maxillo-facial surgeon and eventually neurosurgeon. The majority of these patients must be examined not only with CT of the external and middle ear, but also by CT and MR of the inner ear, 3D-CT of the skull and finally MR for the study of the temporomandibular joint and for searching for brain malformation.[32]

Amongst the most frequent syndromic craniofacial deformities with associated ear abnormalities are Treacher-Collins syndrome, Goldenhar syndrome, branchio-oto-renal (BOR) syndrome, Apert syndrome and Crouzon syndrome.

Treacher-Collins syndrome is an inherited disorder with autosomal dominant transmission characterised by anomalies of the structures developing from the first and second branchial

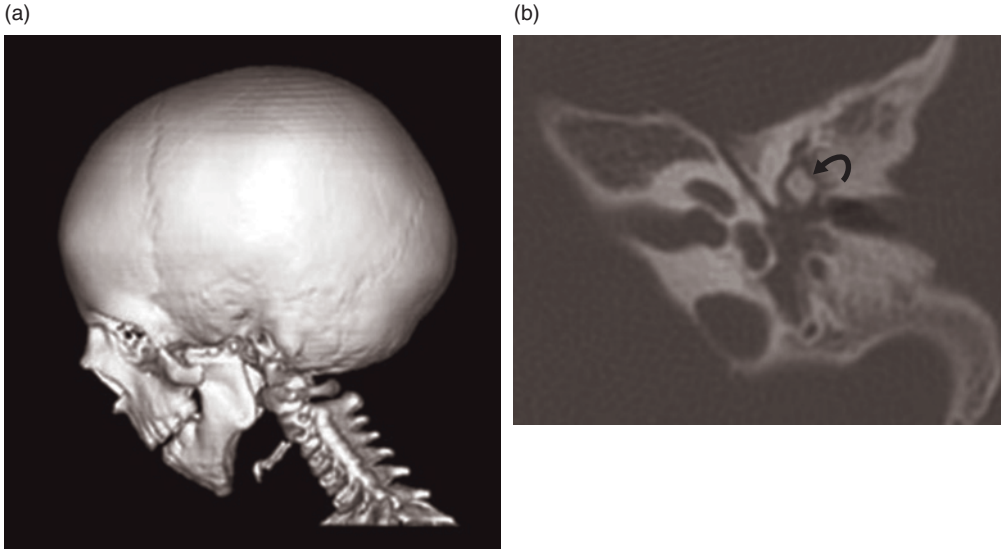

Figure 5.43 A 4-year-old male with Treacher-Collins syndrome. (a) Skull 3D-CT: incomplete zygomatic arch, mandibular hypoplasia and condilar dysplasia, mastoid hypoplasia. (b) CT axial scan: stenosis of the external auditory canal, small tympanic cavity, fusion of the malleoincudal joint (arrow).

arches. In general, there is complete penetrance and variable expressivity of the trait.[72] Abnormalities are frequently bilateral and symmetrical, in particular external and middle-ear malformations (Figure 5.43); inner-ear malformations are exceptional.[6,32,72,73]

Goldenhar syndrome, also known as oculo-auriculo-vertebral dysplasia, is a wide spectrum of congenital anomalies mainly affecting the first and second branchial arches. Its occurrence is predominantly sporadic, but inherited forms, both autosomal recessive and dominant, have been described.[74–76] Various associated malformations can be present: ocular (epibulbar dermoids, coloboma, microphthalmia), facial (median facial cleft), cranial (lipoma and dermoids), spinal (vertebral fusion and spina bifida), cardiovascular (ventricular septal defects, mitral stenosis) and visceral (portal vein absence). Amongst ear abnormalities, not only frequent external and middle-ear malformations, but also inner-ear malformations have been reported; in particular, a case of semicircular canal malformation (absence of the common crus) has been described.[77] Eustachian tube anomalies have been described.[78] Congenital facial nerve palsy is reported in a case of Goldenhar syndrome.[74]

BOR syndrome has an autosomal dominant transmission and comprises preauricular pits, branchial fistulas and ear and renal abnormalities. In some families, the phenotypic expression is limited to branchial anomalies, without renal dysplasia (branchio-otic syndrome); in other families, branchial and renal anomalies occur without hearing impairment.[79] Concerning ear abnormalities, cochlear malformations (Figure 5.22) and EVA are frequently diagnosed, but hypoplasia of the tympanic cavity and ossicular chain abnormalities may also be present.[6,80,81]

Apert syndrome, or acrocephalosyndactyly type 1, is a congenital craniofacial synostosis associated with syndactyly of the hands and feet. Many other multi-organ malformations may be associated. Most cases are sporadic, but autosomal dominant transmission is possible.

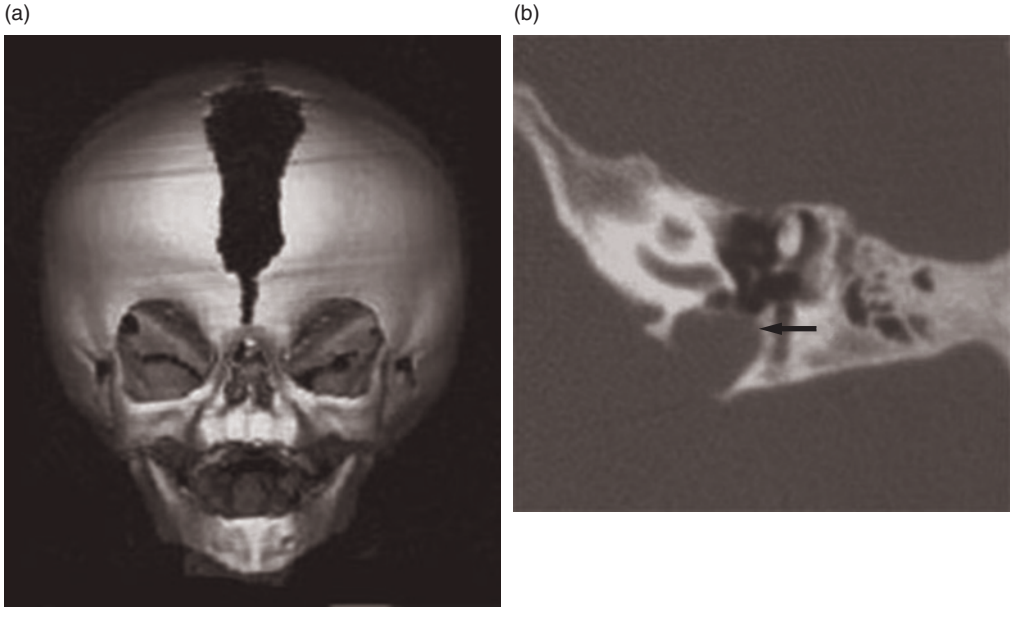

Figure 5.44 Crouzon syndrome. (a) Skull 3D-CT: craniosynostosis with abnormal dilation of the bregmatic fontanel and metopic suture. (b) Axial CT scan: tympanic dehiscence of the jugular bulb (arrow) (reproduced from Calzolari F., 2006,[6] with permission of Omega Edizioni).

Associated ear abnormalities include malformations of the auricle and the eustachian tube, stenosis of the EAC and ossicular chain dysplasia, in particular stapes footplate fixation. Wide cochlear aqueduct has also been described.[82]

Crouzon syndrome is a severe craniosinostosis with hypoplasia of the maxilla, hypertelorism and proptosis. It may be sporadic or autosomal dominant transmitted. Stenosis and atresia of the EAC, tympanic hypoplasia and dysplasia of the ossicles are frequent. Hydrocephalus and absent septum pellucidum may coexist.[32,83] Tympanic dehiscence of the jugular bulb is frequent in Crouzon syndrome (Figure 5.44). These patients have a distorted nasopharynx, which frequently leads to middle-ear secretions and necessitates myringotomy. Consequently, patients with Crouzon syndrome are at risk for inadvertent puncture of the jugular bulb during myringotomy; CT performed previously may be helpful in preventing this complication.[84]

External and middle-ear abnormalities are typical in patients with *hemifacial microsomia*, a disorder characterised by microtia, macrostomia and mandibular hypoplasia; neither facial nor ear abnormalities seem to correlate with the type or degree of hearing loss.[85,86]

Ear malformations have been described in many other genetic syndromes. For instance, radiological abnormalities of the external, middle and inner ear were demonstrated in *Down syndrome (trisomy 21)*[87] and *Klippel-Feil syndrome* (short neck, low occipital hairline, cervical and thoracic vertebrae dysplasia).[88,89] Clumping of the ossicle, stapes fixation and sclerosis of the footplate are described in *cleido-cranial dysplasia*, a rare autosomal dominant skeletal dysplasia affecting both membranous and endochondral bone formation.[90] Abnormalities of the middle and inner ear may be found in other congenital syndromes such as *CHARGE*

association (Coloboma, congenital Heart disease, Atresia of choanae, mental Retardation and/or central nervous system anomalies, Genital hypoplasia and Ear anomalies),[91,92] *Noonan syndrome* (short stature, facial dysmorphism, webbed neck, heart defects)[93,94] and *VATER syndrome* (Vertebral, Anal, TracheoEsophageal, radial and Renal defects).[95] CT and X-ray findings of inner-ear deformity were described in *Wildervanck syndrome* (deafness, Klippel-Feil deformity and ocular motility disturbance – the so-called Duane retraction syndrome).[96,97] Enlargement of the vestibular aqueduct and the vestibule, narrowing of the IAC and hypoplasia of the modiolus were detected by CT in patients with *Waardenburg syndrome*, an autosomal-dominant syndrome characterised by dystopia canthorum, eyebrow hyperplasia, iris heterochromia, white forelock and sensorineural hearing loss.[98,99]

Finally, CT and MR findings of inner-ear malformations are typical in *Pendred syndrome*, an autosomal recessive disorder characterised by goitre and progressive sensorineural deafness.[100] Pendred syndrome is the only known genetic disorder with dilation of the vestibular aqueduct (Figure 5.31) aside from BOR syndrome.[101] Moreover, mutations in the SLC26A4 gene, coding for the protein pendrin, have been implicated in the pathophysiology of both Pendred syndrome and non-syndromic EVA.[102]

CONCLUSION

Nowadays, newborn hearing screening enables early identification of deafness. However, this is of little importance if it is not combined with quality services that can provide children and families with the potential advantages of significantly earlier diagnosis.[103] A newborn with a congenital malformation arouses a great anxiety in his or her parents. Furthermore, if this defect is of the craniofacial region, the abnormality cannot be hidden from the world. So, the parents want to know not only why their child is deaf or affected by a congenital defect, but also what can be done immediately to correct the hearing defect or evident abnormality. The paediatrician plays an extremely important initial role in providing valuable reassurance, direction and surveillance for deaf children with auricular malformations. On the other hand, from the otorhino-laryngologist's point of view, clinical, audiological and diagnostic imaging examinations are essential to manage the child and his or her parents.[16]

Early and complete imaging of ear malformations is necessary not only to give an indication of the eventual aesthetic and audiological therapy, but also to plan surgical treatment. CT represents the imaging technique of choice for showing external and middle-ear abnormalities; CT and MR are complementary in the study of the inner ear.

Imaging shows that external and middle-ear congenital malformations are frequently associated, probably related through a common embryological origin. However, children with microtia and atresia may have severe inner-ear malformations despite the fact that the outer, middle and inner ear develop from embryologically separate structures. Finally, it should be kept in mind that outer-, middle- and inner-ear malformations can be found also in children with normal auricles.

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6 Genetics of hearing loss

A.P. Read

INTRODUCTION

About half of all profound childhood hearing loss, and a significant but unknown proportion of milder or later onset loss, is caused by mutation of a single gene (different in different cases).[1] Remarkable progress has been made in the last 15 years in mapping and identifying the genes involved. I will first briefly discuss the methods that have made this possible, then summarise the data for the more significant genes, and finally consider the implications of this scientific advance for the working clinician.

METHODS: HOW THE GENES ARE IDENTIFIED

This brief overview describes some of the tools and limitations of current approaches. For more detailed discussion of methods, the reader should consult a suitable textbook.[2,3] References are given in the succeeding discussions to the appropriate section of these books.

There is no universal correct way to identify the gene underlying a genetic disease. However, all the many possible methods (summarised in Strachan and Read,[2] fig. 14.1, p. 296) converge on testing a *candidate gene* for mutations. One way or another, one gene is chosen from amongst the 24,000 or so in the human genome. If it is the correct gene, then people with the disease should have mutations in that gene. DNA from patients is analysed to see if the sequence of the gene in the patients differs in any way from the normal sequence. Numerous laboratory techniques are available to answer that question (Strachan and Read,[2] section 18.3; Read and Donnai,[3] chapter 4), but problems are still common. None of the techniques is 100% sensitive, and when a deviation from the normal sequence has been found, it may be difficult to know whether the change is pathogenic or just a coincidental neutral variant (a 'polymorphism'). For that reason it is desirable to have a panel of unrelated patients available for mutation testing. If a good proportion of the patients have a mutation, and if the mutations include several different sequence variants, none of which is found on testing, say, 100 healthy controls, then it is highly likely that the correct gene has been identified. Further confirmation requires functional studies. These might be conducted in a cell-free system, in cultured cells or in genetically engineered mice.

How is the candidate gene chosen? There are many different ways, but almost always the first step is to narrow down the choice by defining the approximate chromosomal location of the gene (mapping it). This is done by studying large families in which the disease is segregating. We need DNA samples from a good number of people (typically 20–30) who could have inherited either the disease gene or its normal allele from their parents, and where we know

by clinical examination which possibility in fact happened. We then study DNA markers (common non-pathogenic DNA variants) to try to find one whose transmission through the family tree parallels the transmission of the disease gene. This is called linkage analysis (see Strachan and Read,[2] section 13.3, or Read and Donnai,[3] chapter 9). If a marker is found that satisfies the statistical test for linkage (measured by the lod score), then the disease gene must be located on the same chromosomal segment as the marker.

Given large families that are good samples to study, this method typically narrows the candidate region down from 3,000 million base pairs of DNA (the size of the whole human genome) down to 1–5 million base pairs. Such a region might contain anything from a handful to a score or more genes. Public databases list the genes in each chromosomal region, and with the completion of the Human Genome Project, the lists are fairly reliable and complete. It then remains to prioritise candidates for mutation testing from amongst the genes on the list. Clues include the temporal and spatial pattern of expression (presumably the correct gene will be switched on in the inner ear, whether or not it is functional elsewhere as well; genes responsible for developmental defects should be active at the appropriate stage of embryonic development). Genes come in families, and another clue might be a relationship to a gene already known to be implicated in a similar phenotype, in man or a model organism. Knowledge of the biochemical function of the gene may also be relevant, though our ignorance of cell biology means that it is often very difficult to guess the clinical result of mutations in a gene from such knowledge.

From the foregoing, it follows that progress in identifying the genes underlying genetic hearing loss depends crucially on collaboration between clinicians and laboratory workers. However clever the molecular geneticists are, they can achieve nothing unless their clinical colleagues can identify good large families, and collect their DNA for linkage and mutation analysis. A particularly important role of the clinicians is in identifying heterogeneity. Undetected heterogeneity in a collection of families can make linkage analysis almost impossible, whilst identification of subtle distinguishing features can point the way to identifying new disease genes. Finally, alert clinicians picking out patients with chromosomal abnormalities as well as a known Mendelian disease have often provided the vital clue to launch successful gene identification projects. In all these ways, one of the pleasures of working in clinical molecular genetics is the opportunities it provides for fruitful collaboration between insightful clinicians and cutting-edge scientists.

SOURCES OF INFORMATION

This is a fast-moving area, and for up-to-date information, the reader should consult one of the excellent Internet resources.

- For general information on simply inherited diseases and the underlying genes, OMIM (Online Mendelian Inheritance in Man)[4] is the first choice. Searching this database for a word or phrase will return a series of numbers, each pointing to one entry, e.g. Waardenburg syndrome Type 1 is 193500. Clicking on the number brings up a brief clinical description, a more detailed summary of the genetics, lists of references and links to other Internet resources. OMIM entries are reliable and generally up to date, but note that new material is usually simply added on to the end of the previous content of a section, so the early parts of an entry for a disease may have been written many years ago.

- Specific information on hereditary hearing loss is collected on the Hereditary Hearing Loss Homepage,[5] a resource maintained jointly by G Van Camp (Antwerp) and R Smith (Iowa). This excellent resource lists all the identified and mapped genes and has links to many other reliable Internet resources.

PROGRESS IN IDENTIFYING THE GENES: SYNDROMIC HEARING LOSS

Gorlin, Toriello and Cohen[6] describe 427 syndromes in which hearing loss is a regular or occasional feature. Most are very rare. Table 6.1 summarises a few of the more frequent syndromes. For further genetic information on all these conditions, consult the OMIM database.[4]

The points of note are discussed in succeeding sections.

Branchio-oto-renal syndrome

This clinically and genetically heterogeneous syndrome has been split into branchio-oto-renal (BOR) and branchio-otic (BOS) syndromes, although both may be caused by mutations in the

Table 6.1 Examples of syndromic hearing loss.

Condition	Locus	Chromosomal Location	Gene (where known)
Alport	*COL4*	Xq22	*COL4A5*
		2q36-q37	*COL4A3, COL4A4*
Branchio-oto-renal	*BOR1*	8q13.3	*EYA1*
	BOR2	19q13.32	*SIX5*
Jervell and Lange-Nielsen	*JLN*	11p15	*KCNQ1*
Neurofibromatosis 2	*NF2*	22q	*NF2 (Merlin)*
Pendred	*PDS*	7q21-q34	*SLC26A4*
Treacher Collins	*TCOF1*	5q32	*TCOF1*
Usher Type 1	*USH1B*	11q13.5	*MYO7A*
	USH1C	11p15.1	*USH1C*
	USH1D	10q22.1	*CDH23*
	USH1E	21q21	*?*
	USH1F	10q21-q22	*PDCH15*
	USH1G	17q24-q25	*SANS*
Usher Type 2	*USH2A*	1q41	*USH2A*
	USH2B	3p23-p24.2	*?*
	USH2C	5q14.3-q21.3	*VLGR1*
Usher Type 3	*USH3*	3q21-q25	*Clarin*
Waardenburg Type 1	PAX3	2q35	PAX3
Waardenburg Type 2	*MITF (15%)*	3p14	*MITF*
	Unknown (85%)	*?*	*?*
Waardenburg Type 3	*PAX3*	2q35	*PAX3 (+/– or –/–)*
Waardenburg Type 4	*EDNRB, EDN3*	13q22, 20q13	*EDNRB, EDN3,*
	SOX10	22q	*SOX10*
X-linked with dystonia	*DFN1*	Xq22	*DDP (TIMM8A)*
X-linked with gusher	*DFN3*	Xq13-q21	*POU3F4 (Brain 4)*

See text for descriptions of some of the genes. See a human genetics textbook, for example section 2.4.1 of Strachan and Read[2] for description of the way chromosomal locations are named.

EYA1 gene on chromosome 14. *EYA1* is the human homologue of the *eyes absent* gene in the *Drosophila* fruit fly – this is an interesting example of the way that a gene can acquire rather different functions during the course of evolution (although *EYA1* mutations have also been found in a few humans with eye abnormalities). A second BOR gene is *SIX5* on chromosome 19. Both these genes encode transcription factors, that is, DNA-binding proteins that control the expression of other genes. Other BOS loci map to chromosomes 1 and 14.

Jervell and Lange-Nielsen syndrome (JLN)

JLN is one of many examples of hearing loss caused by defects in ion transport. It is caused by defects in the IKs potassium channel. The mutations can be in the genes encoding either the alpha or beta subunits (*KCNQ1* on chromosome 11p15 and *KCNE1* on 22q22, respectively). People with no functional KCNQ1 ion channels have JLN, people with 50% of the normal level are clinically normal, whilst people with a level somewhere between 0 and 50% have a heart problem (long QT interval) but normal hearing. JLN mutations simply abolish the function of the gene product. Thus homozygotes have JLN but heterozygotes are normal, and JLN is recessive. Some mutations result in the production of an altered KCNQ1 protein that is not only non-functional, but also partly blocks the function of any normal protein present (a dominant negative effect). People who are heterozygous for such a mutation have the dominant Roman-Ward long QT syndrome, but normal hearing.

Neurofibromatosis 2 (NF2)

All vestibular schwannomas (VS), whether sporadic and unilateral or part of NF2, originate from a cell that has lost both functioning copies of the *NF2* gene. Sporadic VS happens when by pure chance an originally normal cell suffers two successive mutations. This is a rare piece of bad luck, hence sporadic VS are unilateral and usually seen in older people (who have had more time to accumulate mutations). People who inherit one mutant *NF2* gene are perfectly normal because cells can function normally with a single intact copy of this gene. But every cell carries the mutation, and only a single acquired mutation is needed to convert a cell into the precursor of a VS. Given the large number of potential target cells, this is a highly likely occurrence. Hence, NF2 affects younger people and often produces bilateral or multifocal tumours. NF2 is a classic example of Knudson's two-hit mechanism of hereditary tumours (see section 17.4 of Strachan and Read[2] or chapter 12 of Read and Donnai[3]).

Pendred syndrome

The *SLC26A4* gene encodes pendrin, a protein that is involved in transport of chloride and iodide ions. The hearing loss reflects the general importance of ion transport to cochlear function, whilst the iodide transport defect explains the goitre present in this syndrome. Some *SLC26A4* mutations cause the non-syndromic *DFNB4* hearing loss; the reason for this difference is not clear.

Treacher Collins syndrome

The *TCOF1* gene encodes a protein ('treacle') that is involved in nucleolar function. Almost all described mutations are predicted to result in premature chain-termination during protein

synthesis, causing that copy of the gene to produce no functioning protein. How a 50% dosage of treacle protein produces the clinical features of Treacher Collins syndrome is unknown. The great clinical variability, even within families, is typical of conditions caused by such dosage sensitivity ('haploinsufficiency'). Branchio-oto-renal and Waardenburg syndromes provide further examples.

Usher syndrome

Usher syndrome has turned out to be remarkably heterogeneous at the molecular level, with so far six different loci implicated in Type 1 syndrome, three in Type 2 syndrome and one in Type 3 syndrome. This implies that many different molecules have roles in both cochlear and retinal function, so that mutations affect both organs. Interestingly, for four of the genes, mutations can cause either Usher Type 1 syndrome or non-syndromic hearing loss (Table 6.2). This suggests that the cochlea is less tolerant than the retina of mild functional deficits in the encoded proteins.

Waardenburg syndrome (WS)

The label 'Waardenburg syndrome' is applied to a heterogeneous collection of auditory-pigmentary syndromes, all of which have their origin in a dysfunction of melanocytes (see Read and Newton[7] for a review). Apart from their role in pigmentation, melanocytes also form the pigmented intermediate cells of the stria vascularis, and in their absence there is no hearing. Four types of WS are usually listed.

- Type 1 with dystopia canthorum (outward displacement of the inner canthi of the eyes) is caused by mutations in *PAX3*, which encodes a homeodomain-containing transcription factor expressed in the embryonic neural crest (the tissue of origin of melanocytes). WS1 is dominant, patients are heterozygous, and the pathogenic mechanism is haploinsufficiency.
- Type 2 WS is a melanocyte-specific disturbance, caused in some cases by mutations in the *MITF* transcription factor gene (a master gene controlling differentiation of melanocytes), and in other cases by as yet unidentified gene(s). Claims of linkage to 8p23 or deletion of the *SNAI2* gene were mistaken.
- Type 3 WS has the features of Type 1 with additionally limb abnormalities. Most 'WS3' patients have mild muscular hypoplasia of the arms and/or contractures of some joints. This is an occasional variant presentation of WS1, and these patients are heterozygous for *PAX3* mutations similar to those seen in WS1. In very rare cases, patients have a much more severe phenotype with extreme depigmentation, severe dystopia canthorum and amyoplasia of the arms and shoulders. These patients are homozygous for *PAX3* mutations, and in at least two cases WS1 was documented in both parents.
- Type 4 WS, or Waardenburg-Shah syndrome, has the features of WS2 plus Hirschsprung disease. All the affected tissues are derived from the neural crest and WS4 comprises a heterogeneous set of severe neurocristopathies. Three causative genes have been identified. Mutations in endothelin 3 and its receptor EDNRB usually cause isolated Hirschsprung disease in heterozygotes but WS4 in homozygotes; mutations in the transcription factor SOX10 can cause WS4 in heterozygotes (sometimes with additional neurological problems).

Table 6.2 Non-syndromic hearing loss.

Locus	Chromosomal location	Gene (where known)	Identical loci
Autosomal dominant loci (54 described)			
DFNA1	5q31	*DIAPH1*	
DFNA2	1p34	*GJB3, KCNQ4*	
DFNA3	13q12	*GJB2, GJB6*	*DFNB1*
DFNA4	19q13	*MYH14*	
DFNA5	7p15	*DFNA5*	
DFNA6	4p16.3	*WFS1*	
DFNA7	1q21-q23	?	
DFNA8	11q22-q24	*TECTA*	= *DFNA12*
DFNA9	14q12-q13	*COCH*	
DFNA10	6q22-q23	*EYA4*	
DFNA11	11q12.3-q21	*MYO7A*	*DFNB2, USH1B*
DFNA12	11q22-q24	*TECTA*	*DFNA8, DFNB21*
DFNA13	6p21	*COL11A2*	*DFNB53*
DFNA14	4p16	*WFS1*	= *DFNA6*
DFNA15	5q31	*POU4F3*	
DFNA16	2q24	?	
DFNA17	22q	*MYH9*	
DFNA18	3q22	?	
DFNA19	10cen	?	
DFNA20	17q25	*ACTG1*	= *DFNA26*
DFNA21	6p21	?	
DFNA22	6q13	*MYO6*	*DFNB37*
DFNA23	14q21-q22	?	
DFNA24	4q	?	
DFNA25	12q21-q24	?	
DFNA26	17q25	*ACTG1*	= *DFNA20*
DFNA27	4q12	?	
DFNA28	8q22	*TFCP2L3*	
DFNA29	Reserved		
DFNA30	15q25-q26	?	
DFNA31	6p21.3	?	
DFNA32	11p15	?	
DFNA33	Reserved		
DFNA34	1q44		
DFNA35	Reserved		
DFNA36	9q13-q21	*TMC1*	*DFNB7/11*
DFNA37	1p21	?	
DFNA38	4p16.3	*WFS1*	= *DFNA6 / 14*
DFNA39	4q21.3	*DSPP*	
DFNA40	16p12	?	
DFNA41	12q24-qter	?	
DFNA42	5q31.1-q32	?	
DFNA43	2p12	?	
DFNA44	3q28-q29	*CCDC5O*	
DFNA45	Reserved		

Table 6.2 *Continued*

Locus	Chromosomal location	Gene (where known)	Identical loci
DFNA46	Reserved		
DFNA47	9p21-p22	?	
DFNA48	12q13-q14	*MYO1A*	
DFNA49	1q21-q23	?	
DFNA50	7q32	?	
DFNA51	9q21	?	
DFNA52	4q28	?	
DFNA53	14q11-q12	?	
DFNA54	5q31	?	
Autosomal recessive loci (67 described)			
DFNB1	13q12	*GJB2*	*DFNA3*
DFNB2	11q13.5	*MYO7A*	*USH1B, DFNA11*
DFNB3	17p11.2	*MYO15A*	
DFNB4	7q31	*SLC26A4*	*Pendred*
DFNB5	14q12	?	
DFNB6	3p14-p21	*TMIE*	
DFNB7	9q13-q21	*TMC1*	*= DFNB11, DFNA36*
DFNB8	21q22	*TMPRSS3*	*= DFNB10*
DFNB9	2p22-p23	*OTOF*	
DFNB10	21q22.3	*TMPRSS3*	*= DFNB8*
DFNB11	9q13-q21	*TMC1*	*= DFNB7, DFNA36*
DFNB12	10q21-q22	*CDH23*	*USH1D*
DFNB13	7q34-q36	?	
DFNB14	7q31	?	
DFNB15	3q21-q25 or 19p13?	?	
DFNB16	15q21-q22	*STRC*	
DFNB17	7q31	?	
DFNB18	11p14-p15.1	*USH1C*	*USH1C*
DFNB19	18p11	?	
DFNB20	11q25-qter	?	
DFNB21	11q22-q24	*TECTA*	*DFNA12*
DFNB22	16p12.2	*OTOA*	
DFNB23	10p11.2-q21	*PDCH15*	*USH1F*
DFNB24	11q23	*RDX*	
DFNB25	4p15.3-q12	?	
DFNB26	4q31	?	
DFNB27	2q23-q31	?	
DFNB28	22q13	*TRIOBP*	
DFNB29	21q22	*CLDN14*	
DFNB30	10p12.1	*MYO3A*	
DFNB31	9q32-q34	*WHRN*	
DFNB32	1p13.3-p22.1	?	
DFNB33	9q34.3	?	
DFNB34	Reserved		
DFNB35	14q24.1-q24.3	*ESRRB*	
DFNB36	1p36.3	*ESPN*	

Table 6.2 *Continued*

Locus	Chromosomal location	Gene (where known)	Identical loci
DFNB37	6q13	*MYO6*	*DFNA22*
DFNB38	6q26-q27	?	
DFNB39	7q11.22-q21.12	?	
DFNB40	22q	?	
DFNB41	Reserved		
DFNB42	3q13.31-22.3	?	
DFNB43	Reserved		
DFNB44	7p14.1-q11.22	?	
DFNB45	1q43-q44		
DFNB46	18p11.32-p11.31	?	
DFNB47	2p25.1-p24.3	?	
DFNB48	15q23-q25.1	?	
DFNB49	5q12.3–q14.1	*MARVELD2*	
DFNB50	12q23	?	
DFNB51	11p13-p12	?	
DFNB52	Reserved		
DFNB53	6p21.3	*COL11A2*	*DFNA13*
DFNB54	Reserved		
DFNB55	4q12-q13.2	?	
DFNB56	Reserved		
DFNB57	10q23.1-q26.11	?	
DFNB58	2q14-q21.2	?	
DFNB59	2q31.1-q31.3	*PJVK*	
DFNB60	5q22-q31	?	
DFNB61	reserved		
DFNB62	12p13.2-p11.23	?	
DFNB63	11q13.2-q13.32	?	
DFNB64	Reserved		
DFNB65	20q13.2-q13.32	?	
DFNB66	6p21.2-p22.3	*LHFPL5*	= *DFNB67*
DFNB67	6p21.2-p22.3	*LHFPL5*	= *DFNB66*
X-linked loci (5 described)			
DFN1	Xq22	*TIMM8A*	
DFN2	Xq22	?	
DFN3	Xq21.1	*POU3F4*	
DFN4	Xp21.2	?	
DFN6	Xp22	?	
DFN8	Reserved		
Mitochondrial mutations (many described)			
7445insC,		*TRNA*$^{SER(UCN)}$	
A1555G		*12S RNA*	

Note: Mutations in many different genes can cause non-syndromic hearing loss. Autosomal dominant loci are symbolised *DFNA1*, *DFNA2*, etc.; recessive loci are *DFNB1*, *DFNB2*, etc., whilst X-linked loci are *DFN1*, *DFN2*, etc. See the Hereditary Hearing Loss Homepage[5] for more details.

PROGRESS IN IDENTIFYING THE GENES: NON-SYNDROMIC HEARING LOSS

In many families uncomplicated hearing loss segregates in a pattern consistent with determination at a single genetic locus. Prelingual loss is usually autosomal recessive, whilst dominant inheritance is more commonly seen in late-onset loss. X-linked inheritance is uncommon. There is no simple way of working out how many different genes are involved, although indirect estimates based on population genetics suggested there might be 30–100 loci determining autosomal recessive hearing loss.[1] This very great heterogeneity is simply a reflection of the number of different proteins, each encoded by a separate gene, that are specifically required for cochlear function.

For many years non-syndromic hearing loss was regarded as genetically intractable. The dominant forms are mostly of late onset and the pattern in families is confused by the frequent co-occurrence of age-related loss aetiologically unconnected with the familial loss. Families with recessive deafness are usually individually too small for linkage analysis, but families cannot be combined for analysis because of the expected extensive genetic heterogeneity. Moreover, the frequent deaf–deaf marriages can make it impossible to follow the line of transmission of a deafness gene through a family. Further confusion is introduced by family members who are deaf for some other reason, despite not inheriting the family gene (phenocopies). For recessive loss, the solution was to study the large, multiply inbred kindreds that can be found in various societies around the Mediterranean and across the Middle East to the Indian subcontinent. A single kindred can be large enough to give statistically meaningful linkage data, thus avoiding the problem of heterogeneity. Dominant hearing loss of adult onset is amenable to family study where the usual age of onset is relatively early, but little headway has yet been made in identifying genetic susceptibility to presbyacusis or noise-induced loss.

Many groups worldwide have been assiduously collecting multi-case families suitable for linkage analysis, and as a result of their efforts about 70 recessive and 50 dominant loci have so far been mapped (Table 6.2). To avoid conflicts of nomenclature, a central naming system has been set up, and researchers who have identified a new locus can reserve a name even before publishing their data – hence the 'reserved' entries in Table 6.2. Sometimes, once the actual gene has been identified, it turns out that two different entries are both due to mutations in the same gene. In two cases (*DFNA2*, *DFNA3*), the opposite has happened: mutations have been found in two genes at the appropriate chromosomal location in different families. Table 6.2 gives an immediate impression of the extreme genetic heterogeneity of non-syndromic hearing loss.

Progress in cloning the genes is accelerating as the Human Genome Project makes better tools available. Currently, the genes responsible for 26 recessive and 22 dominant forms of non-syndromic hearing loss have been identified. The gene products provide interesting insights into the mechanism of hearing. They include ion channels (GJB2, GJB3, GJB6, CLDN14, KCNQ4, SLC26A4), motor proteins (MYO1A, MYO3A, MYO6, MYO7A, MYO15A, MYH9, MYH14), adhesion molecules (CDH23, PCDH15) and structural components (COL11A2, USH1C, SANS, WHRN, TECTA, OTOA, STRC). The roles of these various genes and proteins have been reviewed by Petit[8] and Snoeckx and Van Camp.[9]

A surprising finding has been that the same gene may be mutated in two or more different types of loss. Sometimes mutations in the same gene can cause either dominant or recessive non-syndromic loss, e.g. connexin 26 (*GJB2*) is mutated in recessive *DFNB1* and dominant

DFNA3, whilst alpha-tectorin (*TECTA*) is mutated in dominant *DFNA12* and recessive *DFNB21*. In other cases, mutations in the same gene may underlie both a non-syndromic and a syndromic form of hearing loss. MYO7A is mutated in Usher syndrome 1B, in recessive *DFNB1* and in dominant *DFNA11*. The explanation here is likely to centre around the distinction between simple loss of function mutations and dominant negative effects, as described earlier in connection with Jervell and Lange-Nielsen syndrome. Such dominant negative effects are especially seen when the protein encoded by the gene functions as a multimer – a multimer containing some normal and some abnormal molecules may be non-functional. Heterozygotes are affected, and so the condition is dominant. This is particularly clear with connexin 26. The protein functions as hexamers to produce connexons, intercellular gap junctions. Mutant versions of the *GJB2* gene that produce no protein are seen in recessive hearing loss – evidently cells can function adequately with a half-dose of connexin 26. The mutations that produce dominant *DFNA3* loss lead to production of a full size but abnormal protein, which presumably can sequester the product of the normal allele in non-functional hexamers.

The distinction between syndromic and non-syndromic hearing loss is not absolute. Several types of non-syndromic loss have special features that can help point suspicion at the relevant gene.

- *Pendrin and enlarged vestibular aqueducts*: Many patients with mutations in *SLC26A4*, the gene-encoding pendrin, have Pendred syndrome (hearing loss with goitre), but others have the *DFNB4* non-syndromic loss. In most cases, there is enlargement of the vestibular aqueducts.
- *Otoferlin and auditory neuropathy*: Patients with otoferlin (*OTOF*) mutations have an unusual form of recessive non-syndromic loss in which otoacoustic emissions are conserved. This is referred to as auditory neuropathy. In a patient this finding would suggest that *OTOF* mutation screening might be worthwhile. However, not all cases of familial auditory neuropathy map to the *OTOF* locus at chromosome 2p22.
- *COCH and Ménière's disease*: Patients with the dominant *DFNA9* form of hearing loss have mutations in the *COCH* gene and show a variety of vestibular and Ménière-like symptoms. However, *COCH* does not appear to be responsible for most cases of Ménière's disease, for which genetic susceptibility factors have not so far been identified.
- *Wolframin and low-frequency loss*: The recessive Wolfram syndrome includes diabetes mellitus, optic atrophy and, usually, hearing loss. However, heterozygous carriers of certain mutations show a non-syndromic hearing loss (*DFNA26*), which is unusual in affecting mainly low frequencies.

Most loci have been implicated in only one or a few families, but a few genes seem to be frequent causes of non-syndromic genetic hearing loss. The most important is connexin 26. Connexins are proteins that assemble into hexameric units (connexons) in cell membranes and bind to connexons on an adjacent cell to form a gap junction, through which small molecules can pass from one cell to another. Mutations in at least three connexin genes (*GJB2*, *GJB3* and *GJB6*) have been implicated in non-syndromic hearing loss. The Connexin-deafness Internet homepage[10] is a good source of further information. By far the major player is connexin 26, encoded by the *GJB2* gene. Mutations in *GJB2* are the cause of *DFNB1* recessive hearing loss and also *DFNA3* dominant loss. As discussed later, *GJB2* mutations are sufficiently common and sufficiently easy to detect in the laboratory that testing for them has become part of normal clinical practice.

MITOCHONDRIAL SYNDROMES

Mitochondria have their own small genome, a 16,569 base-pair circle of DNA containing 37 genes. Mutations in the mitochondrial DNA are the cause of a bewildering variety of disorders, with the hallmark that they are inherited exclusively from the mother. Sperm do not contribute mitochondria to the zygote. Cells contain many mitochondria, and patients with mitochondrial mutations can be homoplasmic (all mitochondria the same) or heteroplasmic (a mixture of mitochondrial types). Heteroplasmy can be transmitted from mother to child because the egg contains huge numbers of mitochondria.

Mitochondrial DNA is rather variable compared with nuclear DNA. Thus, sequence variants are common, and their significance is often hard to assess. Several variants are associated with hearing loss, often as part of syndromes. One variant, A1555G (replacement of nucleotide 1555, normally A, by G) causes extreme sensitivity to the ototoxic effects of aminoglycoside antibiotics. Estivill's group in Spain[11] found A1555G in 17/70 consecutive referrals of familial severe congenital or progressive sensorineural hearing loss with no other identifiable cause; 'familial' here meant that the proband had at least one other affected relative. There was often no documented history of antibiotic exposure. In other countries (except Portugal and Cuba), this variant is similarly frequent in the general population but not amongst deaf families who report no aminoglycoside exposure. It seems likely that this difference reflects the past high consumption of aminoglycosides and their ready availability over the counter in those three countries, but not elsewhere.

PROGRESS IN IDENTIFYING THE GENES: AGE-RELATED AND NOISE-INDUCED HEARING LOSS

Unlike the conditions described in the foregoing, the aetiology of age-related or noise-induced hearing loss is complex. Environmental factors – not least age and noise, but also many other factors – obviously have major roles. For age-related loss, family and twin studies provide strong evidence that people vary in their inborn genetic susceptibility. The evidence that humans differ in their genetic susceptibility to noise-induced loss is weaker, but experiments in mice have shown strong differences between strains.

Identifying the individual genes underlying susceptibility to common complex conditions has been a major strand in genetic research for the past two decades. Until recently the results of these investigations have been very disappointing. Many identifications have been claimed, but few confirmed. Family-based investigations, as described earlier, have very poor power to detect factors that have only modest effects on overall susceptibility. Additionally, for age-related loss there is the obvious problem of finding suitable families with affected people in several generations still available to provide DNA. Such studies have been attempted for hearing loss, and possible susceptibility loci have been mapped, but experience across many diseases suggests that great caution is needed in interpreting such results. An alternative approach is needed, and this is provided by association studies.

The idea here is to test a large collection of independent unrelated affected people, to look for any genetic variant that is significantly more common amongst affected than unaffected people. Such a factor might be directly causative, but alternatively it might simply reside on a conserved and widely shared ancestral chromosome segment that somewhere carries the true susceptibility factor. The study design is simple in principle, but it requires very large numbers

of subjects to get the required statistical power. Additionally, any shared ancestral chromosome segments will be extremely short, so in order to search the whole genome for susceptibility factors, it is necessary to test huge numbers of very closely spaced genetic variants. Until very recently, this was not technically possible. Advances in technology have now made such genome-wide association studies possible, but they remain formidably expensive.

More targeted association studies are less expensive and have already yielded some results. A reasonable hypothesis is that if mutations that totally inactivate a certain gene cause hearing loss, then lesser changes that just reduce the activity of the gene product might increase vulnerability to age-related or noise-induced loss. On this basis, variants in the *KCNQ4* gene (implicated in *DFNA2* hearing loss) and *TFCP2L3* (implicated in *DFNA28* loss) have been tentatively identified as possible susceptibility factors for age-related loss.[12,13] It should be emphasised, however, that even if these associations are confirmed, they have only a modest effect on susceptibility. There is no case at present for screening populations or testing patients for these factors.

IMPLICATIONS FOR DIAGNOSIS

Identifying the genes causing hearing loss has several purposes. Biologists hope to gain insight into normal human physiology and development. The genes mutated in non-syndromic loss presumably encode components of the auditory transduction machinery, whilst those mutated in syndromic loss control developmental processes. Identifying the genes should help elucidate the mechanisms of these various processes.

Identifying a disease gene immediately raises the possibility of molecular diagnosis. A definitive diagnosis is of value in itself to patients and parents who want to know why they or their child is deaf. It can put an end to guilt and soul-searching, and allow accurate counselling about recurrence risks. Whether or not this hope is realistic depends on the precision of the diagnostic question being posed. Consider three possible questions:

1. Does this patient have *any* mutation in *any* gene that will explain his or her condition?
2. Does this patient have *any* mutation in *this particular gene* that will explain his or her condition?
3. Does this patient have *one specified* mutation (e.g. nucleotide A replaced with G at position 1555) in *this particular gene*?

Question 1 is impossible to answer at present. Many people envisage a time when everyone will have their complete genome sequenced as an integral part of their medical record. Even if this does eventually come about, it does not follow that we would know how to interpret the information. Question 3 on the other hand can be answered cheaply and easily, by a single quick laboratory test. Question 2 is in principle always answerable with current knowledge and technology, but answering it may be unfeasibly expensive. Scanning through the whole sequence of even one large gene such as *MYO7A* or *OTOF* is currently too expensive and laborious to be a routine diagnostic procedure. With each passing year, the costs of DNA testing fall, and the scale upon which it can be done increases. Analysis of any one gene should reasonably soon become a routine procedure, but at present this is available only for a limited number of genes (see succeeding discussions). The development of specialised microarrays (‘gene chips’) may soon allow a spectrum of more common mutations to be checked in a single

operation. Nevertheless, it is unlikely that diagnostic laboratories would be able to offer a comprehensive analysis of all the genes in Table 6.2 as a routine service in the foreseeable future. Thus, the key to routine molecular diagnosis is knowing which gene to target.

How practical is it to specify the gene in advance? For syndromic hearing loss, this is usually possible if the syndrome is identified correctly by clinical examination. Sometimes, as for example with Usher syndrome, there may be several suspects, but at least the list is limited. For non-syndromic loss, clinical examination will usually give no clue, and the only hope is if experience shows that one particular gene is mutated in a substantial proportion of all patients. In small isolated populations, this can be quite a common occurrence; it is less likely in large and open populations.

In general, it is not possible to specify the precise DNA sequence change that is sought (i.e. to pose question 3). For most genetic diseases, unrelated affected people have different mutations, and it is necessary to search the whole gene to find a mutation. Genes are long stretches of DNA, thousands or tens of thousands of base pairs long, and this is a major task. However, there are three circumstances in which one can suggest the particular mutation to be tested for:

1. If additional family members are being tested for the presence or absence of a mutation that has already been defined in one affected family member.
2. If the nature of the disease is such that only one very specific alteration in the gene sequence will produce that effect. An example would be sickle cell disease; in hearing loss the only common example that comes to mind is the mitochondrial A1555G mutation in aminoglycoside-induced hearing loss.
3. If one particular mutation, inherited from a common ancestor, has by chance spread very widely through a population. The connexin 26 mutation 30delG (described further in the succeeding discussions) is the prime example of this (that particular sequence is also a mutational hotspot, which no doubt contributes to its high prevalence).

What molecular diagnostic services are available at present for the clinician? This is an area of rapid change superimposed on great differences between countries and even regions. For most of the important syndromes, mutation testing languishes in the gulf between research and service. Mutation screening of *MYO7A*, *EYA1*, *PAX3*, *MITF*, *SLC26A4* and other important genes has been offered by the researchers who initially identified these genes, but once they have published a few dozen mutations, they can rarely justify using research funds for further testing. If they are set up to handle invoicing, they may be able to continue on a fee for service basis. With over 1,000 disease genes identified, routine diagnostic laboratories have to restrict themselves to a limited menu of tests. Laboratories are slowly moving towards establishing consortia where in each country (or for rarer diseases, each continent), two laboratories, one primary and one back-up, agree to provide a fee service for any particular rare disease. A directory of European diagnostic laboratories, searchable by gene or disease, is available.[14]

Connexin 26 is a special case. GJB2 mutations are a major cause of prelingual hearing loss in many populations, accounting for up to half of recessive (unaffected parents, two or more affected children) and 10–25% of sporadic prelingual hearing loss in several studies.[15] Moreover, in different populations, a high proportion of all mutations are one particular sequence change. In Europe, loss of one G from a run of six consecutive G nucleotides (30delG, sometimes called 35delG) is much the major mutation. In East Asia, a different mutation, 235delC,

is common, and amongst Ashkenazi Jews 167delT is frequent. This has important implications for diagnostic testing.

The high frequency of the 30delG mutation in many European countries clearly justifies routine testing of hearing-impaired children, and it is simple and cheap to test for this specific mutation. However, it is important to have a policy in place when a deaf child turns out to have a single copy of the 30delG mutation. This is a common finding – in early studies, it was seen in 7/39 and 10/82 consecutive children in presumed recessive families in France[16] and Spain,[17] respectively. Does the child have a different mutation in his or her other copy of the GJB2 gene, or is he or she deaf for some unrelated reason, but coincidentally a heterozygous carrier of 30delG? Carrier frequencies in many European countries are 1–2%, so the dilemma is a real one. Thus, any laboratory offering testing for 30delG must also be able to offer, or at least organise, screening of the whole GJB2 gene for further mutations. Fortunately, the GJB2 gene is small, and not over-challenging to sequence in its entirety.

Thus, it is clear that the progress of the last 15 years in identifying the specific causes of genetic hearing loss has greatly extended the scope of diagnostic testing. In the longer term, it is hoped that the new knowledge will lead to better treatment and maybe prevention or cures. Gene therapy (replacing or removing malfunctioning and defective genes) could eventually produce cures for diseases where the symptoms stem from malfunctioning of the defective gene here and now, and are reversible. It would not help with developmental defects, where the damage was done long ago and is irreversible. Animal experiments have shown proof of principle that gene therapy could work for some forms of hereditary hearing loss,[18,19] though practical interventions in humans are still many years away.

Maybe some people are genetically sensitive to particular environmental insults (as with aminoglycosides), and if they could be identified by population screening, they could be singled out for protection. Maybe genetic dissection of the mechanisms of development and function of the auditory system will identify novel targets for drug treatments. These are all developments for the long-term future – but a consistent lesson from the past 20 years of molecular genetics has been that the long-term future often materialises remarkably quickly.

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7 Craniofacial syndromes and hearing loss

V.E. Newton

INTRODUCTION

Normal development of the head is a complex process. In order for the skull and facial structures to develop to the right size and shape, and for features to develop in the right position, there needs to be coordination of cell movement and activity during intrauterine life. Signalling networks are involved and regulate such processes as brain patterning, cell migration and tissue fusion. The networks involve classes of genes such as transcription factors, homeobox genes or fibroblastic growth factor receptors, e.g. *Hox* genes and *PAX* genes. As these are regulatory genes, when they are abnormal this has a cascading affect, altering the function of other genes. Mutations in the genes of the signalling networks cause a range of craniofacial defects.[1]

Craniofacial malformations are involved in one-third of all human congenital defects[2] and three-quarters of congenital birth defects in humans affecting the head and neck.[3] Most inherited craniofacial syndromes are due to autosomal dominant genes. The most common mechanism is reported to be haploinsufficiency, but gain of function mutations also occur.[4] Inherited factors are not the only causes of craniofacial malformations. Environmental teratogens can be responsible directly or can cause craniofacial defects indirectly by triggering a new gene mutation or chromosomal abnormality.

The majority of the syndromes with craniofacial abnormalities display external ear defects. Hearing loss may be conductive, sensorineural or mixed, symmetrical or asymmetrical, stable or progressive.

In this chapter, some of the genetic and chromosomal craniofacial syndromes are described with the main emphasis being on the associated hearing impairment. Few studies have reported on vestibular abnormalities in connection with these conditions.

CRANIOSYNOSTOSES

These are conditions in which there has been a premature fusion of one or more cranial sutures. The brain continues to develop and, to allow for this, compensatory growth takes place in the sutures still patent. This results in an abnormal head shape.

Wilkie[5] estimated that premature suture fusion occurs in 1 in 2,500 births. Approximately 8% are familial.[6] Many are inherited as autosomal dominant conditions but some exhibit autosomal recessive inheritance and others are sporadic or secondary to other disorders, e.g. microcephaly, hyperthyroidism or mucopolysaccharidoses. Males are affected more commonly than females (3 : 1).

The syndromic craniosynostoses have been found to be associated with mutations in the fibroblast growth factor receptor family. Mutations may be gain-of-function or loss-of-function mutations.[7] Over 100 craniosynostosis syndromes have been described.[8] Those more commonly associated with hearing loss are Crouzon syndrome, Apert syndrome, Pfeiffer syndrome and Saethre-Chotzen syndrome. Early diagnosis allows for surgical intervention which may be needed to relieve raised intracranial pressure and/or to correct cranial and facial abnormalities.

Crouzon syndrome

This syndrome is characterised by premature and progressive craniosynostosis, hypoplasia of the midface with shallow orbits and ocular proptosis (Figure 7.1). Coronal and sagittal sutures are almost always fused and the lamboidal sutures are involved in 80% of cases.[9] The syndrome exhibits wide phenotypic variability. Prevalence is reported to be about 15 to 16 in one million live births.[10]

Crouzon's syndrome is an autosomal dominant disorder resulting from a mutation in the gene encoding fibroblast growth factor receptor 2 (*FGFR2*). Sporadic cases of Crouzon syndrome are associated with increased paternal age.[7]

More than 50% of those affected by Crouzon syndrome have a conductive hearing loss. This may be attributed to atresia of the external auditory meatus and canal or malformation or fixation of the ossicles. In 19 patients (12 male, 7 female) reported by Orvidas et al.,[11] eight had abnormalities of the external ear including one with atresia and six with a malaligned auricle. Ten of the patients had a hearing impairment: four a conductive hearing loss, two a mixed hearing loss and four a sensorineural hearing impairment. The middle-ear conditions included ossicular fixation and otitis media.

Apert syndrome

This autosomal dominantly inherited condition is found in 9.9/million to 15.5/million births and accounts for about 4–5% of craniosynostosis. The syndrome is caused by mutations in *FGFR2*.[12] Mutations are exclusively of paternal origin.[9] Craniosynostosis is most frequently in the form of bicoronal and cranial base synostosis.[10] There is flattening of the occipital bones and a prominent forehead with mild to moderate exophthalmos. Many have an intellectual impairment. Midfacial hypoplasia is found and a depressed nasal bridge. Characteristically,

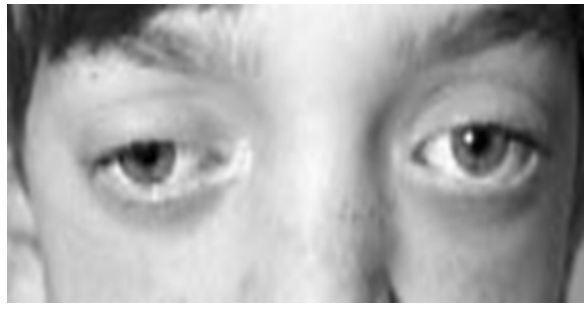

Figure 7.1 Proptosis in Crouzon syndrome (courtesy of Professor D. Donnai).

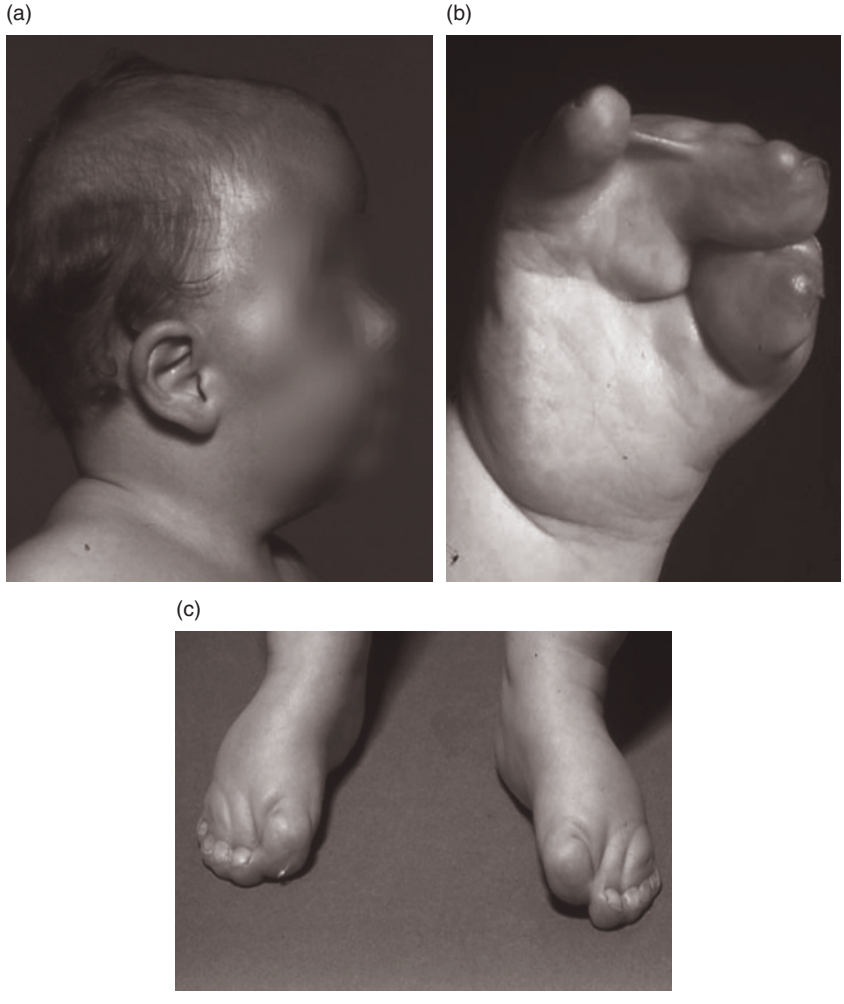

Figure 7.2 Apert syndrome: (a) Cranial shape; (b) syndactyly of hand and (c) foot (courtesy of Professor D. Donnai).

the craniofacial abnormalities are associated with syndactyly of the hands and feet, the hallux being separate from the other toes, and sometimes there are six metatarsals. A cleft lip and palate is present in around one-third (Figure 7.2).

Conductive hearing loss is found. Out of the 70 patients reported by Rajenderkumar et al.,[13] 3–6% had a congenital hearing loss and in 56% of these otitis media was present and persisted until the ages of 10–20 years. Congenital stapes ankylosis has also been described.[14]

Pfeiffer syndrome

In this autosomal dominant craniosynostosis syndrome, the thumbs and toes are broad and there is a partial syndactyly of the hands and toes. The syndrome can result from a mutation

in either *FGFR1* or *FGFR2*, though Cunningham et al.[7] have suggested that in the cases of *FGFR2* mutations, Crouzon syndrome might be the more appropriate designation.

Cremers[15] described a 14-year-old boy with a conductive hearing loss, acrocephaly, minimum syndactyly and broad thumbs and big toes who underwent an exploratory tympanotomy. The incus was found to be fixed to the epitympanum and there was ankylosis of the stapes. The internal auditory meatus was dilated bilaterally.

Nine children, 2 to 12 years, eight of whom had a hearing impairment, formed the group studied by Vallino-Napoli.[16] Conductive hearing loss was found in seven of the children with four having a middle-ear effusion and one had a mixed hearing loss. A CT scan revealed stenosis and/or atresia of the external auditory canal, hypoplasia of the middle-ear cavity and enlargement of the middle-ear cavity, and in a few cases, the ossicles were hypoplastic. All but one had a normal inner ear.

Saethre-Chotzen syndrome

This syndrome is characterised by unilateral or bilateral coronal synostosis, ptosis, ocular hypertelorism, maxillary hypoplasia, a low frontal hairline, a small pinna with a prominent crus and syndactyly.[7] It is inherited as an autosomal dominant condition caused by mutations in the *TWIST 1* gene, a basic helix–loop–helix transcription factor. The mutations lead to a loss of function.[7]

Hearing loss is mainly described as conductive or mixed, but a case of severe to profound sensorineural hearing loss has been reported.[17] In the family described by Ensink et al.[18] with a conductive hearing loss, ankylosis of the stapes and a fixed ossicular chain in a small epitympanum were described.

MANDIBULAR DYSOSTOSES

Treacher Collins syndrome (TCS)

TCS is an autosomal dominant condition with variable penetrance and expressivity. It is estimated to occur in 1 in 50,000 live births. Sixty per cent of cases arise *de novo.*[19]

Clinical abnormalities involve structures derived from the 1st and 2nd branchial arches and are mainly bilaterally symmetrical. The clinical features include downward-sloping palpebral fissures, coloboma of the lower eyelid and absent eyelashes in the outer third; hypoplasia of the maxilla and mandible, cleft palate and abnormal development of the external and middle ear. There may be microtia, ear tags, absent or deformed ossicles. The middle-ear cavity may be dysmorphic or absent. Hearing loss when present is usually conductive, but a mixed hearing loss has been reported (Figure 7.3).[20]

TCS is caused by mutations in the *TCOF1* gene mapped to 5q32–33.1 in 1996. It encodes the protein Treacle. It has been suggested that haploinsufficiency of Treacle affects the proliferation and proper differentiation of specific embryonic cells during development.[21] Treacle controls the production of mature ribosomes and if deficient, this is what causes disruption in neural crest formation and proliferation resulting in the hypoplasia characteristic of TCS.[22]

Bone-anchored hearing aids and prostheses may be used in the management of those with severe bony atresia.

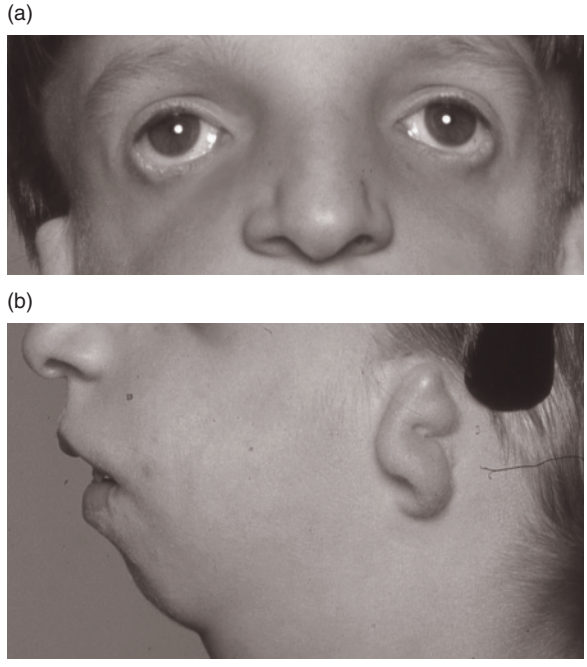

Figure 7.3 Treacher Collins syndrome: (a) facies showing downward slope of the palpebral fissures, coloboma and micrognathia; (b) pinna abnormality and micrognathia (courtesy of Professor D. Donnai).

Nager acrofacial dysostosis

This syndrome, first described in 1948 by Nager and deRenier,[23] is a rare mandibular facial dysostosis with limb abnormalities and hearing impairment. Most cases are thought to be sporadic. Limb abnormalities include hypoplasia of the thumbs, radii and humerii. The facial features include downward sloping palpebral fissures and hypoplasia of the malar bones and the mandible (Figure 7.4).[24]

Conductive hearing loss is described and has been attributed to external ear malformations and ossicular abnormalities.[25,26] In Herrmann et al.'s[25] study, external ear malformations were found in 8 out of the 10 patients enrolled. These ranged in severity from an isolated stenosis to anotia. The ossicular abnormalities included fixation of the malleus and incus to the adjacent temporal bone. Hearing loss was conductive in 90% of their patients. Two patients in the same study were reported to have developed a sensorineural 'dip' at 2 kHz in later childhood.

Hemifacial microsomia/Goldenhar's syndrome (also known as oculo-auriclo-vertebral spectrum or first and second arch syndrome)

This consists of the triad of craniofacial microsomia, ocular dermoid cysts and spinal defects. Occasionally, cardiac and renal anomalies are found. The syndrome is found in 1 in 3,000 live births and in many cases it is unilateral. In a group investigated by Touliatou et al., 70% had unilateral manifestations and these were mainly right-sided.[27]

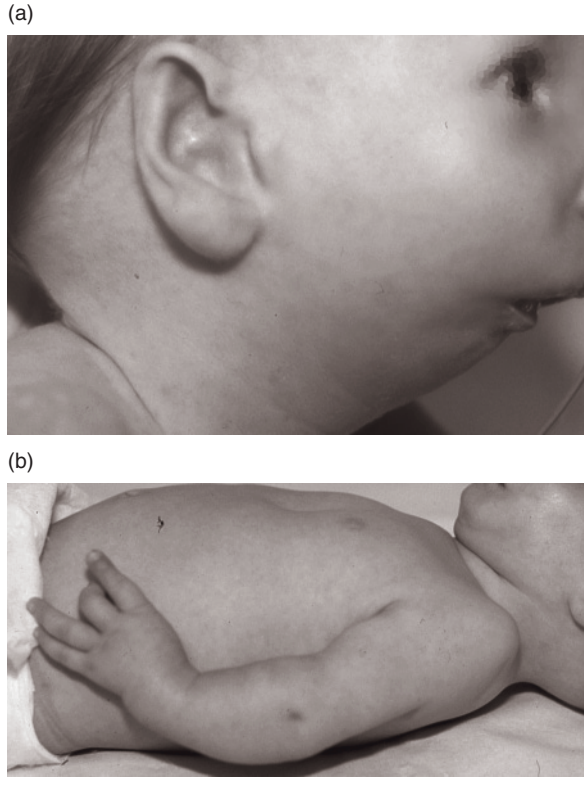

Figure 7.4 Nager syndrome: (a) facial features; (b) upper limb abnormality (courtesy of Professor D. Donai).

Whereas the majority are sporadic, autosomal dominant and autosomal recessive forms have been described. A literature review described by Tasse et al.[28] found that those with autosomal dominant inheritance are more likely to be bilaterally affected than those where the condition was sporadic. Hearing impairment, atresia and epibulbar dermoids were less frequently found in the genetic group than in sporadic cases. Where epibulbar dermoids are present, then ear tags tend to be present bilaterally. Clinical manifestations of the syndrome, as found in 17 patients described by Touliatou et al.,[27] included auricular defects in 94% and ocular abnormalities in 65% (Figure 7.5).

Forty percent are reported to have a conductive hearing loss.[9] A higher proportion of hearing impairment has been reported by others.[27,30] Cavalho et al.[30] found 74 out of 99 paediatric patients (75%) had a hearing loss. There was a statistically significant relationship between auricular abnormalities and conductive and sensorineural hearing impairment.

Hearing loss results from middle-ear abnormalities or external ear atresia.[9] Middle-ear abnormalities described include malformation of the tympanum and of the ossicles.[29] Five percent have a cleft lip and palate.[9]

The presence of a sensorineural hearing impairment has also been reported in this condition,[30–32] and abnormalities of the stria vascularis and semicircular canals have been demonstrated.[29]

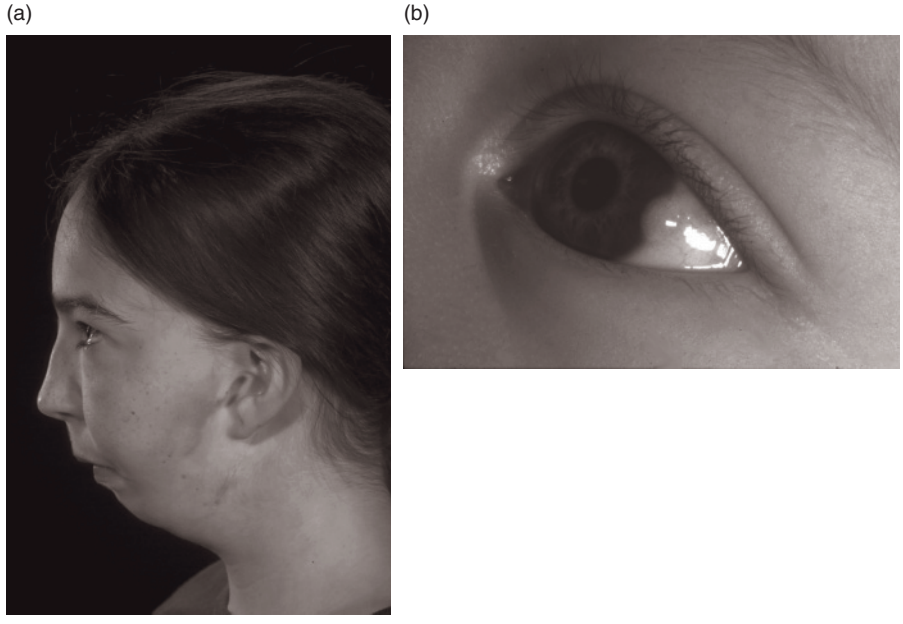

Figure 7.5 Goldenhar's syndrome: (a) facies (courtesy of Dr M. Bitner-Glinditz); (b) epibulbar dermoid.

Isolated microtia may be a marker for unsuspected hemifacial microsomia.[32] Examination of 100 consecutive patients with isolated microtia revealed that 40% had hemifacial microsomia, 31 unilateral and 9 bilateral; 37 had a conductive hearing loss; and one a sensorineural hearing impairment.

INHERITED CHONDRODYSPLASIAS

Stickler syndrome/Marshall syndrome/Marshall/Stickler

Marshall and Stickler syndromes are heterogeneous conditions affecting collagen connective tissue. They are characterised by high myopia, orofacial abnormalities and hearing loss. The hearing loss is usually sensorineural affecting mainly the high frequencies and with a tendency to progress. Conductive and mixed hearing loss can also be found due to the association of a cleft palate and otitis media. Some individuals have no overt signs of the condition. As there are overlapping features, there has been controversy whether Marshall and Stickler syndromes represent different manifestations of the same syndrome or are different syndromes.

Stickler syndrome is mainly autosomal dominant, and types I, II and III have been described. Type I is caused by mutations in the *COL2A1* gene, Type II from mutations in *COL11A2* and Type III from mutations in *COL11A1*.[33] *COL 2A1* is the most common gene causing the condition. Clinical signs depend upon the mutations present. Mutations in *COL11A1* give rise to Marshall phenotypes or overlapping Stickler/Marshall phenotypes. In 2006, in one family of Moroccan origin with Stickler syndrome, an autosomal recessive pattern was identified and the syndrome was found to be due to a homozygous R295X mutation in *COL9A1*.[34]

Reports of the auditory manifestations related to the type of Stickler syndrome present indicate that they are milder in Type I than in Types II and III.[33,35] Hearing loss is reported to be more severe in Marshall syndrome than in Stickler syndrome.[33]

Prevalence of early onset hearing loss is reported to be less common (7.5–19%) in families with exon 2 mutations in *COL2A1* than in other families (70%).[36] Three families identified and investigated by Richards et al.[37] with mutations in exon 2 of *COL2A1* were described as having a predominantly ocular phenotype. Hearing loss was uncommon but was the most frequent systemic finding, and both conductive and sensorineural hearing loss was described. Donoso et al.[38] reported on a large family of 2,384 members who also had a mutation in exon 2. The researchers were able to obtain the clinical records of 165 family members of whom 95 were affected and 70 unaffected. Affected individuals had early onset posterior perivascular retinal degeneration, vitreous degeneration and retinal detachment ($n = 95$). A hearing loss was found in only two (7.5%) of a subset of 28 affected individuals, both of whom were described as needing a hearing aid before the age of 21 years.

The auditory manifestations of Type II Stickler syndrome caused by a *COL11A2* mutation were explored by Admiraal et al.[39] in 15 affected persons. Six had a mixed hearing loss and five of these had a submucous or overt cleft palate. The mean sensorineural threshold was 40 dB HL. Audiograms obtained from 14 affected persons were described as sloping ($n = 6$), flat or gently sloping ($n = 3$), flat ($n = 2$) and U-shaped ($n = 3$). They commented that in this non-ocular form of Stickler syndrome, sensorineural hearing loss had a higher prevalence than in Type I Stickler syndrome. The conductive element was associated with otitis media.

Griffith et al.[40] described hearing loss in three individuals with the Marshall phenotype resulting from mutations in *COL11A1*. The three individuals described had serial audiograms from the age of 5 years. These depicted a cochlear sensorineural hearing loss progressing to severe-to-profound hearing loss by the 6th decade, Vestibular function was explored using electronystagmography, calorics, rotational chair and dynamic posturography. Central dysfunction was reported in two subjects and peripheral dysfunction in the third.

The family described by Van Camp et al.,[34] where Stickler syndrome was inherited as an autosomal recessive condition, had moderate to severe sensorineural hearing gloss, moderate to high myopia, vitreoretinopathy and epiphyseal dysplasia. Heterozygote carriers had no signs of the syndrome clinically.

Hypermobilty of the tympanic membrane was found in 21/46 examined in Szymko-Bennett et al.'s study[33] although the appearance was normal. It was suggested that this finding could be useful as a clinical diagnostic feature.

OTHER SYNDROMES

Waardenburg syndrome

This auditory pigmentary syndrome was first described by Waardenburg in 1951.[41] Subsequently, four syndrome types have been identified. Types I, II and III are inherited in an autosomal dominant manner, whereas Type IV is autosomal recessive. Details of the genetic basis of this syndrome can be found in Chapter 6. The prevalence of this condition has been estimated to be 1.44–2.05/100,000 in the general population.[42]

Variable penetrance and expressivity have been described. The main clinical features are dystopia canthorum, i.e. lateral displacement of the inner canthi (Types I and III only), pig-

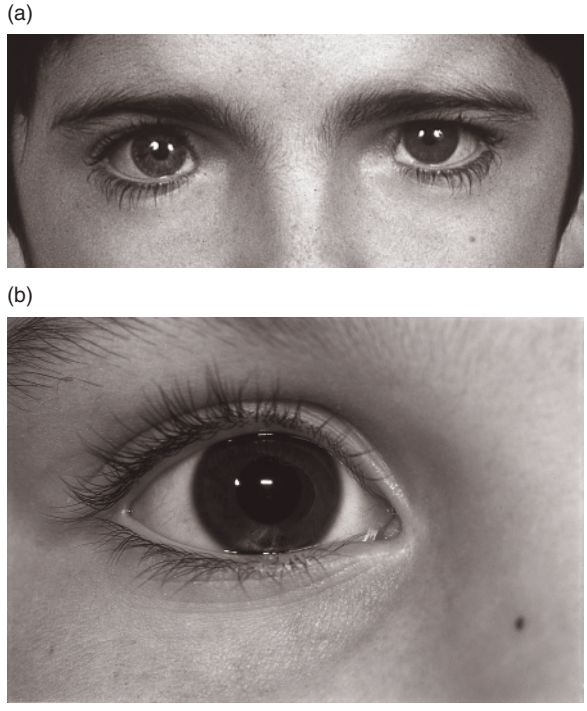

Figure 7.6 Waardenburg syndrome: (a) eyelid anomaly with dystopia canthorum, synophrys and complete heterochromia irides; (b) partial heterochromia irides with a clearly demarcated segment of a different colour.

mentation abnormalities and congenital sensorineural hearing impairment. The displacement of the inner canthi results in the medial sclera appearing smaller than on the lateral side and the lacrimal ducts open opposite the cornea rather than the sclera. Synophrys and hypoplasia of the alae nasi are associated with the presence of dystopia canthorum (Figure 7.6).

The pigmentation abnormalities include a white forelock although occasionally a black forelock has been described. The irises may be of different colours, have segments of one or both irises of a colour different from the rest of the iris, or both irises may be hypochromatic with a deep blue appearance. Hypopigmentation of the skin is an additional feature and some cases of hyperpigmentation have also been described. Hirschsprung's disease is associated with the recessive form of Waardenburg syndrome; it is not clear if it is also associated with other syndrome types.[43]

Congenital sensorineural hearing impairment or normal hearing is found. When hearing loss is present, it can be unilateral but is usually bilateral and varies in degree from mild to profound. The prevalence of hearing impairment is higher in Type II than in Type I.[44] Newton reported that 67% of those with Type I had a hearing loss and 87% with Type II.[44] A variety of audiogram shapes have been recorded including low-frequency ascending, U-shaped, flat and gently sloping or steeply sloping high-frequency hearing loss.[44] In Type II, an asymmetrical hearing loss in which one ear has a low-frequency ascending hearing loss and the other a profound hearing impairment has not been described in Type I. Hearing loss is more frequently

found in association with pigmentation abnormalities than without these features.[45,46] Whereas hearing gloss is generally stable, progression has been described in association with Type II Waardenburg syndrome.[47]

Computerised tomography has indicated that in most instances, the cochlea is normal but a dilated vestibular aqueduct has been described in a small group of children with a profound sensorineural hearing impairment.[48] Vestibular function is believed to be mainly normal, but vestbular abnormalities have been reported.[47,49]

Children with a profound hearing loss as a result of Waardenburg syndrome have been reported to be good candidates for cochlear implantation.[50]

Branchio-oto-renal syndrome

First described in 1975, this syndrome is estimated as occurring in 1 in 40,000 of the population. It is inherited as an autosomal dominant trait and results from abnormal development of the first and second branchial arches. Causative genes have been identified – *EYA 1*, *SIX1* and *SIX5*. Recently, Sanggaard et al.[51] made the observation that renal and temporal lobe malformations seem to be more frequent in the condition when due to *SIX1* mutations than to *EYA 1*–related disease. The syndrome is clinically variable; branchio-oto-renal (BOR), branchio-oto-facial and branchio-otic syndromes have been described.

Abnormalities of the ear are associated with branchial fistulae and renal anomalies. Atresia, an auricular abnormality (lop-ear deformity), pre-auricular pits and tags may be present (Figure 7.7).

Ceruti et al.[52] described inner-ear malformations in all eight patients with BOR in their study. Hypoplasia and dysplasia of the cochlea was consistently found, a wide vestibular aqueduct was a frequent finding, and bilateral hypoplasia of the eighth nerve was found in one patient. The most common features described by Propst et al.[53] were a hypoplastic apical turn of the cochlea, deviation of the facial nerve to the medial side of the cochlea, a funnel-shaped internal auditory canal, a patulous eustachian tube and widened vestibular aqueduct. Others have described enlarged endolymphatic sacs and ducts.[54]

Hearing loss found is variable in degree from mild to profound. It may be conductive (33%), sensorineural (29%) or mixed (52%).[55] It is usually stable (~70%) but can be progressive in

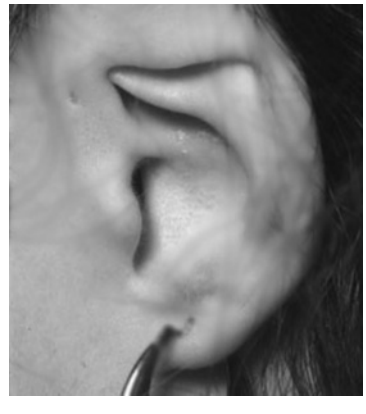

Figure 7.7 Branchio-oto-renal syndrome: preauricular pit (courtesy of Dr T. Sirimanna).

association with a dilated vestibular aqueduct.[55] Progressive fluctuating hearing loss has been reported.[56]

Wildervanck Syndrome (cervico-oculo-acoustic syndrome)

This syndrome encompasses the Klippel-Feil anomalad, Duane's retraction syndrome and hearing impairment. The syndrome is uncommon with prevalence higher in females than males (10:1). It may be unilateral or bilateral. Anomalies include short stature, microcephaly, mental retardation and cleft palate.[57] Radiological examination of one patient with Wildervanck syndrome revealed a conductive hearing loss in one ear and a Mondini defect in the other indicative of a sensorineural hearing impairment.[58]

Klippel-Feil syndrome (KFS)

Fusion of the spine of varying degree affects the cervical, thoracic and sometimes the lumbar spine. Imaging by CT or MR scan of 24 consecutive patients with KFS found that cervical spondylosis or disc herniation was the most commonly associated radiological abnormality ($n = 10$).[59] Clinically, there may be a short neck, limited cervical mobility and low posterior hairline, but these features may not be readily apparent in those with mild manifestations of the syndrome. Sprengel's shoulder is one of the most commonly associated abnormalities.[60] Samartzis et al. observed Sprengel's deformity in 5 out of 30 (16.7%) patients with the Klippel-Feil anomalad; in one it was present bilaterally.[60]

Hearing loss may be mixed, conductive or sensorineural, and both unilateral and bilateral impairment have been described. The causes of a conductive hearing loss include outer-ear malformation and ossicular abnormalities. Sensorineural hearing loss is the most common type associated. McGaughran et al.'s study[61] indicated hearing problems in 35 out of 44 with the syndrome with sensorineural hearing impairment found in 15 and a mixed hearing impairment in 10; there was no evidence of a typical audiometric profile.

Duane retraction syndrome

This involves a lateral rectus palsy with the eye being retracted on adduction. Kirkham[62] described sensorineural hearing impairment in 12 out of 176 patients with this diagnosis.

Noonan syndrome

The incidence of Noonan syndrome is estimated as between 1/1,000 and 2.5/1,000 births. Clinical features include short stature, hypertelorism, ptosis, downward-slanting palpebral fissures, posteriorly rotated auricles, a webbed neck, cardiac defects, cryptorchidism and bleeding problems. The syndrome is inherited as autosomal dominant and mutations in the *PTPN11* gene on chromosome 12 are implicated in around 50% of cases.[63] A small proportion with the syndrome have mutations in the *KRAS* gene.[63]

Hearing loss is often found.[64,65] Ranke[66] recorded that out of 410 cases with Noonan's syndrome, 63% had symptoms relating to the ear. Qui et al.[65] reviewed 20 cases of Noonan syndrome and reported that 50% of ears showed a progressive high-frequency sensorineural hearing loss. Conductive hearing loss has also been reported.[67] The patient described by Cremers et al. had a unilateral conductive hearing loss and tympanotomy revealed absence

of the long process of the incus and an abnormal relative positioning of the malleus and stapes.

Cornelia de Lange syndrome (CdLS)

This autosomal dominant developmental disorder is estimated to affect 1 in 10–30,000 newborns. Features include the typical facial features of thin arched eyebrows and long eyelashes, a low dorsal hairline and low set ears. There is usually mental retardation and there may be microcephaly, hirsutism and limb abnormalities. Some have a cleft palate and stenosis of the external auditory meatus has been found.[68] Features of the syndrome vary from mild to severe.

Mutations in *NIPBL*, *SMC1A* and *CMC3* genes cause the syndrome. Around half are believed to be due to *NIPBL*, which codes for the protein delangin which has a role in regulating the activity of other genes involved in early development.

Temporal bone pathology was reported by Yamanobe and Ohtani,[69] who described anomalies of the middle and inner ear and facial nerves in CdLS. Hearing loss may be conductive, sensorineural or mixed.[70–72] In Marchisio et al.'s[71] investigation of 50 children, 1–18 years with CdLS, hearing loss was found in 40 (80%), with conductive hearing loss alone in 60% and in combination with a sensorineural hearing loss in 20%. Otitis media with effusion was found in 94% and prevalence was reported to be the same in all age groups.

Sensorineural hearing loss was found in two boys examined by Ichiyama et al.[72] One had no responses on an auditory brainstem response test at 100 dB Hl, the other had a wave V threshold of 40 dB HL.

Townes-Brocks syndrome

This is an autosomal dominant disorder with multiple malformations. The gene has been mapped to 16q12.1, and mutations in *SALL1*, a candidate gene, have been reported in one family and in a sporadic case. Clinical defects involve mainly the ear, hands and feet, the anus and kidney.

A review of the clinical features of the syndrome by Powell and Michaelis[73] indicated that the most common limb defects are triphalyngeal thumb and preaxial polydactyly, but a broad distal phalanx of the thumb is also common. Imperforate anus is the most frequent anal abnormality found and renal defects include hypoplastic or dysplastic kidneys. Intelligence is usually normal.

External ear abnormalities include pre-auricular tags or lop ear and there are also ossicular abnormalities, including a hypoplastic head of malleus and a malformed incus. Hearing loss is predominantly sensorineural affecting high frequency thresholds and is slowly progressive, with a mild hearing impairment in childhood progressing to a moderate hearing loss by early adulthood.[74]

Johansson-Blizzard syndrome

This autosomal recessive syndrome was first described in 1971 as featuring aplasia of the ala nasae, deafness, hypothyroidism, dwarfism, absent permanent teeth and malabsorption. Other abnormalities have been described subsequently including microcephaly, cardiac and genitourinary anomalies.[75]

Hearing loss has been described in 75% of those affected and is sensorineural, severe and bilateral. A CT scan of the inner ears of both patients described by Braun et al.[75] revealed a Mondini defect bilaterally together with cystic dilatation of the vestibule. The vestibular aqueducts were shortened and widened and there was narrowing of the round window.

CHARGE association

This spectrum of congenital defects is found in one in 10,000. The acronym CHARGE is based on the presence of Coloboma, Heart malformation, Atretic choanae, Retarded growth and or development, Genital hypoplasia and Ear abnormalities. Facial weakness and orofacial clefting have also been described. Clinical criteria for diagnosing CHARGE association have been proposed, the most recent focusing on coloboma, choanal atresia and abnormal semicircular canals.[76]

The *CHD7* gene on chromosome 8q12.1 is a major cause of this syndrome.[77] Many cases are sporadic but familial occurrence was recorded by Delahaye et al.,[78] who described six patients in two families, one parent and two children in each. There was marked intrafamilial variation in the clinical features present.

The external ear is typically low set, anteverted and cup-shaped. Pre-auricular tags and microtia may be present. In the middle ear the stapedius muscle may be absent, the incus and stapes hypoplastic and fixation of the ossicular chain present.[79] There may be hypoplasia or agenesis of the semicircular canals.[80] Absence of the bony semicircular canals in the presence of a bony cochlea is a characteristic finding in CHARGE association (Figure 7.8).[81]

Hearing impairment is frequently found and the hearing loss is usually severe. Mixed hearing impairment is the most common type of hearing loss but both conductive and sensorineural hearing impairment have been described.

In Morgan's[82] investigation of 50 patients, all had ear abnormalities and 48 had malformed pinnae (96%). Facial nerve palsies were present in 27 of the patients (54%). The most common

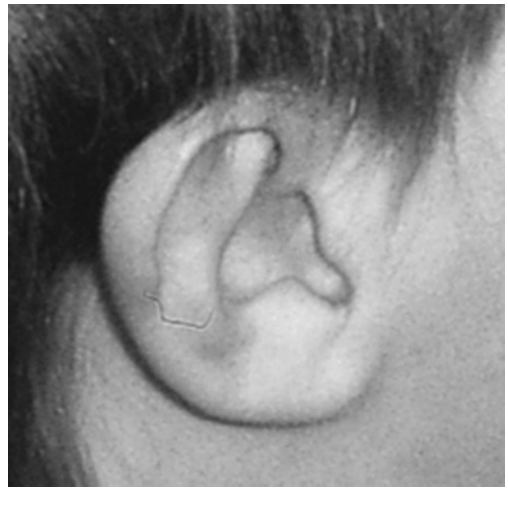

Figure 7.8 CHARGE association: typical auricle.

hearing defect was a severe conductive or mixed hearing loss, only eight (4%) having normal hearing. Amongst the 84% with radiological abnormalities, the characteristic findings were a hypoplastic incus and absent semicircular canals.

Edwards et al.[83] described a group of 21 children, of whom four had a mixed hearing impairment bilaterally, five had a sensorineural hearing loss, and two had a mixed hearing loss in one ear and a conductive hearing loss in the other. Two of the children had cochlear dysplasias.

CHROMOSOMAL ABNORMALITIES

Craniofacial abnormalities are featured in many syndromes caused by chromosomal abnormalities; two of the more common of these are described.

Down syndrome

This chromosomal disorder is due to trisomy 21. It occurs in 1 in 600 live births and is characterised by a number of clinical features. Ear abnormalities are found, including a small pinna with deficient cartilage in the upper pole, a stenosed external auditory canal, ossicular abnormalities and a shortened cochlea. Hearing loss is reported to occur in 38–78%.[84] Whereas otitis media is believed to be the commonest cause of a hearing impairment in young children, a mixed hearing loss or a sensorineural hearing loss have been found.

Otitis media in children with Down syndrome is related to eustachian tube defects. The tube is shaped differently and collapses more easily.[84] A radiographic study of the skull base and nasopharynx reported by Brown et al.[85] revealed that the nasopharynx was narrower than normal in Down syndrome patients and the angle between the base of the skull and the hard palate was significantly less acute than in normal controls.

Inner-ear dysplasia was found to be common in a study described by Blaser et al.[86] Using high-resolution computerised tomography or magnetic resonance imaging, they investigated 59 patients with Down syndrome. They found inner ear structures to be hypoplastic with vestibular malformations common. They described a small bony island of the lateral semicircular canal as ‘highly typical’. Other abnormalities included fusion of the lateral semicircular canal and the vestibule, enlargement of the vestibular aqueduct and endolymphatic sac, stenosis of the internal auditory canal and hypoplasia of the cochlear nerve canal.

In an auditory brainstem evoked response test described by Krecicki et al.,[87] the latencies (peaks I–III and interpeak latencies I–III) were shorter for children with Down syndrome under the age of 1 year than their unaffected peers, and significantly longer in an older age group when compared with a control group. They suggested that reference values obtained from normal children should not be used as a reference for assessing the hearing of children with Down syndrome.

Turner syndrome

Turner syndrome is the most common sex chromosome disorder in females and is due to a partial or total deletion of one of the X chromosomes, the latter being the most common. Phenotypic manifestations differ depending on the parental origin of the intact X chromosome.

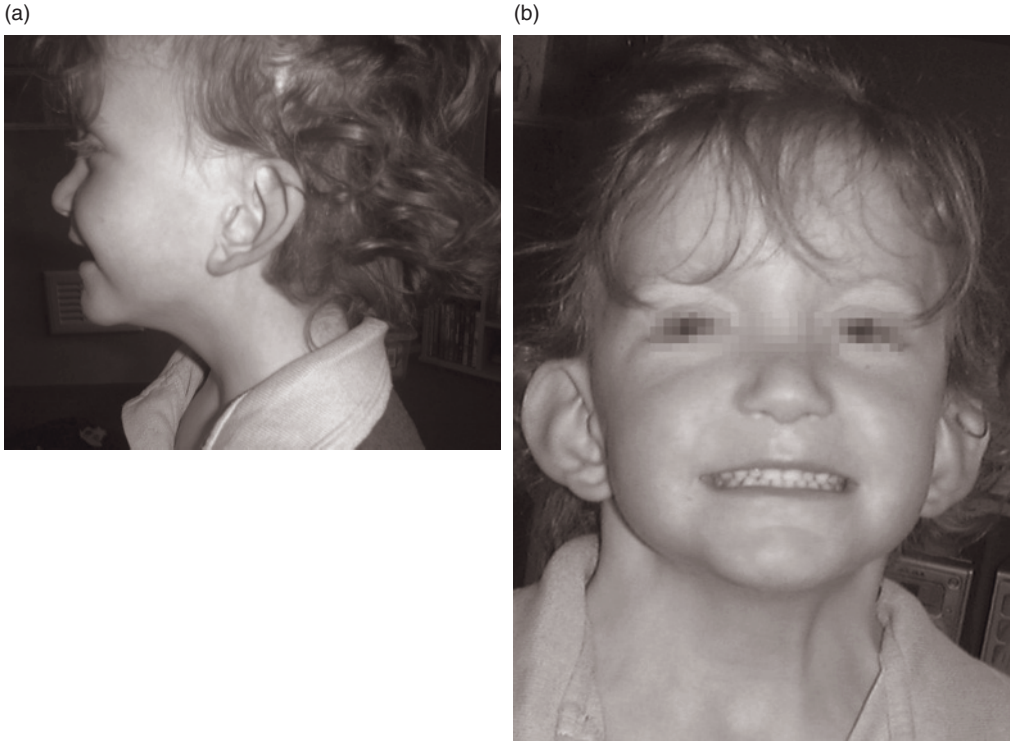

Figure 7.9 Turner syndrome: showing (a) the low set rotated auricles; (b) the auricles and broad neck.

The clinical features of the syndrome include small stature, a nuchal prominence, broad webbed neck, a depressed sternum, cubitas valgus and cardiac, renal and auditory abnormalities (Figure 7.9).

Hearing loss is frequently found and is often conductive.[88,89] Stenberg et al.'s study[89] of 56 girls aged 4–15 years with Turner syndrome revealed that 61% had a history of recurrent otitis media. A sensorineural hearing loss develops from late childhood to early adulthood[88] but a mid-frequency sensorineural 'dip' has been reported in children as young as 6 years.[89] The sensorineural hearing loss is progressive and worse in the high frequencies.[90]

A relationship has been found between the degree of mosaicism and the incidence of auricular anomalies and sensorineural hearing loss. A large investigation of the ear and hearing problems in 119 girls with Turner syndrome was carried out by Barrenas et al.[91] with particular emphasis on the degree of mosaicism. They noted that the prevalence of sensorineural hearing loss and auricular abnormalities increased significantly, the greater the proportion of 45, X cells present in an individual. A recent study by King et al. of 200 females 7–61 years of age revealed significantly poorer air-conduction thresholds in those with a karyotype 46, XdelXp and 46, XiXq groups than in the 46, XdelXq group.[90]

Morimoto et al. reported that in their group of 33 patients, 8–33 years of age, age-dependent high-frequency hearing loss was more prevalent in the XO karyotype than in those with the mosaic type.[92]

A relationship has also been found with the origin of the intact X chromosome. It has been observed by Hamelin et al. that those with an intact X chromosome of maternal origin are less likely to have a sensorineural hearing loss than those with the intact X chromosome being of paternal origin.[93]

CONCLUSION

The association of craniofacial abnormalities with hearing impairment indicates that children with such abnormalities are at risk for a hearing loss and possibly vestibular dysfunction; they should have their hearing examined at the earliest opportunity.

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8 Infectious causes of paediatric hearing impairment

P.J. Vallely and P.E. Klapper

INTRODUCTION

Hearing loss or impairment is a frequent consequence of infection. In particular, bacterial meningitis and otitis media are well known and important causes, but infection with many other pathogens including viruses and parasites can also lead to sensorineural or conductive hearing loss. The impact of childhood vaccination programmes in the developed world means that the burden of hearing loss due to infection is heaviest in developing countries, but many children in all areas of the world are born with hearing deficits as a result of a congenital infection, or develop hearing impairment or loss in childhood due to an acquired infection.

It is crucial to understand and identify infectious causes of hearing loss and raise awareness of them as, unlike for many other causes of deafness, there is a real possibility of intervention to prevent or limit such loss.

INFECTION AS A CAUSE OF HEARING LOSS IN CHILDREN

Epidemiological data

The World Health Organization (WHO) estimate that 80% of deaf and hearing-impaired people live in low- or middle-income countries.[1] As infectious diseases are similarly more prevalent in such developing countries, this statistic suggests that infection is an important contributor to hearing impairment. The WHO are currently compiling a global database on deafness and hearing impairment and this will undoubtedly help in understanding the scale of the problem and allow appropriate targeted interventions. However, there is still an urgent need for good epidemiological data to show the direct proportion of cases of deafness or hearing impairment due to an infectious cause.

Smith and Mathers[2] provided an overview of epidemiological data available from surveys relating to infection and hearing loss carried out within the last 20 years across the 17 sub-regions of the WHO (Table 8.1). These were mostly cross-sectional surveys carried out on representative populations within a particular country or area of a country, although some were conducted within individual schools or hospitals, and included reports of both permanent and temporary (e.g. as a result of chronic otitis media) hearing loss. The data were identified from a total of 12 regions but large parts of Africa, Europe, America and Asia were not represented. Conclusions from these limited data indicate that in sub-Saharan Africa, infection may contribute as much as 50% of the total burden, whilst in Europe, America and parts of Indonesia, the burden due to infection appears to be closer to 10%. It is clearly important that better data

Table 8.1 Infectious burden of hearing loss by WHO Region (adapted from Smith and Mathers, 2006)[2].

WHO Region	Country	Year of study	Ages of participants	Number of participants	Prevalence of hearing impairment	Proportion of hearing loss due to infectious causes (%)	Reference
African	Nigeria	2000	≥ 6 months	8975	18.8%	47.5%	Nwawolo 2003[193]
	Sierra Leone	1992	5–15 years	2015	9.1%	ND	Seely et al. 1995[194]
	Zimbabwe	1998	4–20 years	5528	2.4%	54.1%	Stewart et al. 1998[195]
Americas	USA	1991–1992	3–10 years	324327	ND	10%	Van Naarden et al. 1999[196]
	Brazil	2003	≥ 6 months	2427	28.2%	15.3%	Beria et al. 2005[197]
Eastern Mediterranean	Saudi Arabia	1988–1990	2 months – 12 years	6421	7.7%	70.6%	Al-Muhaimeed 1996[82]
	Oman	1997	≥6 months	11400	5.5%	18%	Al-Khabori and Khandekar 2004[198]
European	UK	1994–1995	1–10 years	552558	0.12%	8.6%	Fortnum and Davis 1997[199]
	Estonia	1985–1990	Followed from birth	144186	0.172%	ND	Uus and Davis 2000[200]
South East Asian	Indonesia	Random cluster study	>6 months	5604	12%	10%	Mackenzie 2002[201]
	India	Random cluster study	>6 months	5428	20.8%	39.2%	Mackenzie 2002[201]
Western Pacific	China	2000	All ages	126876	3.3%	26.8%	Liu et al. 2001[202]
	Vietnam	2001	>6 months	13120	20.4%	6.3%	Dung 2003[203]

are obtained in order to estimate the true global burden of hearing impairment due to infectious causes.

INFECTIOUS AGENTS CAUSING HEARING IMPAIRMENT

Infection is caused by invasion of the body with a pathogenic micro-organism. This may be a bacterium, virus, fungus or parasite. The symptoms associated with disease due to infection may be caused directly by the pathogen, usually by replication of the organism within body tissues or fluids, or indirectly, by damage resulting from toxic substances produced by the organism or from the immune response mounted by the host. As detailed in further discussions, relatively little is known about the specific pathogenic mechanisms that occur within individual infections and lead to hearing loss, but damage occurs either within the ear itself or to the eighth cranial nerve innervating the ear.

Hearing loss due to infection in children occurs either before birth as a result of congenital infection or during childhood as a result of an acquired infection. The major infectious causes of congenital hearing loss are rubella, cytomegalovirus and syphilis, and the major causes of acquired hearing loss are bacterial meningitis and chronic otitis media. Sensorineural deafness resulting from congenital infections such as rubella is understood to be due to nerve damage during organogenesis, whereas the conductive hearing loss typically seen with otitis media is due to build up of immune infiltrate within the inner ear. It can be difficult in individual cases to identify the infectious cause of hearing impairment: often, hearing loss is the only symptom of the infection or the loss may develop after the infection has cleared. This means that it is likely that infectious causes are under-reported. However, wherever possible, identification of an infectious aetiology is desirable as it will help with counselling parents, may allow treatment intervention to limit damage and may also help to anticipate prognosis.

CONGENITAL INFECTIONS

Rubella

Epidemiology

Rubella virus is an enveloped RNA virus classified within the *Togaviridae* family. It infects only humans, is spread via the respiratory route, and its transmission generally requires close contact. Following an incubation period of 14–21 days a mild, self-limiting, red rash illness is typically seen in children. This non-confluent maculopapular rash appears first on the face and spreads centripetally to the trunk and limbs. The rash may be associated with enlarged lymph nodes, sore throat, cough, mild conjunctivitis, a low-grade fever and, particularly in post-pubertal females, arthralgia and arthritis, which may last for several weeks. Serious complications are rare. In contrast to this mild post-natal illness, rubella causes a serious congenital infection if contracted by a woman for the first time during pregnancy, particularly during the first 16 weeks of gestation when the virus will invariably be transmitted to the foetus with an 85% risk of congenital damage, most common amongst which is sensorineural hearing loss (SNHL) (Table 8.2).

Prior to the introduction of routine vaccination, rubella had a worldwide distribution emerging each spring in temperate climates and causing epidemics every 4–7 years. The implementa-

Table 8.2 Likely outcome of congenital infection with rubella virus according to gestational age.

Timing of infection	Possible outcome
Preconception	Minimal risk
0–12 weeks	100% risk of congenital infection, major congenital abnormalities likely, Spontaneous abortion in 20% of cases
13–16 weeks	Deafness and retinopathy in ~15%
After 16 weeks	Normal development, slight risk of deafness and/or retinopathy

tion of vaccination programmes begun in the late 1960s and 1970s mean that the disease is now rare in Australia and large parts of Europe and has been declared eliminated from North America.[3] Many other countries have introduced vaccine programmes more recently, but in Africa and Asia vaccination is more variable and it is estimated that around 100,000 cases of congenital rubella syndrome still occur each year mostly in these regions.[4]

Congenital rubella syndrome (CRS)

Rubella was first described in Germany in the mid-nineteenth century and became commonly known as 'German measles' to differentiate it from classical measles. For the next century rubella was largely ignored as the disease was considered of little significance until, in 1940, a link between maternal infection with rubella during pregnancy and certain specific birth defects was made by Norman Gregg.[5] Gregg reported a series of 78 babies with similar kinds of congenital cataract, some of whom also had heart disease. In 68 of these cases the mother was shown to have been infected with rubella during the first or second month of pregnancy. Further studies showed that in some of these babies, and in others born to rubella infected mothers, deafness and microcephaly were also seen, with a preponderance of cases of deafness occurring in infants whose mothers were infected slightly later in pregnancy (mean 2.1 months gestation; reviewed by Hanshaw et al.[6]). This was the first realisation that viruses could cause congenital malformation.

It is now recognised that congenital infection with rubella produces a range of symptoms including the classic triad of cataracts, heart defects, and sensorineural deafness, recognised by Gregg. Affected infants may also show intrauterine growth retardation, central nervous system (CNS) defects, hepatosplenomegaly, thrombocytopenia, petechia, purpura, as well as late-onset manifestations such as diabetes mellitus, thyroid disorders and progressive sensorineural deafness (Box 8.1).

Congenital rubella and the ear

In contrast to most of the other manifestations, SNHL as a consequence of rubella infection may occur in isolation, particularly when infection occurs after the first 12 weeks of pregnancy.[7] This is because development of the inner ear occurs over a longer period spanning the 2nd to 4th month of gestation compared with the structures of the eye which develop within the first few weeks, and the heart which develops by the second month of gestation. The hearing loss may be unilateral or bilateral, range from mild to profound, and loss may be asymmetric.[8] Hearing loss may be present at birth or, as the virus can persist in the inner ear fluid, it may continue to cause degenerative changes in the organ leading to progressive loss, which becomes

Box 8.1 Symptoms of congenital rubella syndrome.

Transient symptoms

Low birth weight, hepatosplenomegaly, thrombocytopenic purpura, bone lesions, meningoencephalitis, hepatitis, haemolytic anaemia, pneumonitis, lymphadenopathy

Permanent sequelae

Of the heart: peripheral pulmonary stenosis, pulmonary valvular stenosis, patent ductus arteriosus, ventricular septal defect

Of the eye: retinopathy, cataract, microphthalmia, glaucoma, severe myopia

Of the ear: sensorineural deafness

Other organs: microcephaly, psychomotor delay, diabetes mellitus, thyroid disorders, dermatoglyphic abnormalities, dental defects

Late-onset permanent sequelae

Sensorineural deafness, mental retardation, diabetes, thyroid disorders

apparent in infancy or early childhood, or occasionally as a late onset consequence of the disease (Box 8.1).

The overall incidence of deafness following congenital rubella infection is not accurately known. Various studies have reported figures ranging from 30 to 80%.[9–12] The significance of rubella as a cause of hearing loss is shown by studies conducted in the UK in the era before rubella vaccination was given to all infants as part of the measles, mumps and rubella (MMR) vaccine campaign (that began in 1988). Two cohorts of children identified as having congenital rubella infection and reported to the UK National Congenital Rubella Surveillance Programme (NCRSP) were examined. The first[10] comprised 111 children born between 1978 and 1982 and included 68 (61%) described as hearing impaired. The second cohort[13] of 159 children born between 1983 and 1987 included 75 (47%) with hearing impairment. In both studies, a loss of 40 dB or greater in both ears was present (mean loss 93 dB and 96 dB in the respective studies). In agreement with these data, follow-up of all children born between 1971 and 1993 and reported to the NCRSP suggests around half had SNHL. In a number of these cases, the hearing loss was only reported at second or subsequent follow-up, suggesting delayed diagnosis of the hearing loss was common and that delayed onset also occurred.[14] Furthermore, although approximately one-quarter of all children ever reported to the NCRSP had SNHL as the only sequelae of congenital rubella infection, all infants reported in recent years have had severe infection with multiple rubella defects. This may imply recent under-reporting of congenital rubella infection in the UK when SNHL is the only manifestation[15] as congenital rubella may no longer be suspected in an older child who has received the MMR vaccine by the time a diagnosis of SNHL is made.[14]

Control of infection

To date, no effective antiviral drugs have been developed to treat rubella virus infection and the mainstay for control is prevention by vaccination. Rubella vaccine is unusual in that it is not given to protect the individual to whom it is administered, but rather to protect the unborn child with whom that individual may have later contact. Most countries have adopted a strategy of childhood immunisation of both sexes designed to eradicate or at least prevent the epidemic spread of virus in the population. However, this strategy requires a vaccine uptake rate of 80% or higher in the targeted age group. Lower rates than this can lead to an upward shift in the

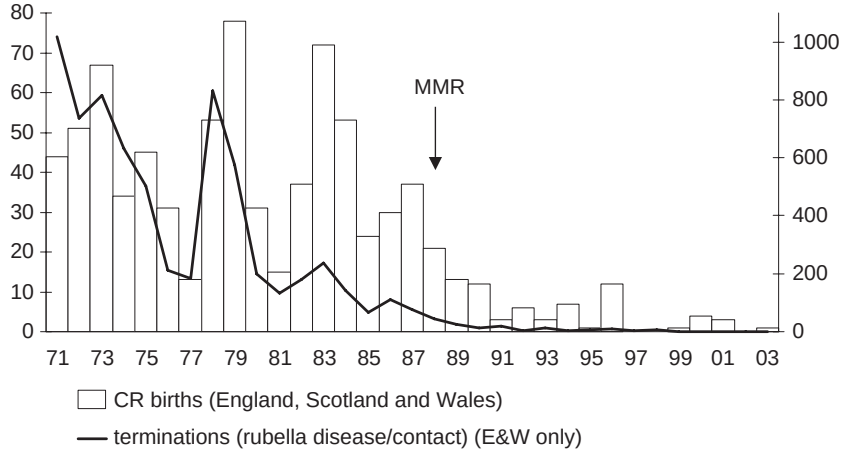

Figure 8.1 Congenital rubella births (NCRSP) and rubella associated terminations (ONS) 1971–2003 (adapted from Salisbury D., Ramsay M., Noakes K. Immunisation against infectious disease. Norwich: The Stationery Office, 2006).

average age of rubella infection with the effect of actually increasing the rates of CRS. The alternative strategy of vaccination of adolescent and adult females provides protection to the pregnant woman, and thus controls CRS but does not prevent or reduce virus circulation in the community, leaving non-vaccinated individuals at risk.

Universal childhood immunisation was introduced in the United States in 1969 and rates of rubella infection declined dramatically from 40 to 60,000 cases annually to only nine reported cases in 2004.[16] Many of the cases reported after the vaccination programme was established were born to non-immunised, Hispanic immigrant mothers[17] but since 2001, rates in this group also fell to <1/100,000 population due to improved vaccine coverage. Epidemiological analysis of rubella and CRS cases occurring between 1998 and 2004 suggested that the disease was no longer endemic in the United States.[16,18] In 1970, the UK introduced a programme of selective immunisation of pre-pubertal females. This led to a large reduction in CRS (Figure 8.1) but did not eliminate it, and around 50 cases of CRS continued to be reported each year. Universal vaccination was introduced in 1988 and has been effective in eliminating epidemic spread with 40 cases reported between 1991 and 2002[19]; most of these children were born to mothers who acquired the infection outside the UK. However, worldwide, CRS continues to be a problem. Of the 192 countries reporting to WHO in 2004, 116 (60%) have a rubella immunisation programme usually targeted at infants before their second birthday.[20] However, although this is a considerable improvement from the 78 countries with such a programme in 1996, coverage still accounts for only 26% of all births and it is estimated that a minimum of 100,000 cases of CRS still occur annually.[21]

Cytomegalovirus

Epidemiology

Cytomegalovirus (CMV) is one of the eight herpes viruses known to infect man. It is a large enveloped DNA virus, and in common with all herpes viruses, primary infection is followed by lifelong latent infection of the host with periodic reactivation and viral shedding. The virus

is present in all human populations studied and seropositivity rates vary from around 60–70% in adult populations in the developed world,[22,23] to >95% in developing countries.[24] Infection typically occurs in childhood, especially in the developing world, and transmission is associated with close physical contact in families or in daycare centres for children. Infants typically excrete large amounts of virus for months or years and exposure of contacts to saliva and other bodily fluids including via breastfeeding results in efficient virus spread. After childhood a further peak of infection is seen during adolescence and early adulthood coinciding with exposure to sexual activity.

Although human CMV infection (HCMV) rarely causes disease in immunocompetent individuals, the virus is responsible for significant morbidity and mortality in immunocompromised individuals and in prenatally and perinatally infected infants. Indeed, HCMV is accepted as the leading cause of congenital infection in the developed world. In a recent meta-analysis of studies where congenital CMV infection had been identified by universal screening, an average of 0.7% (0.3–1.3%) of all live births were found to be infected.[25] It is possible this may be an underestimate of the true rate of congenital CMV as all the available data arise from individual studies in industrialized countries and reliable, national-level data on incidence of congenital cytomegalovirus infection are not available from either developed or developing countries.

Congenital infection may arise as a result of either a primary or reactivated infection in the mother. The risk of transmission of the virus to the foetus is undoubtedly higher during primary maternal infection when approximately 30–40% of all maternal infections will result in a congenital infection.[26,27] However, around 10–30% of seropositive mothers will experience a reactivation of HCMV infection during pregnancy and in 1–3% of such cases the foetus will become infected.[27,28] As most infants worldwide are born to seropositive mothers, the overall number of congenital infections arising from reactivation in the mother could be substantial.[29] For many years, it was assumed that the foetus was at risk of damage from congenital CMV infection only if the mother acquired a primary infection. However, more recently, evidence has emerged that suggests the risk of symptomatic infection during a reactivated or secondary HCMV infection during pregnancy in a seropositive mother is in fact much higher than previously understood: data are available from both the United States and Europe to suggest that congenitally infected infants are born to mothers with pre-existing immunity,[26,27,30,31] and that a significant proportion of these will have an adverse outcome.[32–34]

Congenital cytomegalovirus infection

HCMV is now recognised as the most frequent cause of congenital infection in humans and is the leading non-hereditary cause of congenital sensorineural deafness,[35–37] It is estimated that of all children born with congenital CMV infection, 10–15% will have symptoms evident at birth and 85–90% will be asymptomatic (Figure 8.2). However, a further 10% of these asymptomatic children will develop late sequelae, most common amongst which is SNHL.[38]

Amongst the 10–15% of symptomatically infected infants (Figure 8.2), the range of disease is varied. The most severe manifestations are evident in a child born with cytomegalic inclusion disease, which results from viral interference with intrauterine growth and development, and causes prematurity and/or the birth of a severely affected child with multi-organ disease, particularly involving the reticuloendothelial and central nervous systems. Most affected infants display petechiae, jaundice, purpura and hepatosplenomegaly, and approximately two-thirds show neurological abnormalities including microcephaly. Mortality amongst such severely affected neonates may be as high as 30%[39] with death typically occurring as a result

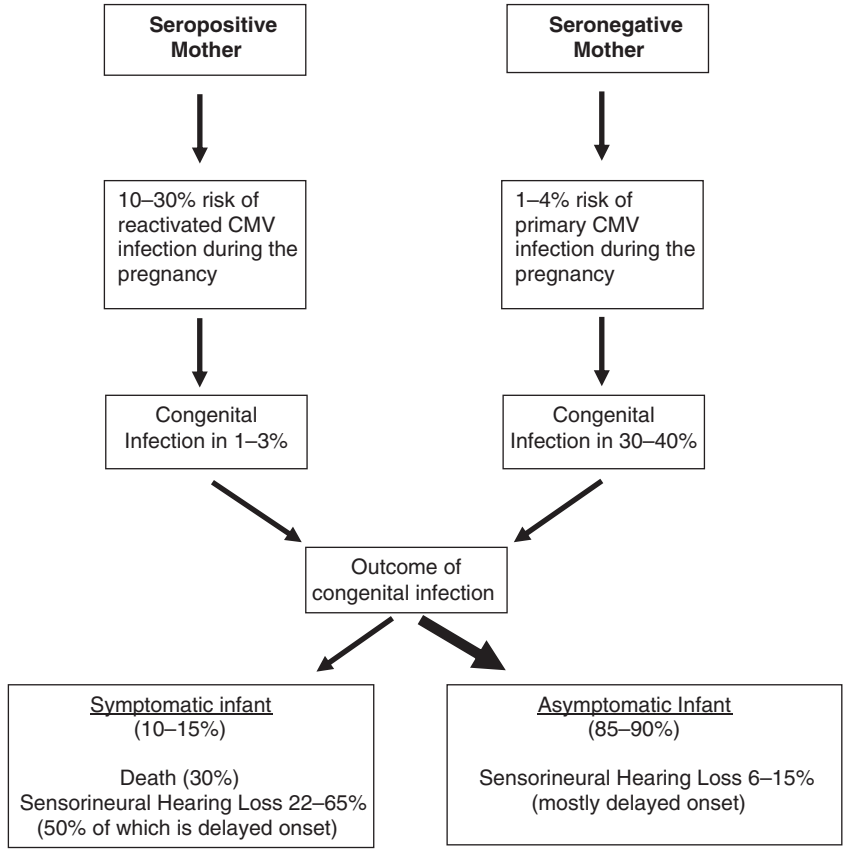

Figure 8.2 CMV infection in pregnancy and risk of congenital CMV infection.

of liver dysfunction, blood disorders or secondary bacterial infection. Permanent CNS sequelae are also common, including SNHL, mental retardation, cerebral palsy, seizures, blindness and other visual defects.

The majority of infected infants are asymptomatic at birth, but a proportion of these will develop late sequelae. The most common amongst the late sequelae is development of some degree of SNHL, which may be unilateral or bilateral.

Congenital HCMV and the ear

An association between congenital HCMV infection and hearing loss was first described more than 40 years ago,[40] and it is now recognised as one of the most important causes of deafness in childhood. Hearing loss is found in both symptomatic and initially asymptomatic infants, although it is apparently more likely to occur in a symptomatically affected infant (22–65%) compared with an infant who is asymptomatic at birth (6–15%).[41–45]

A series of studies carried out in Birmingham, Alabama, USA found that most affected children exhibited severe or profound SNHL (>70 db), and delayed onset of hearing loss was seen in both groups at an average age of 33 months for symptomatic and 44 months for

asymptomatic children. Progression of hearing loss was also noted in more than half of the children from both groups.[44]

As neonates are not routinely screened for HCMV infection at birth, many asymptomatic infections remain undiagnosed. The data from the Alabama study[44] suggest that between one-third and one-half of all CMV-related SNHL is not apparent until after the newborn period.[46] Thus, it is likely that hearing loss that develops in early childhood is often not recognised as being due to congenital HCMV infection.

A number of reports have attempted to estimate the overall prevalence of congenital HCMV-induced hearing loss. These reports are difficult to compare because of a lack of consistency in the definition of hearing loss, but most suggest that around 15–20% of all cases of moderate to profound hearing loss in children are associated with congenital HCMV infection,[47,48] and that this figure rises to around 25% if only profound hearing loss is considered.[28,47–50]

The reason that HCMV affects the ear in some infected infants and not in others and the reasons for the wide variation in severity is still poorly understood. A number of studies have looked for a correlation between various risk factors and development of SNHL. Rivera et al.[51] found that intrauterine growth retardation, petechiae, hepatosplenomegaly, hepatitis, thrombocytopenia and intracerebral calcifications were all associated with hearing loss, but that the presence of microcephaly or other neurological abnormalities was not. The authors conclude that disseminated disease rather than neurological involvement at birth is the most significant risk factor for hearing loss.

In support of this conclusion, a recent study found a relationship between high viral load in urine or peripheral blood of the infant in the neonatal period and development of hearing loss: virus levels in blood and urine from 83 congenitally infected infants were determined. Significantly, higher levels were found in both specimens among the children with hearing loss ($n = 12$) than among those without.[52] This was particularly noticeable in the asymptomatic infants with hearing loss where the viral burden was approximately 10-fold higher than in the asymptomatic infants with no loss. Further studies are needed to determine the importance of viral load in development of hearing loss and whether this can be used to predict children at risk of loss and identify those who may benefit from treatment.

Although the pathogenic mechanisms contributing to hearing loss remain unknown, there is some evidence that the host immune response against the virus may cause acute or persistent inflammatory damage within the ear.[53] Thus, a recent study[54] raises an interesting area for future work. Using a guinea pig model for CMV-induced hearing loss, the authors examined the possibility that virally encoded immunomodulatory genes were involved in the pathogenesis. Viral mutagenesis techniques were used to delete a specific viral immunomodulatory gene; this gene encodes a viral protein that acts as a homologue for a host protein, macrophage immunomodulatory protein 1α (MIP1α). HCMV is a virus that is well known for its ability to modulate the immune response raised against it by its host and this MIP1α homologue is one of the virally encoded proteins involved in modulation. Animals inoculated with the wild-type virus suffered significantly more hearing loss than animals inoculated with virus in which the gene had been deleted. These data suggest a potentially important role for viral immunomodulatory genes in the development of hearing loss, and further studies in this area are needed.

Control of infection

In contrast to rubella where it is possible to predict outcome in relation to timing of maternal infection, HCMV transmission may occur at all stages of pregnancy.[27] The risk of severe

congenital damage is probably higher in the first half of gestation.[38] CNS sequelae are particularly thought to be significantly increased if infection occurs in the first trimester.[55] However, termination of all pregnancies complicated by primary CMV infection cannot be recommended as neither identification of primary maternal infection nor direct evidence of foetal infection is predictive of outcome, and the majority of infected babies will be healthy.

HCMV is one of the few viral infections for which effective antiviral therapy exists. Currently, three antiviral drugs are available for the therapy of life-threatening HCMV infection: ganciclovir, famciclovir and cidofovir. However, none are currently licensed for use in pregnant women or neonates because all are associated with significant side-effects. A trial of ganciclovir in congenitally infected neonates, although reporting significant haematological toxicity, did show some benefit in preventing the incidence or progression of hearing loss in some severely affected infants.[56] More recently an oral formulation of the drug, valganciclovir, has been used to treat a symptomatic infant and was found to be well tolerated.[57] At present, effective antiviral treatment for this congenital infection is not available and further and more extensive studies will be needed before any treatment regime can be justified.

As the risk of transmission is lower in mothers with pre-existing immunity, the development of a vaccine to protect seronegative women from primary infection is desirable and a number of candidate vaccines are in preclinical or early clinical development.[58] An important consideration if an effective vaccine emerges will be the agreement of a vaccination strategy. Currently, there is no consensus amongst developed nations as to whether it would be more effective to vaccinate adolescent females or to incorporate the vaccine into the universal childhood vaccine schedule. The effectiveness of vaccination in developing countries where HCMV seropositivity rates are higher has not yet begun to be explored. A further problem that prevents progress with development of a vaccine for this virus is the general lack of awareness regarding the public health significance of congenital HCMV infection. An increase in public awareness of the problem may provide the necessary driver for progress in the development and testing of effective vaccination and antiviral therapy regimens.

Syphilis

Epidemiology

Syphilis is a sexually transmitted infection caused by the spirochete bacterium *Treponema pallidum* (Figure 8.3). There are three major stages to the infection: primary, secondary and late syphilis. Primary syphilis is characterised by a lesion at the site of inoculation with the organism (usually on the genital organs). This typically begins as a single, painless flat lesion that progresses to an ulcerative papule. Left untreated it will usually heal within 4 or 5 weeks, and consequently many cases of primary syphilis are not diagnosed.[59]

Secondary syphilis occurs 1 to 2 months after the primary infection when the organism establishes a systemic infection. Typical symptoms are a widespread red rash, fever and general malaise. Complications of secondary syphilis, although not common, arise from the vasculitis caused by the dissemination of the organism and include hepatitis, nephritis, gastrointestinal involvement and meningovascular syphilis, which may involve the 8th cranial nerve giving rise to a rare form of acquired deafness. The symptoms of secondary syphilis generally improve over the course of 3 to 6 weeks and the disease enters a latent and asymptomatic period. In around a third of patients in whom the disease is untreated, latent syphilis progresses

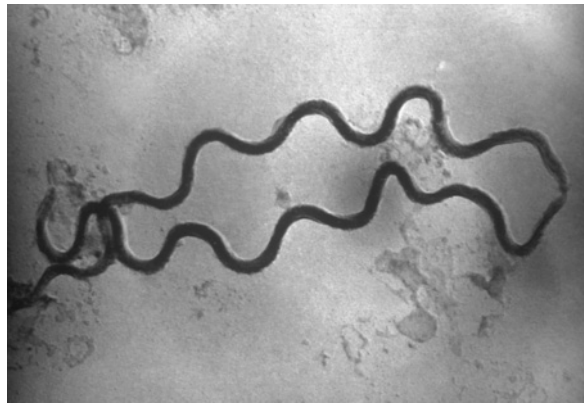

Figure 8.3 Scanning electron micrograph of two spiral-shaped *Treponema pallidum* bacteria (36000x) (courtesy of CDC/Joyce Ayers).

to tertiary syphilis. The duration of the latent period is very variable; 3–15 years is a common range but it may be even longer than this in some patients.[60]

Tertiary syphilis is now very rarely seen outside of developing countries. It has three main presentations: neurosyphilis, cardiovascular syphilis and late benign or gummatous syphilis. The latter of these is the most common, occurring in 50% of patients. The main feature is the appearance of multiple lesions or gummas. These are painless red nodules, which principally arise on the skin but can affect all organs; they often ulcerate and can cause severe tissue damage. The presentation is known as benign because the lesions themselves are not usually life-threatening; however, if an ulcerative gumma occurs in the brain, bone or other vital organs, then serious sequelae including death are likely.

Cardiovascular syphilis reflects invasion of the cardiac tissues with the organism, and angina, aneurysm or heart failure are common. If the spirochete enters the cerebrospinal fluid, it may spread to the vasculature of the meninges and from there to the spinal cord and brain resulting in parenchymatous neurosyphilis manifesting as tabes dorsalis or paresis.[61] As with secondary syphilis, involvement of the 8th cranial nerve may cause deafness. Tertiary syphilis can also result in deafness if lesions occur in the temporal bone or in the auditory canal.

The development of the long-acting penicillins effective against the bacterium in the 1950s and the introduction of serological screening programmes for syphilis in pregnant women brought about a dramatic reduction in cases of both sexually and congenitally transmitted syphilis. However, the WHO estimates that 12 million new cases of infectious syphilis still occur each year and 90% of these are in developing countries[62] with highest rates in sub-Saharan Africa and South and Southeast Asia.[63]

The incidence of syphilis in the United States and Western Europe was in decline for the latter part of the twentieth century and the 1998 data from the United States was the lowest recorded, showing an overall incidence rate of 20 cases of infectious syphilis per 100,000 population, with three cases of congenital syphilis per 100,000 live births.[64] Similarly, in the UK the incidence of syphilis declined to 0.47 per 100,000 population.[65] However, in the last decade a resurgence of the disease has been seen. The re-emergence apparently began in Russia and Eastern Europe[63] and during the late 1990s rates began to rise in many of the large cities

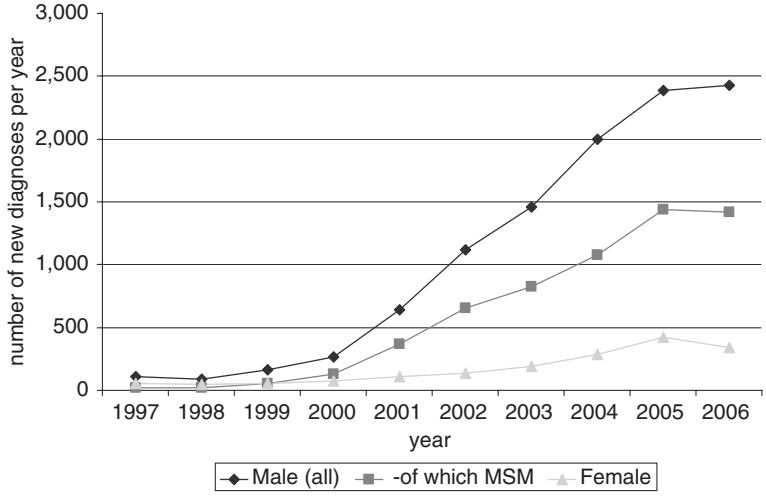

Figure 8.4 New cases of syphilis presenting to GUM clinics in the United Kingdom (compiled from data collected by the Health Protection Agency, UK).

in Western Europe and North America.[66–68] The epidemic is most prominent in homosexual males, e.g. in England, a 3,000% increase in syphilis in homosexual men was seen between 1996 and 2002 (Figure 8.4), but early syphilis is also increasingly recognised in heterosexual males and females.

Congenital syphilis

Estimates for the annual number of congenital syphilis cases are in the range 713,600 to 1,575,000 cases per year, leading to 1.3% of all deaths in children under 5 years old, mostly occurring in the developing world, particularly sub-Saharan Africa.[69]

Congenital syphilis results from passage of spirochetes present in the mother's blood across the placenta to enter the foetal blood and tissues. The disease is usually transmitted during the primary or secondary stages of maternal infection and is rare in pregnancies occurring more than 4 years after the primary infection. An adverse outcome is predicted in around 80% of pregnancies where the mother has syphilis. The effect of congenital syphilis in the infant is severe. Many are stillborn or die during the neonatal period. Babies born alive are often premature and of low birth weight; some show clear multi-organ involvement but most are initially asymptomatic. Symptoms may appear at any time within the first 2 years of life, when they are termed 'early' manifestations, or after 2 years when they are considered as 'late' manifestations.

Early manifestations are similar to the symptoms of secondary syphilis in adults. One of the earliest manifestations is a persistent rhinitis with a profuse nasal discharge (Figure 8.5), which is highly infectious. Other features include hepatosplenomegaly, lesions on the skin, inflammation of the long bones, anaemia and thrombocytopenia together with low birth weight and failure to thrive. Late congenital syphilis occurs when symptoms develop in an infant older than 2 years and is most commonly seen at puberty. Any organ system can be involved but typically manifestations are seen in the bones, teeth and nervous system.

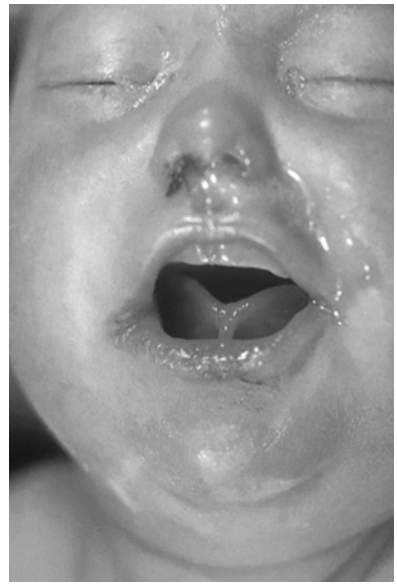

Figure 8.5 Newborn infant with congenital syphilis showing persistent rhinitis with profuse nasal discharge (courtesy of CDC/Dr Norman Cole).

Syphilis and the ear

Sensorineural hearing loss can be a consequence of both acquired and congenital syphilis. In acquired syphilis, SNHL occurs in 17% of early latent, 25% of late latent and 54% of tertiary syphilis cases.[70] In contrast, deafness is a rare complication of congenital syphilis, but may occur in a previously undiagnosed child. It appears as a late manifestation, typically when the child is 8–10 years of age, although occasionally it may be delayed until adulthood. It is often seen as part of 'Hutchinson's triad',[71] which includes notched incisor teeth, interstitial keratitis and eighth cranial nerve deafness. Facial abnormalities (a 'saddle' nose, protuberant mandible), CNS abnormality (mental retardation, optic nerve atrophy) and bone or joint involvement (frontal bossing of the skull, 'saber' shins, hypertrophy of the sternoclavicular joints) may also be present and point to a diagnosis of congenital syphilis.

With both acquired and congenital syphilis, the clinical course of hearing loss is similar: sudden or rapidly progressing, typically bilateral SNHL, sometimes with vestibular symptoms also present. The loss results from damage to the 8th cranial nerve probably from a persistent and ongoing inflammatory response to the infection. Initially, higher frequency sounds are lost, with normal conversational tones affected later.

Control of infection

Congenital syphilis still represents a considerable global health problem and the WHO recently proposed an action plan for elimination of the disease.[72] Comprehensive antenatal maternal screening programmes, coupled with effective treatment of infected mothers and their partners are the key to detection and prevention of congenital syphilis. Such screening programmes have been shown to be cost-effective even when syphilis seroprevalence is relatively low.[73]

When infection is identified, penicillin is the treatment of choice, and whilst antibiotic therapy during pregnancy can be problematic,[74] effective therapy is essential if a favourable outcome to the pregnancy is to be achieved. Implementation and expansion of such programmes in developing countries will be essential if the goal of global elimination of congenital syphilis is to be achieved. However, the recent resurgence in primary and secondary syphilis amongst young women in developed countries suggests constant vigilance is required there also and late congenital syphilis must increasingly be considered as a cause of sudden onset childhood hearing loss.

OTHER CONGENITAL INFECTIONS AND HEARING LOSS

Toxoplasmosis

The role of the protozoan parasite *Toxoplasma gondii* as a cause of hearing loss has been debated for decades.[75] *T. gondii* causes a serious congenital infection when, following maternal parasitemia, the parasites infect the placenta and then the foetus. Estimates for the incidence of congenital toxoplasmosis range from 0.1 to 10 cases per 1,000 live births.[76] Maternal infection in the first trimester of pregnancy carries a low risk of transmission to the foetus (4–25%) but a high risk of severe damage (40%) evidenced at birth.[77,78] As pregnancy progresses the rate of transmission to the foetus increases (to 80% in the late stages of pregnancy) but the risk of severe damage conversely reduces (to effectively zero in the last trimester[77,78]). Severe cases of congenital toxoplasmosis are thus rare but show all or some of the tetrad of symptoms described by Sabin[79] – chorioretinitis, cerebral calcification, hydrocephalus and mental subnormality. However, mild infection with *T. gondii* may be under-diagnosed, and, as is seen with HCMV infection, the majority of infected infants may be born symptom-free but develop late sequelae. Retinal disease is the most common of these but late development of hearing loss and some degree of mental retardation have been suggested to occur, sometimes up to two decades later.

Early studies[80] suggested profound hearing loss was a common finding in severely affected infants with congenital toxoplasmosis. A later study testing a small number of children with congenital infection found that although the toxoplasmosis children suffered eye defects, there was no association with hearing loss.[81] Various other retrospective studies also found an association with hearing loss.[82,83] However, the majority of more recent studies, particularly where larger numbers of children are included and followed prospectively and where other potential secondary causes (such as otitis media) were considered, have found no association between *T. gondii* congenital infection and hearing loss.[84–86] These studies have, however, shown a clear link between congenital toxoplasmosis and retinochoroidal lesions of the eye. Thus, the current evidence suggests that hearing loss should not be considered as a complication of this infection.[75]

Herpes simplex virus

Congenital deafness has been attributed to infection with HSV.[87] Most cases of infection appearing in the neonatal period are a result of perinatal or post-natal acquisition of virus. Baldwin and Whitley[88] considered that only 5% of all babies born with neonatal HSV had been infected *in utero*. This was based on the criteria that such infection would result in symp-

toms (skin vesicles or scarring, chorioretinitis and/or hydrancephaly) within the first 24 to 48 hours of life. The reported incidence of neonatal herpes varies from extremes of 1 case in 2,500 live births[89] to 1.65 per 100,000 live births reported in a UK survey.[90] Thus, *in utero* infection with HSV is most probably a rare occurrence but the mortality rate is high and most children who survive have neurological sequelae including SNHL.

Herpes varicella zoster virus (VZV)

Primary maternal infection with herpes VZV during the first trimester and early part of the second trimester results in transmission of the virus to the foetus in almost 2% of cases.[91] At birth, infants have cutaneous scars and exhibit a range of congenital damage including eye abnormalities, limb deformation, cortical atrophy, mental retardation and deafness. It is possible that milder cases occur with residual auditory morbidity only apparent in later life. Sensorineural deafness following congenital varicella has been reported only rarely;[92] six children were reported with sensorineural deafness whose mothers had either severe chickenpox or severe herpes zoster during the first trimester.

OTHER INFECTIONS

Whilst there are occasional reports of deafness associated with *in utero* infection caused by various other organisms including enteroviruses, lymphocytic choriomeningitis virus, parvovirus B-19, *Borrellia burgdorferi* (Lyme disease), *Psuedomonas aeruginosa* and Mycoplasmas,[93,94] there are no systematic studies and hence no real evidence that any other infectious organisms induce deafness when contracted as a congenital infection. However, many congenital infections are complicated by neurological involvement, and hence, the possibility of damage to the ear and the nervous system associated with it should always be considered in a hearing-impaired infant born to a mother who contracted an infection during pregnancy.

CHILDHOOD INFECTIONS

Bacterial meningitis

Epidemiology

Bacterial meningitis is a serious and life-threatening disease. It is estimated that it is responsible for 170,000 deaths each year and case fatality rates are 5–10% in the developed world and much higher than this in the developing world.[95] Survivors show high levels of residual morbidity including 10–20% who develop permanent sequelae such as epilepsy, mental retardation and hearing loss. At least 50 different species of bacteria can cause meningitis, but more than 90% of cases in children beyond the neonatal period and under the age of 5 are caused by just three organisms: *Neisseria meningitidis*, *Streptococcus pneumoniae* and *Haemophilus influenzae* type B.

H. influenzae was the most significant cause until the introduction of the *H. influenzae* type B protein-polysaccharide conjugate vaccine (Hib vaccine). Between 1992 and 1995 a mass vaccination programme was introduced across most European countries and this quickly resulted in a decrease in annual cases by around 90% so that currently the incidence of Hib

meningitis across Europe ranges from 0.01 per 100,000 population in Denmark and Germany to 0.74/100,000 in Switzerland.[96]

Neisseria meningitides is a common commensal organism of the mucosal membranes in the nasopharyngeal tract. It is transmitted in respiratory droplets from carriers to uninfected individuals and is usually harmless. However, in a small number of cases infection can lead to septicaemia and meningitis in previously healthy children and young adults. Meningococcal meningitis results in death in 9–12% of cases overall but this rises to 40% if septicaemia is present.[97] The incidence of permanent sequelae is 11–19% and includes hearing loss, neurological deficit and loss of limbs from sepsis.[96]

N. meningitides has a number of serotypes and five of these are associated with meningitis: A, B, C, Y, W-135. Although meningococcal disease is a global problem that occurs in all countries the significant serotype varies. Group A are the only type of meningococci to cause epidemic disease. Although rarely associated with disease in developed countries, they are responsible for large-scale outbreaks in developing countries, particularly in sub-Saharan Africa, where a high burden of disease is seen across a so-called meningitis belt running from Senegal in the west to Ethiopia in the east. Mass travel of individuals from this region to Saudi Arabia for the annual Hajj pilgrimage ensures circulation of the virulent strains between countries. There is some evidence that other meningococcal serogroups are gaining importance in this region; serogroup W-135 was particularly associated with infections acquired during the Hajj pilgrimage in 2000. This also resulted in W-135 appearing in Europe (particularly the UK and France) at this time.[98,99] However, the outbreak subsided rapidly and has not so far been repeated. In contrast, types B and C are more predominant in Europe.

The overall incidence of invasive meningococcal disease in Europe between 1995 and 2005 as measured by the European monitoring group on meningococci network (EMGM) ranged from 1.4 to 2.7 cases per 100,000 population.[96] Serotype B is more commonly seen although most deaths are associated with group C infections. Serogroup Y is occasionally seen in Europe.

Streptococcus pneumoniae is a Gram-positive encapsulated coccus. It is spread by the respiratory route and like *N. meningitides* is normally carried as a harmless pathogen.

In the very young, the elderly and in immunocompromised patients it is a major global cause of morbidity and mortality and is responsible for invasive and non-invasive disease. Otitis media, sinusitis and bronchitis are common but rarely life-threatening, whilst pneumonia, febrile bacteriaemia and meningitis are all associated with high mortality. Ninety serotypes have been identified although most disease is thought to be due to 11 of these. Respiratory disease due to *S. pneumoniae* is responsible for the deaths of one million children each year in developing countries and it is the most common cause of bacterial meningitis in infants and young children worldwide, particularly amongst infants younger than 3 months of age.[100–101] Pneumococcal meningitis has the highest mortality rate of all forms of bacterial meningitis, being approximately twice that of meningococcal meningitis. Up to 50% of survivors have permanent neurological deficits, the most common of which is SNHL.[102,103]

Bacterial meningitis and the ear

Bacterial meningitis is the most important cause of potentially preventable hearing loss acquired post-natally. Overall SNHL occurs in approximately 10% of all cases of bacterial meningitis,[100] with a range from 3.5 to 37.2% reported in a review by Fortnum et al.[104] However, the incidence is highly dependent on age, with young children more susceptible, and on the causative

organism: a prospective study carried out in the United States[105] found 31% of children with pneumococcal meningitis had hearing loss, compared with 10.5% with meningococcal and 6% with *H. influenzae*. Subsequent studies have reported similar rates for each organism and confirmed that *S. pneumoniae* is particularly associated with auditory complications.[106–108]

Damage to hearing is thought to occur early in the infection, probably within the first 24–48 hours.[109,110] As this is typically around the time of admission to hospital, hearing loss is usually apparent within 6 hours of first assessment.[111] Both clinical studies and animal models have shown that duration of infection before antibiotic treatment begins is strongly linked to development of auditory impairment, and early administration of therapy is essential to prevent permanent loss.[111–113] In some cases the hearing loss is temporary, with improvement noted over a 2-week recovery period. No improvement in recovery of hearing beyond the initial 2-week period has been observed.[110,114] Conversely, no reports of 'late' onset deafness have been described, although hearing loss in a neonate occurring in the course of the meningitis may not be apparent until the child is older. The pattern of hearing loss varies, it may be unilateral or bilateral, severe and permanent involving both high and low frequencies.[105,110,114–116]

The mechanism by which the organism spreads from the meninges into the inner ear is not completely understood; however, histopathological and auditory brainstem studies have shown that the cochlea is the site of the lesion in meningitis associated hearing loss and it is likely that the bacteria enter the cochlea from the cerebrospinal fluid following inflammatory damage to the blood–labyrinth barrier.[117]

Control of infection

Bacterial meningitis is always a medical emergency necessitating prompt, and aggressive, antimicrobial therapy. Antibiotics should be started as soon as meningitis is suspected and should cover the three main organisms listed above; thus, a third-generation cephalosporin, such as cefotaxime or ceftriaxone, is recommended. However, there is considerable evidence that deafness due to meningitis is partly or wholly due to an inappropriate host inflammatory response to the bacteria or its products. For this reason, administration of adjunctive corticosteroid therapy, such as dexamethasone, is recommended, particularly for pneumococcal meningitis.[110,118,119]

As with most infections, prevention of bacterial meningitis is preferable to, and more successful than, treatment. The epidemiology of meningitis, at least in the developed world, has been transformed in recent years by the introduction of routine vaccination against several of the major causes. The vaccine against *H. influenzae* type B is now widely administered in childhood and has resulted in a greatly diminished incidence of this type of meningitis.

Meningococcal conjugate vaccines effective against serotypes A, C, Y and W-135 are now routinely given and, in the UK, have reduced serogroup C meningitis by more than 80% in vaccinated populations[120] and have brought about a general reduction in unvaccinated populations due to the effect of herd immunity.[121] Vaccine for serogroup B is not yet available but is under active development.

In 2000 and 2001, two polyvalent pneumococcal vaccines were licensed for use in the United States. The first of these induces immunity against 23 of the 90 known serotypes of *S. pneumoniae.* The second is directed against the seven most common serotypes causing childhood disease and was introduced into the UK childhood immunisation schedule in late 2006.[122] It is expected that this will bring about a similar reduction in pneumococcal meningitis.

Otitis media (OM)

OM is a major cause of hearing loss in children. This subject is covered in Chapter 10 in this book and will be dealt with only briefly here. Acute OM is inflammation within the middle-ear space initiated by infection and characterized by a red and bulging tympanic membrane. It is now widely recognised that the initiating event is usually a viral infection of the upper respiratory tract, typically respiratory syncytial virus (RSV), a common infection of infants.[123,124] The viral infection causes congestion of the nasal and nasopharyngeal mucosa; this leads to congestion in the eustachian tube, which alters the pressure equilibrium between the nasopharynx and the middle-ear cavity. As a result, drainage of secretions from the middle ear into the nasopharynx and ciliary clearance of invading bacterial pathogens is reduced, allowing them to gain access to the middle ear where they multiply and cause OM.

The three major pathogens involved in acute OM are *Streptococcus pneumoniae, Haemophilus influenzae* and *Moraxella catarrhalis.*[125,126] As with bacterial meningitis, *S. pneumoniae* is the most common cause (found in 30–50% of cases) and is associated with more severe clinical findings and complications. Introduction of the pneumococcal vaccine has had little impact on the overall incidence of acute OM.[127,128] This is because 'replacement phenomenon' has occurred, whereby elimination of the serotypes present in the vaccine has allowed other serotypes or different organisms to fill the niche so that the overall incidence of OM has not greatly decreased.[129–131] *H. influenzae* is found in 15–25% of cases of OM, particularly bilateral OM and in cases where concurrent otitis conjunctivitis syndrome is seen. The currently available vaccine against *H. influenzae* is a conjugated type B vaccine and as virtually all OM caused by *H. influenzae* is due to non-typeable strains, the vaccine is ineffective in preventing OM.

M. catarrhalis is found less often (10–15%) and is associated with milder symptoms, but may be more commonly associated with a mixed infection.

Other acquired childhood infections associated with hearing loss

Bacterial meningitis and OM are the principal causes of acquired hearing loss in childhood. However, many other childhood infections are also associated with deafness to varying degrees and these are described below.

Mumps virus

Mumps is a distinctive childhood illness characterised by swelling of one or both parotid glands. In areas where vaccination is administered routinely the infection is now rare. However, mumps remains endemic in many areas of the world. The typical salivary gland swelling usually occurs within 24 hours, progresses for 2–3 days and subsides within 1 week. The most common complications are orchitis, pancreatitis and aseptic meningitis.

Hearing loss is a rare but serious complication, occurring in 5 out of every 10,000 cases of mumps. The onset of hearing loss is sudden and typically unilateral (80% of cases). It occurs as part of the acute infection, usually in association with aseptic meningitis and is often accompanied by vertigo, tinnitus, ataxia and vomiting. The overall severity of the infection does not determine whether hearing loss will be a feature. Mumps deafness tends to be profound and permanent, preferentially affecting the higher frequencies. Unilateral hearing loss may remain undetected for years following the infection, especially if the child is very young.[132–134]

Asymptomatic mumps virus infection has also been demonstrated to be a cause of sudden and total bilateral deafness[135] and it has been suggested that such asymptomatic infection may be responsible for some cases of unexplained, mild, sudden hearing loss.[136]

Although mumps sequelae such as hearing loss are rare, the consequences when they do occur are severe. For this reason, mumps immunisation, given as part of the MMR vaccine, is now routine in most developed countries. However, there are still many parts of the developing world where the vaccine is unavailable and mumps infections continue to be a significant cause of hearing loss.

Measles virus

Measles virus infection (rubeola) is the most contagious of all the childhood diseases. It is a major global health problem resulting in 20 million cases each year occurring mostly in children in the developing nations, and in 2006, 242,000 of these cases resulted in fatality.[137] The characterising features of the infection are the appearance of the pathognomonic enanthem, Koplik's spots, on the buccal mucous membranes, followed within 1–2 days by a generalised maculopapular rash. Infectious virus continues to be shed until approximately 4 days after the appearance of the rash, contributing to the epidemic spread. The disease is usually self-limiting, but occasionally complications are seen. These include blindness, encephalitis, pneumonia, diarrhoea and ear infections, particularly OM which occurs in 5–15% of cases.

OM, caused by a bacterial infection secondary to measles is a significant cause of conductive hearing loss following measles infection. However, SNHL directly induced by virus replication or following measles encephalitis is also reported. Permanent hearing loss following measles virus infection occurs in up to 1 in 1,000 cases. The loss is usually sudden and bilateral, and occurs at the same time as the appearance of the measles rash. Prior to the introduction of measles vaccine, the virus was thought to be the cause of 3–10% of all acquired deafness in children.[132,138]

Complications resulting from childhood measles infection remain a problem, particularly in developing countries. However, measles vaccine, either presented alone or given as a combined measles, mumps and rubella (MMR) vaccine or measles, mumps, rubella and varicella (MMVR) vaccine, have been demonstrated to be safe and effective. In 2006 the WHO estimated that 80% of the world's children were covered by measles vaccine, but infections continue to be severe in the areas where coverage is lacking. Consequently, the Global Immunisation Vision Strategy, presented by WHO and UNICEF in 2005, includes measles with the aim of ensuring 90% of the world's children are vaccinated by 2010 and that measles mortality is reduced by 90% compared with mortality levels in the year 2000.[137]

Human immunodeficiency virus

There are currently 33 million people in the world infected with human immunodeficiency virus (HIV) (UNAIDS 2007). The virus has a tropism for T-helper lymphocytes and destruction of these cells causes a breakdown in immunity leaving the individual susceptible to infection with other opportunistic organisms. Most HIV infection in children results from perinatal transmission. The HIV-positive child will encounter those infections that are common in childhood, but impaired immunity may render them severe, chronic and more frequently recurrent. Thus, the child is potentially susceptible to all of the infectious causes of hearing impairment described previously.

There are few epidemiological studies investigating hearing loss in HIV-infected individuals but where they have been undertaken up to 49% of participants have been shown to have some degree of audiological abnormality.[139,140] It must be assumed that the majority of hearing loss identified in HIV-infected or AIDS patients is due to secondary viral or bacterial infection resulting from impaired immunity. In a minority of patients, though, HIV has been suggested to be an ototoxic virus since hearing loss is seen in HIV-infected patients who do not have evidence of significant immunosuppression[141] and for whom no other clear cause of hearing loss can be identified.[142,143] The virus is certainly neurotropic as well as lymphotropic and most AIDS patients show some degree of neurological involvement, especially if they have not received effective antiviral therapy. It is thus plausible to suggest that the virus may occasionally invade and damage the auditory nerve, and further studies in this area are warranted.

However, most hearing loss in HIV-infected patients is likely to result from opportunistic infection with another pathogen. As described earlier, congenital or secondary syphilis is an important cause of hearing loss and syphilis has a higher incidence in HIV-infected patients. There is also evidence that progression of hearing loss is accelerated in these patients.[144]

Similarly, infection with VZV is common in immunocompromised patients. Primary infection with VZV causes chicken pox and is usually acquired in childhood. VZV then establishes a latent infection in the trigeminal or dorsal root ganglion of the host. If reactivation occurs, the result is a zoster or 'shingles' outbreak. Patients with HIV are more prone to zoster and if reactivation occurs in the 7th cranial nerve a condition known as Ramsay Hunt syndrome, or herpes zoster oticus, results. Blisters develop in the ear canal and auricle and involve the nerves innervating the inner ear, causing facial paralysis, hearing loss and vertigo.

In the absence of prophylactic treatment, almost all AIDS patients show some evidence of CMV involvement and in approximately one-third the virus enters the CNS.[145] HCMV retinitis and encephalitis are common late-stage manifestations of AIDS in untreated patients and there are individual reports of the virus entering the 8th cranial nerve and causing permanent hearing loss.[146]

Fungal infections are also more common in HIV-infected patients and *Aspergillus*,[147] *Cryptococcus neoformans*[148] and *Pneumocystis carinii*[149] have all been identified as causes of hearing loss in this setting.

Mycobacterium tuberculosis

Cases of *Mycobacterium tuberculosis* infection are increasing globally. The disease is epidemic in many African countries[150–152] and has reached high prevalence even in some developed countries.[153] This resurgence of the disease is linked with the HIV pandemic, and emergence of drug-resistant forms of the bacterium have increased the mortality rate and threaten existing control of the disease.

Tuberculosis (TB) is associated with hearing loss in both children and adults and occasionally deafness is the presenting feature of infection.[154] TB can induce chronic ottorhea and perforations of the tympanic membrane are seen. Without prompt anti-tuberculous therapy complications of infection including the development of tuberculoma may result in permanent hearing loss.[155] In addition, hearing loss is reported following tuberculous meningitis. In one US study, severe hearing loss was seen in 25% of children with neurological deficit who survived TB meningitis.[156] BCG vaccination of infants is recommended in countries where prevalence is high, for both HIV-infected and uninfected children[157] and this does seem to reduce the incidence of TB meningitis and hence hearing loss.

Borrelia burgdorferi (Lyme disease)

Lyme disease, caused by infection with the tick-borne spirochaete *Borrellia burgdorferi*, results in frequent neurological complications and permanent SNHL.[158] In common with other spirochaete infections, the manifestations of Lyme disease are varied and may be characterised as early, intermediate or late stages of the disease. Neurological symptoms, including hearing loss, are a part of the late manifestation of the disease. The exact mechanism of hearing loss is unknown, but it is possibly due to direct damage to the auditory centre, the 8th cranial nerve, or labyrinthitis.

Fungal infections

Fungal infections resulting in deafness are rare, but meningitis caused by *Candida albicans* has been noted as a causative factor, particularly in babies of low birth weight.[159] In addition, as noted earlier, *Cryptococcus neoformans*, *Aspergillus* sp. and *Pneumocycstis carinii* may all cause meningitis with residual auditory impairment in immunocompromised patients, particularly AIDS patients.

Visceral leishmaniasis

A number of protozoal infections are capable of producing CNS infections, which may include hearing impairment amongst their residual morbidity. For example, visceral leishmaniasis caused by *Leishmania donovani* can cause retrocochlear hearing loss. Nerve conduction studies suggest demyelination as a principal cause of hearing loss, which resolves after successful treatment of the infection.[160]

Rubella

Post-natal rubella rarely affects hearing. Where it has been reported it affects adults rather than children and presents as unilateral SNHL,[161] contrasting with the profound bilateral deafness seen following congenital rubella infection.

Herpes simplex virus and VZVs

Neonatal HSV infection is rare but can have severe manifestations. It usually occurs in neonates without protective maternal HSV antibody; infection with HSV-2 is typically more severe than that caused by HSV-1. If untreated, mortality exceeds 50% and children with disseminated infection have the worst prognosis. Even with prompt administration of specific antiviral chemotherapy, survivors may have various degrees of psychomotor retardation and hearing loss is common amongst these.

An analogous disease, severe neonatal chickenpox (varicella) may occur when maternal varicella presents within 5 days of delivery. Neonatal VZV infection is fatal in approximately 30% of cases, and, as with neonatal HSV infection, survivors often exhibit some degree of hearing loss.

Other infections

It is likely that many cases of sudden hearing loss are attributable to infection and the causes will undoubtedly emerge as diagnostic methodology continues to improve. In addition to

congenital infection, both perinatally and post-natally acquired *T. pallidum* infection can result in loss of hearing. Clinical symptoms are of unilateral or bilateral hearing loss but may also present as Ménière's syndrome.[162]

Many viral infections of the CNS can produce residual auditory impairment as a result of central or peripheral nerve damage. Epidemics of CNS infection may have, as late sequelae, increased occurrence of auditory impairment such as has been observed in tick-borne encephalitis in children.[163] Damage incurred through such infection is unpredictable and in many cases an abberant immune response triggered by a severe virus infection is a more plausible explanation of audiological deterioration than direct viral involvement.

Microbial-associated toxicity (e.g. in pneumococcal infection) or toxicity indirectly caused as a result of infection (e.g. in fulminant viral hepatitis where excessive bilirubinaemia has been suggested as a cause of cochlear damage) presents a further source for infection-associated hearing loss.

THE PATHOLOGY OF HEARING LOSS ASSOCIATED WITH INFECTION

Just as there are many infectious causes of hearing loss, there are likely to be many pathogenic mechanisms involved, and in most cases, little is known about them. Indeed, this is an area of infection that is currently neglected and where further research would undoubtedly yield information that could be helpful in preventing, treating and managing hearing loss resulting from infectious causes.

Hearing loss following infection may result from direct cytolytic effects of the pathogen within the inner ear, from immune-mediated damage due to the inflammatory response to the organism, or as a consequence of damage to the auditory nerves following CNS infection.

A number of early studies examined the temporal bones taken post-mortem from infants congenitally infected with rubella or CMV or who died after measles or mumps infection.[164–169] These studies described common histopathological features, principally of endolymphatic labyrinthitis, with pathologic changes limited almost entirely to the membranous labyrinth, in particular, the cochlear duct, saccule and utricle. However, although the common pathology indicates a common mechanism of damage to the specific audiological structures, it is evident from the detail of the reports that the significant lesions may be mediated differently depending on the individual pathogen involved.

Lindsay and Hemenway[166] reported the case of an infant who had died after measles complications. The structures in the cochlea showed degeneration, which was greater at the basal coil and diminished towards the apex so that in the basal coil only slight remnants of stria remained and Corti's organ was absent. Only a small fraction of the normal number of nerve fibres remained and the ganglia were greatly reduced. In the middle and apical coils, more of the stria and organ of Corti were present and the nerve cells were approximately normal in number, but there were areas in the remaining stria vascularis of the apical coil where a localised inflammatory reaction around actively proliferating foci of infection was evident. They suggested that the viral infection of the inner ear occurred via the stria vascularis, beginning at the basal coil. Release of virus and inflammatory cells into the endolymph caused infection further into the coils of the cochlea, with resultant destruction of the nerve cells secondary to the initial infection. The stria vascularis in the cochlea is a site rich in capillaries and, therefore, a likely portal of entry for the blood-borne virus.

Lindsay et al.[164] also described the temporal bone pathology in a case of mumps deafness. Similar to the measles case described earlier, they found that the pathologic changes were confined primarily to the cochlear duct and consisted of degeneration of the stria vascularis, the organ of Corti and the tectorial membrane in the basal coil of the cochlea, with damage diminishing progressively towards the apex. However, the case described was unusual for mumps deafness in that it was bilateral. Earlier work[170] suggested that hearing loss resulting from mumps infection more usually occurred because of viral invasion of the meninges. Encroachment of the localised meningeal lesion on the acoustic nerve would produce a pathology more likely to lead to unilateral hearing loss.

Congenital rubella is now thought to be unique in that its principal effect is that of a teratogen interfering with the genesis of normal organs. Inner-ear malformation manifests as a lack of development of hair cells and supporting cells especially in the apical coil of the cochlea. The tectorial membrane is found rolled up against the limbus in contact with Reissner's membrane and is enclosed in a sheath of flattened cells. The saccular membrane may be hypertrophic and adherent to a degenerated macula or collapsed and the stria vascularis may be partly or totally absent.[165,167] It is likely that rubella acts on the epithelial cells of the developing labyrinth resulting in the cochleosaccular degeneration described.[169]

Myers and Stool[167] were first to examine the inner-ear pathology in a fatal neonatal case of cytomegalic inclusion disease. In contrast to the changes noted in some other viral inner-ear infections, no obvious involvement of the organ of Corti or curling of the tectorial membrane was noted in this or later studies.[168]

For most viral infections, demonstration of the actual virus within the ear has not been achieved, presumably because the temporal bones are examined at too late a stage in the infection. In the case of CMV, however, characteristic cytomegalic nuclear inclusions have been described, located in the epithelial cells of the cochlea, saccule, utricle and semicircular canals and demonstrating the susceptibility of the entire ear to this virus. Indeed, evidence for more extensive distribution of virus than the areas of manifest cellular damage has been provided by the use of CMV-specific immunofluorescent antibodies, which demonstrated the presence of viral antigen in the organ of Corti and in neuromas of the 8th cranial nerve.[81]

Davis et al.[173] isolated CMV from the perilymph of an infant who died from congenital CMV. A later study[174] reported a congenitally CMV-infected infant who showed no evidence of CMV-induced hearing loss and no histopathologic changes to the inner ear. The child died of encephalitis of undetermined aetiology, but which was presumed to be due to HSV. Postmortem CMV was isolated from the inner-ear fluid, but not from the adjacent brain tissue. The infant was 5 months old and the authors suggested that CMV is capable of persisting in the ear for prolonged periods without causing destruction to the cochlea. They speculated that the delayed and progressive hearing loss following congenital infection may have been due to the virus either slowly causing direct damage to the critical inner ear cells or to a delayed host immune response causing immunopathologic damage.

The presence of an inflammatory cell response in most cases which have been investigated has raised the possibility that the damage to the inner ear is partly due to an immunopathological response as well as direct viral cytopathology. In support of this hypothesis, Harris et al.[175] described an animal model for CMV labyrinthitis wherein a positive relationship between the degree and extent of inflammatory reaction in the cochlea and hearing loss was found. No such correlation existed between CMV antigen level and hearing loss. The authors suggested that the inflammatory response may be of more importance in causing inner-ear damage than is the direct effect of the virus. More recently, a case was reported which described the temporal

bone histopathology in a child who died at 14 years of age from the sequelae of congenital CMV infection.[176] No virus could be isolated from the inner ear (or from any other tissues) but the damage to the ear structures was more extensive than that previously reported from the infant cases. Atrophy of the stria and loss of cochlear hair cells was noted along the entire length of the basilar membrane. There was evidence of damage to vestibular as well as cochlear structures and the overall finding was of chronic immunopathologic damage.

Collectively, the histopathology of the CMV-infected inner ears suggests that hearing loss in the acute stage of the disease is caused by viral cytolysis, whereas the delayed or progressive hearing loss often associated with congenital infection may result from damage caused by the immunological response to the infection.

There are few recent reports describing the pathology of the inner ear in cases of hearing loss following meningitis in humans,[115,177] but together with some early studies and data derived from animal models, they provide evidence that the principal cause is a cochlear lesion.[178–183] It is known from human studies that bacteria accumulating in the sub-arachnoid space can invade the cochlea via the cochlear aqueduct,[178] along the 8th cranial nerve or perhaps even via the blood vessels of the blood–labyrinth barrier[117] and that the resulting labyrinthitis causes damage to the inner ear.

Experimental data show that introduction of antigen into the inner ear of an animal previously sensitised systemically produces an immune response in the ear against the antigen. The resulting cellular infiltration and inflammation, release of cytokines and triggering of the complement cascade are all likely to damage the delicate cochlear tissue. However, it is not evident why this immune-mediated pathology should cause damage to the auditory processes more frequently than to other neurological structures, nor why the pneumococcal organism causes hearing loss more often than do other aetiological agents of meningitis.

Although there is little doubt that inflammatory-mediated damage plays an important role in hearing loss resulting from infection, further data suggest that in some infections, the bacterial products themselves are directly ototoxic. In particular, the pneumococcal toxin pneumolysin has been found to induce severe damage to the inner ear in experimental meningitis. When the toxin was introduced directly into the scala tympani, lesions appeared on the hair cells within a few minutes suggesting that the toxin is able to cross the basilar membrane and that its effect is caused by a direct, rather than an indirect action.[184] A similar experiment using the *E. coli* endotoxin also produced lesions but the effect was much less severe and occurred more slowly. It is possible that the potency of pneumolysin may account for the increased ototoxicity of pneumococcal meningitis.

Whether directly or indirectly mediated, pathological damage to the various cell types in the cochlea will result in loss of hearing. As transduction of sound pressure into electrical impulses is dependent on intact stereocilia, damage to the hair cells would inevitably disrupt this process. Similarly, damage to the nerve endings at the base of the hair cell would interfere with the generation of nerve impulses along the auditory nerve.[185] As auditory hair cells are not thought to be capable of regeneration in mammals, including humans, damage to the hair cells would lead to permanent hearing loss. Damage to nerve endings, on the other hand, is more likely to be temporary. Stereocilia have been shown to be highly susceptible to pneumolysin[113] which might explain the higher incidence of permanent hearing loss following pneumococcal meningitis.

In summary, hearing loss resulting from infection, although not completely understood, is likely to be mediated by a number of mechanisms, which are dependent on the organism involved, the stage of disease and the immune response mounted against the pathogen. Rubella

is the only organism known to cause deafness by interfering with foetal inner ear development. Where CNS infection occurs, damage to the acoustic nerves is possible. Alternatively, the organism may invade the inner ear from the meninges or from the blood system and cause direct cytopathic damage, in particular to the delicate cochlear structures. This damage may be compounded by the indirect, immunopathologic effects of the immune response mounted against the pathogen. In some cases, particularly perhaps following congenital CMV infection, the virus persists in the inner ear, and a delayed immune response occurring several years later may be responsible for late-onset hearing loss.

LABORATORY DIAGNOSIS

The identification of an infectious cause of hearing loss is often problematic. In many instances, intrauterine and perinatal infections are inapparent or produce symptoms that are only mild and non-specific. Associated hearing impairment may not be discerned for months or even years after the initial infection. In such circumstances conventional diagnostic procedures designed to detect the pathogen may fail unless the hearing deficit is associated with chronic infection. Serological investigation may be used but because of the time elapsing between infection and the observation of hearing impairment, results may be difficult to interpret.

Initial and very valuable information may be obtained through careful examination of antenatal immunisation records. A detailed clinical and family history should be the first step in any investigation including detail of any foreign travel and possible exposure to likely causal agents. Physical and audiological examination with neurological and ocular tests should be performed as these can guide laboratory investigation. Relevant information should then be communicated to the diagnostic microbiology and virology services to enable appropriate analyses to be carried out.

INTRAUTERINE AND CONGENITAL INFECTIONS

Overt maternal infection during pregnancy may lead to investigation that alerts the clinical staff to the possibility of congenital infection. Rubella in pregnancy is usually manifest as a morbilliform rash. Examination of suitable specimens (throat swab, saliva, urine or blood) will reveal the presence of virus. Isolation of virus using cell culture is possible in specialised monolayer cell cultures but the yield is low and nowadays direct detection of the virus using molecular amplification and detection techniques (e.g. the polymerase chain reaction (PCR) technique) would more usually be employed. Examination of blood will reveal the presence of rubella virus-specific immunoglobulin M (IgM) antibody. Immunoglobulin M is the first antibody to be produced in response to infection and is also the shortest lived antibody persisting for 3–6 months following acute infection.

Arising soon after the appearance of IgM, immunoglobulin G (IgG), is the antibody that persists and is a marker of past infection. In the early stages of infection the strength with which the IgG antibody binds to the disease agent, the so-called avidity of the antibody, is low. The strength of binding of this antibody increases with time and reaches a maximum within about 3 months of the acute infection. Measurement of virus specific IgM antibody together with virus-specific IgG antibody and measurement of the avidity of the IgG antibody detected can provide precise information about the timing of the infection, if the investigation

is carried out within the first 3 months after the acute episode. Beyond that time, IgM antibody may or may not be detectable (depending upon the sensitivity of the IgM assay utilised) and IgG will be detected but will be of high avidity and will thus only indicate infection at some time in the past.

It is always necessary to confirm the presence of IgM antibody using a second independent immunoassay technique, as false positive test reactivity is commonly encountered in detection of IgM antibody. Immunoglobulin M antibody as the first antibody to arise in an acute infection is inherently broadly cross-reactive. An acute infection of any cause can lead to the production of IgM antibody and this non-rubella antibody may cause low-level reactivity to be detected in a rubella-specific IgM antibody assay simply because of this cross-reactivity. Rheumatoid factors (IgM anti IgG antibodies) can, depending upon the immunoassay format used to detect rubella-specific IgM antibody, produce false positive test reactions. False negative test results may also, again depending upon the immunoassay format used, occur when high levels of rubella-specific IgG antibody are found together with IgM antibody. Careful and expert interpretation of immunoassay results is thus always required.

Using a constellation of serological tests it is possible to pinpoint the time of infection. This is particularly critical in rubella virus infection as the stage of pregnancy will define the risk to the foetus. If rubella virus infection occurs in the first 12 weeks of pregnancy, up to 90% of patients will have some manifestations of the congenital rubella syndrome. For infection at 12–16 weeks, the risk is approximately 20%.[7] For women wishing to continue with the pregnancy foetal infection can be investigated in various ways. Foetal blood samples obtained by fetoscopy can be examined for rubella-specific IgM. However, the foetus does not produce sufficient IgM for detection before 22 weeks gestation. Virus may be isolated from amniotic fluid or chorionic villus biopsies using specialised cell cultures or detected by PCR amplification of the viral nucleic acid (RNA). The sensitivity of PCR testing of amniotic fluid is reported as being between 87 and 100%.[186] Chorionic villus biopsies must be interpreted with care as the presence of placental rubella virus might not reflect foetal infection.

Cytomegalovirus infection during pregnancy can present considerable difficulty in its management. In contrast to rubella virus, which is clearly teratogenic if infection occurs during the first trimester, cytomegalovirus can cause both teratogenic and cytolytic damage to the developing fetus that makes infection at any stage of pregnancy a risk. Primary infection with the virus is associated with the highest risk of congenital damage but as described earlier reactivation of a maternal latent infection can also affect the foetus. Maternal HCMV infection produces, in the majority of cases, mild infection that may not warrant investigation at the time it occurs. Where laboratory investigations are instigated, PCR examination of blood, urine or saliva and/or virus culture will allow detection of virus. In a primary infection cytomegalovirus-specific IgM antibody will be detectable in blood together with cytomegalovirus IgG antibody of low avidity.

Prenatal diagnosis should be attempted only where there is a strong suspicion of primary CMV infection and/or ultrasonographic abnormality.[187] Virus may be detected in amniotic fluid or chorionic villus biopsies using PCR. The sensitivity of prenatal diagnosis was found to be 50, 76.2 and 91.3% when infection occurred at <8, 9–12, and >13 weeks of gestation, respectively, in the largest series published to date.[187] However, as outlined earlier, virological confirmation of foetal infection is not in itself a predictor of outcome.

All women should be screened serologically for evidence of syphilis during the early stages of pregnancy. Where risk of infection is thought to be high, repetition of serological testing during the third trimester is suggested. Transmission of infection can be almost completely

prevented by treatment before the 16th week of pregnancy.[188] Diagnosis of primary syphilis infection, before the appearance of antibody, depends upon the direct detection of *Treponema pallidum* in syphilitic mucocutaneous lesions or lymph node aspirates, using dark ground microscopy, direct fluorescent antibody staining, silver staining or PCR.[189]

Serology remains the mainstay of laboratory diagnosis. In early infection, diagnosis may be achieved by the detection of IgM antibody to *T. pallidum*; IgG antibody appears at almost the same time. Serological screening for syphilis usually utilises a combination of IgG- and IgM-specific ELISA tests coupled with *T. pallidum* particle agglutination assays and non-treponemal tests such as the rapid plasma regain test or the venereal disease research laboratory test.[189] Detailed guidelines on the management of suspected congenital syphilis are available.[190]

Intrauterine infection with either herpes simplex virus or herpes VZV is rare. Primary maternal infection with either agent can be investigated via detection of IgM-specific antibody but more invasive procedures to demonstrate intrauterine infection are not usually attempted. Maternal infection that becomes apparent close to or immediately after delivery requires thorough investigation utilising virus culture, PCR and electron microscopic examination of vesicle fluids. Maternal serology will usually reveal the presence of virus-specific IgM in initial samples, followed by IgG in later sera. The neonate may initially be seronegative with the appearance of IgM antibody delayed for 6 months or more. Negative neonatal serology does not therefore preclude a diagnosis of HSV or VZV infection. Investigation of possible maternal infection is as important and urgent as investigation of the neonate. The maternal antenatal serological specimen should be retrieved wherever possible to allow comparative pre- and post-natal serology. Diagnosis of congenital infection is best established by comparison of the results obtained in the maternal and the neonatal specimens (Table 8.3).

Investigation of possible congenital infection in the neonatal period or early infancy

During the first 6–9 months of life caution must be exercised in interpretation of serological test results since the detection of antibody may merely reflect passively transferred maternal antibody rather than infection (Table 8.4). Serological investigation of a child that reveals *T. pallidum* infection after 6–9 months of age provides supportive evidence for congenital or early infection as this is a rare infection of childhood. At 12–15 months of age detection of antibody to rubella may or may not reflect congenital infection since immunisation against rubella is given at this time.

Similarly, the detection of HCMV, HSV or VZV antibody cannot provide clear evidence of congenital infection since most primary infections with these viruses occur in childhood. An extremely valuable avenue of diagnosis of congenital infection is afforded by examination of Guthrie card dried blood spot specimens. In many developed countries, a programme of screening neonates for inborn errors of metabolism is in place. The Guthrie card test involves the collection of blood from heel prick performed in the neonatal period; the blood is collected and dried onto filter paper. After the use of a small sample of blood to test for enzyme deficiency, the cards are usually stored. Examination of blood samples eluted from these cards for IgM antibody or for the nucleic acid of the disease-causing agent provides a snapshot in time of the status of the baby close to the time of birth. Using this technique, it has proved possible to define congenital infection many years after the time of birth.[191,192]

Table 8.3 Investigation of congenital infection and infection evident in the neonatal period: diagnostic specimens and appropriate tests.

Infant

	PCR	Culture	Serology (IgG and IgM)**
Urine*	+	(+)	–
Throat swab	+	(+)	–
CSF	+	(+)	+
Blood	+	(+)	+
Vesicles or other lesions	+	(+)	–

Note: In newborn infants, investigation of both mother and baby is essential.

(+) Test may be appropriate in certain circumstances.

*Important specimen for detection of HCMV congenital infection. Virus excretion can occur for 2 or more years post-natally.

**Determination of IgG avidity is a valuable adjunct to IgM detection in defining recent infection. In recent infection low avidity IgG antibody will be detected, while in old or recurrent infection high avidity IgG antibody will be present.

Mother
Serological investigation for recent infection with:
Herpes simplex virus
Herpes varicella-zoster virus
Human cytomegalovirus
Human immunodeficiency viruses 1 and 2
Hepatitis B
Rubella virus
Treponema pallidum

Where possible examine routine antenatal blood specimen in parallel with post-natal blood specimen.

Table 8.4 Acquired infection: diagnostic specimens and appropriate tests.

	PCR	Culture	Serology (IgG and IgM)*
Urine	+	(+)	–
Blood	+	(+)	+

*Serological diagnosis is likely to be the mainstay of diagnosis in the asymptomatic child or in a child being investigated at a late stage. Serological investigation should include investigation of both child and mother and, if appropriate siblings or other immediate family members. Where possible, examine mother's routine antenatal blood specimen in parallel with post-natal blood specimen.

The availability of this test is invaluable in investigating hearing loss in children where damage to auditory function may have been present at birth. It must be emphasised, however, that while detection of a causal organism is valuable, failure to detect a disease-causing agent does not rule out the possibility of congenital infection. The preservation of the agent in blood stored on cards may vary case to case, the efficiency of elution of dried blood from the card may vary, and the efficiency of detection of antibody or nucleic acid will vary between laboratories.

Post-natal infection

Bacterial meningitis particularly when presenting with bacterial septicaemia is a major medical emergency. Whilst meningitis is usually secondary to a bacteraemia, infection can also result from spread from a focus of infection in the ear, sinuses or from a skull fracture. A variety of rapid diagnostic procedures including antigen detection, nucleic acid hybridisation and PCR techniques have been developed to supplement conventional blood and CSF culture procedures. However, not all cases are amenable to diagnosis. Prompt administration of antibacterial agents, though essential for patient management, may preclude identification of the causal agent, whilst cryptic sites of infection may limit the availability of organism in conventional diagnostic specimens (blood and CSF). A wide variety of organisms may cause such infection and the type of organism involved will vary with age. In contrast to viral and parasite serology, bacterial serology is rarely, if ever, useful in retrospective determination of cause.

Acute stage viral infections in children may be diagnosed by virus culture, identification of specific viral antigens using ELISA-based techniques or by detection of specific nucleic acids using molecular assays such as PCR. For diagnosis of measles or mumps virus infection throat swabs, saliva, blood and urine are valuable specimens. It must be remembered, however, that it is a feature of most virus infections that by the time symptoms are evident, the peak of viral replication is over, and thus, if a virus is to be identified, early collection of appropriate specimens is essential. Serological investigation of infection can be achieved later, either via detection of virus specific IgM or by detection of a rising level of virus-specific IgG antibody in suitably spaced specimens. Oral fluid, collected using specialised swabs, has been developed for MMR virus antibody detection. The sensitivity of testing using this procedure, however, is lower than can be achieved by testing whole blood samples.

CONCLUSION

Although further epidemiological studies are urgently needed, it is clear that infection, both congenital and post-natal, is a significant cause of paediatric deafness. Hearing loss is rarely the only feature of the infection, but the delayed onset of deafness attributed to some infections, particularly congenital, make it likely that the role of infectious organisms in the epidemiology of hearing loss is presently underestimated. A number of viruses, only briefly considered here, have been occasionally associated with loss of hearing in children or young adults and it is likely that as diagnostic methods improve further associations will be made. The decline in hearing loss attributed to rubella, measles and mumps viruses and to some forms of bacterial meningitis demonstrates the success of an effective immunisation programme. Improvements in the diagnosis of paediatric infection, implementation of vaccine programmes into developing countries and future vaccine development promise continued decline in the incidence of preventable childhood hearing loss.

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9 Adverse perinatal factors and hearing loss

T. Sirimanna

INTRODUCTION

Approximately 50% of all congenital hearing loss is from acquired aetiology, which includes infective and other causes that lead to a hearing loss during pregnancy, such as rubella and cytomegalovirus infections (see Chapter 8), and adverse factors that lead to a hearing loss during the perinatal period. The birth of a baby from the secure environment in the mother's womb with a steady supply of essential nutrients and oxygen and the swift removal of unwanted products of metabolism involves a process which has many stages that can go wrong, leading to disastrous outcomes, including deafness.

A number of factors relating to the baby have been causatively connected to hearing loss. These include low birth weight and gestational age, intrauterine growth retardation (IUGR), chronic hypoxia, acute hypoxic episodes during labour, hyperbilirubinaemia from various origins such as rhesus, ABO incompatibility and G6PD deficiency, ototoxicity, neonatal infections, prolonged ventilation and noise exposure from sources such as incubators. However, the evidence that these factors act independently to cause hearing loss is scant; in most cases more than one aetiological factor is normally found, although this is true for some factors more than others. Therefore, in most cases it is difficult to pinpoint an individual factor as being the sole cause of the hearing loss in a baby. Furthermore, in some cases, there may have been a genetic predisposition, e.g. 1555A>G mitochondrial mutation, which causes abnormal susceptibility to aminoglycoside ototoxicity and leads to a hearing loss from an agent that would not normally have caused the hearing loss. In addition, a child who has been exposed to a number of adverse factors perinatally may have suffered a hearing loss from a genetic cause, e.g. autosomal recessive deafness as in connexin 26 mutation, which may not be apparent unless systematic aetiological investigations are carried out. Often the aetiological investigation of hearing loss does not take place soon after birth for a number of reasons and therefore keeping a very detailed and accurate record of perinatal and post-natal adverse events is extremely important.

The phased introduction of the National Newborn Hearing Screening Programme (NHSP) in England (completed in 2006 and established in other countries in the United Kingdom) has provided every newborn baby with the chance to have a hearing screen at or soon after birth. Those who do not pass the hearing screen are referred for further assessment. This assessment is expected to be completed in 80% of the babies by 6 months of age; 98% of them will be assessed by 12 months of age. In addition, it is expected that the parents of children with a permanent hearing loss will be offered an opportunity to have their baby's hearing loss investigated aetiologically (Quality Standards of the NHS Newborn Hearing Screening Programme, www.hearing.screening.nhs.uk). This means that when seen by a medical officer for aetiological investigations, some babies could be between 6 and 12 months of age, which will require

the difficult task of collating detailed information about the pregnancy and perinatal events. This relies on accurate and descriptive medical notes, e.g. of the dose and blood levels of ototoxic medication, closely recorded degrees of hypoxia, acidosis and the detailed breakdown and serial blood levels of bilirubin, to maximise the chance of an accurate aetiological diagnosis.

EVIDENCE FOR PERINATAL FACTORS CAUSING HEARING LOSS

There is little doubt that adverse perinatal events can lead to neurological sequelae including deafness, and the evidence supports increased vulnerability of the pre-term baby. Studies have quoted rates of 6.1% to 14.9% for perinatal factors to be the cause of hearing loss.[1,2] These perinatal events may range from low birth weight and extreme prematurity to hypoxia and hyperbilirubinaemia.[3] However, there is some difficulty in confidently determining the audiological effect of adverse factors in the neonatal period. This is mainly because of the complexity of possible causative factors. Those infants who have been subjected to one factor may have, in fact, had several other aetiological factors: e.g. a child who is pre-term and has had a difficult birth may have had hypoxia, an increased likelihood of having hyperbilirubinaemia, and sepsis that required ototoxic antibiotics. In a review of 56 publications relating to adverse perinatal conditions and hearing loss in Western countries, Newton[4] found 6–14% sensorineural hearing loss that could be attributed to problems related to birth (see Table 9.1).

Furthermore, it is difficult to compare publications because the accuracy of audiological assessment in the newborn was likely to have been incomplete and less reliable in the past; it is only over the past few years that more accurate techniques have been available for frequency-specific hearing threshold determination in this group of children. Marlow et al.[9] studied 15 children born at less than 33 weeks gestation and a matched control group of 30 children and found that children with sensorineural hearing loss had 'longer periods of intubation, ventilation, oxygen treatment and acidosis, and more frequent treatment with dopamine or aminoglycosides', which showed the complexity of the aetiological factors. Review of data from a targeted newborn hearing screening programme in two London districts from 1995 to 2003 where there was a three to four times higher incidence of hearing loss compared with

Table 9.1 Proportion of sensorineural hearing impairment attributable to perinatal causes.

Author	Year	Hearing loss criteria (dB HL, better ear)	Frequency range (kHz)	Type of study	% perinatal causes
Parving[5]	1984	>35	0.5–4	Population	14
Newton[4]	1985	>25	0.5–4	Population	13.5
Parving[6]	1988	>35	0.5–4	Population	10.1
Das[7]	1991	>25	0.5–4	Population	12.7
Van Rijn and Cremers[2]	1991	>35	0.25–8	School-based	14.9
Fortnum and Davis[1]	1997	>35	0.5–4	Population	6.1
Sutton and Rowe[8]	1997	>25	0.5–4	Population	6.8

Adapted with author's permission from Newton VE, Adverse perinatal factors asssociated with hearing impairment. In: Newton VE (editor) Paediatric Audiological Medicine. London. Whurr; 2002.[4]

Table 9.2 Number of babies diagnosed with permanent hearing loss in a cohort of 1,737 babies referred through a targeted newborn hearing screening programme per risk factor (Sirimanna, unpublished data).

Main risk factor	Number of babies	Number with mild to moderate hearing loss	Number with severe to profound hearing loss
Low birth weight	485	30	6
Toxic aminoglycoside levels	165	10	1
Hyperbilirubinaemia	155	12	2
Hypoxia	39	2	1

the national average showed that out of a total of 1,737 referrals from two maternity units, there were 485 referrals with birth weights less than 1,250 g, 165 with exposure to high levels of aminoglycosides, 155 with hyperbilirubinaemia where exchange transfusions were given or considered and 39 babies who had hypoxia with Apgar of 3 or less at 5 min. Although these were the main factors, a significant proportion of babies had more than one risk factor. Table 9.2 shows the number of children with bilateral permanent hearing loss in this cohort. For the risk factors ototoxicity and hyperbilirubinaemia, there was no significant difference in the peak and trough levels of gentamicin and maximum bilirubin levels among children with normal hearing and children with hearing loss.[10] This may suggest an individual susceptibility to hearing loss in some babies or an unrecognised factor which plays a part in the causation of hearing loss.

Olusanya and Okolo[11] carried out a multivariate logistic regression analysis of perinatal factors associated with hearing loss and showed that birth asphyxia, difficult delivery, neonatal jaundice and seizures, consanguinity and a family history of hearing loss lead to a higher risk of deafness in the baby. A significant percentage of extreme pre-term babies survive with the advancement of neonatal management and technologies, but a considerable proportion of these end with neurodevelopmental impairment, including hearing loss.[12]

HYPOXIA

Hypoxia in the perinatal period can result from birth-related factors or post-natal causes such as apnoeic episodes, respiratory distress syndrome, meconium aspiration and pneumonia or cardiac events. Temporal bone studies have shown changes in the cochlea as a result of hypoxia. In one such study, Koyama et al.[13] examined temporal bones of four asphyxiated babies with a gestational age of 24–36 weeks and normal foetal growth, who died between 1 and 13 days of age. They found degeneration and loss of outer hair cells and oedematous changes in striae vascularis in the baby who had severe asphyxia. However, the evidence for hypoxic cochlear damage in the newborn is not straightforward and is complicated by the fact that often these babies have other factors that may cause a hearing loss. A number of studies using either transient evoked or distortion product otoacoustic emissions, or auditory evoked responses have recently shown reliable evidence of cochlear and central auditory pathway abnormalities in babies who have had significant hypoxia. Jiang and colleagues[14,15] examined the effect of hypoxia on outer hair-cell function by using distortion product otoacoustic emissions (DPOAEs) in 46 term infants who suffered from hypoxia. They found that compared with a control group of term non-hypoxic babies, the hypoxic group had poor DPOAEs in the

1–5 kHz range ($p < 0.01$) 3–5 days after birth with the impairment remaining at 1 month of age. In another study of 339 children, Das[16] found that 12.8% (43 children) had hearing loss due to perinatal factors. Of these, 35 had a hearing loss of 80 dB HL or more. As shown by Anand, Gupta and Raj in a study of 24 children,[17] there is also evidence suggesting that babies with grade II hypoxic ischaemic encephalopathy (HIE) are more likely to have a hearing loss,

Borg[18] reviewed 53 published papers that not only favoured the foregoing discussion but also said that pre-term babies are more vulnerable than term babies to hypoxic damage and found that the length of artificial ventilation correlated well with hypoxic hearing loss. He also found that the presence of severe HIE also increased the risk of having a hearing loss. In a large study, Karjalainen et al.[19] examined 10,000 babies of which 20 had hypoxia during the intrauterine period due to placental insufficiency. None of these 20 children had a sensorineural hearing loss. However, this study focused on chronic milder hypoxia rather than sudden severe hypoxia, and may not be a true representation of hypoxic injury seen perinatally. In an interesting study, Sawada et al.[20] showed that the effect of chronic mild hypoxia is different from an acute anoxic episode. In the former, the picture was similar to that seen in auditory neuropathy whilst the latter tended to produce outer hair-cell damage leading to a typical cochlear hearing loss. In another study of 17 surviving neonates following severe hypoxemia, Cheung, Robertson and Finer[21] found that hyperlactaemia correlated with neurodevelopmental sequelae including sensorineural deafness. Recovery of hearing following severe hypoxia has also been reported by Jiang et al.,[22] studying 51 term newborn infants who suffered perinatal hypoxia, using maximum length sequence evoked potentials. They found that the hearing loss noted in these babies progressed until the third day and started to improve from then on, continuing for up to a month.

NEONATAL INTENSIVE CARE UNIT (NICU) AND PROLONGED NEONATAL VENTILATION

Prolonged ventilation of the newborn has been shown to be associated with sensorineural hearing loss (SNHL). Fortnum and Davis.[1] studied a cohort of children with bilateral sensorineural hearing loss of >40 dB HL, born between 1985 and 1993 in the Trent Health Region in the UK, and found that the prevalence of SNHL was six times higher in those babies who spent more than 48 hours in the NICU. In a similar study by Hille et al.[23] of 2,186 NICU babies, the author reported that ventilation for 5 or more days was a risk factor for developing a sensorineural hearing loss.

INCUBATOR NOISE AND HEARING LOSS

Noise is a well-known cause of hearing loss, and there is an abundance of literature supporting this from both animal and human studies. There is also evidence from adult studies on individual susceptibility,[24,25] and a genetic predisposition,[26,27] to noise exposure, and a synergistic effect from other factors such as ototoxic drugs.[28] It also appears that the cochlear sensory cells in the newborn are more susceptible to noise especially at a lower intensity level. Douek et al.[29] showed that when regular noise levels found in neonatal incubators were continuously applied to guinea pigs in their second week of life, the noise caused histological damage to a

portion of sensory cells in the cochlea. The same noise level applied to adult guinea pigs did not cause cochlear damage. Noise levels from incubator motors have improved over the years, but sudden peaks of sound generated from other forms of activities within the NICU have remained unchanged.[30] Chen and Chang[31] measured the peak noise distribution in the NICU in Southern Taiwan and found a distribution of less than 59 dBA to more than 80 dBA at times. A similar study by Kent et al.[32] in a Canadian NICU found noise levels within the incubator averaging 61 dB, significantly higher than the noise level outside in the room (55 dB). They also found peak noise levels in excess of 120 dB. A similar finding was observed by Benini et al.[33] in 1996, with background noise from the incubator motors measuring 74.2–79.9 dB and impulsive noise levels over 80 dB. A more recent study by Surenthiran et al.,[34] who measured the noise generated by ventilatory systems in the post-nasal space using probe microphone measurements, showed that in those who receive continuous positive airway pressure, the noise levels can reach up to 102 dB SPL with high flow rates.

HYDROCEPHALUS AND HEARING

It is important to remember that those infants with hydrocephalus may have abnormally raised or absent auditory brainstem response (ABR) thresholds that are likely to improve after normalisation of the cerebrospinal fluid pressure following shunt insertion[35] and may not represent the actual state of the infant's hearing.[36] This may currently be classified as auditory neuropathy/auditory dys-synchrony (AN/AD) but the ABR morphology and threshold is likely to improve with normalisation of the intracranial pressure in these infants.

HYPERBILIRUBINAEMIA

Bilirubin is a by-product of the breakdown of haemoglobin, released in its unconjugated form, whenever there is destruction of red blood cells that normally takes place in the spleen. Unconjugated bilirubin, insoluble in water, is bound to albumin and is transported to the liver for conjugation with glucuronic acid, thus making it water soluble. Most of it ends up in the small intestine via bile, broken down by colonic bacteria and excreted in the stool whilst a small amount gets reabsorbed and excreted in urine as urobilin and urobilinogen. When there is excessive haemolysis of red blood cells with surplus production of bilirubin, e.g. rhesus or ABO incompatibility, and also reduced conjugation of bilirubin in the liver because of immaturity and lack of conjugatory enzyme activity, there is accumulation of unconjugated bilirubin in the circulation in large amounts.

A conjugated bilirubin molecule is too large to cross the blood brain barrier (BBB), but this is not so with unconjugated bilirubin. Immaturity of the BBB increases with prematurity. This, along with conditions that make it weaker or more permeable such as hypoxia and acidosis, lead to the BBB becoming more permeable to low molecular weight substances. Unconjugated bilirubin has a high affinity and toxicity towards certain parts of the brain especially in those areas such as the basal ganglia, where there is high metabolic activity. Concentration of bilirubin in the brain and the length of exposure are the most important determinants of neurotoxicity in kernicterus. In a study by Oh et al.,[37] the neurological sequelae including hearing impairment correlated well with the peak total bilirubin levels during the first 2 weeks of life of babies with very low birth weight.

Hearing abnormalities are reported to be common in kernicterus (bilirubin in the brain, bilirubin encephalopathy)[38–40] and are usually caused by deposition of unconjugated bilirubin within the auditory nerve and/or the cochlear nucleus. Some authors have questioned this. For example, Oun et al.,[41] in a prospective study, compared a group of neonates with severe hyperbilirubinaemia with an age-matched control group and found no difference in their audiological status using ABR. However, other authors have found convincing evidence of hearing loss following hyperbilirubinaemia.[42] Boo et al.,[43] studying 128 jaundiced term neonates found that 28 (22%) had a hearing loss when tested using ABR. Of the 128 babies, those who were born pre-term and required exchange transfusions were more likely to have had a hearing loss. The hearing loss seen in hyperbilirubinaemia is due to a neural cause[44] and is different from the most common form of sensorineural hearing loss, which is secondary to involvement of the cochlea, and is called AN/AD (for more details refer to Chapter 12). In AN/AD, the cochlear outer hair-cell function is preserved whilst ABR to sounds is abnormal due to dys-synchronous firing of the eighth nerve fibres. In some patients with hyperbilirubinaemia, the hearing loss is due to cochlear damage most probably following secondary hypoxia and these present with typical features of a cochlear hearing loss. A review by Shapiro[45] suggested that AN/AD should be considered as supportive evidence of kernicterus.

There are case reports suggesting improvement of audiological effects of hyperbilirubinaemia with time, i.e. improvement in hearing thresholds and dys-synchrony.[46] Bhandari et al.[47] studied 30 newborn babies with hyperbilirubinaemia and found that those who were treated showed an improvement in their hearing. On the other hand, there are also reports of normal hearing immediately after hyperbilirubinaemia, progressing to a sensorineural hearing loss in a few months.[48]

INTRAUTERINE GROWTH RETARDATION (IUGR)

IUGR from placental insufficiency has been reported to be associated with sensorineural hearing loss[49] although the number of studies is sparse. It is possible that there is a common aetiology in these babies with the same causative factor leading to both the hearing loss and IUGR. These factors may include chronic hypoxia, placental insufficiency and infective causes such as cytomegalovirus.

PRE-TERM AND LOW BIRTH WEIGHT, AND HEARING LOSS

Using ABR, Chen et al.[50] showed in 194 high-risk babies who had either HIE, hyperbilirubinaemia or a low birth weight that low birth weight was associated with a hearing loss more than hyperbilirubinaemia. The group that had HIE had the least incidence of hearing loss. Pre-term babies are also more prone to having neurological sequelae as the developing brain is more susceptible to injury.[51] Grade I or II intracranial haemorrhages are common in this group of children with periventricular leucomalacia[52] and may lead to developing central deafness. Another study from Israel by Ari-Even Roth et al.,[53] examining 346 infants born between 1998 and 2000 with very low birth weight and using transient evoked otoacoustic emissions (TEOAEs), found only one case of bilateral sensorineural hearing loss. However, as they used only TEOAEs, it is possible that cases of AN/AD were missed. Pre-term babies can have both peripheral and central hearing loss, and this was illustrated in a study of 70 pre-term babies

with a birth weight of less than 1,500 grams.[54] In this cohort of babies, 14% had peripheral hearing loss, 17% had central impairment and 4% had both. In a review of 83 publications, Lorenz[55] found an overall prevalence of 20–25% with at least one major disability in the surviving 22–26-week pre-term babies with deafness in 3–5% of those babies. There is some evidence that the development of the central auditory pathways are delayed in babies born prematurely, as shown by Jiang et al.,[56] who studied 30 babies born at 30–32 weeks gestation with maximum length sequence evoked response audiometry and compared this group with 38 normal controls born at 38–42 weeks of gestation. They found an increase in Wave V latencies and I–V interpeak interval in the test group. They also found that this was more so in pre-term babies who had perinatal complications.[57]

OTOTOXICITY

The ototoxic effects of drugs have been noted for many years, from the days of quinine (for the treatment of malaria) and streptomycin (since its discovery in 1944 for the treatment of tuberculosis). It is now known that there are a significant number of drugs that can be toxic to the inner ear, and these include aminoglycosides, loop diuretics, platinum drugs (e.g. cisplatin), salicylates and macrolides (e.g. erythromycin). These not only can cause hearing loss but also may produce vestibular toxicity leading to a delay in physical milestones in the newborn. Normally, the basal turn of the cochlea is affected first, and the hearing loss may progress even after the withdrawal of the causative agent. There is also increased susceptibility of individuals to ototoxicity. Certain potentially ototoxic drugs given to the mother can cross the placental barrier to appear in sufficient concentrations in the foetal circulation and may cause cochlear damage because of vulnerability of pre-term cochlea to adverse substances. Aminoglycosides are widely used to treat neonatal sepsis including meningitis and are effective for gram-negative bacteria. The ototoxic properties of aminoglycosides have been known for a considerable period of time. Histopathological studies have shown that damage to outer hair cells occurs early in aminoglycoside ototoxicity.[58] Animal studies have clearly demonstrated the effect of aminoglycosides on the inner ear and hearing. Using guinea pigs, Kalkandelen et al.[59] demonstrated that all aminoglycosides were ototoxic when given systemically and locally through the eardrum. However, the dose used was 10–20 times higher than that is recommended for use in humans, and this may have influenced the findings. They found gentamicin to be the most ototoxic, followed by amikacin, streptomycin and netilmicin. In another study using guinea pigs, Halsey et al.[60] explored the possibility of predicting aminoglycoside ototoxicity with efferent mediated adaptation of distortion product otoacoustic emissions and found that this was a good predictor of gentamicin ototoxicity. Some studies have suggested that aminoglycosides are not ototoxic. One of these is a review of seven published prospective, controlled studies totalling 1321 newborn infants, by McCracken[61] that showed little evidence of ototoxicity to aminoglycoside drugs except in one study where there were flaws with regard to the number of patients studied and the duration of follow up. In another study by the same group of researchers where netilmicin or amikacin was used to treat sepsis in a group of babies in an intensive care unit, there was no significant difference in the incidence of hearing loss on longitudinal follow-up.[62]

On the other hand, Matz[63] participated in two randomised prospective studies using aminoglycosides where the investigator was blinded. In the first 108 patients, 54 received gentamicin and the others were given amikacin. In the second study of 163 patients, 61 received genta-

micin, 52 tobramycin and 50 netilmicin. The toxic effects noted in this study included hearing loss as well as vestibular toxicity. Gentamicin was more ototoxic (11% in the first study and 18% in the second) followed by amikacin (12.9%). Ototoxicity with tobramicin was 11.5% whilst netilmicin produced only 2% ototoxicity. Black et al.[64] studied a group of patients 1 year after treatment with gentamicin. Thirty-three subjects had vestibular dysfunction on tests with all having a feeling of imbalance. In this group of patients there was no correlation between vestibular toxicity and the dose of gentamicin or the serum gentamicin levels. There is also a suggestion that certain conditions, such as cystic fibrosis, can offer some protection against aminoglycoside ototoxicity, as the incidence of ototoxicity in this group is considerably lower than expected in spite of repeated courses of aminoglycosides.[65]

In a study from Bristol, UK,[66] 24 children receiving a gentamicin once-daily regimen were serially monitored using TEOAEs and pure-tone audiometry (PTA) (or when this was not possible, ABRs). Eleven children had gentamicin for up to 7 days and the others (13) were treated for 8–29 days. No significant change in hearing levels was noted in the 7-day group, but the group that received more than 7 days of gentamicin showed significant reduction in the TEOAEs mean response level with prolonged treatment. No change in PTA or ABR was noted in the same group. They suggested that TEOAEs could be used for early detection of ototoxicity although this study did not use a control population, and there was no audiometric confirmation of a measurable hearing loss in those with reduced TEOAEs. Ertl et al.[67] published a case-controlled study where 164 premature infants treated in an NICU were screened using TEOAEs. Thirty-two infants were referred for further hearing assessments using ABR, and of these, 22 were found to have a bilateral hearing loss. This group had significantly poor Apgar scores, lower pH and pO_2 values, and had had higher aminoglycoside doses and hyponatraemia. They concluded that aminoglycoside treatment and hyponatraemia were the most significant factors for developing hearing loss on multivariate regressive analysis.

Research over the past 10 years or so has shown that reactive oxygen species (oxygen ions, free radicals and peroxidase), which are by-products of cellular metabolism, do play a significant role in ototoxicity and may explain why high frequencies are affected initially compared with low and mid frequencies.[68] In fact, there is evidence from animal studies suggesting that neurotrophins, lectins and antioxidants can be used in the prevention of ototoxicity.[69–71]

There is considerable evidence for abnormal susceptibility to aminoglycoside antibiotics in some individuals (see also Chapter 6) and most of these have a mitochondrial mutation (1555A>G).[72] In addition, 4309G>A, another mitochondrial mutation, has also been associated with increased susceptibility to aminoglycoside ototoxicity.[73] Tono et al.[74] suggested that the striae vascularis in these patients can be primarily affected. The main defect appears to be related to mitochondrial protein synthesis required for cellular function and survival.[75] The mutation is maternally inherited, i.e. mother-to-child transmission. Those members of the family who have not been exposed to aminoglycosides also are likely to show evidence of sensorineural hearing loss beginning in their 30s. Therefore, in families with a maternally inherited pattern of hearing loss, it is important to check for these mutations before commencing aminoglycosides.

Vancomycin, a glycopeptide that has been used for over 50 years as an anti-staphylococcal antibiotic, has also been reported to cause hearing loss occasionally but the publications are scant and the evidence is not strong. Bailie and Neal[76] reviewed 28 publications on vancomycin ototoxicity and concluded that the hearing loss normally involved the high frequencies; they could not determine whether it was a permanent or a temporary loss. Brummett and Fox[77] also carried out a literature review and concluded that there is occasional permanent sensorineural

hearing loss that improved in some cases after stopping vancomycin. Tange et al.[78] carried out an animal experiment using Mongolian gerbils that were treated with vancomycin 80 mg/kg per day for a 2-week period and found no significant difference between ABR thresholds and no cochlear damage on electron microscopy. Although the publications on glycopeptides ototoxicity cast a doubt about its existence, because of the uncertainty it is sensible to be cautious.

Loop diuretics are also reported to be responsible for hearing loss and most evidence comes from animal studies. In one such study, Rybak et al.[79] found that four loop diuretics – furosemide, piretanide, bumetanide and ethacrynic acid – caused significant reduction of endocochlear potentials in adult chinchillas. However, in a different study of 57 neonates who received furosemide compared with 207 neonates who did not, Rais-Bahrami et al.[80] found no significant difference of hearing loss between the two groups. At least some of those who received diuretics had a degree of renal failure, which itself may have increased the chance of ototoxicity due to elevated serum levels of these antibiotics from reduced clearance. Close monitoring of blood levels, especially in those with renal failure, must be undertaken, and adjusting the dosage of any ototoxic antibiotics accordingly is extremely important in order to avoid ototoxicity.

SYNERGISTIC EFFECTS OF PERINATAL FACTORS

There is evidence to support the fact that perinatal factors, which can potentially cause hearing loss, have a synergistic effect when they act simultaneously. Tan et al.,[81] using a guinea pig model and distortion product otoacoustic emissions, showed that ototoxicity can occur with subdamaging doses of amikacin when the animal is exposed to noise compared with those who did not receive amikacin and the same level of noise exposure.

CONCLUSION

A large number of adverse perinatal conditions, acting individually or more often collectively, can lead to temporary or permanent hearing loss that is sensory or neural in nature, with varying degree of severity. High frequencies appear to be affected more often, especially in ototoxicity and hypoxia, but other frequencies can be involved in some patients. There appears to be a susceptibility to hearing loss, which is most probably genetically determined. Often more than one perinatal factor is seen in babies who have a hearing loss, and it is likely that these factors have a contributory or synergistic effect. By identifying early those babies who are susceptible to ototoxicity, hearing loss may be prevented by selecting alternative antibiotics. A detailed history of all perinatal events and the pregnancy is extremely useful in making an accurate diagnosis of the cause of the hearing loss. Better management of the birth and avoidance of those factors that may lead to a hearing loss, such as hypoxia, hyperbilirubinaemia and ototoxicity, certainly will minimise the incidence of perinatally acquired hearing loss.

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10 Acute otitis media and otitis media with effusion

E. Raglan

CLASSIFICATION AND DEFINITIONS

A review of the literature reveals an entire spectrum of inflammatory conditions of the middle ear, frequently with different terminology used to describe similar conditions, which can create confusion for researchers and clinicians alike. Bearing this in mind, a committee of experts reached a consensus on the terminology used relating to inflammatory conditions affecting the middle ear.[1] They have classified terms concerning acute otitis media (AOM), otitis media with effusion (OME), as well as eustachian tube dysfunction and its complications.

Otitis media (OM) is inflammation of the middle-ear cleft. AOM has a rapid onset; the signs and symptoms of inflammation can be ear pain (otalgia), fever, a bulging tympanic membrane which may appear opaque or more frequently erythematous, in addition to ear discharge (otorrhoea) through the formed perforation (Figure 10.1). Frequently, it is associated with upper respiratory tract infections, presenting with additional symptoms of cough, nasal congestion and nasal discharge. The clinician frequently has difficulties in differentiating OME from AOM; OME may precede or follow a bout of AOM, and this confusion may lead to the inappropriate use of antibiotics as this is not the recommended treatment for OME.

OME is a condition in which fluid accumulates in the middle-ear cleft without the signs and symptoms of inflammation, whereas AOM, with acute presentation of symptoms and signs of inflammation, is accompanied by middle-ear effusion (MEE). The fluid may be serous or mucoid and it may contain bacteria.[2,3] The condition may follow a bout of upper respiratory tract infection or indeed an ear infection such as that seen in AOM. It is a fluctuating condition that may occur in a child several times throughout childhood or may persist over longer periods of time, especially in children with cleft palate or Down syndrome. In most cases, it disappears with age without any long-term sequelae. In some children, it may lead to various complications requiring more active management.

Eustachian tube dysfunction has similar symptoms to OM, such as hearing impairment and ear pain (without signs of MEE) with tympanographic evidence of negative pressure in the middle-ear cleft.

EPIDEMIOLOGY

OM is one of the most common conditions in childhood; incidence decreases with age. Half of the children studied had OM onset within the first 6 months of life, with peak occurrence in the first 18 months.[4] Prevalence figures show the decline of OM to 3–4% by 10 or 11 years

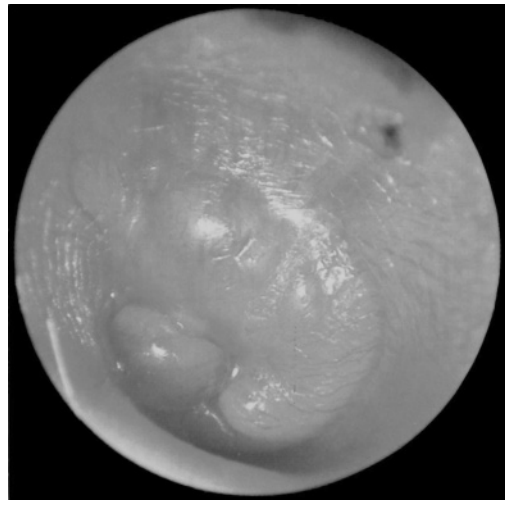

Figure 10.1 Acute otitis media. The tympanic membrane is bulging laterally under the pressure of the infected purulent debris in the middle ear (Left ear) (courtesy of Hawke M., Bingham B., Stammberger H., Benjamin B. In: Diagnostic Handbook of Otolaryngology. London: Martin Dunitz, 1997, p75).

of age.[4] OME is more prevalent than AOM, as MEE tends to persist after AOM episodes. OME is especially common in children, with its peak around the ages of 2 and 5. Its prevalence at age 2 is about 20% and about 15% at 5 years of age.[5] With increasing age, it becomes less frequent and of a shorter duration.[6] It is generally difficult to assess the duration of effusion as the episodes may be asymptomatic. However, there is a tendency for spontaneous recovery.[7] The resolution of the effusion in the majority (60–94% of children) occurs within 3 months, although in about 10% it may persist for more than 1 year.

RISK FACTORS FOR OM

A variety of risk factors have been associated with recurrent OM and OME; they can be subdivided into host-related and environmental factors. These are: eustachian tube dysfunction/occlusion, nasopharyngeal infection, allergy, defective or impaired immunological status, craniofacial abnormalities, genetic predisposition, passive smoking, bottlefeeding instead of breastfeeding and attendance at a day nursery.

Eustachian tube dysfunction/occlusion

The eustachian tube is positioned between the nasopharynx and the middle-ear cavity and the entire area is covered by a mucous membrane. In infants, the tube is more horizontal and shorter than in adults[8] and allows for pressure equalisation in the middle-ear cleft (with atmospheric pressure).

Eustachian tube dysfunction may lead to impairment of the middle-ear pressure regulation thus leading to the development of AOM or MEE. For example, the tube can be mechanically

obstructed by the inflammatory exudates secondary to infection or allergy,[9,10] or a cartilaginous portion of the tube can be compressed by an adenoidal mass.[11,12] The eustachian tube may fail to open during swallowing activity, as was described in infants with unrepaired cleft palates[13] who had chronic OME with effusion and in children with middle-ear disease.[14–16] This could be due to collapse of the tubal cartilage. In infants, there is less cartilage and it is less dense than in adults.[17,18]

Infection originating in the nasopharynx leads to the development of changes in the mucosa lining the middle-ear cleft and the Eustachian tube, and results in the production of mucoid or serous fluid. As a result, negative pressure increases within the middle-ear cleft. Blockage of the Eustachian tube leads to absorption of air from the middle-ear cavity into the blood vessels of the mucosa, leading to a reduction in middle-ear pressure and thus to a retraction and restriction of movement of the tympanic membrane.

Allergy and immunology

The role of allergies in OM has been a subject of ongoing discussion. OM frequently occurs in children suffering from allergic rhinitis; its prevalence is 16.3% higher in those suffering from allergies as opposed to 5.5% in normal controls.[19,20] A common cause of allergic rhinitis is an immunoglobulin E (IgE)-mediated allergic reaction, but it is only one of many aetiological factors for OM.[21] Both bacterial and viral infections may be influenced by the allergic response. The biochemical mediators released during the nasal allergic reaction produce oedema and inflammation of Eustachian tube, leading to the formation of OM.[22] The same cellular and cytokine profiles were found in the middle-ear mucosa and the nasopharynx, supporting the hypothesis that the middle ear may be an integral part of the united airway concept,[23] where the impaired function of the upper airways due to allergic rhinitis that causes nasal obstruction plays an important role in the development of lower airways symptoms.[24] In some patients, atopy may be responsible for the recurrence and maintenance of middle-ear disease. There are a number of hypotheses explaining the mechanism by which allergies may contribute to OM. The possible mechanisms to be considered are: middle-ear mucosa functioning as the target,[25] allergies leading to inflammatory swelling of the eustachian tube mucosa, inflammatory nose obstruction and aspiration of bacteria-laden allergic nasopharyngeal secretions into the middle-ear cavity.[26] The other hypotheses are based on a possible increase in circulating inflammatory mediators from local allergic reactions in the nasal mucosa, which may alter middle-ear mucosa permeability and lead to altered gas exchange.[27] The immunological status of a child may play an important role in the predisposition to recurrent infections.

Children with a history of recurrent AOM (at least four or more episodes within the first year of life) were found to have subtle immunological abnormalities predisposing them to such infections.[28] A recurrence of bilateral OME after grommet insertion was found to be more likely in children with a combination of low IgA and IgG2 levels.[29] It has been postulated that immature or defective immunological responses in children with recurrent OME might have contributed to the pathogenesis of the condition.[30]

Craniofacial abnormalities

OM is very common in infants with an unrepaired cleft palate,[31] which can contribute to poor eustachian tube function and thus failure of its opening mechanism.[32] The condition has a

prolonged recovery and high incidence of late sequalae.[33] Its occurrence is reduced in children after palate repair.[31] OM is also common in children with various craniofacial abnormalities and Down syndrome.[34]

Genetic susceptibility

The observation that some children develop recurrent episodes of OM and chronic MEE suggests increased susceptibility of those children to those conditions, with a similar pattern of recurring episodes reported in their families. This has been confirmed by recent studies on twins and triplets.[35–37] Work on the mouse model has allowed it to become a tool with which the genetic basis for human OM can be unravelled.[38] The development of the animal model (in mice) allowed for creation of an absence of one of the genetic factors resulting in the abnormal structure of the Eustachian tube, and thus allowing OM to occur. This paves the way for a greater understanding of OME and points us further towards improvement of patient management.[39]

Social and environmental factors

Children who attend daycare and have many siblings in their family tend to have a higher rate of the particular upper respiratory tract infections that cause eustachian tube dysfunction, thereby increasing the likelihood of developing OM. According to some authors,[40] children who are passively exposed to smoking also have a significantly higher risk of recurrent OM.

Breastfeeding has a protective function against OM, and while it is not clear whether the length of time a mother nurses her child influences the incidence of OM, there is some evidence to suggest that the longer the mother is able to breastfeed, the fewer the number of episodes of OM.[41]

DIAGNOSIS OF OME

OME in a child is most often clinically suspected by the child's parents, their caregivers or teachers. Changes that are often seen by those involved in the child's care and which lead to the child's presentation to the doctor include: the child becomes unresponsive to everyday sounds, often asks others to repeat their words, may be seen to be unresponsive when in noisy surroundings, may listen to the television or audio equipment at very loud levels or may appear to be withdrawn, inattentive and eventually may become disruptive, with a reported lack of concentration and poor educational progress. The infant may also be observed not to turn when trying to locate the source of a sound. Eventually, the child may have spelling difficulties or the child's speech may become indistinctive. There may be some difficulties with balance, or the child may appear clumsy or may complain about having tinnitus. There will typically be a history of repeated ear infections or respiratory tract infections. Such observed symptoms should prompt suspicion of possible hearing impairment which would require confirmation through appropriate assessment and, if diagnosed, follow-up with the appropriate management.

Confirmation of diagnosis

The diagnosis of OME is made on the basis of clinical history, clinical examination and appropriate tests of hearing and tests of the status of the middle ear. The suspicion of hearing impairment caused by OME is confirmed by a detailed medical, otological, developmental and behavioural history. Clinical history should reveal details of the child's respiratory and otological health, the degree of severity of hearing impairment as well as the child's general medical health and the effects of the impairment on speech, language or behavioural development. Clinical examination of the nose and throat, as well as observation of the child's behaviour, should contribute to establishment of the factors predisposing the child to glue ear and its potential effects. The examination of the ear, inclusive of otoscopy and tuning forks where appropriate, should further confirm not only the diagnosis of the condition but also the presence of conductive hearing loss. Some clinicians tend to use pneumatic otoscopy when available, which allows for assessment of the colour, appearance and mobility of the tympanic membrane. Otoscopy may show the typical appearance of the tympanic membrane from the presence of an air-fluid level (Fig. 10.2) or bubbles behind the translucent tympanic membrane which create a bulging, dull appearance with the absence of the light reflex (Fig. 10.3). The mobility of the tympanic membrane is restricted by the presence of fluid in the middle ear, thus causing conductive hearing loss.

The tuning fork tests are able to confirm conductive hearing loss. Consequently, developmentally appropriate behavioural tests of hearing together with tympanometry should confirm the presence of hearing impairment of a conductive type (Fig. 10.4).

A Type B tympanogram should indicate the presence of MEE. Type C, showing the presence of negative pressure within the middle-ear cleft, may be found in the early stages of OME

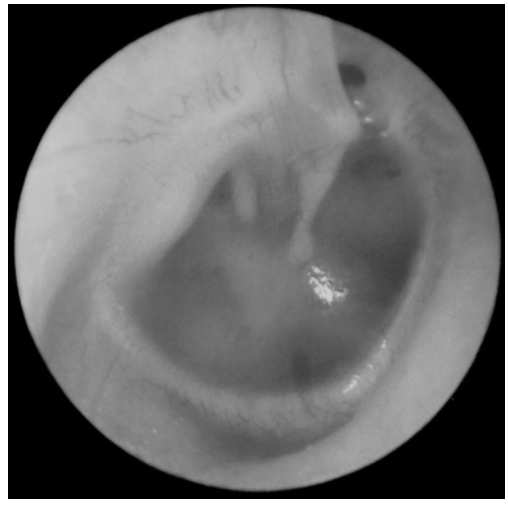

Figure 10.2 Otitis media with effusion. OME accumulation of a clear straw-coloured thin uninfected watery serous fluid within the middle ear. The tympanic membrane shows a yellowish discolouration from the fluid within (Right ear) (courtesy of Hawke M., Bingham B., Stammberger H., Benjamin B. In: Diagnostic Handbook of Otolaryngology, London: Martin Dunitz, 1997, p79).

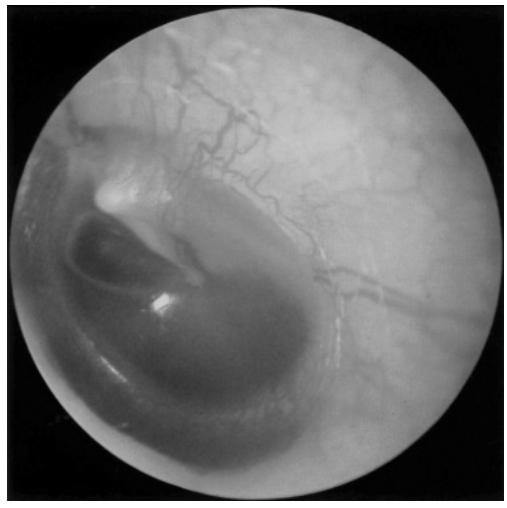

Figure 10.3 Otitis media with effusion. The mucoid fluid in the middle ear has an orange tinge. Note the tiny air bubble anterior to the malleus handle (Left ear) (courtesy of Hawke M., Bingham B., Stammberger H., Benjamin B. In: Diagnostic Handbook of Otolaryngology, London: Martin Dunitz, 1997, p78).

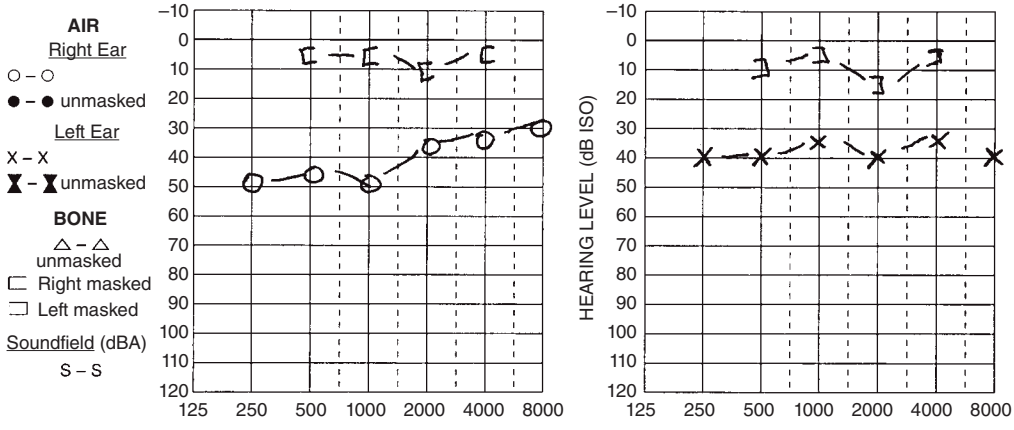

Figure 10.4 PTA showing a bilateral conductive hearing loss.

or in eustachian tube dysfunction (Figure 10.5). Occasionally, if the behavioural tests are not conclusive, the electrophysiological tests will provide objective evidence about the level and type of hearing impairment in conjunction with tympanometry.

OME-causing conductive hearing impairment may coexist with sensorineural hearing loss and needs to be diagnosed and managed separately.

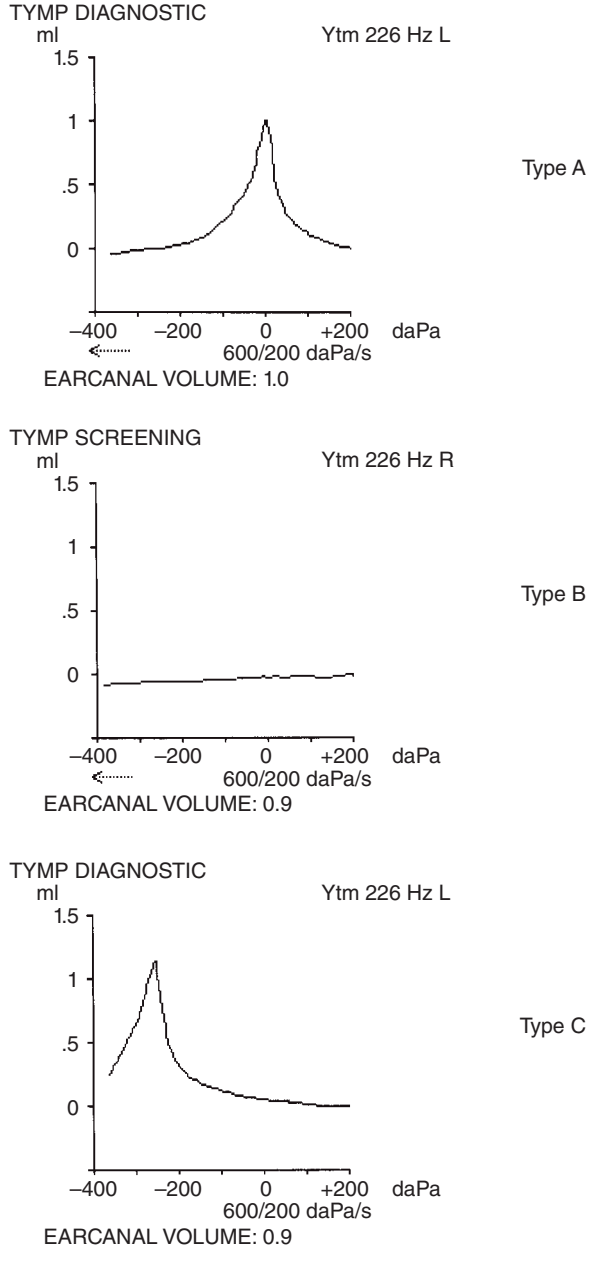

Figure 10.5 Classification of tympanogram shapes: tympanogram types A, B and C.

EFFECTS OF OME

The effect on speech, language and behaviour

A collection of fluid in the middle ear will impair sound transmission, leading to conductive hearing impairment. This may have a fluctuating nature or, in the case of recurrent OME, the hearing impairment may become more pronounced. It may involve not only low frequencies such as seen in early presentation, but will also involve higher frequencies leading to a picture of a flat configuration across a spectrum of frequencies. The resulting hearing loss can be expected to have an impact on speech and language development with secondary communication and behavioural difficulties. The complexity of this relationship depends upon the child's degree and duration of hearing impairment, the child's stage of development, as well as the level of existing vocabulary skills, cognitive and linguistic perceptual abilities and the degree of support in the child's home environment. Conductive hearing impairment in the early stage of a child's development (i.e. within the first or second year of life) will alter the acquisition of sounds, semantic, syntactic and pragmatic rules of language.[42] The distorted acquired sound is stored in the child's language database, and later on, when this situation continues, such children are unable to decipher incomplete auditory messages and are unable to refer to the established language code. The child is unable to use contextual clues or previous experience to decipher the auditory message. In conversations, the child will ask to have sentences repeated and is likely to mishear, and similarly, is likely to ask for the volume of the television to be raised. A child may shout whilst trying to communicate or be observed to lip read. There may be poor pronunciation or difficulties in being understood by those outside the immediate family. The child may be shown to have difficulties in learning, slowness, inattentiveness and poor concentration.

The associated OME conductive hearing impairment may also worsen sound localisation and binaural processing.[43] Both animal and human studies have indicated that the attenuated/delayed auditory input due to prolonged MEE leads to the impairment of binaural hearing temporal resolution sensitivity to short tones in the presence of background noise.[44] However, with age, some aspects of the disordered central auditory processing recover spontaneously once the MEE has resolved,[45] or they may be reversed by active learning.[46] At a younger age, such a child may show symptoms of withdrawal, lack of concentration and disordered behaviour arising from frustration. At school, when background noise is present, the child may present with difficulties listening, leading to poor academic progress and additional behavioural difficulties, such as poor concentration, withdrawal, inattention, further poor educational progress and finally, disruptive behaviour.

Balance problems

Balance difficulties in children with longstanding OME[47] are considered to be attributable to less common complications of this condition, as compared with hearing impairment and speech and language difficulties. They tend to be reported by parents as clumsiness and frequent falls.[48] The most recent study of 24 vertiginous children,[49] compared with age- and sex-matched healthy children, found that the predominant diagnosis amongst benign paroxysmal vertigo of childhood and migraine associated dizziness, was OM-related vertigo. Balance platform studies found an increased sway in those children as compared with non-sufferers,[50] and the rotational chair study showed increased abnormalities in the sufferers.[51,52] However, resolution of MEE

leads to improvement of the balancing function. Bower and Cotton.[53] and Goltz et al.[54] found that symptoms of imbalance in children with chronic OME resolved after evacuation of MEE through the insertion of grommets. The postulated mechanisms for such abnormalities in those children are secondary movements of the inner-ear fluids due to the transfer of negative middle-ear pressure changes through the labyrinthine windows,[55,56] irritation of the vestibular receptors by serous labyrinthitis[54] and a transfer of ions through the round window membrane leading to changes in the chemical composition of the endolymph via the perilymph, which leads to ionic changes in the vestibular hair cells resulting in the observed balance disturbance.[50]

MANAGEMENT OF GLUE EAR

The method of management of glue ear very much depends upon the duration of the effusion, the degree of hearing impairment that the glue ear causes, the effects of this impairment on other aspects of the child's well-being (such as behavioural, developmental, speech effects and balance disturbances), as well as the presence of other medical conditions and syndromes such as Down syndrome, or the presence of a cleft palate (with or without craniofacial abnormalities).

If on initial presentation hearing loss is not greater than 25–30 dB HL, averaged across the main frequencies of 0.5, 1, 2 and 4 kHz, and there are no speech, balance or behavioural abnormalities, then a period of observation of 3 months is recommended during which the glue ear may resolve spontaneously or it may become worse. During the period of active monitoring, educational advice should be given: the child should have preferential seating in the classroom, parents/caregivers should make sure that they speak clearly to the child at a close proximity, without raising their voices to be heard. If there are sequelae of hearing impairment, as mentioned earlier, or if the hearing is worse than the symptoms indicate and are persistent, then surgical management with the insertion of grommets should be offered. If surgery is contraindicated, hearing aids should be considered. In the latter case, treatment via application of hearing aids or myringotomy with the insertion of grommets should be proposed. In addition, adenoidectomy should be recommended if there are symptoms and signs of recurrent upper respiratory tract infections and adenoidal symptoms and signs.

Management of AOM

Medical management consists of treatment of the pain, with an initial 48–72-hour observation period (with delayed antibiotic treatment in some cases), followed by antibiotic treatment. Pain relief should be obtained through use of analgesics such as ibuprofen or other systematic or topical agents as soon as this is possible – hopefully within the first 24 hours of an infection. Treatment with antibiotics (type, dose, time of application) will vary according to the child's age and severity of illness.

In its recent guidelines, the American Academy of Paediatrics and Family Practitioners recommends that infants below the age of 6 months, whether diagnosis of AOM is certain (rapid onset, presence of MEE, signs and symptoms of middle-ear inflammation) or uncertain, should be offered antibacterial therapy because of the danger of serious infection and its sequelae.[57] However, those children between 6 months and over 2 years of age should only be offered antibacterial therapy when the diagnosis is certain. If there is any uncertainty, then therapy should only be offered in cases of severe illness (defined as a temperature higher than

39˚C and accompanied by severe pain). All other children should be observed for at least 48–72 hours before antibacterial treatment is given. Until then, they should be treated only symptomatically.

The recommended first line antibiotic is amoxycillin, which is sensitive to the most common pathogens causing middle-ear infections such as *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Moraxella catarrhalis*.[58] The recommended dose is 80–90 mg/kg/day.

For patients with more severe illness, a 10-day course[59] of a combination of amoxycillin with clavulanate is recommended (90 mg/kg/day of amoxicillin and 6.4 mg/kg/day of clavulanate in multiple doses).[60] This is for children below the age of 2 who are suffering from a more severe form of the illness. For children 6 years and older with milder disease, a 5–7-day course of antibiotics is recommended. If, after 48–72 hours, there is no improvement when taking the initial agent or a deterioration of symptoms is noted when under observation, then an antibiotic or combination of antibiotics should be prescribed. Alternative antibiotics might be tried, especially in patients with a history of allergy to penicillin, such as cefdinir, cefpodoxime, cefuroxime, azithromycin, clarithromycin, erythromycin or ceftriaxone.[61]

PREVENTION OF AOM

The prevention of AOM and its recurrent attacks may be achieved by providing the patient, his parents and/or caregivers with information and advice, as well as by the use of immunoprophylaxis. The identification of the susceptibility genes may provide for better understanding of the pathogenesis of the condition, hence ensuring better methods of prevention and treatment.[62] It has been postulated that variation in innate immunoresponse genes may result in an alteration of the production of cytokines, leading to altered inflammatory responses, which ultimately contributes to recurrences of acute OM in otitis-prone children.[63]

Preventative environmental factors such as the exclusion of passive smoking, the implementation of breastfeeding within the first 6 months of life (along with avoidance of supine bottlefeeding) as well as the predisposing environmental factor of a child's attendance at day nursery all need to be discussed and shown to be possible factors relating to the recurrence of AOM. In cases of recurrent AOM, investigations should centre on exclusion of such possible underlying conditions as allergy or immunological defects.

The introduction of the pneumococcal conjugate vaccines brought about a decrease in frequent childhood pneumococcal infections, but it has been found by some authors[64–67] to be less effective in the prevention of AOM. The 7-valent pneumococcal vaccine has been 90% effective in reducing pneumococcal disease due to vaccine serotypes and has also been effective in reducing carriage. However, as vaccine serotypes are no longer able to compete for colonisation of the nasopharynx of those who are vaccinated, their place has been taken by serotypes which are not targeted by the vaccine.[68] Not all literature supports the reduction in effectiveness of the vaccine.[69] The latter found that children were actually less likely to suffer recurrent episodes of otitis media, or undergo grommet insertion after the introduction of the heptavalent pneumococcal conjugate vaccine. These findings were confirmed in 2001 by Eskola et al.[70] in a randomised double-blind trial of a heptavalent pneumococcal conjugate vaccine in 1,662 infants. Eskola et al. found that vaccination reduced the number of episodes of AOM due to any cause by 6%. Cultures obtained confirmed that pneumococcal episodes leading to AOM decreased by between 34% and 57% due to the serotypes contained in the vaccine. However, there was an increase of 33% of the number of episodes due to all other serotypes. A further

study by Block et al.[71] confirmed that community-wide vaccination with the heptavalent pneumococcal conjugate vaccine significantly altered the microbiology of AOM. In vaccinated children, the proportion of gram-negative bacteria was seen to be twice as frequent as *S. pneumoniae* in AOM. This change in the microbiology of AOM, together with the development of antibiotic-resistant strains following the widespread use of antibiotics for the treatment of AOM, is likely to have an important impact on future management of OM and vaccine implementation.

AOM may be followed by a persistent OME, and this condition, after a period of active monitoring, should be treated appropriately either with grommet insertion alone or with an adjuvant adenoidectomy in cases where adenoidal symptoms are present or with the application of hearing aids if this is appropriate. This is especially true when there is a history of recurrent upper respiratory tract infections.

The new method for treatment of persistent middle-ear infections could be vaccines made with biofilm-specific molecular targets. The biofilms are multicellular networks of bacteria found near the mucosal surface encased in a matrix, found in the middle ear and are resistant to the host immune system. They express different genes and are phenotypically different from their planktonic counterparts. In biofilms, bacteria exist in their own microenvironment, and aerobic and anaerobic bacteria can exist alongside each other.[72] Biofilm bacteria with lower metabolic rates showed increased resistance to antibiotics, which affected their metabolism.[73] There are current studies to demonstrate how biofilms can impede successful antibiotic therapy.[74–76]

In a study of 5-year-old children in daycare, xylitol, a sugar found in birch trees, raspberries and plums, and used as sweeteners in chewing gum and in toothpaste, has been found to have a beneficial effect in preventing episodes of AOM in children who used it, as compared with children using sucrose gum.[77] It inhibits growth of *Streptococcus pneumoniae in vitro* and *Streptococcus mutans*, and is used in toothpaste to prevent dental infection. It is well tolerated by small children[78] and is the subject of further clinical trials.

MEDICAL TREATMENT OF OME

A variety of agents have been used in the treatment of chronic OME such as antibiotics, topical or systemic decongestants, antihistamines, steroids, dietary modifications, immunostimulants, homeopathy, osteopathy, acupuncture, massage, autoinflation and the application of hearing aids. There are no published studies about the usefulness of acupuncture, cranial osteopathy, massage, immunostimulants or dietary modification, nor are there any systematic reviews about the effects of homeopathy or hearing aids; however, there are good systematic reviews available that look at the effectiveness of steroids, antihistamines, decongestants, antibiotics and autoinflation. The effectiveness of intranasal steroids is unknown, but one systematic review has showed that oral steroids alone or in combination with antibiotics led to a quicker resolution (within 2 weeks) of OME, but there is still no evidence of a long-term benefit from treating hearing loss associated with OME.[79] In the short term, researchers reported some adverse effects of oral steroids, such as vomiting, diarrhoea and dermatitis, but there were no longer-lasting effects.

Similarly, well-conducted systematic reviews on the usage of decongestants and antihistamines alone or in combination,[80] as well as the meta-analysis of studies of the use of antibiotics,[81] did not show any statistical evidence of the benefits of these forms of treatment in children

presenting with OME. The limited benefit of autoinflation was shown in one particular systematic review,[82] but children's compliance with this method of treatment was poor. However, it is reasonable to recommend this form of treatment during the period of watchful waiting, pending natural resolution of OME because of its low cost and absence of adverse effects. There are no good studies evaluating the benefit of homeopathy in the management of OME. Results from a pilot trial[83] carried out at two general practice centres in the UK compared the benefit of homeopathy with the standard GP care of watchful waiting, autoinflation, and in some children, a low-dose course of antibiotics for 4–6 weeks. After 1 year of homeopathic treatment, the tympanograms of those children showed improvement, with no observable benefit for the other interventions.

REHABILITATIVE MANAGEMENT OF OME

In the case of persistent OME and hearing loss children should be offered hearing aids as an alternative to surgical management or when surgery is contraindicated. This will allow for an improvement in hearing, and hence reduction in disabilities arising from hearing impairment, such as impairment in speech and language. The results of two surveys[84,85] carried out in the UK showed good acceptance and fairly good compliance with hearing aids by parents and children with improvements in hearing, speech and behaviour. However, there are no good-quality comparable studies which would evaluate the effectiveness of amplification in those children. Management with hearing aids requires careful monitoring of the middle ears to prevent structural damage to the middle ear as a result of OME.

SURGICAL MANAGEMENT OF OME

The decision for surgical management of OME is taken on an individual basis, but there are broad guidelines which should help to make such a decision.[57] The indications for surgery are as follows:

1. bilateral OME persisting more than 3 months;
2. persistent OME leading to secondary complications arising from the effects of long-term effusion. The effusion may lead to functional consequences or structural changes within the middle-ear cleft. These are as follows:
   (a) hearing impairment, leading to delayed development of speech and language;
   (b) delayed development may in turn cause communication and behaviour difficulties;
   (c) clumsiness and balance difficulties;
   (d) tinnitus;
   (e) poor educational progress;
   (f) structural changes within the middle ear, such as ossicular involvement, perforation, adhesive changes and retraction pockets of the tympanic membrane;
3. recurrent episodes of AOM where the medical/antibiotic treatment has failed.[86]

The decision on the timing of intervention is based on evidence arrived from a well-conducted systematic review of the natural history of OME in a number of cohort and random clinical

trial studies.[7] The research indicates that the resolution rate of OME is 56% at 3 months, 72% at 6 months and 81% at 9 months; hence it can be presumed that at least half or more of these children should have a natural resolution of the condition without any need for surgery.

The effectiveness of surgical intervention was also evaluated in several studies where the effects of grommet insertion were compared with myringotomy and active monitoring. The results of these comparisons were presented as a systematic review in which outcome measures such as the presence of a hearing impairment, the duration of MEE and prevention of the development of complications arising from hearing impairment and behaviour were taken into consideration.[87]

The conclusion of the review was that bilateral grommet insertion leads to an improvement of hearing of 4–10 dB within the first 6 months after the procedure, with the effect diminishing over time. There was no effect on expressive language development of grommet insertion as compared with active monitoring. Between 20% and 50% of children who have had grommets inserted suffer further episodes of OME after their extrusion.[88] The grommets are effective in correcting conductive hearing impairment as long as they remain in place and are functioning. However, there is also evidence to suggest that there may be long-term complications of grommet insertion, including increased risk of pathological abnormalities of the tympanic membrane (perforation, focal atrophy, retraction, tympanosclerosis) and raised hearing thresholds 6 to 10 years following the surgery as compared with children who did not have grommet insertion.[89,90]

Adenoidectomy is frequently performed at the time of grommet insertion, especially for recurrent episodes or chronic persistent OME. However, evidence of its benefit is not entirely clear in terms of improvement of hearing levels, especially in children who do not have a history of recurrent upper respiratory tract infections associated with OME. Additionally, the reported trials comparing hearing levels in patients following the insertion of grommets alone versus a combination of grommet insertion and an adenoidectomy have a number of problems. Primarily, they fail to mention the duration of OME, their design suits the efficacy and not the effectiveness of the intervention, the size of the sample population is small and there is a failure to collect data on hearing levels in the long term.[91,92] There is a further need for good-quality randomised controlled trials on a larger population in order to document the difference between these two types of surgical intervention. The decision for an adjuvant adenoidectomy should be made carefully and considered especially in the presence of persistent, recurrent and frequent upper respiratory tract symptoms.

SUMMARY AND CONCLUSION

Inflammatory middle-ear conditions, especially OME, are frequent in childhood. They may lead to fluctuating or persistent conductive hearing impairment with associated sequelae such as a delay of speech and language development, behavioural problems, balance difficulties, clumsiness and educational problems. They may also impact on the structure of the middle ear. AOM is predominantly treated with antibiotics; in the older child, treatment is recommended after a period of 48–72 hours, whereas immediate treatment should be provided to a younger infant. The treatment of OME requires a minimum 3-month period of active monitoring, following which, if the condition persists, consideration should be given as to whether the child will be treated with application of hearing aids or with the insertion of grommets. An adjuvant adenoidectomy is recommended in cases where a history of OME associated with

persistent, frequent upper respiratory tract infections is present. There is no good evidence that other forms of non-surgical intervention, such as antihistamines, decongestants, steroids, antibiotics, acupuncture, dietary modifications or homeopathy are at all helpful in the treatment of OME.

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11 Central auditory processing disorders

J.S. Martin and R.W. Keith

INTRODUCTION

Some children, despite having normal peripheral hearing sensitivity, continue to face considerable difficulties in how they process auditory information. For more than 30 years, this general condition, commonly referred to as central auditory processing disorder (CAPD), or simply, auditory processing disorder (APD), remains an expanding area of professional interest amongst hearing-care professionals. The purpose of this chapter is to provide readers with a broad construct of APD and highlight current areas of professional interest and controversy. The underlying nature of APD and approaches to diagnosis and intervention are admittedly complex. Many aspects of the disorder at the time of this writing are still openly debated, making comprehensive reviews on the subject challenging. Readers interested in more in-depth information on topics presented in this chapter are referred to a number of recent textbooks devoted entirely to the subject.[1–5]

We begin by briefly highlighting several of the more controversial issues involved in the assessment of individuals suspected of having this enigmatic disorder. Next, we review common behavioural and electrophysiological characteristics of children at risk for APD. We conclude by reviewing different approaches to management and remediation.

CURRENT CONTROVERSIES IN APD

Although several consensus and position statements have been offered to help guide the diagnosis and intervention of APD,[6,7] most approaches, particularly those involved in the assessment of children, continue to evoke vivid debate among clinicians and researchers. A number of factors contribute to the differing viewpoints on APD, but most fundamentally reflect the fact that different conceptual frameworks of the disorder have been introduced over the years. Thus, prior to reviewing specific test strategies for APD, it is appropriate to briefly discuss a few of the more controversial issues on the topic.

When considering the nature of a central-auditory perceptually based deficit (ergo APD), the issue of modality specificity often arises. Some researchers imply that the APD label be reserved for individuals with a demonstrated deficit restricted to the auditory system,[6,8] whereas others question whether modality specificity is a necessary component in APD diagnostic criteria.[7] The most recent position statement from the American Speech-Language-Hearing Association[7] stipulates that APD arises from the 'difficulties in perceptual processing of auditory information within the CNS' (p. 2) as opposed to deficits in higher-order processes

(e.g. attention, memory and language), although such problems may coexist. Whilst we make no attempt here to completely resolve this issue, converging evidence from cognitive neuroscience would suggest that a complete dissociation of auditory 'abilities' from general cognitive functions, even on the basic tasks, is both impractical and perhaps impossible. We do not mean to suggest, however, that the diagnostic tests employed should disregard the extent to which other non-auditory-specific factors influence results. Indeed, more reliable and valid diagnostic measures capable of identifying a primary deficit attributable to the auditory mechanism while approximating the contribution of other general task or stimulus variables are needed.

Since there is no universally agreed upon criterion or 'gold standard' for the diagnosis of APD, the prevalence of the disorder is still unclear. Some clinical reports estimate the occurrence of APD in children to be between 2% and 3%.[9] In this regard, the question of how to best validate diagnostic procedures for the purpose of deriving sensitivity and specificity data remains controversial. One traditional approach taken to validate experimental test batteries for APD has been to administer tests on individuals with known lesions of the auditory central nervous system.[10] Whilst the assessment of individuals with demonstrated brain lesions can be immensely helpful in our understanding of APD, it has not been entirely effective. Such studies are often difficult to carry out on a larger scale, and the extent of the lesion(s) may not be entirely localized to auditory areas.[8] Another method for validating APD tests may be to take a 'population-based' approach.[11] This approach assumes that only a small percentage of the general population (about 5%) is believed to have genuine APD. By conducting comprehensive testing on a very large group of children, 'outliers' in performance are identified for the purpose of deriving which auditory processing and/or cognitive tests are sensitive and specific to those outliers. Studies incorporating principles of construct validity may also be useful for the purpose of validating APD test procedures. It is also anticipated that the combined utility of auditory evoked potentials and functional brain imaging techniques will be involved in the validation process.

Finally, much concern has been focused on the differential diagnosis of APD from other common childhood disorders, such as attention deficit hyperactivity disorder (ADHD), dyslexia, or expressive and receptive language impairments. Indeed, the question of whether APD and ADHD, for example, are two separate and distinct clinical entities, or whether they are indistinguishable behaviours or simply co-morbid conditions has received much attention. Readers interested in this topic are encouraged to see a review by Cacace and McFarland.[12] Their review addresses a wide range of intellectual, behavioural, educational, psychological, medical and social issues often associated with APD. For this reason, the importance of a transdisciplinary approach to APD management involving audiologists, speech-language pathologists, development psychologists and educators cannot be overstated, and speech-language services are designed to assess areas of strength and weakness for all aspects of language including discrimination, phonology, receptive and expressive language, prosody, semantics and pragmatics. Psychological measures can be used to establish the current level of functioning in different cognitive domains and various aspects of behaviour. As important as these additional measures may be, the diagnosis of APD is not made on the basis of such assessments. Information obtained on speech-language and psychological tests, however, can be helpful in guiding the selection of appropriate APD test measures and approaches to management.

BEHAVIOURAL CHARACTERISTICS OF POTENTIAL CANDIDATES FOR TESTING

Audiologists are initially faced with identifying those who should be tested. Most children are referred for testing in light of their poor academic achievements in reading, writing, spelling or delays in reaching other developmental milestones. Whilst problems in these areas do not directly indicate the presence of APD, these children are certainly potential candidates for central auditory testing. Children who report past or current auditory-specific difficulties, either by direct examination or parent report, certainly warrant further investigation. Keith[13] summarises the more commonly reported characteristics. These include:

- Demonstrates normal-hearing sensitivity but with significant history of prolonged middle-ear disease (e.g. otitis media).
- Responds inconsistently to auditory stimuli. Responds inappropriately on many occasions, but, at other times, follows auditory instructions in a normal manner.
- Difficulty with localising sounds, including the inability to tell the distance of the sound source, or the inability to differentiate between soft and loud sounds.
- Becomes frightened and upset when exposed to certain sounds (usually loud in nature) to the point that they cover their ears or remove themselves from the listening environment.
- Difficulty in discriminating between different sounds.
- Shows deficiencies in remembering phonemes and manipulating them on tasks such as reading, spelling, and phonics, as well as phonemic synthesis or analysis.[14]
- Difficulty understanding speech in the presence of background noise.
- Difficulty with auditory memory, either span or sequence, and poor ability to follow multiple instructions.
- Demonstrates poor listening skills and shows decreased attention for auditory information, distractibility or restlessness while listening in difficult listening situations.
- Frequently requests that information be repeated. Often responds to direct questions with ‘huh’ or ‘what’.
- Difficulty understanding rapid speech or individuals with an unfamiliar dialect.

Not every child suspected of APD will exhibit all of the characteristics mentioned above. The severity of characteristics, ranging from mild to severe, is unique to each child. Professionals should place great importance on the manner in which information is collected from the child or child’s caregiver. To aid in this process, screening questionnaires can be used to supplement the case history in order to objectively evaluate a child’s difficulties in various auditory dimensions.

Case history

A carefully conducted case history provides an invaluable source of information to help the clinician (1) differentiate between other conditions that masquerade as APD, (2) supplement results obtained on diagnostic tests and (3) guide decisions about management. Several working guidelines should be adopted when giving the case history. Caregivers should be given sufficient time to state their concerns about their child, describe their child’s auditory behaviours,

and express any other related concerns. Specific inquiries about a child's history should be made with the aim to clarify caregivers' responses rather than to guide the nature of their answers. The case history should also be taken in a systematic fashion to avoid missing important information about (1) hearing, language and learning problems that may exist in the family; (2) birth history; (3) the child's growth and development, health and illnesses; (4) general behaviour and social-emotional development; (5) speech and language development; (6) hearing and auditory behaviours and (7) educational progress.

Standard audiometric procedures

Although research suggests that prolonged periods of conductive or sensorineural hearing loss may alter the central representation of auditory information, it is generally advised to initially rule out such conditions in order to facilitate a clearer interpretation of test results. To this end, normal peripheral hearing sensitivity (i.e. thresholds of 15 dB HL or less at octave frequencies between 500 and 4,000 Hz) should be established via routine air-conduction audiometry. Bone conduction testing may be appropriate if the hearing thresholds for both ears differ by as much 10–15 dB. In such cases, tympanometry should also be performed to establish normal middle-ear function. Acoustic reflex testing can be helpful in establishing the neural integrity of the auditory pathways. Standard speech audiometry should also be carried out on both ears using age-appropriate materials. In the absence of hearing loss, children who show abnormal findings on such basic speech measures should heighten the clinician's suspicion of APD and/or other related auditory conditions, such as auditory dys-synchrony.

Otoacoustic emissions (OAEs)

OAEs reflect the non-linear properties of the cochlea, specifically the active processes of the outer hair cells, in response to sound. At this time, the principal rationale for including OAE testing in the APD test battery is to substantiate normal cochlear function. However, variations of the basic procedure, such as the contralateral suppression of OAE amplitude via stimulation of the olivocochlear bundle (OCB), may provide additional information concerning those mechanisms underlying the subject's ability to hear in noise.[15] It has been demonstrated that this suppression effect does not occur in situations where the OCB is compromised[16] or in cases of neural involvement.[17] Muchnik et al.[18] reported less suppression of transient evoked OAE activity in a group of children diagnosed with APD as compared with a control group. These results indicate that children with APD may show low activity of the medial OCB system, which mediates the suppression effect. Future research is needed to determine the clinical value of the OCB suppression phenomenon.

BEHAVIOURAL TESTS OF CENTRAL AUDITORY FUNCTION

The idea that only sufficiently challenging listening situations are capable of revealing central auditory weaknesses forms the basis of most behavioural tests of APD. While there is certainly no shortage of 'sensitised' test procedures that have been introduced over the past several decades to fill this role, we highlight a few of the more common tests used in the evaluation of children.

Tests of temporal processing

Since auditory information is experienced over time, deficits in 'temporal processing' within the auditory mechanism can affect speech-understanding abilities in different ways. Auditory discrimination, for example, is dependent not only the ability to distinguish differences in voice onset time for consonants such as /p/ versus /b/, recognising formant frequency transitions in contrasts such as /da/ versus /ga/, or deciphering between the duration of silent intervals between words (boundaries), but also in one's ability to discriminate and remember different patterning (sequencing) in auditory stimuli. Temporal patterning is fundamental to the identification of both segmental and suprasegmental aspects of speech. Any circumstance that interferes with the perception of these temporal intervals, either internally or externally, may reduce speech understanding or affect the child's learning.

Different clinical procedures have been introduced to assess auditory temporal processing. Some procedures evaluate the listener's ability to detect short temporal gaps in acoustic stimuli whereas others require the listener to discriminate the temporal order or sequence in stimuli. The basis for many of these procedures comes from research which shows that disturbances in the temporal aspects of audition have been linked to cortical dysfunction.[19–22] Two commonly used tests of temporal processing include the Frequency Patterns and Duration Patterns tests[23] and Random Gap Detection test (RGDT).[24]

In the Frequency Patterns test, listeners are presented at a comfortable listening level sets of three tones that vary in terms of pitch, either 'high' (H) or 'low' (L). Listeners report the pattern heard following each trial (e.g. HHL, LHL, HLL, etc.). If the child shows difficulty in verbally labeling the tones, he or she can hum or sing the pattern. Practice items are given to ensure that listeners understand the task. Results are evaluated in terms of the number of correct responses for stimuli delivered to each ear and are referenced to normative values.[23] The Duration Patterns test is administered and scored in a similar manner except that listeners are asked to respond to tone sequences in terms of their duration, either 'long' or 'short'. Whilst both the Frequency and Duration Patterns tests appear to be resistant to the effects of peripheral hearing loss, a high degree of variability may be present for younger listeners and may not be appropriate for those below the age of 7.[1]

The RGDT[24] is designed to measure one aspect of audition, namely temporal resolution or 'gap detection'. The procedure consists of several subtests. The first subtest (Subtest 1) consists of a practice and screening procedure to determine the appropriateness of the task for the individual listener. In Subtest 1, listeners are asked to identify whether a temporal gap exists in 500 Hz tone pairs. Stimuli are presented binaurally and at a comfortable listening level. The interstimulus interval (ISI) changes incrementally from 0 to 40 ms. In the second subtest (Subtest 2), the tone pairs are presented at octave points from 500 to 4,000 Hz; however, the ISI (0 to 40 ms) is randomly selected. The remaining subtests (Subtests 3 and 4) are similar to Subtests 1 and 2 with the exception that white noise click stimuli are used. In all cases, listeners are asked to verbally indicate how many stimuli were heard, or in special circumstances, respond manually by raising the number of fingers corresponding to their judgements. The average gap detection threshold is obtained using the individual RGDT thresholds at each test frequency. According to the criteria reference score provided by Keith,[24] thresholds greater than 20 ms are consistent with a problem in temporal processing.

Monaural degraded speech tests

A hallmark characteristic of children with APD is their difficulty communicating in background noise. In situations where the acoustic speech signal becomes degraded, successful listeners are able to extrapolate the meaning of a verbal message via the extrinsic redundancy inherent in the speech signal and the intrinsic redundancy via the auditory central nervous system in coding such information. Many authors suspect that children with APD lack these abilities. To confirm these suspicions, different types of monaural degraded speech tests have been proposed over the years.

Speech-in-noise tests make up one class of monaural degraded speech tests. These tests involve the presentation of speech stimuli in the presence of competing acoustic signals. The type of competing stimuli used varies across different tests, but speech-shaped noise or single and multi-talker babble are common examples. An important variable in speech-in-noise tests is the intensity of the speech message relative to the intensity of the competing stimulus, or message-to-competition (MCR) ratio. Since the type of competing stimulus and the MCR ratio are known to influence results, it is important to know the cut-off for 'normal' performance for both of these variables. Two standardised speech-in-noise tests designed for assessment of children include the Paediatric Speech Intelligibility test[25] and the Auditory Figure Ground subtest of the SCAN-C: Test of Auditory Processing Abilities in Children.[13]

Another class of monaural degraded speech tests includes procedures that reduce the extrinsic redundancy in speech by filtering out part of the speech spectrum. Early reports by Willeford and Billger[26] showed that children suspected of APD performed more poorly on low-pass filtered word tests, a trend that has subsequently been verified by a number of investigators. Presumably, the child with APD is unable to resist acoustic distortions of speech, resulting in poor listening abilities in acoustic environments that are less than optimal. When examining behavioural performance, however, it is important to consider the differences in filter parameters, such as the cut-off frequency and attenuation rate, which exist across different filtered word tests. Since performance on filtered word tests can be affected by high frequency loss, it is advisable to rule out peripheral hearing loss prior to testing. It is equally important to consider how other non-auditory-specific factors, such as attention or facility with the linguistic materials on the test, might also influence results. Common low-pass filtered word tests used in paediatric assessment include the Filtered Words subtest of the SCAN-C [13] and the Auditec recording of the filtered NU-6 word lists.

A final class of monaural degraded speech tests discussed here are Time-Compressed Speech tests. These tests aim to explore the capacity of the auditory system to handle rapid changes in the acoustic signal. Different approaches to time compression have been explored, but most accomplish the task by re-sampling or deleting small segments of the speech signal. The amount of time compression, i.e. the amount of the original speech signal eliminated, is often expressed as a percentage (e.g. 0%, 40% and 60%). In addition to the amount of time compression, the type of speech stimulus used (e.g. words or sentences) can also influence results. Two standardised Time-Compressed Speech tests include the NU-6 words on the US Department of Veterans Affairs compact disc[27] (Tonal and Speech Materials for Auditory Perception, 1992) and the Time-Compressed Sentence test.[28]

Dichotic tests (binaural tests of separation or integration)

Dichotic listening (DL) tests are commonly used in research and clinical practice to examine hemispheric specialisation and interhemispheric interaction.[29] Listeners simultaneously receive

competing auditory signals, usually syllables, words or sentences, with one or more being presented at a comfortable listening level to each ear. In general, listeners respond to dichotic stimuli under one of two different types of instructional sets: free report and/or directed report. In free report, listeners are asked to respond to stimuli heard in either ear and in no specific order. In terms of the allocation of attention, such instruction is often referred to as a divided-attention DL paradigm. An example of this test approach is the Dichotic Digits test.[30] In pure directed report, listeners are asked to respond only to the stimuli presented to a predetermined side. Such instruction is sometimes referred to as a directed-attention DL paradigm. An example of this test approach is the Competing Sentences subtest on the SCAN-C test.[13] Some procedures incorporate a combination of the two approaches; listeners are asked to report stimuli heard in both ears, but in a certain order. This situation is referred to as divided attention with a pre-cued direction of report.[31] An example of this test approach is the Competing Words subtest on the SCAN test.[13] In most dichotic tests, the dichotic signals are recorded with simultaneous alignment of onset and offset of stimuli. An exception to this is the Staggered Spondaic Word test,[32] where the onset and offset of each spondee differs in time. In this approach, the examiner can obtain a score for each ear when stimuli are presented in a non-competing or competing (i.e. dichotic) listening situation.

Irrespective of the linguistic material and DL paradigm used, most individuals are slightly more accurate in their report of right-sided inputs than left-sided inputs. This direction of interaural asymmetry, commonly referred to as the dichotic right-ear advantage (REA), has been used extensively to study hemispheric dominance for language[33] and various aspects of attention. Although the neural mechanisms underlying ear advantage on DL tests are still under investigation, most reports attribute the phenomenon to (1) cerebral lateralisation in auditory function,[33] (2) bottom-up processing biases due to static asymmetries in the structural organisation of the central auditory nervous system[34] and (3) top-down or 'controlled' factors relative to the asymmetric allocation of attentional resources.[35]

There is a long line of research to suggest that patients with various brain disorders (e.g. lesions of the temporal lobes, frontal lobes and corpus callosum) show abnormal performances on DL tests.[29,36–40] Whilst overall DL performance by these patients is often poor, many demonstrate substantial discrepancies between the right-ear and left-ear scores, generally in the form of a substantial left-sided deficit (LED). Interestingly, an LED on DL tests has also been reported in cases of children showing signs of APD.[41–44] Studies using auditory event-related potentials (ERPs) to explore interaural asymmetry in these children have obtained similar results.[44,45] Given the similarity in test performances between these two different clinical populations, it has been hypothesised that the LED arises from a disruption or maturational delay in the interhemispheric transfer of auditory information.[13,46–48]

Prior to the selection of any DL test for the evaluation of APD, it is important to consider the various methodological factors and listener characteristics known to influence results. Of most concern is how attention influences the magnitude of interaural asymmetry (i.e. REA). Since the degree of ear advantage serves as an important index in the assessment of central auditory function, the most reliable and valid estimate of interaural asymmetry is critical. Studies using directed-attention or 'focused' procedures have revealed that the degree of asymmetry obtained on divided-attention tasks can be susceptible to the influence of spatial attentional factors.[49] Some authors suggest that the combined utility of both divided-attention and directed-attention procedures may be valuable in approximating the relative contributions of bottom-up and top-down factors on DL function in individuals.[31,50] Behavioural procedures that aim to minimise, or at least approximate the contribution of non-auditory-specific factors, such as attention, on DL test results in children are still needed.

Binaural interaction procedures

As implied by the name of this general category of tests, the basis of binaural interaction procedures come from the fact that both ears interact in diverse ways to influence our perception. A variety of tests fit this category. Some procedures target specific lateralisation abilities and rely on the assessment of interaural timing or intensity differences set up between the stimuli delivered to each ear,[51,52] whereas others examine auditory localisation function using stimuli presented in the sound field.[53] Still others rely on the fact that acoustically modified stimuli, when presented to each ear in isolation are difficult to discern, but when presented dichotically, for example, the stimuli become easily understood or 'fuse' together.[54] Variations of the general idea have been proposed by Willeford and Billger[26] and Wilson and Mueller.[55]

The most common binaural interaction procedure used for the assessment of APD in children is the masking level difference (MLD) test. The MLD refers to the difference in thresholds for speech or tonal signals when presented in two different binaural masking paradigms. In one listening situation, referred to as a homophasic test condition, the signals and noise maskers in both channels are in phase with one another. In another listening situation, referred to as the antiphasic condition, either the signals or the noise maskers are out of phase. Although a variety of stimulus arrangements are possible, it is generally expected that a listener's ability to detect the signal will improve (i.e. in decibels) when either the signal and/or noise masker presented to the ears differ on some dimension (e.g. phase or timing). The MLD can be 10–15 dB for pure tones and is frequency dependent, with the largest effects usually observed in the lower frequencies (300–600 Hz).

The neural mechanisms underlying binaural interaction phenomena are believed to be primarily housed in the lower levels of the brainstem.[56] The amount of hearing loss is known to reduce the MLD effect, and brainstem lesions can substantially reduce or eliminate MLD altogether.[57] The MLD effect has also been reported to be reduced in children with suspected auditory perceptual problems.[58] They found that tonal MLDs were more effective in distinguishing children with auditory perceptual dysfunction from normal children, but speech MLDs were not. Although the MLD has been discussed in reference to specific auditory abilities, such as listening to speech in noise, it is best to utilise the procedure as a behavioural measure of brainstem integrity. For this purpose, the MLD can serve as a valuable tool.

ELECTROPHYSIOLOGICAL TESTS OF CENTRAL AUDITORY FUNCTION

The utility of electrophysiological procedures to examine the integrity of auditory central nervous system has a rich history. A family of auditory evoked responses (AERs) can be used to establish neural function at all different levels of the auditory system, from the brainstem through the auditory cortex. AERs are often grouped according to whether they represent mostly exogenous or endogenous aspects of information processing. The auditory brainstem response (ABR), auditory middle latency response (AMLR) and portions of the auditory late response (ALR) are often referred to as exogenous potentials, as they occur in response to external events and reflect primarily changes in the acoustic features of the stimulus. Other longer latency evoked responses, sometimes referred to event-related potentials (ERPs), reflect

more cognitive aspects in information processing or changes in task demands. Common ERP examples include the P300 or P3, N400 or N4, P600, and contingent negative variation (CNV). Readers interested in more detailed information on AER acquisition and interpretation are encouraged to review recent textbooks devoted to the topic.[59,60]

Auditory brainstem response (ABR)

The ABR represents neuroelectrical activity from the auditory nerve and nuclei predominately along the ascending auditory brainstem pathway.[61] When elicited with a relatively intense stimulus with abrupt onset (a click), the traditional ABR appears as a series of five major waveform peaks within the first 10 ms post stimulus onset. Because the ABR reflects pontine-mesencephalic transmission of neural activity, it is generally regarded as a measure of central auditory processing at the brainstem level.

Whilst the ABR is widely carried out on clinical populations with confirmed neuro-otologic disorder, head trauma, auditory deprivation or individuals who present 'hard' neurological signs, it is still not often utilised for routine assessment of APD in children. Presumably, this reflects the fact that the ABRs recorded using standard recording techniques in children suspected of APD are typically normal.[9] Only a few reports have indicated some abnormalities.[62,63] Given the increased awareness of auditory neuropathy/auditory dys-synchrony, however, including the ABR in the APD test battery may be helpful to rule out such conditions.

Methodological changes have been proposed in an effort to make the auditory evoked brainstem potentials more effective in the identification of children at risk for APD. Jirsa[64] evaluated the clinical utility of the maximum length sequence (MLS) of the ABR in children diagnosed with APD. Results showed that the ABR wave V latency was prolonged in this group as compared to an age-matched control group on the same MLS procedure. Gopal and Pierel[65] and Delb, Strauss, Hohenberg and Plinkert[66] investigated the diagnostic value of the binaural interaction component (BIC) of the ABR in cases of suspected APD. The BIC is considered an electrophysiological index of binaural interaction and is computed as the arithmetic difference between the sum of the evoked potentials obtained for each ear separately and the evoked potentials evoked with binaural stimulation. Compared to a group of control listeners, Gopal and Pierel[65] found that the amplitude of the BIC component was significantly reduced in the group of suspected APD listeners, indicating possible weakness between the groups in how they process binaural inputs. In a similar approach, Delb et al.[66] investigated BIC measurements in a group of children at risk for APD, but with the additional goal of deriving some sensitivity and specificity data. According to these authors, a 76% sensitivity and specificity was achieved using the BIC measurements.

More recently, Kraus and colleagues have begun to uncover the relationship between the acoustic structure of speech processing and brainstem functioning using speech-evoked auditory brainstem potentials called the BioMAP™.[67,68] Their approach utilizes a complex speech stimulus (/da/) to elicit potentials that correspond to both transient (i.e. ABR) and sustained aspects (i.e. frequency-following response) of stimulus encoding at the level of the brainstem. Their findings indicate that approximately one-third of children with demonstrated language-based learning problems exhibit a unique pattern of neural activity that is different from children with other types of learning problems. Whilst different aspects of the clinical utility of this procedure are still under investigation, the implication of their research is important – brainstem function in some children suspected of APD may be more 'atypical' than once originally suspected.

The auditory middle latency response (AMLR)

The AMLR is a series of waveform components that occur within the first 10 to 60 ms following stimulus onset and reflect the interaction of a number of neural generators, including primary auditory cortex, thalamocortical projections, reticular formation in the midbrain and thalamus, inferior colliculus and medial geniculate body of the thalamus.[1] Several reports in the literature indicate that the AMLR can be useful for the assessment of individuals suspected of APD.[69–73] More recently, Purdy, Kelly and Davies[74] compared the AMLR obtained from a group of children diagnosed with learning disabilities (LD group) and those from a control group. They found that the latency of the Na component was significantly prolonged and the amplitude of the Nb component was significantly reduced for the LD group as compared to control listeners, thereby indicating possible APD. In principle, the AMLR is well suited to examine central aspects of auditory function due to loci of the neural generators underlying the response. Various methodological factors, particularly maturational and level of arousal issues, as well as the continued lack of sensitivity and specificity of the AMLR in the assessment of APD[9] have minimised its routine administration. More research in this area is still warranted.

Auditory late responses (ALR)

The ALR can be divided into subcategories to reflect differences in exogenous or endogenous states of information processing. Earlier appearing waveform components of the ALR, such as the P1 component, which some consider to be part of the AMLR, and the N1 component, are often regarded as 'obligatory' responses since they are generated without conscious attention or active participation of the listener. There is considerable evidence to suggest that the neural generators of these components originate from multiple neural ensembles located at both subcortical and cortical levels of the central nervous system (e.g. thalamocortical projections, associative auditory cortex and primary auditory cortex). For this purpose, the P1 and N1 components appear to represent basic sensory encoding aspects of the evoking acoustic stimulus at higher levels of the auditory system. Other commonly elicited components of the ALR, such as the P2 and N2, do not appear to solely reflect basic sensory encoding of the auditory system per se, but rather, represent higher-order processes relative to further stimulus evaluation and analysis or changes in the subject's state.[1]

A number of studies have reported differences in ALR morphology between children with various language-based learning problems (LP) and their typically developing peers.[75–79] However, as Gilley et al.[77] have shown, the morphology of individual ALR waveform components can change as a function of age and stimulation rate. Thus, before the ALR is to be carried out for the routine assessment of APD, the complex maturational patterns of the ALR need to be thoroughly addressed. When these factors are accounted for, the results from Gilley et al.[77] suggest that the ALR could be used to identify abnormal patterns of central auditory maturation.

Of the more endogenous components of the auditory ERP, the P3 (P300) component is the most widely used by audiologists in the clinical setting. Elicited in an 'oddball' paradigm, the P3 is most robust when rarely occurring stimuli (i.e. target tones) are pseudorandomly presented (10–20%) amongst frequently occurring stimuli (i.e. non-target tones). Listeners are typically asked to keep a mental count of the number of 'targets' they encounter. Although it

is still unclear as to what exact mental operations the P3 indexes, its amplitude and latency appear to reflect a listener's ability to recruit a number of processing resources (i.e. attention, auditory discrimination, memory) to either arrive at a decision about a contextually important stimulus or to classify it into task-relevant categories.[80,81] It is important to remember that the P3 is multimodal in nature and can be elicited via auditory, visual and tactile stimulation. A variety of diverse brain regions (e.g. frontal, parietal and temporal lobes and hippocampus) contribute to its generation.[82,83]

There is sufficient evidence to suggest that the P3 component appears sensitive to a host of neurological conditions, cognitive impairments, and focal brain (cerebral) lesions. Some reports also indicate group differences in basic P3 measures for children with various language-learning impairments, such as APD[74,84] and attention deficit hyperactivity disorder.[85] Other researchers have noted group differences in the topographic distribution of the P3 in both adult and paediatric populations. [86,87] A recent study by Martin and colleagues[50] also examined the P3 and N4 ERP components in a group of children identified with deficits on a commonly administered test of dichotic listening (DD group). These authors were particularly interested in separating, experimentally, the contributing factors underlying their poor dichotic performance. Stimuli were presented using diotic, divided-attention and directed-attention dichotic paradigms. Results showed that the P3 component for the DD group, as compared with age-matched control listeners, was delayed in latency and reduced in amplitude when stimuli were presented under a divided-attention mode. When stimuli were presented under a directed-attention or diotic mode of administration, P3 measures for the two groups were similar. Overall, their results suggest that a principal factor contributing to the difficulties that some children face on DL tests may stem from an inability to allocate appropriate attentional resources.

Mismatch negativity (MMN)

Another ERP that has also received considerable interest in the study of auditory processing is the MMN. The MMN is elicited in a similar paradigm as that used on the P3 task, with the general exceptions that (1) listeners are not asked to specifically attend to the stimuli or provide any behavioural response and (2) smaller differences usually exist between the rare and frequent stimulus in some respect (e.g. frequency or duration contrast). In this paradigm, the MMN waveform is derived by taking the difference (subtraction) between the rare and frequent stimulus events, which appears as a prolonged negativity over approximately 100–300 ms (or longer). The difference between these two waveforms, the MMN, is believed to reflect the brain's 'automatic' or pre-attentive response to stimulus change.[88] The MMN can be elicited with a variety of complex stimuli, such as speech, and can be observed even when the physical difference between the evoking stimuli approaches psychophysical boundaries.[89]

The precise anatomic locations underlying the MMN is still growing, but the non-primary thalamus, primary auditory cortex and association auditory cortex have been implicated.[90–92] Some frontal contributions have also been suggested.[93]

The auditory-specific nature of the MMN makes it an attractive tool to study APD. Indeed, a number of researchers have used the MMN to examine basic sensory discrimination abilities (i.e. speech perception) in children who exhibit APD-like characteristics and to monitor changes following behavioural training.[94–97] More recently, Sharma et al.[98] examined MMN responses in a group of children identified predominantly for reading problems but who also

failed a test of auditory processing (frequency patterns), thus also meeting their diagnostic criteria for APD. Compared to a group of control listeners, the speech-evoked MMN was significantly reduced in amplitude for group with diagnosed reading disorders. Naturally, the significance of their results speaks to the co-morbidity of APD and reading disorders.

The clinical utility of the MMN, however, has been questioned. Issues regarding its accurate detection, reliability, and validity on an individual basis have been raised.[99,100] Two general problems have tempered the initial excitement regarding the clinical utility of the MMN. First, the MMN is not always present in individuals who can easily distinguish, behaviourally, between the stimuli contrasts used to evoke the response.[99] A study by Dalebout and Stack[101] showed that the MMN was not present in one-third of normal young adult listeners, suggesting the possibility of false positive outcomes in a clinical situation. A second problem relates to the difficulty in quantifying whether a MMN response is actually present. Fox and Dalebout[102] evaluated the median method to signal averaging in an attempt to better define the MMN. In most cases, the median method proved to be of no additional benefit. Cacace and McFarland[103] reported a statistical method to quantify the signal-to-noise (SRN) ratio of the MMN based on the Pearson product moment correlation coefficient (Pearson's *r*). Given the poor SNR of the response, their results suggest that more research is needed to improve the detection of the MMN amongst individual listeners before it transitions to the clinical setting.

MANAGEMENT AND REMEDIATION

Different approaches to intervention are taken to address the auditory processing deficits unique to a particular child. Irrespective of the intervention strategies selected, an important question is whether they are efficacious. Whilst some children respond favourably to intervention, other children with similar behavioural profiles do not. The variability observed in intervention arises from different sources. Foremost, it likely reflects the aforementioned difficulties in identifying the true nature of APD in individuals. Many have argued that without accurate diagnosis, the effectiveness of treatment is difficult to measure. To a lesser extent, it may also reflect fundamental differences between the professional groups (e.g. audiologists, speech-language pathologists, educational psychologists and educators) in how they approach management. Given that children with APD often experience deficits beyond basic perceptual auditory-based functions, such as developmental delays in integrating processes relative to language and literacy and weaknesses in cognitive/metacognitive abilities, an interdisciplinary approach to intervention is essential.

Two terms are commonly used in discussing intervention following an APD diagnosis: management and remediation. Whilst the terms are often used interchangeably, some authors believe that they have distinct meanings and implications for APD. The term 'remediation' reflects an actual altering of central auditory nervous system function whereas 'management' involves the modification of behaviour, performance or environment with compensatory or cognitive techniques. Whilst the appropriateness of terms used to describe a given intervention strategy can be argued, both aspects are viewed as necessary components of a successful intervention plan. For this purpose, intervention strategies for management and remediation of APD are classified into different subgroups. Irrespective of the type of APD diagnosis, Ferre[104] suggests that intervention should include techniques to address three different areas: (1) modification of the environment, (2) use of compensatory strategies and (3) direct remediation. The following is a brief overview of these different strategies.

Environmental modifications

Children with APD face a number of challenges to auditory processing whilst in the ordinary classroom. Here, they experience a substantially impoverished speech signal due to reverberation, reflections and competing signals. The overall SNR is generally less than to be desired. There are things that can be done, however, to enhance the listening environment and improve the child's ability to use auditory information. From Ferre[104] some of the more common environmental modifications include:

1. Noise abatement through modification of the classroom to reduce sound reflections and noise. Carpeting floors, placing curtains, drapes or acoustic tiling along the walls and ceiling, isolating extraneous noise sources inside and outside the classroom, or strategically placing bookcases or objects in the classroom to create baffle and minimise noise can be helpful.
2. Signal enhancement via assistive listening devices (ALDs) such as FM systems. Whilst the overall goal of these devices is to improve the SNR of the auditory signal to improve speech perception, improvements in auditory attention and memory have also been reported. FM systems can also be tailored to the individual needs of the child.
3. Changes in oral message presentation. Slowing the overall speaking rate, repeating and rephrasing information, or emphasising key points can improve both acoustic and linguistic saliency.
4. Classroom seating configurations or preferential seating. These modifications can improve signal audibility by maximizing both acoustic and visual cues to the listener.

Compensatory strategies

Compensatory strategies are often used to strengthen 'listening' abilities and teach specific academic skills. There are many different approaches to teaching children how to recognize those skills that will assist them in their academic and social endeavours. In essence, these strategies aim to teach children that their auditory comprehension can be improved through their active participation. Once a particular weakness is recognised, steps can be taken to minimise those barriers contributing to their failures in communication. Readers are encouraged to see Bellis[1] for a review of specific metacognitive, linguistic and metalinguistic compensatory strategies that can be used following the diagnosis of APD.

Direct remediation

The final group of intervention strategies briefly discussed here are those that directly attempt to alter the brain's ability to organise or reorganise information. For this reason, they are generally the more controversial techniques used in APD intervention. Direct remediation therapies are based primarily on research suggesting that the brain's response can be modified and lead to a change in behaviour with systematic and adaptive training. Various therapies have been introduced to strengthen or alter specific auditory processes within the central nervous system. Of course, the selection of the appropriate therapy programme becomes challenging when the nature of the auditory processing deficit remains uncertain.

Ferre[104] identifies two broad types of direct remediation therapies for APD: those that target the bottom-up or the top-down processes. In general, bottom-up therapies aim to remediate

specific auditory deficits whereas top-down therapies target concept-driven abilities or the application of higher-order linguistic rules, such as auditory closure, schema induction and prosody training.[104] Bottom-up therapies focus on perceptual training to address the integral aspects of audition. Some programmes focus on the listener's ability to discriminate between different speech sounds through manipulation of the spectrotemporal characteristics of the acoustic signal. Recent commercially available computer-assisted programmes that aim to improve temporal processing skills (and other skills) include Fast ForWord™,[105] Earobics™,[106] and SoundSmart™.[107] These adaptive training programmes employ a variety of games designed to train discrimination abilities.

Other training strategies focus on binaural processing and localisation abilities by examining how information presented to both ears interacts at the cortical level. For this purpose, dichotic listening exercises are often incorporated. Training programmes that aim to improve the listener's ability to use temporal (or patterning) attributes of auditory information are also available. These programmes use both speech and non-speech stimuli and typically incorporate activities that focus on sequencing either by direct identification or imitation of auditory patterns by humming or tapping. Programmes of this type include the Processing Power[108] and the Lindamood-Bell Phoneme Sequencing programme.[109] Still other remediation techniques examine related skills, such as auditory attention, interhemispheric communication, speech recognition in noise and lip-reading. Readers interested in learning more about exercises targeting these areas are encouraged to see the review by Ferre.[104]

CONCLUSION

Professional interest in APD in children has steadily increased over the past several decades. Although some gains have been made in our understanding of APD in this population, questions about how to best differentially diagnose the problem or which approaches to intervention are efficacious still remain. Similarly, whilst a number of behavioural tools are available for the assessment of APD, the extent to which other variables might influence results, such as intelligence, facility with stimulus materials, memory and attention, deserve additional consideration. More research is needed to reveal the underlying nature of APD in children. Through better understanding, the efficacy of various approaches to assessment and intervention can be determined.

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12 Auditory neuropathy

D. Bamiou

INTRODUCTION

The 8th nerve lies in the internal auditory canal (IAC) and consists of three segments: (a) the auditory (cochlear) nerve, which contains afferent auditory fibres; (b) the vestibular nerve, which contains afferent vestibular fibres; and (c) efferent fibres, which travel in the olivocochlear bundle. The auditory nerve occupies the anteroinferior part, the vestibular nerve occupies the posterior half and the facial nerve occupies the anterosuperior quadrant of the IAC. The 8th nerve enters the pontomedullary junction of the brainstem at the cerebellopontine angle (CPA).

The auditory nerve has approximately 30,000 auditory nerve fibres, which synapse with the hair cells in the organ of Corti. Ninety-five per cent of these fibres will synapse with the inner hair cells (IHC), i.e. the cells that generate electrical current in response to sound, and the rest will synapse with the outer hair cells (OHC), i.e. the motile hair cells, which amplify the incoming sound. Each IHC synapses with 10–30 afferent endings of type I neurons, whilst each OHC synapses with 4–10 afferent endings of type II neurons.[1] The cell bodies of these neurons form the spiral ganglion that lies in the Rosenthal's canal. IHC are divergent, in that many type I neurons receive input from a single IHC, whilst each neuron synapses with many cells in the cochlear nuclei. Fibres from the low-frequency encoding apex of the cochlea occupy the core of the nerve trunk and end in the ventral portion of the cochlear nucleus, whilst fibres from the high-frequency base are at the periphery of the nerve trunk and arborise in more dorsal parts of the cochlear nucleus.

The vestibular nerve has approximately 15,000 fibres and consists of a superior division that innervates the anterior and horizontal canals, the utricle and part of the saccule, and an inferior division that innervates the posterior canal and the main part of the saccule. The cell bodies of vestibular neurons form the Scarpa's ganglion and vestibular nerve afferents terminate within the vestibular nuclei. The ventral component of the inferior division of the vestibular nerve consists of the olivocochlear bundle, which carries efferent innervation to the OHC of the contralateral cochlea by the medial bundle and to radial afferent fibres in the ipsilateral cochlea by the lateral bundle.

OVERVIEW OF TESTS OF 8TH NERVE FUNCTION

When there is a clinical suspicion for pathology of the 8th nerve, comprehensive assessment of all its three divisions, i.e. the auditory, vestibular and olivocochlear branches, is required. At present, there is no single diagnostic procedure which will identify the cochlear or vestibular

nerve involvement, as current audiovestibular procedures do not allow the differentiation between lesions affecting the receptor hair cells, the hair cell to nerve synapses or purely neural lesions. Diagnostic evaluation of suspected 8th nerve pathology requires a test battery approach, with application of several behavioural and electrophysiological tests, and with interpretation of the results based on a good understanding of the relevant physiology.

BACKGROUND OF THE TERM 'AUDITORY NEUROPATHY'

In 1996, Starr et al.[2] described ten subjects (five adults and five children) with mild to moderate sensorineural hearing loss, with onset of the hearing complaints in childhood or early adulthood. These subjects had normal OHC function, as indicated by the presence of intact otoacoustic emissions and/or electrocochleography, with absent or severely abnormal auditory brainstem evoked potentials, absent stapedial reflexes and absent suppression of otoacoustic emissions by contralateral noise (which is mediated by the olivocochlear bundle). The hearing impairment preceded the onset of a peripheral neuropathy in eight of their cases. Starr et al. proposed that this presentation was indicative of an auditory nerve lesion, which might have a neuropathy of the auditory nerve as one of its causes, and thus, coined the term 'auditory neuropathy'.

'Neuropathy' is the term that refers to pathology of peripheral nerve fibres, which may be divided into three major types: demyelinating, axonal and mixed. 'Neuronopathy' refers to pathology in the cell bodies of origin of the neural fibres. Current clinical tests, however, do not allow differentiation between an end organ versus neural or ganglion dysfunction. To complicate matters more, pathology, which affects a specific part of the auditory pathway, will not remain 'pure' in the long term, as degeneration may take place both in an anterograde and retrograde fashion. Thus, damage of the IHC or of the peripheral afferent dendrites will lead to death of the spiral ganglion neurons with these dendritic connections because these neurons will be deprived of the hair cells' trophic influences, and this will lead to long-term degeneration of the auditory nerve ('trans-synaptic degeneration'), which may lead to degeneration in higher parts of the ascending auditory pathway.[3] Cochlear neuronal degeneration may similarly take place due to central pathologies (at the level of the brainstem or above), but as these pathologies also tend to affect the vascular supply of the end organ, this is less well-studied.[4] Several authors feel that current use of the term of 'auditory neuropathy' may be inappropriate, in that it may group together a wide range of auditory pathologies.[3] Others have proposed the broader term of 'auditory dys-synchrony',[5] which would include both a true neural abnormality, and other possible underlying mechanisms, which would result in neural dys-synchrony, such as possible delayed maturation of the lower-level auditory pathway, in the case of newborns.[6] However, it must be noted that in the case of delayed maturation, recovery would be expected to take place within 12–18 months.[5] In order to acknowledge the ambiguities regarding the site of lesions due to current tests limitations, this chapter will use the term 'auditory neuropathy/auditory dys-synchrony' (AN/AD).

DEFINITION AND DIAGNOSIS

Auditory neuropathy/dys-synchrony (AN/AD) refers to the hearing impairment seen when cochlear amplification (i.e. OHC function) is relatively preserved, but afferent neural conduc-

tion in the auditory pathway is disordered.[2,5] Sininger and Oba[7] proposed that diagnosis of AN/AD ought to be based on all three of the following criteria:

(1) Evidence of poor auditory neural function, with the inability to record evoked neural activity at the level of the auditory nerve (compound action potential), abnormal auditory brainstem response (ABR) (i.e. latencies >2 s.d. beyond the normal range, and/or amplitudes significantly below normal and/or abnormal waveform morphology[8]), and elevated or absent other auditory brainstem reflexes such as stapedial reflexes and otoacoustic emissions (OAEs) suppression by noise.
(2) Evidence of normal OHC function, such as normal OAE or cochlear microphonics on electrocochleography (ECochG). However, OAEs can disappear over time, whilst cochlear microphonics may remain present.[9]
(3) Evidence of poor hearing in the presence of normal or abnormal audiometric thresholds. AN/AD can be associated with any degree of hearing loss or audiometric configuration and is characterised by marked hearing fluctuation[7] with characteristic temporal and speech perception deficits which are disproportionate to the audiometric thresholds.[10]

Finally, Sininger and Oba[7] proposed that exclusion of other potential causes ought to be made by appropriate investigations, such as a brain magnetic resonance imaging (MRI) to establish the structural integrity of the auditory nerve. A recent study identified hypoplastic or absent 8th nerve on brain MRIs in just under 20% of children with clinical characteristics of AN/AD.[11] This was more likely to be true if AN/AD was unilateral and/or the associated hearing loss was profound.

For newborn infants and babies up to the age of 4 months, in particular, the UK Newborn Hearing Screening Programme (UK NHSP) Protocol[6] recommends that assessment for suspected AN/AD should include:

- OAEs – either transient evoked (TEOAE) or distortion product (DPOAE).
- Click-evoked ABR. If the ABR is absent or severely abnormal (i.e. with only wave V identifiable), separate runs of condensation and rarefaction clicks at 80 dB nHL ought to be conducted in order to identify a CM. Testing for the CM is necessary if OAEs are absent (as the CM may still be present), whilst if OAEs are present, the identification of a cochlear microphonic, which inverts with the change of click polarity, may further confirm the presence of AN/AD. In addition, it is recommended that the CM is recorded with 'tube' insert earphones, which will introduce a time delay between the electrical signal at the transducer and the acoustic signal at the ear canal and may thus enable separation of the CM (which is temporally close to the sound stimulus) from the electromagnetic stimulus artefact. Once the CM has been identified, an additional control run ought to be performed, with the tube insert clamped, as this would eliminate the true CM but not an electrical artefact (Figure 12.1).

The use of stapedial reflexes is debatable for this age group. Berlin et al.[12] reported that acoustic reflexes are either absent or observed at higher levels than 100 dB HL (which would be the levels expected in light of the normal OAEs) in 133 out of 136 AN/AD subjects tested. They proposed that ipsilateral stapedial reflexes ought to be tested at least at 1 and 2 kHz in any perinatal hearing screening that depends solely on otoacoustic emissions. However, at present there is no evidence to show that stapedial reflexes are recordable in infants younger than 4

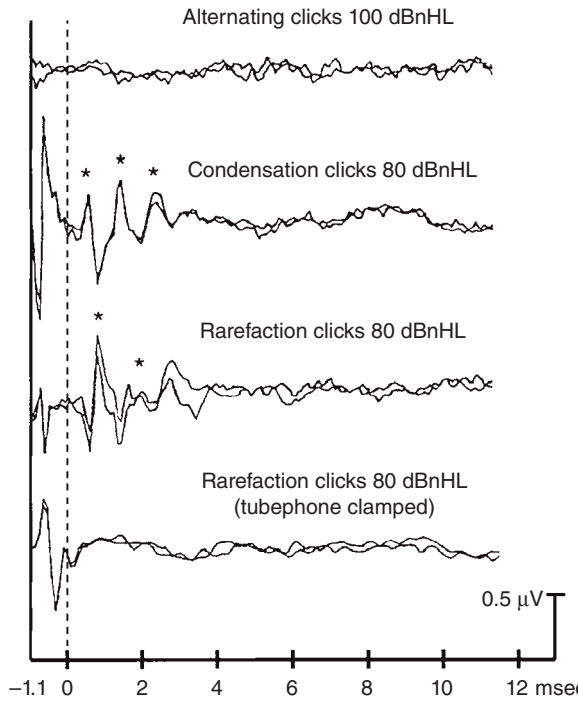

Figure 12.1 ABR recordings for a 3 year-old child with AN/AD type hearing loss. The dotted line represents the point at which the stimulus reached the cochlea. The top tracings show no repeatable potentials to alternating clicks presented at 100 dB nHL. The middle tracing pairs show repeatable cochlear microphonic responses but absent brain stem response waveforms to unipolar stimuli at 80 dB nHL. The asterisks indicate the positive peaks in the cochlear microphonic waveform. The final tracings, in which only the stimulus artefact is evident, were obtained to rarefacting clicks presented with the tubephone clamped. Reprinted from Rance 2005, with kind permission.

months, where tympanometry and stapedial reflexes must be recorded with high-frequency probe tones (1,000 Hz) and stapedial reflexes are not part of the UK NHSP standard protocol for this age group. Babies older than 4 months should have stapedial reflexes at 1,000 and 2,000 Hz stimuli, preferably with contralateral measurements.

HISTOPATHOLOGY AND PATHOPHYSIOLOGY

Post-mortem biopsy of the auditory nerve in an adult case of auditory neuropathy accompanied by peripheral neuropathy showed two major changes of the auditory nerve[13]:

(1) Demyelination. It has been argued that this change would lead to disruption of the temporal synchronicity of discharges to the sound stimulus, as fibres that are demyelinated to a different degree will transfer the neural signal with different speeds (Figure 12.2). In addition, these fibres will have prolonged refractory periods, and repetitive activation may result in a progressive increase in the conduction time. The ABR may be very sensitive to stimulus rates, i.e. it may be elicited by a low stimulus rate but may disappear at higher rates.

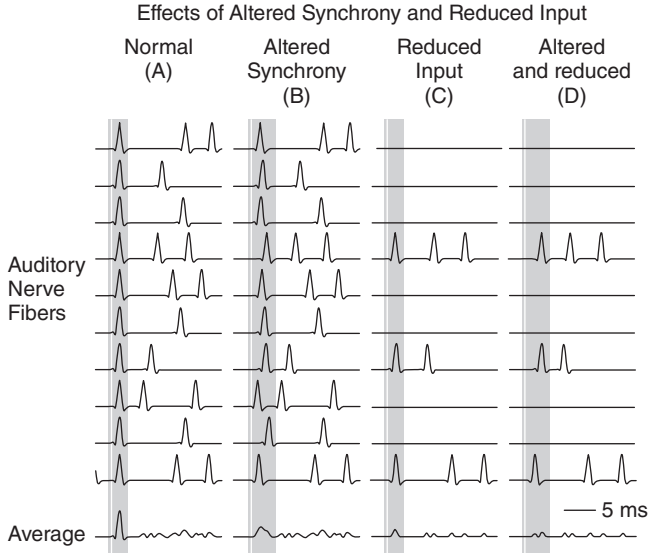

Figure 12.2 A computer model of the discharges of ten auditory nerve fibres synaptically related to a single inner hair cell. The fibres are spontaneously active and also discharge at short latency to a transient sound stimulus presented at the vertical line. The average potential of these fibres is shown below each column: (A) normal, all fibres discharge synchronously to the acoustic stimulus; (B) altered synchrony, all fibres discharge but their latency is delayed up to 1 ms from A; (C) reduced input, only three fibres are active without a change in synchrony; (D) combination of altered (synchrony) and reduced input. Note that the averaged nerve activity is both reduced in amplitude and prolonged in duration in B; reduced in amplitude in C and reduced in amplitude and prolonged in duration in D, making it indistinguishable from spontaneous activity levels. Reprinted from Starr et al., 2003, with kind permission.

(2) Axonal loss and reduced numbers of auditory fibres, which conduct the auditory input to the cortex. This loss would be compatible with a reduction in the amplitude of the neural action potential and ABR rather than an increase in latency (Figure 12.2).[14]

There are several human genetic models for this type of AN/AD, such as mutations in the MPZ gene or the connexin 32 gene (see aetiology section for more details).

AN/AD could also be due to a disorder of the synapse between IHC and the auditory nerve, as seen in cases with mutations in the otoferlin gene, which encodes a protein at the base of the IHC that is thought to be involved in synaptic vesicle recycling.[15] In these cases, the ABR may also be particularly sensitive to fast stimulus rates.[13]

Finally, AN/AD may be due to selective IHC loss, as demonstrated by animal models of AN/AD.[16] Animal genetic models for this possible presentation include the Bronx Waltzer mouse and the Beethoven mouse models.[8] In addition, a recent histopathological post-mortem study[17] on 15 neonatal intensive care baby unit cases identified three cases with isolated IHC loss (Figure 12.3), none of whom had a recordable ABR at 40 dB nHL before they died, and two babies with a mixed inner and outer hair cell loss, all of whom had an intact auditory nerve. The ABR absence in the presence of IHC lesions (as well as in cases of neuropathies with axon loss) is thought to be primarily caused by a reduction in the numbers of neural elements which contribute to the volume-conducted response rather than to disrupted synchronicity of

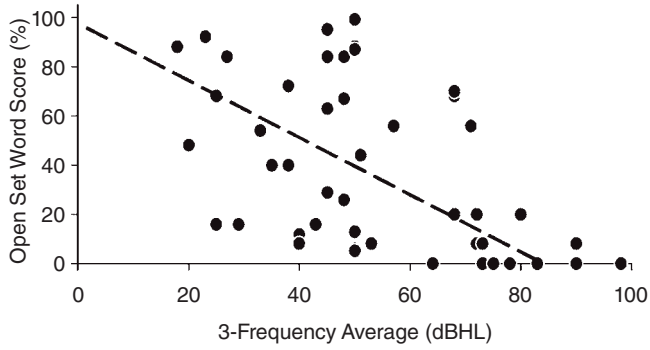

Figure 12.3 Meta-analysis of open-set word/average hearing level comparisons for children with auditory neuropathy/dys-synchrony type hearing loss ($N = 46$) from ten different studies. The dashed line represents the minimum expected score for ears with sensorineural hearing loss. Reprinted from Rance 2005, with kind permission.

the fibres.[8] It has been suggested that cases with isolated IHC damage would be expected to show behavioural and electrophysiological thresholds that are elevated by a similar degree.[18]

In all these cases with AN/AD, the afferent pathway may not be able to provide sufficiently high or sufficiently synchronised discharges to activate the motor neurons of the stapedius muscle[19] and to show suppression to ipsicontra- or bilateral stimulation by noise.[20]

Thus, AN/AD is likely to be a group of diverse disorders that result in the same clinical presentation. In addition, this group may also include cases with mixed neural and more central damage, e.g. AN/AD seen after hyperbilirubinaemia, in which case the brainstem auditory nuclei are primarily affected, as well as the cell bodies of the auditory nerve in the spiral ganglia.[21] Despite the limitations of the current tests and the ambiguities in what would constitute an AN/AD, every effort ought to be made to differentiate between 'peripheral' (up to the root entry zone of the brainstem) and more 'central' lesions (from the brainstem upwards), or for documentation of additional central damage of the auditory pathway (which can be sometimes seen on the brain MRI after hyperbilirubinaemia). When possible, it is important to make this distinction, so as to address the hearing needs of these patients appropriately and to better understand the rehabilitation outcomes in the long run.

THE AUDIOLOGICAL PICTURE

Degree and configuration of the hearing loss (HL)

Hearing thresholds may range from normal to profound hearing loss, and thresholds may fluctuate on repeat audiograms.[18,22] In babies and toddlers with AN/AD, the audiometric configuration may be flat, particularly if thresholds are normal or profoundly impaired, or the low to mid frequencies may be more affected than the high frequencies in cases with moderate to severe HL.[18] Another study on children with AN/AD aged 4–15 years found thresholds to be higher in low to mid frequencies rather than in the high frequencies in about half of the cases, high-frequency hearing loss in about a quarter of cases and a flat configuration audiogram in the remaining cases.[22]

High-rate SSEPs are poorer predictors of behavioural hearing thresholds for children with AN/AD than those with sensorineural loss. In some children with AN/AD, the SSEP may be obtained at levels below the behavioural threshold and this may indicate that the auditory pathway may produce a phase-locked response to sound at levels below those required for sound perception.[18] Similarly, AERPs may be recordable in children with AN/AD hearing loss and the presence of AERPs does not correlate with the degree of the hearing loss.[23] Some children with severe AN/AD-related hearing loss may thus have preserved AERPs, whilst others with mild or moderate hearing loss may have absent AERPs.

Preneural responses

Otoacoustic emissions may be either present in about half of babies with AN/AD; in these cases, the OAE levels are similar to normal, or absent in the remaining cases, despite the presence of a CM, and the presence or absence of emissions does not correlate with the degree of HL.[18] This finding could be due to subtle middle ear problems which may affect the recording of OAE; alternatively, the OHCs of these children may have been sufficiently damaged so as to disrupt active cochlear amplification but not to the point where the CM was abolished. A third possibility, that the CM was generated solely by the IHCs, is less likely, as the observed CM is quite robust in neonates with AN/AD.[18] OAEs may become degraded or completely abolished over time in some children with AN/AD who have not been exposed to amplified sound.[9]

Speech perception measures and cortical responses

A meta-analysis compared open-set word recognition scores for children with AN/AD versus average audiometric thresholds (at 1, 2 and 4 kHz) to normative data, and found that when ears with threshold averages exceeding 80 dB HL were excluded, the word recognition scores were within the expected range in 44% ($N = 18$) of these ears and 56% (23 in 41) ears were either borderline abnormal or significantly poorer than expected in view of the hearing thresholds (Figure 12.3).[8] An earlier paper by Rance et al.[18] reported that up to 40% of paediatric cases with AN/AD may not show measurable speech perception, either aided or unaided, and after a period of hearing aid use of more than 1 year. Speech perception may be significantly better in the aided versus the unaided condition for about half of their cases, but no different in the two conditions for the other half.[18] These results may be due to the severity of the degradation of the acoustic signal because of the AN/AD, but also to other linguistic and developmental factors, particularly in the presence of generalised neurologic abnormality which can be due to the same cause as the AN/AD.

Approximately 50% of children with AN/AD will show AERPs of normal latency, amplitude and morphology, and this finding correlates with substantial speech perception ability, whilst the absence of the AERPs correlates with extremely poor speech perception.[23] The presence of AERPs may indicate that there is sufficient residual neural synchrony to encode important timing aspects for speech perception. The dissociation in findings between ABR and AERP may be due to a ‘smearing’ of the response by the loss of temporal synchronicity, to which the ABR will be more vulnerable, as its peaks are usually only separated by approximately 1 ms. The ‘smearing’ of the response may thus cancel the averaged ABR, but not the AERPs, which have broader peaks separated by at least 50 ms and which may thus be less vulnerable to fluctuations in the timing of individual responses.[8]

Psychoacoustics

Zeng and Liu.[24] conducted a psychoacoustic study on 21 cases with AN/AD, which included four children younger than 10 years old and five children younger than 17. They reported that loudness discrimination, pitch discrimination at high frequencies and sound localisation using interaural level differences were unimpaired. In contrast, pitch discrimination at low frequencies, temporal integration, gap detection, temporal modulation detection, backward and forward masking, signal detection in noise, binaural beats and sound localisation using interaural time differences showed severe deficits.

CLINICAL PRESENTATION

Children with pre-lingual onset of auditory neuropathy/dys-synchrony are at risk of significant speech perception deficits which may lead to delays in speech and language acquisition,[8] whilst extreme difficulty in background noise seems to be a hallmark of this condition.[24] About 7% of the cases of children identified with AN/AD because of abnormal ABR early in life will develop normal hearing and language within 12 to 18 months after identification, and they may only complain of difficulties hearing speech in noise.[25] These cases may later be suspected of suffering from central auditory processing disorder (CAPD), and this highlights the need to test stapedial reflexes and ABR in suspected cases of CAPD. Some other children with AN/AD may behave as if they have severe/profound hearing loss with occasional episodes of hearing sensitivity, whilst other cases may occasionally show unexpectedly good hearing abilities.[25] Hearing fluctuations may be present in approximately one in three cases and may produce marked differences in overall functional hearing as well as in speech understanding, as reported by parents and teachers of affected children.[7,18] In addition, some children with AN/AD may also present with additional symptoms to the hearing impairment, as the underlying pathogenetic mechanism for AN/AD may have affected other systems. Thus, about 12% of AN/AD cases will develop peripheral neuropathies later in life, and in the patients reported by Starr et al.,[2] the peripheral neuropathies were diagnosed after the age of 10.

PREVALENCE

Rance et al.[18] reported a prevalence of 0.23% for AN/AD within a neonatal population whose neonatal or family histories placed them at increased risk for hearing loss, whilst the prevalence of AN/AD within the group of children with permanent hearing loss was 11.01%.

AETIOLOGY

Causes include genetic (syndromic and non-syndromic) as well as acquired (such as infections, toxic, neonatal illness or idiopathic). These may affect

(a) only the afferent auditory pathway, from the IHCs up to the brainstem;
(b) the afferent auditory as well as the afferent vestibular pathway (again from the level of the receptor cells up to the brainstem);

(c) the afferent audiovestibular pathway up to the brainstem as well as other peripheral nerves as a peripheral neuropathy syndrome (in which case vestibular nerve involvement may be more common);
(d) finally, there may be additional damage at different levels of the auditory pathway. This may affect the outer hair cells, e.g. in some paediatric cases who only have CM but no OAEs present (despite normal middle-ear function), or who lose OAEs over time, or higher levels of the central auditory pathway, e.g. in the case of kernicterus (see further discussions).

GENETIC FORMS OF AN/AD

Non-syndromic disorders

AN/AD may be an isolated finding in some families. The gene *otoferlin* encodes a protein at the base of the IHCs that is thought to be involved in synaptic vesicle recycling. Varga et al.[15] reported nine affected children with a clinical diagnosis of AN/AD from four families with mutations in the *otoferlin* gene. The hearing levels ranged from moderate to severe and from severe to profound, speech discrimination was worse than expected for the audiometric thresholds and vestibular function (on the basis of the rotation test) was normal. None of these children had evidence of a peripheral neuropathy. There was evidence that the OAEs deteriorated with age in one of these families. Varga et al.[26] also reported that an *otoferlin* gene allele may be responsible for the temperature-sensitive auditory neuropathy phenotype. In two siblings with AN/AD and a mutation in the *otoferlin* gene, one reported that her mild low-frequency hearing loss when she did not have a fever deteriorated to profound hearing loss in the low frequencies, and to severe hearing loss in the high frequencies when her temperature was at 38.1°C. When her temperature was at 37.8°C, she showed a mild to moderate hearing loss whilst her auditory function returned to baseline after the fever subsided. The child had reported to her parents that her hearing became worse when she had a fever, whilst her brother was similarly affected.

Kim et al.[27] similarly mapped a gene responsible for autosomal dominant auditory neuropathy in a US family of European descent to a locus on 13q14-q21. Hearing loss had an average age of onset of 18.6 years; however, it was lower (8 and 9 years) for two homozygous individuals of consanguineous parents. However, with the exception of an age of onset at the lower end of the range, there were no apparent clinical features differentiating their phenotype from that of the heterozygotes.

Delmaghani et al.[28] investigated two multigeneration consanguineous families from two different regions of Iran, with autosomal recessive, non-syndromic, bilateral, pre-lingual sensorineural hearing impairment, which met the clinical criteria for AN/AD. The hearing loss was profound in one and severe in the other family. They identified mutations in a new gene, the *DFNB59* gene, in both these families. The *DFNB59* gene encodes pejvakin, which is expressed in all the relays of the afferent auditory pathway from the cochlea to the midbrain.

Syndromic presentations

Mutations in genes that encode gap junction channels, i.e. channels between neighbouring cells that permit the rapid exchange of certain molecules, may be responsible for peripheral neu-

ropathy as well as auditory neuropathy resulting in hearing loss. These include connexin 31, which has been associated with AN/AD in adulthood,[29] and connexin 32 responsible for the X-linked form of Charcot-Marie-Tooth (CMT).[30] Abnormalities of peripheral protein 22 on chromosome 17p11.2 are responsible for type I Charcot-Marie-Tooth disease,[31] where the primary histopathological lesion is loss of cochlear spiral ganglion cells and hypertrophic changes in nerves 7 and 8.[32] Audiological evaluation has identified features of both auditory neuropathy and cochlear involvement in affected individuals, with increasing clinical severity and younger age of onset of CMT and hearing loss in each progressive generation.[31] Hereditary motor and sensory neuropathy, Lom type,[33] is an autosomal recessive form of CMT, with peripheral neuropathy, auditory neuropathy related hearing loss and absent vestibular caloric responses.[34] There are reduced numbers and size of myelinated fibres and the responsible gene, *NDRG1*, may have a role in the peripheral nervous system, possibly in the Schwann cell signalling necessary for axonal survival.[35] ABRs were found to be abnormal in children with hereditary motor and sensory neuropathy-type Lom.[36]

Wang et al.[37] studied an extended five-generation Chinese family with type I auditory neuropathy involving primary degeneration of the auditory nerve (i.e. axons) accompanied by late-onset peripheral neuropathy. The pattern of inheritance, as indicated by the pedigree, was X-linked recessive. Most affected patients had suffered from hearing loss and speech discrimination difficulties from the age of 10 to 16 years. Neurological examination revealed diffuse peripheral sensory neuropathy later in life. A novel X-linked auditory neuropathy locus/region (AUNX1, Xq23-q27.3) was identified.

The heterozygous R445H mutation in OPA1 was identified in five patients with progressive autosomal dominant optic atrophy and progressive hearing loss consistent with AN/AD. The hearing loss first presented at the ages of 6 and 9 years in two of these patients, and at the age of 17 in another.[38]

Auditory and vestibular neuropathy may also be associated with peripheral neuropathy as well as with cerebellar abnormalities in olivopontocerebellar and spinocerebellar degeneration and in Friedreich's ataxia. Friedreich's ataxia is characterised by severe loss of cochlear and to lesser extent vestibular neurons, but with intact sensory epithelia.[39]

Koskinen et al.[40] described early-onset spinocerebellar ataxia between 1 and 2 years of age in 19 previously healthy Finnish infants, who first presented with clumsiness and loss of ability to walk, with clinical findings of ataxia, athetosis, muscle hypotonia and loss of deep tendon reflexes, whilst hearing loss and ophthalmoplegia were diagnosed by school age, and sensory neuropathy by adolescence. The main finding on brain MRI was cerebellar atrophy.

Cerebro-oculofacio-skeletal syndrome is a rare autosomal recessive disorder of childhood with dysmorphic features, hypotonia, osteoporosis and peripheral neural degeneration, which shows accelerated cochlear and to a lesser degree vestibular nerve degeneration that resembles the histopathology findings in Friedreich's ataxia.[41] Apart from cochlear abnormalities, primary 8th nerve degeneration is also present in Usher syndrome, i.e. recessively inherited deafness and retinitis pigmentosa.[42] There is also some evidence to suggest auditory neuropathy in some children with mitochondrial disorders.[43]

Acquired AN/AD

Kernicterus is a prime example of a clinical condition which is due to pathology both in the auditory nerve (as indicated by an absent wave in the ABR) as well as in the brainstem, as bilirubin predominantly affects the brainstem nuclei. It has been proposed that kernicterus is

defined in term and near-term infants when the total serum bilirubin exceeds 20 mg/dl and when there are clinical findings of abnormal muscle tone on examination, and/or an audiological diagnosis of AN/AD, and/or an MRI shows bilateral lesions of the globus pallidus and/or the subthalamic nucleus.[21]

Abnormal ABRs may improve or even become normal with exchange transfusion. Other causes of acquired AN/AD may include infection, such as meningitis (Rance et al.[18]), hydrocephalus, hypoxia, prematurity.[17,18] AN/AD may also be caused by drugs such as carboplatin, which primarily destroys IHCs,[44] and trauma.

DIFFERENTIAL DIAGNOSIS

AN/AD may need to be differentially diagnosed from the congenital absence or hypoplasia of the 8th nerve[11] as well as from lesions that compress or infiltrate the vestibulocochlear nerve. A brain MRI should be included in the diagnostic investigations of AN/AD. Osteopetrosis,[45] craniodiaphyseal dysplasia[46] and other similar conditions may cause a progressive osseous lesion of the internal auditory canal which may compress the 8th nerve and give symptoms/signs from all its branches. Vestibular schwannomas (VS) are benign, encapsulated tumours with a nodular surface, which arise from the Schwann cells of the vestibular nerve, and initially tend to compress rather than invade the nerve fibres. VS may be due to a genetic disorder, such as neurofibromatosis 1 (von Recklinghausen disease – NF1) and neurofibromatosis 2 (NF2) which are clinically and genetically distinct disorders. Other lesions which may compress the 8th nerve and lead to a clinical presentation similar to AN/AD may include meningiomas, haemangiomas, granulomas and hamartomas.

Management strategies

AN/AD includes a range of disorders, and the management approach ought to be individualised. In some cases, appropriate medical management of the causative disease will be required, e.g. exchange transfusion for hyperbilirubinaemia, which may lead to normalisation of the abnormal ABRs.[21]

At present, there is no consensus regarding amplification in children with AN/AD. Sophisticated hearing aids may benefit some cases.[47] Similarly, Rance et al. reported that about half of the children with AN/AD may develop significant open-set speech perception after the provision of hearing aids.[23] There is no correlation between aided speech perception ability and the results of clinical tests, with the exception of AERPs, which may predict speech perception.[23] Assessment of unaided speech perception 'performance-intensity' functions may be useful, as the presence of 'rollover' may be a contraindication for amplification.[47] Conversely, the presence of OAEs in AN/AD may not necessarily be a contraindication for the provision of a high-gain hearing aid, as the OHCs may not contribute to hearing ability, since neither pure-tone sensitivity nor speech perception correlates with the presence of OAEs.[9,18] In addition, OAEs may become degraded or completely abolished over time in some children with AN/AD who have not been exposed to amplified sound.[9] Zheng and Liu[24] propose that amplitude compression should be avoided, as it will reduce the amount of temporal modulation in the acoustic signals and that linear amplification should be considered instead. They have also proposed several signal-processing strategies which may hold promise for successful amplification of AN/AD, such as expansion of temporal modulation and filtering out of low-frequency

signals and/or shifting these to high-frequency regions. Early amplification may be important, and may be responsible for the better results with amplification in paediatric versus adult cases with AN/AD, although the underlying pathophysiology may also play a role for the observed differences.[8]

There are also reports of improved listening and communication skills, and restoration of the desynchronous ABR following cochlear implantation in some, but not all, cases with AN/AD.[47] In some implanted cases, speech perception in noise is significantly better in the implanted versus the non-implanted ear in the same individuals with AN/AD. It has been argued that in cases of AN/AD, electrical stimulation may lead to better neural synchrony in the auditory pathway than acoustic stimulation. Thus, if the AN/AD-related pathology affects the IHC or the synapse, electrical stimulation by the cochlear implant will stimulate the spiral ganglion cells directly.[18] Cochlear implantation may be very successful in these cases, and Gibson and Sanli[48] reported that children with AN/AD and with an early positive summating potential on round window ECochGs who had normal electrical ABR, and in whom the site of lesion was more likely to be pre-neural, performed significantly better after cochlear implantation than a control group of children with sensorineural hearing loss who had absent OAEs but appropriate ABR results. In cases with AN/AD due to neural pathology with myelin deficiency, electrical stimulation of the 8th nerve may generate and propagate action potentials, albeit at an increased threshold and with decreased dynamic range.[49] There are reports of successful restoration of electrically evoked ABR after cochlear implantation in patients with AN/AD.[50] However, in the case of progressive neural pathology, the neural activity generated via electrical stimulation may decrease over time due to conduction block, and these patients may at one point no longer benefit from their cochlear implant.[18] Similarly, cochlear implantation is less likely to be effective if there is extensive loss of auditory neurons.[13]

Additional signal enhancement techniques, such as clear speech may be of help for cases with AN/AD. Clear speech leads to significantly improved speech perception than for conversational speech, both in quiet and in background noise, in children and adults with AN/AD.[24] This is because talkers who are instructed to speak clearly produce more intelligible speech than in normal conversation, and this reflects both acoustic and phonetic differences between the two styles of speech, e.g. reduced speech rate, improved temporal modulations, lengthened vowel space. In addition, auditory training and other strategies for management of the functional deficit[47] may also be beneficial. Finally, in some cases, a conservative approach may be more appropriate. Berlin et al.[25] propose that children with AN/AD should first be exposed to sign language, coupled with observation of the child, whilst hearing aids may make these children more aware of environmental sounds. However, others are proponents of a more aggressive approach, e.g. with cochlear implantation, and there is no consensus at present about which approach is indicated for which case.

CONCLUSION

Auditory neuropathy/dys-synchrony is an umbrella term, in that it encompasses a range of pathologies which may affect different levels of the auditory system. Identification of AN/AD can be challenging and may require a multiple test battery and a high index of clinical suspicion. However, making a diagnosis of AN/AD may be of paramount importance, as it may help identify the underlying cause, which may require treatment, may help clarify the genetics of the disease and may enable the most appropriate rehabilitation intervention. At present, the

rehabilitation plan for children with AN/AD ought to be made on an individual basis and every effort ought to be made to ascertain the site of the lesion. It is to be hoped that in the near future more site-specific tests will emerge, and this will in turn help inform better the management decisions.

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13 Progressive hearing loss

D. Lucas

INTRODUCTION

After the initial shock of confirmation or identification of their child's hearing loss, parents tend to ask three questions: (1) 'When will he talk?', (2) 'Why me?', (3) 'Will it get worse?' This last question is almost impossible to answer unless the diagnosis (aetiology) has been accurately determined. Professionals may be as shocked as parents to discover that the hearing loss is progressing, especially where research and experience have previously suggested that the hearing is likely to remain stable. With the advent of newborn hearing screening, parents may have little opportunity to enjoy their baby, adjust to the deafness and accustom themselves to recognising how their baby responds to sound, before addressing the uncertainties posed by yet more unwelcome information. Although it is clearly wrong to assume that the hearing will not change, confirmation or even suspicion of change presents both parties with significant problems.

For families, the resolution of grief following the confirmation of deafness may become impossible. They and their child dread each visit to the audiology department, anticipating further bad news. They must constantly adjust to the changes in the ability to communicate and use residual hearing, to the type of hearing aids provided, to participation in school and to the eventual future in society. They may have to contemplate a decision about cochlear implants, often struggling with the delicate balance between reduced access to sound and the criteria for implant, which emphasise measurement of hearing thresholds and speech perception more than functional hearing and participation. They are in a state of constant uncertainty, enhanced by a sense of powerlessness, which may fuel anger directed towards the professionals responsible for the child's care, towards each other, towards other family members or towards previous professionals whom they may blame for the lack of identification or effective treatment. Young children are remarkably stoical about changes in their disease or disorder; older children, however, may become extremely distressed, tearful and depressed as they struggle to cope with the change in their ability to participate.

Although recognising the needs of child and family to be paramount, thought must be given to the dilemmas facing professionals. Gauging how to share information with families is professionally challenging. Early progression may not be identified with confidence; late presentation or identification of hearing loss raises both doubts and assumptions about the onset and natural history. Frequently, professionals will question the validity of their previous findings and must, therefore, deal with their own uncertainties, especially if the diagnosis is not clear and only retrospectively is there thought to have been the possibility of active and potentially successful intervention.

PRESENTATION

Progression may be detected on routine review which may be the only way to identify early progression in those infants whose deafness was detected as newborns. Follow-up continues through an age at which accurate diagnostic testing requires great expertise, experience, time and often repeated visits. Deterioration in behaviour may be the first sign of hearing loss progression in young children, with the child becoming naughty or disruptive, persistently tired or aggressive, increasingly demanding or withdrawn, rejecting hearing aids even where use has been consistent, refusing even to use sign support. Later in childhood, the child, a parent or teacher may remark upon poorer responses to sound or instructions. Older children may complain of reduced speech perception, a complaint that must be taken seriously, even in the presence of an unchanged pure tone audiogram. They may become reluctant to attend school or socialise or have apparently irrational fears, for example of shopping or the dark. The clarity of speech may deteriorate or speech and language not develop as expected; conversely, speech or voice quality may be better than anticipated in a child who presents with severe to profound hearing loss in early childhood. Occasionally, the first symptoms may be of the underlying disease. Only rarely is there a clear history of a precipitating event.

Tinnitus, vertigo, headache or noise intolerance may be directly related to progression or fluctuation. Some children describe, if asked, how tinnitus interferes with their sound perception on testing; it is a source of some dismay that there are still times when children languish with unacknowledged progressive hearing loss because professionals do not believe the changes or variability found on behavioural testing.

PREVALENCE

Given that the prevalence of permanent childhood hearing impairment approximately doubles from one per thousand at birth to two per thousand by the age of 9 years,[1] it is no longer surprising to find permanent hearing loss in children who passed newborn screening. Accurate information on the proportion of children who have progressive (and/or fluctuating) hearing loss is difficult to obtain and relates almost entirely to sensorineural hearing loss (SNHL). There is no universally accepted definition of progression, which is reported to occur in 2% to 32% of children depending on the criteria used (10, 15, 20 dB HL at one or more frequencies measured on more than one occasion) and the population studied. Various researchers have surveyed general or selected populations, seeking to link progression to aetiology, age and audiometric configuration: most studies are retrospective, often including children who were not identified to be deaf until after 18 months of age; so early information about hearing thresholds is missing and conclusions are speculative. Our practice is to look carefully at children whose hearing changes by 10 dB at more than one frequency or 15 db at one frequency, measured without middle ear dysfunction, on more than one occasion (see Figure 13.1).

Studies using 10 dB progression have reported 6% of 365 ears and 32% of 106 children to have progressive losses; of the 365 ears, a further 57% fluctuated with gradual progression.[2,3] Nine per cent of 177 children were thought to be an underestimate in one study using 15 dB as the criterion,[4] whereas another the same year found progression in 2% to 4% of a cohort of 138 children but as many as 16% based on a 10 dB difference.[5] The stricter measure of 20 dB at two or more frequencies yielded 6.2% of 178 children.[6] A retrospective review of 92 children

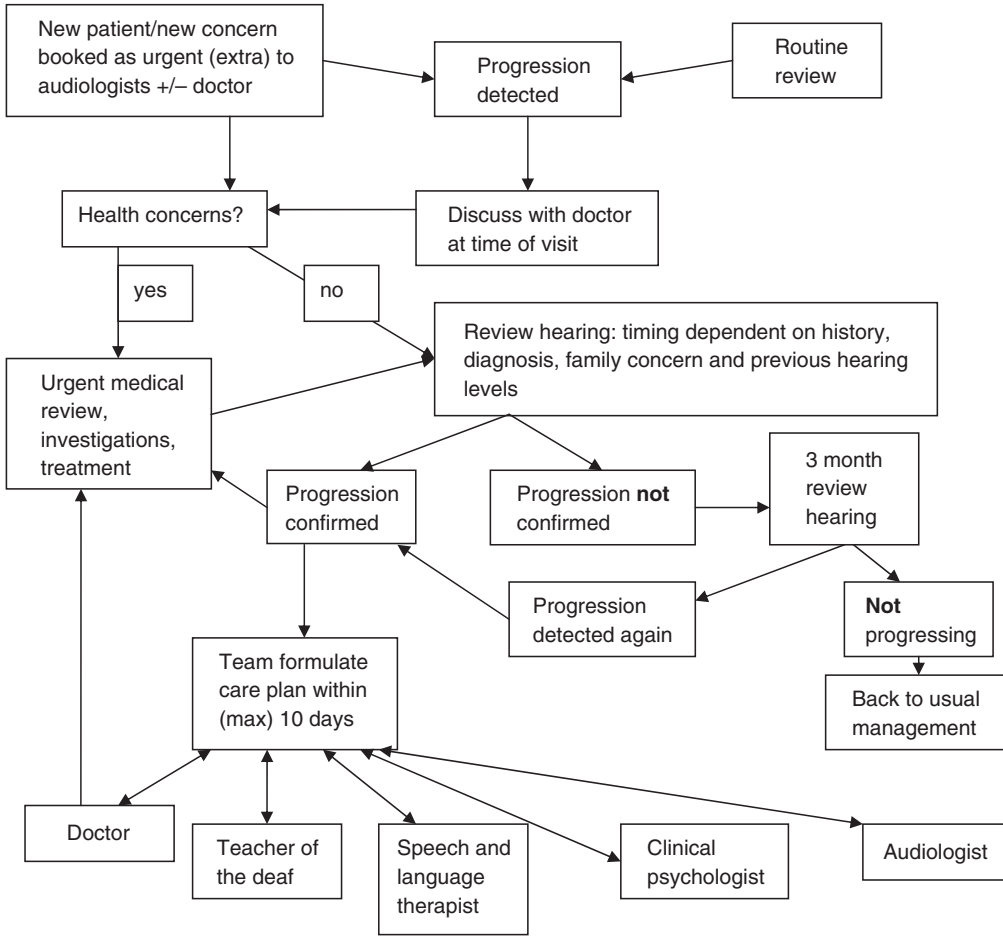

Figure 13.1 Care plan for management of progressive SNHL.

over 15 years found 22.8% with progressive loss, all of whom had a mild hearing loss when first identified.[7] Another retrospective published 10 years later found 11% to be progressive or acquired.[8]

Progression is found to be faster in children under 5 or 6 years of age,[4,9] especially between 1 and 2 years of age.[3] Progression between 2 and 4 years of age was confirmed in 60% of those survivors of neonatal intensive care who were deaf at 2 years.[10] A carefully structured prospective study looked at 688 children who had a change in hearing of more than 20 dB at any frequency on pure tone audiometry: 43.9% became progressive in the early years, only 5.7% of progression occurred after 4 years of age and the frequency of detected progression increased the longer the observation period.[11] If data from before the children's 6th birthdays had been available, the hearing loss of more than just 13% of 132 children might have been shown to progress[12] (see Table 13.1).

Deterioration is described across all frequencies equally although high frequencies may deteriorate first or be most vulnerable.[11] Experience suggests that further loss of hearing is

Table 13.1 Commonest ages of onset of progressive hearing loss in children.

Age band	Aetiology
0–5 years	Autosomal recessive
	X-linked
	Jervell and Lange-Nielsen syndrome
	Perinatal events
	Congenital cytomegalovirus
	Congenital rubella
	Mucopolysaccharidoses
5–10 years	Autosomal dominant
	Osteogenesis imperfecta
	Alport syndrome
	Alström syndrome
	Marshall syndrome
	Noonan syndrome
10–20 years	Otosclerosis
	Usher Type 3
	Mitochondrial
	Down syndrome
	Turner syndrome
	Norrie syndrome
	Congenital syphilis
	Autoimmune
	Noise
Any age	Bacterial meningitis
	Ototoxic drugs
	Widened vestibular aqueducts
	Tumours
	Trauma

likely to occur where there is an 'island' of much better hearing remaining: deterioration is greatest in the better hearing ear in asymmetrical hearing loss,[4] the least affected frequencies tend to deteriorate the most,[6] and an unusual configuration of the hearing loss seems linked with progression.[12] Progression tends to be gradual rather than sudden, although a combination of progressive and fluctuating is possibly more common.[2] There does not seem to be any sex difference.

Prevalence is likely to be changing with immunisation, neonatal detection, improved diagnostic acumen and facilities, advances in genetics, family size limitation, population demographics and treatment.

ASSESSMENT: HISTORY AND EXAMINATION

Professionals still have a tendency to describe as 'congenital' all severe to profound hearing losses with onset before 2 years of age. In areas where identification of hearing loss remains

late or where infants have passed the newborn hearing screening and have no recognised risk factors directing review, the crucial information needed to enable identification of early progressive hearing loss will be missed. Parents are generally correct in their observations of their child's response to sound and perplexed by the changes they have observed, only to be dismissed by professionals at a time which is already very distressing for them. A careful history will include asking for the family's story and observations over time, the perinatal and developmental history with particular reference to motor skills and vision, family history (including a sensitive enquiry about consanguinity), use of hearing aids and satisfaction with them, methods of communication and any change therein and, if not a new event, reflections back on the original discovery of the hearing loss. Associated features (e.g. tinnitus, vertigo, headache, fever, rash) and events (e.g. head trauma, illness, red eye) may provide keys to diagnosis. Enquiries into general physical health, growth, sleep and feeding are often overlooked by non-paediatricians but give essential information about a child's well-being. Where possible, a detailed history should be taken from the child first, using sign support where necessary. Children will often give a very clear description of their symptoms, uncontaminated by the fear and worry that frequently clouds the comments of their parents. It is not unknown for them to have omitted to inform their parents of change in their hearing acuity until after the clinic has identified the further loss.

Clinical examination is mandatory. It is interesting to reflect on how often children with significant SNHL have their hearing accurately measured but no steps are taken to identify other clinically relevant signs: examination of the ears, nose and throat is frequently all that is considered necessary. Basic paediatric parameters include height, weight and head circumference, all of which can be plotted on age-related charts. Much more than this, however, is relevant to the clinical examination of any patient with an unexplained worsening condition (Table 13.2). No list can be comprehensive; much of the examination will depend on the carefully elicited history.

Table 13.2 Clinical examination of child presenting with progressive hearing loss.

Clinical part	Looking for
General inspection of face	Unusual features, setting and asymmetary
External ear anomalies	Setting, shape, pre-auricular pits/sinuses and epidermoids
Eyes	Setting, shape, sinuses, colour, cataract, coloboma and retinal examination
Palate	Cleft
Teeth	Shape, number and stage of dentition
Neck and spine	Branchial pits and sinuses, goitre, webbing, head tilt, scoliosis and kyphosis
Skin	Lentigines, café au lait spots, freckling in the axillae, haemangiomata and dryness
Cardiovascular	Heart, pulse and blood pressure
Hands and feet	Polydactyly, creases, webbing, other digital anomalies and nails
Hair (including eyebrows)	Colour, texture and distribution
Neurology	General neurological examination including cranial nerves and peripheral reflexes
Neurotology	Gait, balance and eye movements
First degree relatives	General physiognomy

INVESTIGATIONS

Further investigation will be dictated by the findings on anamnesis and examination and by what may have been undertaken for an already identified hearing loss. The basic investigations of imaging, ophthalmic examination, connexin 26 and a hearing test on first-degree relatives must be offered to the family if not already undertaken.

Magnetic resonance imaging (MRI) is probably the single most useful examination in the assessment of SNHL. It gives excellent imaging of the petrous bones, the cochleae and vestibular apparatus, the size and shape of the nerves and whether they enter the cochlea; sagittal oblique imaging may be particularly useful in progressive SNHL. Where the loss is suspected to be conductive, computed tomography (CT) remains the radiological investigation of choice, with axial and coronal scans being more useful for precision, although spiral CT (with reformatting) is a much faster examination for a younger child. If CT is the only imaging available, it will be essential to have 1.2-mm cuts if the widened vestibular aqueduct is to be demonstrated. Densitometry will clarify the extent of otosclerosis or cochlear otospongiosis.

As up to 64% of children who are deaf have some sort of eye or visual defect, all children should be examined by an ophthalmologist who is familiar with the visual and ophthalmic implications of childhood progressive hearing impairment.[13] Any previous findings should be reviewed if hearing changes significantly.

Blood tests will be dictated by knowledge and clinical findings. Young children identified before immunisation or exposure should have rubella and cytomegalovirus (CMV) antibodies, with toxoplasmosis, syphilis and CMV prompted by later onset. Late onset (rapid) progressive loss, especially with general health concerns or findings, will dictate investigations for autoimmune disorders (see below). Anaemia has been linked with progressive hearing loss[8] as have various haemoglobinopathies. The discovery of a goitre indicates thyroid function tests, including thyroid antibodies. Other blood biochemistry for renal or metabolic function will be determined by history and examination.

DNA investigations must include not only connexins 26 and 30 but also a search for mitochondrial anomalies of which m.1555A>G is arguably the most relevant in a well patient with no specific history. Routine screening for SCL26A4 (Pendrin gene) has also been suggested.[14] Chromosomal analysis may be relevant in dysmorphic children.

A search for cells, protein and blood in the urine is mandatory. The presence of renal anomalies is demonstrated simply by renal ultrasound and is essential in children with preauricular or branchial pits or sinuses. A urinary metabolic screen is of debatable value unless dictated by clinical suspicion.

Many young children will tolerate detailed neurovestibular testing by a knowledgeable clinician with appropriate tools in child-friendly surroundings.

AETIOLOGY

A number of conditions are now widely recognised to be associated with progression or fluctuation of SNHL, which may be the presenting feature. Early identification of hearing loss through newborn hearing screening has enabled much earlier diagnosis (where parents are willing), but there is still a dearth of accurate prospective population studies from this cohort. There is now improved recognition that ‘adult’ disease may present in childhood. Many studies

predate the recognition of the features and onset of a variety of genetically and adventitiously determined progressive hearing losses.

Genetic causes are reported to be the most common[11] especially in the congenitally deaf: late-onset progressive SNHL remains a particular feature of dominant inheritance and early onset of recessive inheritance. One study suggested that only 14% were genetic, although another 7% had a family history.[2] In another, with all recognised syndromal hearing losses excluded, five out of eleven patients were found to have an inherited hearing loss, one was due to CMV, one due to an anatomical abnormality (widened vestibular aqueducts which we now know to be largely genetically determined) and four were of unknown aetiology.[6] A number of authors from 20 and more years ago pointed out the frequent finding of progression in association with intrauterine infection with CMV and rubella.[4] Others did not find progression to be linked with any specific aetiology[7] or excluded all known causes to give an idiopathic group, the only identified possible correlation being a mild iron deficiency anaemia in those less than 6 years old[9]; iron deficiency is known to occur in up to a quarter of this age group.

The definition and aetiology of the auditory neuropathies are in their infancy, especially in association with progressive hearing loss. The combination of the presence of otoacoustic emissions (OAE) or cochlear microphonic with abnormal or absent auditory brainstem response (ABR) implies a multitude of pathological processes in the intervening microstructure. Some will be due to hypoplasia of the cochlear nerve which may coexist with other structural anomalies. Reports suggest that late onset and progressive change in hearing can occur with or without peripheral neuropathies and in combination with progressive cochlear loss, that OAE may subsequently disappear and that the progressive hearing loss associated with other neuropathies (e.g. mitochondrial cytopathy, Brown-Vialetto-van Laere syndrome) may be at least partly due to auditory neuropathy.

CONFOUNDING ISSUES

General considerations

Otitis media with effusion is as common in the child with permanent hearing loss as in the general population; glue ear may add 40 dB to the hearing threshold. Increased demands on language may be confused with deterioration of hearing. Anxious children may add another variable, that of spurious (non-organic) overlay. Behavioural and electrophysiological tests are not directly comparable, and tests in very young children can be difficult to interpret and reproduce, especially in those with additional needs. Even ABR testing can be misleading and requires careful knowledge and experience. The recognition that small or tortuous canals or soft pinnae may collapse with headphones has led to more reliable testing with insert earphones. A child's skill and reliability at testing develops over time and sometimes only after the consistent use of hearing aids. Poorer performance may be due to malfunction of the hearing aids. More powerful hearing aids may be needed to deliver the same gain as the ear grows and canal resonance changes. Much will depend on the expertise and knowledge of the tester; consistency of personnel may be particularly important in very young or handicapped children.

Perinatal events

It has long been known that there is a ten-fold risk of hearing loss in those who have been in neonatal intensive care although whether there is a direct cause and effect is debatable.

Survivors of severe neonatal respiratory distress may develop late-onset progressive SNHL, onset after 2 years of age being likely to result in a less severe predominantly high-frequency SNHL.[10] Extracorporeal membrane oxygenation (ECMO), a technique used only in very sick neonates with severe cardiorespiratory failure, has been implicated in SNHL, but more recent work suggests that it is the underlying morbidity which is important, not the technique.[15] Parker (personal communication) found that intermittent positive pressure ventilation of more than 5 days duration and the administration of ototoxic antibiotics were positively correlated with subsequent progressive SNHL in infants from neonatal intensive care units who had passed the hearing screen. A combination of (loop) diuretic use for more than 14 days and a neuromuscular blocker drug dose of more than 0.96 mg/kg was found to be associated with the late-onset progressive hearing loss identified in 53% of 81 survivors of neonatal intensive care. Many of these infants had also received other drugs including aminoglycosides and vancomicin but this group did not find evidence that these or illness severity were contributory.[16] Hyperbilirubinaemia is known to be associated with SNHL: there are reports of improvement over time in the auditory neuropathy which characterises this pathology although OAE may no longer be recordable.

The lack of a family history may be misleading, Some of these children may subsequently be found to have the m.1555A>G mutation in which hearing loss may rapidly occur after as little as one dose of aminoglycoside. Some will have reasons for their deafness which are the same as those of their peers born at term, others may have hearing loss associated with whatever underlying condition resulted in perinatal disadvantage.

GENETIC NON-SYNDROMAL PROGRESSIVE HEARING LOSS

Autosomal dominant sensorineural

Up to 25% of congenital profound sensorineural hearing loss may be dominantly inherited. The majority are non-syndromal and, therefore, without any warning signs or features; hence it is important to carefully review those children identified to be at risk through an accurate history of the onset of the hearing loss in the adult members of the family. Very few dominantly inherited non-syndromal hearing losses are accompanied by vestibular hypofunction.

Variability of expressivity is characteristic of dominantly inherited disorders, so it is unsurprising to find that, although the shape of the audiogram may be similar between family members, the level of hearing loss and speed of change can be variable. Almost every configuration of audiometric pattern has been described. Although most dominantly inherited progressive losses are said to commence post-lingually in the high frequencies, the stigma of deafness and the relative paucity of facilities may have prevented earlier identification in older family members.

In the past, discrimination between these hearing losses was made primarily by the time of onset and pattern of progression. Nowadays, however, it is the identification of different gene loci that enables a more accurate classification, although this is often confined to specific families who have agreed to participate in a study. A number of different linkages have been demonstrated on a variety of chromosomes with increasing frequency over the past few years and mouse models are now available to enable further study (see Table 13.3). Excitingly, many overlaps with recessively inherited conditions or other disorders are being identified. Apart from research programmes, there is no facility for routine genetic investigation of individuals who present in the clinic.

Table 13.3 Some identified chromosome linkages in dominantly inherited non-syndromal progressive sensorineural hearing loss.

Linkage	Chromosome	Onset	Progression
DFNA1	5q31	Low-frequency 1st decade	Profound 4th decade (Monge's deafness)[17]
DFNA2 DFNA2/12	1p32	High-frequency early ?congenital	Mid- and high-frequency 1st–5th decade[18,19,20] Profound 4th decade[21]
DFNA3	13q12	Moderate/severe pre-lingual	Severe/profound[22]
DFNA4	19q13	2nd decade	Profound by 5th decade[23]
DFNA5	7p15	High-frequency early childhood	Low-frequency 4th–5th decade[24]
DFNA6 DFNA6/14	4p16.3	Low-frequency pre-lingual/2nd decade	Slow progression[25] Gradually other frequency[26]
DFNA7	1q21-23	High-frequency pre-lingual	Moderate (not profound) in 2nd decade[27]
DFNA9	14q12-q13	With vestibular disturbance	Profound in 5th decade[28]
DFNA10	6q23	Sloping high-frequency 1st decade	Flat, moderate stable from middle age[29]
DFNA11	11q	? high-freq sl worse 1st decade	Bil moderate/profound[30]
DFNA13	6p	Mid- and high-frequency 2nd–4th decades	To include low frequencies 6th decade[31,32]
DFNA14	4p16.3	Low frequency 1st decade	Becoming flat in 4th decade?[33]
DFNA16	2q23-24	Rapidly progressive/fluctuating 1st decade	? steroid responsive[34]
DFNA17	22q11.2	Mild high-frequency 1st decade	Moderate/severe 3rd decade[35]
DFNA 20/26		Sloping 1st–2nd decades[36]	
DFNA 21	6p21-22	Mid frequency 1st, usually 3rd–5th decade[37]	
DFNA24	4q35-qter	Moderate high frequency birth	Moderate to profound throughout life[38]
DFNA25	12q21-24	High-frequency by 2nd decade (maternal)	Slowly progressive[39]
DFNA36	9q13-21	High-frequency 1st decade	Rapid to profound by 2nd decade[40]
DFNA44	3q28-29	Moderate low and mid-frequency 1st decade	Flat profound 6th decade[41]

Autosomal dominant conductive: otosclerosis and otospongiosis

The typical history of otosclerosis is of onset of progressive conductive hearing loss in the middle years of life, hastened by pregnancy. Onset in childhood is said to be rare but may occur more commonly than is recognised, the disease being reported in 5-year-olds. Some children present with a sensorineural or mixed hearing loss, CT scan demonstrating cochlear otospongiosis. There is debate, however, as to whether cochlear otospongiosis and the progressive anterior stapediovestibular joint fixation of otosclerosis are the same condition. Various pharmaceutical treatments have been tried in adults but without sustained benefit and limited by side effects. There are risks of progression attached to surgical intervention, although results

are encouraging with 90% maintaining an air–bone gap of less than 20 dB over several years. Cochlear implant is an option for those with cochlear otosclerosis but is unlikely to be an issue for children.

Autosomal recessive sensorineural

There are a number of non-syndromal recessive progressive sensorineural hearing losses described, but ascertainment is poor due to the wide clinical heterogeneity in these families. There seem to be two patterns: one of early onset with progression to profound hearing loss by 5 years of age, the other predominantly high frequency. A number of apparently non-syndromal recessive hearing losses turn out to be syndromal when carefully and fully investigated or with the passage of time.

Connexin 26

Mutations in the GJB2 gene (and less commonly GJB6) are now known to account for up to 50% of congenital hearing loss in some populations. It is widely thought that the hearing loss, of whatever degree or configuration, is stable. Recent publications have reported non-penetrance at birth or well-documented change in hearing thresholds, not associated with any particular mutation or any precipitating factor.[42,43] A variety of heterozygous mutations in *GJB2* have been reported in 23% of patients with apparently idiopathic progressive SNHL, compared with only 5% of controls without hearing loss.[44]

On current evidence, progression in connexin 26 deafness is rare but it must be possible that there are other alleles influencing the phenotype or that this is actually a progressive loss which is nearly always of prenatal onset.

X-linked deafness

A number of X-linked non-syndromal hearing losses have been described starting in infancy or early childhood, often with high-frequency loss. The most widely recognised association is that of progressive mixed hearing loss with perilymphatic gusher. The hearing loss becomes evident in the first year of life and is accompanied by vestibular hypofunction. Imaging may show the typical findings of a bulbous internal acoustic meatus with a deficiency in the bone between the lateral end of the meatus and the basal turn of the cochlea.

Mitochondrial deafness

Mitochondrial DNA encodes the mRNA necessary for effective cell metabolism and is transmitted maternally with the exception of Kearns-Sayre syndrome (KSS) which usually occurs sporadically. The hearing loss starts post-lingually and is generally bilateral, symmetrical and progressive, the high frequencies being affected first. Mitochondrial disorders may be syndromal or non-syndromal and are more common than previously thought. Mitochondrial DNA mutations were found in 30% of 10 subjects with maternally transmitted congenital or childhood onset hearing loss.[45] In a random group of patients with child or early adult onset of non-syndromal post-lingual progressive hearing loss, recognised mitochondrial mutations were found in 7.5% of the UK and 4.2% of the Italian patients, although just 10% had a family history.[46]

Despite the commonly held belief that mitochondrial cytopathies are diseases of adults, progressive SNHL often presents in the second decade of life and sometimes earlier. Forty-two per cent (12 of 29) of children with known mitochondrial cytopathies were found to have SNHL, mostly symmetrical and possibly progressive, cochlear and retrocochlear.[47] The SNHL commences in the high frequencies in 30% to 50% and may be the only early manifestation of the disease, e.g. KSS (progressive ophthalmoplegia and retinitis pigmentosa, structural mitochondrial rearrangements), MERRF (myoclonic epilepsy, ragged red fibres, ataxia, dementia, optic atrophy, m.8344A>G and m.8356A>G), NEPPK (familial non-epidermolytic palmoplantar keratoderma, m.7445A>G). Some cytopathies include vestibular disturbance, e.g. MELAS (myopathy, encephalopathy, lactic acidosis and stroke-like episodes, m.3243A>G). Reports vary as to whether the onset of the hearing loss of MIDD (maternally inherited diabetes and deafness, short stature, m.3243A>G and others) precedes that of the diabetes mellitus. In general, progression of the SNHL correlates with neither the phenotype nor the severity of the mitochondrial disease.[47]

Onset is often at that vulnerable age when hearing loss is commonly thought to be spurious (non-organic). Diagnosis is often delayed, especially when the condition has not been recognised in the mother, or when other life events (such as leaving school) have greater importance. Drug treatments to prevent further progression of hearing loss are being investigated. Cochlear implant seems to offer significant benefit to the majority of those implanted, even when MRI scan has shown cerebral white matter abnormalities.

Particular attention is currently being paid to the m.1555A>G mutation and its association with SNHL modified by aminoglycoside antibiotics. Susceptible family members who are not known to have been exposed to aminoglycosides may have an initially mild high-frequency loss which is slowly progressive; not all become deaf, however, and the variable penetrance may relate to exposure to another environmental prompt or the presence of a modifying nuclear genetic mutation. True prevalence of this gene is difficult to determine: 1/4000 (2–5%) of the congenitally deaf in the UK may have it;[45] 1/206 random samples in New Zealand was positive;[48] 1/1161 screened neonates in the USA were positive;[49] it has been found in 4/68 postlingually deafened persons presenting for cochlear implant;[50] in Spain, 27% of families with at least two deaf persons were found to have the gene, conferring a 96.5% probability of becoming deaf by the age of 30 years if treated with aminoglycosides as opposed to 39.9% if not.[51] Aminoglycosides are used for the treatment of severe life-threatening disease or febrile neutropenias in immunocompromised patients. They are easily obtained across the counter in some countries where injectable drugs are regarded as being more efficacious and are being suggested as a pre-operative preparation (in patients who are allergic to penicillin) to prevent *Clostridium difficile* infection in the UK. This is an opportunity to use evidence to prevent deafness in individuals who may not know they are susceptible; at the least, testing could be undertaken in those whose aminoglycoside treatment is planned in advance.[52]

GENETIC SYNDROMAL PROGRESSIVE HEARING LOSS

Many of the syndromes associated with progressive hearing loss are complex multisystem disorders where initial presentation, the type of hearing loss and the rate of progression vary. Hearing loss may be the initial symptom especially if other features are mild, unrecognised or only become evident in later life. The picture is confused by uncertainties in early diagnosis

and by advances in genetics where similar mutations are found in disorders previously thought to be distinct but with similar or overlapping phenotypes. Perrault syndrome has been reported as progressive hearing loss with gonadal dysgenesis in females, but only deafness in affected males: further evidence is emerging of progressive motor and sensory neuropathy which may be a different form. Brown-Vialetto-van Laere syndrome – progressive pontobulbar palsy with progressive hearing loss and progressive vestibular dysfunction – may have a variable phenotype within one family. Progressive high-frequency loss to about 65 dB has been found in the dominantly inherited otodental syndrome. A moderate to severe progressive hearing loss from early childhood is often the presenting feature of the dominantly inherited HDR syndrome (hypoparathyroism, sensorineural deafness, renal dysplasia). Progressive SNHL starts in the second decade in X-linked adrenal hypoplasia and gonadal dysgenesis, becoming profound within 7 years.

Progressive conductive hearing loss due to stapes and malleus fixation is also described in Beckwith-Wiedemann syndrome, a rare foetal overgrowth syndrome, demonstrably inherited in some 15% of cases, characterised by macroglossia and organomegaly, post-natal hypoglycaemia, abdominal wall defects and increased incidence of Wilms and other tumours.

Chiari malformation is often reported in association with progressive hearing loss and unsteadiness. There are anecdotal reports of progressive high-frequency SNHL associated with hydrocephalus, although it is most likely that this is associated with the events leading up to the hydrocephalus.

Disorders of bone

Progressive mixed hearing loss during childhood is an almost universal feature of the craniotubular dysplasias, inherited variably in a dominant or recessive manner, which affect the temporal bone. A particular example of this mix of inner and middle ear pathology is to be found in the bone disorder Camurati-Engelmann disease where treatment with steroids has been advocated to alleviate the symptoms. Facial palsy and deafness due to cranial nerve entrapment develops in childhood in over 80% of patients with sclerosteosis. Progressive mixed loss is also reported in the chondrodysplasias. Progressive conductive hearing loss occurs in fibrodysplasia ossificans progressiva where ectopic bone forms post-natally in soft tissues.

Osteogenesis imperfecta

Families with type 1 autosomal dominantly inherited osteogenesis imperfecta (OI) (a genetic disease of connective tissue involving coding for type 1 collagen) show the typical triad of multiple fractures, blue sclerae and hearing loss, the latter said to commence most commonly at the end of the first decade. The hearing loss was previously thought to be invariably conductive due to stapes fixation, fracture or anomalous ossicular articulation. It is now realised that, despite the variable expressivity of the gene, a sensory component is the rule rather than the exception, being progressive in up to 50%, starting at under 30 years of age as a mild high-frequency loss and continuing to include the low frequencies. A 5-year study found that 73% (17 of 22) children with OI had a hearing loss: most had otitis media with effusion (OME) which resolved with treatment, but two children had conductive losses unrelated to OME and three (13.6%) had SNHL, one of which was detected at 1 year of age.[53] The hearing loss does

not seem to be related to the severity or the frequency of fractures, nor is it mutation specific. More severe degrees of hearing loss are often accompanied by vestibular symptoms.

Cleidocranial dysostosis

Cleidocranial dysostosis may present with progressive hearing loss. More often, the children are seen with a history suggestive of OME, which masks the underlying mixed hearing loss. This disorder of membranous and endochondrial bone formation is best known because of the aplasia or hypoplasia of the clavicles and persistence of open fontanelles and sutures with mild short stature. The prevalence is about 1 per million with a phenotype which can be overlooked. The pathology of the conductive hearing loss includes mastoid sclerosis and narrowing of the external auditory canal, stapes fixation, footplate sclerosis and other ossicular anomalies while speculation suggests that slowly progressive SNHL is due to narrowing of the internal acoustic meatus.

Ophthalmic problems

Pigmentary disorders of the retina are the best known of the variety of eye disorders that are reported to be associated with childhood progression of genetic hearing loss, some being clinically manifest after the onset of the hearing loss and some the primary symptom. Pigmentary retinal degeneration is usually the presenting feature of Alström syndrome, a complex recessively inherited condition, which includes the later appearance of progressive SNHL, truncal obesity and growth retardation, acanthosis nigrans and abnormalities of lipid and glucose metabolism. Edward syndrome is phenotypically similar but is accompanied by significant learning difficulties.

Other syndromes include corneal dystrophy (autosomal recessive), cataracts (autosomal dominant), macular dystrophy, ophthalmoplegias, optic atrophies or high myopia leading to retinal detachment. The onset of progression ranges from early childhood to the second decade and from slow progression in the high frequencies only to severe to profound deafness within 10 years.

X-linked syndromes with progressive SNHL are linked to a variety of neurological defects or optic atrophy. The SNHL of Mohr-Tranebjaerg syndrome is usually the first sign of the disorder, progressing rapidly to a profound loss by 10 years of age with later progressive neural degeneration affecting brain and optic nerves. In Norrie syndrome (congenital blindness due to maldevelopment of the retina with a variety of other ocular features), some 30% of patients develop a progressive hearing loss, usually in adult life, but onset may be masked by deteriorating cognitive function.

It is important to remember that a variety of eye development disorders are found in association with intrauterine infection including chorioretinitis, pigmentary change and cataracts.

Usher syndrome type 3

The rare Usher type 3 is characterised by progression of the initially mild hearing loss, which is not usually congenital. Retinitis pigmentosa develops at the end of the second decade resulting in night blindness and visual field defects. It is linked to chromosome 3q and thought to represent between 1% and 4% of patients with Usher syndrome. There seems to be a far higher prevalence in certain parts of Finland. There have been reports of progression in other types of Usher syndrome.

Marshall syndrome and Stickler syndrome

Progressive mixed hearing loss may be the presenting feature of the dominantly inherited Marshall syndrome, a disorder of connective tissue, in conjunction with myopia, cataracts and saddle nose. Stickler syndrome has similar features: high myopia associated with retinal detachment, cleft palate and spondyloepiphyseal dysplasia. The differentiation between these two syndromes remains uncertain: early onset and progressive hearing loss with vestibular hypofunction is reported to be a feature of Marshall syndrome but is also described in Stickler. There are a number of types of Stickler syndrome with varying degrees of hearing loss possibly related to the underlying genetic mutation. The importance of the correct diagnosis relates to the risk of retinal detachment, glaucoma and cataract in the untreated eye in Stickler.

Metabolic Disorders

The advent of treatment for some of these conditions underpins the need for awareness.

Mucopolysaccharidoses (MPS)

The progressive deterioration in physical, neurological and mental function in most of these lysosomal enzyme deficiency disorders supersedes concerns about hearing, but children may present with conductive hearing loss and apparent OME, usually within the first three years of life. Symptoms include deterioration in developmental progress and coarsening of the facial features with others depending on the enzyme involved. All are autosomal recessively inherited, with the exception of Hunter syndrome, which is X-linked.

At least five types are known to be associated with conductive hearing loss but progressive SNHL is less well recognised. The pathology is complex, involving both the middle ear and semicircular canals. The progressive conductive hearing loss of Hurler syndrome (MPS1-14) is now known to be associated with a progressive SNHL in many children. About 25% of those with Maroteaux-Lamy syndrome (MPS VI) have a progressive conductive loss as do 25% to 50% of those with Hunter syndrome (MPS II). Morquio syndrome (MPS IV) is generally milder than the other MPS but virtually all patients with type A develop a progressive SNHL by the second decade.

Glycosphingolipidoses

The X-linked Fabry's disease (lack of lysosomal alpha-galactosidase A) is characterised by severe neuropathic pain with transient ischaemic episodes, angiokeratoma, fibromyalgia, progressive renal disease and a variety of other symptoms. Progressive SNHL is a recognised feature in the majority of affected males and in female carriers.[54]

Biotinidase deficiency

Progressive sensorineural hearing loss develops in at least 75% of children who become symptomatic with biotinidase deficiency and can be profound. Children may present with hearing loss and episodic or progressive ataxia, or other symptoms including neurological features such as seizures, hypotonia, developmental delay and visual problems and cutaneous features such as skin rash, alopecia, and conjunctivitis. With biotin replacement, the neurological and

cutaneous manifestations resolve but the hearing loss and optic atrophy are usually irreversible. Biotinidase deficiency is secondary to absence of the water-soluble B-complex vitamin biotin.[55]

Renal disorders

A number of nephritides have progressive SNHL amongst their features: in Epstein syndrome, hearing loss and macrothrombocytopaenia develop before the age of 10 years with renal symptoms developing later in most families. The SNHL of Charcot-Marie-Tooth disease (nephritis with motor and sensory neuropathy) is often slowly progressive from childhood. Infantile renal tubular acidosis presents typically with failure to thrive in the first year of life with a severe and possibly progressive SNHL.

Alport syndrome

Typically, the hearing loss commences in the mid frequencies at the end of the first decade of life, but often commences earlier and may be one of the first symptoms. The syndrome includes specific glomerulonephritis, SNHL, axial myopia, anterior lenticonus (progressing to cataracts) and macular or perimacular flecks. The patient usually presents with haematuria. Progression to the high frequencies and further often parallels the deterioration in renal function due to renal failure. Males are more severely affected, but female carriers may also develop deafness, proteinuria and renal failure. The pattern of disease progression tends to be individual to the family; nearly all cases involving distinct mutations because of poor male fitness. The most common variety is X-linked (85%) with the remainder being mainly autosomal recessive. The basic pathology lies in changes in collagen formation affecting basal membranes.

Widened vestibular aqueducts: Pendred syndrome and branchio-oto-renal syndrome

The progression of SNHL is well recognised in structural anomalies of the inner ear. The commonest and best recognised of these is the widened vestibular aqueduct (WVA), first described in association with incomplete cochlear partition by Carlo Mondini in 1791.[56] Hearing loss occurs in early childhood or is congenital, commonly severe to profound and characterised by stepwise progression or fluctuation, often precipitated by minor head trauma or pressure change, such as air travel. These fluctuations with sudden drops of hearing are significantly associated with progression and sudden permanent total deafness and characteristic of Pendred syndrome.[57] Many patients also complain of dizziness and tinnitus. Confusingly, some patients will also have a mild to moderate conductive hearing loss, attributed to inner-ear fluid pressure on the round window. The WVA may be seen on 1.2 mm cuts on CT scan but is usually demonstrable on MRI scan as an enlarged endolymphatic duct and sac, often associated with the Mondini cochlea. A higher incidence of fluctuation and better hearing level has been reported in the 74% of 114 ears with inner ear malformations which were discovered to have the combination of WVAs, Mondini cochlea, large vestibule and semicircular canal dysplasia.[58] They do not necessarily co-exist, however, and do not form a syndrome in their own right. In over 70% of patients, the WVA is bilateral, although the hearing loss may

be asymmetrical. Even where a WVA is only found unilaterally, hearing loss is generally bilateral. There is no robust evidence that ultimate hearing level, pattern of loss or rate of progression is related to the size of the aqueduct at either the midpoint (>1.6 mm) or the external aperture. Some patients have the bony anomalies without enlarged endolymphatic sacs and without progression of the hearing loss, but there is no defined correlation. Normal hearing in ears with WVAs has been reported.

Pendred syndrome combines SNHL with goitre, the latter reported to occur in about one-third in the second or third decade, with abnormal uptake of iodine to the gland as measured on a perchlorate discharge test. Although examination of the neck should be undertaken regularly in deaf children, goitre is difficult to detect in those under 5 years old, and some children with goitre will turn out to have Hashimoto's thyroiditis. Perchlorate discharge may be misleading and is not recommended in children under the age of 10 years. The characteristic SLC26A4 mutation in the large Pendrin gene is described in a variety of homozygous, heterozygous and compound heterozygous alterations in over 50% of patients with WVAs.[59] It is associated phenotypically with bilateral WVAs and progression of the hearing loss, even in heterozygotes, although it is postulated that the latter may have more stable hearing.[60] Vestibular hypofunction occurs in approximately two-thirds of patients with Pendred syndrome, either unilaterally or bilaterally, and not consistently within families nor related to the labyrinthine structure.[61] Surgical occlusion has no benefit in WVAs and is adversely linked with further irretrievable progression.

A WVA is among the radiological features of branchio-oto-renal syndrome, a dominantly inherited syndrome of variable phenotype with a high prevalence of fluctuating and progressive SNHL. Other radiological findings include short bulbous internal acoustic meati, and labyrinthine and cochlear hypoplasia. The co-existence of OME in children and early surgery to address troublesome sinuses has been known to obscure the diagnosis. Careful family history and clinical examination searching for evidence of persistent conductive or mixed hearing loss, preauricular pits and sinuses, lacrimal sinuses, branchial cysts and sinuses and renal anomalies should enable differentiation. The discovery of the common mutation in the *EYA1* gene will help to confirm the diagnosis.

Those described are the more common of the causes of inner-ear anomalies and relatively well documented in the literature. Progressive hearing loss may also occur in other conditions, which include labyrinthine structural anomalies (e.g. CHARGE association). Common cavity anomalies are recognised to be at risk of recurrent meningitis as well as progression of the hearing loss. Although children with Mondini dysplasia do well with a cochlear implant, caution must be exercised in more primitive anomalies where reduced dynamic range and unwanted stimulation via the facial nerve may occur.

CHROMOSOMAL SYNDROMES

There are many chromosomal anomalies which include hearing loss but documentation is not comprehensive and most losses, as reported so far, seem to be non-progressive.

Down syndrome

The commonest hearing impairment in children with Down syndrome is related to otitis media with effusion and its complications. A progressive high-frequency hearing loss commences in

the teenage years in as many as one-third, continuing to affect nearly all of those in middle age. Proper information to families and regular hearing review are principles of good practice.

Turner syndrome XO and mosaics

Some 40–60% of children with Turner syndrome are reported to have a progressive mid-frequency dip or high-frequency hearing loss; most will have a history of recurrent ear infections and conductive hearing loss which may obscure the onset of the SNHL. Although described as typically occurring in the teenage years, the sensory loss has been identified in younger children and may be universal in monosomic individuals compared with mosaics.[62]

The hearing loss may masquerade as non-syndromal in children as the characteristic features of Turner syndrome are not always found due to the variability of the chromosomal configuration and the frequency of mosaicism.

INTRAUTERINE INFECTION

With increasing international availability of immunisation, the pattern of hearing loss is changing. Whereas 20 years ago, intrauterine rubella was a leading cause of SNHL, it is now found relatively rarely and interest has become more focused on cytomegalovirus (CMV). The hearing loss in other viral embryopathies has not been proven to progress.

Cytomegalovirus

There is now no doubt that hearing deteriorates in congenital CMV. Only 10% of those affected will be symptomatic at birth: approximately half of these will have significant hearing loss, often severe to profound with some evidence that the more severe initial hearing loss is predictive of progression.[63,64] Of asymptomatic infants, some 10% to 15% will later have symptoms of the disease, SNHL being the most common. A large study of 307 children with asymptomatic congenital CMV found that 7.25% had SNHL of whom 50% deteriorated between the ages of 2 and 70 months. Fluctuation was a significant finding in 22.7% of those with proven SNHL. Furthermore, some 18.2% of children did not appear deaf when first screened but became deaf between 25 and 62 months.[65] Children with unilateral hearing losses may develop a significant hearing loss in the better hearing ear, and both may become worse. Progression occurs most commonly in the first 6 years of life but may continue throughout childhood and into adolescence. CMV DNA can now be identified by PCR on the Dried Blood Spot (Guthrie) card allowing the thought that 20–30% of hearing loss might be attributed to CMV, and be mostly unrecognised: 10% of 87 children with hearing loss identified in the first 2 months of life; 34.2% of 38 children who presented later; 42.7% of 28 children with hearing losses greater than 70 dB.[66] Pre-existing maternal seroconversion does not protect against hearing loss but may have an impact on severity and progression.[67] Twin pregnancies may have different outcomes for each infant.

Current recommendations to identify congenital CMV include urine tests and mouth swabs (only diagnostic in infants aged under 3 weeks), blood testing for CMV IgG and examination of the Dried Blood Spot if these are positive or if all other common causes of SNHL have been excluded. False negative urine tests means that urine testing should be repeated if there is high clinical suspicion. Imaging by MRI for leukomalacia with ultrasound or CT scan for calcifica-

tion may help diagnosis. Vestibular dysfunction is increasingly recognised as a significant symptom of congenital CMV and has been observed more frequently than hearing loss.[68]

It is not yet clear whether progression is related to reactivation of the latent virus, to immunologic competence or to the manifestation of damage already done. Hearing loss is associated with increased virus in urine and viral load in blood[69] and current research is looking at the persistence of virus in perilymph.

Other exciting developments in congenital CMV include the possibility of treatment, the development of a vaccine[63] and, more recently, reports suggesting some relationship with genetic mutations.[70,71] Prolonged treatment with intravenous ganciclovir in the neonatal period is reported to prevent deterioration in hearing but at a significant cost in morbidity.[72,73] Oral valganciclovir demonstrably reduces viral load with fewer side effects.[74] Cochlear implants provide useful access to sound for those profoundly deaf, with performances on speech detection being comparable to peers, although full outcomes may vary depending on the child's other deficits.

Rubella

Progression of the hearing loss is thought to occur early in life in at least 25%, although recent information is lacking. Congenital rubella has declined due to immunisation and documentation of early progression was less certain when rubella was more prevalent and it was more challenging to measure hearing accurately.

Toxoplasmosis

Although progressive late-onset hearing loss has been reported in congenital toxoplasmosis, robust evidence is difficult to find. Neurological dysfunction and eye disease are the most common clinically relevant concerns.

Congenital syphilis

Although congenital syphilis is traditionally associated with a progressive SNHL beginning at the end of the second decade. the time of onset varies from early childhood to middle age. The hearing loss may be unilateral, bilateral, asymmetrical, of gradual onset over several years or occur with devastating rapidity as a sudden profound loss. Patients may also complain of tinnitus and vertigo. The classical neonatal findings of snuffles, rash, anaemia, jaundice and osteochondritis are easily overlooked and the later appearance of features such as saddle nose, Hutchinson's teeth and mulberry molars may not be recognised. Suspicion is most likely to be raised by the finding of interstitial keratitis, present in most patients. Spirochaetes may linger in the labyrinthine fluids for some time; osteitis and middle-ear thickening are also described. Active disease should be treated with penicillin and steroids.

Syphilis remains the great imitator in the pantheon of disease. Its incidence is increasing with the spread of HIV positive people and the increasing prevalence of unprotected sexual activity in young people. It must always be considered in progressive and sudden SNHL.

POST-NATAL INFECTION

Measles, mumps, Lyme disease and syphilis are described as causative agents of SNHL but evidence of progression in children is lacking.

Bacterial meningitis

Progression following bacterial meningitis is unpredictable and variable in both onset and time. Nearly a quarter of children were found retrospectively to have a fluctuating or progressive hearing loss which stabilised between 3 months and 4 years after the initial illness; no predictive factors for progression were identified in the initial illness.[75] Fluctuations may be attributed to secondary endolymphatic hydrops. Labyrinthitis ossificans is thought to account for many of the cases of progression; an early MRI scan is mandatory to demonstrate the filling defects indicative of early fibrosis so that the window of opportunity for a cochlear implant is not missed.

AUTOIMMUNE INNER EAR DISEASE

Autoimmune disorders remain uncommon, or perhaps unrecognised, representing less than 1% of SNHL, and are usually bilateral, sudden or rapidly progressive over only weeks or months, with occasional fluctuations. Diagnosis may be challenging in the absence of systemic symptoms or signs, but some 50% of patients will have vestibular symptoms (over 90% of those tested have vestibular hypofunction), and many will have tinnitus and aural fullness.[76,77] Cogan's syndrome in children is rare, often atypical (without interstitial keratitis or with a long gap between the onset of ophthalmic and auditory symptoms) with red eye predominating as the presenting feature, usually initially misdiagnosed.

There is a lack of information about the specificity of investigations even in adults: a pragmatic approach would include ESR, CRP, full blood count, ANA, ANCA, AECA, antiphospholipid, anticardiolipin and antithyroid antibodies, C3 and C4 and noting the response to treatment.[78] Tissue-specific antibodies such as hsp70 (68 kd) are not yet demonstrated to be useful (Agrup, 2007, personal communication). Progressive rather than sudden SNHL is reported to be more common in children and young adults with antiphospholipid antibodies.[79]

Prompt treatment with steroids is recommended to prevent further deterioration in hearing and possibly promote restoration but recent reports have found that the initial improvement seen in Cogan's syndrome is not sustained.[80] There is some evidence that those patients (adults) with antibodies to the inner-ear supporting cell antigen gained more improvement with steroids than those without.[81] Progressive hearing loss may be amenable to treatment with steroids and cyclophosphamide, methotrexate and plasma exchange.[6] The severe potential side effects of treatment with steroids may cause debate among clinicians and families and prejudice compliance. Cochlear implant has been successful in these children.

MENIERE'S DISEASE

The classical triad of tinnitus, episodic vertigo and fluctuating hearing loss is rare in children although it is suggested that a secondary endolymphatic hydrops may be more common than is realised. Temporal bone studies of infants and children showed bulging in Reissner's membrane in the cochlear duct in 16.9%, more commonly in those with congenital anomalies.[82]

INTRACRANIAL TUMOURS

Clinical experience suggests that tumours should be in the differential diagnosis of progressive SNHL, especially if unilateral.

Acoustic neuroma

Most cases present with unilateral symptoms of the neuroma including progressive hearing loss, tinnitus and, occasionally, disequilibrium; some present with other types of schwannoma or intracranial or spinal tumours. It is uncommon to find vestibular schwannomas presenting in childhood: almost exclusively they will occur in children with neurofibromatosis type 2 (NF2), probably first described by Wishart in 1820.[83] Childhood cataracts may occur in 20% and this, together with a family history, may alert the clinician. Café-au-lait spots are uncommon but may be found in the axillae. Management will be related to tumour size, growth and symptoms. Surgical removal is commonly necessary, with new techniques being more likely to preserve hearing. Brainstem implants are now being considered for those young patients who sustain bilateral profound hearing losses.

PERILYMPH FISTULA

Progressive hearing loss and dizziness have been reported as due to fistulae even in very young children. Nonetheless, the significance of perilymph fistula in progressive SNHL remains under debate. Most protagonists firmly believe that early identification followed by surgical intervention will stabilize hearing.[84] All children with relevant symptoms should undergo a CT scan to identify any radiological abnormalities including fractures, as well as a careful fistula test.

HEAD INJURY

A total of 32 (74%) of 43 patients over 14 years of age with closed head injuries were found to have hearing loss which progressed by more than 15 dB with risk factors including age and temporal bone fracture. The worse the initial hearing loss, the greater the progression.[85]

MIDDLE-EAR DISEASE

It has been suggested that chronic middle-ear disease may lead to SNHL in as many as 50% of patients, but there is no substantial evidence to confirm that this affects general communicative ability in children.[86] Some children with chronic or recurrent acute otitis media have been found to have impaired hearing in the very high frequency range (12 to 20 kHz) with normal hearing across the recognised speech frequencies.[87] Speech reception thresholds of some children operated on for cholesteatoma, both acquired and congenital, may became worse postoperatively;[88] it is not clear whether this is related to the site of the cholesteatoma, preceding disease or the surgery itself. Congenital cholesteatoma has also been reported in association with branchio-oto-renal syndrome where progressive hearing loss due to WVA is now well recognised.[89]

OTOTOXICITY

Drugs

Drugs associated with progressive SNHL include aminoglycoside antibiotics, salicylates, loop diuretics and chemotherapeutic agents. Research continues into the genetic predisposition to

ototoxicity, prompted in part by the m.1555A>G gene and aminoglycosides. It is important to reiterate that aminoglycoside ototoxicity may be dose related, without known genetic predisposition and devastating in its effects on the vestibular system with preservation of hearing.[90]

Platinum compounds are known to induce a dose-related progressive sensory loss, enhanced by previous irradiation; over 60% are likely to be affected, and at least one case of sudden total hearing loss has been reported following a single dose of cisplatin. Distortion product otoacoustic emissions may predict who will be affected and may detect early changes, as may extended high-frequency audiometry.[91]

Noise

Exposure to sudden or prolonged noise has long been recognised to be a potent inducer of SNHL in adults. With the advent of 'clubbing' and pop concerts (where the success of the evening is partly measured by temporary threshold shift), of personal sound systems and mobile phones, of noise-making toys and robots, increasing attention is being paid to noise as a hazard in young people.

Squeaky toys for babies (often cited by parents as an indicator of good hearing) may emit sounds from 78 to 110 dBA. Toy pistols and other 'weapons' can emit levels as high as 150 dB.[92] There have been anecdotal reports of noise levels as high as 90 dB in incubators, although 60 to 70 dB is more likely. The effects of noise on the young ear and whether damage is potentiated by ototoxic drugs remains unclear.

In young adults, outer hair-cell damage occurs first in the region of 2 kHz as measured by transient evoked otoacoustic emissions.[93] Large studies have identified notches in at least one ear of 12.5% of 6- to 19-year-olds.[94] Indeed, it has been estimated that 40% of 10 to 17-year-olds might have significant hearing loss after 10 years if they continue to experience their current noise exposure, with a higher relative risk in the less educated.[95] Alberti has suggested that noise-induced hearing loss is rare before the age of 10 years and states 'if ringing persists more than 30 minutes after listening to intense sound or if fullness of hearing persists for more than a very few hours, the young person almost certainly has hypersensitive ears and is at risk of hearing loss from levels of sound which are generally not damaging'[96].

Hearing aids

Given the evidence about noise, it is natural to consider powerful hearing aids as a factor in progressive hearing loss. Although a number of authors have addressed this both retrospectively and prospectively, evidence is lacking.[97] There may be differences in response between those with normal hearing and those with a hearing loss: one child who had been prescribed high-powered post-aural hearing aids at another centre for a presumed progressive profound hearing loss had absent transient evoked otoacoustic emissions (TEOAEs) and a 50 dB nHL threshold to high-frequency click on ABR testing; the TEOAE were robustly recordable and ABR thresholds measured 20 dB nHL, 36 hours after she stopped wearing her hearing aids (Crowhen, 2007, personal communication).

TREATMENT

A variety of generic treatment regimens have been proposed over the years with little in the way of evidence to support them. The suspicion is that physicians are treating their own natural

anxiety and that of the parents, rather than expecting to find any change. Thus, betahistine, ubiquitin (extract of calf thymuses), glycerol, calcium antagonists, carbogen, magnesium and cerebral vasodilators have all been tried but without notable or sustained success; the use of many of these substances might be frowned upon in children where research is lacking. High-dose oral steroids, hyperbaric oxygen and bed rest remain a standard protocol during any identified acute phase of deterioration but there is no evidence of success and confining an otherwise healthy young child to bed is a challenging proposition. There is no reliable evidence of the benefit or optimal methodology for direct instillation of steroids into the middle ear, nor of what might be treated in this manner, nor for what duration. Children with symptoms suggestive of endolymphatic hydrops may benefit from restriction of salt and caffeine intake and the judicious use of diuretics.

Surgical approaches have now mostly been discounted except where there is a clear history of perilymph fistula, where ventilation of the middle ear in intercurrent glue ear is necessary for health and effective amplification and where there is a risk of meningitis in severe Cock's anomaly of the inner ear in combination with progressive hearing loss.

MANAGEMENT

The aim of management must be to maintain the child's confidence and rate of progress in development and learning. The mainstays of management, therefore, are continuous rehabilitation and support. Addressing the child's needs as well as the parents', introducing psychological support, reviewing hearing aid provision, communication mode and educational environment must all be undertaken sensitively and promptly.

The child and family will be under a number of increased stresses at this time. They must deal with the increasing impairment in the child and the impact this has on the child's ability to function and the reaction of those around them. They may have to address other major health or ability problems. More frequent visits to the audiology centre result in time away from work and school, reduced academic achievement and reduced confidence; teasing may occur both because of poor achievement and because of increased hearing difficulty. Parents may cling onto the child's earlier hearing potential and be reluctant to adapt to additional needs, especially if this involves a change of school or the introduction of signing. Feelings of guilt may supervene, especially where the loss is recognised to be genetically determined or due to some potentially avoidable cause. From research on children with chronic illness, much depends on how the child's mother reacts, whether there is a network of family or friends to support them or whether support is distant and fragmented. This is more challenging where the family originates from another area, culture or country or is constantly relocating. Siblings may perceive the extra attention as 'unfair' and have a higher incidence of emotional behavioural problems with poorer identity and self-worth.

Children cope with bad news in a variety of ways, depending on their age. They need more than just the facts about their increased hearing loss – they need tactics and strategies to maintain their self-esteem, to be able to explain their situation to others and to address the conflict that may arise within their personal and social lifestyles. The often unpredictable nature of progression makes it difficult for the child and family to adjust. Their sense of belonging to a common community may be disturbed. Many older children ask to talk to peers who have had a similar experience, as do their parents. They may wish to meet a member of the Deaf community.

In pre-school children, additional emotional and practical support is often provided by the visiting teacher of the deaf who is often already a family confidant; finding support is much

more difficult for the families of children already attending school who have less contact with their child's peers and teachers. It is often difficult to encourage services to respond quickly to a change in a child's circumstances and all too often the school-age child is left floundering long after he or she has requested additional support or a change to special education. Specialist speech and language therapists for the deaf are in short supply but their help is invaluable in determining speech perception, evaluating speech and language acquisition and supporting its development. Clinical psychologists with knowledge and experience of deaf children are thin on the ground but have the skills to encourage resolution of grief in parents, children and siblings, advise on behaviour, build self-esteem and confidence and address specific issues such as tinnitus. There may be a social worker for the deaf available to offer wider experience or a counsellor with specific training. Explanation to a child's peers and siblings may moderate the social effects of the greater loss.

Digital hearing aids offer flexibility in programming and microphone facilities, which enable a greater consistency in the provision of amplification. Assistive listening devices should not be overlooked: the frustration and grief of previously independent children will only increase if they find that they are now totally dependent on family or peers for everyday activities and alerts. Children with progressive profound SNHL find cochlear implant of considerable benefit, especially if they have had good language development and access to sound prior to implant. Some families regard the initial discussion about implant with horror as it underlines the unthinkable and removes all expectation of recovery. Cochlear implant may be rejected outright by a reluctant and distressed adolescent or seen inappropriately as of great benefit or even 'cure' by distressed and anxious parents. Close working with the implant team will allow families a realistic and carefully timed empathetic assessment, coordinated with local services, before the child is really struggling.

Appropriate investigation, onward specialist referral (where needed) and genetic counselling must always be offered to the child and family. Where the diagnosis is in doubt, careful and regular medical review is essential as is regular review of the hearing of siblings who may develop a progressive loss at a different age from the recognised child and go undetected for significant periods, especially if previously they have been found to have normal hearing.

CONCLUSIONS

The identification and management of progressive hearing loss in children remains one of the most difficult and challenging aspects of audiovestibular practice, requiring a high index of suspicion, robust audiometric and clinical facilities with sound medical knowledge of related disease and disorder and professional honesty with both families and children. Fortunately, the integrated working of the multidisciplinary team of professionals in paediatric audiology departments facilitates an effective and supportive approach, underpinned by careful liaison with local professionals, agencies and voluntary organisations.

The advent of newborn hearing screening theoretically reduces doubt about the age of onset of hearing loss and its natural history but risks inducing a false sense of security in both parents and professionals when babies pass the screen. Immigrant families may not have had this early screening nor benefited from comprehensive immunisation programmes, and thus remain a particularly vulnerable population. The suspicion that progressive hearing loss is more common than previously may be a false premise related to earlier age of identification, more carefully structured review, more reliable audiometry and greater knowledge about the

natural history of hearing loss and related conditions. There is increasing realisation that some children may have positive results for more than one condition associated with progressive hearing loss.

Treatment to prevent the onset or progression of hearing loss is now available for a number of conditions, although success is variable. Future treatments through gene therapy and viral delivery of neurotrophins or antioxidants (with or without cochlear implant) are tantalisingly close but will depend on the robust identification of cause and pathophysiology. It is not obvious whether there will be a possibility of reversing damage already done, of preventing onset of hearing loss or of preventing progression.

Much of the current research is through geneticists whose endeavours not only help to clarify the diagnosis but also open up possibilities for pre-symptomatic identification, prevention or direct treatment. A number of mouse homologues have enabled clarification of mechanisms. The knowledge that genetic predisposition may affect the adverse effects of environmental factors underlines the importance of further research, public health information and prevention. There are a multitude of symptoms and syndromes associated with progressive deafness, many of which will present to paediatricians in other fields: clinical collaboration, research, publication and dissemination of information will enable the needs of children and their families to be met.

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14 Children with unilateral sensorineural hearing loss

D.P. Sladen, A. Rothpletz and F.H. Bess

INTRODUCTION AND BACKGROUND

The past two decades of research have demonstrated that unilateral hearing loss in children may produce communicative or psychoeducational deficits. Specifically, children with unilateral hearing loss can be at risk for a number of complications including communicative deficits, social or emotional problems and academic failure.[1–9] This chapter offers an overview of theoretical and clinical knowledge of children with unilateral sensorineural hearing loss. To this end, the chapter focuses first on background information pertinent to unilateral hearing loss and includes such topics as binaural versus monaural listening, speech understanding in adverse listening situations, and learning and educational issues. Second, a review of the current status of this population and recommendations for identification and management is provided.

BINAURAL VERSUS MONAURAL LISTENING

Some of the problems experienced by children with unilateral hearing loss can be explained, in part, by their lack of binaural processing mechanisms. That is, two ears provide a distinct listening advantage over one ear alone. Factors that contribute to a binaural listening advantage include: binaural summation,[10] localisation,[11–13] head shadow effects[14] and binaural release from masking.[15]

Binaural summation

When a sound is presented simultaneously to two ears it is perceived louder than if the same sound is presented monaurally. Research has demonstrated that binaural thresholds for pure-tone and speech stimuli are better than monaural thresholds by approximately 3 dB. That is, when two ears are equated for hearing sensitivity, individuals with normal hearing receive binaural gains for both tonal and speech stimuli. Although a 3 dB binaural advantage may seem unimportant, it has considerable effects on speech understanding. A 3 dB increase can result in an 18% improvement in monosyllabic word recognition scores and a 30% improvement for sentence materials.[12] This binaural advantage is even greater at suprathreshold levels. For example, a stimulus presented at a level of 30 dB SL to one ear has about the same loudness as a 24 dB SL stimulus presented simultaneously to both ears – a 6 dB effect. Binaural gains can be as large as 10 dB for stimuli presented at 90 dB SL.[16–17] Depending on the severity of their hearing loss, individuals with unilateral hearing loss may not experience

binaural summation – a phenomenon which is also thought to contribute to the ease of listening.

Localisation

Another binaural phenomenon is the ability to localise a sound source on the horizontal plane. Predictably, individuals with unilateral hearing loss exhibit considerable difficulty on localisation tasks. Interaural time differences (ITDs) and interaural intensity differences (IIDs) provide a physical basis for the localisation of sound on the horizontal plane. Sounds are localised on the side that receives the more intense signal level or earlier stimulation. The cue (ITD or IID) that predominates in sound localisation depends on the frequency of the sound stimuli. Specifically, the ITD is the predominant cue for low-frequency sounds and the IID is the predominant cue for high-frequency sounds. In summary, the localisation of sounds is largely dependent upon a listener's ability to process between-ear differences in the time of arrival or the intensity of the auditory stimulus.

Individuals with unilateral hearing loss have limited access to IID or ITD cues. Without such cues, they must depend on less-reliable cues (loudness, pinna effects, head movements) to localise sounds on the horizontal plane.

Head shadow effect

The 'head shadow effect' occurs when the head serves to attenuate sounds propagating to the ear farther from the sound source. It is most salient when a sound is directed at a 45-degree angle towards the listener. This head shadow causes a reduction in the intensity of the signal at the far ear. Specifically, for a sound source at 45 degrees, the head shadow can attenuate speech (complex) signals 6–12 dB in the far ear relative to its level in the near ear.

For a normal-hearing listener, the head shadow effect does not generally affect speech recognition. Regardless of the origin of the primary and competing signals, listeners with normal hearing can attend at will to the ear having the better signal-to-noise (S/N) ratio. In contrast, the impact of the head shadow effect for individuals with unilateral sensorineural hearing loss depends upon the orientation of the listener. The effect is most profound when the primary signal comes from one source and a competing message or noise comes from a different source. If the primary signal originates from the side of the impaired ear and the noise originates from the listener's good ear, the resulting listening condition is most adverse for individuals with unilateral sensorineural hearing loss.

The head shadow effect can create problems in speech understanding for individuals with unilateral hearing loss. The effect is greatest for high-frequency sounds. Because high-frequency consonants carry 60% of speech intelligibility, individuals with unilateral hearing loss experience significant speech recognition difficulty when the signal source is initiated on the side of the impaired ear.

Binaural release from masking

Consider the situation in which a noise masker and a signal are presented binaurally to a normal hearing listener over headphones. If different interaural manipulations are imposed on the masker from those imposed on the signal, the signal becomes more detectable than if the same manipulations are imposed on both masker and signal or if the masker and signal are presented

to only one ear.[18] When masking noise is presented identically to both ears whilst speech signals are presented interaurally phase reversed, the release from masking is approximately 3–8 dB.[19] This binaural release from masking is believed to result from the auditory system's ability to compare the stimuli to the two ears and effectively reduce or cancel the masking noise, thereby yielding a better S/N ratio than occurs in either ear alone.[20]

Binaural release from masking is one factor underlying the real-world phenomenon known as the 'cocktail party effect'. The cocktail party effect refers to the ability of an individual to tune in to one conversation in a room of competing conversations. Under these circumstances, the auditory system is able to take advantage of the fact that the primary conversation has a different ITD from that of the competing conversations because of their different spatial locations.[21] Clearly, a person with a unilateral hearing loss would be unable to take advantage of this difference.

Several studies have demonstrated the positive effects of binaural release from masking on speech recognition in adverse listening situations.[22–25] For normal-hearing subjects, speech recognition improves for binaural stimulation over monaural stimulation, even if in the monaural case the ear is in a favourable position for the primary signals. Also, binaural speech recognition is superior to monaural recognition in reverberant conditions; a 3 dB S/N ratio advantage exists for binaural listening over monaural listening.

In summary, binaural hearing offers a number of important listening advantages over monaural hearing. These advantages have clear implications for understanding speech under routine daily living activities. The benefits of binaural hearing are particularly apparent in communicative situations where background noise and/or reverberation exist.

SPEECH UNDERSTANDING IN ADVERSE LISTENING SITUATIONS

Similar to normal hearers, individuals with unilateral hearing loss are often confronted with listening in a variety of adverse listening situations – situations that can interfere with the ease of listening. These difficult listening situations can presumably impose a deleterious effect on classroom learning.

A child's ability to hear the teacher and make fine-grained auditory discriminations depend highly on the acoustical conditions of the classroom, particularly the S/N ratio. The S/N ratio is the relationship between the primary speech or signal of interest (e.g. the teacher's voice) and background sounds (e.g. other talkers, hallway noise, air-conditioner noise and classroom clatter). A poor S/N ratio and reverberant conditions not only degrade speech perception ability, but also negatively effect behaviour, concentration, attention, reading/spelling ability and academic outcomes.[26–32]

Unfortunately, the classroom is not an ideal listening environment for normal-hearing students, much less for students with unilateral sensorineural hearing loss. Finitzo-Hieber and Tillman[30] reported that, based on the monosyllabic word discrimination performance of normal-hearing children, an adequate classroom listening environment would have a S/N ratio of at least +6 dB (preferably +15 dB) and reverberation time of less than 0.04 seconds. Gengel[33] reported that children with hearing loss require a S/N ratio of +20 to 30 dB for maximum speech understanding. Saunders,[34] however, found that the S/N ratio in a typical classroom ranged from +5 to +1 with a reverberation time of 0.6–1.2 seconds. Other studies have also reported S/N ratios of +5 to −7 dB.[27–29,32,34] These findings suggest that the S/N ratio and reverberation time commonly found in classrooms is unacceptable even for children without educational disabilities.

It is possible that the listening conditions of classroom environments interfere with the communication and learning of many children with unilateral hearing loss. In fact, normal-hearing children yield significantly more errors in speech discrimination when classroom noise is present than in quiet conditions. The noise and reverberation levels, typical of many classroom settings, can mask many of the important cues needed for speech understanding. Therefore, children who already miss some of the salient acoustic cues for speech because of their unilateral hearing loss will experience even greater difficulties under acoustic conditions often encountered in schools.

Several studies have demonstrated the undermining effects of noise and reverberation on the speech understanding of both normal hearing children and children with hearing impairment – including those with unilateral hearing loss. These studies have demonstrated that children with hearing loss experience greater debasement in word recognition as noise and reverberation increase.[28,30,35–37] In addition, children with even minimal degrees of hearing loss experience more difficulty than their normal-hearing peers. Finally, the more adverse the listening situation, the greater the disparity in speech perception performance of normal-hearing children and children with hearing loss.[28]

LEARNING AND EDUCATIONAL ISSUES

When one considers the probable adverse effects of noise on the speech understanding of persons with unilateral sensorineural hearing loss, it is not surprising that many of these children experience academic problems as well. Evidence suggests that fine-grain speech perception skills are critical to language development and learning. Elliott, Hammer and Scholl[38] found that measures of fine-grained auditory discrimination classified nearly 80% of children, 6 to 8 years old, as progressing normally or demonstrating language learning difficulties. Others have reported that weakness in the auditory discrimination of speech sounds is one of the most frequent causes of poor reading skills.[39–40] Such findings suggest that academic achievement is highly dependent on the student's ability to perceive and discern word–sound differences.

It seems probable that speech-understanding difficulties in noisy conditions contribute to the academic problems experienced by many children with unilateral hearing loss. Downs and Crum[41] reported that processing demands during auditory learning are significantly greater under competition than under quiet conditions. Classroom noise may produce deleterious effects on the learning performance of children, particularly children with hearing loss. Increased effort is required to attend selectively to an auditory signal when the acoustical environment is adverse. If this energy is not expended, there is a concomitant decrease in learning performance. That is, optimal learning may be compromised if the processing demands of a task are increased. It is possible that unilateral hearing loss accompanying noise and reverberant conditions typical of most classrooms makes learning a highly demanding task, resulting in reduced academic performance of many children with unilateral sensorineural hearing loss.

AUDITORY ASYMMETRIES AND SPEECH LATERALISATION

The terms 'laterality' and 'lateralisation' are often used to describe differences between the left and right cerebral hemispheres and imply the dominance of one hemisphere over the other

with regard to a specific brain function.[42] A long-standing premise has been that speech lateralises to the left cerebral hemisphere and non-speech sounds lateralise to the right cerebral hemisphere in most people. This assertion is based on early animal studies, research examining individuals with brain damage, and performance of adults and children on dichotic listening tasks.[43–46] In dichotic listening tasks, different speech stimuli are presented to each ear simultaneously. Studies have shown that children and adults demonstrate better recognition of speech stimuli presented to their right ear than to speech stimuli presented to their left ear.[45,47–48] This right-ear advantage on dichotic listening tasks is thought to support the theory that the left hemisphere is specialised for language and the right ear has privileged access to the left hemisphere. Privileged access of the right ear to the left hemisphere is attributed to the fact that contralateral auditory pathways are stronger than ipsilateral pathways, and activity in contralateral pathways suppresses the ipsilateral pathways. Current research employing electrophysiology and neuroimaging techniques has generally supported the concepts of a specialised role of the left hemisphere for processing language and favoured access of right-ear input to the left hemisphere.[49–51] However, a few studies suggest that left/right differences in auditory processing may be based on acoustic properties of the input rather than based on whether the input is speech or non-speech stimuli per se.[52,53] Specifically, temporal information (critical for processing rapidly changing signals such as speech) is thought to be favourably processed in the left auditory cortex and tonal or spectral information is thought to be favourably processed in the right auditory cortex.[53]

Interestingly, in normal-hearing infants, asymmetries in auditory function have been identified at the most peripheral levels of the auditory system. Specifically, click-evoked stimuli (a rapidly changing signal) presented to the right ear elicit larger otoacoustic emission responses and larger and more rapid auditory brainstem responses than click-evoked stimuli presented to the left ear.[52] These investigators have suggested that processing of sound in the auditory system at the level of the cochlea and brainstem during infancy may serve to facilitate later development of hemispheric specialisation for sound processing.[52]

Given that unilateral hearing loss results in a deprivation of auditory input to one side, it is reasonable to speculate that asymmetries in the auditory system may be affected. Auditory evoked potential and functional magnetic resonance imaging (fMRI) studies have demonstrated a reduction of hemispheric asymmetries in individuals with unilateral hearing loss compared with normal-hearing individuals.[42,54,55] These studies suggest that there may be some reorganisation of the central auditory system when unilateral hearing loss occurs. How reorganisation of auditory processing affects the development of children with unilateral hearing loss is not well understood at this time. However, as is discussed in the next section, differences between children with right- and left-sided hearing loss have been documented in the areas of speech perception, academic achievement and intellectual abilities.

Children with unilateral sensorineural hearing loss

Given the discussion presented in the previous section concerning the advantages of binaural hearing, the problems children have understanding speech in noise, and learning and educational issues, it is not surprising to note that some children with unilateral hearing loss experience a variety of communicative, psychoeducational and psychosocial problems. This section addresses our current knowledge on unilateral hearing loss in the areas of epidemiological considerations, auditory performance, educational performance, language, cognitive skills and functional health status.

Epidemiological considerations

The epidemiology of unilateral sensorineural hearing loss is an important consideration when examining the nature of the problem, determining methods for identification and planning strategies for intervention. Epidemiological issues pertinent to unilateral sensorineural hearing loss include: prevalence, age of identification and aetiology.

Prevalence

The prevalence of unilateral hearing loss has been examined in both newborn infants and school-age children. With regard to infants, aggregate data reported by 37 state newborn hearing screening programmes in 2004 to the Directors of Speech and Hearing in State Health Welfare Agencies (DSHPSHWA) and the Center for Disease Control (CDC) indicated that the prevalence of permanent unilateral hearing loss* was 0.354 per 1,000 babies screened.[56,57] However, other data suggest that the prevalence of permanent unilateral hearing loss in newborns may be much higher than state-sponsored newborn hearing screening programmes indicate. For example, a study by Prieve et al.[58] examining the New York State Universal Newborn Screening Demonstration Project reported a prevalence of permanent unilateral hearing $loss_1$ of 0.83 per 1,000, which is over twice the prevalence indicated by the DSHSHWA/CDC data. This discrepancy in prevalence estimates of permanent unilateral hearing loss between research data and reports from state health departments has led some experts to speculate that the number of infants with unilateral hearing loss are significantly underidentified or under-reported at the present time.[59]

Shifting attention to school-age children, it is surprising to find the prevalence rates of permanent unilateral hearing loss to be nearly 100 times the prevalence rate in newborns reported by DSHPSWA. For example, Bess et al.[2] reported a prevalence of unilateral sensorineural hearing loss of 3 per 100, or 3% in children (aged 8–15 years), when using an average threshold criterion of ≥20 dB HL in the affected ear. Even higher prevalence rates of unilateral hearing loss have been reported by the National Center for Health Statistics. Specifically, data collected from 1988 to 1994 through the Third National Health and Nutrition Examination Survey (NHANES-III) estimated that the prevalence of low-frequency unilateral hearing $loss_2$ was 5.6% and the prevalence of high-frequency unilateral hearing $loss_2$ was 9.6%.[60] The higher prevalence rates reported by NHANES-III may be attributed, in part, to the fact that they used a lower average threshold criterion to define hearing loss (i.e. >15 dB HL rather than >20 dB HL), and to the fact that the survey did not include immittance measures and bone-conduction audiometry to separate transient conductive hearing loss (i.e. resulting from otitis media) from sensorineural and non-transient conductive hearing loss. Nevertheless, data from multiple sources indicate that the prevalence of unilateral hearing loss in school-age children is much higher than prevalence estimates reported in the newborn population.

So, what accounts for the remarkable discrepancy between prevalence rates reported in the newborn period (i.e. 0.35–0.83%) and prevalence rates reported in school-age children (i.e. ≥3%)? The answer is likely manifold. First, a number of school-age children with unilateral hearing loss presumably had normal hearing at birth and then acquire hearing loss during childhood. Factors associated with late-onset unilateral hearing loss include bacterial

* Permanent hearing loss was defined as sensorineural hearing loss, non-transient conductive hearing loss (e.g. resulting from craniofacial anomalies, ossicular fixation, etc.) or mixed conductive and sensorineural hearing loss of >20 dB HL in the affected ear.[56,58]

meningitis, head trauma, certain genetic mutations, cytomegalovirus (CMV) and noise-induced threshold shift. One cannot assume, however, that the jump in prevalence estimates between infants and older children is attributed entirely to late-onset hearing loss. Rather, it is likely that a significant number of infants with congenital unilateral hearing loss are not identified until later in life because either they are 'lost to the system' or are missed by current technology used in newborn hearing screening programmes. As demonstrated by data reported by state health departments, over half of all infants who fail their newborn screening never return for diagnostic testing.[56] Children with unilateral hearing loss who do not return for follow-up testing after failing their newborn screening often will not be identified until they are 5–6 years of age and fail a routine school hearing screening. In addition to infants who are lost to follow-up, infants with mild hearing loss (unilateral and bilateral) may constitute another group missed by newborn hearing screening programmes. Current newborn screening technologies (otoacoustic emissions and automated auditory brainstem response measures) do not reliably differentiate between normal hearing and mild hearing loss and, therefore, may miss a significant number of infants with mild hearing loss.[61,62] Finally, inconsistencies in the reporting of newly identified cases of hearing loss to public health departments may lead to underestimation of the prevalence of infants with unilateral hearing loss. Specifically, it is possible that some infants with unilateral hearing loss are identified as a result of newborn screening but are not included in prevalence estimates reported by public health departments because some diagnostic centres do not report the newly identified cases to government and health officials.

Demographic factors such as gender and ethnicity appear to have some influence on the prevalence of unilateral hearing loss. A number of studies have shown that children with unilateral hearing loss are somewhat more likely to be male than female.[6,8,63–65] With regard to ethnicity, Lee, Gómez-Martin and Lee[68] examined prevalence rates of unilateral hearing loss of various ethnic groups in the United States and Puerto Rico and found the prevalence of unilateral hearing loss (>30 dB HL) in children (aged 6–19 years) to be 11.8% in African-Americans, 12.3% in Cuban-Americans, 6.4% in Mexican-Americans, 6.9% in Puerto Ricans and 7.9% in white non-Hispanic-Americans.

Age of identification

In years past, unilateral hearing loss was identified much later in life than bilateral hearing loss. Because most young children with unilateral hearing loss do not have conspicuous speech and language deficits, the hearing loss typically went undetected until the child failed a routine hearing screen in kindergarten or first grade.[66] However, with the recent widespread implementation of universal hearing screening, many children with congenital unilateral hearing loss are now identified during early infancy. To date, there have been no published reports documenting the average age of identification of unilateral hearing loss in children since the adoption of mandated universal newborn hearing screening programmes in the UK and the USA.

Aetiology

The cause of unilateral hearing loss is idiopathic in 35–66% of cases, according to various studies.[6,63,67–70] Congenital factors known to be associated with unilateral hearing loss include connexin-related genetic mutations (*GJB2* gene and *GJB6* gene), CMV, enlarged vestibular aqueduct, craniofacial anomalies (syndromic hearing loss) and non-syndromic cochlear nerve

aplasia. Unilateral hearing loss may also be acquired during childhood. Factors associated with late-onset/acquired unilateral hearing loss include CMV, meningitis, seizures, and head trauma. Prior to the introduction of the mumps vaccine in 1967, the mumps virus was also a common contributor to acquired unilateral hearing loss. It has been reported that most developed countries have experienced a 90% to 95% decrease in the incidence of mumps due to immunisation efforts;[6] however, there have been recent outbreaks of mumps documented in the USA, the UK and other developed countries.[57,71] Another possible cause for acquired unilateral sensorineural hearing loss is middle-ear disease with effusion. Young children with an early history of ear disease can experience hearing loss in the high frequencies, especially if a history of multiple intubations exist.[72,73] It is theorised that bacterial products are transmitted through the round window causing damage to the basal end of the cochlea. Finally, exposure to hazardous noise levels may also be a contributing factor in cases of acquired unilateral hearing loss in children. In an analysis of NHANES-III data, nearly 10% of the 5,249 children (aged 6–19 years) surveyed demonstrated a noise-induced threshold shift in one ear.[60]

Auditory performance

Localisation

Children with unilateral sensorineural hearing loss experience significant problems localising sound on the horizontal plane. Bess, Tharpe and Gibler[4] reported on the localisation scores of a group of children with unilateral hearing loss ($n = 25$) and contrasted these data to a matched group of children with normal hearing. Localisation scores for the unilaterally hearing-impaired children were significantly poorer than the normal-hearing children at the two frequencies tested (500 and 3,000 Hz). Predictably, localisation scores were positively correlated with the degree of hearing loss; that is, as hearing loss in the impaired ear worsened, localisation errors also increased.

Speech recognition

Given that they are not able to reap the benefits of binaural listening, noisy environments pose significant challenges for children with unilateral hearing loss. Accordingly, studies have shown that children with unilateral hearing loss have more difficulty understanding speech than normal-hearing children when background noise is present. For example, Bess, Tharpe and Gibler[4] examined the speech recognition skills of children with unilateral sensorineural hearing loss and a matched group of normal listeners ($n = 25$) at different S/N ratios using nonsense syllables. Unilaterally hearing-impaired children exhibited significantly greater difficulty understanding speech than did their normal-hearing counterparts under all listening conditions. A summary of the data reported by Bess and co-workers is shown in Figure 14.1.

This figure illustrates the mean nonsense syllable recognition scores (percent correct) across several S/N ratios for a group of normal-hearing children and children with unilateral sensorineural hearing loss. The hearing-impaired children were assessed in monaural direct and monaural indirect conditions, whereas the normal hearers were tested in the monaural direct condition only. Interestingly, it is seen that the unilaterally hearing-impaired children performed worse than the normal hearers across all monaural direct conditions; that is, when the primary signal is directed to the good ear with noise striking the poor ear at full impact, unilaterally hearing-impaired children did not perform as well as their normal peers. Moreover, the more adverse the listening situation, the greater the discrepancy between the unilateral

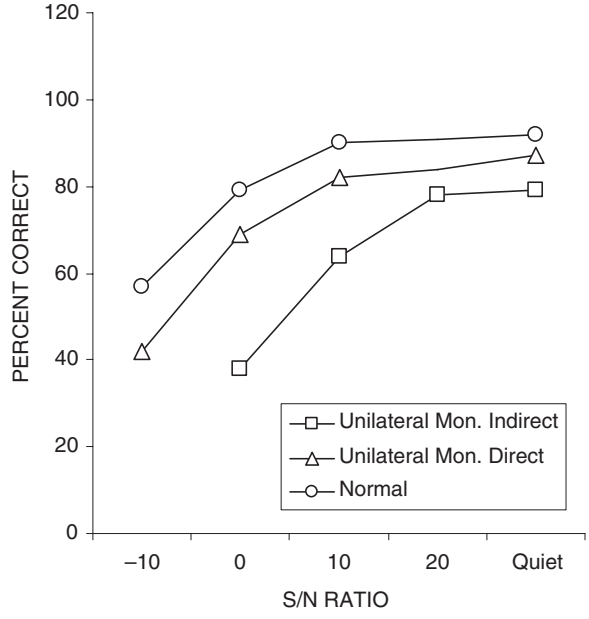

Figure 14.1 Data from Bess, Tharpe and Gibler[4] that show the speech recognition skills of children with unilateral sensorineural hearing loss and a matched group of normal listeners ($n = 25$) at different S/N ratios using nonsense syllables.

subjects and their normal-hearing counterparts. Specifically, in the monaural indirect condition, the children with unilateral hearing loss show a marked decrease in speech recognition even under the most favourable S/N ratios. The ability to understand speech varied based on the degree of hearing loss; children with the more severe impairments performed less well than children with the milder hearing losses. Clearly, children with unilateral sensorineural hearing loss experience far greater difficulty understanding speech in the background of noise than children with normal hearing, especially in the monaural indirect condition. Other studies support these conclusions. Ruscetta, Arjmand and Pratt,[74] for example, found that children with unilateral hearing loss required significantly greater S/N ratios to perform as well as children with normal hearing on sentence and syllable recognition tests, particularly in the monaural indirect condition where the children with unilateral hearing loss required almost a 9 dB greater S/N ratio to perform equally as well as their normal-hearing peers.

Several studies have suggested that children with right-sided unilateral hearing loss have more difficulty on speech perception tasks than children with left-sided impairment. As seen in Figure 14.3, data from Bess et al.[4] demonstrated a definite trend of right-ear-impaired subjects performing more poorly than left-ear-impaired subjects across all listening conditions. Right-ear-impaired subjects performed more poorly than normal-hearing children and more poorly than left-ear-impaired subjects.

Finally, research suggests that a significant relationship exists between poor speech perception abilities and academic failure in children with unilateral hearing loss. Specifically, Bess and co-workers[4] examined the speech recognition abilities of children with unilateral hearing loss as a function of those who had failed a grade and those who had not failed a grade. The findings from this analysis are shown in Figures 14.2 and 14.3. The speech recognition

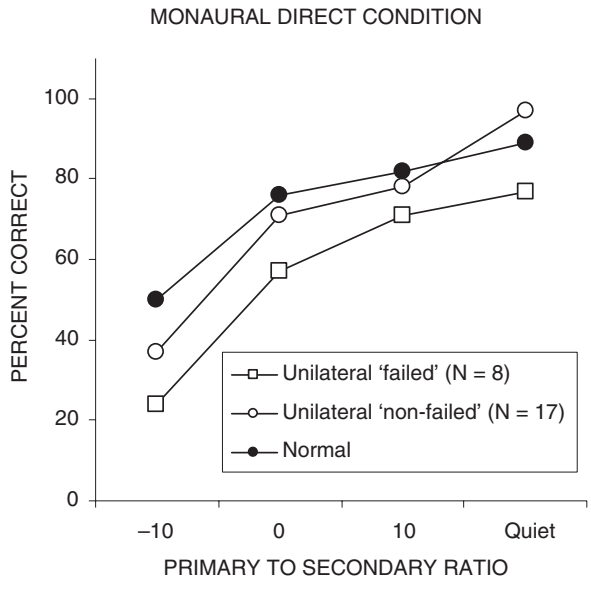

Figure 14.2 Results from Bess[66] show the speech recognition abilities of children with unilateral hearing loss as a function of those who had failed a grade and those who had not failed a grade.

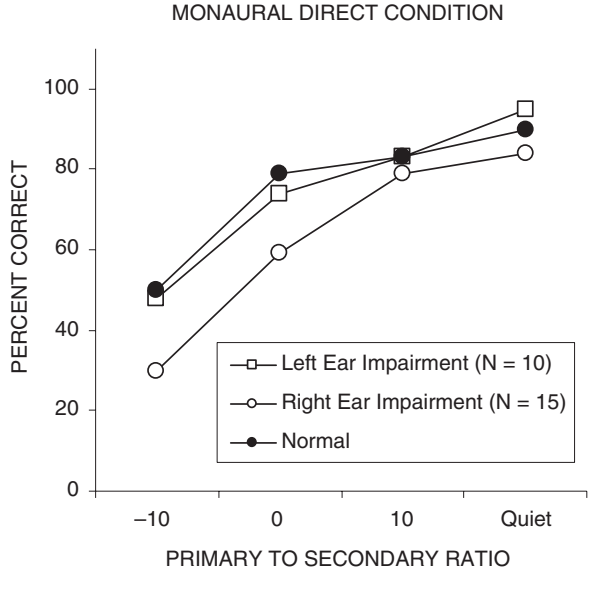

Figure 14.3 Data from Bess et al.[4] showing right- and left-ear-impaired subjects in quiet and various S/N ratios.

performance scores for children who had failed a grade versus those who did not fail are shown in Figure 14.2. There is a tendency for children who experience difficulty in school to perform more poorly in the monaural direct position than children who perform satisfactorily in school. Note, for example, that children who have failed a grade perform worse than normal listeners and children who have not failed a grade across all listening conditions.

EDUCATIONAL PERFORMANCE

Children with unilateral hearing loss experience far greater difficulty in school than children with normal hearing. In fact, children with unilateral sensorineural hearing loss are ten times greater at risk for academic failure than their normal-hearing counterparts.[3,8] A breakdown of the grades typically failed by children with unilateral loss is shown in Figure 14.4.

Note that the largest number of children failed in the first grade; however, many children failed grades two through seven. Boyd[75] was one of the very first to examine the effects of unilateral hearing loss on educational performance. Boyd reported that 38% of their children with unilateral loss exhibited reading problems, 31% exhibited spelling problems and 23% had problems in arithmetic. Subsequent studies have supported Boyd's findings. Brookhouser et al.,[6] for example, reported that 59% of 173 consecutive children with unilateral hearing loss evaluated at Boys Town National Research Hospital had a history of either academic or behaviour problems at school. Bess and colleagues[4] reviewed case history data from 60 school-age children with unilateral hearing loss and found that 35% failed at least one grade; these data rank in comparison to an overall failure rate of 3.5% for the school district norm. Overall, 48.3% of the sample of unilaterally hearing-impaired children experienced significant academic problems that required either resource assistance or grade repetition. These data have been validated from a number of other studies in the USA and Europe.[5,8,76,77]

It appears that teachers also perceive children with unilateral hearing loss to be at risk in school. In a study by Dancer, Burl and Waters,[78] children with unilateral hearing loss received lower teacher ratings than their peers on the Screening Instrument for Targeting Educational Risk (SIFTER) in the areas of academics, attention, communication, participation and behaviour.

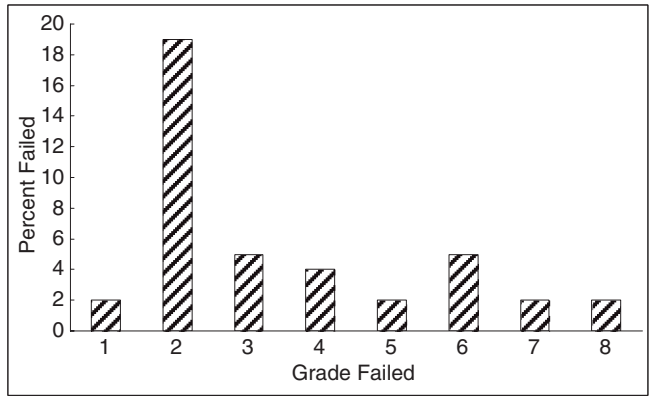

Figure 14.4 The percentage of children who failed each grade. Re-drawn from Bess et al.[4]

Table 14.1 Data from Bess[66] comparing children with unilateral hearing loss and their matched controls on the Wide Range Achievement Test (WRAT).

WRAT (standard scores)	Unilateral group	Normal group	Significance
Word recognition	100.4	109.3	*p = 0.03*
Spelling	97.5	107.5	*p = 0.01*
Arithmetic	99.2	96.3	NS*

*Not significant.

Performance on academic achievement tests has also been examined in children with unilateral hearing loss. Bess[66] compared children with unilateral hearing loss and their matched controls on the Wide Range Achievement Test (WRAT) and revealed that the children with hearing loss showed significantly greater problems than the normal-hearing children on word recognition (decoding) and on spelling. No differences were found between the two groups, however, on arithmetic (a nonverbal task – see Table 14.1).

With regard to the relationship between the degree of unilateral hearing loss and educational risk, findings from various studies are mixed. Some studies suggest that children with severe to profound hearing loss are at greater risk for academic problems than children with mild to moderate unilateral hearing loss.[3,8] Other studies, however, have not found a relationship between the severity of unilateral hearing loss and risk of academic problems.[6] Nevertheless, all of these studies suggest that even children with mild unilateral hearing loss are at greater risk for academic failures than their normal-hearing peers.

Finally, it appears that right-ear-impaired subjects are at greater risk for academic failure than left-ear-impaired subjects,[3,8] In fact, according to Oyler and Matkin,[8] unilaterally hearing-impaired children with right-ear impairment are five times greater at risk for academic failure than children with left-ear impairment.

Language and cognitive skills

Many children with unilateral hearing loss exhibit good language skills, and the children who do have deficits tend to exhibit ones that are subtle – two facts which make it difficult to propose standard early intervention recommendations for this population. For example, Kiese-Himmel[68] collected retrospective data regarding language development milestones from parents of children with unilateral hearing loss and found, overall, that the children were not delayed in acquisition of their first spoken word, but they were an average of 5 months delayed in the acquisition of their first two-word phrase. Sedey, Stredler-Brown and Carpenter[77] reported outcome data on 15 children with unilateral hearing loss on a battery of speech and language tests. Ten (66%) of the children demonstrated normal language development, one (7%) had a borderline language delay and four (27%) had significant language delays. Klee and Davis-Dansky[79] compared performance on a battery of standardized language tests of 25 children with unilateral hearing loss to a matched control group of 25 children with normal hearing. No significant differences were found between the two matched groups on any of the measures included in the language battery.

The Klee and Davis-Dansky[79] study also examined cognition of children with unilateral hearing loss. A comparison of performance scores on the WISC-Revised for unilateral-hearing-

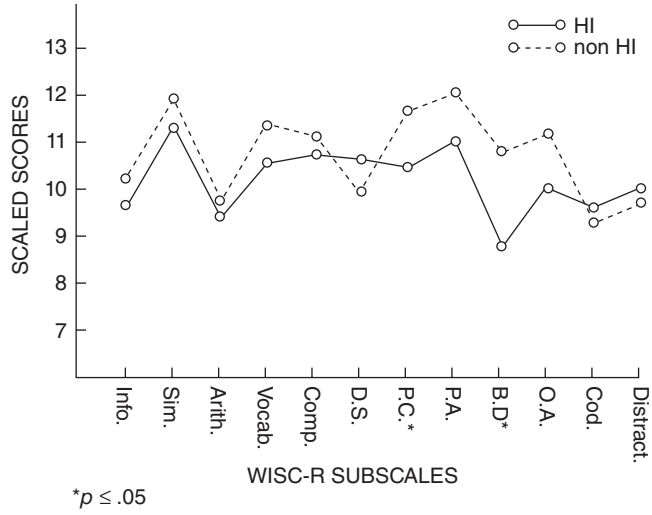

Figure 14.5 Data from Klee and Davis-Dansky show WISC-R scores for a group of children with mild hearing loss and a matched group of children with normal hearing.

impaired children ($n = 25$) and a matched group of normal-hearing children is summarised in Figure 14.5.

It is seen that few differences exist between the two groups; significant differences are noted only for the subscales picture completion and the block design. However, an analysis within the group with unilateral hearing loss revealed that (1) children with severe to profound hearing losses exhibited significantly lower full-scale IQs than children with the milder hearing losses and (2) children with unilateral sensorineural hearing loss who failed a grade in school exhibited verbal IQs significantly lower than unilaterally hearing-impaired children who had not failed a grade.

Niedzielski, Humeniuk, Blaziak and Gwizda[80] also used the WISC-R to measure intellectual skills in children with unilateral hearing loss and reported full-scale IQ levels in the normal range, but found that the side of the hearing impairment had a significant effect on performance. Specifically, children with right-sided hearing loss scored more poorly than children with left-sided hearing loss on the verbal subtests of similarities, vocabulary and comprehension. In contrast, on the non-verbal subtests of block design and object assembly, the children with left-sided hearing loss scored more poorly than the children with right-sided hearing loss. Earlier studies also support the influence of right- versus left-sided impairment in children with unilateral hearing loss. Hartvig Jensen, Børre and Johansen[81] reported poorer performance on the similarities and auditory digit span subtests of the WISC by children with right-sided unilateral hearing loss than children with normal hearing and children with left-sided unilateral hearing loss. On the Rapid Alternating Stimulus Naming (RAN) test, a verbal test reported to be highly predictive of future reading ability, children with right-sided impairment performed more poorly than children with left-sided impairments and children with normal hearing on a number of the subtests. It has been speculated that the performance deficits on verbal tests of children with right-sided hearing impairment on verbal tests may be attributed to the reduction of auditory input to the left cerebral hemisphere, which is the dominant hemisphere for language in most individuals.[80–81] Collectively, these studies suggest that children with right-sided

impairment are at greater risk for cognitive deficits and academic failure and may require closer monitoring than children with left-sided unilateral hearing loss.

Psychosocial

Finally, many children with unilateral sensorineural hearing impairment also exhibit functional difficulties. Bess and co-workers[4] noted that unilaterally hearing-impaired children experienced difficulties in such general areas as dependence/independence, attention to task, emotional ability and peer relation/social confidence. Some unilaterally hearing-impaired children misbehave to gain attention and often appear frustrated or anxious. In the peer relation/social confidence category, children with unilateral hearing loss were more often rated being aggressive towards peers and not initiating interaction with their peers. Overall, when teachers were asked to rate whether the children were average, above average or below average academically, there were marked differences between the children with unilateral hearing loss and those with normal hearing. Only 39% of the hearing-impaired group versus 53% of the normal-hearing children were rated as average. Twenty-two per cent of the hearing-impaired children versus 42% of the normal-hearing children were rated as above average. In direct contrast, 39% of the hearing-impaired versus 5% of the normal-hearing children were rated as below average. In another study, Bess, Dodd-Murphy and Parker[2] examined the functional status of school-age children with minimal sensorineural hearing loss – many of these children had unilateral losses. Children with minimal sensorineural hearing loss exhibited greater dysfunction than normal-hearing children on such psychosocial domains as behaviour, energy, stress, social support and self-esteem.

IDENTIFICATION AND MANAGEMENT OF CHILDREN WITH UNILATERAL SENSORINEURAL HEARING LOSS

Conventional management of children with unilateral hearing loss has been limited. In fact, preferential seating and the use of frequency modulation (FM) systems have historically been the only mechanisms available to children with unilateral hearing loss. However, in view of the current data, it is evident that a more aggressive approach to identification and intervention is needed. Whilst many children with unilateral hearing loss may do quite well with little or no intervention, there does appear to be a cohort of children with unilateral hearing loss who need more specialised intervention.

Early identification and appropriate intervention of unilateral sensorineural hearing loss in children may serve to minimise any disparaging effects on academic and social-emotional development. In addition, if we could target children with unilateral sensorineural hearing loss who appear to be at greater academic or functional risk and in need for early intervention, educational resources may be distributed more prudently. Three disciplines of professionals are likely to encounter children with unilateral sensorineural hearing loss – audiology, speech-language pathology and psychology. The following review represents some identification and management considerations for each of these disciplines.

Audiological considerations

Early intervention efforts are dependent upon early identification of unilateral sensorineural hearing impairment. As previously noted, unilateral sensorineural hearing losses have

traditionally been identified much later in life than bilateral hearing loss due to the inconspicuous nature of the disability. However, with the onset of universal neonatal hearing screening, children with congenital unilateral hearing loss that is at least mild in severity are being identified at birth. Beyond newborn hearing screening, the hearing loss ideally should be identified before the child enters school, preferably by 2 years of age. It is important to remember that behavioural assessment procedures conducted in sound-field settings are not sufficiently sensitive to identify unilateral impairment, and should, therefore, be supplemented with behavioural assessment under earphones and/or electrophysiological measures, such as immittance, otoacoustic emissions and auditory evoked responses.

When a child is identified with a unilateral hearing loss, otological follow-up is recommended. Medical evaluation of the hearing loss may yield important information for monitoring the normal and impaired ear. For example, if the hearing loss is attributed to an enlarged vestibular aqueduct, some recommendations for protecting the normal-hearing ear may be provided. Furthermore, middle-ear status should be closely regulated, especially in the early years. A conductive overlay, secondary to otitis media with effusion can cause significant problems for children with unilateral sensorineural hearing loss, particularly if the good ear is affected. This is especially critical for younger children who are most prone to experience recurrent bouts of otitis media, and are typically transitioning through a time of rapid speech and language learning.

Decisions regarding audiological management of unilateral sensorineural hearing loss should be made upon a case-by-case basis, depending upon the individual needs of the child. Therefore, deliberate audiological, speech-language and educational monitoring of the child's progression is essential. If a child is performing well in school and seems to be adjusting well to the impairment, then preferential seating in the classroom and routine monitoring of his or her progress may be sufficient. For many children with monaural hearing impairment, however, more aggressive intervention is warranted.

Effective management of children with unilateral hearing loss will require a communicative link between the audiologist and the child's parents, teachers, physicians and other professionals who come in contact with the child. Each of these individuals obtains important information about the child, and their expertise is essential to maximise the child's education and intervention. Audiologists have expertise in the specific nature of the child's hearing impairment and are, therefore, in a good position to provide practical information and recommendations to parents and teachers. Furthermore, they can serve as an advocate for the child by attending Individualized Education Plan (IEP) meetings and making specific suggestions regarding classroom seating, amplification, the acoustical environment and the possible needs of speech-language assessments.

Conversely, the child's teachers and other professionals (e.g. child-care providers or speech-language pathologists) have an abundance of knowledge about a given child that may not be readily available to the audiologist, such as the child's academic progress and social skills. In addition, because they observe the child on a daily basis, teachers, speech-language therapists, and child-care providers may be the first to recognize a change in performance suggesting a change in hearing status or the need for more aggressive management. Finally, parents may be the most instrumental experts on their child. Children interact with their parents off and on many hours of each day; therefore, parents are sources of abundant information about their child and typically are the most vested in the child's success. Parents should not be treated as onlookers, but should be encouraged to become actively involved with their child's audiologic and educational management. To a large extent, the child's

success in school will depend upon the cooperation, understanding and support given by parents.

Depending on the severity of the hearing loss, the child may use a hearing aid on the impaired side, although the efficacy of such fittings has not been thoroughly researched. Fitting a hearing aid to the impaired ear may have effects that extend beyond simply providing amplification. It is possible that leaving an impaired ear unaided may deprive the ear of important stimulation and have negative effects on auditory development. Previous research has shown that speech recognition abilities can decrease in the unaided ear for individuals with bilateral hearing loss but unilateral amplification.[82] Given the potential for auditory deprivation, providing auditory stimulation via amplification to the impaired side may be worth considering. Researchers speculate that binaural hearing is a skill learned early in childhood and cannot be recovered later in life.[83] It is also possible that aiding a unilaterally impaired ear will facilitate the development of binaural neurons. Research to date has not confirmed a critical or sensitive period for the development of binaural hearing in humans; however, animal research suggests that a critical period exists for the development of binaural interaction in newborn rats.[84]

As previously discussed, children with unilateral hearing loss are likely to have the greatest difficulty listening in adverse listening conditions but have little difficulty in quiet conditions. Intervention, therefore, should address listening conditions with high levels of background noise (e.g. the classroom) in which these children experience the most difficulty. Unfortunately, hearing aids do nothing to rectify a poor S/N ratio or reverberation; these devices merely amplify all sound in the environment. Therefore FM technology is the preferable choice for children with unilateral sensorineural hearing loss in classroom environments.

The purpose of FM systems is to improve the S/N ratio reaching the child's ear. This goal is achieved by placing a microphone on the person speaking and transmitting his or her voice via FM signals to a headset. This system maintains an optimal S/N ratio by allowing the child to receive the teacher's voice from any location in the room at an intensity that is stronger than the ambient noise.[32] If fitted properly, a personal FM system can improve the S/N ratio by 20–30 dB.[85] The situation is comparable to the teacher speaking within 6 inches of the child's ear, no matter where the teacher or the child is located in the classroom.[86] FM systems significantly enhance the speech recognition abilities of children with unilateral hearing loss in classroom-like listening conditions.[87,88] In order to minimise the risk of over-amplification, it is important that an audiologist fit and maintain these devices. It is recommended that the FM system be coupled with an open earmould and that the high-frequency gain is set at 12 to 15 dB at one-half the volume rotation, with an SSPL-90 not to exceed 105–100 dB. Of course, the use of sound-field amplification is another effective way in which to overcome the problem of poor acoustical conditions.

Speech and language considerations

Although unilateral hearing loss does not always produce obvious effects on speech and language acquisition, children with monaural hearing impairment should be closely monitored for subtle speech and language problems. Customary protocols for speech-language evaluations for children include both screening and full-scale assessments. The purpose of screening is not to diagnose a speech or language problem or to make specific recommendations, but to identify children on whom more comprehensive testing is warranted. Screening can take the form of standardised screening tests, parent referrals or professional referrals. It is important to note that formal screening tests may not be sensitive to some of the subtle language problems

that children with unilateral sensorineural hearing loss may exhibit. Though slight, these problems may have detrimental effects on how well the child copes with the demands of the classroom where verbal skills such as reading, writing, and oral discussion are emphasised. It is recommended that information from parents and teachers regarding the child's progress supplement formal screening tests.

A full-scale speech-language evaluation is indicated if a child fails a screening test or if there is concern from the child's parents, teacher, audiologist, or physician regarding speech or language development. Comprehensive speech-language tests serve to ascertain the child's current level of speech and language functioning, to diagnose a speech or language disability, and to determine appropriate intervention strategies. Full-scale speech and language tests should assess language production (e.g. spontaneous language sample, or formal articulation/expressive language tests), receptive vocabulary, syntactical understanding, language comprehension, and if necessary, non-verbal cognitive functioning.

Like standardised screening tests, comprehensive language batteries can also be insensitive to the subtle speech-language deficits that children with sensorineural hearing loss may experience. For this reason, it is recommended that specific information from parents and teachers regarding the child's speech and language development be obtained to complement formal test batteries.

It is possible that speech-language deficits in many children with unilateral sensorineural hearing loss are not the result of the hearing loss, per se, but secondary to the causal agent of the hearing loss. For example, research has indicated that in a significant percentage of children with unilateral hearing loss who exhibit speech-language or educational deficits, the suspected aetiology of the hearing loss was bacterial meningitis, viral infection or other casual factor associated with neurological damage.[26] These aetiologies may also have considerable behavioural impact on the child. The importance of a thorough case history, therefore, cannot be overstated in assessing the child's current state of functioning and risk for developing speech-language, academic or social/behavioural problems. These factors should also be considered when constructing intervention strategies for the child.

Educational and social considerations

As previously discussed, many children with unilateral sensorineural hearing loss will encounter significant academic and/or psychosocial problems requiring comprehensive assessment and resource services. As with the audiological and speech-language management, a thorough case history should be the foundation of the assessment. The case history should tap into medical information from the pre-natal period, past illnesses which might have some effect upon the child's learning or behaviour, social/family data, records of school performance and descriptive social/behavioural information. The case history may suggest some aspect of the child's development that may be related to the current problems and may provide some information regarding the specific nature of the child's difficulties.

There exists no universal assessment battery for children with unilateral hearing loss; therefore, the assessment should be based on a hypothesis-testing approach. That is, the battery of assessments should be based upon the presenting symptoms of the child and should be intended to evaluate a specific hypothesis regarding the child's strengths and deficits. In addition, the battery of assessment should be completed with the end goal of obtaining a profile of documented strengths and weaknesses for each child that can form the basis for future educational planning. One commonly used tool for evaluating educational performance for children with all types of hearing loss is the SIFTER.[89]

Once diagnostic information on the specific nature of the child's deficits has been obtained, educational programming becomes the key to successful intervention. Since the passage of Public Law 94–142 in the United States, now the Individuals with Disabilities Education Act (IDEA), public schools have been required to provide a variety of special education services, tailored to the individual needs of the child. These services may include self-contained classroom placement, resource training for part of the day combined with a regular classroom placement for the remainder of the day, or itinerate services from a resource teacher who provides consultation to the child's regular classroom teacher regarding appropriate educational modifications. Of paramount importance to the success of educational intervention is the involvement of the child's family. Under IDEA, parents have the right, through the IEP, to actively participate in the planning of the goals and types of services their child receives. Audiologists should inform parents of their rights and should empower them to become advocates for their child.

CONCLUSION

Although not every child with unilateral hearing loss will exhibit communication or academic problems, research has indicated that many children with unilateral hearing loss will experience significant problems. A recurring profile for unilaterally hearing-impaired children who are at risk for communicative and psychoeducational problems include (1) early age of onset, (2) perinatal or post-natal complications, (3) severe to profound hearing loss and (4) right-ear impairment. For these children, preferred seating is often not sufficient to effectively manage their educational needs. Children with unilateral sensorineural hearing loss comprise a heterogeneous group with variable needs, and therefore, require individualised assessment and intervention strategies. Planning of appropriate education strategies depends on detailed evaluation and input from professionals of multiple disciplines. Intervention efforts should be based on the specific strengths and weaknesses of the child and should be modified according to the child's progress. Finally, the involvement of parents is an essential component to the successful intervention of any child with unilateral hearing loss.

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15 Medical management of the deaf child in a multidisciplinary context

B. Mac Ardle and C. Munro

INTRODUCTION

Management of deaf children involves many professional disciplines and agencies at different stages of the child's life. Professional teams may not be based within the same organisation, share the same working principles or receive funding from the same sources.

Effective management should enable children and their families to experience seamless service from professionals working with the family. To achieve this goal, professionals across the disciplines must be highly motivated and effective in their communication skills and have a good understanding of the input the families are receiving from other professional groups.

The immediate professional team focusing on management of deafness for infants and children is likely to include an audiological physician or paediatrician with a special interest in childhood deafness or an otorhinolaryngologist, audiologists, and teachers of the deaf (TODs). How these professionals work with each other, and with the individual family and child, will have significant implications on short- and long-term outcomes.

This chapter will provide a framework for this complex multidisciplinary management, the context in which professionals may be working, the active inclusion of the parents in all aspects of planning and decision making, and how emotional and individual needs of the families and children can be effectively understood and met. The role of the audiological physician within this context is expanded in this chapter.

THE CONTEXT IN WHICH A DOCTOR IS MANAGING DEAF CHILDREN

There are a variety of doctors who work with the deaf child and their families. These may include audiological physicians, paediatricians with a special interest, phoniatricians and otorhinolaryngologists. As well as having excellent generic skills,[1] there are highly specific competencies that the audiological physician or paediatrician must acquire in order to deliver the best care to children and their families.

THE CONTEXT

In United Kingdom in the past decade, several external factors have greatly influenced the role of the doctor. These include:

- Increasingly explicit description about the duties of a doctor.[1] There is an expectation that doctors will implement evidence-based medicine with development of clinical care pathways and follow best-practice guidelines.
- Delivery of services to children which are influenced by Department of Health initiatives some of which include the National Service Framework for Children[2] as well as increasing patient input to care.[3]
- Production of quality standards from voluntary organisations such as the National Deaf Children's Society (NDCS), who are active advocates for deaf children and their families. NDCS produce Quality Standard documents which set standards for delivery of 'family friendly services' to hearing-impaired children and their families.[4]
- The introduction of the national Newborn Hearing Screening Programme (NHSP)[5] has presented physicians with a new set of challenges which include correct interpretation of audiological assessments, earlier interventions and earlier aetiological diagnosis of deafness in very young babies.
- Developments in digital hearing aid technology and earlier age of cochlear implantation have also improved the quality of habilitation and outcomes for the deaf child.

All of these external drivers raise parental expectations about the standard of care delivered by the doctor and multidisciplinary team working with the deaf child and his or her family.

THE ROLE OF THE DOCTOR

What are the specific skills and knowledge required to work with the deaf child and their family?

In order to be effective, the doctor working with deaf children and their families requires specific knowledge, skills and attitudes. In the UK, doctors training in audiological medicine obtain their knowledge base by completing a master's degree in audiological medicine as well as completing a specific curriculum and undergoing regular assessments of competency[6] before being awarded their Certificate of Completed Training (CCT) in audiological medicine. Many other doctors who work with children acquire further knowledge and skills as part of their continuing professional development.

It is essential that doctors must also work in teams and this includes respecting the skills and contributions of colleagues; communicating effectively with colleagues within and outside the team; ensuring that patients and their parents understand the role of the doctor and their responsibility within the team and who is responsible for each aspect of patient care; participating in regular reviews and audit of standards and performance of the team, taking steps to remedy any deficiencies; and supporting colleagues who have problems with conduct or health.[1]

Audiological physicians work in a team with experienced paediatric audiologists and TODs.

Paediatric audiologists

Paediatric audiologists provide detailed assessments of hearing and balance function using a wide variety of objective and subjective tests following national protocols.[5] They also prescribe

hearing aid systems according to national guidelines[7] and regularly monitor both hearing aid function using both real ear measurements as well as behavioural assessments including free-field aided responses and, more importantly, speech perception testing to assess outcomes from intervention. The paediatric audiologist is a key member of the multidisciplinary team, and it is essential that the audiological physician has a close working relationship with the paediatric audiologist.

Teachers of the deaf

TODs work in a variety of settings – from within the child's home with the child and the family to mainstream inclusive-based teaching and advice, and special school provision. The majority of TODs are employees of the local education service (LEA). However, TODs also work for the health service on UK paediatric cochlear implant multidisciplinary teams.

TODs are educational specialists who have expertise on how deafness can impact on communication and language acquisition development from birth onwards. Additional postgraduate training is a requirement for the profession.

The role of a TOD includes:

- early input to support family knowledge, understanding and feelings about deafness;
- support of the parent–child interaction and language access across the range of communication approaches;
- establishing the use of amplification, and associated early listening skills/sound awareness;
- specialist teaching approaches in many educational settings (mainstream, with inclusion, hearing-impaired units, special school settings);
- training and support for mainstream teachers/support staff for curriculum differentiation and deaf awareness/accessibility;
- effective introduction of additional or alternative amplification systems;
- input for Individual Education Plans (IEPs) and statutory assessment.

Multiprofessional and agency working is a fundamental professional skill.

TODs and speech and language therapists often work closely together, with distinct, but overlapping roles.[8]

Notification of identification of deafness goes to the local TODs team and should (with parental permission) take place within 1 day of identification of deafness.[4]

WORKING IN A MULTIDISCIPLINARY TEAM

Context framework for professional practice

Professionals working with families with deaf children must be well informed and be able to work effectively within established good practice, legal and ethical guidelines.

The introduction of the NHSP has sharpened awareness of the fundamental issues associated with family management of young deaf children – parental access to clear and accurate information, parental understanding of the impact of hearing loss and communication

approaches, effective communication between professionals and availability of quality habilitation and specialist support locally.

These fundamental principles recognise that families are individual in their feelings about hearing loss, and how they would wish their child to be managed. Services should be aware of the meaning of being 'family friendly hearing services'.[9]

Overview of initiatives to support young children (England)

Government mechanisms to achieve coordinated services for families with disability across the professions and support services are developing using the common assessment framework.[10]

The Early Support Programme[11] for pre-school children seeks to provide a coordinated approach for monitoring the development of young children across England. A monitoring protocol for deaf children[11] is in use across many specialist educational services.

Sure Start Early Excellence Centres provide a one-stop-shop integrated education and day care, and aim for multiagency working, social inclusion, and local school links in the early years. It has a partnership approach to working with families and bringing together early education, childcare, health and family support. This framework is described in 'Birth to Three Matters'.[12]

The NDCS's guidelines and quality standards for professionals working with deaf children and their families seek to provide a high level of consistent professional practice across England. This effective working practice should 'lessen the impact on the family' and provide 'appropriate management'. Quality standards consider 'sensitive follow up care and appropriate habilitation [to be] available' as fundamental to the process.[4]

The Disability Discrimination Act (DDA) 2005 requires that service providers ensure equal access to their service for the disabled. In the context of hearing services and management of deaf children, one example would be the availability of language interpreters (spoken and signed) as fundamental to fulfilling the legal obligation.[13]

The Special Educational Needs (SEN) Code of Practice[14] provides a framework for developing strong partnerships between parents and professionals and promotes a consistent approach to meeting special educational needs, placing the child at the heart of the problem.

Parental informed choice

Early identification of deafness and intervention requires that health education and social services work with families to promote informed choices from the initial stages and onwards.

Making informed choices requires access to honest, accurate and unbiased information. Professionals should be working within an approach where parents (and children as appropriate) are integral to decision making. This level of partnership cannot exist without individual and multiteam commitment to ensure pathways and interventions are jointly agreed and the parents are, by right, central to the process.

Experienced clinicians are aware that families are individual in their skills and abilities to process audiological and medical information about their child and understand the potential

impact on auditory and communication development. Clinicians need to be skilled at listening to parents.

Personal value systems will have a significant influence on parental attitudes to their child's deafness. Parents make decisions within their own context and according to their own values and attitudes. These attitudes will influence management decisions, and in some circumstances may need longer-term discussion and psychological support. However, parental autonomy must be respected.

The process of working with parents towards informed choice at each transition is not the role of one professional. Depending on the child's history there may be numerous other services involved, each offering information and guidance within their specialty. Multiteam working and sharing of information (with parental permission) is required. Communication between involved professionals gives the best opportunity for the parents to access coherent information.[15]

Principles of habilitation of deafness

Habilitation is the means by which the impact of deafness is reduced by parental and professional input. What form habilitation takes depends on many factors which may include: assessment of the nature of the child's hearing loss on daily living both at home and at school, as well as the recording of the impact of the environment on the deaf child's functioning;[16] age at which hearing loss was identified; family circumstances feelings and needs; child's feelings and needs; and additional disabilities the child may have.

To ensure the best outcome decisions about management of the hearing-impaired child should be based on information about hearing levels, assessment of functional listening, developmental progress and educational performance.

All of the aforementioned require discussion with the audiologist, TOD, the child (where appropriate) and parents prior to any agreed interventions.

Recommendations should always include implementation of good hearing tactics by all adults and other children who have contact with the deaf child; when appropriate, each child should be encouraged to develop self-help skills and tactics for coping with their hearing and communication difficulties; and opportunities to meet other deaf children (and their families) to have as role models.

Effective habilitation is developmentally appropriate and responsive to the individual child's functional listening and communication needs. Professional input should be supportive of a collaborative partnership with the parents.

The long-term aims of a habilitative team approach will be to enable deaf children and deaf young people to become confident, self-sufficient and effective in their language and learning abilities.

ESTABLISHING THE LEVEL AND TYPE OF HEARING LOSS AND ITS IMPACT ON THE CHILD

It is essential to establish the degree and type of deafness as well as any possible history of fluctuation or progression in hearing levels in each child. Every behavioural test performed should document the testing conditions and degree of subject cooperation.

On a regular basis, the doctor should observe testing with parents present. Demonstrating to parents a child's performance on speech perception testing and/or sound-field testing may frequently highlight the child's previously unrecognised hearing difficulty.

Assessments should include a range of tests which may include behavioural tests of hearing, tympanometry, acoustic reflexes as well as transient evoked otoacoustic emissions. Using the crosscheck principle is essential in paediatric practice.[17] Further tests may include evoked response audiometry and/or speech perception testing.

Upon the identification of deafness, or in clinical discussions at any point in the child's life, clinicians should be sensitive to changes in level of deafness. Other factors that can have a significant impact on auditory access include changes in classroom environment or teaching style. It is important not to dismiss the link with the hearing issues as improbable, based on level or configuration of loss. Significant life events can have an impact on emotional development and also affect auditory access.

The impact of hearing loss cannot be assessed by the audiogram alone. Assessment of the functional use of hearing for speech discrimination skills in a range of listening situations provide evidence of the impact of a hearing loss.

Assessment of the child's functional use of hearing (depending on the age and developmental level of the child) should be available, usually by the local TOD.

Assessment tools may include:

(1) Listening Inventories for Education UK (LIFE UK) questionnaire.[18] This includes classroom listening scenarios in picture form. The child (7+ years) rates how well they would hear the teacher or peer words – from 'always easy' to 'always difficult', when used pre and post amplification it is an effective measure of functional benefit in school/learning situations for children. This tool is also useful for identifying and managing children and young people who have auditory access difficulties or experience intrusive tinnitus in school.
(2) Screening Instrument for Targeting Educational Risk (SIFTER).[19] There are pre-school, school-age and secondary-aged versions. A teacher questionnaire identifies five areas of school performance (academic, attention, communication, class participation and school behaviour, rated in comparison to peer group) related to auditory access issues and educational risk.
(3) Use of the hierarchy of auditory functioning is helpful in making baseline assessments and monitoring progress in young deaf children and in children with multiple sensory difficulties.[20]
(4) The Listening Progress Profile (LIP)[21] is based on the hierarchy of auditory skills. Abilities are assessed using informal play activities, so it is appropriate for very young children. The scale starts with ability to detect sound (and no sound) to discrimination skills between two familiar names (different syllabic length).
(5) Parental report/observation and the Meaningful Auditory Integration Scale (MAIS)[22] can provide information that may not be evident in a clinical setting. There are three versions of MAIS: a standard one for parents, one for teachers and one for children under 5 (IT-MAIS), which were developed to evaluate meaningful use of sound in everyday situations. The questions cover the child's adaptation to amplification (hearing aids or cochlear implant) to developing auditory skills in everyday situations. The child is rated on a 5-point scale: always, frequently, occasionally, rarely or never.

Table 15.1 Outcome measures for listening pre- and post-cochlear implantation.

Assessment	Pre-cochlear implant	Initial connection	3-month post-cochlear implant
IT-Mais	42%	60%	77%
LIP	14%	66%	85%

Information and training in assessment tools are available from The Ear Foundation.[23] See Table 15.1 for an example of the use of IT-Mais and LIP scores to measure progress in a 2-year-old child with auditory dys-synchrony pre- and post-cochlear implantation.

For some children, a specific request to the local TOD for a structured observation of the child's hearing and listening behaviour in the classroom may be indicated to clarify difficulties and plan for future management.

Children who experience auditory access difficulties related to hearing loss, fluctuations in hearing, difficulties in noise, or other factors, are frequently found to have in place strategies that effectively mask the extent of their problem. An experienced observer is needed to see these strategies being used, and to assess the extent of the difficulty.

To ensure a satisfactory habilitation outcome, especially when considering amplification it is mandatory to have the views of the child, parent and multidisciplinary team before intervention. With some families this may take some time.

Historically, the potential impact of unilateral, mild and fluctuating losses (sensorineural and conductive) was regarded as insignificant and not requiring intervention. More recently, this stance has been challenged by studies that show association between these hearing issues and lack of achievement academically, socially and linguistically.[24,25]

In order to maintain best practices, it is important for doctors to be aware of the limitations of test results, for example, in delayed auditory maturation in pre-term infants, as great care is required in interpreting both objective and behavioural test results. Assessment of children with complex special needs also provides a challenge to the physician and team.

Many children with hearing loss will also have vestibular areflexia/hypofunction and knowledge of how this may impact on activity and participation is essential.

COMMUNICATION APPROACHES

Professionals need to be aware of the different communication approaches available for deaf children. It is likely a child will use a range of communication approaches, over time, as their language and learning needs change. When parents choose a particular approach it is essential the family and the child experience good practice in level of support and the use of such approaches.

Communication approaches may include auditory-oral approaches, sign bilingualism and total communication.

Practical expression of total communication at an early years level may include use of natural gesture and early sign, alongside establishing hearing aid use, at home, with support from a visiting TOD.

For most families, their main source of information about communication options will be from the local TOD teams. Information should be unbiased and balanced whatever the source. Professionals working with deaf children and their families should be aware of the polarisation of views still found when communication systems and deafness are discussed.

Communication approach over time will influence school placement, and some parents will experience local education policies that make true educational choice difficult to achieve. Some families may find themselves appealing for alternative placements.

Parents will continue to need pragmatic and sensitive support to work out which direction(s) are right for their child and to understand that these choices may change over time in tune with their child's needs.

Some fundamental principles for professionals to keep in mind are:

- Does this child have optimal amplification? What about functional responses/receptive and expressive language skill progression?
- Where hearing levels impact on access to sounds for speech or additional needs impact on access to making sense of sound, does the child have communication support through visual communication as well?
- How do the parents feel about learning new ways to communicate, and do they have family or other support? Are the parents and the child able to communicate with each other now, and will they be able to do so in the future?
- Does educational placement use an appropriate communication mode for access to learning for the child?

IDENTIFICATION OF DEAFNESS FOLLOWING NHSP

Implementation of newborn screening nationally offers early identification of deafness, and with this opportunity, especially for children with severe to profound deafness, to obtain significant benefit for language development.[26]

For health and education teams working with the parents of these infants there has been a sharp professional learning curve as a result. Parents are inevitably at their most vulnerable in the few weeks following delivery, and for most there are no outward signs of deafness in their newborn baby.

Management by health and education hearing services must be supportive and attentive to the individual family and child's emotional, psychological and social needs. This is part of the habilitation process – outcomes for parents and children are more positive – where early interaction with professionals has been grounded in family-focused initiatives.[4]

Models of such team approaches provide helpful insight.[27] Table 15.2 describes our approach at the Nuffield Hearing and Speech Centre at the first assessment appointment following diagnostic auditory brainstem response testing.

Principally, the parent is recognised as the key influence and provider of habilitation, with support and guidance from the clinic-based and local professional team for babies and young children.

Table 15.2 Initial stages of multidisciplinary management of the newly diagnosed deaf child post-NHSP.

First assessment	**Time line: within 2 weeks of referral (max 4 weeks)**	**Time taken: $2^{1}/_{2}$–3 hours**
Consultant in Audiovestibular Medicine **Action:** • Team discussion prior to appointment • Full clinical history and examination • Behavioural observational audiometry if appropriate • Interpretation of the findings in the light of the clinical presentation • Discussion of abnormal results + management plan with audiologist + TOD • Counsel the parents appropriately + discussion of management plan • Initiate aetiological investigation • Ensure the parents have a contact number to phone and ask questions • Contact the family doctor and referrer within 24 hours by telephone and/or letter • If appropriate, child's name to be placed on newly diagnosed deaf children's list for discussion of management plans at multidisciplinary team meetings (MDT) • Participate at weekly multidisciplinary meetings to discuss newly diagnosed deaf children.	Audiologist/Scientist **Action:** • Team discussion prior to appointment • Otoscopy • Brief history • Auditory brainstem response testing • High-frequency tympanometry • TEOAEs • Record results in notes • Feedback/discussion of results with audiological physician • Participate at weekly multidisciplinary meetings to discuss newly diagnosed deaf children.	Teacher of the Deaf (TOD) Clinic/hospital based **Action:** • Team discussion prior to appointment • Alerted and involved as appropriate following clinical and test results • To provide family/child-centred information and advice • Counselling and guidance • Ensure the parents have a contact number/email to phone and ask questions • Liaise as appropriate with local TOD, with parental consent • Child's name to be placed on newly diagnosed list for discussion of management plans at weekly multidisciplinary meetings • Participate at weekly multidisciplinary meetings to discuss newly diagnosed deaf children.

THE ROLE OF THE DOCTOR POST-NHSP

Audiological physicians and paediatricians are now seeing deaf children from birth. This has presented a unique series of diagnostic challenges which include establishing with certainty the level and type of hearing loss. National protocols for hearing assessment of no clear response on NHSP babies are available.[5] In general, severe to profound hearing losses are more easily identified when compared with milder hearing losses. However, confirmation of deafness can be delayed up to 8 months of age for milder hearing losses.[28] Babies presenting with features of auditory dys-synchrony, delayed auditory maturation or auditory neuropathy require very careful evaluation prior to any suggested habilitation or investigation.[5]

It is particularly important to follow up with babies with glue ear post-NHSP as it is likely to be more persistent. It is important to remember that the child with glue ear may have an atopic tendency. Always check the palatal integrity especially if a craniofacial syndrome is suspected. Rarer causes of glue ear include children with immunodeficiency or ciliary dyskinesia.

It is important that the doctor understands the ethical dilemmas posed by a number of interventions such as newborn hearing screening, genetic testing and pre-natal diagnosis as well as the implications of interventions such as early cochlear implantation and the stress this places on families.

The doctor must respect the views of Deaf culture. In addition, hearing parents of deaf children need to be informed about the perspective of the Deaf community, and opportunities for parents to meet a broad spectrum of families with deaf children should be facilitated as early as possible for each child after confirmation of diagnosis of deafness. Access to other families may be possible via the local TOD service.

Doctors must demonstrate excellent communication skills at all times and in particular at the time of confirmation of diagnosis of deafness. Standards for 'sharing concerns' are set for doctors to follow.[29,9]

Audiological physicians must ensure that they allocate sufficient time in their work programme to accommodate the frequent consultations that are required both for the deaf child and their family as well as time for multidisciplinary team discussions. Time should also be allocated for wax removal as good ear mould fit is essential for all children.

MEDICAL INVESTIGATION OF DEAFNESS

Aetiological investigations

NHSP has resulted in earlier introductions of aetiological investigations which lead to much earlier diagnoses of many conditions, e.g. congenital cytomegalovirus (CMV) infection, connexin 26 deafness, Pendred's syndrome, Usher syndrome, mucopolysaccharidoses and other metabolic conditions. Some of these diagnoses may have genetic implications, and this places a further burden on parents who may be at the initial stages of accepting their baby's deafness. Families therefore require easy access to the doctor who has special responsibility for managing children referred from the NHSP.

Information about aetiological investigations should not be biased and should given in language that the family can understand.[30] In addition, families should be given clear information about the possible implications of positive test results. The doctor must respect the parents' decision if they decide against investigation or recommended interventions.

Doctors require a high level of skill in history taking, developmental assessment and physical and dysmorphology examination. Integrating findings from history and examination with the audiovestibular assessment enables the doctor to make a correct diagnosis of syndromal or non-syndromal deafness.

Understanding normal development and knowing about how deaf children develop language and communication is essential. The latter will ensure that the most appropriate tests for assessment are requested and that there is a correct interpretation of the level of hearing loss as well as its impact on the child, especially those with complex needs.

GUIDELINES FOR GOOD PRACTICE ON AETIOLOGICAL INVESTIGATION OF PERMANENT HEARING LOSS IN CHILDREN (ADAPTED FROM THE BRITISH ASSOCIATION OF AUDIOLOGICAL PHYSICIANS GUIDANCE [WWW.BAAP.ORG.UK])

Level 1 investigations

Level 1 the timing of investigations should be considered for every child with bilateral severe to profound permanent sensorineural deafness

Timing of investigations will depend on several factors, including the family's agreement to proceed with tests.

In cases where the aetiology has not been found, further aetiological investigations may need to be arranged and for some children may need to be repeated in the future.

Level 1 investigations include the following:

(1) Detailed paediatric history:
- onset of audiovestibular symptoms
- pregnancy, delivery and post-natal period
- developmental milestones including speech, language, motor milestones as well as social development
- history of exposure to risk factors, e.g. exposure to noise, ototoxic medications/radiation, head injury, ear disease, meningitis, bacterial and viral illness, immunisation status
- family history of deafness or risk factors associated with hearing loss in first- and second-degree relatives
- document consanguinity and always identify the ethnic origin of the family.

(2) Clinical examination:
- height, weight and head circumference; inspection of craniofacial region and physical measurement
- examination of the ears, neck, skin and nails, limbs, chest, abdomen and gait
- development assessment.

(3) Family audiograms of parents and first-degree relatives.

(4) An electrocardiography (ECG) for prolongation of the (corrected) QT interval is essential in children with evidence of vestibular hypofunction which may manifest in form of delayed motor milestones e.g. head lag, delayed sitting without support and delayed age at walking.

(5) An ophthalmological assessment should include assessment of visual acuity, fundoscopy and discussion with the ophthalmologist about indications for electro-retinography if motor milestones are delayed to detect Usher type 1, unless the child has adequate explanation for vestibular problems, i.e. vestibular malformation.

(6) Urine examination for microscopic haematuria.

(7) Cytomegalovirus screen:
- <1 year of age:
  Urine CMV DNA Polymerase Chain Reaction (PCR) × 2 (separate occasions):
  if positive, request neonatal blood spot for CMV DNA testing

- >1 year of age:
  Urine CMV DNA PCR:
  if positive, request neonatal blood spot for CMV DNA testing
  if negative, request IgG or serum IgG:
  if positive, request neonatal blood spot for CMV DNA testing.
- Any age: consider testing mother's serum CMV IgG if not already known.

(8) Blood test for connexin 26 mutations with consent from parents, an explanation that DNA is stored afterwards in lab, that genetic testing can take a long time. Permission should be sought to share results with other family members/professionals.[31]
(9) Magnetic resonance imaging of internal auditory meati or computed tomography scan of petrous temporal bone.

Level 2 investigations

Level 2 investigations will be indicated from history and clinical findings. As with level 1 investigations, timing will depend on the family's readiness to proceed with tests, availability of local test facilities and how well the child can cooperate with tests.

Level 2 in investigations include the following:

(1) Serology to exclude congenital infection and also to include maternal stored (booking) serum
(2) Haematology and biochemistry where clinically indicated, e.g. thyroid function tests indicated if there is a family history of thyroid disease, a goitre is present or there is a widened vestibular aqueduct or Mondini deformity of the cochlea.
(3) Investigation into autoimmune diseases where clinically indicated.
(4) Metabolic screen on blood and urine where clinically indicated.
(5) Renal ultrasound if child has preauricular pits or sinuses, deformity of ear, branchial cleft or cysts; Mondini defect on imaging; permanent conductive or mixed hearing loss.
(6) Clinical photography.
(7) Chromosomal studies are indicated with a history of developmental delay or dysmorphic features (most development paediatricians have an investigation protocol for developmental delay).
(8) Further genetic testing if indicated after discussion with the geneticist; consider referral to clinical geneticist especially if the parents are consanguineous, a syndrome is suspected, the child has multiple problems, or at parental request.
(9) Vestibular investigations; consider in all cases where motor milestones are delayed or where a syndrome, e.g. Usher type 1, Jervell Lange-Nielsen, is suspected as well as in cases of progressive deafness.

It is essential that doctors keep up to date with their practice and constantly review their aetiological workup on the hearing-impaired children as some investigations may need to be repeated, e.g. vision assessments,[32] or as new information about the child or family or new tests become available.

MANAGEMENT OF HEARING AID FITTING

Hearing aid fitting is part of a well-informed and planned habilitative approach decided with parents and, where appropriate, with the child.

At The Nuffield Hearing and Speech Centre, we provide a multidisciplinary approach to hearing aid fitting appointments. Once aiding is agreed with the family, a series of joint appointments are arranged. The child's notes are jointly reviewed by the audiological scientist and the hospital-based TOD prior to the appointment, and the team involved plan in detail how the appointment(s) will be structured appropriately for each family.

The hearing aid fitting appointment includes:

- Discussion with the parents and/or child to confirm parental understanding about the nature of the hearing loss, potential impact on auditory access, benefits/limitations of amplification.
- Initial responses, e.g. eye gaze to amplified sound, are observed and demonstrated to parents using sound makers and voice or the child may self-report.
- Communication and listening ideas are modelled/discussed in the session and written information is provided.
- Discussion about the importance of visual information (from eye-level facial expression to use of sign) to support listening.
- Joint report to local TODs service with parental agreement, including request for updates on progress.

Follow-up appointments may include:

- Parental observation and local TOD reports on auditory responses at home and hearing aid usage – these inform hearing settings and progress.
- Observation of child's functional responses to sound stimuli/voice.
- Appropriate guidance in response to information and discussion of parental concerns and questions about child's communication progress.
- Audiology/TOD reports to local advisors/teachers and request for updates on progress.

These appointments are frequently combined with an appointment with the audiological physician in order to monitor overall progress and feedback results from medical investigations.

MANAGEMENT OF CONDUCTIVE HEARING LOSS

Fluctuating conductive hearing loss is most commonly caused by recurring otitis media.

Fluctuations in hearing levels can have a significant impact on auditory access especially in children with additional sensory impairment. The impact on learning abilities in the school-aged population is well documented. Long-term auditory processing issues are noted even in those children where the hearing levels are subsequently stable.[33]

Management may include:

- Information and discussion with parents about minimising the impact of the hearing loss
- Practical guidance on classroom management to pre-school/school provision, preferably via the local TOD team
- Watchful waiting
- Medical management
- Grommet insertion
- FM system in the classroom
- Temporary amplification.

Permanent conductive hearing loss

Children with a permanent conductive hearing loss require a thorough medical history and examination as well as investigation.

It is important to consider the following aetiologies:

- Craniofacial syndromes, e.g. Treacher Collins syndrome; it is important to arrange detailed imaging of the middle and inner ears as well as renal tract ultrasound.
- Congenital malformation of the outer and/or middle ear structures.
- A history of long-term middle-ear disease.
- Dilated vestibular aqueducts.

Management options may include a headband bone-conductor hearing aid, behind-the-ear hearing aids or later, where appropriate, a bone-anchored hearing aid system alongside specialist advice for management in classroom and at home.

CHILDREN WITH HEARING DIFFICULTIES WHO HAVE NORMAL TESTS OF PERIPHERAL HEARING

Some children are referred to physicians with a tentative diagnosis of non-organic hearing loss. It is essential in these cases to take a very thorough case history to clarify what the nature of presenting complaint is. Clarification about the child's hearing ability as well as concentration, listening and auditory memory is important. In school-age children it is particularly important to establish whether they have any specific learning difficulties. It is also important to establish with non-directive questioning whether the child has tinnitus as it can be provoked during behavioural testing and cause confusion for the child when test stimuli are presented.

Many children find it difficult to cooperate with audiometric assessment and for these children the audiologist requires a lot of skill with the alteration of stimulus type, using ascending techniques and encouraging the child during the testing.

The test battery for such children should include behavioural testing, tympanometry with ipsilateral and contralateral stapedial reflex thresholds, otoacoustic emission testing as well as speech perception testing. Some children may require diagnostic and threshold auditory brainstem testing.

The outcome depends on test results. If the tests of peripheral hearing are normal and the child does not have any relevant features in his history, further investigation is not required. However, if there are features to suggest specific learning difficulties, auditory processing disorder or auditory neuropathy then further audiological and multidisciplinary assessment will be necessary.

Some children may need referrals for assessment by a clinical or educational psychologist, speech and language therapist and/or a paediatrician.

OTHER SPECIFIC ROLES FOR THE DOCTOR

Assessment of children with complex needs

The audiological physician will frequently be asked to assess children with complex special needs and those who are difficult to test. These groups provide a great challenge to assess. It

is important to decide what the expected outcomes from assessment might be as they may well be different from more routine assessments. A holistic developmental approach is essential in order to correctly interpret the results of both behavioural and objective tests and what the results mean for each child. A variety of behavioural assessments with a hierarchical approach, e.g. auditory functioning,[20] auditory behaviour index and early auditory behaviour,[34] as well as development of attention[35] and speech perception skills[36] are helpful in building the child's profile.

There are essential components of the assessment of a child with complex needs. It is vital to collect detailed knowledge of the child's developmental profile including whether the child has additional sensory handicap.

(1) Prior to the appointment – information collection:
- organising a very structured assessment, i.e. sending out questionnaires to parents and caregivers and obtaining detailed reports from the paediatrician before the appointment;
- parental observation of child's response to sound as well as completion of questionnaires about auditory behaviour;
- observations by professionals who are involved with the child as well as completion of questionnaires about auditory behaviour.

(2) During the assessment:
Allocate adequate time for the assessment as it takes considerably more time for these children to adapt to different people and the test environment. Testing and assessment also requires a lot more time. If the child has visual or motor impairment, special consideration should be given to adequate seating and lighting conditions during testing, as well as knowledge of the child's visual ability, visual fields, cortical blindness etc.

Performing the assessment with appropriately trained team members is essential. Modifying the behavioural testing is essential,[37] for instance, by agreeing on what types of stimuli are appropriate and the duration for each stimulus. It is important to acknowledge that many children may demonstrate a slower response to sound and that some children with motor handicaps may not be able to localise the sound source.

It is important for testers and parents to agree on what a response to sound is and whether it was reproducible.

Objective testing of hearing is also an important component of assessment and the crosscheck principle should always be used. It is particularly important to exclude auditory neuropathy in this group of children. In some children with severe brain malformations or damage, the waveforms on evoked response audiometry may be very difficult to interpret.

At the end of an assessment based on knowledge of the child's developmental profile, auditory behaviour and auditory development, as well as audiological test results, it should be possible to make some clear statements about the stage of the child's auditory development and give a clear description of the child's functional hearing.

Management of deaf children with multiple special needs should include advice that will always include good hearing and communication tactics based on the child's developmental and cognitive levels. Decisions about assistive listening devices and/or hearing aids should be informed by consultation with the parents and main caregivers. There are a variety of issues relating to the fitting of hearing aids in this group.[38]

Post intervention assessment of the deaf child who is not making expected progress

With such children a holistic approach is essential. It is necessary to review existing diagnoses and carefully review the history focussing on:

- Whether or not the child and family have accepted the deafness, amplification and intervention programme.
- Audiovestibular function. It is important to remember how insidious the signs of progressive hearing loss can be. An additional fluctuating conductive hearing loss in association with a sensorineural loss can be very distressing for the child and family.
- Changes in behaviour may be a clue to difficulties the child has in coping at school. Environmental issues can impact on listening, e.g. change of teacher or classroom. Insufficient support in the classroom, poor school attendance or inappropriate school placement may all lead to poor progress.
- Childhood depression and poor self-image as well as bullying can have a major impact on a child's progress.

Careful physical examination is indicated.

A review of previous investigations including genetics is essential to exclude the possibility of an undiagnosed syndrome or metabolic disorder, chromosomal disorder, neurological or degenerative disorder or vision impairment.

Many children with hearing impairment have additional special needs and these may include language disorders, autism and behavioural problems, specific or general learning difficulty.

In addition to reviewing the medical diagnosis, further specialist involvement may be indicated e.g. assessment by TOD specialist, a speech and language therapist, clinical or educational psychologist or child psychiatrist. Such assessments may inform changes in educational placement and communication approaches.

Non-organic hearing loss

Non-organic hearing loss has been defined in many ways, e.g. pseudohypoacusis, functional hearing loss, psychogenic deafness, conversion hearing loss or deafness. A preferred definition is 'responses to a hearing test indicating a deficit greater than can be explained by organic pathology'.[39] Jerger and Jerger[40] estimated a prevalence of 7% of non-organic hearing loss in children aged 6 to 17 years. Presentation of erratic audiometric thresfolds may not reflect their speech perception outcomes. Non-organic hearing loss may present at any age and is more common in girls. A typical age of presentation is between 10 and 12 years. It can occur in the following clinical scenarios:

- children with known sensorineural hearing loss
- children with progressive hearing loss
- children with a past history of middle-ear disorder
- children who present with acute unilateral or bilateral profound hearing loss
- children who are experiencing stress either within the family, academic stress, or it may be a presenting feature of child abuse.

History taking needs to be very skilled and requires a holistic sensitive fact-finding approach in which the doctor displays excellent listening skills and is able to identify and empathise with the child's complaint. It is important on system review to detect any pointers to organic disease and to understand the family dynamic as well as what is going on for the child both at home and in the educational environment. A detailed physical examination is essential, and Stenger's test can be helpful in the clinical setting.

Audiological assessment should include pure-tone audiometry, speech audiometry, objective tests including stapedial reflexes, otoacoustic emission testing and auditory brainstem/cortical evoked response testing. Some children may require central imaging.

Management

Austen and Lynch[39] recommend avoiding confrontation with the patient but providing 'a benign explanatory model and the option of a dignified "recovery" '.

A team approach is essential, and some children and their families will require referral to a clinical psychologist. For children with a presentation of a conversion deafness or psychogenic deafness referral to a child psychiatrist is essential.

SPECIAL EDUCATIONAL NEEDS GUIDANCE – INPUT OF DOCTOR

Audiological physicians receive requests to write reports about the child's health and medical profile for education departments. Such reports should contain information presented in a practical way in order to clarify the type of deafness and any associated medical diagnoses. The audiological and medical management should be clearly stated. There should be some description of the impact of the deafness on the child and ways in which access to their peer group and the curriculum can be improved.

Emotional support for children and families

Physicians should be alerted to the needs of families of children (and the child) with deafness at various stages in the child's life:

- at time of diagnosis
- deciding on amplification
- choosing communication mode
- deciding whether cochlear implantation is appropriate
- choosing school placements
- if child develops progressive hearing loss
- if child develops behavioural problems
- if child is not making expected progress
- transition from primary to secondary education
- allowing the young adult to gain independence skills prior to transfer onto adult services.

The doctor should be available to discuss parental and child concerns and act as an advocate for the family. When in agreement, families should be referred to the most appropriate expert local resource.

TRANSITIONAL MANAGEMENT

There should be a clear policy agreed between the paediatric and adult audiology departments about transfer of audiological care. This should include age at transfer and how medical and audiological information is transferred to the adult service.

In addition, written information should be made available to young adults about when they will be transferred and how the adult service is accessed.

Some centres have a transition support worker, often a hearing therapist who has direct contact with the young adult and helps with their adjustment to change.[41,42]

CONCLUSION

The Audiological Physician is an essential member of the multidisciplinary team that provides holistic family friendly services to the deaf child. The physician must have adaptable skills, attitudes and appropriate competence in order to provide a quality medical service that is evidence based and meets national standards. It is essential that parents of deaf children are provided with unbiased information that relates to medical investigation and management as well as surgical intervention. The physician will have a more specific role in assessment of children with complex needs, those with non organic hearing loss, and investigation and management of children with progressive hearing loss. Management of transition of care to Adult services is an increasingly important role for the physician.

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16 Selecting amplification for children

D. Toe

INTRODUCTION

There is no doubt that we live in an age of rapid technological development. The proliferation of digital technology in telecommunications, entertainment and information technology confronts us daily in work and play. Similarly, the pace of technological development in hearing instruments can be overwhelming. Audiologists are faced with a wide array of sophisticated new hearing aid features often accompanied by steep price tags. In many settings, particularly in developing countries, budget constraints are tight, and professionals must make decisions about whether a more expensive hearing instrument will provide significant additional benefit for the children whom they support. The process of managing these choices for children and guiding parents through the maze is challenging but can be simplified by maintaining a clear view of the purpose of hearing aid fitting. That is, to provide each child with optimum access to amplification for the purpose of developing communication skills. This chapter will outline critical aspects in the theory and practice of fitting hearing aids to children.

BASIC COMPONENTS OF A HEARING AID

All hearing aids operate upon similar principles. Modern hearing aids can be categorised as either analogue or digital depending upon how they process the incoming sound signal. A simple block diagram for an analogue hearing aid is shown in Figure 16.1. A microphone converts mechanical acoustic energy into electrical current. The amplifier in the hearing aid increases the amplitude of the electrical current, which corresponds to an increase in the intensity of the sound energy. The receiver or earphone converts the amplified electrical signal into acoustic energy and delivers it to the ear via an earmould (air conduction) or a bone vibrator (bone conduction).

Figure 16.2 shows a digital hearing aid. In a digital aid, the sound is converted to an electrical current by the microphone (an analogue procedure). An analogue/digital converter then turns the electrical signal into a series of binary numbers in the form of positive or negative electrical voltage. The central processing unit of the hearing aid is a computer. It is instructed by the programming computer to manipulate the data. The manipulation of a series of numbers provides great flexibility thus allowing the aid to be adjusted more precisely to meet the needs of the hearing aid user. The computed digital output is converted back to analogue electrical impulses by a digital/analogue converter, which in turn is converted by the receiver back to sound. The computer chip in a digital aid is usually very small, and with significant capacity, allowing a range of features and alternative programmes to be built into the hearing aid.

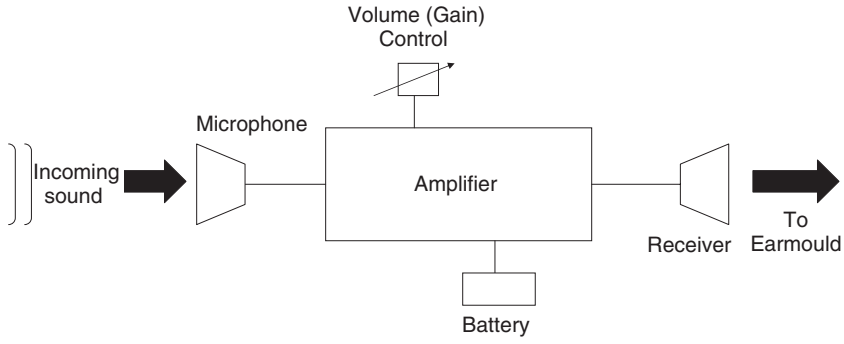

Figure 16.1 Block diagram for a simple analogue hearing aid.

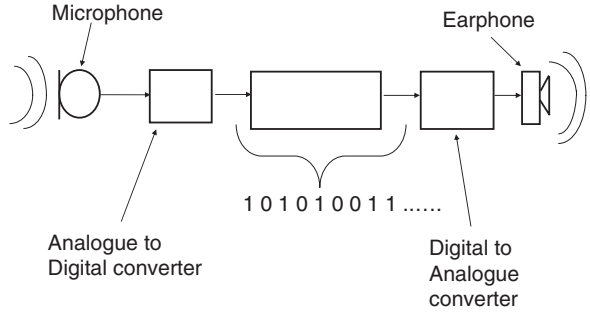

Figure 16.2 Block diagram for a digital hearing aid.

Hearing aids may be worn on the body, whereby the microphone and amplifier are worn on the chest and a separate receiver is worn at ear level, or at ear level. Ear-level aids can be behind-the-ear (BTE), in-the-ear (ITE) or in-the-canal (ITC). The most practical and commonly worn aid for small growing ears is the BTE aid worn with a custom earmould.

PRINCIPLES AND AIMS OF AMPLIFICATION

The main purpose of a hearing aid is to amplify speech so that it is audible and comfortably loud. Audibility is critical – speech that cannot be heard cannot be understood – but it is not everything; audibility is not perfectly correlated with intelligibility. There is little point amplifying sound to the point of discomfort, because it will inevitably lead to aid rejection or a reduced volume setting. Byrne and Ching[1] point out that as hearing loss increases, the contribution made by a given amount of audibility decreases. Increasing audibility in the high frequencies may even result in reduced speech recognition.[2] What appears to be critical is the balance between low-frequency and high-frequency amplification and the contribution of each to the overall loudness and comfort associated with aided listening.

DEFINING HEARING AID PERFORMANCE

Fitting hearing aids to children involves a careful process of selecting and adjusting a particular hearing aid to an individual child. A first step in understanding this process involves defining hearing aid performance, i.e. establishing how much amplification a hearing aid provides. The characteristics of hearing aids are described using the terms 'gain', 'frequency response', 'saturation sound pressure level (SSPL)' and 'distortion'. A brief description of each of these electroacoustic hearing aid characteristics is presented in Table 16.1.

Measuring hearing aid performance

Hearing aid measurement in the 2cc coupler

The electroacoustic characteristics of an individual hearing aid can be measured by attaching the hearing aid to a 2cc coupler to simulate some of the properties of an average adult ear. The 2cc coupler is placed in a soundproof test chamber. A tone or speech weighted noise of a specified input level is generated in the test box, and the specified level is maintained at the hearing aid microphone. The output of the hearing aid is measured in the 2cc coupler. Gain is calculated by subtracting the input from the output. The gain of the hearing aid will vary across the frequencies tested; hence the frequency response of the aid is measured in the test box to

Table 16.1 Terminology for the electroacoustic characteristics of hearing aids.

Terminology	Electroacoustic characteristics of hearing aids
Gain	Gain is the difference in decibels between the input signal and the output signal. In a hearing aid test box, the gain of an aid is measured by presenting a signal of 50 or 60 dB to the hearing aid microphone and measuring the output of the hearing aid in a 2cc coupler. By subtracting the input from the output, gain can be calculated at individual frequencies. Gain can be assessed with the aid on full volume (full on gain) or on a child's user volume (user gain).
Frequency response	Hearing aid frequency response is determined by measuring hearing aid gain across a wide range of frequencies (e.g. 125 Hz to 10,000 Hz) using a constant input level. All sound systems are limited in the range of frequencies that they can amplify and typically hearing aids amplify across the range of 200 to 6,000 Hz.
Saturation sound pressure level	The saturation sound pressure level, also known as the maximum power output and the SSPL90, is the greatest sound pressure that can be produced by a hearing aid. It is measured with the aid on full volume using an input signal of 90 dB SPL.
Distortion	Distortion occurs in a hearing aid when the sound leaving the hearing aid varies from the sound entering the hearing aid. The addition of distortion to the amplified sound may reduce speech intelligibility for the hearing-impaired listener. Harmonic distortion occurs when new frequencies are generated in the amplifier that are whole number multiples of the fundamental frequency of the input signal. Harmonic distortion increases with increases in volume setting making it undesirable to fit children with an aid that is to be worn near maximum volume.

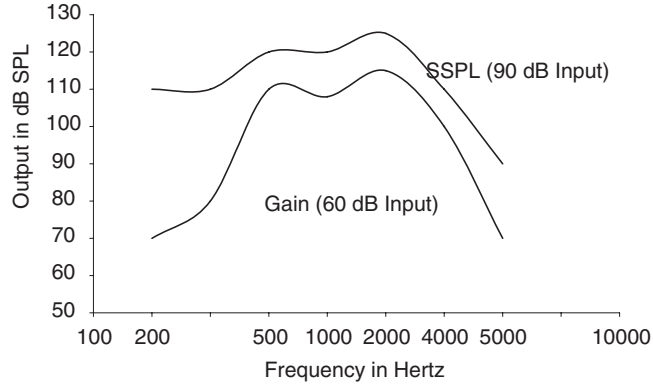

Figure 16.3 Gain and frequency response (60 dB input) and SSPL (90 dB input) characteristics of a BTE hearing aid measured with a 2cc coupler in a hearing aid test chamber.

establish the amount of gain in the hearing aid at each frequency. Saturation sound pressure level can also be measured in the hearing aid test box by using a high input level of 90 dB that aims to fully saturate the hearing aid. The aid is turned to full volume. SSPL is tested across the frequency range. The graphs shown in Figure 16.3 show gain, frequency response and SSPL for a hearing aid on full volume. Distortion can also be measured in the hearing aid test box.

The 2cc coupler measures serve several useful purposes. They are critical for quality control to ensure that individual hearing aid match up to manufacturers' specifications. In 1977, a set of standards was established by ANSI (American National Standards Institute) that required the hearing aid industry to undertake a set of 2cc coupler measures and express hearing aid performance according to a standardised set of rules. The manufacturer specifications provide an important guide to hearing aid performance. However, each individual aid can vary by up to 12 dB from the specification sheet and still be within acceptable tolerances.[3] It is therefore important to measure each aid in the hearing aid test box prior to commencing the selection process.

The 2cc coupler measures provide a valuable starting point for hearing aid selection. They allow the audiologist to select an appropriate hearing aid to try with a hearing-impaired child. However, 2cc coupler measurements will not accurately predict hearing aid gain or SSPL in the real ear, particularly in a small child's ear. Consequently, further real ear measurements of the hearing aid characteristics are required.

Real ear probe microphone measurements

The development of equipment to measure hearing aid performance via a microphone placed near the tympanic membrane represents a small revolution in hearing aid fitting. Using this equipment, the audiologist is able to measure the real ear gain, output, SSPL and harmonic distortion of a hearing aid whilst it is worn by a hearing-impaired child or adult. The procedure is quick, objective, accurate and reasonably non-invasive.

A small rubber probe tube attached to a measurement microphone is placed into the unaided and unoccluded ear canal. The child is seated beside or in front of a loudspeaker. The

Table 16.2 Terminology used for real ear measurement of hearing aid performance.

Terminology	Description of real ear measurement
Real Ear Unaided Response (REUR)	The SPL as a function of frequency in the unoccluded, unaided ear canal with the probe tube at a specified point in the ear canal and in a specified sound field.
Real Ear Unaided Gain (REUG)	The difference in decibels between the SPL in the ear canal and the SPL at the field reference point (reference microphone) in the unaided, unoccluded ear with the probe tube at a specified point in the ear canal and in a specified sound field.
Real Ear Aided Response (REAR)	The SPL, as a function of frequency, with the hearing aid in place and turned on and the probe tube at a specified point in the ear canal and in a specified sound field.
Real Ear Aided Gain (REAG)	The difference in decibels, as a function of frequency, between the SPL in the ear canal and at the field reference point (reference microphone) with the hearing aid in place and turned on and the probe tube at a specified point and in a specified sound field.
Real Ear Insertion Gain (REIG)	The difference in decibels between the REAR and the REUG, made at the same measurement point and in the same sound field.
Real Ear Saturation Response (RESR)	The SPL, as a function of frequency, with the hearing aid in place, and as close as possible to full volume, at a specified point in the ear canal with a specified sound field and at a stimulus level sufficiently intense to operate the hearing aid at SSPL.
Real Ear to Coupler Difference (RECD)	The difference in decibels, as a function of frequency between the output of the hearing aid in the real ear and the output of the hearing aid in the 2cc coupler, taken with the same input signal and hearing aid volume setting.

Adapted from Mueller and Hall.[4]

loudspeaker generates a test signal, such as a frequency sweep of warble tones, a speech weighted broadband signal, or real speech input, and the output is measured in the ear canal. By comparing the SPL in the ear canal to the SPL at a reference microphone placed near the pinna the real ear unaided response (REUR) can be established.

Keeping the probe microphone in place, the hearing aid is then placed on the child's ear, set on user volume and the procedure is repeated, this time measuring the SPL at the eardrum when the hearing aid is worn. By comparing the SPL in the ear canal to the SPL at the reference microphone, the real ear aided response (REAR) of the aid can be established, showing how much amplification the aid provides at each frequency in the real ear. Alternatively, the REAR can be compared to the REUR to generate a measure of real ear insertion gain (REIG). Insertion gain is the amount of gain delivered to a child or adult wearing a hearing aid that he or she did not have prior to the fitting of the hearing aid. It is the net effect of placing an earmould into the child's ear and amplifying the sound with a hearing aid. Table 16.2 contains a summary of the terminology for probe tube real ear measurement.

Hearing aids behave differently in the real ear than in the 2cc coupler. Real ear probe microphone measurement offers a very reliable tool for verifying hearing aid performance in the real ear, but real ear measures are not hearing tests. They measure how the hearing aid amplifies sound in an individual ear, thus reflecting the acoustic characteristics of that ear. Young children may not tolerate repeated real ear measures. Real ear aided gain can also be accurately predicted by using an individualised real ear to coupler difference (RECD) measure

and then applying this to coupler measurements.[5] The RECD is the difference between the SPL in the individual's ear canal and the SPL in a 2cc coupler. Precise instructions for measuring the RECD are often provided in instruction manuals that accompany real ear analysers. Other real ear measurement systems include the RECD as a standard procedure, and it may even be internally incorporated into the most recent digital hearing aids.[6]

Sound-field thresholds and functional gain

A third means of measuring hearing aid gain involves obtaining a child's unaided thresholds in the sound field and then comparing these with the child's aided thresholds measured with their hearing aids on the selected user volume.

The formula for calculating functional gain is

$$\text{Functional gain} = \text{Unaided thresholds} - \text{aided thresholds}$$

Where unaided thresholds have been measured in HL under headphones, they can be converted to SPL in the free field using the figures in Table 16.3.

In the past, sound-field thresholds have enjoyed great popularity as a means of verifying hearing aid performance in the real ear. However, they have a number of significant disadvantages. Test–retest reliability for aided thresholds has been established with adults by Hawkins, Montgomery, Prosek and Walden[7] as ± 15 dB. Unaided thresholds have similar test–retest confidence limits. Consequently, a measure of functional gain may vary from one day to another by as much as 30 dB with no real change in the amplification provided by the hearing aid. Test–retest reliability is likely to be even larger for young children.[8] Obtaining a full set of aided thresholds can be time consuming. Moreover, testing only at octave frequencies often masks the presence of peaks and troughs in the hearing aid frequency response.

Aided sound-field testing with non-linear hearing aids is highly problematic. Non-linear hearing aids provide different amounts of prescribed gain for different inputs. Typically they will provide higher gain for soft inputs and less gain for louder inputs. Since aided thresholds show the softest sounds that the child can hear when aided, they must be a poor predictor of gain for typical speech inputs with non-linear aids.[9] Most digital hearing aids are non-linear, and many children in developed countries are fitted with digital hearing aids (e.g. in Australia, all children under the age of 18 receive free digital non-linear hearing aids from Australian Hearing, a government-funded statutory authority). Consequently, aided threshold testing is not valid as a means of verification for many hearing aid fittings.

With the development of real ear probe microphone measurement, there is no need to rely upon sound-field threshold testing. In most cases, hearing aid performance can be verified using probe tube measures with very young children, particularly if the RECD procedure[5] is adopted. However, under some circumstances, aided sound-field threshold testing remains of

Table 16.3 Minimum audible field (MAF) figures for transforming dB HL to dB SPL.

250	500	750	1000	1500	2000	3000	4000
13	6	4	3	2	0	–3	–4

Source: ISO Draft International Standard 389-7 (December 1993) corrected for monaural listening and semi-diffuse field
Note: To convert HL to SPL: HL + MAF = SPL

value: (1) where queries remain regarding the unaided thresholds; aided sound-field thresholds can assist in confirmation of the accuracy of unaided thresholds when consistent with the coupler gain in a child's hearing aid, and (2) where a child has a corner audiogram and the presence of true hearing is in question. A child with only vibrotactile thresholds may appear to be well aided when tested using real ear probe tube measures but obtain very little benefit from amplification if their responses are all vibrotactile and they have no true hearing.

Processing the signal

Hearing aids are no longer simple amplification devices. They offer a wide choice of features and methods of processing the incoming sound so as to, at least theoretically, meet the individual needs of the child with a hearing loss.

Linear hearing aids

With linear hearing aids, the input/output function maintains a 1 : 1 ratio until the limiting level is reached. Hence, with every 10 dB increase in input there is a corresponding 10 dB increase in output. Linear hearing aids increase output for all input levels by a fixed amount of gain; for example, an aid with a gain of 50 dB at 1,000 Hz will amplify a 45 dB input sound, a 65 dB input sound and an 80 dB input sound by the same gain of 50 dB. The most basic form of linear hearing aid may employ peak clipping to limit the hearing aid output.

Output limiting: peak clipping

All hearing aids have a maximum power output (MPO) that limits the output of the hearing aid for comfort and safety. Peak clipping aids clip or remove the peak of the signal when the MPO is reached. With peak clipping amplifiers the ratio between the sound coming into the aid and output of the aid, the input/output ratio, remains constant until the limiting level is reached. As saturation is reached, the peaks of the amplified sound signal are clipped. This results in a squaring of the waveform and adds harmonic and intermodulation distortion to the sound signal. Figure 16.4 shows the effect of peak clipping upon a sinusoidal waveform.

The distortion added to the signal by peak clipping has a significant impact upon sound quality and may also affect speech intelligibility.[10] Today, it is rarely the output limiting system of choice.

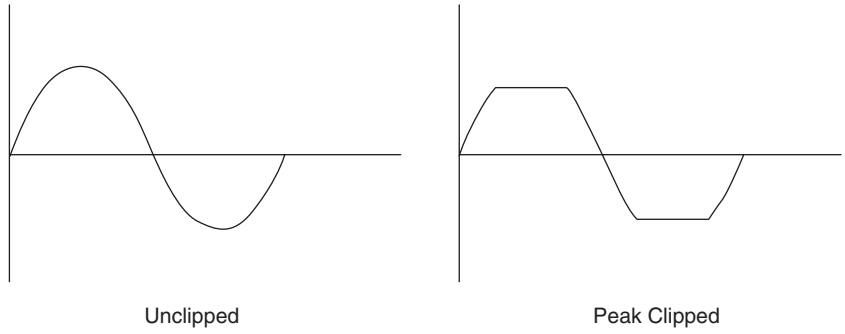

Figure 16.4 The effect of peak clipping upon a sinusoidal waveform.

Output limiting: compression limiting

Many linear hearing aids use output compression limiting to provide an upper limit to hearing aid output. Automatic gain control or compression is a way of controlling the gain and output of a hearing aid. It uses an electronic feedback system to monitor either the level of the input signal (input-controlled compression) or the output of the signal (output-controlled compression) to prevent the signal from reaching saturation when confronted with high input levels. Most output limiting compression circuits utilise output-controlled compression systems. An output-controlled compressor with a high-compression ratio is used to quickly and automatically reduce the gain of any input signals that exceed the hearing aid's MPO. The attack time (the length of time from the point at which input exceeds the limiting level to the point at which gain is stabilised) and the release time (the time lag from that moment to when the system returns to normal gain function) of the compressor is set to be very short so as to maximise comfort and minimise loss of sound input following the loud sound. This compression or automatic gain control only occurs with loud sounds that exceed the MPO of the hearing aid; all other sound inputs results in linear amplification.

Non-linear hearing aids

In contrast, non-linear hearing aids utilise compression circuits to increase audibility and comfort for the hearing aid users. These aids typically provide more gain for softer sound inputs, medium gain for average inputs and lower gain for high-level inputs. Wide-range dynamic compression hearing aids use a low-compression knee point so that compression occurs over a range of inputs, and a portion of the dynamic range of speech is in compression. Such systems aim to increase the listener's comfort, increase the audibility of soft phonemes, normalise loudness and reduce noise. Whole range syllabic compression circuits are similar but have a shorter release time. The theoretical benefits of non-linear amplification primarily relate to improved audibility of soft sounds with less need to adjust the volume control in a wider range of listening conditions. Research on the benefits of wide-dynamic-range compression (WDRC) compared to linear hearing aids has provided mixed results. Some studies have shown improved speech perception skills and higher ratings of speech quality for hearing aids with compression as compared with linear hearing aids with adults[11] and children.[12] Other studies have failed to show any improvement in speech discrimination or speech quality ratings[13,14] with WRDC aids when compared with linear aids. These ambivalent results may be explained by differences in fitting procedures, the type of compression systems and the number of channels in the aids assessed.[12]

Multichannel devices

Digital non-linear hearing aids may offer the listener as many as twenty channels in which compression parameters can be independently adjusted. This type of flexibility is theorised to reduce the impact of recruitment in individual frequency bands and improve the signal-to-noise ratio with adjustments to the lower frequency bands. As with WDRC, research findings comparing single channel and multichannel devices are equivocal. Hickson's[15] review of 21 studies showed that half supported the benefits of at least two-channel compression while the other half found no benefit when compared to a single-channel device. More recently, Keidser and Grant[16] compared one-, two- and four-channel devices under laboratory conditions and found no significant differences in speech perception scores. In contrast, their field trials in a range

of real listening environments, comparing single- and two-channel devices indicated a clear preference for the two-channel device for individuals with steeply sloping hearing losses. Individuals with flat, moderate to severe hearing losses preferred the single-channel hearing aid. There is little evidence to suggest that hearing aids with more than four channels provide any additional benefits for speech perception.[11]

Studies that have sought to compare different types of signal processing have often found discrepancies between speech perception scores and speech quality ratings.[16,12] One explanation for this finding is that the signal-processing algorithm may be slightly degrading the signal by reducing spectral and temporal contrast but providing greater perceptual listening comfort in noisy listening environments. An alternative explanation may partly lie with the findings of Bentler et al.[17] They studied the impact of digital labelling on outcome measures. Hearing aid preferences and sound quality ratings were significantly influenced by the 'digital' label. Participants were much less likely to prefer the unlabelled digital hearing aid over a conventional analogue hearing aid than when it was labelled as a digital aid. This finding emphasises the need for caution when evaluating any research that purports to show a clear benefit for digital technology based upon participant preference or ratings.

Are high technology hearing aids necessarily high performance?

Given the high cost of many modern digital hearing aids with WDRC and multiple channels, these results are sobering. On balance, it would appear that there are small benefits for non-linear hearing aids that use WDRC in terms of speech perception and subjective ratings of sound quality. This may be impacted by the degree of the individual's hearing loss, the fitting procedure and other aspects of signal processing. Marriage et al.[12] conclude that 'it appears safe to use well-designed WDRC for children with severe or profound hearing loss; there is unlikely to be a penalty in terms of reduced ability to understand speech'. Multiple channels appear to provide limited additional benefits for children with severe and profound hearing losses, who appear to prefer single-channel devices.[16] Two-channel devices may benefit individuals with steeply sloping hearing losses.

There are other benefits associated with digital signal processing in hearing aids. The computer chip inside a digital aid can offer the listener many additional features that are not available in analogue hearing aids. These features include feedback cancellation (FBC) systems that can reduce the risk of acoustic feedback and therefore allow the listener to use more gain. FBC circuits continually monitor the output of the hearing aid to ascertain if the amplified signal has any components with the characteristics of acoustic feedback. If these are detected, the feedback circuit determines the frequency, amplitude and phase of the feedback component and then generates signals of the opposite phase that cancel or substantially reduce the feedback component. Consequently, it is possible to achieve target gain requirements that may have been previously limited by acoustic feedback. It is also possible for listeners with moderate and severe losses to wear more open and comfortable earmoulds without risking acoustic feedback.

Other features offered by digital signal processing hearing aids include multiple memories, whereby the listener can choose different gain settings depending upon whether they are listening in a quiet or noisy environment, and noise-reduction systems that monitor the modulations in the sound input and reduce those frequencies where noise is present. Another feature is directional signal processing where the hearing aid reduces the input from sound coming in on either side of the listener and enhances signals received from the front.

In the near future, it is inevitable that all hearing aids will be digital and offer a range of signal-processing strategies. According to Jenstad and Souza, in 2005,[18] about 85% of hearing aids fitted in the USA used some form of compression to process the incoming signal in a non-linear fashion. It is, therefore, comforting to know that digital non-linear multichannel hearing aids will be at least as good as analogue models, particularly for children with severe and profound losses. However, there is very little evidence to support the view that digital, multichannel non-linear aids will dramatically change a child's capacity to access speech. Where funds are limited, as they often are in developing countries, no parent should ever feel that they have disadvantaged their child by not being able to afford the latest high technology hearing aid. What is more important is that the hearing aid has been carefully selected, fitted and validated for the individual child.

FITTING AMPLIFICATION TO CHILDREN: SPECIAL CONSIDERATIONS

Amplifying sound for small ears

The residual volume of air between the tip of the earmould and the eardrum is significantly reduced in a child as compared to an adult. Consequently, the output of a hearing aid worn in a small ear is likely to be higher than in the ear of an adult. Feigin et al.[19] demonstrated that RECDs were larger in children, and the magnitude of that difference varied with age, with larger RECDs found in young infants. This is of particular relevance with recent developments in universal newborn hearing screening and very early identification of hearing loss. Coupler measures may significantly underestimate the SPL in the real ear. This highlights the importance of probe microphone measures to assess the RECD and establish real ear gain and SSPL. In the past, assessment of the real ear MPO or real ear SSPL has been particularly neglected. This hearing aid characteristic has frequently only been assessed in the 2cc coupler. It seems likely that there have been young children fitted with unnecessarily high real ear SSPL, possibly resulting in further deterioration of their hearing.

Amplification and auditory development

Normal-hearing infants can make very fine speech discriminations early in infancy. Pioneering work by Eimas[20,21] using the high-amplitude sucking procedure showed that 1- and 4-month-old babies could discriminate between syllables from different phonetic categories. Many more studies have since extended this body of knowledge regarding infants' considerable capacity to make fine speech discriminations. Nozza's[22–24] work in this area has particular relevance to the fitting of hearing aids to young children.

Nozza et al.[22] showed that infants were capable of discriminating speech in noise but they required a signal-to-noise ratio of 6–12 dB higher than adults for the same task. Nozza and his colleagues also studied speech perception in infants whilst simulating a hearing loss using a reduced signal intensity[23] or by filtering the speech signal. They showed that infants required an additional 8–10 sensation level to discriminate speech sounds level as compared to adults. Nozza[24] concluded that infants require greater signal intensity and a better signal-to-noise ratio than adults to perceive speech optimally. One untested conclusion that could be drawn from these studies is that a young child requires more gain and better listening conditions for speech perception than an adult with the same degree of hearing loss.

Amplifying speech for young language learners

Adventitiously deafened adults are usually competent users of spoken language with a great capacity to compensate for their gradual loss of sensory input. In comparison, children who are congenitally deaf or hearing-impaired are faced with the dual task of learning to listen with amplification and learning language simultaneously. In this situation, every piece of sensory input becomes critical.

APPROACHES TO THE SELECTION AND VERIFICATION OF AMPLIFICATION FOR CHILDREN

The modern hearing aid market seems full of heavily promoted new technology including digital feedback reduction, multiple listening programmes, adaptive noise suppression and noise reduction through directional microphones. However, the wonders of new technology are of little use to a hearing-impaired child if an appropriate procedure is not used to match the hearing aid selected to the characteristics of the child and his or her hearing loss. Having decided that a child is a hearing aid candidate, the first and most essential step in the process is the selection of the hearing aid characteristics for the child.

Historically, hearing aid fitting with both adults and children was undertaken using what is sometimes known as the 'traditional' or 'evaluative' approach. Having established the individual's hearing thresholds, these were used to pre-select one or more hearing aids, which were then fitted with a customised ear mould. Pre-selection of aids was often based on the audiologist's previous experience of what might be appropriate for the degree and shape of hearing loss. This was followed by some form of assessment of performance with the aid or aids involving aided sound-field thresholds, speech perception testing and subjective evaluation (i.e. how does it sound?). This approach may well have been effective at a time when there was a very restricted range of hearing aids available, but it has many disadvantages. It is time consuming, speech perception tests are insensitive to quite large differences in frequency response,[25,26] sound-field thresholds have limited test–retest reliability, and repeated assessments are highly impractical for young children. In addition, there remains the dilemma of pre-selecting the most appropriate aids. Can a clinician really be sure that they have compared the most suitable aids for the child? Although clinical experience is highly valuable, without some theoretical model for ascertaining what is an optimum hearing aid fitting, it will always be difficult to be absolutely confident in the aids pre-selected for trial. Although traditional approaches have been popular in the past,[27] their usage has declined.[28] The complexity of modern hearing aids requires a more refined and scientific approach.

PRESCRIPTIVE APPROACHES TO HEARING AID SELECTION

Prescriptive approaches use a model or theoretical rationale that aims to optimise the audibility of speech according to the degree and configuration of hearing loss. A set of figures is derived from this model. These permit the audiologist to precisely select the electroacoustic characteristics of a hearing aid based upon the child's unaided hearing thresholds. These may be expressed as 2cc coupler figures, sound-field-aided thresholds, or as real ear gain. There many advantages associated with choosing to use a theoretical or prescriptive approach

to select a hearing aid for a child: The use of a theory or model for optimising the child's reception of speech is clearly stated and open to scrutiny, there are clearly specified criteria available for pre-selection of hearing aid and verification following fitting, the approach can be implemented by both experienced and inexperienced audiologists, it is time efficient.

Several well-documented prescriptive approaches have been developed over the past 50 years. Some of the more well known include Prescription of gain/output (POGO) II,[29] the Desired Sensation Levels method[30,31] and the National Acoustic Laboratories' revised procedure (NAL-RP).[32,33] These approaches have been used to fit linear analogue hearing aids. More recently, prescriptive procedures for fitting non-linear hearing aids have been developed. Two of the most well known and frequently used with children are the DSL (input/output [i/o]) approach[34] and NAL-NL1 approach.[35]

The desired sensation levels approach to hearing aid selection for children

The earliest versions of this approach appeared during the1980s. Its stated goals are to provide children with amplified speech that is audible, comfortable and undistorted across the broadest relevant frequency range possible.[36] This approach has been specifically developed for children and is cognisant of the arguments regarding the importance of adequate amplification during the early language-learning years and the differences between young children's and adults' speech perception skills.

Gain and frequency response selection in the desired sensation levels (DSLs) approach originates from a set of DSLs across the frequency range for the long-term speech spectrum. These DSLs are dependent upon frequency and degree of hearing loss. From these DSLs, targets for real ear aided gain, real ear insertion gain, and sound-field thresholds have been generated. In addition, the approach specifies targets for real ear saturation responses based upon theoretical work by Pascoe.[37]

The DSL approach has incorporated the use of probe microphone measurements so that all measurements are specified in the ear canal. Hence, there are no conversions or corrections, and a child's resulting hearing aid fitting can be clearly presented as his or her dynamic range for speech showing unaided thresholds, the amplified speech spectrum and the real ear saturation response of the aid as if measured in the ear canal.

A significant, innovation in the DSL approach has been the use of the RECD procedure, whereby the only measurement performed with the child is to establish their individual RECDs.[38] Targets for selection of gain and frequency response and SSPL setting are generated efficiently using the DSL method software.

The DSL (i/o) prescriptive procedure was first described by Cornelise, Seewald and Jamieson in 1995.[34] This procedure built upon the original DSL method to provide prescriptive targets for the fitting of non-linear hearing aids that use wide-dynamic-range compression. The DSL (i/o) algorithm uses a curvilinear map to fit a range of input levels to a set of output targets across frequencies. It aims to normalise loudness so that the hearing-impaired listener would hear narrow-band test signals at different input levels at the same loudness that would be perceived by normal-hearing listeners. The most recent version of this software is DSL v5.0,[39] which incorporates a multistage algorithm that is reported to adapt to the different needs of listeners with congenital versus acquired hearing loss and also to accommodate the different listening requirements of quiet and noisy listening environments.[39]

The National Acoustic Laboratories (NAL) approach to hearing aid fitting

The first version of the NAL procedure was introduced in Australia in 1976. This approach aims to amplify the long-term speech spectrum so that is comfortable and equally loud across the frequency range. Byrne and Tonnison[40] based their estimate of the overall required real ear gain on research by Byrne and Fifield[41] that had shown that moderate to severely hearing-impaired children preferred to use, on average, 4.6 dB of gain for each 10 dB of hearing loss. Added to this was a frequency-dependent correction figure that aimed to account for loudness differences in speech across the frequency range and for the shape of the long-term speech spectrum. This approach has undergone several modifications.

In 1986, Byrne and Dillon[32] revised the procedure as the NAL-R approach following extensive evaluation with hearing aid users fitted using the original NAL procedure. They added an X-factor to take into account the slope of the audiogram and modified the formula used to calculate required gain. In 1990, the profound correction factor was added for individuals with severe and profound hearing losses[33] prescribing more gain overall for 60+ dB losses and more low-frequency gain for people with a hearing loss greater than 95 dB at 2,000 Hz. In 1998, selection of SSPL90 in the 2cc coupler was added to the NAL procedure, and experimental data were published on the derivation and validation of that procedure.[42] The NAL-RP procedure has been systematically validated with adults[32,33] and to some extent with school-aged children.[43] Targets for the NAL-RP approach can be found in most real ear measurement equipment.

Unlike the DSL (i/o) approach and many other non-linear prescriptive fitting procedures, the NAL prescriptive procedure for fitting non-linear hearing aids (NAL-NL1) does not attempt to normalise loudness at each frequency. The NAL-NL1 procedure aims to maximise speech intelligibility for hearing-impaired listeners while accounting for the effects of hearing loss desensitisation at high input levels. It also aims to model the effects of a sensorineural hearing loss on the perception of loudness rather than assuming the listeners with hearing loss will perceive loudness in the same way as those with normal hearing.[10] The NAL-NL1 software program generates targets for a 2cc coupler and the real ear. At speech input levels of 70 dB SPL, it provides similar gain to the NAL-RP linear procedure.

The NAL procedures are frequently used with children despite the fact that they were not designed specifically for this population. This could be a disadvantage of the NAL approach when fitting hearing aids to very young children; however, there are no research data available to draw this conclusion.

The availability of so many choices in digital signal processing hearing aids has also seen the development of many proprietary fitting algorithms by hearing aid manufacturers. Hearing aid manufacturers encourage audiologists to fit their hearing aids using their own algorithm rather than one of the more established generic algorithms such as DSL (i/o) or NAL-NL1. Keidser, Brew and Peck[44] showed that the use of manufacturers' algorithms can lead to big differences in gain fitted as compared with generic algorithms. The lack of data available to assess the underlying model used in the development of these proprietary algorithms is worrying, particularly when used with young children who cannot provide feedback about sound quality or complete speech perception tests.

A risk that can be associated with the use of proprietary algorithms for hearing aid fitting relates to the individual validation of the aid fitting. Digital non-linear hearing aids are complex, and some clinicians may believe that it is not possible to verify an aid fitting because

of the many features that the aid utilises, such as noise cancellation and adaptive directionality.[45] The software provided for selecting the gain and MPO in digital non-linear aids is really only a more sophisticated version of the screwdriver used with basic analogue hearing aids. Setting up the non-linear aid with the computer software is just the first step in the process of aid fitting. It is essential that the aid fitting be verified on the listener using probe tube real ear measurement and target values. This is possible and highly recommended with all hearing aids, including those employing sophisticated noise cancellation systems. Newer real ear measurement systems offer simulated speech signals or may even use real speech for accurate real ear measurement with any hearing aid. Verification is important for all individuals with hearing loss, but it is critical for young children who are learning language with their hearing aids.

The underlying models upon which the NAL and DSL procedures are based also result in significant differences between the gain prescribed by each. Deciding which prescriptive procedure is best depends upon the target population, the clinician's personal philosophy and some careful evaluation of the theoretical models that underpin each procedure. Both the DSL and the NAL approaches have been carefully developed to systematically prescribe both linear and non-linear hearing aids for hearing-impaired individuals. They represent two excellent theoretical models for hearing aid selection. Some comparative studies have been published,[46–48] but there is no conclusive evidence yet available to allow clinicians to confidently select one approach over the other. Scollie et al.[46] and Wigney et al.[48] both independently demonstrated that children with prior experience of one approach to aid fitting (either NAL or DSL) are most likely to prefer that approach when offered a choice in a preferred listening task. There are significant benefits for the profession in the existence of two strong generic fitting procedures as they stimulate each other and encourage further development and refinement that can only benefit children who are deaf or hearing-impaired.

When clinicians select an aid based upon a hearing aid prescription, whether the DSL or NAL approach, they might assume that their work is done. Once the procedure's targets have been achieved during the validation process, the hearing aid fitting is seen as complete, not to be altered until some change occurs in the child's acoustic or audiological characteristics. However, although the selection and validation process is critical to the aid fitting process, it is just the first step. All prescriptive approaches are based upon theoretical averages. On average, the targets that the audiologist seeks to achieve may be appropriate for a child with that degree of hearing loss. Fine-tuning will be necessary to obtain an optimum match between the aid and the performance and preferences of an individual child. What is often overlooked in prescriptive procedures is the importance of feedback from the child and his or her parents and teachers to evaluate hearing aid benefit. Audiologists need to be willing to make some modifications to an aid fitting based upon data gathered after the aid is fitted. It is a process frequently entered into with adults but often neglected for children. Parents and teachers can play a vital role in this process particularly if they are involved in intensive auditory habilitation with the child.

EVALUATING CHILDREN'S PERFORMANCE WITH AMPLIFICATION

Ongoing evaluation is an important step in both traditional and theoretical approaches to paediatric hearing aid fitting. It is critical for initial aid fittings and also when a child is changed

over to new hearing aids. Teachers and parents must work very closely with audiologists to provide the kind of feedback the audiologist needs to judge the effectiveness of the aid fitting. This is particularly important for children with severe and profound hearing loss who are being considered for cochlear implants and who are undergoing hearing aid trials to assess their implant candidature.

Feedback from teachers and parents can come in several formats. For beginning listeners, their initial responses to sound may well be quite crude and gradually become more specific. If the child and parent or teacher are engaged in intensive auditory/oral or auditory/verbal habilitation, then responses to sound and speech can be monitored closely. The development of vocalisations and speech can provide many clues about the child's access to audible speech. The Parents Evaluation of Aural/Oral Performance of Children (PEACH) is a questionnaire designed to record parents' observations in a systematic way for the purpose of evaluating hearing aid fitting. Developed by the NAL, this instrument has been used to evaluate hearing aid fittings using both the NAL and DSL procedures.[49]

Older children can be observed in the pre-school or school environment to assess their responsiveness to speech and environmental sounds, their visual attention to the speaker and their verbal repetitions. These can take the form of informal reports by a teacher, parent or child or a more formalised checklist or classroom observation tool such as the Screening Instrument for Targeting Educational Risk (SIFTER).[50] Self-reporting by older children can be very valuable. When a child is changed over to new hearing aids, gradual changes to the frequency response may be required to accommodate his or her listening comfort. Parents and teachers can use functional tools such as the Meaningful Auditory Integration Scale (MAIS),[51] to provide feedback to audiologists about the child's use of residual hearing.

At the more formal end of the spectrum, speech perception tasks can be used to document the development of speech perception skills. Speech perception test results can show which sounds can be discriminated by a child and assist with a review of a child's hearing aid fitting and habilitation programme.

OTHER CONSIDERATIONS IN HEARING AID FITTING

Binaural hearing aid fitting

Choosing two hearing aids for a child with a binaural hearing loss is not new wisdom or sophisticated technology, but it needs highlighting. In many ways, even considering fitting one aid to child with a symmetrical binaural hearing loss seems as absurd as optometrists recommending monocles to their clients. As Pascoe has observed, 'Hearing aids should be chosen to help restore binaural hearing. They should, in fact, be sold in pairs, just like eyeglasses' (Mueller and Hall,[4] p. 135). The advantages associated with binaural hearing aid fitting include improved sound quality, improved speech discrimination in noise, reduction of the head shadow effect, loudness summation, sound localisation and spatial balance. Moreover, research in the area of hearing aid acclimatisation suggests that auditory deprivation is a real risk with monaural fittings whereby an individual will actually show decreased speech perception scores over time in the unaided ear.[52–54]

Monaural hearing aid fitting is not a legitimate way to reduce amplification costs with children. If funds are pressed, it is far better to choose two less expensive hearing aids than

one high-technology hearing for child with binaural hearing impairment. Monaural fitting should only be considered when it can be clearly shown that speech perception in one ear is so poor as to significantly decrease the overall speech intelligibility for the child.

Earmould selection

Good impression taking technique and the manufacture of a well-fitting custom-made earmould is a critical component in the paediatric hearing aid fitting process. Acoustic feedback and earmould discomfort are constant problems for young soft ears. Table 16.4 contains a list of tips for obtaining a good earmould impression.

An audiologist must also select the most appropriate acoustic characteristics for the child's earmoulds. All acoustic modifications to a child's earmould will affect the gain, the frequency response and the SSPL of the hearing aid. The three main options available to a clinician are venting, damping and fitting an acoustic horn.

Venting

Inserting a parallel vent into an earmould allows children to utilise any good low-frequency hearing they may have, reduces low-frequency real ear gain, aerates the ear canal and relieves pressure. Vents are unlikely to be viable with severe and profound losses due to the risk of acoustic feedback.

Damping

Acoustic filters or dampers of wool or sintered metal are often used to reduce the resonant peaks created by the original response of the hearing aid earphone. In theory, dampers can be placed at various points along the tubing to damp peaks at different frequencies. In practice with children, dampers are unlikely to stay in position unless they are placed in an ear hook. Common places are the tip or nub of the ear hook where they will have most effect around 1,000 Hz. As a minimum, a low-resistance damper is recommended with most hearing aid fittings to reduce some of the bigger peaks and troughs of the frequency response and improve the natural quality of the sound.

Table 16.4 A guide to taking earmould impressions.

Taking an earmould impression
1. Carefully inspect the ear using an otoscope.
2. Always insert a foam or cotton wool canal block into the ear canal before inserting the earmould impression material.
3. Fill the syringe with impression material and gently squeeze the plunger until a drop or two emerges from the syringe.
4. Insert the syringe into the ear canal and fill the ear canal without removing the syringe. Always keep the nozzle of the syringe buried in the impression material.
5. Fill the helix and the concha completely.
6. After a few minutes, test with a fingernail. Gently remove impression when material bounces back.
7. Inspect ear with otoscope to ensure that all material has been removed from the ear.

Table 16.5 Predicted response of Libby acoustic horns as compared to the response of 2-mm constant diameter tubing.

Frequency	250	500	1,000	1,500	2,000	3,000	4,000	5,000
Libby Horn 4 mm	–1	–2	–3	0	–2	6	10	7
Libby Horn 3 mm	–1	–2	–2	1	0	6	8	8

Acoustic horns

An acoustic horn is a stepped piece of tubing that gradually increases in diameter from 2 mm at the ear hook to 3 or 4 mm at the end of the sound bore. The Libby horn was patented in 1982. It works by matching the resistance of the ear canal more closely to the resistance of the earphone and tubing, thus enhancing the amplification of high frequency sounds (see Table 16.5).

Ordering an acoustic horn in a child's custom earmould may assist an audiologist in meeting a child's high-frequency prescription targets. However, acoustic horns may not be viable for very young children. The ear canal must be able to accommodate at least a 3 mm sound bore. Acoustic feedback and loudness discomfort can also be risks, depending upon the shape of the hearing aid frequency response and the features offered in the child's hearing aid. The development of digital non-linear hearing aids with an improved high-frequency response has resulted in reduced reliance upon acoustic horns to reach prescription targets.

HEARING AID MANAGEMENT

There seems little point in taking great care to select appropriate high-technology hearing aids for a child if little or no attention is paid to daily aid monitoring and maintenance. Over the past 25 years, a number of hearing aid performance studies have shown that hearing aids malfunction at an alarming rate. In a study of a residential school, where staff had been trained to perform daily listening checks, 45% of aids failed to pass the adequate performance criteria either due to low battery voltage, acoustic feedback or inadequate volume settings.[55] Other early studies have reported similar results.[56–58]

Targeted school training programmes have shown improved results in hearing aid function and maintenance,[59] particularly when the programme is directed at both the students with hearing aids and their teachers. Without an intensive approach to daily maintenance, hearing aid malfunction can be a substantial barrier to ensuring that children are optimally amplified. A team effort is required with the child as the key player developing self-management skills and good reporting strategies from an early age. Such programmes need to be well resourced. From a cost–benefit perspective, there is little value in spending a substantial amount of money on hearing aid fitting if there is no educational audiology support for children, teachers and families to keep everything working. Daily aid maintenance and the development of efficient repair systems are not glamorous aspects of audiology but they are critical components in the process of maximising audibility for every child.

Future developments in aid technology

With the recent rapid development in hearing aid technology, it is exciting to look into the future and speculate on the new developments that may take place. Some of the prospects

include trainable hearing aids that can learn to adjust to the range of environments regularly encountered by a particular individual, hybrid implant/hearing aids devices that incorporate the benefits of both electrical stimulation and amplification, and wireless communication between binaural hearing aids and between hearing aids and other devices such as mobile phones, computers and MP3 players.[60] Many of these new developments are already close to production and will see a much more integrated approach to the utilisation of the hearing sense not only for individuals with hearing loss but possibly for children and adults with normal hearing.

Other developments that will impact on the effectiveness of amplification include much more precise diagnosis of hearing loss including the role of inner- and outer-hair cells, the detection of auditory processing difficulties of input signals beyond the cochlea and the identification of dead spots in the cochlea.[60] Technology itself, i.e. the Internet, will make it much easier to keep up with new options for children who wear hearing aids.

CONCLUSION

This chapter has focused upon providing a practical guide to the selection of amplification for children for audiological professionals. It has emphasised the importance of systematically selecting a hearing aid to match the audiological and acoustic characteristics of each child, particularly in an environment of rapid technological change. However, the fitting of amplification will always be but a first step in the process of audiological habilitation. Without the child's, parents' and teachers' participation in ongoing evaluation and habilitation and without dedicated management of amplification in educational settings, it may be effort wasted. Only a team approach will allow every child with a hearing loss to make maximum use of new developments in the design and fitting of hearing aids.

Resources

Software

The Desired Sensation Level Method: Version 5.0

For computer software to implement the DSL approach to hearing aid selection contact:
The Siemens Child Amplification Laboratory,
The National Centre for Audiology,
University of Western Ontario,
Elborn College, London,
Ontario, Canada
N6G 1H1.
http://www.dslio.com

National Acoustic Laboratories (NAL-NL1)

Hearing Aid Selection Procedure

For hearing aid fitting computer software to implement the NAL-NL1 hearing aid fitting procedure contact:
Research Administrative Officer
National Acoustic Laboratories

126 Greville Street
Chatswood NSW 2067
Australia
Telephone: +61 2 9412 6872 or +61 2 9412 6862
Facsimile: +61 2 9411 8273
Email: Research@nal.gov.au
http://www.nal.gov.au/

Books

Dillon, H (2001) *Hearing Aids*. New York: Thieme

This book is a complete reference book on the topic of hearing aids. It provides detailed discussion of a wide range of issues relating to hearing aids and hearing aid fittings. It is very clearly written and set up in such a way that readers can choose to read a summarised or full version of each chapter.

Mueller, HG and JW Hall (1998) *Audiologists Desk Reference Volume II: Audiologic Management, Rehabilitation and Terminology*. San Diego, CA: Singular Publishing Group Inc.

An excellent resource for all professionals involved in hearing aid fitting. This reference book contains excellent tables, summaries, diagrams and quick guides to every conceivable aspect of amplification.

Video/DVD Resources

Pediatric Hearing Instrument Fitting

Phonak Video Focus 2 (1997)

Hearing Care for Infants: Strategies for a Sound Beginning

Phonak Video Focus 4
http://www.phonak.com/pediatrics/focus

Non-linear Hearing Aid and the NAL-NL1 Prescription Procedure

Set of 2 VHS videos with notes
$75 (AUS)
Telephone: +61 2 9412 6872 or +61 2 9412 6862
Facsimile: +61 2 9411 8273
Email: Research@nal.gov.au
http://www.nal.gov.au/

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17 Cochlear implants in children

R.T. Ramsden and P. Axon

INTRODUCTION

Cochlear implantation is without doubt the most exciting development in otology in recent years, and many would reasonably argue of all time. During the first half of the twentieth century, many of the challenges of middle-ear hearing impairment were met and to a considerable degree solved by operations such as the tympanomastoidectomy and the stapedectomy. With a few exceptions, however, the solutions to the problems of inner-ear deafness did not seem to lie in the surgical domain. Individuals who had lost their hearing after they had acquired speech and spoken language would have little alternative to rehabilitative strategies that relied on the use of hearing aids and lip reading.

Children born with a profound hearing impairment or those developing a profound hearing impairment before the acquisition of spoken language would never be able fully to gain this skill. Education for such children has been based on either an oral tradition, employing lip reading and amplification of residual hearing, on signing, or on total communication, which uses a combination of both philosophies. Whichever educational path is followed, and however successful it might be in an individual case, the fact remains that the social, educational and psychological progress of these children is often held back, and integration into the culture of normal hearing is impossible.

Cochlear implantation is based on the replacement of the lost cochlear transducer with an electrode system that delivers to the auditory pathways a processed electrical signal resembling closely the essential characteristics of speech, which the brain is capable of decoding. The development of the technique owes much to remarkable advances in a wide variety of scientific fields: engineering, microcircuitry, neurophysiology, surgery, the cognitive sciences and education. In many ways the success of cochlear implantation has been due to unique cooperation and teamwork between the scientists, manufacturers, implant clinics and educationalists. Implantation has had its opponents, and controversy has often arisen as the signing Deaf community has seen it as a threat to their identity. Because of the costs incurred in the provision of cochlear implants (CIs) to children, their rehabilitation purchasers of healthcare were initially cautious to support this surgery, but now that the results of timely implantation have been seen to be so good, funding is very much less of an issue, and the trend is now towards provision of bilateral implants as a routine.

HISTORY AND THEORETICAL BASIS OF COCHLEAR IMPLANTATION

Benjamin Franklin (1706–1790) was supposedly the first person to suggest that if an electric current was applied to the ear a sound would be heard. It is certainly true that in 1800, Alessandro Volta, who was given to self-experimentation with electricity, reported to the Royal Society of London that when he placed a metal rod in each ear and connected them to his 'new electrical apparatus', the Voltaic pile, he 'experienced a commotion in my head and a few moments later I began to hear a sound or rather a noise in my ears'. It was a sort of rhythmic crackling or sizzling as though a paste or thick substance was boiling. This sound continued uninterrupted as long as the conductive circle was complete. 'The disagreeable sensation which I feared could be dangerous if the shaking in my brain meant that I did not repeat this experiment.' If one assumes that the stimulus was delivered to the organ of Corti, and not to more central structures, this graphic description is the first to show that electricity can be conducted from the periphery of the auditory system to the cortex.

The next landmark was the operation performed in Paris in 1957 by Djourno and Eyries. They placed a simple copper ball electrode on the eighth nerve of a man previously operated upon for cholesteatoma, who was undergoing further surgery for repair of his facial nerve. Electrical stimulation of this device produced an auditory percept, which the patient likened to the sound of crickets chirping or a roulette wheel. He could detect changes in the stimulus frequency up to 1,000 Hz, was aware of environmental sounds and had improved recognition of the prosodic patterns of speech.

A number of workers in North America were encouraged by this case to look more methodically at the effects of electrical stimulation of the inner ear. These early workers included William House in Los Angeles, Blair Simmons at Stanford and Robin Michelson in San Francisco. They faced opposition from basic scientists who felt that, on theoretical grounds, cochlear implantation should not work, and from sections of the Deaf community who felt that the operation was unethical. Their work led to the development of an increasing number of centres throughout the world looking at experimental and clinical aspects of cochlear implantation throughout the 1970s, '80s and '90s. Prominent amongst these experimenters were Clark in Melbourne, Burian in Vienna, the Hochmairs in Innsbruck, Chouard in Paris, Eddington in Utah, and Fraser and Douek in London. Early problems to be solved included the siting of the electrode (intracochlear or extracochlear), number of channels (single or multiple) and the signal-encoding strategy (analogue or digital).

Psychoacoustic tests to try to predict neuronal survival and potential benefit from implantation were studied in great depth. In addition, a vast number of animal studies were necessary to establish the safety and reliability of the devices, before widespread use in humans could proceed. The problem that cochlear implantation sets out to solve is that of severe, profound or total deafness due to partial or total absence of the organ of Corti. This loss may be congenital or acquired. The organ of Corti acts as a transducer that detects the incoming physical sound waves in the cochlear fluids and converts them into electric currents that pass down the auditory nerve and through the central auditory pathways to the primary auditory cortex in Broca's area. Association fibres project to other parts of the brain, conferring significance, meaning and an emotional overlay to the incoming signal.

The CI takes the place of the damaged organ of Corti and delivers to the inner ear a processed signal that stimulates more central neural structures, probably the spiral ganglion. In the typical CI system, there is an externally worn microphone that delivers the raw electrical

Figure 17.1 Nucleus ear level speech processor in situ.

signal to a so-called speech processor in which the signal is manipulated to enhance speech recognition (Figure 17.1). The individual speech processing strategies vary from manufacturer to manufacturer. The refined signal is transmitted through the skin by a process of inductive coupling. The power source for the system is a battery housed in the external component. The implanted component of the device decodes the incoming digitised signal and directs it as a series of discrete stimuli to the electrode array. It is now universally recognised that intracochlear placement of a multichannel device is the strategy that gives the best results (Figure 17.2). There are now very few if any indications for extracochlear placement, or for single-channel devices.

The CI takes advantage of several known anatomical and physiological features of the normal cochlea. The very tight tonotopic arrangement of the cochlea, with low-frequency perception at the apical end and high frequencies at the basal end, is utilised in the design and stimulation strategy of multichannel systems. For example, the Nucleus system features 22 intracochlear electrodes reaching from the region of the round window to the middle turn of the cochlea.

The more frequently the incoming signal is sampled, the greater the fidelity of information transfer to the central nervous system, and sampling rates of up to approximately 100,000 per second are now possible. The problems of crosstalk between electrodes at this rate of stimulation have been minimised by the strategy of continued interleaved sampling (CIS), which staggers the arrival of the stimulus at the electrodes so that they are not stimulated synchronously.

An early and ingenious stimulation strategy employed by the Nucleus company depended on recognising the specific formant frequencies of speech and extracting them from the rest of the signal. This has been superseded by a simpler strategy by means of which only the electrodes with the greatest energy peaks at any precise moment of stimulation are chosen to activate the auditory nerve. The patterns of these peaks will fluctuate rapidly from moment to

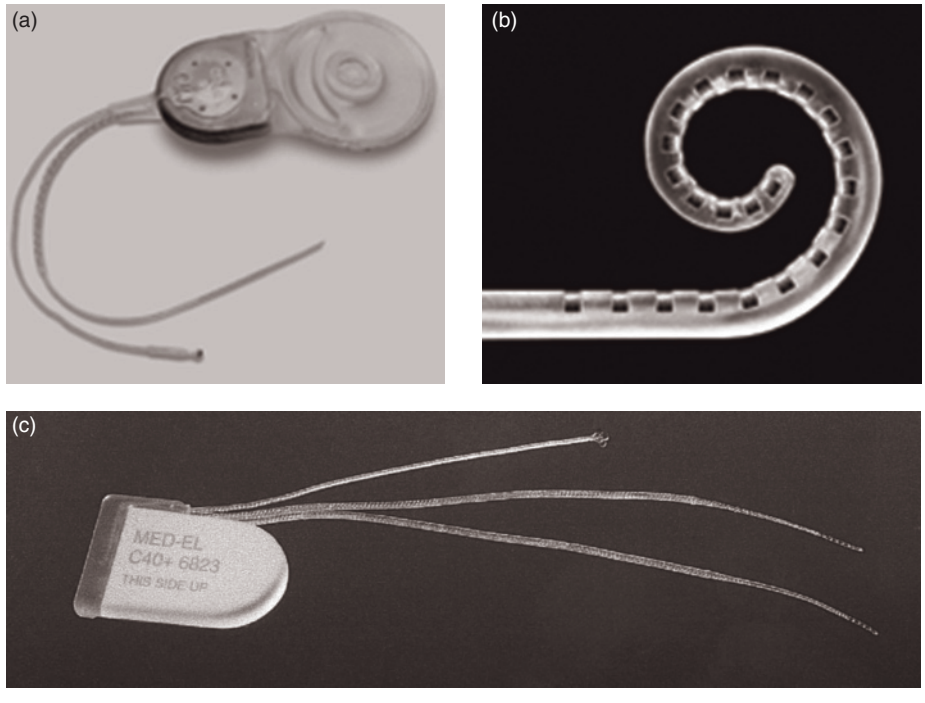


Figure 17.2 Devices (a) Nucleus CI24M with (b) magnification of electrode array. (c) Medel split electrode for use in ossified cochlea.

moment depending on the incoming signal. Consonant recognition is very important for speech understanding and as many consonants contain a lot of high-frequency information (e.g. sibilants and fricatives) it is important that the implant system transfer as much high-frequency information as possible. Speech processing is a very complex subject and further examination of the topic is beyond the scope of this chapter.

Most of the improvements in CI performance over the past 20 years have come from refinements in signal processing rather than changes in the electrode system. A notable advance has been the reduction in size in the speech processor so that the circuitry can now be contained in an ear-level package rather than a body-worn box. For some years the prospect of a fully implantable CI system has tantalised workers in the field, but as yet it remains no more than a fond hope.

Recently, CI manufacturers have developed electrodes that position themselves close to the spiral ganglion in the modiolus – so-called modiolus hugging electrodes. These have the theoretical advantage of more precise frequency tuning and less power consumption and so longer battery life. New generation implant systems also have the facility for neural response telemetry (NRT), which allows the clinician to record information from the auditory nerve. It is hoped that data acquired in this way may be of value in the mapping process.

Understanding of auditory processing in the central nervous system has increased as a result of CI research – in particular, the recognition of the complex phenomenon of neural plasticity

as it relates to speech and language acquisition. The newborn child does not speak. During the first year of life, its exposure to the sounds and patterns of speech is of immense significance, and at about the age of 12 months babbling commences.

Vocabulary and language acquisition proceed rapidly during the period between 18 months and 3 years in parallel with other cognitive and motor skills, and by the time the 'average' child goes to school at the age of 4, he or she will have an immeasurably large vocabulary and will speak fluently and more or less grammatically most of the time. This 'window of opportunity' for acquiring speech and language is not, however, open indefinitely. There are a few examples of children with normal hearing who have grown into adult life without hearing speech – for example, feral children or normal-hearing children of signing deaf parents. If these children are introduced to speech in their adult life, they are unable to learn speech because that critical period for speech acquisition has gone.

These mechanisms are highly relevant when we think about implanting individuals born deaf or who become deaf before language has been acquired. For such a child to have a chance of learning to speak with the implant, the device must be inserted before the window of opportunity closes. An adult who was born deaf is, like the normal-hearing feral child, highly unlikely to learn to assign meaning to the auditory stimuli coming from the implant and will not acquire speech or spoken language. On the other hand, an adult deafened in adult life will usually be a good candidate because the central auditory pathways have been previously stimulated. The primary auditory cortex and association areas have been previously programmed, and the CI in a sense simply reawakens it from its dormant state.

SELECTION AND ASSESSMENT OF CHILDREN

The first recipients of CIs were all adults, but as successful outcomes were obtained, and as safety and reliability were seen to be acceptable, the attention of implant teams turned to the greater challenge of children. The causes of profound hearing impairment in children have been examined in Chapter 1. Graeme Clark's Melbourne team was the first to implant children with the multichannel CI in 1985. Since then, many thousands of children have been implanted worldwide. The experience of the past two decades has taught us much about the selection criteria for this operation. It is clear that these criteria are changing as time goes by and that implant teams have become less conservative as they have learned more about the cognitive processes involved in language acquisition in the young child. Diagnostic techniques have become more sophisticated, most surgical challenges have been overcome and speech-processing strategies have enhanced the fidelity of information transfer. Ethical and cultural dilemmas have been faced and largely resolved.

Children who are considered for implantation fall into two main groups. The post-lingually deafened group comprises those children who have gone deaf after the acquisition of spoken language, for example, from meningitis, head injury or from the side effects of ototoxic drugs. They will be aged 3 or older. In linguistic terms and in the context of post-surgical rehabilitation, the post-lingually deafened child is not very different from the post-lingually deafened adult. The pre-lingually deaf children include those who are born deaf, either as a result of a genetically determined abnormality or from an intrauterine event, such as rubella or drug toxicity. In addition, post-natal illness in the first two years of life, such as meningitis and viral damage to the cochlea, may cause a profound hearing impairment before speech has been

acquired. Deafness occurring around the time that language is being learned is sometimes referred to as perilingual deafness.

Severity of hearing impairment

In the early days of adult implantation, a potential patient was one with virtually no measurable hearing in either ear on pure-tone audiometry. With time it was realised that pure-tone thresholds were less important than performance on speech audiometry, and at the time of writing in the UK, a maximum speech discrimination score of 40% or less would be acceptable to most implant teams. With children there are of necessity a different set of values. It should be recalled that the object of the assessment process is to select for implantation children who are likely to perform better with an implant than with a conventional hearing aid, but as Dowell et al.[1] have pointed out, it is difficult to define what the audiological profile of a child, and particularly an infant, should be. Clearly a speech audiogram has no place in the assessment of the pre-lingually deaf child, and even in the post-lingually deafened child, there may not be enough reliable speech perception information to allow one to predict post-operative performance.

Threshold assessment depends on the age of the child. In the child of 9 months or older, behavioural techniques performed with meticulous care provide the most reliable estimate of threshold, and of course, threshold estimations should be made in the best aided condition as well as unaided. Auditory brainstem response (ABR) is essential and may rapidly confirm the presence of a profound hearing impairment, but there are shortcomings to the test – in particular, its inability to give an accurate low-frequency threshold. The persistence of some low-frequency hearing in a congenitally deaf child is important because it tells the surgeon that there must be an auditory nerve present. In almost all instances, the child will have a trial with the most appropriate hearing aids for a period of some months, and a child will commonly be referred for assessment for implantation, having already had a trial with aids. It is felt that even if the benefit has been slight, this early stimulation of the auditory pathways is of value and has a beneficial effect on outcomes with the implant. One group that is recognised as being likely to benefit from implantation comprises those who have previously gained some speech and language using a hearing aid but whose hearing has deteriorated to a point where powerful aids no longer help. This is known as the ‘changeover’ group.

Age and duration of deafness

As with adults, a post-lingually deafened child will be implanted as soon after diagnosis and satisfactory assessment as possible. The minimum age for implanting a congenitally deaf child has come down progressively over the years as a result of earlier referral of potential candidates to implant teams, an increased awareness on the part of implant teams of the importance of the critical period for speech and language acquisition, as well as more rapid availability of funding on the part of purchasers of healthcare.

Until recently, the target age at which most teams would aim to implant a congenitally deaf child seemed to be 2 years old. The advent of universal neonatal screening has, however, obliged paediatric implant teams to review their protocols. Deafness may now be suspected at birth and confirmed on electrophysiological tests soon thereafter. It is becoming increasingly common to have a firm diagnosis of deafness, trials of hearing aids and pre-operative radiological assessments completed by the age of 6 months. As result, more and more children are

being implanted under the age of 1 year, and the results of such early intervention are being studied with great interest. At the other end of the scale, most implant teams would be reluctant to implant a congenitally deaf child over 7 years old, or would at least warn the parents of such a child that the benefits of implantation were likely to be limited.

Speech and language ability

As the numbers of congenitally deaf or pre-lingually deafened children coming for assessment increase and the age of implantation decreases, the relevance of speech and language assessments seems to be lessening. However, many of these children have a linguistic substrate based on some form of signing strategy, and this communication skill is increasingly being evaluated before implantation. In older post-lingually deafened children, there are many assessment tools available to evaluate speech production skills and language performance.

Middle-ear disease

Young children with a severe sensorineural hearing impairment are no less likely to suffer from middle-ear disease than other young children, and the most common condition to be recognised is otitis media with effusion (OME). This should be corrected, and the effects of any superimposed conductive hearing impairment negated before an accurate threshold estimation is attempted. The presence of OME would not be seen as bar to surgery by most surgeons. There is some difference of opinion as to whether an implant can be inserted in the presence of a ventilation tube. The instinctive feeling that a tube may act as a route for infection reaching the implant seems unjustified and many surgeons are happy to insert an implant with the tube still in place. It was asserted by some surgeons that by performing a cortical mastoidectomy and posterior tympanotomy in the approach to inserting the implant, one reduced the likelihood of the recurrence of glue ear. This claim has been refuted by the work of Migirov et al.,[2] who compared a group of children whose implant was inserted via the traditional transmastoid approach with a group whose implant was inserted via the suprameatal approach which does not entail mastoidectomy. There was no difference in the incidence of OME in the two groups. If glue ear occurs in an ear already implanted, there appears to be no contraindication to inserting a ventilation tube. Otitis-media-prone children should be treated appropriately prior to cochlear implantation with adenoidectomy and ventilation tubes.[3] If acute otitis media or mastoiditis occur following cochlear implantation, they must be recognised and treated promptly with the appropriate intravenous antibiotics and myringotomy if necessary. Subperiostial abscess requires drainage to avoid infection of the implant site and parents should be advised of the importance of a prompt medical opinion if the implant is not to be lost.[4,5]

Any more serious middle-ear condition such as perforation or cholesteatoma would require corrective surgery in a separate operation or at a staged surgery before the implantation could be performed.

Children with complex needs

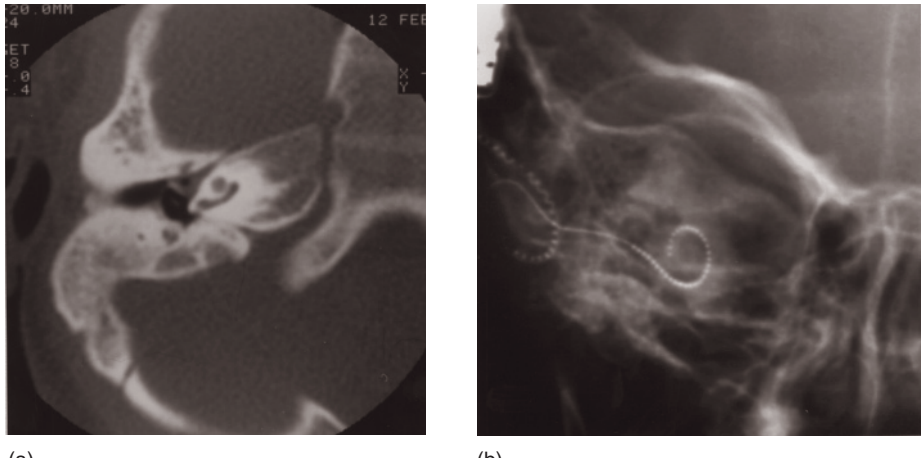
In recent years, an increasing number of children receiving CIs have other complex needs in addition to their deafness. They fall into two groups: those whose additional disabilities have been identified prior to implantation and those whose difficulties become apparent later.[6] In

the former group are conditions such as CHARGE association and the Jervell Lange-Nielsen syndrome (see Chapter 7). The latter group includes children who go on to develop autism, or with time reveal the extent of multisystem disability seen in Special Care Baby Unit children, resulting from prematurity, anoxia, hyperbilirubinaemia, and the effects of aminoglycoside antibiotics. Congenitally inherited progressive conditions, such as Usher syndrome, may be diagnosed at birth but not present their effects until later in life. The full central effects of infections such as meningitis and cytomegalovirus (CMV) may not be apparent until some time after implantation.

The existence of multiple handicaps is one of the factors that adversely affects the outcome of implantation, especially visual handicap and central language processing difficulties. Nevertheless, it may be that even limited benefit may be of value to these children, and most implant teams would now be prepared to consider these children for implantation always assuming that it is made clear to the families that there may be a limited benefit.

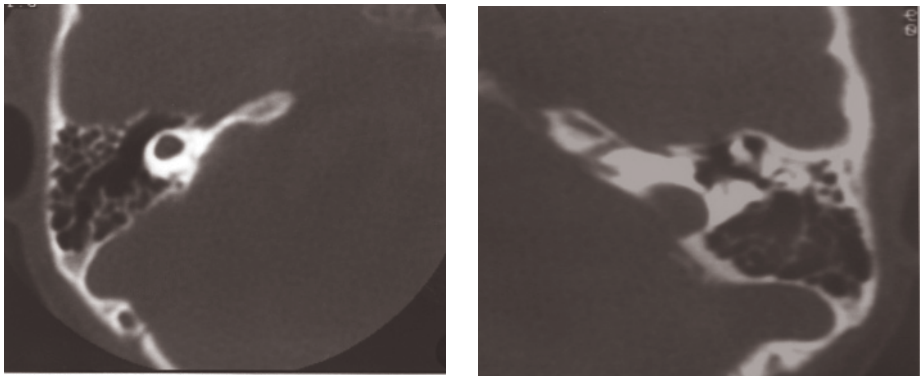
RADIOLOGY

Radiological imaging of the inner ear is essential in the evaluation of a potential implantee to establish whether any developmental or acquired abnormality of the inner ear is present (Figure 17.3). High-definition computed tomography (CT) and magnetic resonance (MR) imaging are the investigations of choice, enabling accurate delineation of cochlear anomalies. Not all dysplasias preclude implantation, and Mondini's deformity, the large vestibular aqueduct syndrome and some common cavity abnormalities have all been successfully implanted. However, pre-operative diagnosis might influence the pre-operative advice given to the child's parents regarding hearing outcome. The most important anomalies to prevent implantation are total agenesis of the cochlea and agenesis of the auditory nerves (Figure 17.4). Stimulation of the auditory nerve within the modiolus is a prerequisite for cochlear implantation, and the early diagnosis of agenesis of the nerve, though rare, is essential. (Figure 17.5).



(a) (b)

Figure 17.3 (a) CT scan of normal cochlea. (b) Full insertion of a multichannel electrode.



(a) (b)

Figure 17.4 CT scan showing (a) severe dysplasia (primitive otocyst) on right side and (b) total cochlear agenesis on the left side (same patient).

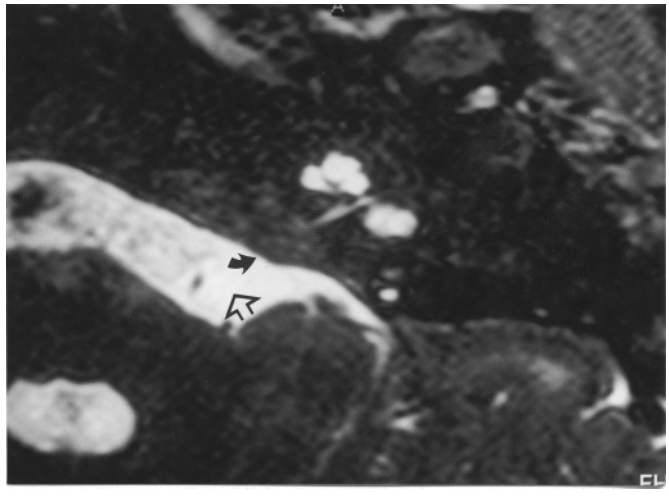
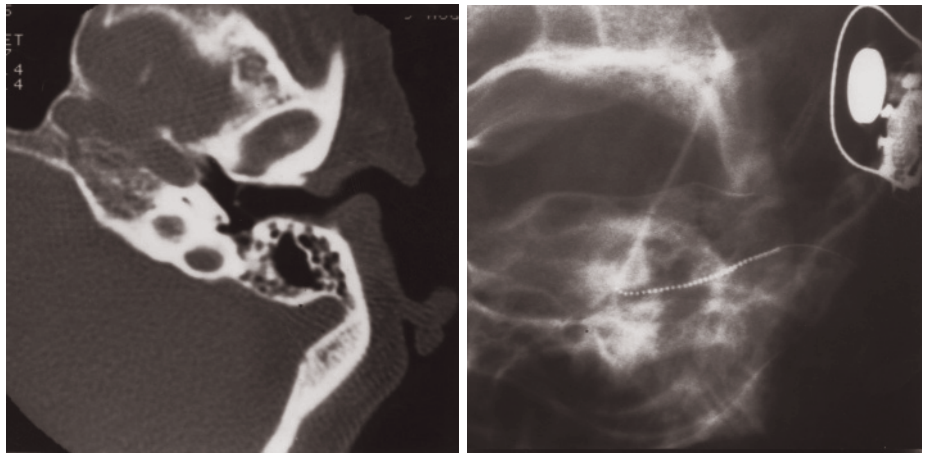


Figure 17.5 MRI scan showing almost complete agenesis of the internal auditory meatus (solid arrow) and the absence of the cochlear nerve in the cerebellopontine angle (open arrow).

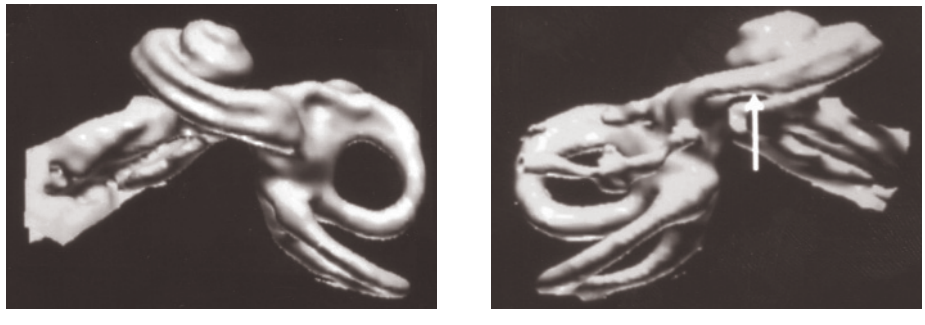
Acquired deafness is rarely an impediment to insertion of the electrode array, but labyrinthitis ossificans after meningitis is the notable exception. Progressive ossification within the cochlear lumen can rapidly prevent full insertion of the electrode array and, although not barring surgery, can reduce implant performance (Figure 17.6). Early patient referral for radiological evaluation is paramount, enabling urgent implantation should ossification be seen to be developing.

Recent advances in MR imaging allow differentiation of luminal fibrosis from endolymph, which is thought a possible precursor of ossification. Three-dimensional (3D) imaging of the cochlea now allows precise localisation of ossifying foci, valuable information that might



(a) (b)

Figure 17.6 (a) CT scan showing advanced obliteration of the cochlear lumen by new bone. (b) Partial electrode insertion in the obliterated cochlea.



(a) (b)

Figure 17.7 3D MRI images showing (a) normal cochlea and (b) partial loss of the image from the scala tympani as a result of obliteration.

advise the surgeon of possible difficulties and direct him or her to the use of a modified electrode (Figure 17.7).

FAMILY AND PSYCHOSOCIAL FACTORS

Cochlear implantation is expensive. The device itself costs a lot of money (approximately £15,000). Evaluation and surgery are expensive, and rehabilitation is a very costly, labour-intensive process. It is therefore essential that the implanted child has a loving and supportive home environment, with a family that recognises the investment in money and time and agrees to do its part in getting the most out of the technique. Parental expectations have to be care-

fully evaluated. There may be totally unrealistic ideas often picked from the 'gee whiz' tabloid press about how well they think their child is likely to perform with an implant. A series of counselling sessions with opportunities to meet children who have already been implanted and their parents is essential. They must realise the time scale within which progress may be expected and must be committed to long-term involvement with the child, his or her school and the implant team. The process of assessment must involve the educationalists with whom the child and family will be working and also the community paediatricians and general practitioners.

It is important to recognise the emotions that parents experience when confronted with the diagnosis of profound deafness in their child. Anagnostou et al.[7] highlight the grief felt by parents of very young infants and the fact that fathers tend to adopt a state of denial more than mothers. There is, in effect, a sense of bereavement. Parental depression is commonly observed due to child–parent communication difficulties that can lead to feelings of inadequacy and depression.[8] There may also be a sense of self-recrimination on the part of parents especially if the child has a genetically determined hearing loss. Whilst coming to terms with the diagnosis, parents then quickly have to reconcile themselves to the fact that their infant will have to undergo a major operation very soon if the deafness is to be helped. In evaluating a child's candidacy, the professionals involved in the process must be aware of the parents' preferences, goals, values, beliefs and expectations.[9]

SURGERY

Surgical implantation of the receiver unit and electrode array represents only a small part of the child's hearing habilitation. Nevertheless, it requires careful surgical planning and dedicated attention to detail in order to optimise implant performance whilst minimising complications. The objective is to attain full insertion of the electrode array within the cochlea and placement of the subperiosteal receiver unit in a position that allows comfortable placement of the overlying magnetic transmitter coil. Fortunately, the cochlea and the middle-ear structures are of adult size at birth, so there is no problem (a question often raised by parents and professionals alike), of having to put in a bigger implant as the child grows.

The design of the implant increasingly reflects the importance that is placed on patient comfort. The receiver unit is thin and conforms to the convexity of the skull. Its position, behind the ear, is carefully planned with post-operative placement of the magnetic transmitter coil in mind. Most surgeons now employ a small postauricular skin incision rather than the large extended endaural incision of previous years. It is sited away from the receiver unit to minimise the risk of wound breakdown. A separate periostial flap is elevated, and this is useful at the end of the operation to help stabilise the receiver stimulator. To gain access to the middle-ear cleft, a limited cortical mastoidectomy and posterior tympanotomy is drilled. The cortical mastoidectomy has an overhanging lip to facilitate stable placement of the electrode wires.

During the child's growth, the mastoid increases in size, slowly drawing on the available slack in the electrode lead within the mastoid bowl without displacing the electrode array. The posterior tympanotomy is performed with extreme care because the facial nerve lies in close proximity at this point in the operation. The occurrence of facial nerve injury is very rare indeed, but clearly this type of surgery should only be carried out in centres

with wide otological experience. Facial nerve monitoring is recommended. The next step is the creation of an opening into the scala tympani of the basal turn of the cochlea. Before the cochleostomy is performed, the stapes, promontory and round window niche are identified within the middle-ear cleft through the posterior tympanotomy. The cochleostomy is carefully drilled through the promontory, just in front of and slightly below the round window niche, about 2 mm inferior to the stapes. A slow speed drill is used in order to minimise vibration-induced trauma to the cochlea. The endostium of the scala tympani is identified as a white membrane and is gently incised with a fine microknife. Some surgeons like to inject a small quantity of hyaluronic acid (Healon) at this stage to act as a lubricant to ease the passage of the electrode into the cochlea, and to prevent the entry of blood and bone dust into the cochlea.

The electrode array is carefully threaded along the scala tympani with the assistance of a specially designed fine claw-like instrument. Some resistance may be met as the tip of the electrode reaches the anterior end of the basal turn. Modiolus hugging electrodes are designed in such a way as to minimise possible injury to the cochlear partition at this point (Figure 17.2). A muscle plug is inserted into the cochleostomy, to help support the array and prevent the potential for infection spreading from the middle-ear cleft.

At the end of the operation it is possible to stimulate the electrode and observe an electrically evoked stapedial reflex (ESRT).[10] This tells the surgeon several things: the electrode is in the right place, the electrode is functioning, there is a functioning eighth nerve, and the facial nerve has not been damaged. More robust information about position and function can now be obtained using neural response telemetry (NRT), or by eliciting the electrically evoked auditory brainstem response (EABR). Before wound closure, the receiver unit is secured within a bony well, flush with the outer table of the skull. It may be secured with ties or simply placed in a pocket under the periostium/temporalis muscle. In the immediate post-operative period, the correct electrode position is confirmed by plain radiograph. The slow process of hearing habilitation begins as soon as the surgical wound has healed.

Special surgical problems

Meningitis

Meningeal infection can spread to the cochlea through a persistently patent cochlear acqueduct, which enters the scala tympani close to the round membrane. In addition to causing deafness, the infective process leads to changes in the endosteal lining of the cochlea that may lead to partial or total obliteration of the cochlear lumen with new bone, which may make insertion of an implant difficult or indeed impossible. Minor degrees of ossification confined to the region of the round window are common and do not present much of a problem. The cochleostomy may bypass the affected area, but if not, it is a simple matter to drill past it to a clear cochlear lumen. Occasionally, the scala tympani is completely obliterated by new bone formation. There are various possible solutions to this problem. One option is to attempt an insertion into the scala vestibuli, which is less likely to be affected by ossification. This is done by removing the incus and stapes, and drilling just in front of the oval window. An alternative is to cut a gutter in the bone of the promontory and simply lay the electrode in it in the manner described by Gantz, McCabe and Tyler.[11]

The manufacturers have come up with two modifications in electrode design that also address this problem. One is the compressed electrode array, which is shorter than the standard

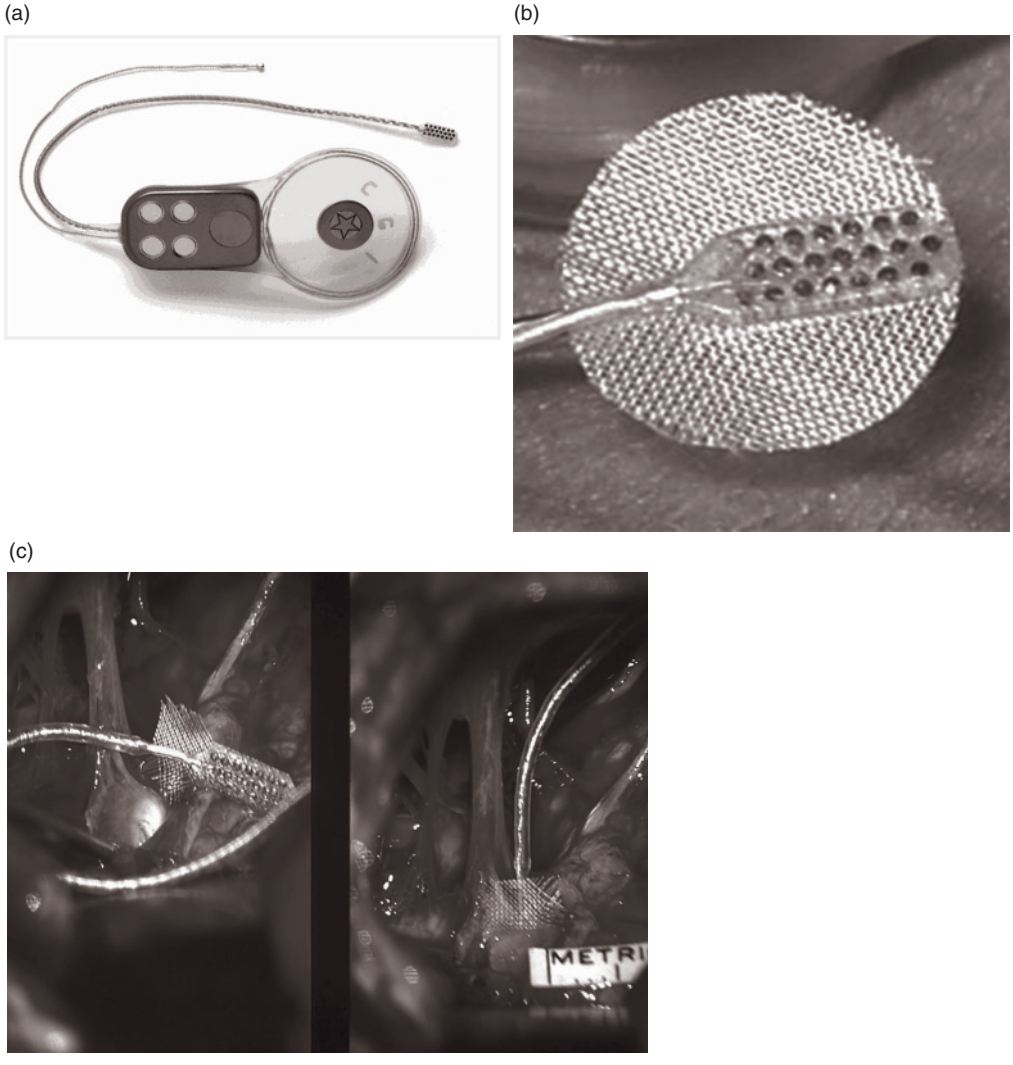


Figure 17.8 An auditory brainstem implant (a) shows the ABI low power, (b) is a close-up of the electrode array and (c) shows the device about to be inserted (L) and in position (R).

array but still has the same number of electrodes. This is clearly an advantage if there is only a very limited space in the cochlea. Alternatively, one can use a so-called split electrode array. There are two short arrays, each carrying half the electrodes. One is inserted into a gutter in the basal turn and the other through a separate cochleostomy into the middle turn. The other possibility, and a fairly controversial one, is to insert an auditory brainstem implant (ABI) (see further discussions). The most important solution to the problem of complete ossification is avoidance if possible. This necessitates early identification of hearing loss after meningitis, and early MR imaging to pick up early ossification. Cochlear implantation may become an urgent priority for these children.

The dysplastic cochlea

There is a wide spectrum of dysplasias and the classification of these has been clarified recently by Sennaroglu and Saatchi.[12] At one end of the scale, a true Mondini dysplasia, with a normal basal turn and common middle and apical turns, is not a difficult problem, and the results are as good as in normally formed cochleas. The same is true of the isolated large vestibular aqueduct commonly seen in Pendred's syndrome. Common cavities can be implanted but the problem here is knowing the extent of neuronal survival and the location of the neural elements that are to be stimulated. On the assumption that any surviving neural elements are situated round the periphery of the cavity, modified electrode arrays have been designed to deal with this specific problem. CSF fistula is also a risk because of communication between the subarachnoid space and the inner ear through a dehiscent lateral wall of the internal meatus. This is usually easily controlled with packing. Severe degrees of cochlear dysplasia are more of a problem, notably cochlear hypoplasia or total aplasia (Michel deformity). These ears are not implantable, and the ABI may offer a possible alternative.

Complications of cochlear implantation

Surgical complications are rare. Facial nerve injury almost never occurs. Wound breakdown can occur but is not common. If the device does get infected then it almost inevitably has to be removed because of biofilm formation in the silastic casing. The risk of device failure in children, though higher than in adults, is low at about 6%.[13] Children may be more prone to head trauma than adults, and the implant may be more vulnerable for this reason, but re-implantation in the event of device failure does not usually present a problem for the surgeon. In 2002, concerns were raised that cochlear implantation might predispose a child to meningitis. The very few cases that did occur seemed to be associated with the use of the Clarion device, which had an electrode positioner that was thought to cause microfractures from the cochlea into the internal auditory meatus. This device is no longer in use.

SWITCH ON AND MAPPING

Before the implant system can be switched on, it must be customised so that the current levels reaching each electrode during stimulation are correct for the individual patient. Each electrode has a threshold at which the stimulus just becomes audible (T level). Similarly, each electrode also has a second level above which the electrical stimulus causes discomfort or pain rather than hearing (C level). The difference between the T and C levels is the dynamic range for that electrode. Stimulus levels for each electrode should lie within the dynamic range. Below the T level, there will be no auditory percept. Above the C level, there will be pain. T and C levels should ideally be worked out for every electrode in the array, and there may be considerable variation in these levels and in the dynamic range along the array.

The process of establishing these levels is known in CI jargon as 'mapping'. Mapping is usually easy in linguistically competent adults who will tell you very quickly if something is audible or painful. Pre-lingually deaf children cannot do this. Furthermore, a child who associates the implant with pain or discomfort is not likely to want to wear the system and is, indeed, likely to reject the implant. Accurate mapping of a little child is one of the greatest skills in audiology, requiring great patience and expertise in conditioning. A good team will often be able to map the complete electrode array of a toddler in no more than a few sessions, although

it may be felt prudent to introduce only a few electrodes at a time until the child has become accustomed to the experience of sound. Electrodes that appear to produce unwanted (non-auditory) effects can be simply left out of the map.

Work with adult CI patients has revealed a remarkably high incidence of non-auditory effects on one or two channels, and one must assume that the incidence is at least as high in children. Data from NRT and EABR recordings may prove of value in estimating thresholds, but many experienced teams feel this is unnecessary. Once a map has been established the information has to be programmed via a computer interface into the speech processor where it will remain permanently or until the map is changed at a subsequent habilitation session. At this stage, the implant system is finally switched on. At future mapping sessions T and C values are checked, additional electrodes introduced, rogue electrodes are eliminated and general fine-tuning of the system carried out. It is common to find that these psychophysical values change with time and adjustments have to be made.

HABILITATION

Habilitation has been defined as 'the process by which professionals support a child and family in adapting to hearing impairment, getting used to a hearing device and developing the child's language and communication skills'.[14] This definition embodies a general principle that is as applicable to hearing aids as it is to CIs. The habilitation of a child with an implant is based on this principle with modifications and additions that are specific to the use of an implant.

The basic aim of habilitation is to give the child access to education through spoken language. The individuals involved in the process are the child's family, teachers of the deaf, and the audiologists, speech and language therapists and scientists on the CI team. The family has to have a realistic idea of the time frame within which progress may be expected and to structure communication strategies appropriately. For example, a 2-year-old who has been deaf from birth and has only had the implant for only 6 months will not immediately acquire the linguistic skills of a normal-hearing 2-year-old. This may seem self-evident, but parents need to understand this and to appreciate the period of listening and learning that is essential in the process of catching up. They must know that listening and learning are continuous processes that should be integrated seamlessly into the normal day and not consigned to an allocated half-hour period in the day. The child should be learning from his or her implant during every activity of the day, for example, dressing, eating, playing and helping in the house. Within each of these routine activities specific linguistic goals can be set.

The implant team and the teachers of the deaf need to ensure that goals are set that are appropriate to the age of the child at implantation, the duration of deafness and the length of implant use. For the child of school age, it is important that these goals are incorporated into the general educational programme. The motivation of the child has to be sustained, and the hope is that increased enjoyment of listening and hearing will provide that motivation rather than artificial rewards for success, which carry the negative corollary of no rewards for failure. The implant team's involvement lies in ensuring that the device is functioning properly and is correctly programmed. It has a major role in ensuring that realistic goals are set and in liaising with teachers of the deaf and other professionals. The team will frequently visit the child in his or her domestic or educational environment and give advice and guidance to family and professionals. It provides major support to families who, despite extensive pre-operative coun-

selling, may feel that progress should be faster than it is. The team also reviews all children at regular intervals to assess progress and modify schedules as necessary.

OUTCOMES OF COCHLEAR IMPLANTATION IN CHILDREN

Cochlear implantation in children has been controversial, and it has certainly been expensive. From both ethical and economic standpoints, it is essential that the outcomes are held up to close scrutiny. Adult cochlear implantation in the UK has been the subject of a very rigorous examination by the Medical Research Council.[15] It was shown to be an effective and cost-effective treatment for certain profoundly deafened adults. In children, the results are most dramatic. Outcomes are evaluated by a series of assessments that have been developed specifically for CI children

The most important skills that are assessed are speech perception, speech production and language. Auditory skills may be categorised on a hierarchy of increasing difficulty: detection, discrimination, recognition and comprehension of speech. Various different types of test material are employed, which, of course, have to be appropriate to the linguistic abilities of the child. Closed-set tests involve the selection of a correct choice from a limited choice of usually four pictorial options. They can be designed to assess specific information about vowel or consonant discrimination. They do not provide information about abilities to discriminate connected speech. This may be obtained from open-set sentences, such as BKB sentences or CID sentences or from continuous discourse tracking.

Speech perception performance at any time, as well as progress with time, may be categorised on a scoring scale. The Melbourne scale described by Dowell and Cohen[16] has seven steps from Category 1 (detection of speech sounds only) to Category 7 (good open-set speech perception >50% phoneme score). The Manchester scale recognises 10 levels of performance. The point of entry on to the scale and the rate of progression to higher levels depend on the previous linguistic experience of the child. A post-lingually deafened child may progress to higher levels rapidly, whereas in the case of the congenitally deaf child, the process may take 3 or 4 years.

There is now no longer any doubt about the spectacular success of cochlear implantation in children. As previously stated, post-lingually deafened children rehabilitate like post-lingually deafened adults. The most dramatic results, however, are seen in the congenitally deaf or pre-lingually deafened children, for whom the acquisition of open-set speech perception should be regarded as the norm.[1] There are a number of variables that may have an influence on outcome; the most important of which is most certainly age at implantation.

Kirk et al.[17] reported that congenitally deaf children implanted by the age of 3 years had significantly faster rates of language development than children implanted later. In an excellent study, Govaerts et al.[18] demonstrated a startling difference in auditory performance in children implanted at the age of 2 years compared with those implanted at the age of 4. After 4 years of implant use, the 2-year-olds were achieving ceiling scores on central auditory processing (CAP) evaluation, whereas the 4-year-olds lagged badly behind. The majority of the early implanted children were able to take their places in mainstream education often with some extra support.

More recently there have been an increasing number of reports of infants implanted under the age of 2 years. Looking at children implanted between the ages of 5 and 20 months, Schauwers et al.[19] reported that prelexical babbling commenced at only 1–4 months after

activation of the implant. In the youngest infants, babbling commenced at a similar chronological age to normally hearing children. Children implanted in the first year of life showed normal CAP development after only 3 months of implant use. Identical outcomes were reported in the study reported by Colletti et al.,[20] and Dettman et al.[21] reported that the rates of receptive and expressive language growth for children receiving implants before the age of 1 year were significantly greater than the rates achieved by children implanted between 1 and 2 years and matched the growth rates achieved by normal-hearing peers.

The importance of age at implantation has emphasised by many authors including Geers et al.,[22] who found that children implanted between their first and second birthdays achieved oral receptive vocabulary levels within one standard deviation of their hearing peers. Speech intelligibility also seems to be influenced by age at implantation. Svirsky et al.[23] have demonstrated that cochlear implantation before the age of 2 years leads to significantly better speech intelligibility outcomes than does later implantation.

Other variables that have an influence on outcome are cognitive ability, family environment, pre-implant hearing levels and the use of hearing aids, as well as duration of implant use and appropriate mapping. Cognitive impairment can be a factor in children with multiple handicaps either congenital or acquired (e.g. as a result of meningitis or CMV infection).

The evidence is, therefore, driving early implantation, but there is a certain disadvantage that should be highlighted. A number of children will be implanted before there has been time for co-morbid conditions associated with the hearing loss to emerge, in particular central processing or cognitive disorders. Autism, which may have an unpredictable effect on a child's performance with an implant, is a condition that is rarely diagnosed before the age of 2 years. For these reasons, it is always best to be guarded in predicting outcomes with the parents of any child who is about to have an implant.

BILATERAL COCHLEAR IMPLANTS

The question of whether second-side implantation might confer a worthwhile additional benefit over unilateral implantation is one which is receiving considerable attention at present. There are still many unanswered questions. In adults, studies have demonstrated a marked improvement in sound localisation with binaural implantation.[21–23] When considering one of the other main benefits of binaural hearing, there is an advantage from a second-side implant in overcoming the head shadow effect. There is little evidence, however, of a worthwhile squelch effect.[24–26] Of course, with bilateral implantation one knows that the better ear will be implanted. Still to be clarified is the question of whether there is any difference in benefit if the two ears are implanted simultaneously or serially, and if serially how important the time interval between implants is.

In children, there is conflicting evidence regarding localisation. Litovsky et al.[27] and Beijen et al.[28] reported significant benefit, but Galvin et al.[29] found no evidence of improved localisation in the bilateral condition. As yet there are no long-term data on the effect of binaural input to the maturing auditory pathways, in terms of language development. Such information inevitably would take years to gather and a powerful prospective randomised controlled trial comparing unilateral with bilateral implantation presents considerable organisational difficulties.

Wolfe et al.[30] looked at a series of 12 children with bilateral implants and found better speech discrimination in noise compared with unilateral performance. There appeared to be an age-related effect with those receiving bilateral implants under the age of 4 years perform-

ing better than those over the age of 4 years. Peters et al.[31] looked at 30 children with a wide age range and found that sequential implantation has the potential to improve speech perception abilities and overcome the head shadow effect. Younger implanted children achieved higher scores than the older subjects. Again, the issue arises of simultaneous or sequential implantation.

Considering the concepts behind activity-dependent developmental plasticity in the auditory system, it would seem, in theory, to be more sensible to implant both sides at the same time, rather than to try to impose a new input from a delayed second-side implant on to the pathways laid down from the first implant. Children who are young at first implantation and have a short (or no) interval between implantations should be better able to integrate the two inputs. These dilemmas still need further study ideally with functional brain imaging such as positron emission tomography (PET) scanning.[32]

THE AUDITORY BRAINSTEM IMPLANT

The ABI is a development from CI technology and was originally produced for patients who had lost their cochlear nerves as a result of neurofibromatosis type 2 (NF2). In NF2, tumours (acoustic neuromas, vestibular schwannomas) develop on both audiovestibular nerves and lead to a progressive and usually profound bilateral deafness. Surgery to remove the tumours contributes to the cochlear nerve damage in many cases.

Cochlear implantation demands an intact auditory nerve to transmit the signals from the ear to brainstem, so is not appropriate in NF2. The ABI bypasses the lost nerve by stimulating the auditory pathway at the level of the cochlear nucleus in the brainstem. This is located in the lateral recess of the fourth ventricle just inside the foramen of Luschka, and can usually be identified without too much difficulty at the end of the operation to remove the vestibular schwannoma. A silastic carrier with 21 small disc electrodes is placed on the surface of the nucleus. Its precise location can be confirmed by stimulating it peri-operatively and recording electrical responses (EABR) from scalp electrodes.

The results from ABI in NF2 are less spectacular than cochlear implantation in appropriate patients.[33,34] Most recipients gain a valuable awareness of environmental sounds. Open-set speech discrimination scores using the ABI alone are not good, however, but when combined with lip reading, the performance is encouraging and can approach 100%, although the range of performance is wide. The performance using ABI and lip reading together is generally significantly better than the sum of the ABI alone and lip reading alone. There are a number of possible reasons for this relatively unimpressive outcome when compared with cochlear implantation. The cochlear nucleus may sustain damage with cell loss as a result of pressure from the tumour, from surgical manipulation or from the effects of stereotactic radiosurgery. Furthermore, the tonotopic arrangement in the nucleus is not as favourable as that in the cochlea. Instead of running across the surface of the nucleus, which would make it accessible to a surface electrode array, the map runs obliquely into the depths of the nucleus. Attempts to access these deeper areas using a special penetrating electrode have as yet met with little success.

Interest is, however, growing in offering ABI to certain totally deaf patients without NF2 who are not suitable for cochlear implantation. There are two groups of patients under consideration: adults with acquired hearing loss and children born with cochlear nerve aplasia, severe dysplasia or severe degrees of cochlear dysplasia.[35] The former includes adults with unimplantable cochleas from advanced otosclerosis or post-meningitic obliteration and patients

with bilateral cochlear nerve avulsion after severe head injury. Early results from these patients who, of course, have normal cochlear nuclei indicate that very good outcomes are possible, with speech discrimination approaching levels seen the in best of multichannel cochlear implantation.[36] Because of the good results from these non-tumour post-lingually deafened adults, a small number of teams have been encouraged to implant young children with cochlear nerve aplasia or severe cochlear dysplasia or severe degrees of inner-ear malformation that prevent cochlear implantation.[37,38]

Bearing in mind the critical period for language acquisition, it is clear that to be most effective, the surgery should ideally be carried out by the age of 2 years. Although it is clear that these children can respond to sound, and begin to imitate elements of speech, it is as yet too early to say whether they will be able to acquire speech and language using the ABI. This should become clearer over the next few years. Surgery to insert an ABI is considerably more invasive than cochlear implantation and carries more risk. Programming and mapping of an ABI in an infant requires considerable skill and experience. The nature of the surgery, together with the as yet uncertain outcomes, presents the surgeon and the parents of these small children with difficult ethical decisions in deciding whether to proceed with the surgery.

THE FUTURE

One of the greatest challenges is the identification of the factors that affect the changing plasticity in the auditory pathways as the child ages. Neurotrophins are assumed to be responsible for the establishment of neural networks in the primary auditory cortex and association areas during the first few years of life. They are also assumed gradually to be switched off as the child gets older, with the result that older children and adolescents gain little speech recognition from implantation.

Clark[39] has suggested that auditory plasticity might be restored by delivery of the critical neurotrophin to the auditory system by means of the implant device itself. This might in turn trigger the release of neurotrophin in the cochlear nucleus, which might then reactivate the gene for the neurotrophin. It is envisaged that neurotrophin release might cause neural sprouting to occur at higher levels in the auditory system and encourage neural connections in the auditory pathways. If the window of opportunity could be reopened by pharmacological means, certain individuals at present debarred from implantation might once again be deemed suitable.

Less ambitious might be the use of nerve growth factors or neurotrophins to increase the population of neurones in the cochlear nerve available for stimulation with the implant. Of course, it is equally possible that a similar approach, by stimulating regeneration of the hair cells of the organ of Corti, could provide a more effective solution to the problem of sensorineural deafness than cochlear implantation itself. The experimental evidence at present suggests that limited regeneration of hair cells may be possible in the guinea pig vestibular system, but there is no evidence to indicate that regeneration of cochlear hair cells is an imminent likelihood.[40]

CONCLUSION

Cochlear implantation has revolutionised the management of severe to profound deafness in children, whether congenital or acquired. Assuming implantation is carried out whilst the

auditory system still retains plasticity, most children can be expected to gain open-set speech discrimination and to go to mainstream schooling. Implantation is safe and the surgery is routine. Assessment and habilitation require the skills of a multidisciplinary team and are expensive and labour intensive. Although the technique has progressed from the experimental to the routine in a relatively short period of time, familiarity should not lead to complacency. It should only be available in a relatively small number of dedicated units, equipped and staffed at all levels to the highest standards.

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18 Managing the listening environment: classroom acoustics and assistive listening devices

D. Toe

INTRODUCTION

Hearing aids are usually selected, verified and fitted in an audiological clinic, a quiet non-reverberant world where people face each other and speak clearly. However, children have to acquire communication skills and be educated in the real world. Homes and classrooms are often characterised by reflective surfaces that absorb critical high-frequency sounds and reflect low frequencies. Background noise is always present, whether it is the hum of traffic noise or the babble of other voices. Teachers and parents speak at less than optimal distances and may neglect to face the child. For these reasons, hearing aids alone often cannot provide a child with a clear speech signal. Attention to room acoustics, an understanding of the needs of developing listeners and an assistive device are needed to ensure that children can maximise their auditory development. Difficult listening conditions impact on all children. Efforts to improve the listening environment and enhance teacher input not only benefit the child with a hearing loss, but may impact on academic performance, on-task behaviour and stress levels for all students.[1]

LISTENING IN THE REAL WORLD

Sensation level, noise, reverberation and stage of development all impact on the perception of speech by children and young people. Moreover, these factors combine to have a synergistic effect on speech perception such that the combined effect of two factors may be greater than the sum of the individual effects seen when each factor is measured alone. Difficulties with receiving sensory input are likely to be further exacerbated by the dynamics of classroom interaction whereby students need to attend to many speakers, manage small groups and interpret both direct and implied messages from teachers and peers.

DEVELOPMENTAL ASPECTS OF SPEECH PERCEPTION

Several studies have shown that speech perception is a developmental skill in children, including those with and without a hearing impairment.[2,3] It has been suggested that young children show poorer sensitivity than adults to the acoustic cues needed to make consonant discriminations and may have immature sensory processing skills at both the peripheral and the central level of the auditory system.[4] The perception of consonants in quiet at adequate sensation levels was shown to be significantly more difficult for young children aged 6–7 years than for

older children and adults.[2] Stelmachowicz et al.[3] reported similar findings using sentences with high predictability and low predictability. They demonstrated a developmental continuum from 5-year-olds through to adults and showed that young children require a higher level of audibility than do older children and adults in order to perceive speech. The very young children in this study were also unable to make use of context to assist with the speech perception task, reflecting the interaction between language skills and speech perception. Stelmachowicz et al. also investigated the speech perception skills of children with mild to moderate hearing loss and showed a similar pattern of development, with very young children requiring higher signal input levels in order to achieve 100% word recognition. Interestingly, these children performed very similarly to hearing children of the same age. A much bigger difference was observed in speech perception performance between all of the children tested and adults. These studies suggest that children do not hear speech in the same way as adults. They require greater audibility to perceive sentences, and they show a clear developmental trend in their capacity to discriminate consonant phonemes. Children may not demonstrate adult-like speech perception skills until their mid- or even late teen years.[2]

UNDERSTANDING SPEECH IN NOISE

Background noise is generated by a range of sources. It may be internal to a room, created by either equipment or people, or it may be external to a room from a range of natural and artificial sources. What is most relevant to speech communication and listening is not so much the absolute noise level but the relationship between the signal (i.e. the level of the speaker's voice) and the background noise. This is known as the signal-to-noise (S/N) ratio. Children, including infants and early adolescents, appear to need a higher S/N ratio than do adults with well-developed auditory systems and communication skills.[5–7] A study by Elliott[5] using sentences with high and low predictability indicated that normally hearing children aged between 11 and 17 years performed more poorly at 0 and –5 S/N ratios on high-predictability sentences than did adults, but there were no differences in performance for low-predictability sentences. This suggests that noise levels impacted on these young people's abilities to use semantic context clues in the presence of high levels of background noise. In contrast, 9-year-olds performed more poorly at these S/N ratios on both low-predictability and high-predictability sentences, suggesting that young children with normal hearing also experience a masking effect for words in sentences.

Special groups of hearing children have also demonstrated the need for an improved signal-to-noise ratio. These include children for whom the language of the classroom is not the language of the home. In noisy classrooms, where the S/N ratio is 10 dB or less, processing English is significantly more difficult for children for whom it is a second language as compared with their native English-speaking peers.[8]

Children with hearing loss are even more likely to struggle with speech perception in the presence of background noise. This includes children with unilateral hearing loss who do not wear hearing aids. Ruscetta, Arjmand and Pratt[9] showed that children with severe to profound unilateral hearing losses required an improved S/N ratio of approximately 3–4 dB to hear sentences and nonsense syllables as well as their hearing peers. Finitzo-Hieber and Tillman[6] investigated monosyllabic word discrimination in quiet and at S/N ratios of +12 dB, +6 dB and 0 for children with mild to moderate sensorineural hearing loss and children with normal hearing. All children experienced a reduction in speech perception scores as the S/N ratio

decreased, but children with impaired hearing experienced the biggest drop, from 87% in quiet to 42% at a S/N ratio of 0. In high levels of noise, even children with only mild losses are receiving less than half of the sensory input. This must have a significant impact on learning.

UNDERSTANDING SPEECH IN REVERBERATION

Reverberation refers to the way that sound waves reflect off various surfaces in an enclosed space before reaching the listener's ear. A listener hears the initial sound directly from the source followed by its reflected waves. For most listeners, these reflections are merged together so that the listener interprets reverberation as a single sound. Reverberation is measured in a room as reverberation time (RT); this is the amount of time it takes for a signal to reduce or decay by 60 dB once the sound has terminated. High levels of reverberation (1.2 seconds plus) usually degrade speech perception because the reflected energy overlaps with the direct signal and acts as a masker.

There is convincing evidence that young children do not understand speech well in reverberant listening conditions.[6] It has been suggested that young children have not yet developed the precedence effect that allows adults to suppress the sound reflections in a reverberant room and hear a single unified sound. This ability improves speech understanding in reverberant conditions.[10] Litovsky[11] has suggested that normally hearing children start to develop the precedence effect at about 5 years of age. Up until this age, they still exhibit much poorer speech understanding in reverberant conditions than adults because they continue to hear some echoes or reflections as independent sounds rather than blending them with the sound heard directly from the sound source. This suggestion is supported by empirical research. Neuman and Hochberg[12] studied the perception of nonsense syllables in groups of children aged 5 to 13 years under two conditions of reverberation, 0.4 and 0.6 seconds. Longer reverberation times resulted in poorer speech perception scores for all ages, with 5-year-olds showing the biggest impact of the poorer RT, with a mean phoneme perception score of 63% as compared with the speech perception score in quiet of 96% correct. A developmental continuum was observed, peaking at age 13 years and suggesting that speech perception scores of normally hearing children in reverberation are like young adults by the time such children reach 13 years of age.

For listeners with hearing impairment, the impact of reverberation is even more significant. Finitzo-Hieber and Tillman[6] compared speech perception scores in reverberation for normally hearing children and children with mild to moderate hearing loss. Mean monosyllabic word scores of 87.5% for children with hearing impairment and a hearing aid dropped to 74% at a 0.4-second RT and then decreased dramatically to just 45% when tested in a room with a 1.2-second RT. Normally hearing children exhibited a much smaller reduction in speech perception scores, from 94.5% down to 76.5% at a 1.2-second RT. Classrooms with poor acoustics are likely to make listening very difficult for students with hearing impairment and cannot help but have a negative impact on learning.

COMBINED EFFECTS OF REVERBERATION AND NOISE

Finitzo-Hieber and Tillman's study[6] made a significant contribution to our understanding of the devastating impact of classroom noise and reverberation on speech understanding by both

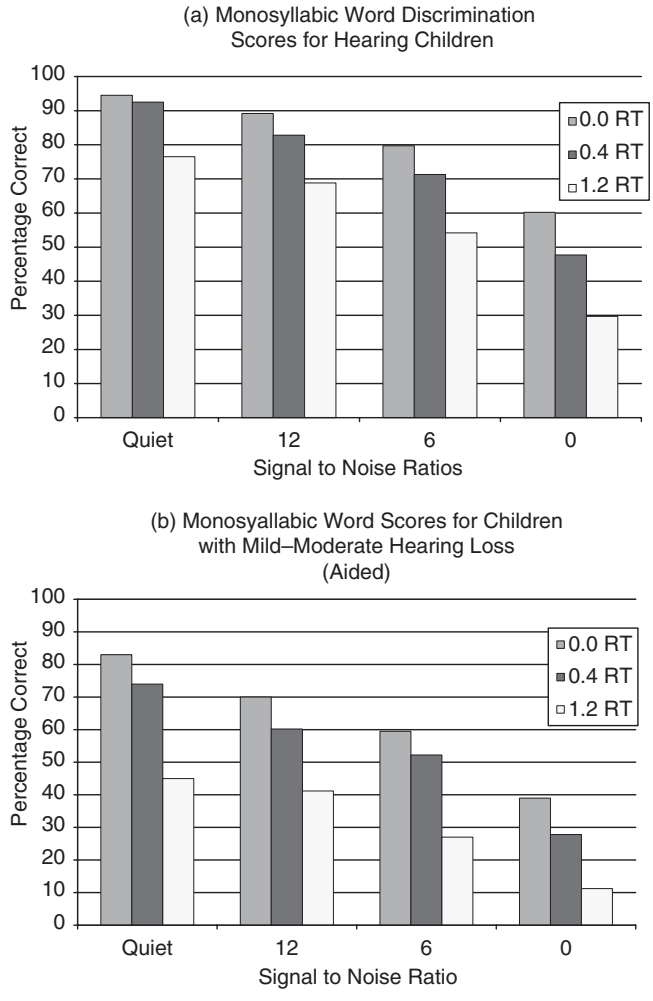


Figure 18.1 The impact of noise and reverberation on speech perception for (a) hearing children and (b) children with mild to moderate hearing loss and hearing aids. Data adapted from Finitzo-Hieber and Tillman.[6]

hearing and hearing-impaired children. A remarkable finding from this study is the way that noise and reverberation combine to impact on speech perception skills. Results from Finitzo-Hieber and Tillman's study are reported here as a column graph to clearly show the synergistic effects of noise and reverberation (see Figure 18.1).

For the hearing group, if the impact of noise (34.3% decrease from quiet to 0 S/N ratio) is added to the impact of reverberation (19% decrease from 0 to 1.2-second RT), then the combined impact of noise and reverberation would be expected to be 53.3%. However, the actual impact of listening in a 1.2-second RT at a 0 S/N ratio was measured at 29.7%, indicating a reduction in scores of 64.8%. This was more than 10% greater than would have been predicted by just adding the effects of noise and reverberation together. A similar pattern was observed for children with impaired hearing. It appears that high levels of noise and reverberation interact to create listening conditions that make listening very difficult for hearing children. With

a mean score of just 11.2%, correct listening would appear to be almost impossible for children with mild to moderate hearing loss. With the vast majority of children with hearing loss now learning in regular classrooms,[13] it is essential that close attention is paid to assessing classroom acoustics and improving them where necessary.

In summary, listening to speech in the classroom may be a very demanding task for a child with hearing impairment. For many children, it is likely to be a case of double or triple jeopardy for learning, particularly if they are very young, speak a different language at home and must cope with poor classroom acoustics and high levels of background noise. Each child's learning environment must be assessed and compared with appropriate standards so as to ensure that children have some chance of meeting their own potential.

RECOMMENDATIONS FOR NOISE AND REVERBERATION IN CLASSROOMS

This body of evidence outlining the impact of poor acoustics in classrooms has led to the development of an American standard on classroom acoustics (ANSI S12.60-2002 American National Standard Acoustical Criteria, Design Requirements and Guidelines for Schools[14]). This standard was developed by a working party representing a wide range of professionals. It recommends that unoccupied classroom background noise levels should not exceed 35 dBA. This would allow a speaker's voice to reach the child's ear at an S/N ratio of 15 dB. The recommended reverberation time for classrooms is between 0.2 and 0.6 seconds. Over 25 years ago, Fourcin et al.[15] made similar recommendations for facilities for deaf children; however, the ANSI standards acknowledge that all children are affected by poor classroom acoustics and require clear speech input to learn. In addition, with so many children with hearing loss, auditory processing disorders, learning difficulties and language delays learning in regular classrooms it is essential that such a set of standards is applied in all classrooms and all educational systems.

NOISE AND REVERBERATION FOUND IN REAL CLASSROOMS

Classrooms can be noisy places, even though teachers work hard to ensure that students listen quietly when important instructions are given. Several researchers have investigated classroom acoustics found in real classrooms. Knecht et al.[16] measured the noise levels and reverberation time in 32 unoccupied regular elementary classrooms in three districts in Ohio, USA. Noise levels ranged from 34.4 to 65.9 dBA. Only four classrooms met the ANSI standard of 35 dBA. The most likely noise offenders were heating and air-conditioning systems or noisy equipment such as fish tanks. Reverberation times ranged from 0.2 to 1.27 seconds. Thirteen of the 32 classrooms exceeded the ANSI standard of 0.6 seconds. Classrooms that exceeded the standard were likely to be large with high ceilings. All rooms that had ceilings of 10 feet (3.3 metres) or less had acceptable reverberation times. Earlier studies have also reported high levels of noise in classrooms, ranging from 41 dBA in unoccupied classrooms to 65 dBA in occupied rooms.[17,18] More importantly, the S/N ratio measured in classrooms where children with hearing impairment are learning has been consistently found to be below +15 dB, ranging from −7 dB to +5 dB.[19,20] In general, noise levels are higher in classes of younger students and particularly high in kindergarten classrooms.[21] Long reverberation times in classrooms have also been reported in several previous studies, with RTs found to range from 0.6 to 1.2 seconds

(see Crandell and Smaldino[1] for a review). Given these findings, it is essential that any professional supporting children with hearing difficulties should assess the acoustic characteristics of the child's classroom.

MEASURING NOISE LEVELS AND REVERBERATION IN CLASSROOMS

Noise levels

Audiologists, teachers and other professionals need to be able to ascertain if a classroom meets the criteria set out in the ANSI standard and to check if the classroom is a suitable learning environment for a child with hearing impairment or other learning difficulties. Measuring background noise levels is straightforward and only requires access to a sound-level metre of reasonable quality and a tripod.

The ANSI standard (S12.60–2002 American National Standard Acoustical Performance Criteria, Design Requirements and Guidelines for Schools[14]) provides guidelines for measuring noise in classrooms. These are outlined in Box 18.1.

Box 18.1 Guidelines for measuring noise in classrooms. Adapted from ANSI S12.60-2002 American National Standard Acoustical Performance Criteria, Design Requirements and Guidelines for Schools.

GUIDELINES FOR MEASURING NOISE LEVELS IN UNOCCUPIED CLASSROOMS

1. Background noise levels should be measured while adjacent spaces are also unoccupied.
2. Background noise levels should be measured during an hour when noise levels are expected to be at their maximum. Doors and windows should be closed and lights should be on.
3. Heating and ventilation systems should be in normal operation in the classroom.
4. A sound level meter (SLM) with frequency weightings of A and C and a SLOW time weighting should be used for measuring background noise. To be acceptable the lowest level of noise measurable by the SLM should be at least 5 dB below the noise level measured in the room.
5. Noise measurements should be undertaken in areas of the classroom used for listening. These are usually student seating areas and the area used by the teacher.
6. Measurements can be taken at a maximum of six locations within the room. Three measurements should be taken at ear height for a seated student and three at ear height for a standing student.
7. The sound level meter should be mounted on a tripod for all measurements.
8. At each location select the slow time weighting and the A filter and record the sound level every 30 seconds for five consecutive 30-second intervals. Note the highest and lowest reading and the average for the five readings. This can be repeated with the C weighting.
9. If the average background noise level from the A weighted measurements at any location are at least 3 dB higher than the ANSI standard of 35 dBA, then it can be concluded that the classroom does not meet the standard.

REVERBERATION

Direct measurement of reverberation requires more sophisticated equipment and it is usually a task undertaken by acoustic engineers in the process of building design or renovation. It is, however, possible to calculate a fair estimate of reverberation time in a classroom using a formula and a table of absorption coefficients. A step-by-step guide to calculating reverberation time is outlined in Box 18.2.

The calculated reverberation time is an estimate. It does not take into account the furniture and other objects in the room. Consequently, a margin of error should be included when comparing the result with the ANSI standard[14] of 0.6-second reverberation time.

Box 18.2 Calculating reverberation time (RT) in a room.

GUIDELINES FOR CALCULATING REVERBERATION TIME IN A ROOM

Introduction

Reverberation time (RT) in a classroom can be estimated by using a simple formula. It involves the volume of a room (V), the total absorption (A) of the room surfaces and a constant (0.16).

Formula

$$RT = \frac{0.16 \times V}{A}$$

RT is measured in seconds
V is measured in cubic metres
A is measured in Sabins

Calculating the RT

I **Calculating V**

1. Measure the length of the room
2. Measure the height of the room
3. Measure the width of the room

Volume = length × width × height in metres

V = _____

II **Calculating A**

The absorption coefficient of each room surface (ceiling, floor etc.) is the product of each area of that surface and the absorption coefficient of the surface lining it.

Area of a surface, e.g. Floor Area = Width × Length in metres

Absorption coefficients are provided for the frequency 500 Hz. in the Coefficients Table below.

For example, absorption coefficient of a floor made of concrete covered with carpet on foam rubber padding (underfelt).

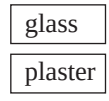
Absorption coefficient = 0.57

Floor Area = 3 m × 3 m = 9 m

$$9 \times 0.57 = 5.13$$

Box 18.2 *Continued*

Note: Where surfaces are made of two distinct materials, e.g. a wall is half glass and half plaster, treat each half as a separate surface and calculate the absorption accordingly.
e.g. Wall = 3 m wide



Area for glass half is 3 m × 1.5 m = 4.5 × absorption coefficient
Area for plaster half is 3 m × 1.5 m = 4.5 × absorption coefficient

Add the separate absorptions for each surface, e.g. floor, ceiling, walls, and obtain the total absorption (A) in Sabins.

A = _____

Finally enter A and V into your formula

$$RT = \frac{0.16 \times V}{A}$$

and obtain the reverberation time in seconds for your classroom.

Sound absorption coefficients at 500 Hz for common materials

Material	**500 Hz**
Walls	
Brick	0.03
Concrete, painted	0.06
Window glass	0.18
Marble	0.01
Plaster on concrete	0.07
Plywood	0.17
Concrete block, coarse	0.31
Heavyweight drapery	0.55
Fibreglass wall treatment, 1 inch (2.5 cm)	0.99
Fibreglass wall treatment, 7 inch (17.8 cm)	0.99
Wood panelling on glass fibre blanket	0.80
Floors	
Wood on concrete	0.07
Linoleum	0.03
Carpet on concrete	0.14
Carpet on foam rubber padding (underfelt)	0.57
Ceilings	
Plaster, gypsum or lime on lath	0.06
Acoustic tiles 5/8 inch (1.6 cm), suspended 16 inches (40.6 cm)	0.46
Acoustic tiles 1/2 inch (1.2 cm), suspended 16 inches (40.6 cm) from ceiling	
The same as above, but cemented directly to ceiling	0.61
High absorptive panels, 1 inch (2.5 cm), suspended 16 inches (40.6 cm) from ceiling	0.75

Coefficients table adapted from
Berg F (1987). *Facilitating Classroom Listening*. Boston: College Hill (p.104).

IMPROVING CLASSROOM ACOUSTICS IN EXISTING CLASSROOMS

The ANSI standard[14] provides considerable detail for architects and builders engaged in building schools with classrooms that will meet the guidelines. In contrast, there are many existing classrooms that may need treatment in order to achieve this goal. Where noise levels are found to be too high in a classroom, several modifications can improve the listening environment. Knecht et al.[16] highlighted the big impact that acoustic treatment of ceilings and reduction of ceiling height and can have on reducing reverberation time. Carpet on floors can also absorb noise from the movement of chairs and may reduce background noise levels. Additional suggestions for reducing noise and reverberation in classrooms are as follows:

- Carefully maintain heating and ventilation systems
- Update noisy heating and/or ventilation systems
- Place rubber tips on chair legs or desks, if carpet is not available or practical
- Install drapes for windows and walls
- Use cork board or carpet for bulletin boards (care should be taken with display of work as shiny paper or cardboard may counteract some of the benefits of the cork or carpet surface)
- Use bookshelves as room dividers to create a quiet classroom area
- Use upholstered chairs to absorb sound reflections
- Position mobile cork or carpet bulletin boards at an angle (not parallel) to walls
- Use landscaping to reduce outside noise
- Close doors to hallways and classroom dividers to reduce noise
- Insulate walls between classrooms.

Improving classroom acoustics and reducing background noise will improve the learning environment for all children, but it may not be sufficient to ensure clear input for children with hearing loss or other learning difficulties. They may also need to use an assistive listening device to enhance the signal received from the teacher.

ASSISTIVE LISTENING DEVICES

Assistive listening device is a term that can include a vast array of auditory and non-auditory devices. In this chapter, the term will be used to refer to devices that work specifically to enhance the speech signal for face-to-face communication. Systems that have been used with children include hardwire systems, loop systems and FM systems. Hardwire systems can be used in special classrooms for hearing-impaired students. Each child wears a set of headphones with boom microphone, attached to a desk or console. The teacher wears a microphone linked into the amplification system. Limited adjustments can be made to individualise the headset for each child. Students receive a relatively noise-free signal from the teacher and from each other. Hardwire systems are simple and potentially inexpensive systems that can work very well with small groups of hearing impaired students educated together. However, the dominance of inclusive educational placement (in 2003, 84% of children with hearing impairment were learning in regular classrooms in Australia[13]) and more activity-based learning has made

hardwire systems an impractical form of classroom amplification. Moreover, hearing aids must be removed during the time the system is used, and the issue of learning to listen with two different forms of amplification has concerned educators and audiologists.[2]

Loop systems were used briefly in classrooms as educational amplification during the 1970s. Children switched to the telecoil setting on their hearing aids and the teacher spoke into a microphone that sent the signal via a room-loop using electromagnetic conduction. These systems fell out of favour due to the presence of dead spots in classrooms, signal overspill from room to room and alterations to the hearing aid frequency response that occurred on the telecoil setting. Loop systems continue to be useful at home when watching TV, for focused dedicated listening in cinemas or school auditoriums and for short duration signal reception in noisy settings such as railway stations, banks and other public spaces. In educational environments, the FM system has become the assistive listening device of choice for students both with and without hearing impairment.

FM SYSTEMS

FM systems, also known as radio aids, radio frequency aids and FM aids, are two-part systems that use a microphone/transmitter worn by a teacher or parent and a receiver worn by the child. Sound is picked up via the microphone, worn close to the lips, and converted into frequency modulated (FM) radio waves that are transmitted on a specific frequency to a receiver. The signal is then transformed back to a speech signal and delivered to the student's hearing aids (personal FM system) or directly via button receivers (all-in-one FM aid). Sound-field FM systems use a similar system but instead of personal receivers the sound is transmitted to a speaker or set of speakers set up in the classroom, thus amplifying sound throughout the whole classroom. Table 18.1 contains a brief description of FM systems currently used with children.

Table 18.1 Brief descriptions of FM systems in use with hearing-impaired children.

Types of FM systems	Description
All-in-one FM systems (Body worn) (Auditory trainer)	A combined body-worn hearing aid and FM system usually worn with button receivers. May also be worn with lightweight headphones for students with central auditory processing disorders.
Personal FM systems: Body worn	A personal FM system connected to a child's personal BTE hearing aids via direct audio input or via a neckloop. This type of system can also be used with body-worn cochlear implant.
Personal FM systems: Ear level	Either a combined BTE hearing aid and FM receiver or an audioshoe that attaches to a BTE hearing aid or ear-level cochlear implant and utilises the hearing aid battery as a power source.
Soundfield FM systems	Utilise a conventional transmitter/microphone but sound is transmitted to a loudspeaker or set of loudspeakers placed around the room. It has many applications, including classes containing children with fluctuating conductive losses, central auditory processing disorder, cochlear implants and normal hearing.
Personal sound-field FM systems	Transmitter/microphone transmits teacher's speech to a portable desktop speaker.

Personal FM systems

Personal FM aids can be worn with hearing aids and cochlear implants or used with a set of light headphones by children with mild loss or auditory processing disorders. Hearing aid and cochlear implant users can receive the FM signal via a body-worn receiver. Many children now use an ear-level FM receiver, which is attached to their aid or implant via a shoe or it may be built into the child's sensory device. Personal FM systems have consistently been shown to improve speech perception and listening behaviour in noise and reverberation for children with hearing loss.[22–25] Boothroyd and Iglehart [22] compared hearing aids alone with FM + hearing aids for 13 severely and profoundly deaf adolescents in quiet and noise. They demonstrated a clear FM advantage that equated to an average improvement of 25% in phoneme recognition. The children who showed the greatest benefit were children with good speech perception skills (between 40 and 60%) with hearing aids alone. Toe[25] also assessed FM aid benefit for 50 adolescents with severe to profound hearing loss listening to sentences in background noise and showed an FM + hearing aid benefit of 18% for auditory speech perception compared with hearing aids alone. Toe[26] showed that the use of an FM system in regular secondary school classrooms resulted in increased visual attention to the teacher by students with profound loss.

Boothroyd and Iglehart[22] found that body-worn FM aids performed slightly better than ear-level FM aids. This may relate to the output limit of the child's hearing aid, as the children in their study all had severe to profound hearing losses. They suggest that the body-worn FM aid advantage may not be evident for children with mild to moderate losses, who have a bigger dynamic range; however, this suggestion has not been verified by empirical research. When compared with sound-field systems, personal FM systems with direct audio input provide the best signal-to-noise ratio for classroom listening by children with mild and moderate losses.[27] Desktop sound-field systems also performed well, suggesting that the closer the output of the FM system to the child's ear, the better the outcome.

Assessing FM aid benefit and usage

FM benefit has been shown to vary between individuals and appears to be affected by a range of factors, including listening experience, degree of hearing loss and auditory awareness.[23–25,28] Following electroacoustic calibration, FM benefit should be assessed in a number of ways. At a minimum, speech perception skills should be assessed with and without the FM aid using an age appropriate standardised speech perception test. FM benefit should also be assessed in the classroom through systematic classroom observation by either an audiologist or a teacher. Self-report is also a useful and often overlooked tool, empowering students at a young age to provide feedback about their own listening needs.

Fitting personal FM systems

Selecting an FM system for a child is a complex process that requires careful pre- and post-evaluation of the child's listening situation at home or in the classroom. The goal of FM aid fitting is to allow a child to hear a primary speaker (often a teacher or parent) at an enhanced signal-to-noise ratio so that their speech is consistently above the background noise. It is also highly desirable that the child is also able to hear and monitor their own voice and hear the voices of other people who are not wearing the FM aid transmitter. This is critical for young

children who are still developing their communication skills. The ease with which both goals can be achieved depends upon several factors: the child's listening environment, the FM system fitted, the coupling option selected and the child's degree of hearing loss.

There are four possible listening modes that may be selected with different FM systems: (a) FM microphone only, (b) FM microphone and hearing aid/implant microphone combined, (c) FM plus mode or voice operated switching and (d) hearing aid/implant microphone only. In the FM only mode, the hearing aid/implant microphones are inactive and the child hears only the speaker with the FM transmitter. This mode offers the best possible S/N ratio and suits a lecture situation quite well. In a combined or FM + hearing aid listening mode both FM and hearing aid microphones are simultaneously active. The child hears the transmitter wearer at a consistently loud level and also hears his or her own voice and the voices of others within hearing distance. If the listening conditions are poor, the noise entering the hearing aid microphones may significantly reduce any advantage offered by the FM system.[29,30] This mode is most suitable for discussion and less structured educational settings such as the pre-school.

An alternative to the combined setting that still offers children some access to their own and other speakers' voices is the 'FM plus', FM precedence or voice-operated switching (VOX) listening mode. With this option, the hearing aid or environmental microphones are attenuated by 15–30 dB only whilst the transmitter wearer is speaking. When the transmitter wearer stops speaking the hearing aid microphones are reactivated and the child can hear his or her own voice and the voices of children around them. This option represents a compromise between the FM only and combined listening modalities. It offers a good S/N ratio without eliminating a child's access to his or her own and other children's speech. It may not work so well in an unstructured discussion where very quick interchanges occur. There is a necessary hang time that occurs between when the transmitter-wearer stops speaking and when the hearing aid microphone is attenuated. As a consequence, the hearing-impaired child may miss the beginnings of comments made in a rapid discussion. Some FM systems also offer the option of listening via the environmental microphones alone. With most personal systems, this may simply be achieved by turning off or removing the FM system. The many FM systems on the market offer different combinations of these listening modalities. It is critical, when selecting an FM system for an individual child, that the child's needs are carefully matched to the listening modes available in the FM aid selected.

A simple guide to FM aid fitting and balancing

Once an FM system is selected, it is necessary to set up or balance the system appropriately for the child who will wear it. As with hearing aid fitting, it is critical that the FM signal is not received at a level that is either too high for comfort and clarity or too low to offer any S/N ratio benefit for the child. Boothroyd and Iglehart[22] have demonstrated that substantial FM aid benefit is lost in a noisy classroom setting if the FM aid output is matched to the hearing aid output. They recommend that the gain of the FM system should be matched to the gain of the child's hearing aids. A compromise is often recommended whereby the output of the FM aid is set to be 10 dB higher than the output of the child's own hearing aids.[31] Particular care must be taken when fitting FM aids with digital hearing aids. Bamford, Hostler and Pont[32] showed that the electromagnetic interference processor in digital hearing aids can interfere with the FM signal. They recommend that only digital aids with a low electromagnetic interference processor should be used with FM aids. Box 18.3 contains a brief guide to fitting FM systems.

Box 18.3 A guide to setting up FM systems with personal hearing aids.

A GUIDE TO SETTING UP PERSONAL FM SYSTEMS

Make all measurements in dB SPL.
A broad-band speech-shaped signal is highly recommended for these measurements.

1. Using a 70 dB SPL input measure the output of the child's hearing aid in dB SPL.
2. Place the FM microphone in position in the test box. The hearing aid is coupled to the FM receiver and then attached to the 2cc coupler. Isolate these from the test chamber at a distance of at least 50 cm.
3. Measure the output of the FM system using an input level of 80–85 dB (chestworn microphone) or 90 dB (headworn microphone).
4. Compare the output of the aid via the FM system with the output of the aid alone. The output via the two systems should be equal, at least at 1,000 Hz. The two outputs may not be perfectly matched across the entire frequency response. If the FM system is to be worn in combined mode, then it is preferable to set the output received via the FM aid to be 5–10 dB higher than the output of the hearing aid. This will ensure an FM aid advantage in noise. This may not be possible with more severe and profound hearing losses.
5. Evaluate the SSPL90 of the FM system using a swept pure tone signal. Compare results with prescription targets to ensure comfort and safety when the FM aid is worn.
6. Performance of the FM system can be verified using probe tube real ear measurement or by applying a child's individual RECD to FM aid measurements made in the hearing aid test box.

Source: Lewis D. (1997), Selection and assessment of classroom amplification. In: W. McCracken and S. Laoide-Kemp (eds.), *Audiology in Education*, London: Whurr Publishers Ltd and *FM Systems for Children: Rationale, Selection & Verification Strategies*, Phonak Focus Video: Sound Foundations (1998). Running Time: 19 minutes

FM AID MANAGEMENT IN THE CLASSROOM, AT HOME AND AT PRE-SCHOOL

Use of a two-part amplification system adds a degree of complexity to the communication situation that requires careful management by the child and his or her teacher or parent. The FM transmitter is worn near the teacher's or parent's lips. They cannot simply walk away from the child if they do not want to be heard. The child and the teacher must work together to ensure that the child receives relevant spoken input. In the classroom, a teacher and student may stay switched on whilst the teacher is addressing the class or talking to the student. However, once the class is working in groups or on individual work it is important that the student switch the receiver off so that they can focus upon their classmates or concentrate on their work. This is particularly important if the teacher is roaming the class and conversing with individual students. Similarly at home, a parent needs to view the FM aid as an aid to face-to-face conversation. It should not be used as a means of unnaturally monitoring distant behaviour or left on while the parent is having conversations with other adults that may interfere with the child's communication with others. This might provide confusing

spoken input that may impair the development of an already fragile construct of spoken communication.

FM aids can play an important role in ensuring that very young children with hearing loss have increased access to incidental and overheard language. Very early identification of hearing loss and early hearing aid fitting or cochlear implantation should mean that parents may be ready to cope with management of an FM aid whilst their child is a toddler. Ear-level FM aids reduce the need for excessive cords and bulky body-worn systems. Careful use of the FM system can ensure good language input during a wide range of activities, when the child is in the car, in a stroller or a backpack or is exploring outside.

Some of the biggest management risks are associated with FM aid use in pre-schools. Burnip and McGuire[33] demonstrated that if a pre-school teacher were to wear the FM transmitter for a whole pre-school session, as much as 70% of the spoken language heard by a hearing impaired child may be irrelevant. So that, whilst young Mary is playing in the block corner with some peers, trying to negotiate the intricacies of early socialisation and communication, she may be being forced to listen to her teacher telling John what a beautiful painting he has done. With such confusing input, cracking the code of spoken language may become an impossible task. Recognition of the complexities of using FM systems in pre-schools has often necessitated limited use of these aids, whereby they are only worn during group activities. For this purpose, new developments in sound-field FM technology may have possibilities for pre-schools that have not yet been fully explored.

Sound-field FM systems

Sound-field FM systems for classrooms operate in a similar way to a public address system. The teacher or speaker wears the FM transmitter/microphone. His or her speech is transmitted by FM radio waves to a receiver and the sound is delivered to the room via several small loudspeakers. Some systems use only one loudspeaker. All children receive the benefit of the amplified voice of the teacher and the teacher is not required to strain his or her voice to be heard. These systems have been found to be beneficial in classrooms with significant numbers of students with special listening needs. These include Aboriginal children in Australia[34] and Inuit children in Canada,[35] who have high levels of otitis media.

Sound-field amplification systems have the potential to provide benefit for all students, including second-language learners, children with auditory processing disorders, young developing listeners, those with mild or unilateral loss and normally hearing children.[36] As with personal FM systems, care must be taken in the management of sound-field systems. Teachers need to be sensitive to the dominance of their voice level during individual independent work and small group sessions. Sound-field systems may be problematic in highly reverberant classrooms. There is a complex relationship between background noise, speaker position and reverberation. If the noise source is farther from the listener than the speech source (loudspeaker of sound-field system), then noise levels may increase more with reverberation than does the speech level, leading to decreased speech intelligibility in classrooms with poor acoustics.[37] Consequently, sound-field FM systems are not a solution for classrooms with high reverberation times. It is necessary to treat the room first, prior to the installation of a sound-field system.

The distance between the sound source and the ear of the child has been shown to impact on speech intelligibility. Anderson and Goldstein[27] demonstrated that students with mild to severe hearing loss had superior speech perception with either a personal FM system or a desktop sound-field system as compared with an infrared sound-field system. Proximity to

loudspeaker is implicated. Personal FM systems are recommended for children with significant hearing loss as they require the best signal-to-noise ratio. This can be delivered with direct audio input. Anderson and Goldstein observed that students were more confident and responsive when using personal FM systems.

NEW DEVELOPMENTS IN ASSISTIVE LISTENING DEVICES

Many of the recent developments in assistive listening technology might be seen as further accessorising a good idea. Bluetooth technology offers individuals with hearing impairment a cordless method to connect to their FM system that is compatible with a wide range of other inputs. Phonak have developed the Wall Pilot that frequency synchronises Phonak multi-frequency FM receivers. When students enter the room, the wall pilot sets their receivers automatically to the nominated room frequency. With this device there is no need for students to manually change frequency when they change classroom. There is an overwhelming range of microphone/transmitters, ear-level and body-worn devices and ear-level receivers designed for students who do not wear hearing aids but require an improved S/N ratio. It is not practical to list all of the functions and features of available assistive listening devices here. Websites of individual manufacturers provide detailed information to assist audiologists and other professionals to select the best device for an individual child.

CONCLUSION

This decade has seen substantial development in our understanding of the devastating impact that poor classroom listening conditions can have on learning for all children and particularly for children with hearing impairment. Classroom acoustic modification is critical for ensuring that students can hear their teachers. Current noise cancelling systems in digital hearing technology are no match for the benefits of placing the microphone close to the speaker. Adults with hearing impairment have recently begun to realise the value of FM technology for listening in the real world. This can only benefit children with hearing loss as more research and development funds are likely to flow from this larger potential market. Professionals must stay committed to promoting FM hearing aids, not as an extra or an accessory, but as a basic piece of equipment for every child with special hearing needs.

RESOURCES

American National Standards Institute (2002) Acoustical performance criteria, design requirements and guidelines for schools. ANSI S12.60. These standards can be downloaded at no cost from the following website, http://asastore.aip.org

Crandell C, Smaldino J, Flexer C (1995) Sound-field FM: Amplification Theory and Practical Applications. San Diego, CA: Singular Publishing Company. This book provides detailed information about sound-field amplification.

FM Systems for Children: Rationale, Selection & Verification Strategies. Phonak Focus Video: Sound Foundations (1998). Running Time: 19 minutes. This is an excellent video that provides a rationale for the use of FM aids and a step-by-step fitting guide.

Phonak Website: The hearing aid company Phonak has a very informative website that provides conference proceedings, detailed FM aid manuals and a range of other valuable information. It is available at http://www.phonak.com/

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19 Balance disorders in children

K. Harrop-Griffiths

INTRODUCTION

Balance, as a sense, requires the integration, within the central nervous system, of sensory input from the vestibular hair cells of the inner ear, vision and proprioception. This information, when compared with previous learnt experiences, allows one to sense position and movement in space. Pathology affecting function in any of these three senses (vestibular, vision and proprioception), their central pathways and associated neural connections can lead to balance disturbance. In addition, balance, as a motor function, requires an intact peripheral nervous system and musculoskeletal system, whilst an upright posture is also dependent on the function of the cardiovascular system and the autonomic nervous system as well as the integrity of other homeostatic processes. The range and complexity of difficulties that can give rise to disorders of balance are considerable, and the clinician needs a careful and logical approach in order to reach an accurate diagnosis and thus to determine optimal management.

Balance disorders in children are relatively common but largely unrecognised. The term is broad, encompassing problems such as clumsiness, delayed motor milestones and motion sickness as well as acute attacks of vertigo, dizziness or syncope. Symptoms are often difficult to describe, particularly so for younger children who lack the vocabulary; thus presentation is often through a third party, the parent, who is largely reliant on observation and his or her own experiences to interpret the child's difficulty. The child's first-hand description of the problem is invaluable and should always be carefully sought where possible. Most diagnoses are defined by a detailed history, with examination, testing and investigation providing confirmatory evidence. Revisiting the problem as the child matures can unearth a clearer picture as the child's language and ability to describe the problem, and tolerate testing, mature.

A DEBILITATING PROBLEM

Disorders of balance can lead to significant morbidity in children. Delayed locomotor development in infants can adversely affect learning by reducing access to play materials and exploration. The ability to walk with ease opens up an exciting world of opportunity and discovery denied to the child who is still learning to sit. For school-age children delay in locomotor skills and poor postural coordination affect social integration and feelings of self-esteem – which captain will choose the clumsy child for the team? The hustle and bustle of a busy school playground can be a frightening challenge for the unstable child, affecting both confidence and independence. Episodic vertigo leads to time off school, with inevitable consequences for

academic attainment and social integration, and in addition can lead to anxiety and panic. The combination of visual impairment and balance disorder such as in Usher I syndrome, Alström syndrome, and DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness) leads to considerable restriction of activity, which is often underestimated. Even common visual difficulties, such as strabismus, can affect a child's ability to compensate from an acute vestibular pathology such as vestibular neuronitis.

EPIDEMIOLOGY

Balance disorders in children are often not reported, making it difficult to assess the true prevalence in the community. Mild or transient dizziness may be ascribed to a viral infection or 'tummy bug' rather than to a vestibular disorder. Many peripheral balance disorders resolve quickly in children – the desire to return to normal play is often paramount and probably enables rapid compensation. Chronic instability is often seen as clumsiness or general developmental delay, and delayed locomotor skills in the Deaf population are regarded as a 'fact of life'. Presentation of more persistent or debilitating problems may be to the general practitioner (GP), paediatrician, paediatric neurologist, otologist or audiovestibular physician, making it difficult for any one group to understand the full extent of the problem.

Several attempts have been made to explore the epidemiology of vertigo and balance disorders. Jayarajan and Rajenderkumar[1] in a postal survey of GPs in four areas of the UK determined that 0.8% of the population consulted their GP with symptoms of dizziness in a 12-month period; of these only 1.3% were between 5 and 20 years of age. A postal questionnaire administered to 2,165 school-age children in Glasgow[2] found that 314 (14.5%) reported at least one episode of dizziness in the preceding year, with 92 (4.2%) reporting three or more attacks. Niemensivu et al.[3] in a questionnaire returned on 938 children of 1 to 15 years of age in Finland reported that 8% had experienced vertigo with an increasing prevalence with age. Twenty-seven per cent of these children (approximately 2% of the whole) stated that the vertigo was bad enough to stop their current activity, and 8% (0.6% of total) had had an attack at least once a week. In addition, they stated that 2% reported balance difficulties, 1% recurrent falls, 2% difficulties in walking and 3% clumsiness.

Several surveys amongst the Deaf population have estimated the prevalence of vestibular hypofunction as occurring in approximately 30 to 40% of deaf children,[4] with increased prevalence in those with more profound losses. Möller,[4] in a study of 74 children with severe and profound losses, reported that more than 50% had a walking age of more than 18 months and that up to 75% reported balance difficulties, particularly with sports. All those with absent caloric and rotatory responses reported a walking age of greater than 18 months, indicating that a record of locomotor milestones can be helpful in identifying those likely to have significant vestibular hypofunction.

EMBRYOLOGY

The inner ear develops from the otic placodes which develop into otocysts. The ventral part of this structure will form the saccule and cochlear duct; the dorsal part will form the utricle,

semicircular canals and endolymphatic duct. The component parts are formed by about 8 weeks' gestation, with the lateral semicircular canals being the last part of the vestibular organ to develop. By 11 weeks, the neuroepithelium has developed and the whole labyrinth is fully formed by 20 weeks gestation. Vestibular tracts are myelinated during the second trimester but central connections are immature at birth. Structural abnormalities of the inner ear are explained by arrested development.

PRESENTATION

Children with balance disorders generally either present with imbalance plus delayed or deteriorating locomotor skills or with episodic dizziness/vertigo. Whilst both complaints can occur together, it is often useful to consider these presentations separately. Likewise, the presence or absence of a hearing loss can help differentiate possible diagnoses; however, this dividing line is arguably less discriminating and the author has managed several deaf children with coincident migrainous vertigo (see Table 19.1 and Boxes 19.1 and 19.2).

Table 19.1 The working diagnoses of 209 cases of vertigo presenting to a specialist clinic to illustrate the relative frequency of the different causes of vertigo in children (personal figures).

Diagnosis	Number	%
Migrainous vertigo	55	26.2
BPVC	6	2.9
Peripheral Vestibular Disorder – unknown cause – 24 with PCHI	28	13.4
Psychogenic (definite abuse = 3)	20	9.6
Widened vestibular aqueducts	11	5.3
Otitis media with effusion/chronic suppurative otitis media	7	3.3
Labyrinthitis	7	3.3
Neuronitis	7	3.3
Postural hypotension	6	2.9
Vestibular processing/sensory integration difficulties – motion sickness	5	2.4
Progressive SNHL with vertigo – unknown cause	5	2.4
Trauma (head injury = 2, radiotherapy = 1, previous neurosurgery = 1)	4	1.9
Visual (oculo-motor apraxia = 2, visual vertigo = 2)	4	1.9
Endolymphatic hydrops – secondary	4	1.9
Episodic ataxia	3	1.4
Heredodegenerative disease (DIDMOAD = 2, Refsum = 1)	3	1.4
Central vestibular pathology – unspecified	3	1.4
Auto-immune (Cogan's S. = 1, unspecified auto-immune = 1)	2	1.0
Epilepsy	2	1.0
Ménière's disease	2	1.0
Caffeine ingestion	1	0.5
Cardiac arrhythmias	1	0.5
Hypnogogic	1	0.5
Tullio phenomenon with PCHI	1	0.5
Incomplete testing	12	5.7
Unknown	10	4.8

PCHI = permanent childhood hearing impairment

Box 19.1 Acute or episodic vertigo – differential diagnoses.

- Middle-ear disorders
  - Otitis media with effusion (+HL)
  - Cholesteatoma (+HL)
- Inner-ear disorders
  - Viral labyrinthitis (+HL)
  - Bacterial labyrinthitis – otogenic/meningitic (+HL)
  - Vestibular neuronitis (–HL)
  - Widened vestibular aqueduct (+HL)
  - Trauma (+/– HL)
  - Auto-immune disease (+HL)
  - Perilymphatic fistula (+HL)
  - Ménière's' disease (+HL)
  - Benign paroxysmal positional vertigo (–HL)
- Central disorders
  - Migraine and migraine equivalents
    - Benign paroxysmal vertigo of childhood
    - Migrainous vertigo
  - Epilepsy
  - Posterior fossa tumours
  - Episodic ataxia
- Other
  - Visual vertigo/strabismus
  - Psychogenic (+HL in a deaf child)
  - Postural hypotension
  - Cardiac arrhythmias
  - Toxic – alcohol, recreational drugs

Box 19.2 Causes of imbalance or instability – differential diagnoses.

- Middle ear
  - Otitis media with effusion
- Vestibular hypofunction
  - Genetic – associated with hearing loss
  - Meningitis with labyrinthitis
  - Congenital CMV
  - Ototoxicity
  - Auto-immune disease
- Developmental
  - Dyspraxia
  - Vestibular processing
- Psychogenic
- Neurological disorders
  - Cerebral palsy
  - Intracranial tumours
  - Post-traumatic
  - Heredo-degenerative disorders
- Musculo-skeletal disorders
  - Muscular dystrophy
  - Arthritis

CAUSES OF BALANCE DISORDERS

There are many causes of dizziness and imbalance in the paediatric population and the following will dwell on the more common or important of these. The differentiation of peripheral from central vestibular disorders is an important aspect of neuro-otology. Peripheral disorders characteristically tend to give rise to acute or episodic vertigo and conversely, central vestibular disorders tend to present with chronic disorientation and instability rather than acute episodic vertigo. However, migrainous vertigo, benign paroxysmal vertigo of childhood, epilepsy and episodic ataxia, all central disorders, can all present with acute episodic vertigo and thus mimic peripheral disorders, whilst bilateral peripheral vestibular hypofunction can present with instability and delayed locomotor milestones in the young or oscillopsia and disorientation if acquired later in life.

PERIPHERAL VESTIBULAR DISORDERS

Disorders of the middle ear

Otitis media with effusion (OME)

Imbalance, dizziness and vertigo are common complaints with middle-ear effusions. Jones et al.[5] confirmed parental reports of poor balance by finding significantly increased body sway in children with OME compared to controls, and reported improvement following grommet insertion. Vertigo is not generally part of the symptomatology with young children but is a recognised symptom in older children and adults with middle-ear dysfunction. The mechanism has not yet been satisfactorily explained; it is possibly due to a mechanical effect on the round window. Concern for the effect of vestibular dysfunction on development has prompted several authors to advocate balance disturbance as an indication for early surgical intervention.

Acute otitis media

This can lead to otogenic bacterial labyrinthitis with vertigo and hearing loss. This is rare in developed countries but can occur in those with reduced immunity, including the very young.

Cholesteatoma

Erosion of the lateral semicircular canal giving rise to vertigo and imbalance is well recognised in cases of cholesteatoma, both congenital and acquired. Darrouzet et al.[6] reported a series of 215 cases of cholesteatomas in children of which 24 (16%) were considered to be congenital in origin. The incidence of lateral semicircular canal erosion was found to be 8.8% in the congenital compared with 3.3% in the acquired cases in children. Other papers quote an incidence of a lateral semicircular canal fistula as being approximately 7% of acquired cases overall without distinguishing different age groups. Gersdorff et al.[7] found that 57% of 54 patients aged 6 years to 83 years with acquired cholesteatomas and erosion of the lateral semicircular canal presented with vertigo.

Disorders of the inner ear

Structural abnormalities

Structural abnormalities of the inner ear are commonly found in association with permanent childhood hearing loss. The aetiology can be genetic (e.g. brancho-oto-renal syndrome), unknown (e.g. Goldenhar) or acquired (as in thalidomide ingestion or maternal iodine deficiency). CT scans are valuable in identifying bony abnormalities whilst MRI scans give additional information about the cochlear and vestibular nerves and endolymphatic duct and sac.

Bamiou et al.,[8] in a study of the findings of 116 CT scans, performed as part of the aetiological investigation for sensorineural hearing loss in children attending a specialist unit, found that 28.4% had identifiable abnormalities. These were more commonly found in those with progressive or profound hearing losses and/or craniofacial abnormalities. Abnormalities of the vestibular labyrinth were more common than those affecting the cochlea. Similar findings have been reported by other authors. Structural abnormalities affecting the vestibular labyrinth are commonly associated with vestibular hypofunction or areflexia.

Widened vestibular aqueducts (WVA) or large vestibular aqueducts

This particular anomaly of the inner ear often presents with episodic vertigo associated with fluctuating and progressive hearing loss precipitated by head trauma which is often quite trivial. The vertigo is of variable length from a few seconds to hours and a drop in hearing is often coincident. It is considered to be the most commonly occurring anatomical anomaly of the inner ear. The clinical picture may be caused by pressure changes transmitted down the widened and straighter endolymphatic duct following minor trauma.

WVA are characteristically found in Pendred syndrome and have been reported in brancho-oto-renal syndrome and CHARGE association, and can also occur as part of a non-syndromic hearing loss. They are associated with cochlear abnormalities, in particular incomplete partition, as in the Mondini deformity. Vestibular findings vary. CT scans or MRI scans are diagnostic of WVA and an MRI scan can also identify the size of the endolymphatic sacs, which are often enlarged in Pendred syndrome (see Figures 19.1 and 19.2).

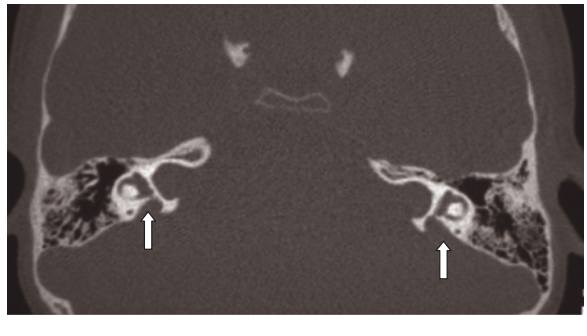

Figure 19.1 CT scan showing widened vestibular aqueducts.

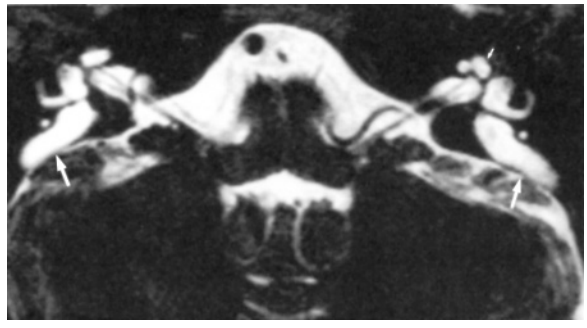

Figure 19.2 MMI scan showing widened vestibular aqueducts and enlarged endolymphatic ducts.

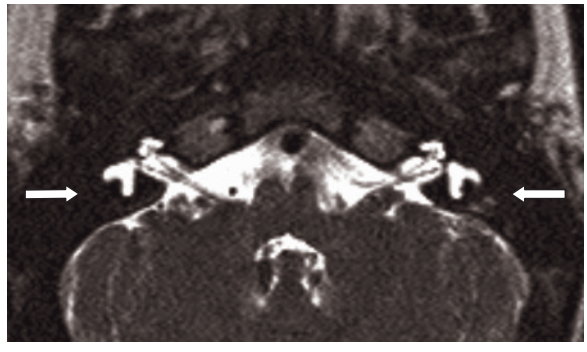

Figure 19.3 MRI scan of bilaterally dysplastic lateral semicircular canals.

Dysplastic semicircular canals

Another commonly found abnormality, which is invariably associated with poor vestibular function. The lateral semicircular canal is most commonly affected, frequently in isolation; although an absent or rudimentary canal is usually associated with cochlear deformity. Malformation of the posterior canal is associated with widened vestibular aqueducts. Johnson and Lalwani[9] in a retrospective analysis of 28 ears (15 patients) with semicircular canal anomalies found that 71% had sensorineural hearing loss, 14% with conductive or mixed hearing loss, 1 (<4%) with microtia and 11% with normal hearing. In 27 out of 28 cases, only the lateral semicircular canal was involved. Clinical presentation is of delayed locomotor milestones and imbalance (see Figs. 19.3).

Agenesis of semicircular canals

This is a finding characteristic of CHARGE association in which abnormalities of the cochlea can coexist. Clearly in cases where development of the inner ear structures remains at a very primitive level the canals do not form, e.g. Michel deformity or otocyst.

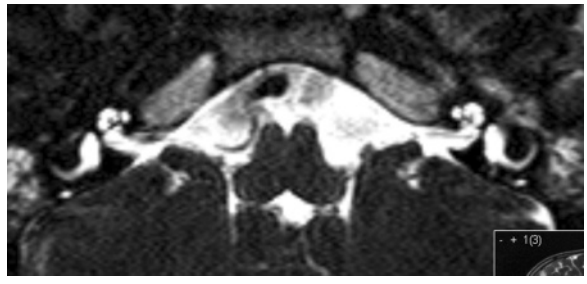

Figure 19.4 MRI scan demonstrating normal inner ear anatomy. Note the 'signet rings' of the lateral semicircular canals.

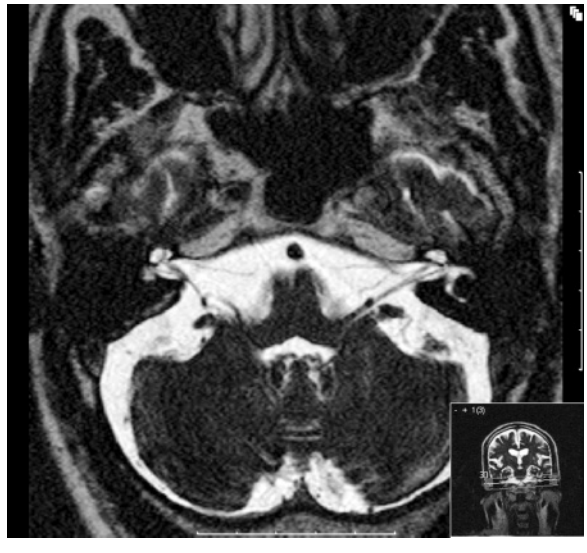

Figure 19.5 Obliteration of the right lateral semicircular canal and part of the left as a result of fibrosis secondary to meningitis.

Hypoplasia/aplasia of the vestibulo-cochlear nerve

Thin or absent eighth cranial nerves can be identified on an MRI scan in some profoundly deaf children. Vestibular hypofunction occurs if the vestibular nerves are involved. The aetiology is unknown.

Superior semicircular canal dehiscence

This rare structural abnormality has been identified as a cause of Tullio phenomenon. A CT scan is diagnostic and surgery has a place in severe cases.

GENETIC HEARING LOSS

Vestibular pathology is well recognised as occurring in both syndromic and non-syndromic hearing loss, whether congenital or late-onset. It is also reported as occurring without hearing loss. Whilst vestibular pathology is clearly associated with the structural abnormalities detailed earlier, it can occur with normal radiological findings. With the exception of WVA, which presents with episodic vertigo, other conditions generally present with delayed motor milestones and imbalance and with findings of vestibular hypofunction or areflexia. The value of vestibular assessment in discriminating between different phenotypes, thus enhancing our understanding of genetic hearing loss, is becoming increasingly recognised. Vestibular assessment should form part of the investigation of every deaf child from the point of view of aetiology as well as habilitation.

Pendred syndrome

WVA is recognised as occurring in 90% of patients with Pendred syndrome,[10] a recessive genetic condition characterised by sensorineural hearing loss and thyroid goitre. The endolymphatic sac is also frequently dilated. The underlying genetic defect is a mutation in the *SLC26A4* gene on chromosome 7, which codes for Pendrin, an anion transporting protein expressed in the thyroid and the endolymphatic duct and sac. Pendrin is probably involved in regulation and resorption of endolymph, and it has been hypothesised that individuals with Pendred syndrome may have marked pre-natal hydrops. Vestibular findings are variable with bilateral hypofunction in about one third, unilateral peripheral vestibular deficit in a third and normal function in a third.[10]

CHARGE association

This is a rare condition characterised by Coloboma, Heart defect, Atresia of the choanae, Retarded growth/development, Genital hypoplasia and Ear anomalies/deafness. Two genotypes have been recognised with mutations affecting chromosome 8 or 7. Ear abnormalities are seen in 91% and deafness in 62% of affected infants[11] with the radiological findings of absent semicircular canals considered pathognomic if present. Balance is severely affected in these children although residual otolith function has been identified and attributed to an abnormal otolith organ in the hypoplastic posterior labyrinth.[12]

Usher syndrome

Sensorineural hearing loss and retinitis pigmentosa characterise this recessively inherited condition caused by several different genotypes. At least three phenotypes have now been recognised. Radiological findings are normal.

Usher type I presents as a congenitally profound sensorineural hearing loss with vestibular areflexia and an early onset of retinitis pigmentosa.

Usher type II is characterised by a congenital moderate to severe sensorineural hearing loss, normal vestibular function and a later onset of retinitis pigmentosa.

Usher type III is generally rare but relatively common in Finland and presents with a progressive sensorineural hearing loss, variable vestibular function and retinitis pigmentosa by the second decade.

Jervell and Lange-Nielsen syndrome

This syndrome presents with severe to profound congenital deafness, vestibular hypofunction and cardiac arrhythmias, which can result in cardiac arrest and sudden death as early as the first year of life. It is a rare recessive genetic disorder caused by mutations on the KCNQ1 or *KCNE1* (potassium channel) genes, giving rise to a prolonged QT interval and subsequent increased risk of arrhythmias. Children may present to a balance clinic with 'funny turns' as a consequence of syncope precipitated by the cardiac arrhythmias. Prolongation of the QTc interval (>440 msec) identified on ECG indicates the need for a cardiology opinion for diagnosis and consideration of an implanted cardioverter/defibrillator.

Waardenburg syndrome

A dominantly expressed disorder, Waardenburg syndrome is characterised by abnormal pigmentation and a variable degree of sensorineural hearing loss. Four different phenotypes have been identified.[13] Abnormal vestibular findings have been described[14,15] as have a variety of anatomical anomalies.[14]

Other genetic disorders

Peripheral vestibular dysfunction has been reported in a number of other genetic syndromes, including Marshall syndrome, Hurler's syndrome, Alport syndrome and Alström syndrome[13,16] as well as some non-syndromic hearing losses. Bilateral vestibular schwannomas are characteristic of Neurofibromatosis 2 and endolymphatic sac tumours can occur with von Hippel-Lindau syndrome, but presentation in childhood is very rare. Vertigo and poor vestibular function can occur in children with mitochondrial disorders.[17]

Several of the central causes of balance disorders have a recognised genetic aetiology, e.g. episodic ataxia, migraine, Refsum syndrome, Wolfram syndrome (DIDMOAD) – see sections on episodic ataxia, migraine and heredodegenerative diseases.

INFECTIVE CAUSES

Congenital

Vestibular hypofunction is recognised as occurring in congenital rubella syndrome. In a study of children in a school for the deaf Huygen et al.[18] reported the finding in five out of eight children with known congenital rubella syndrome.

There is evidence, through case reports[19] and a small study of fourteen confirmed cases,[20] that congenital cytomegalovirus (CMV) infection can give rise to vestibular dysfunction occurring in association with or independent of hearing loss. Congenital CMV is probably prevalent but until recently confirmation has been dependent on serological testing within 3 weeks of birth; however, recent advances in diagnosis using stored neonatal blood spots (Guthrie cards) are enabling more definite diagnoses in children presenting later in childhood.

Congenital syphilis can give rise to hydropic symptoms of episodic vertigo, a fluctuating hearing loss and tinnitus in the second decade. These symptoms are similar to acquired tertiary syphilis. The spirochete, *Treponema pallidum*, can infect all parts of the vestibular system and pathways.

Acquired

Bacterial labyrinthis

Bacterial labyrinthitis leads to fibrosis within the membranous labyrinth and subsequently labyrinthitis ossificans with hearing loss and vestibular hypofunction. There are two mechanisms by which infection can occur:

- Otogenic – either as a consequence of acute otitis media or erosion of the bony labyrinth by cholesteatoma, see section on disorders of the middle ear.
- As a consequence of meningitis when bacteria enter the perilymphatic space through either the cochlear aqueduct or the internal auditory canal.

Bacterial meningitis

Loss of vestibular function commonly occurs as a consequence of bacterial meningitis. The most common pathogens giving rise to labyrinthine pathology are *Streptococcus pneumoniae* and *Neisseria meningitidis. Haemophilus influenzae* meningitis can also lead to hearing and vestibular loss, but the incidence of this infection in the UK has declined dramatically following the introduction of Hib vaccination in 1992. Overall, bilateral vestibular loss after bacterial meningitis is estimated to occur in 40–80% of cases.[4] Peripheral vestibular damage usually accompanies severe to profound deafness but can occur in the absence of hearing loss and can be unilateral or bilateral.

The onset of balance difficulties occurs whilst the child is still unwell and receives less attention than the finding of a significant hearing loss, whilst the reduced tone associated with a sudden vestibular loss can be mistaken for neurological damage. Regression in locomotor skills in the younger child is an important indicator of significant damage. Permanent profound hearing loss is invariably associated with vestibular areflexia.

The onset of labyrinthitis ossificans can be rapid, particularly after pneumococcal disease. Characteristic changes on MRI scan of fibrosis, or on CT scan of ossification, are often first identified in the lateral semicircular canals and can be a sign of progressive obliterative disease, indicating the need for urgent cochlear implantation if deafness is evident. These changes can be seen within a few weeks of the meningitis, making it imperative that children with suspected deafness and motor regression are referred urgently for careful audiovestibular evaluation (see Figure 19.5).

Viral labyrinthitis and vestibular neuronitis.

An acute onset of marked rotatory vertigo, often following an upper respiratory tract infection, suggests either of these two diagnoses: vestibular neuronitis occurring without hearing loss and labyrinthitis with sensorineural hearing loss as part of the presentation. A viral aetiology is assumed. The acute symptoms resolve over a few days and most children feel well enough to resume normal activities after about a week. However, residual symptoms can be reported over the first year or two. Although this is probably quite a common condition it rarely presents to the specialist unless there is delayed compensation. A unilateral peripheral vestibular lesion is commonly found early but Taborelli et al.[21] in a study of twenty-one children with vestibular neuronitis found resolution of symptoms and of vestibular findings in the majority over a 2-year period and in all within 5 years.

Lyme disease

Lyme disease, due to infection by the spirochete *Borrelia burgdorferi* found on deer and some other animals, can give rise to vertigo as part of the presentation and should be considered as a differential diagnosis with a history of exposure to tick infested land in an endemic region. Diagnosis is confirmed by identifying specific antibodies in blood, and treatment with antibiotics is effective. The presentation is similar to that of syphilis.

Traumatic

Fractures

Fractures of the petrous temporal bone, whether longitudinal or transverse, which involve the labyrinth or the internal auditory canal usually give rise to a total hearing loss and total unilateral vestibular loss. Vertigo is present acutely but resolves with compensation.

Labyrinthine concussion

This can occur following head trauma with or without a fracture, giving rise to symptoms of vertigo and dysequilibrium with findings of acute peripheral vestibular failure. Positional nystagmus is common. Symptoms usually resolve over several weeks.

Perilymph fistula

A perilymph fistula occurs when there is an abnormal connection between the inner and middle ear leading to a leakage of perilymph. It presents with fluctuating vertigo in addition to fluctuating and progressive hearing loss. The symptoms are exacerbated by straining or coughing. Opinion is divided as to causation; most clinicians consider that a perilymph fistula arises only as a consequence of trauma, be it accidental or iatrogenic, cholesteatoma or congenital malformation of the inner ear, rather than occurring spontaneously; others consider spontaneous occurrence a valid cause. The fistula test is positive in about 50% of cases of perilymph fistula. Treatment is surgical.

Barotrauma

Flying rarely precipitates vestibular disturbance due to barotrauma, but diving can give rise to acute vestibular symptoms either because of decompression sickness or a failure to equalise middle-ear pressures, which can lead to a perilymph fistula.

Iatrogenic

With surgical procedures on the middle or inner ear, including cochlear implant, there is always the risk of vestibular trauma giving rise to post-operative vertigo. Compensation usually occurs quickly.

Benign paroxysmal positional vertigo (BPPV)

This condition is rare in children although common in adults. It presents with recurrent episodes of acute rotational vertigo related to positional change of the head. It is ascribed to the

presence of loose otoconia in the vestibular system and is considered to be a degenerative condition showing an increasing incidence with age. Post mortem findings have shown a significantly low incidence of debris in the labyrinth in children compared to adults. BPPV can, however, occur in children as a consequence of trauma. The Dix-Hallpike positioning test demonstrates classic findings, which are diagnostic: transient torsional nystagmus towards the downmost ear accompanied by symptoms of vertigo or distress with delay in onset, reversed nystagmus on sitting up and fatigue with subsequent tests. Treatment is with particle repositioning manoeuvres.

Auto-immune

Auto-immune inner-ear disease is a rare cause of vertigo and vestibular hypofunction as well as progressive hearing loss at all ages. Cogan's syndrome is the best known and is defined as a non-syphilitic interstitial keratitis associated with a cochleo-vestibular deficit. It usually occurs in early adult life but can affect children. Hearing loss is bilateral and often quite rapidly progressive, although fluctuation can be reported. With accompanying tinnitus and vertigo the presentation can be similar to Ménière's disease. Investigations indicate non-specific inflammation with a raised ESR, CRP and complement but there is no specific diagnostic test. Labyrinthine fibrosis and ossification can occur early. The eye symptoms and signs are important in determining the diagnosis.[22] An associated vasculitis can be life-threatening and steroids and immunosuppressive drugs are the mainstay of treatment.

Vascular

The arteries to the inner ear are end arteries and vascular occlusion can give rise to sensorineural hearing loss and vestibular damage; however, such events are rare in childhood. Acute vestibular dysfunction has been described as occurring in children with sickle cell crises.[23]

Ototoxic

Vestibular ototoxicity is an important, but often overlooked, side effect of treatment with drugs such as aminoglycosides, cis-platinum and ethacrynic acid or exposure to heavy metals or chemical solvents. The effect of aminoglycosides on the vestibular hair cells can be more marked than on those of the cochlea. The unsteadiness or ataxia occurring as a consequence of medication is often attributed to the severity of the illness.[24] Whilst hearing loss is an acknowledged side effect of these medications and regular screening of hearing is advocated, particularly with cis-platin, vestibular damage is largely ignored but can be debilitating for the patient.

Neoplastic

Vestibular schwannomas are very rare in children, unless as a consequence of neurofibromatosis 2 (NF2), a dominantly inherited condition caused by a defect in chromosome 22q and characterised by multiple meningiomas and schwannomas. Presentation may be as a peripheral lesion or as a cerebro-pontine angle tumour and thus central. In a review of the world literature of 39 cases in children of 16 years or younger, Pothula et al.[25] reported hearing loss and tinnitus to be presenting symptoms in 58.9%, other neurological symptoms suggestive of raised intra-

cranial pressure in 35.8%, cerebellar dysfunction in 17.9% and facial nerve palsy in 25.6%. Boys predominate in this age group and only nine had presented in the first decade. In a study of 483 patients, with vestibular schwannomas in North West England over a 10-year-period Evans et al.[26] identified 12 patients under 20 years of age (2.5% of cases) of whom only 5 (1% of total) were symptomatic.

MRI scan is diagnostic, the tumour is benign and treatment is surgical excision.

Endolymphatic hydrops

Ménière's disease

Ménière's disease or idiopathic endolymphatic hydrops, is rare in children. Presentation is similar to adults with episodic vertigo, fluctuating low-frequency sensorineural hearing loss, tinnitus and aural fullness. Symptoms progress with time. Choung et al.[27] report an incidence of 2.6% of the total number of patients with Ménière's disease and 2.0% of children presenting with vertigo. Episodic vertigo is reported as an early symptom with hearing loss becoming evident over time. The pathogenesis remains unknown although both an auto-immune basis and channelopathies have been considered. A low salt diet can be helpful and early management with diuretics is advocated. Surgical management has been described as having value in severe refractory cases.

Familial Ménière's disease accounts for about 7–15% of cases overall. Inheritance is autosomal dominant with incomplete penetrance and demonstrates anticipation (earlier age of onset with successive generations). Whilst it has been considered to be due to a mutation in the *COCH* gene on chromosome 14,[28] more recent papers have thrown doubt on this.

Migrainous vertigo is an important differential diagnosis in children and differentiation can be difficult. Migrainous vertigo is much more common in children and is the more likely diagnosis for episodic vertigo even if there is an accompanying mild low-frequency sensorineural hearing loss.

Delayed or secondary endolymphatic hydrops

This is described as occurring in ears previously affected by long-standing hearing loss. Presentation is of episodic vertigo accompanied by fluctuating deterioration in the low-frequency sensorineural hearing, tinnitus and aural fullness. In unilateral hearing loss the ipsilateral ear is usually affected, although cases have been reported as affecting the contralateral ear. The underlying aetiology of the hearing loss has been ascribed to viral labyrinthitis, including mumps, trauma, meningitis, congenital CMV and congenital hearing loss. Onset in childhood is rare, particularly in the first decade, and other causes of episodic vertigo should be considered first.[29]

CENTRAL VESTIBULAR DISORDERS

Dizziness and imbalance are both quite common accompaniments of neurological disease in general. The following are specific entities likely to come to the attention of the neuro-otologist rather than an exhaustive list.

Migraine and migraine equivalents

Migraine and its equivalents are the most common cause of episodic vertigo in children. The association of vertigo with migraine is well recognised and dizziness is reported as a complaint in about a third of children with migraine. Studies on adult patients have identified that vertigo is about three times more common in migraineurs than in controls, whilst of those presenting with vertigo approximately 32% will give a history of migraine.[30,31] The International Headache Society classification of headaches includes vertigo as part of the aura of *basilar migraine* and also recognises *benign paroxysmal vertigo of childhood (BPVC)* and *cyclical vomiting* as migraine equivalents in childhood; however, many authorities consider this classification inadequate with regard to migrainous vertigo[31,32] and benign paroxysmal torticollis of infancy (BPTI). BPTI, BPVC, migrainous vertigo and migraine (with or without aura) probably form part of a continuum of a similar pathology presenting differently at different ages.

Benign torticollis of infancy (BPTI)

BPTI is uncommon and characterised by episodic torticollis or head tilting with cervical dystonia. First described by Snyder in 1969,[33] the onset is in the first year of life with spontaneous resolution occurring usually after a matter of months or a few years and certainly by 5 years of age. The recurrent episodes are sudden in onset and often short-lived, although the tilting can be present for hours or even days in some. Vomiting, pallor, ataxia, irritability and drowsiness can accompany BPTI and a positive family history of migraine is reported in more than 50%. There are no neurological signs between episodes and EEG and neuro-vestibular investigation are reported as normal. The underlying pathogenesis has been considered as being due to transient ischaemia of the brainstem and debate has suggested a channelopathy as the root cause. Reassurance is the mainstay of management. In some children there is progression into BPVC and migraine as time progresses.

Benign paroxysmal vertigo of childhood (BPVC)

The first description of this uncommon condition is attributed to Basser in 1964,[34] although there are earlier reports. It presents at about the age of 18 months as short-lived episodes of vertigo (a few minutes) of dramatic onset when the child is frightened and unsteady. Characteristically, they will cling to the cot sides or mother for support. Recovery from the episode is quick and complete without instability, and the child is symptom free between attacks. The children are described as eloquent and can usually give a good account if old enough. There is no hearing loss and vestibular function tests are usually normal. A family history of migraine occurs in more than 50%. This is a self-limiting condition and spontaneous resolution usually occurs by the age of 5–7 years. Although headache is not a feature of the condition BPVC is categorised as a migraine equivalent and symptoms often evolve into those of migrainous vertigo with time. Treatment with anti-histamines can be helpful.

Migrainous vertigo

Migraine is a common condition, with a reported prevalence in children of 2.7 to 10%. Episodic vertigo, with or without headache, is a recognised presentation of migraine in this age group.

The episodes can be variable in length, from minutes up to hours or even a couple of days, and can be strikingly regular in periodicity. Nausea and vomiting sometimes accompany the vertigo. Headache, which can be bilateral or unilateral, tends to be more common in the older child. The patient may predict the onset of the attack, which is often characterised by photophobia, phonophobia, prostration and post-ictal drowsiness. A family history of migraine occurs in more than 50%. A fluctuating mild low-frequency sensorineural hearing loss can occur which resolves with anti-migrainous treatment. Motion sickness is common amongst children with migraine as is a marked sensitivity to rotation, sometimes making vestibular assessment difficult.

The pathogenesis of migraine is not fully understood, but current hypotheses suggest an ion channel dysfunction (a channelopathy) leading to a spreading depression followed by a prolonged vasoconstriction. With the high incidence of a family history (50 to 77% in reported series) a genetic link seems likely although this has only been proved for familial hemiplegic migraine (see section on episodic ataxia). In migrainous vertigo it seems likely that the changes that occur due to vasoconstriction affect the brainstem in the region of the vestibular nuclei and the vestibular pathways.

Clinical examination is usually unremarkable, and interictal vestibular findings are generally normal although mild peripheral or central vestibular abnormalities may be present on testing. Diagnosis is dependent on the history, the exclusion of other causes of vertigo and the response to treatment. First line management includes dietary exclusion of caffeine, chocolate, cheese, citrus fruits and monosodium glutamate; remembering that several very popular carbonated drinks contain caffeine. Medication is useful and whilst simple analgesia is effective for some, those with more frequent episodes often respond to migraine prophylaxis such as pizotifen or beta blockers; in older children specific migraine medication, such as selective 5HT agonists, may be useful. The advice of a paediatric neurologist is recommended.

Epilepsy

Vertigo is recognised as occurring in association with epilepsy.[35]

- Vertigo as an aura preceding an epileptic seizure (grand mal, petit mal or psychomotor) is a relatively common occurrence and can help determine the epileptic focus – usually the temporal lobe.
- Vestibulogenic epilepsy is a seizure evoked by a vestibular stimulus such as a bithermal caloric and is rarely encountered.
- Vertiginous epilepsy is rare and more difficult to diagnose. The epileptic fit presents as episodic vertigo of sudden onset and often short duration. The symptoms cease abruptly with no interictal symptoms. Loss of consciousness can be transient and is usually observed by others rather than the patient. Diagnostic EEG abnormalities may not be present between seizures but have been reported as occurring more commonly in sleep or sleep-deprived EEGs. The temporal lobe is considered the most likely origin although other areas of the cortex can be implicated.

Whilst the first two types of epileptic seizures are relatively easy to diagnose vertiginous epilepsy can be a diagnostic challenge requiring a very careful history. Neurological and vestibular findings are normal interictally.[35]

Episodic ataxia

There are two types of episodic ataxia either of which may present in a vestibular clinic. They are rare, dominantly inherited channelopathies and both respond to treatment with acetazolamide. Channelopathies are due to genetic mutations which affect ion channels in the nervous system.[36]

Episodic ataxia type 1 (EA1) is characterised by intermittent ataxic episodes that may occur spontaneously but are often precipitated by exercise, fever, stress or even sudden movement. Vertigo is considered not to be a feature. The attacks may last seconds to minutes and may occur several times a day. Myokymia, a rippling of muscle, is usually observed in the periorbital and small hand muscles and can persist interictally. EA1 maps to human chromosome 12p13, and the disease is caused by mutations in the potassium channel gene *KCNA1*.

Episodic ataxia type 2 (EA2), also known as familial or hereditary paroxysmal cerebellar ataxia or acute intermittent cerebellar ataxia, is characterised by intermittent attacks of ataxia and dysarthria, lasting from a few minutes to a few days, triggered by exercise or stress. Additional symptoms include nausea, vertigo, diplopia and oscillopsia. Neurological findings include permanent gaze-evoked nystagmus and mild cerebellar ataxia. Evidence suggests that familial hemiplegic migraine, EA2 and spinocerebellar ataxia type 6 are allelic disorders occurring as a result of mutations in the calcium channel gene, *CACNA1A*, on chromosome 19p13, with the nature of the mutation affecting the functional activity of the calcium channel differently, thus dictating the clinical phenotype.

Multiple sclerosis

Multiple sclerosis is a very rare cause of vertigo in childhood. Between 2.7 and 5%[36] of cases are reported as occurring in children, predominantly during the second decade. The aetiology is unknown and diagnosis is dependent on the history and clinical findings, including abnormal eye movements and central vestibular abnormalities. Confirmatory evidence consists of abnormal evoked potentials, changes on MRI scan (T2 weighted) and the presence of oligoclonal bands and other changes in CSF. The presentation can include vertigo and ataxia due to involvement of the vestibular pathways, brainstem or cerebellum. In a prospective study of 54 children with MS under 16 years of age brainstem dysfunction was noted in 27.7%, motor and sensory disturbance in 27.7 and 16.8% respectively, optic neuritis in 13.2% and cerebellar disturbance in 13.2%.[37] The long-term prognosis does not appear to be more severe than for adults.

Congenital abnormalities of the skull base

Arnold Chiari I, the mildest form of hindbrain herniation through the foramen magnum, can be associated with symptoms of impaired vestibular function. This usually presents in the third to fourth decade but has been described in children during the second decade. Dizziness, imbalance and vertigo are among a variety of neurological symptoms such as headache, upper limb weakness, pain and sensory deficits. Kumar et al.[38] presented findings in 77 patients, aged 11 to 67 years, in which they reported imbalance in 45%, dizziness in 32% and vertigo in 21%. Findings include sensorineural hearing loss and central vestibular abnormalities including downbeat nystagmus. The diagnosis is confirmed by MRI scan (sagittal sections) and

management, either conservative or surgical decompression, is dependent on the clinical severity.

Infections

Intracranial infections, e.g. cerebral abscess, encephalitis, meningitis, can present with vertigo as a symptom.

Trauma

Vertigo and unsteadiness can occur following significant head injury, neurosurgery or cranial radiotherapy and may need specialist rehabilitative physiotherapy. Vestibular symptoms can occur following whiplash injury.

Neoplasia

Intracranial tumours are a rare but important cause of imbalance or vertigo in children. The majority of childhood brain tumours occur in the posterior fossa and include medulloblastomas, cerebellar astrocytomas, brainstem gliomas, ependymomas and, rarely, vestibular schwannomas. They can present with unremitting dizziness or vertigo, positional vertigo and progressive instability or ataxia. Abnormal neurological findings or central vestibular signs indicate the need for an urgent MRI scan and neurological opinion.

Heredodegenerative diseases

Heredodegenerative disorders are rare conditions which can include progressive ataxia and imbalance as part of the clinical presentation. In some, e.g. Refsum disease, Wolfram syndrome, the child can also present with deafness. Diseases affecting the brainstem and cerebellum give rise to abnormal neurological signs and central vestibular abnormalities.

Infantile Refsum disease, one of the leukodystrophies, is a rare recessively inherited peroxisome biogenesis disorder in which toxic levels of phytanic acid accumulate in the brain, leading to progressive ataxia as part of the symptomatology. The presentation includes retinitis pigmentosa and sensorineural deafness.

Wolfram syndrome, or DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness) is a rare recessive genetic disorder the presentation of which also includes brainstem atrophy with increasing unsteadiness and findings of central vestibular dysfunction.

Motion sickness

This is more common in children than in adults but is usually relatively mild. It is generally considered to be due to a mismatch between vestibular, visual and proprioceptive input during travel and is rare before the age of 2 years. Girls are more commonly affected. It occurs more frequently in children with a history suggestive of migraine and can be a significant problem in those with vestibular processing difficulties, in which case it can occur at a younger age. Management involves practical tips: be prepared for the vomiting, avoid travelling on a full stomach, break the journey up, open a window near the child, allow sips of water during the journey etc. Anxiety about being sick will increase the likelihood of vomiting because of hyperventilation. It is often the unpredictability of the car's movement that triggers the nausea,

and sitting an older child so that they can see the road ahead and predict turns and stops can help. Distracting a child by looking at the scenery in the middle distance, so that the visual input can more closely match the vestibular, is useful, while reading a book can have the opposite effect. Ginger can be a valuable treatment and anti-histamines are useful for the older child.

Vestibular processing deficit/sensory integration dysfunction

Jean Ayres, an occupational therapist, in 1972,[39] described problems with balance in children which she considered to be due to poor central integration of visual, proprioceptive and vestibular input. The concept can explain difficulties seen in some children with poor balance. They can present with severe motion sickness from a very young age or unexplained falls and clumsiness. These children may have poor saving reflexes and gravitational insecurity, with inordinate fear in challenging situations, e.g. stairs, uneven surfaces and playground swings. They often give a history of other processing difficulties such as dyslexia or dyspraxia. They may have visual problems such as vergence deficits or strabismus and may have mild neurological deficits such as hypotonia or learning difficulties. Hearing is unaffected. Vestibular hyper-responsivity, particularly of otolith function, has been reported. Assessment by an occupational therapist is indicated and treatment options include specific therapy to facilitate integration of sensory inputs. This condition has limited recognition and needs further consideration.

NON-VESTIBULAR CAUSES

Many non-vestibular conditions present with imbalance or dizziness and can offer important differential diagnoses.

Psychogenic, hyperventilation, abuse

It is well recognised that hyperventilation gives rise to dizziness due to the effects of hypocarbia in cerebral perfusion. In turn, anxiety or panic will promote hyperventilation as part of the sympathetic response – a vicious cycle. The history is often vague and other symptoms suggesting anxiety may be present. A test of hyperventilation in the clinic may precipitate symptoms. Vestibular tests are generally normal unless the presentation is of anxiety-delaying compensation from a peripheral vestibular disorder, when a canal paresis or directional preponderance may be evident. Reassurance and explanation often help but psychological or psychiatric input may be needed.

Abuse can precipitate symptoms of anxiety in children who then present with dizziness. It must be remembered that abuse of children with disabilities is relatively common and it is in these children that a history of dizziness may be the most difficult to unravel.

Cardiac arrhythmias

Cardiac arrhythmias can give rise to syncope and dizziness. Of particular importance to the audiovestibular physician is Jervell and Lange-Nielsen syndrome, which can present with syncope; see above, in the section on genetic hearing loss. An ECG is an important investigation in the assessment of any child with vertigo.

Visual disorder

Poor vision can present as clumsiness, particularly in a child. If in addition there is a vestibular disorder, e.g. Usher I, Alström, the problem is compounded. A history of improved balance following prescription of strong glasses is not uncommon in children with vestibular hypofunction and significant refractive errors.

Prolonged use of computer games or television can provoke dizziness or vertigo in susceptible children. Anoh-Tanon et al.[40] propose, in a study of 27 children, that vergence insufficiency or latent strabismus can present with otherwise unexplained vertigo which may be provoked by use of computer games. Optic correction or orthoptic intervention has been shown to be an effective treatment in these children.

Vasovagal episodes and orthostatic hypotension

These can be common, particularly in pubertal girls. Dizziness or light-headedness on standing up quickly or feeling faint with prolonged standing can usually be clarified by a careful history, and advice should be given on avoidance of precipitants.

Miscellaneous

- *Musculo-skeletal*
  e.g. muscular dystrophy, Still's disease
- *Neurological*
  e.g. cerebral palsy, dyspraxia, Friedrich's ataxia, Dandy-Walker syndrome etc.
- *Metabolic*
  e.g. hypoglycaemia, diabetes mellitus, endocrine abnormalities
- *Toxic*
  Vertigo is a common side effect of many drugs, including piperazine, ibuprofen, anticonvulsants, alcohol, caffeine and recreational drugs.
- *Other*
  e.g. breath-holding spells, night terrors, anaemia.

MANAGEMENT

Acute vertigo

Children usually compensate very well from acute vestibular insult. This is possibly because of a number of factors: in general children enjoy vestibular stimulation and seek it out, e.g. being swung round, roundabouts, theme parks; children are accustomed to falling over when learning new skills such as walking, riding a bike, gymnastics etc. and do not feel it to be degrading as do adults should they fall; they are used to someone else being in control and so the loss of control with vertigo that adults fear does not have the same impact on a child. Younger children are also inclined to live in the present and have less fear of what might happen. This is not to say that children are not frightened by the perception of spinning that accompanies acute balance disorders but that they are more inclined to be active once the sensation diminishes. They therefore return to normal physical activities as soon as they can and in this way actively compensate. This may explain why we do not see many children with

uncompensated peripheral vestibular lesions in tertiary clinics, particularly amongst the younger age group; older children are more likely to be anxious about the symptom and its cause. Factors that delay compensation can be visual difficulties, anxiety and problems in other areas of development such as cognitive, locomotor or processing skills. Delayed compensation after an acute episode may need the support of a physiotherapist to reinforce age-appropriate exercise sequences designed to encourage eye, head and neck movements. This will have the effect of facilitating central compensation in much the same way as vestibular rehabilitation in adults.

Acute vertigo is a frightening experience, which can engender anxiety in both the parents and the child. Whilst most children compensate well, particularly if the episode is mild and short-lived, there are others who are frightened by the experience and may develop school refusal or illness behaviour. Recurring episodes of acute vertigo can be particularly distressing and disruptive to normal family life, and the situation is often exacerbated by the comparative rarity of the problem and the difficulty the family might face in obtaining a definitive diagnosis. Much is to be gained by a careful empathetic assessment of the problem, a lucid explanation, gentle reassurance, practical coping strategies and a positive approach. Addressing your explanation, advice and plans clearly to the child, if old enough, as well as to the parents, ensures that the patient understands the problem, its investigation and its management. Asking the child or parents to keep a simple diary provides the physician with useful information about exacerbating and relieving factors, periodicity, severity and accompanying symptoms and often facilitates the child's account. Both relaxation exercises and breathing exercises are invaluable as management techniques for the anxiety that usually accompanies vertigo, particularly in the school-age child, and are particularly valuable strategies for coping with episodic vertigo. Anxiety as an underlying factor delaying compensation, or affecting a child's ability to cope with more chronic vestibular difficulties, may need psychological intervention. Visual problems may exacerbate or precipitate dizziness or instability and should be considered in all children.

Although not often required, medication can be of value; anti-histamine vestibular sedatives have their place during the acute vertigo associated with viral labyrinthitis, vestibular neuronitis or following trauma but should be kept to a minimum in order to allow compensation to occur. They are also useful for motion sickness and BPVC. Specific treatment for migrainous vertigo, EA2 and Ménière's disease is outlined earlier in this chapter.

Advice about avoiding dangerous situations is important, and recurrent vertigo may affect the child's ability to learn to drive or operate machinery.

Chronic imbalance

Chronic instability with delayed motor milestones, frequent falls and clumsiness is a different problem from acute vertigo and may require considerable therapeutic input from a multidisciplinary team, which could include an audiovestibular physician, neuro-developmental paediatrician, physiotherapists, occupational therapists and psychologists. Occupational therapy, particularly, can offer a lot for these children, enabling them to improve everyday functional balance. The importance of good vision cannot be underestimated in these children.

Children with congenital vestibular hypofunction associated with hearing loss often fail to get extra therapeutic help because slow locomotor skills are so common in this population. There is also a tendency to consider a child who is late with locomotor activities as having a 'general developmental delay' rather than a specific vestibular problem, overlooking the need

for specific therapeutic input. These children eventually learn to function so well that after early childhood it is difficult to see that they have a problem until they are challenged with a darkened environment and uneven surface. In spite of their ability to learn to cope, they do report difficulties such as never being very good at sports, being unable to join friends in more adventurous activities or the social embarrassment of having to hold someone's hand as they negotiate darkened stairs in a cinema. Early habilitation for balance difficulties has been shown to be of value[41] in improving functional balance. More understanding of the effects and management of vestibular problems will facilitate care for these children.

Children with significant vestibular hypofunction should not swim unsupervised and should be warned about swimming in deep murky water, where the lack of vision and proprioception can lead to disorientation and a real risk of drowning. Other situations, such as a skiing white-out or an uneven surface in the dark can put the child at risk of injury. Although children have a tendency to avoid situations they find difficult, career advice may be important, particularly for the older child with an acquired problem.

CONCLUSION

The differential diagnosis of balance disorders in children can be complex and requires a methodical and detailed medical approach with a careful history and thorough examination backed up with pertinent testing, investigations and assessments within a multidisciplinary team setting. Specific treatment and rehabilitative input should be considered for all children with persistent or debilitating disorders of balance.

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20 Vestibular testing in children

C. Möller

INTRODUCTION

Disclosure and diagnosis of vestibular dysfunction are generally difficult. In adults, the anamnesis and a thorough test battery are of uttermost importance. In children, it is more difficult to get good case stories from the child or the parents. The very young child when subject to attacks of dizziness often responds with crying, pallor, sleepiness etc. These symptoms can be caused by a variety of disorders. The test battery that can be applied is, however, quite extensive; today, it is possible to perform many vestibular tests in even very young children.

At birth, the newborn infant experiences a new world, where it suddenly is exposed to new kinds of movements and positions. The sensory systems are, however, fully developed at birth; from then on it is a matter of adaptation and learning. The task of keeping good balance is performed by three different systems:

1. The proprioceptive (somatosensory) system
2. The visual system
3. The vestibular system.

Afferent signals from all three systems are conveyed into the brainstem, pons and cerebellum; there they are processed and then transmitted through efferent nerve fibres mainly to muscles and eyes to maintain coordinated movements.

Assessment of these three systems and the central nervous processing is essential when evaluating children with balance disorders. Thus, the assessment of a child should include tests of the vestibular organs, the eyes, proprioception and central nervous system (CNS) integration.[1]

When a child is suspected of having a balance disorder, it is wise to develop a strategy of 'down-to-earth questions', balance assessment and, most importantly, observation of the child.

Children with a congenital or early-acquired genetic non-syndromal or syndromal deafness might have an additional bilateral vestibular hypofunction or areflexia. One of the first symptoms in a young child with bilateral vestibular arflexia or severe vestibular dysfunction is delayed motor milestones. 'Normal' motor milestones are displayed in Table 20.1. Many children with a bilateral vestibular areflexia have, before a correct diagnosis, been described as 'floppy infants' with consequent suspicions of other more severe CNS disorders.

Questions concerning a child with late motor milestones should concentrate on addressing certain 'events' which parents might remember. Examples of questions directed towards a suspicion of bilateral vestibular hypofunction or areflexia are shown in Box 20.1.

Table 20.1 Early motor milestones.

6 weeks	Holds the head in the plane of the body
12 weeks	Holds the head above the plane of the body
16 weeks	Good head control
6 months	Sits unsupported
10 months	Stands up with support
12 months	Walks

Box 20.1 Questions for indicating vestibular dysfunction.

At what age did the child lift his head, roll around?
At what age did the child sit unsupported?
At what age did the child walk unsupported?
Did the child experience difficulties in learning to bicycle?
Does the child have problems when walking in darkness and on an uneven surface?
Does the child experience motion sickness?
Does the child have problems in gymnastics and sport activities?
Is the child considered to be clumsy?

Box 20.2 Diary records.

Symptoms
Pallor, vomiting, unsteadiness, headache, falling etc.
Frequency of attacks: increasing, decreasing
Duration of attacks: hours, minutes, seconds
Time of day
Other symptoms: seizures, hemiparesis etc
Drugs: aminoglycosides, chemotherapeutics, diuretics etc.

In a child with a complete bilateral vestibular loss, all questions except motion sickness should be answered ‘yes’. A person with bilateral areflexia cannot experience sea or car sickness.

When a child is having attacks of vertigo or dizziness, suggesting a variable vestibular dysfunction, a suggestion is to have the parents make a diary, where they should note the information shown in Box 20.2.

Balance assessment is, of course, dependent on the age of the child, but many tests can be performed from a very young age.

OBSERVATION OF THE CHILD DURING PLAY

This takes time but will often give extremely useful information. It might be advisable, in some cases, to ask parents to perform this at home by DVD-video recording. These field studies

Figure 20.1 Observation of the child during play.

should be performed in different settings and light conditions since even small visual cues might mask a vestibular disorder (see Figure 20.1).

EAR, NOSE AND THROAT (ENT) AND DYSMORPHOLOGY EXAMINATION

This is strongly advised in order to detect dysmorphology of the face, palate, outer ear, ear canal, tympanic membrane and middle ear which, in some cases, might suggest an inner-ear abnormality as well.

CRANIAL NERVES, DEEP TENDON REFLEXES AND DEVELOPMENTAL REFLEXES

These can be tested even in very young children. If more than one cranial nerve is dysfunctional, this might suggest a syndromal disorder, not only affecting the auditory and vestibular organs.

Romberg testing

Standing on one leg can be performed in children older than 4 years. The performance in younger children shows, however, a large variation and depends on cooperation amongst the examiner, the child and the parent. A pathological test result is non-localising and can depend on many various disorders.

Smooth Pursuit and saccade tests

The Smooth Pursuit Test is important to assess the possibility to follow a slow-moving target. In young children, these tests are best performed by direct observation of eye movements

Figure 20.2 The Gaze Test.

whilst moving toys in front of the child. Saccadic testing (rapid change of fixation) is difficult to perform, but some information can be obtained by having the child look at various visual cues from left to right and vice versa. With some experience, it is possible to assess the accuracy of the smooth pursuit and saccades, and detect signs of eye muscle paralysis.

Gaze (fixation) Test

The purpose of the Gaze Test is to identify the presence of nystagmus during visual fixation. The child is asked or stimulated to fixate an object in a position 30 degrees to either side of centre gaze. A gaze nystagmus, especially if it is present in different positions, might indicate a lesion within the brainstem and/or cerebellum. If, however, a unilateral nystagmus is at hand, the cause might be an acute unilateral vestibular lesion[2] (see Figure 20.2).

Vestibular Head Impulse Test

This is a fairly new test, making it possible to discover a large unilateral vestibular lesion. The test can be performed from 4 to 5 years of age. The child is asked to fixate the examiner's nose whilst the head is rapidly turned approximately 50–60 degrees to one side. During the test, a close observation of the eyes is maintained. If the vestibular function of the lateral canal and/or the nerve is weak compared with the other side, small correcting saccades will appear.[3]

Video-oculography (videonystagmography, VNG)

Video-oculography will give the clinician very good information concerning pathology of the vestibulo-ocular reflex, noting possible spontaneous, positional and rotatory nystagmus. Most devices have goggles fitted with infrared TV cameras in order to assess the eye movements in complete darkness without visual fixation. These devices are easy to handle, and not too expensive for office use. With VNG, rapid screening can be performed by rotating a child with

hearing loss or profound deafness in the office chair, noting a possible bilateral vestibular areflexia.

Electronystagmography (ENG)

Formerly widely used, ENG can be performed in small children but is subject to difficulties with calibration and irregularities of eye movements, such as eye blinks. As in VNG, the eye movements are best recorded in darkness with a direct current (DC), electro-oculography (EOG) technique.

In order to assess one balance organ at a time, the best method so far is bithermal binaural calorics.

Bithermal binaural calorics (250 cc 30°, 44°C)

This test has been the mainstay in vestibular testing for many years. The test induces endolymph flow in the semicircular canals (primarily the horizontal), thereby creating an excitation or inhibition of the electrical discharge. Bithermal binaural calorics should be performed with eyes open in darkness. The velocity of the slow phase is the best parameter to assess. An inter-aural difference of >20% is considered pathological and a total sum of four irrigations of <40°/sec is considered hypoactive. Calorics can usually, with thorough preparation, be performed from 4 to 5 years of age. Ice-water calorics (50 cc, 8°C), performed binaurally, can be performed in older children who do not show responses during ordinary caloric tests. This test, however, cannot be quantified, but is merely a sign of some vestibular function. Caloric abnormalities usually reflect a vestibular end-organ lesion but can, of course, also be seen in an isolated brainstem lesion, although this is rare.

Rotatory tests

Sinusoidal Rotatory Chair Tests are by far the best test in order to evaluate possible bilateral vestibular loss or a large unilateral weakness in small children. This can be performed using infrared TV-monitoring. The tests should be performed in darkness and if a vestibular function is present, a resulting nystagmus will immediately appear. The tests are performed with different accelerations, usually from 0.02 to 0.64 Hz. This test is, however, difficult to quantify, and it will not differentiate between a moderate or compensated unilateral vestibular loss and bilateral normal vestibular function with certainty.

Dynamic posturography (balance and stability whilst in motion)

This consists of both sensory and motor components. Devices such as computerised dynamic posturography (Equitest) evaluate the integration of vision, proprioception (somatosensation) and vestibular input. The central integration of these inputs will result in different sway patterns. The Equitest system uses a movable force platform capable of quantifying sway and shear forces. A visual surrounding is also movable through tilting, thus creating six different visual and surface support conditions. The test might be helpful in assessing children with histories of imbalance and unsteadiness, bilateral vestibular dysfunction and delayed motor development and in monitoring progress or recovery. The test has large variations and cannot, with certainty, be applied before 6 years of age.[2]

New tests

New tests, in order to examine different parts of the vestibular organ (e.g. sacculus, utriculus, n. vestibularis sup and inf) are today available in large specialised clinics. One test that is quite widely used called Vestibular Evoked Myogenic Potentials (VEMP), assessing saccular function, has, however, not yet been documented in small children. Another test called Subjective Horizontal, assessing utricular function, could probably be used in children from the age of 5 to 6 years.[4]

CONCLUSION

Objective testing of balance problems and vestibular disorders in small children is not available for most clinicians. Most of the time, thorough anamnesis and office testing will be the best solution.

These clinical tests will solve problems in diagnosing most known acute vestibular disorders in children. In order to use vestibular tests in the process of diagnosing hearing disorders, a vestibular test protocol was produced by the European working group on genetics of hearing impairment (www.gendeaf.org).[5] The test protocol has been applied to different hearing disorders and syndromes. From a genetic perspective, vestibular tests can help discriminate between different non-syndromic deafness with identical audiometric patterns. It is likely that a phenotype reflects an underlying defect which is gene-specific. If so, hereditary deafness should also be categorised according to whether the labyrinth is involved or not. If vestibular involvement is consistent within families, then deafness with or without vestibular symptoms represents two major clinical categories of hearing impairment.

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21 Management of tinnitus in children

C.B. Coelho and R.S. Tyler

INTRODUCTION

In this review, a general framework for understanding and treating tinnitus in children is provided. Much of what is written about tinnitus in adults is directly applicable to children. Therefore, the reader is referred to these reviews for further details. In this chapter, the focus is on tinnitus that is distinct to children. First, studies reporting tinnitus in children are documented. Second, the causes of tinnitus in children are discussed. Third, the evaluation, treatment and prevention of tinnitus are reviewed. For other summaries of tinnitus in children, see Leonard, Black and Schramm[1] and Hegarty and Smith.[2]

THE EXISTENCE OF TINNITUS IN CHILDREN

Children experience tinnitus, but generally, they do not mention the symptom unless they are asked about it. In some cases, they might present similar suffering as observed in adults.[3] Tinnitus can cause a significant influence on children's lives which will inevitably affect their families as well.[4]

Investigating tinnitus in childhood is challenging because of its subjectivity and because children are different from adults in several ways. The rate at which children seek professional help might not reveal the true prevalence because children rarely report tinnitus spontaneously[5,6] and the presence of tinnitus is seldom an item in routine paediatric otolaryngological examinations. Therefore, it is difficult to get good estimates of the prevalence of tinnitus in children.

Population studies on the epidemiology of tinnitus amongst children have disclosed prevalence rates from 6 to 59% (Table 21.1). Such rates might be related to the significant difference amongst the studies on their methods of data collection, diagnostic criteria and age groups.

One important aspect is the differentiation between the perception of a sound (tinnitus sensation) and the impact of tinnitus on a person (tinnitus suffering). Information about the prevalence of tinnitus suffering in children is rare.

Clearly, children do have tinnitus. They describe their tinnitus as ringing,[7] as buzzing,[13] as 'beeping' or 'buzzing'[8] and as a 'high-pitched noise' or 'whistling'.[14] This is not unlike the tinnitus described by adults. Many young adults arrive in the clinic and note that they have had tinnitus as long as they can remember.

Hyperacusis and difficulty in concentration, sleeping and hearing are the most frequent complaints associated with tinnitus in children.[3,11,12,15] Interference with many leisure activities and with sports was reported by Coelho et al.[12] A decrease in school performance is mentioned by Drukier[16] and Kentish et al.[17] Tinnitus in children is reported to be more common in older

Table 21.1 Prevalence of tinnitus in children.

Author	Place	N	Age	Diagnosis based on	Prevalence	
Nodar[7]	USA	2000	10–18	Questionnaire	13.3% normal hearing	58.6% hearing-impaired
Stouffer et al.[8]	Canada	161	7–12	Interview	13% in normal hearing 6% after an answer consistency criteria	29% in hearing-impaired 24% after an answer consistency criteria
Holgers[9]	Sweden	964	7	Questionnaire	13% in normal hearing	8.8% in hearing-impaired
Holgers and Pettersson[10]	Sweden	671	13–16	Questionnaire	53% tinnitus perception	27% tinnitus annoyance
Aksoy et al.[11]	Turkey	1020	6–16	Questionnaire	9.2 % tinnitus perception	5.8% tinnitus annoyance
Coelho et al.[12]	Brazil	506	5–12	Interview	tinnitus perception: normal hearing 37.7% hearing-impaired 50%	tinnitus annoyance: normal hearing 19% hearing-impaired 17.8%

children than in younger children[7,11,12] and is probably associated with an increase of exposure to risk factors. Coelho et al.[12] also note an association with tinnitus, headaches and dizziness in some children. Exposure to impulse noise has been significantly associated with tinnitus. Holgers and Pettersson[10] and Coelho et al.[12] found that exposure to sounds in concerts, in discos, from firecrackers and from guns is associated with tinnitus.

HYPERACUSIS AND TINNITUS

Hyperacusis and tinnitus have been well described as related symptoms[18] and this relationship is apparently also present in children. Gabriels[15] has described complaints about annoyance hyperacusis in 30% of children presenting with tinnitus. The presence of hyperacusis has been demonstrated to be the highest risk factor for tinnitus suffering amongst children.[12]

CAUSES OF TINNITUS IN CHILDREN

Congenital tinnitus

Children with congenital tinnitus have had tinnitus since birth or infancy, and to them tinnitus can be considered ‘normal’. Typically, it is not until later in life, perhaps in conversations with a friend, that they discover that not everyone has ringing in their ears.[19] Graham[20] has proposed that children with congenital tinnitus can habituate to it at an early age.

Acquired tinnitus

Children who develop tinnitus later in life have acquired tinnitus. They are aware that the tinnitus represents a change – it was not there before. This experience is similar to what many

adults experience. Like adults with tinnitus, the child could habituate to the tinnitus, or the tinnitus can become a focal issue and a handicap.

Middle-ear tinnitus

Nearly all children experience at least one episode of otitis media. Whether this infection causes tinnitus is debatable, and if tinnitus does occur, its pathophysiology is unclear. One explanation is that the associated conductive hearing loss caused by a middle-ear effusion attenuates external sounds that normally mask low-level tinnitus. Removal of these external sounds 'unmasks' an already existing low-level tinnitus.[1]

In addition to otitis media, other middle-ear disorders can result in tinnitus in children. The list of these disorders is large and includes all tinnitus-inducing middle-ear problems of adults. Table 21.2 reports a variety of studies in which tinnitus is mentioned. As in adults, some forms of middle-ear tinnitus can represent a more general health problem, such as an intracranial tumour. In these cases, treatment of the underlying medical condition can alleviate the tinnitus.

Sensorineural tinnitus

Tinnitus can also be associated with sensorineural hearing loss from any cause. This is true in adults as well as in children. Children with moderate hearing loss tend to present tinnitus more frequently than children with severe and profound hearing loss.[12,19,42]

In Table 21.3, some of the more common causes are listed. Older employed children are susceptible to noise-induced tinnitus. Ototoxic drugs and recreational noise and music exposure are particularly noteworthy in children.

Ototoxicity

Ototoxic drugs are often administered to children, including newborns. As with adults, this type of exposure is associated with a risk for hearing loss and tinnitus. Although drug dose is adjusted for small body size, children may be particularly at risk for tinnitus.

Recreational noise- and music-induced tinnitus

Children can acquire hearing loss and tinnitus from recreational noise, such as snowmobiles, water jetskis, gunfire, toys or fireworks.[48,49] In addition, they are often at risk for music-induced

Table 21.2 Reports of middle-ear tinnitus in children.

Aetiology	Reference
Aberrant carotid artery	Glasscock et al., 1993[21]
Arteriovenous fistula	Lalwani et al., 1993;[22] Cataltepe et al., 1993[23]
Dehiscent jugular bulb	Rauch et al., 1993[24]
Glomus tumours	Bartels et al., 1988;[25] Thompson et al., 1989;[29] Jacobs et al., 1994;[27] Jackson et al., 1996;[26] Magliulo et al., 1996[28]
Middle-ear myoclonus	Howsam et al., 2005[30]
Palatal myoclonus	Quarry, 1972;[32] Deuschl et al., 1990;[31] MacDonald, 2007[33]
Patulous eustachian tube	Kavanagh and Beckford, 1988[34]
Transmitted bruit	Levine and Snow, 1987 (adults)[35]
Venous hums	Meador, 1982 (adults)[36]

Table 21.3 Reports of common causes of sensorineural tinnitus in children.

Aetiology	Reference
Air bag deployment	Mittal et al., 2007[37]
Head trauma	Gabriels, 1996[15]
Ménière's disease	Ménière, 1861;[38] Nodar and Graham, 1965[43] Parving, 1976;[41] Meyerhoff et al., 1978;[40] Hausler et al., 1987;[39] Gabriels, 1996[15]
Noise exposure	Gabriels, 1996;[15] Davis and El Refai, 2000;[44] Holgers and Petterson, 2005;[10] Coelho et al., 2007[12]
Neurofibromatosis type 2	Miyakawa et al., 2007[45]
Perilymph fistula	Parnes et al., 1987[46]
Sudden deafness	Chen et al., 2005[47]

tinnitus as either players or listeners if they are exposed to intense music (>80 dBA) for long time periods (>2 or 3 hours) on a routine basis (>3 or 4 days a week).

Generally, sound must be above 80 dB A for over 8 hours per day to be considered potentially damaging to hearing. Exposure at higher levels for shorter durations can also cause hearing loss. In fact, single bursts of very loud sound can damage hearing and produce tinnitus. There are no published guidelines for noise-exposure limits for tinnitus. Segal et al.[50] reported that 25% of children (n = 13) who sought medical care after being exposed to noise from toys and firecrackers complained about tinnitus.

Musicians

Hearing loss and tinnitus are not problems solely for rock musicians – they can occur in classical musicians as well. Risk is increased when the musician is playing next to the sound source, such as another instrument or a loudspeaker, for several hours over an extended time period.

Listeners

Hearing loss and tinnitus can occur in children who listen at rock concerts and 'dances', or through personal wearable or non-wearable sound playback systems. Rock concerts can certainly produce damaging noise levels[51,52] and can last for several hours. Incredibly, Yassi et al.[51] reported that 60% of attendees reported tinnitus immediately after a concert. Wearable headphones also produce high sound levels[53] that can produce a temporary threshold shift.[54]

EVALUATION

In general, the evaluation of children with tinnitus parallels that of an adult with tinnitus. There is perhaps one exception – children are frequently less verbal about their health conditions, and on questioning, they may attempt to please the health-care worker. Therefore, open communication and caution should be exercised during the examination. Children with tinnitus can have difficulty concentrating. Academic problems, difficulty sleeping, behavioural problems and hyperacusis can ensue.[15] A detailed physical and radiological evaluation has been described by Hegarty and Smith[2] for children, and Perry and Gantz[55] and Tyler and Babin[56]

for adults. Savastano[57] proposes a specific protocol to investigate tinnitus in children over age 8 years, including a general and otological history, subjective description of tinnitus, pure-tone audiometry, tympanometry, tinnitus measurements including tinnitus loudness and spectral composition, masking effectiveness and residual inhibition. However, our approach is somewhat different. We recommend that you use only those otological and audiological tests that are needed to determine diagnosis and treatment.

TREATMENT

Surgical treatment can be pursued for some types of middle-ear tinnitus as is the case for adults.[2,55–56] However, no safe medication has been shown to help large numbers of tinnitus patients in controlled investigations.[58,59]

Children with significant hearing loss and sensorineural tinnitus should be fitted with hearing aids. Hearing aids help tinnitus because they:

- improve communication and therefore reduce the stress of listening (reducing stress helps the patient cope with tinnitus);
- produce some background noise that facilitates masking or habituation; and
- amplify background noise that facilitates masking or habituation.

Treatment for young children

If a young child becomes aware of sensorineural tinnitus but shows no concern, it is advisable not to create concern. Many adults (and presumably children) have tinnitus and lead happy and productive lives.

If a young child is concerned about sensorineural tinnitus, a general discussion about the background and treatment is advisable (see Table 21.4).[60] Information must be offered at an age-appropriate level for each child.

Gabriels[15] noted that the effect of tinnitus on school and social life must be considered in the management of children. She noted one 5-year-old child with tinnitus and hyperacusis who attacked a schoolmate who shouted in his ear.

It also is important to engage the parents and siblings as they need to understand difficulties and problems related to tinnitus. Because tinnitus is usually subjective, in some situations it can be difficult for parents to accept their child's complaint.[15]

Kentish and Crocker[4] have described a technique for psychological treatment in those children that present tinnitus suffering and need intervention. This protocol includes a wide range of strategies, including relaxation techniques and educational assessments that have been developed from narrative techniques (see Tyler[62]).

Treatment for older treatment (and adults)

Tinnitus treatment for older children is similar to treatment for adults. When a child does complain of tinnitus, this should be taken seriously.[2] Not only can tinnitus be debilitating, but it can also reflect an underlying treatable disease. Graham[20] noted that tinnitus can result in behavioural problems in school. Table 21.5 lists some treatments that can be helpful in adults and could be easily adapted for older children.

Table 21.4 Background and treatment information that can be discussed with children (adapted from Tyler, 2000[61]).

	Information area	Content
1.	Prevalence	• About 1 in every 10 adults experiences some tinnitus • About 1 in every 200 adults is severely bothered by tinnitus • Many children have tinnitus
2.	Causes	• Noise- or music-induced hearing loss • Unknown • Medications • Head injury • Almost anything that causes hearing loss
3.	Treatments	• No magic pill • Strategies to help yourself • Low-level background sound • Hearing aids, counselling, relaxation, maskers, habituation

Table 21.5 Different tinnitus treatments available for adults that could be adapted for children (adapted from Tyler and Erlandsson, 2003[60]). See Tyler, 2006[61].

	Treatment	References
1.	Hearing aids	Tyler and Bentler, 1987[62]
2.	Counselling	Tyler, Stouffer and Schum, 1989[63] Wilson and Henry, 2000[64] Tyler et al., 2006[65]
3.	Relaxation	Erlandsson, 2000[66]
4.	Cognitive behaviour modification	Sweetow, 1986[67] Wilson and Henry, 2000[64]
5.	Masking	Vernon, 1998[68] Tyler and Bentler, 1987[62]
6.	Habituation	Hallam., 1989[69] Hallam and McKenna, 2006[70]
7.	Sound therapy	Tyler and Erlandsson, 2003[60]
8.	Refocus therapy	Tyler and Erlandsson, 2003[60]

PREVENTION

Even better than treating tinnitus, is preventing it. There are at least two causes of tinnitus in children that can be preventable: drug-induced tinnitus and noise-induced tinnitus. The use of ototoxic mediations should be carefully monitored using drug-specific parameters and encouraging the child to report changes in hearing or the onset of tinnitus. The potentially harmful consequences of exposure to loud music and noise (continuous and impulse) can be minimised by education on the importance of hearing protection and avoidance. Modelling the judicious use of hearing protection by siblings, peers and parents is beneficial. At-risk children also can be warned of the implication of the onset of tinnitus – it may be the harbinger of permanent noise-induced hearing loss.

GENETICS

The exact role of familial and hereditary factors on tinnitus has not been established yet. This gives us evidence that tinnitus genetics is not controlled by single genes (monogenic or Mendelian traits). Tinnitus is most likely to be controlled by interactions amongst multiple genes variants and environmental risk factors, the so-called multigenic or genetically complex traits.

CONCLUSION

Tinnitus does occur in children. In general, it seems to present itself in the same fashion it does in adult and is diagnosed and treated similarly. However, there are some differences and/or noteworthy comments, specifically:

- Children with tinnitus do not always report this symptom
- Children with congenital tinnitus can consider this 'normal'
- Children with tinnitus can present with academic, social, sleep and concentration difficulties
- Children with tinnitus can have hyperacusis
- Children are at risk for tinnitus secondary to ototoxic medications and intense noise or music
- In a young child with tinnitus, it is important to avoid creating a significant problem if none exists
- In managing children with tinnitus, it is important to consider the school and social environment and to engage the support of the family
- Tinnitus in children can be prevented with education about ototoxity, hearing protection and wise listening strategies.

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22 Development of early vocalisation and language behaviours of young hearing-impaired children

E.A. Tobey and A.D. Warner-Czyz

INTRODUCTION

Twenty years ago, David Luterman[1] published *Deafness in the Family* describing the emotional impact on a family when discovering their child had a profound sensorineural hearing loss. Using poignant samples of conversations with family members and didactic professional discussions, he reviewed the stages of grief (denial, anger, bargaining, depression and acceptance). He also noted the importance of affection issues of parents of children with sensorineural hearing losses by quoting a mother of a 2.5-year-old child, who said:

> I am glad I did not tumble to the fact that my child was deaf until she was 14 months old. I really enjoyed her for that first year. I was very carefree in my mothering. When I found out she was deaf, I became stiff and very self-conscious. I was terrified about the hearing aids and I was afraid of making any mistakes. There was no fun or joy then. (p. 37)

International efforts to establish newborn hearing screening programmes have changed the dynamics of services, expectations, technological assistance and outcomes in young children identified with sensorineural hearing losses. In the following pages, we will reflect on the changing landscape for hearing-impaired children by briefly reviewing how early intervention services impact young children, how parental interactions and expectations impact early communicative development, how modern technology enhances communication development and how communication outcomes in young children are influenced by the constellation of variables that impact their communication environment. This new paradigm shifts attention away from simple mother–child dyads to increased attention to fathers, siblings, extended family members, educational settings and community settings outside the home. A broader scope of attention is needed as identification of hearing loss in young children moves from parent-initiated models of diagnosis to institution-initiated models. As described in more detail later in this chapter, movement towards institution-initiated detection and identification of hearing losses in young children lowers the age of identification, the age of first intervention, the age of first hearing aid use, the age of first cochlear implant use, the age of first FM assistive listening device use and the developmental milestones associated with communication acquisition. It remains unclear, however, what the impact of institution-initiated mandates will be on long-term family dynamics.

CHANGING THE SERVICE DELIVERY LANDSCAPE

In the past, parent-initiated models of diagnosis were motivated by family observations over time that a young baby failed to respond to auditory stimuli within the family environment.

On parent-initiated visits, typically to their family paediatrician, a hearing loss would be detected and confirmed by additional testing by an audiologist. An example of a parent-initiated visit is reviewed in Luterman[1] from an interview with a mother who stated:

> When Nancy was 12 months old, [my mother-in-law] would say to me, 'Gee, Nancy doesn't seem to know her name', but because this was my first child and I thought that Nancy heard, I really didn't know what my mother-in-law meant. So when we went to the pediatrician, he clapped his hands several times and she turned around. He told us to watch her during the next week, so we began testing her but she didn't respond, so the pediatrician sent her to Children's Hospital. I remember sitting in the waiting room listening to the audiologist explain to another family that their child had nerve deafness. That was the first time it dawned on me that my child could be deaf. (p. 81)

A major characteristic associated with parent-initiated diagnosis was a delay in recognition of the hearing loss and, consequently, delays in initiating family- and child-oriented intervention programmes. For example, prior to state-mandated newborn hearing screening established in 1999, the Texas Department of State Health Services indicated the average age of identification of a hearing loss in a young child was 56 months in Texas, 31 months across the United States, and between 6 and 7 months in England and Israel (Texas Department of State Health Services). Following legally mandated newborn hearing screening to detect hearing losses, initial attempts to implement newborn hearing screening procedures reduced the identification ages to between 12 and 24 months.[2] Further improvement in screening procedures, false-positive identifications and family follow-up are leading to even earlier identifications, sometimes as early as age 3–6 months[3] (see Figure 22.1).

The changing service delivery landscape is being driven, in part, by the recognition of professionals that the first years of life are driven by an explosion of capabilities and any diminishment to the nurturing of those capabilities has major long-term effects. Perhaps one of the most significant observations made in the last 15 years was by Yoshinaga-Itano and her colleagues, who observed a reliable positive relationship between early identification and intervention of hearing loss on early language performance in children.[4] Yoshinaga-Itano and colleagues[4] compared the language abilities of 72 children whose hearing loss was identified by age 6 months with 78 children whose hearing loss was identified after age 6 months. Results

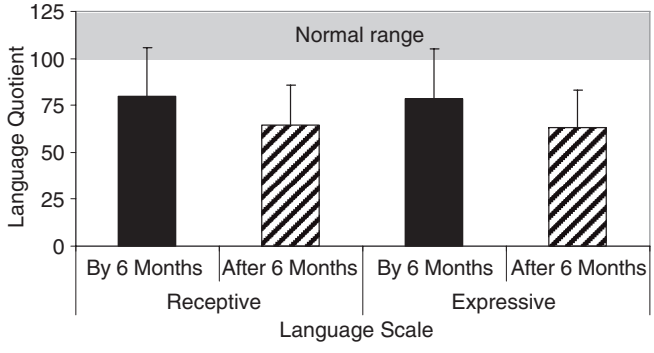

Figure 22.1 Language quotients are shown for children identified with a hearing loss before versus after 6 months chronological age. Although children identified by age 6 months demonstrate higher language quotients than those identified after age 6 months, all hearing-impaired children perform below the normal range for receptive and expressive language. Figure adapted from: Yoshinaga-Itano C., Sedey A.L., Coulter D.K., Mehl A.L. Language of early- and later-identified children with hearing loss. Pediatrics 1998.

from the *Minnesota Child Development Inventory* were reported as a language quotient, or the child's language age divided by chronological age and then multiplied by 100. A language quotient of 100 suggested appropriate language skills for a child's chronological age, whereas a language quotient less than 100 indicates delayed language development relative to the child's chronological age. Children with normal cognitive abilities identified before age 6 months achieved significantly higher receptive, expressive and total language skills than children identified after age 6 months. Age of identification outweighed all other variables, including the degree of hearing loss (mild, moderate, severe or profound), communication mode (an emphasis on listening/speaking or on using visually based communication systems), educational settings (special education, mainstream, private or public), socio-economic status, gender and the presence or absence of additional disabilities. Subsequent investigations continue to demonstrate increased benefits on communication development if early detection of a hearing loss occurs at even younger ages.

THE CHANGING LANDSCAPE OF EXPECTATION

As institution-based detection and intervention models gain momentum, the focus of professional efforts continue to hone in on the very young child. Efforts in this direction are aimed at impacting the child and family at critical early ages with the expectation that early intervention will lead to language performance approaching that of chronologically equivalent typical, normal-hearing peers. But what are the early indicators of communication development? What seems to be their natural course of development? In order to examine these expectations, it is necessary to review the concurrent and parallel development of maturing anatomical, physiological and behavioural systems contributing to the earliest forms of communication outputs (see Table 22.1).

Neurologically intact infants produce reflexive vocalisations and vocal play during the first 6 months of life.[5,6] In the earliest stages (birth through age 2 months), infant vocalisation

Table 22.1 Progression of early vocal behaviours leading to first words in normal-hearing infants.

Stages	Chronological age (months)	Vocal behaviour	Example
Pre-canonical	0–2	Crying	
	2–3	Cooing and gooing	/ku/ and /gu/
	4–6	Vocal play	Growls, raspberries, yells and squeals
Canonical	7–8	Canonical babbling	/bababa/
	7–12	Reduplicated and variegated babbling	Reduplicated: /baba/; Variegated: /babi/, /badi/
Lexical	12–18	Single words, 25–50% intelligible	
	18–20	Vocabulary spurt	
	20–24	Two-word combinations, 50% intelligible	/gi sal/ for *green slide*
	36	Three-word sentences, 75% intelligible	

outputs are composed of crying, fussing, coughing, sneezing and burping.[5] During this early time period, the corticobulbar and corticospinal tracts are not well formed and vocal outputs appear to be reflexive in nature. Kent[7] describes a newborn infant as being basically 'split-brained' due to underdeveloped cortical fibre tracts within and across hemispheres. Anatomically, a young infant's tongue consumes the oral cavity; the larynx lies high behind the tongue; and the soft palate and uvula lie parallel to the tongue and jaw. Contact between the epiglottis and uvula may occur, limiting a very young infant to a vocal tract consisting of a single tube. The tongue and jaw do not act as independent agents, leading to the production of quasivowels produced with the jaw and tongue acting as a single unit.[8] By age 2–3 months, infants enter a 'cooing stage' characterised by the prevalence of isolated back vowels like /u/ and syllables combining dorsal (produced in the back of the mouth) consonants like /k/ and /g/ with vowels also produced in the back of the mouth (i.e. /ku/ and /gu/).

Age 4–6 months marks a child's entry into the 'vocal play' period, during which they begin exploring the capacity of their vocal system. Anatomically, the larynx drops, forming a pharyngeal cavity, and the face grows down and forward, forming an oral cavity. The soft palate and uvula initiate their downward trajectory and, thus, separate contact between the uvula and the epiglottis.[9] During vocal play, vocalisations are characterised by raspberries, yells, squeals and growls. These vocal outputs allow the infant to explore vibratory patterns of the lips and larynx, aerodynamic contributions to intensity, increases and decreases in laryngeal frequency and alterations to vocal quality. The child also produces prolonged vowels (e.g. *aaaaah*) or prolonged syllables (e.g. *baaaaa*) termed 'marginal syllables' because, as in the cooing/gooing period, the timing of the opening and closure of these syllables is not yet adult-like.[8]

Hearing infants begin to produce rhythmic, syllable-like canonical babbling around age 7–8 months and continue producing babbling behaviours through the first word period until approximately age 18 months.[6,8,10,11] Oral consonants like /b/ and /d/ outnumber nasal consonants like /m/ and /n/ by a ratio of 3:1. Hearing infants favour producing sounds articulated in labial (e.g. /b, p, m/) and coronal (e.g. /d, t, n/) consonantal positions. Stops, nasals and glides are the most common manner of production, and lax vowels (e.g. /ɛ, æ, ʌ /) are the most common vowel types. Vowels with low (e.g. /ae/), mid (e.g. /ʌ, a/), front (e.g. /i, ae/) and central (e.g. /ʌ, a/) tongue placements predominate.[10,12,13] During this period, few dorsal consonants (e.g. /g, k/) and back vowels (e.g. /u, o/) requiring basic shaping of the tongue dorsum are observed.[7,14] A traditional perspective of the canonical babbling period suggests an initial phase of *reduplicated babbling*, in which identical consonant–vowel syllables are repeated (e.g. /baba/), followed subsequently by a period of *variegated babbling*, in which consecutive syllables are composed of different consonant and vowel segments (e.g. /badi/).[15–17] However, more contemporary studies of early vocal development indicate that reduplicated and variegated patterns co-exist from the onset of canonical babbling through early words.[10,18,19] When infants combine consonants and vowels into syllables, they maintain an articulatory compatibility such that the consonant and vowel segments share the same anterior–posterior position of the tongue.[10] Infants are more likely to produce a syllable with two segments in the front of the mouth (i.e. coronal /d/ with a front /i/) compared with a syllable that requires movement of the tongue between the consonant and vowel components (i.e. coronal /d/ with back /o/). Infants demonstrate considerable overlap in favoured phonetic patterns and syllable structures from canonical babbling through the single word period.[10,12,15,16,20–23] The numerous phonetic similarities between babbling and early words persist in studies of a variety of languages.[12,24,25–27] Phonetic similarities in vocal qualities represent universal aspects of early speech acquisition based on characteristics of the infant production system development (see Figure 22.2).

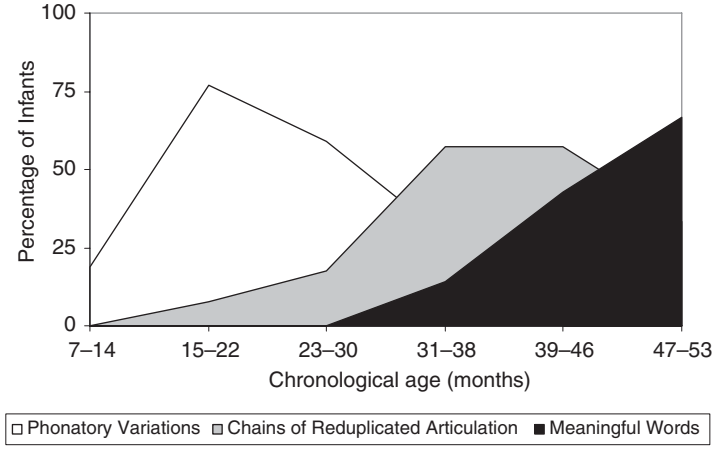

Figure 22.2 The proportion of typical, normal-hearing infants producing phonatory variations, strings of reduplicated articulations and meaningful words at various chronological ages during the first 5 years of life are shown. The greatest proportion of young children produce phonatory variations between ages 15 and 22 months. Figure adapted from: Koopmans van Beinum F.J., van der Stelt J.M. Early stages in the development of speech movements. In: Lindblom B., Zetterstrom R. (editors), Precursors of Early Speech. New York: Stockton Press, 1986; 37–49.

The most frequently produced consonants and vowels also tend to be the most accurate in the early word period. For example, children produce labial (e.g. /b, m/) and stop (e.g. /b, d/) consonants most accurately. Front and central vowels, particularly /i/ and /a/, are accurate most frequently. Word position affects how well young hearing children produce the consonants and vowels in their words. Consonants are most accurate in initial position whilst vowels are most accurate in medial position. When children do not produce the consonant or vowel in the target word, they tend to omit consonants and substitute vowels.[28] Fewer studies have detailed acquisition of accuracy in typically developing children in early words.[10,29–33]

First words herald the beginning of an explosive, dynamic acquisition of language constructs and elements. Evidence from large studies indicates 47% of typical, normal-hearing children produce two-word sentences between the ages of 12 and 23 months.[34] Three-word sentences (approximately 20%), and three- to four-word sentences (approximately 9%) occur less frequently during this early time frame. Most children between the ages of 24 and 35 months show considerable increases in the presence of two-word sentences (98%), three-word sentences (90%) and three- to four-word sentences (84%). Three-fourths (76%) of 2-year-olds achieve utterances judged 50% intelligible. Three-year-old children (between ages 36 and 47 months) produce more complex sentences composed of three to four words (approximately 98% of the children). At age 3 years, nearly 100% of typical, normal-hearing children produce adjectives and half of the children demonstrate over-generalisation of verb conjugations. Eighty per cent of typical, normal-hearing 3-year-old children are understood by others at least 75% of the time. Nearly all typical, normal-hearing 4-year-old children accurately name their colours and produce compound sentences. Over half of typical, normal-hearing 4-year-old children use adult-like language structures, with the majority of children being understood 100% of the time (see Figure 22.3).

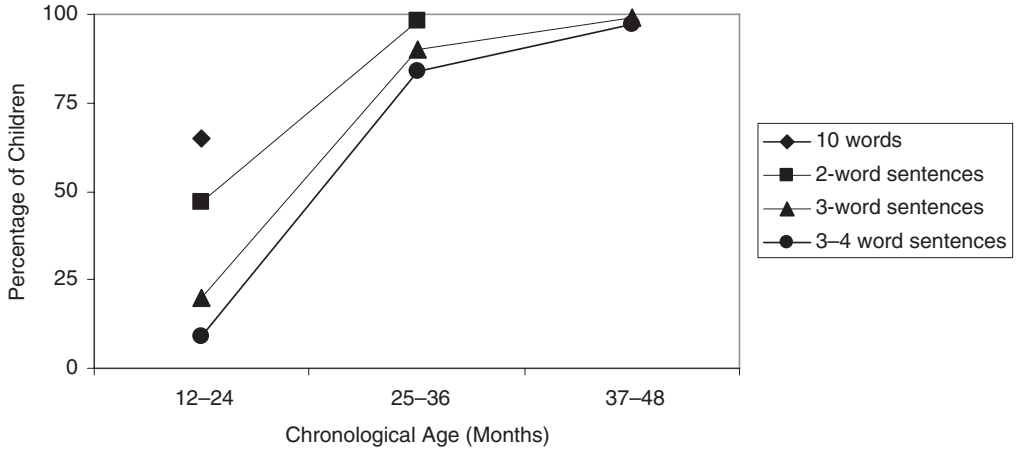

Figure 22.3 The percentage of typical, normal-hearing children demonstrating words and sentences of different lengths at different chronological ages is shown. Chronological age is positively associated with sentence complexity in children under age 4 years. Figure adapted from: Luinge MR, Post WJ, Wit HP, Goorhuis-Brouwer SM. The ordering of milestones in language development for children from 1 to 6 years of age. J Speech Lang Hear Res 2006; 49(5): 923–940.

THE CHANGING LANDSCAPE OF COMMUNICATION CONSEQUENCES OF HEARING LOSS

The broad impact of auditory perceptual input on early speech behaviours is striking, as it affects both the quality and quantity of vocal output.[35–37] As shown earlier, age at identification and degree of hearing loss significantly affect speech and language acquisition.[4] Identification and intervention efforts occurring before 6 months of age result in better oral language outcomes. Degree of hearing loss also affects speech and language performance. Children with mild hearing losses show little difference in early vocalisation behaviours compared with their hearing counterparts.[29] Speech–language abilities of children with moderate to severe hearing losses lag behind children with mild to moderate degrees of hearing losses, but the vocal abilities of the two groups converge by 5 years of age. Children with severe to profound hearing losses, however, exhibit different output patterns than their typical, normal-hearing or hard-of-hearing peers which are likely related to their inability to fully access the acoustic properties of speech. Other intact feedback systems (e.g. visual and kinaesthetic systems) provide cues to placement and movement of articulators; however, these systems are limited due to incomplete visual access to articulatory characteristics (e.g. tongue position) and the child's inability to interpret kinaesthetic cues.

Very early vocalisations of children with severe to profound hearing losses mirror those seen in typical, normal-hearing children through age 6 months. The similarity in early vocal behaviours between children with normal hearing and children with hearing losses contributes to the inability of parents to detect communication difficulties until age 8 months for children with severe to profound hearing losses and age 15 months for children with mild to moderate hearing losses.[38] At these critical early ages, when syllabic formations dominate babbling

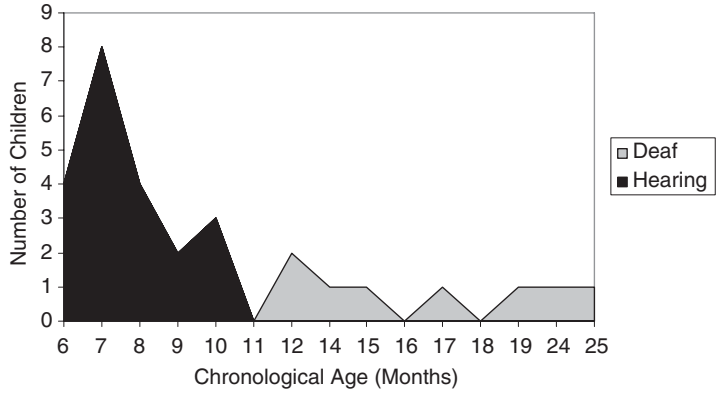

Figure 22.4 The chronological age of the onset of canonical babbling behaviours differs in children with normal hearing compared with children with sensorineural hearing loss. Typical, normal-hearing infants acquire canonical babbling by age 11 months, whereas children with sensorineural hearing losses fail to initiate canonical babbling behaviours within the same time scale as normal-hearing children. Figure adapted from: Oller D.K., Eilers R.E. The role of audition in infant babbling. Child Dev 1988; 59(2): 441–449.

behaviours, infants with hearing losses begin to deviate from the course of vocal behaviour observed in typical, normal-hearing children. Oller and Eilers[36] explored the onset of canonical babbling in infants with normal hearing and with severe to profound hearing losses. All infants with normal hearing began canonical babbling by age 10 months. In contrast, the onset of canonical babbling in infants with severe to profound hearing losses fails to begin until after age 11 months. Infants with hearing losses do not babble on average until age 16 months, nearly 8 months later than typical, normal-hearing children, with essentially no overlap in the onset distribution of canonical babbling between the two groups[21,36] (see Figure 22.4).

Children with severe to profound hearing losses begin canonical babbling with lower volubility than the 300 syllables per hour typical of young hearing children.[29,39] These early babbling behaviours in children with hearing losses occur with less consistency and with canonical babbling comprising less than 20% of total vocalisations per session.[21,36] Vocal output differs with respect to the regularity of syllable timing expected in canonical babbling.[29,36] Rather than maintaining a rhythmic speech-like cadence, infants with hearing losses often employ prolongation of sounds with rapid rises and falls in amplitude creating irregular, inappropriately timed vocal patterns.[21,29,36] These early babbling patterns parallel the vocal play and marginal syllables produced by typical, hearing infants at 4–6 months chronological age.

The types of consonants and vowels typically used by children with severe to profound hearing losses are often limited compared with their hearing contemporaries.[29] Because children with profound hearing losses cannot access all the sounds and sequences in their language environment, these children tend to rely on other intact feedback systems such as the visual and kinaesthetic systems to provide cues to placement and movement of articulators (such as the tongue and velum). Dependence on these visual and kinaesthetic cues results in inventories composed of consonants with more visible (i.e. /b, m/) and tactile (i.e. /m, n/) cues. Children with severe to profound hearing losses produce vowels that have neutral qualities (i.e. /Λ/) rather than distinct front, back, high or low placements requiring additional less visible movements of the tongue.[40–42] Their consonant and vowel repertoires are considerably limited

compared with those seen in children with lesser degrees of hearing losses, children with normal hearing of the same chronological age and children with normal hearing of the same hearing age.

IMPACT OF TECHNOLOGICAL INTERVENTION ON EARLY COMMUNICATION BEHAVIOURS

The international emergence of institution-initiated detection and identification of hearing losses in very young children has lowered the age of identification, the age of first intervention, the age of first hearing aid use, the age of first cochlear implant use, the age of first FM assistive listening device use and, consequently, the acquisition of the developmental milestones associated with communication acquisition. In particular, the advent of multi-channel cochlear implants for use in individuals with severe to profound hearing losses and the technological advances associated with these devices have impacted intervention goals and strategies; this has afforded young children with hearing losses more viable opportunities for developing communication milestones in a manner paralleling that of typical, normal-hearing children. The development and availability of multi-channel cochlear implant devices have allowed children with severe to profound hearing losses access to consistent auditory input in the speech spectrum at earlier ages. Many children using cochlear implant technology develop oral communication behaviours approaching levels associated with those of typically developing normal-hearing children.[39–48]

In this section, we will explore the early behaviours of children with cochlear implants to highlight how technology interfaces and enhances communication development. One can imagine that the impact of technology would be evident in multiple ways. For example, it is possible that young infants with hearing losses who appear to diverge from normal developmental patterns at the onset of the babbling period would begin to babble when technology providing a consistent auditory input is provided. But is it possible that a constellation of factors, including experience, will determine when and how young children with hearing losses using cochlear implantss will babble? That is, does 'hearing age', the length of time a child has had audition provided by an implant, give a better time marker of development than chronological age? Moreover, will early babbling behaviours influence the development of phonological structure, first words and language complexity? In order to examine these issues, we will first review how cochlear implants work and what communication information they convey to a young developing child with hearing loss (see Figure 22.5).

Briefly, the technology of a cochlear implant electrically stimulates remaining auditory nerve fibres, providing access to sound patterns.[49,50] Externally worn hardware, consisting of a microphone and speech processor, converts speech and other environmental sounds into an electrical code that preserves the frequency, intensity and duration of the input signal. The speech processor divides the input signal into different frequency bands, which are subsequently assigned to the electrodes in the array to maximise the tonotopic (i.e. frequency-specific) organisation of the afferent auditory nerve.[51] Output from the speech processor is sent to the external receiver, which is magnetically coupled across the skull to the internal receiver. The internal receiver sends electrical pulses to an electrode array surgically placed within the basal end of the cochlea. The electrical pulses from the electrodes stimulate remaining, intact spiral ganglion cells or nerve fibres to send information from the auditory periphery to the cortex.[44,45,51]

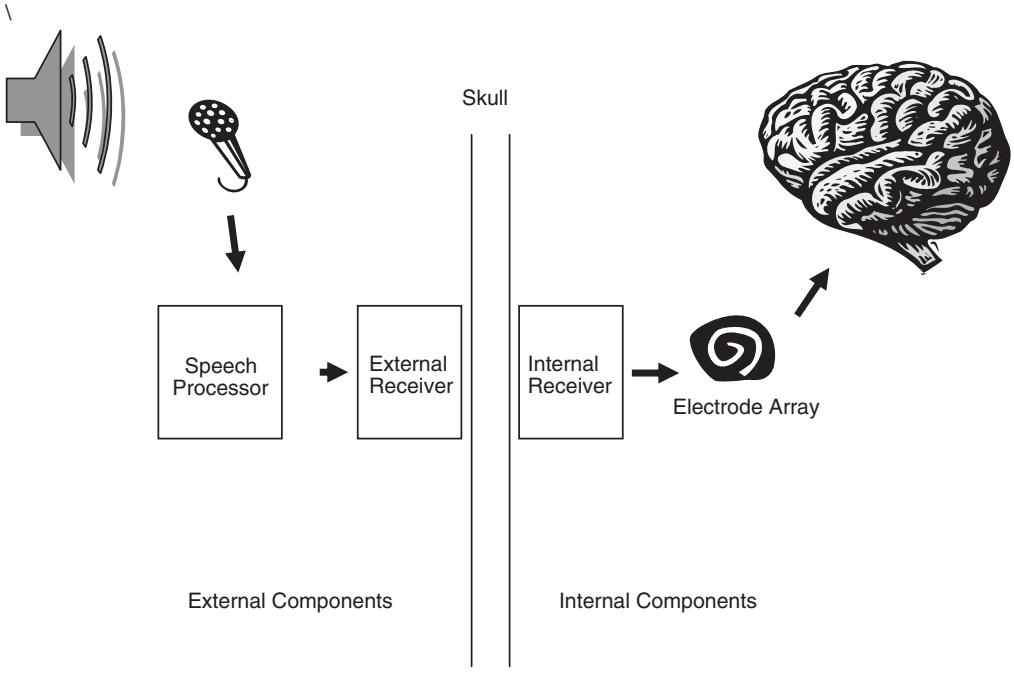

Figure 22.5 All cochlear implant devices have common features. The devices consist of features located external to the skull and features located internally in the middle and inner ear.

Cochlear implant technology allows perceptual retrieval of pitch, loudness and timing aspects of the speech signal. The electrical coding and perceptual retrieval of the speech signals allow young children who use cochlear implants to integrate audition into their communication system, to develop more intelligible speech and more sophisticated language structures than previously observed in hearing-impaired children using conventional hearing aids. For example, infants with severe to profound hearing losses fail to enter the babbling stage in a similar time frame to typical, normal-hearing infants.[21,36] In contrast, children who receive cochlear implants before age 2 years demonstrate more promising patterns of early vocal development. Moore and Bass-Ringdahl[46] found the average age of onset of canonical babbling was 6.5 months post-activation of the cochlear implant in twelve children implanted between ages 18 and 20 months. It is striking to note that the 6.5-month onset time parallels the 6- to 10-month average time frame for the onset of canonical babbling in typical, hearing infants. This finding is consistent with the concept of maturational 'thawing' of a sensory, perceptual or motor system, where the onset of deafness 'freezes' development of the auditory cortex at a particular maturational state.[52–55] Once auditory stimulation is received, maturation proceeds at a rate consistent with typical development.

Application of a 'thawing' metaphor, however, may not serve as the best description of speech acquisition in children implanted under the age of 18 months. Canonical babbling appears in some infants at a 'hearing age' of 1.6 to 4 months after activation. Thus, some children implanted very early may acquire canonical babbling behaviours at younger 'hearing ages', reflecting a shorter time period of sensory experience to achieve babbling

behaviours at similar chronological ages as typical, hearing children.[56] The onset of canonical babbling marks an important milestone in communication, but time of babbling onset alone does not capture the entire picture for hearing-impaired children. When young infants with hearing losses babble – even those infants who receive cochlear implants – vocal patterns often deviate from hearing infants. These differences are evident in a restricted formant frequency range, limited phonetic inventories for consonants and vowels, longer segmental durations and fewer syllable-based vocalisations meeting the timing requirements for speech.

IMPACT OF HEARING LOSSES ON EARLY ACQUISITION AND USE OF SPEECH SOUNDS

Children with profound hearing loss often exhibit different consonant and vowel inventories in early vocalisations compared with hearing infants[4,24,37,39,41,42,57–59] and compared with children having lesser degrees of hearing loss.[4,60] Data suggest the variety of segments used by children with severe to profound sensorineural hearing losses include fewer vowels and consonants than children with lesser degrees of hearing loss[4,60] (see Figures 22.6 and 22.7). Prior to implantation, repertoires include labial, nasal and stop consonants as well as mid and central vowels.[47,61–63] Auditory access to speech via sophisticated technology sparks expansion of phonetic repertoires.[60] Paediatric cochlear implant recipients implanted before age 5 years show use of consonant repertoires diversified to include coronal and dorsal place as well as fricative, glide and liquid manner after 1 year of cochlear implant listening experience.[61,62] Vowel inventories expanded from primarily neutral vowels towards increased use of front vowels and all three vowel heights 1 year post-implant.[61,62]

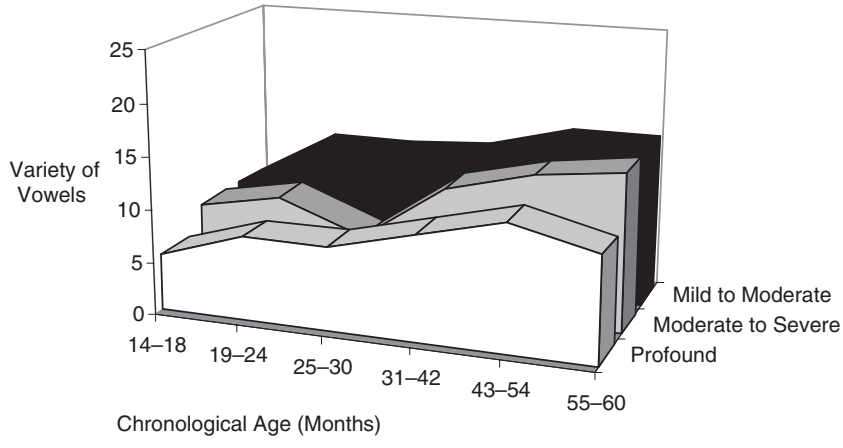

Figure 22.6 The number of different vowels produced by children of various ages with varying degrees of hearing loss is depicted. A wider variety of vowels are produced by older children than by younger children, regardless of the degree of hearing loss. Fewer vowels are produced by children with profound losses than by children with less restrictive losses. Figure adapted from: Yoshinaga-Itano C., Coulter D., Thomson V. The Colorado Newborn Hearing Screening Project: Effects on speech and language development for children with hearing loss. J Perinatol 2000; 20: S132–S137.

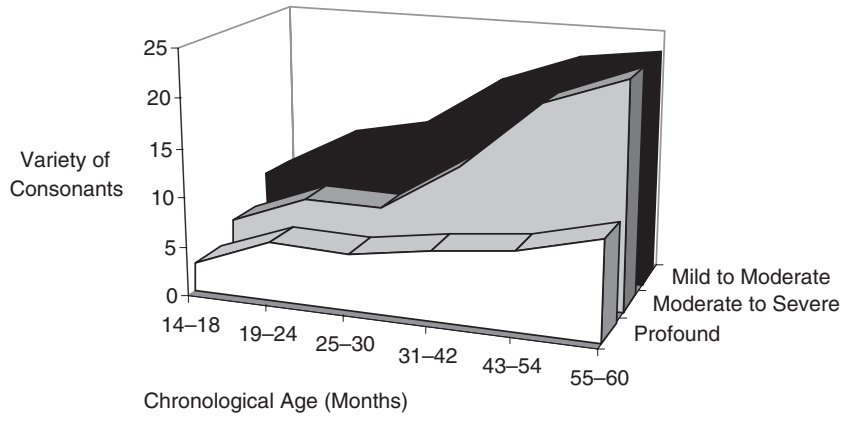

Figure 22.7 The number of different consonants produced by children of various ages with varying degrees of hearing loss is depicted. A greater number of different consonants are produced by older children than by younger children, regardless of the degree of hearing loss. Data further suggest that children with profound losses produce fewer consonants than children with less restrictive losses. Figure adapted from: Yoshinaga-Itano C., Coulter D., Thomson V. The Colorado Newborn Hearing Screening Project: effects on speech and language development for children with hearing loss. J Perinatol 2000; 20(8 Pt 2): S132–S137.

Children who receive their devices earlier exhibit similar pre-implant vocalisation patterns, but differences in the rate at which they expand their segmental repertoires.[39–41] Children implanted before age 2 years produced high, front and central vowels within 6 months of implant activation. Labial, nasal and glide consonants dominate early inventories, but stops, fricatives and liquids emerge after 6 months of auditory experience via the cochlear implant. Children implanted younger than 2 years of age exhibit expansion patterns for both consonants and vowels that are faster than later-implanted children and more consistent with those of hearing children.[37,40,41,43,58,59]

One major difference between children with normal hearing and those who receive cochlear implants is the proportion of nasal consonants in their repertoires. Children with cochlear implants show, across studies, dramatically decreased nasals with concurrently increasing oral production as they gain auditory experience with the cochlear implant device. The prevalence of nasal consonants in children with hearing loss differs from typical vocal patterns for hearing infants and may relate to decreased visual access to the tongue and velum position as well as restricted acoustic cues for place and nasality. Without acoustic information to signal change from the velum's resting position, infants with hearing loss do not learn to consistently elevate the velum to close the velopharyngeal port for oral productions.[10,24,39,57,64] Another proposal related to the nasal production mode in children who are deaf or hard of hearing has been tactile stimulation in an attempt to create feedback through a non-auditory modality.[24,37,64]

These studies demonstrate that children implanted early, particularly those who receive their devices before age 2 years, expand their vowel and consonant repertoires to mimic developmental patterns in hearing children. However, the developmental processes of normal-hearing children and young cochlear implant recipients may not be exactly the same. Comparison of segmental inventories of cochlear implant recipients with those of hearing children may seem logical with respect to the starting point (i.e. predominance of labials) and end point (i.e.

expanded phonetic repertoires). However, examination of phonetic inventories via transcription analysis focusses more on the *product* rather than the *process* of speech development, which may not accurately represent speech acquisition in this population.

Researchers have reported that children with hearing loss – particularly those identified later, those implanted later and those with profound hearing losses – can develop maladaptive speech production strategies like ingressive labial stops[65] and non-native contrasts[45] based on the relative contributions of the visual, kinaesthetic and auditory systems. That is, these children may produce the percept of a /b/, but the technique used to articulate the phoneme may not match that of hearing children because of exaggerated or inaccurate kinaesthetic feedback or inappropriate timing. Consequently, the assumption of identical developmental processes may not accurately describe early speech acquisition in hearing and hearing-impaired children because these two groups might differ not only in auditory levels, but also in levels of physical and cognitive maturation and dependence on kinaesthetic cues and movement patterns.

Box 22.1 Notation used for referring to the 'age' of children with sensorineural hearing losses.

Chronological age refers to the time (years and months) a person has lived since birth. In children with sensorineural hearing losses, this metric allows comparison with children with equivalent maturational status of basic systems such as motor development, vision etc.

Age at onset of hearing loss characterises the chronological age (years and months) when the hearing deficit occurred. The age at onset of hearing loss may be identified by a specific age (e.g. 6 months) or by categorical notations: congenital (hearing loss present at birth) versus acquired (hearing loss occurring after birth) or pre-lingual (hearing loss acquired before language acquisition) versus post-lingual (hearing loss acquired after language is learned).

Age at identification of hearing loss represents the chronological age (years and months) when the hearing loss was diagnosed.

Age of amplification refers to a child's chronological age at the time hearing aids are first fitted.

Age at cochlear implantation refers to a child's chronological age at the time of surgery or denotes a child's chronological age when the cochlear implant was activated, the initial hook-up. Depending on the operational definitions assigned to this term, the age at cochlear implantation may vary by 3–6 weeks.

Hearing age denotes the length of time (years and months) since auditory intervention. When the child wears a hearing aid, hearing age refers to the length of time since the hearing aid fitting. When a child first wears a hearing aid and later receives a cochlear implant, hearing age refers to *either* the time since the hearing aid fitting *or* the amount of auditory experience with the cochlear implant.

Duration of deafness describes the difference in time (years and months) between onset of sensorineural hearing loss and when assistive hearing intervention occurs.

Current research suggests that some experience with audition is needed to achieve babbling milestones following cochlear implantation. However, the length of this period is unclear because neither hearing age nor chronological age alone can predict when babbling or other early speech behaviours will emerge in young cochlear implant users (see Box 22.1). There appears to be an interaction between chronological age and hearing age such that children implanted earlier (i.e. before age 5 years) outperform later-implanted children in expansion of phonetic inventory. The older children may perform better than the younger group in the pre-implant measures and the early post-implant measures due to the advantage of previous speech–language therapy and practice. The younger group, though, seem to excel at a faster rate such that they outperform the older group between 2 and 5 years post-implantation.

The interaction between age at implantation and cochlear implant experience may reflect the neural plasticity within the auditory system. The auditory sensory mechanism is functional at birth and ready to establish neural connections based on auditory experience.[66] Maturation progresses from the periphery to the central auditory nervous system during the first 6 months of life. Interestingly, this 6-month time period coincides with the finding that children with hearing loss who receive intervention prior to age 6 months outperform their peers with hearing loss on language measures. Previous research has demonstrated that normal auditory brainstem development continues through the first 3 years of life whilst the cortical auditory system can change until puberty, although maximum plasticity of the auditory system occurs during early sensitive periods.[66] Because the introduction of sensory input after auditory deprivation creates system reorganisation of inputs, the changes may occur more quickly for the younger group than the older group during the early sensitive periods of neural development. Earlier access to conversational speech via the cochlear implant may spark broader dendritic growth, which could accelerate the integration of audition in the younger children's multimodal speech perception and, ultimately, speech production.

Expansion of consonant and vowel inventories is central to the development of a kinaesthetic and auditory sense of sound production. Early-implanted children often reach canonical babbling milestones by 3–4 months and word onset between 6 and 10 months post-implant activation. The goal of cochlear implantation is the acquisition of intelligible oral communication. This means that the child must learn not only to produce a diverse set of consonants and vowels, but also to produce them correctly in the appropriate order with respect to a word target. We call this phonetic accuracy.

As in hearing children, young cochlear implant recipients require production experience prior to achieving phonetic accuracy.[42,43,67–69] Early segmental forms consist mainly of labial consonants and central vowels. Not surprisingly, these phonetic categories represent the most accurately produced phonetic segments in early words in young cochlear implant users.[42,63,68–71] As production inventories expand, accuracy increases for other consonant and vowel types. Early-implanted children are most accurate for nasal, stop and glide consonant manners and labial place during the first word period, with accuracy scores ranging from 55 to 75%. Consonant types produced with less frequency – fricatives, coronals and dorsals – are produced with far less accuracy during the same time frame. Word position also influences accuracy patterns, with greater accuracy in initial position than in medial and final positions, similar to the pattern in hearing infants.[33] Vowel accuracy is best for front, central and low vowels, although accuracy scores for mid and high vowels approach 60%. Overall, vowel accuracy exceeds consonant accuracy.[40,41,68] This may correspond to the acoustic highlighting of vowels over consonants as a result of increased intensity, prolonged production (e.g. 300 versus 100 ms), and the steady state quality of vowels.[72]

These studies have found more correct productions for visible over less visible consonants and for the most frequently produced consonants and vowels, suggesting that intact feedback systems and production experience guide early phonetic accuracy. Error patterns have received less attention, but findings suggest a tendency to omit vocalic segments,[48,73] replace them with a neutral vowel or change the vocal quality of the vowel through prolongation or nasalisation.[74] Consonant inaccuracies primarily include omissions or errors of voicing, stopping and cluster reduction.[74] The finding that phonetic segments emerge in a child's inventory prior to the attachment of a sound–meaning relationship in early words occurs for both consonants and vowels and mirrors patterns seen in hearing children at the same vocal development age.[67] Thus, it appears that many hearing-impaired children display communication behaviours resembling those of typical, normal-hearing children.

IMPACT OF TECHNOLOGY ON LANGUAGE DEVELOPMENT AND ACHIEVEMENT

The changing landscape of technology and the rising expectations of communication capacities in children with hearing losse impact many levels of communication, including speech recognition, speech perception, early vocalisation behaviours, sound patterns and language acquisition. Early vocalisation behaviours form the underpinning for the expression of more complex oral language constructs, including the building of a lexicon, development of grammatical skills, expansion of discourse abilities and acquisition of literacy knowledge. Institution-initiated identification and intervention programmes are designed to ensure children receive assistance during the formative years when early language constructs are formed. Technological advances in hearing aids, FM systems and cochlear implants continue to improve the quality and integrity of speech signals presented to impoverished auditory systems by scaffolding the processes underlying the emergence of language. In the preceding sections we reviewed early language behaviours associated with infants and young toddlers who experience hearing losses, but difficulties with many aspects of language development are influenced by hearing loss.

Language equips a child with skills to interact in a bidirectional fashion as a receiver and sender of communication intent, as shown in Figure 22.8. Persons with hearing losses may fail to understand a message, resulting in a communication breakdown. Communication breakdowns also may occur when the hearing-impaired person acts as a speaker and is unintelligible to another listener. Language is important for both receiving and conveying the information in messages. The intent of a message may be conveyed in multiple fashions, e.g. speaking, gesturing or writing. The intent of a message may be received in multiple fashions, e.g. listening, watching or reading.

The use of language thus carries profound implications for a child with hearing loss with regard to cognitive growth, emotional and behavioural adjustment, scholastic performance and later vocational options. Access to acoustic–phonetic patterns of speech forms the perceptual foundation for normal linguistic development, initially by enabling phonological representations in the first year of life and, subsequently, facilitating phonological representations of words. The representation of speech sounds serves as an integration point for receptive and expressive vocabulary. Children with early auditory deprivation are at significant disadvantages in establishing the early precursors of oral language reflected in vocabulary, morphology and syntax.

Figure 22.8 Language equips a child with the skills necessary to interact in a bidirectional fashion as a listener and a speaker. Persons with hearing losses may fail to understand a message resulting in a communication breakdown. Communication breakdowns also may occur when the hearing-impaired person acts as a speaker and is unintelligible to another listener. Language is important for both receiving and conveying the information in messages. The intent of a message may be conveyed in multiple fashions, e.g. speaking, gesturing or writing. The intent of a message may be received in multiple fashions, e.g. listening, watching or reading.

One of the major consequences of profound hearing loss on oral language skills in children is a reduction in overall speech intelligibility. Speech intelligibility appears related to a variety of linguistic and extra-linguistic characteristics.[75] For example, Weston and Shriberg[76] report speech intelligibility is associated with utterance length, word position, intelligibility of adjacent words, phonological complexity, grammatical form and syllabic structure. Tobey et al.,[63] building on an earlier model of speech intelligibility proposed by Kent,[9] suggest that a child becomes a good oral communicator by relying on oral speech, demonstrating few communication breakdowns that require repair or clarification, communicatively appropriate use of language in conveying social cues and high speech intelligibility (see Figure 22.9). Individuals who use speech that is clear and easily understood by a listener experience few communication breakdowns. These communication features scaffold the language processes necessary for a child with a sensorineural hearing loss to communicate by relying on auditory-based versus visually based communication. Children who receive cochlear implants after a period of auditory deprivation ranging from 36 to 44 months demonstrate expanded phonetic repertoires and improvement in the accuracy of their articulation.[47,63,73,77–79]

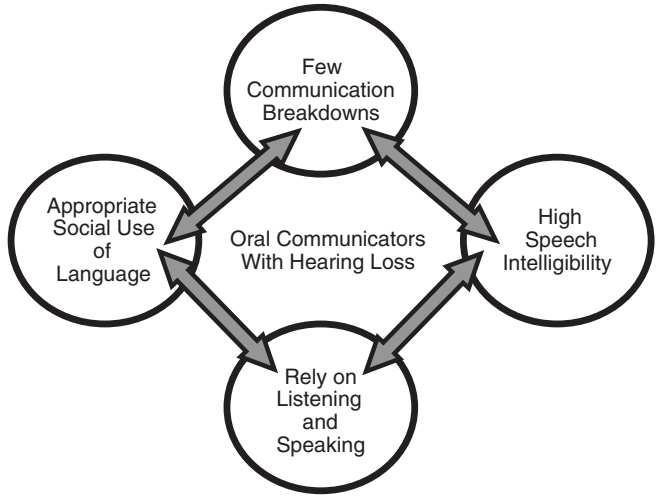

Figure 22.9 Persons who rely on listening and speaking for communication achieve high levels of performance when they demonstrate an appropriate use and understanding of language by relying heavily on listening and speaking rather than on more visually based communication systems. Individuals who use speech that is clear and easily understood by a listener experience few communication breakdowns. These communication features scaffold the language processes necessary for a child with a sensorineural hearing loss to communicate by relying on auditory-based versus visually based communication.

Speech production in deaf children acts as a significant independent predictor of English language competence, even when both speech and sign language are considered together.[80,81] Accuracy of speech sound production is positively related to reading levels in children using hearing aids[82] and cochlear implants.[81] Several researchers suggest access to the phonological form of printed words is related to deaf individuals' speech intelligibility.[82–84] Speaking rate also is positively related to speech intelligibility, with more intelligible talkers completing sentences in shorter durations.[63] Reading scores are positively correlated with intelligibility and negatively correlated with speaking rate in 8- to 9-year-old children who use cochlear implants.[81]

Over the past decade, the average age of cochlear implantation for deaf children has been steadily decreasing.[85] Language growth rates of children who receive cochlear implants before age 5 years resemble growth rates of typical, normal-hearing children.[78,86,87] Spoken language performance in children with cochlear implants demonstrates an increasing spoken language advantage with each month of increasing auditory experience with a cochlear implant, such that children implanted between ages 12 and 18 months achieved language quotient scores comparable to normal-hearing peers by 4.5 years.[88] Increased experience listening with cochlear implants results in steady acquisition of different root vocabulary with greater strides in vocabulary acquisition occurring in children implanted at younger ages. Over a broad age range, children who receive cochlear implants at younger ages develop more intelligible speech.[75]

Studies examining early language development generally report that children with cochlear implants display a developmental gap relative to hearing age-mates. However, if children receive a cochlear implant by age 2 or 3 years, they start developing language at a near-normal

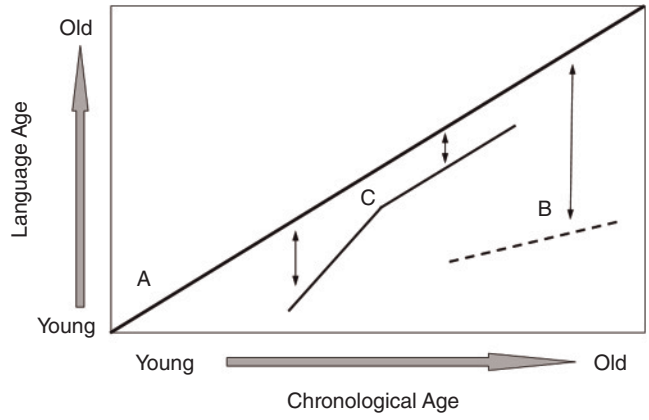

Figure 22.10 This schematic illustrates many of the principles underlying investigations and findings in studies of language acquisition in children with sensorineural hearing losses. Chronological age is located on the abscissa, ranging from young to older ages. Language ages, ranging from young to older ages, are shown on the ordinate. Parts of the diagram are identified as follows: A, idealised trajectory of language performance for a typical, normal-hearing child who acquires language at a rate commensurate with chronological age; dashed line, language acquisition trajectory for a child with a hearing loss who is identified and fitted with hearing aids at older chronological ages; B, The arrows represent gaps between the language performance of normal-hearing and hearing-impaired children; C, language acquisition trajectory for a child with earlier identification and intervention.

rate and the developmental gap no longer increases with age.[86] Figure 22.10 diagrams language acquisition in children with hearing loss who use an auditory prosthesis such as a hearing aid or cochlear implant. Chronological age is shown on the abscissa, ranging from young to older ages. Language ages, ranging from young to older ages, are shown on the ordinate. The solid diagonal line labelled 'A' represents the idealised trajectory of language performance for a typical, normal-hearing child who acquires language at a rate commensurate with their chronological age. The dashed line indicates the language acquisition trajectory for a child with a hearing loss who is identified and fitted with hearing aids at older chronological ages. Children identified late display poor language skills relative to their normal-hearing peers of comparable chronological ages and acquire additional language skills at a slower rate. The slower rate of acquisition results in an increasing 'gap' between the language performance of normal hearing and hearing-impaired children, as indicated by 'B'. Earlier identification and intervention, as represented by 'C', also may result in children with lower language performance relative to their normal-hearing peers; however, these children may show an accelerated acquisition immediately following intervention, followed by a rate of acquisition similar to their hearing mates. Note, however, the rate of language acquisition may parallel normal-hearing children but still remain delayed or disordered, as indicated by the continuation of a gap in performance.

Language growth is more rapid in children implanted as infants than in children implanted as toddlers.[89] Early implantation may be associated with improved prognosis for literacy development,[90,91] but a large sample of long-term cochlear implant users at ages 8 and 9 years found no advantage associated with earlier implantation for speech perception,[92] speech production,[63] language[81] or reading performance.[93]

Speech communication conveys lexical and indexical information. Lexical information represents the word sequence from a speech signal. Indexical information provides information

such as talker identity, gender, age, emotional state and regional dialect. Adult cochlear implant users are minimally competent at gender discrimination,[94,95] talker discrimination[94] and emotion identification. Poor performance in talker discrimination[96] and tone identification tasks[97,98] is found in young cochlear implant users. Perception of lexical and indexical information does not occur independently.[99,100]

Children with severe to profound hearing losses plateau in reading skills, reading when completing high school at a 4th-grade level.[101] Several studies report higher reading performance from groups of children who used cochlear implants than from groups of students with profound deafness who used hearing aids. Spencer, Tomblin and Gantz[90] report that 45% of cochlear-implanted children enrolled in total communication programmes read within 8 months of their mainstream grade level. Spencer, Barker and Tomblin[91] note cochlear implant users performed within one standard deviation of normal-hearing, age-matched children on measures of language comprehension, reading comprehension and writing accuracy. Moog[102] reported that 70% of cochlear-implanted children enrolled in an oral communication setting scored within the average range for their hearing age-mates on standardised reading tests.

There is considerable debate about how deaf students access written materials. Children learning a sign-based language may use a top-down model of reading comprehension.[103] In this case, deaf children process print on the basis of semantic cues that depend on vocabulary knowledge and their ability to bring sufficient world knowledge to the task. For children learning a speech-based language, bottom-up strategies may be important. Bottom-up models emphasise the development of sub-lexical processing, where readers use phonological decoding strategies to translate the printed text into previously acquired acoustic units. Bottom-up strategies rely on the child's ability to use letter–sound generalisations to decode words. Most normal-hearing children are competent language users when they begin to map reading onto existing phonological, syntactic and semantic skills. The deaf child brings very different sets of language experiences to the reading task. Children who have impaired hearing demonstrate average delays of 4–5 years in language development by the time they enter high school.[42]

Vocabulary plays an important role in reading acquisition in hearing children. Poor readers with normal hearing are characterised by deficits in vocabulary. Deaf children approach reading with a more limited vocabulary than their hearing age-mates,[70] particularly for words with multiple meanings. Therefore, lexical access is restricted, even if decoding is successful. A limited vocabulary may further affect reading by tying up processing capacity at the expense of higher level syntactic and text comprehension abilities. Syntactic or grammatical knowledge underlies the acquisition of mature literacy. Poor syntax is associated with reading delays in typical, normal-hearing children.[82] Grammatical competence in kindergarten is highly predictive of literacy levels attained by typical, normal-hearing children in 2nd and 4th grades.[104] Individuals with pre-lingual deafness typically display poor syntax in expressive language and exhibit greater difficulty in recognising grammatical errors than their hearing age. Deaf students use syntactic information poorly when making inferences about words, presumably because they are unable to use context for sentence comprehension. Deaf readers are unable to capitalise fully on their vocabulary knowledge if they have poor grammar.

WHERE WILL A CHANGING LANDSCAPE OF SERVICE DELIVERY TAKE US?

Changing the service delivery landscape from parent-initiated to institution-initiated may assist in identifying children with hearing losses at earlier ages. Earlier identification leads to earlier

intervention opportunities that may include advancing technologies, such as cochlear implants or digital hearing aids. Technological interventions of these types provide great potential for allowing children to experience auditory information during the formative years of language development. The advances associated with such technologies will change the fabric of service delivery for hearing-impaired children worldwide. Clinical and parental expectations will change, reflecting the evolving landscape. Perhaps the following parental impressions will characterise the impressions of many parents in the future when welcoming a hearing-impaired child into their home and family:

> At birth, Emma passed her OAE newborn hearing screen before we were discharged from the hospital. She seemed like any other newborn responding to our voice and sounds around her. As a few months went by, we noticed that Emma was not developing speech. In November 2003, Emma was referred for an Auditory Brainstem Evoked Response Test. We were shocked to learn that our daughter had a bilateral severe to profound sensorineural hearing loss. We immediately questioned the results and accuracy of the test. This led to a second objective opinion that arrived at the same exact conclusions. Even then, we had a hard time accepting the results. We searched for anyway possible to prove that the tests may be inaccurate.
>
> Our first thought about cochlear implantation was that we would absolutely not put our child through the risks of elective surgery. We accepted Emma as she was and could not fathom the idea of putting her through surgery at such a young age. But after a few months of in-depth research on the benefits of a cochlear implant, we realized that the risks were low when compared to other pediatric surgeries and the rewards were infinite. . . . The decision to go with a cochlear implant was not based so much on us wanting our daughter to hear, as it was based on wanting to give Emma a choice; a choice of embracing both hearing and non-hearing worlds. We wanted to give Emma a chance at all options and take maximum advantage of those options. The cochlear implant has maximized her options.
>
> Emma says something or learns something new everyday! At first, we counted the words that she was learning. Now, we can not keep up with the number of words and phrases she is learning! Less than a year ago, Emma could not say a word. Today, we hear Emma in the other room yelling 'mama!' or 'dada!' or 'no!' (at her brother). The only problem we have encountered is Emma has learned to answer the phone when she hears it ring and she is learning to talk back (like a typical two-year-old). Emma's receptive language (understanding speech) started out being better than her expressive language (talking). However, that gap seems to be closing. We can now call Emma from another room in the house and she will come running. We can tell her to wash her hands or go get her shoes and she will do it (without any visual cues). Emma has exceeded all expectations. The only time Emma can not hear us is when she is swimming, taking a bath, or sleeping. During those times we have to remind ourselves that we must rely on some visual cues.
>
> (Excerpt from Testimonials contained on the Dallas Cochlear Implant Program website: http://www.DCIP.org.)

Language skills such as literacy, lexicon and grammatical skills, emerge from the interface of multiple elements, including but not limited to parental interactions, technology sophistication and parental expectations. The potential for more advanced communication capacities for children with hearing loss has changed with the implementation of effective newborn hearing screening and intervention programmes. Earlier identification of hearing loss affords earlier interventions, which may be more effective in taking advantage of natural learning processes and the neural plasticity of the auditory system as an individual and integrative unit within the child. Our expectations for this population will continue to transform with the advances afforded by new technology, the resulting changes in communication development and the subsequent effect on family dynamics. As institution-initiated detection and identification of

hearing loss gains acceptance, clinicians and families will need to anticipate advances in oral language that have not been previously seen in children with hearing losses. The challenges are exciting and suggest the need for the development of new interaction and intervention approaches, to enhance the strides made in communication development for children identified through early identification programmes. Future studies will need to explore how the changing service delivery landscape impacts sophisticated language applications associated with reading, spelling and writing; acquisition of multiple languages; academic performance; family adjustment and dynamics; and social interactions in early identified children with sensorineural hearing losses.

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23 Deaf children and communication approaches

L. Watson

INTRODUCTION

Communication is a dynamic process, involving meaningful exchanges between two (or more) individuals. How best to facilitate this for children who are born deaf or who acquire deafness early in life is a topic that continues to generate lively debate in many countries. The situation in the UK is no exception and can serve as an example of the nature and scope of the debate, with the pendulum of opinion swinging between the use of British Sign Language (BSL) as the child's first or primary language at one extremity and the use of spoken language exclusively at the other, with many different positions to be met along its trajectory.

In the 1960s in the UK, oralism was the approach used almost exclusively in educational provision for deaf children, although there was not simply one oral approach, and there was discussion between advocates of different oral methods.[1] The Lewis Report[2] was set up to consider the 'place, if any, of fingerspelling and signing in the education of the deaf' but did not seek to impose the use of either. However, gradually during the 1970s the exclusive use of spoken language began to be challenged, fuelled in part by the work of Conrad,[3] whose report that almost 75% of his sample of deaf children had unintelligible speech, and concern about the low levels of literacy of deaf pupils raised questions about the effectiveness of the oral approach. During the 1970s and 1980s, some educational establishments for deaf children in the UK introduced the use of signs alongside speech. These were signs for key words, which were used in conjunction with speech (referred to as Total Communication (TC) in the UK or as Manually Coded English (MCE) or Simultaneous Communication (Sim Com) in the US). A more recent change has been the introduction of sign bilingualism, which comprises the use of BSL and written/spoken English, an approach supported by the recognition of BSL as a language in 2003. A second relatively recent change has been in the nature of oralism, with the introduction of different oral approaches, building on greater understanding of how children learn language coupled with a new generation of hearing aids (including cochlear implants).

The current situation in the UK is both complex and fluid, and the debate is multifaceted. It has gone beyond a discussion of whether it is more desirable for deaf children to learn the (spoken) language of the wider, hearing community in which they live or the signed language of the Deaf* community, to encompass debate around the concept of a critical or sensitive period for language acquisition; the social and emotional development of deaf children; their sense of identity; and questions of choice, both parental choice made on behalf of their child and the child's own voice, or right to choose for themselves. The reality of the situation as it

*The use of the capital D indicates the community of deaf people who use BSL as their language and identify with other deaf people who share their language, culture and history.

is lived by deaf children and their families is complicated and diverse. It is not static and is subject to variation in accordance with deaf children's changing communication needs and preferences.

In this chapter, I will first consider three communication approaches individually: oral approaches, TC and sign bilingualism. I will provide an explanation of each of these as separate communication approaches in order for the reader to gain an understanding of them. I will explain how each approach is currently defined and used, in order to give a clearer picture of the current situation. I will continue the chapter with a consideration of possible changes in the future, in the light of the introduction of the newborn hearing-screening programme (NHSP) in the UK, advances in technology and the implications of cochlear implantation. I will conclude with a consideration of the question of choice and some implications will be drawn and speculation offered for the future.

COMMUNICATION APPROACHES

Oral approaches

There are different oral approaches to communication used in the UK. They vary in the relative emphasis that they give to audition or vision, the extent to which they follow a structured language programme and their views on where deaf children should be educated.[4] All recognise the need for professionals and parents to work closely together to promote the deaf child's language development, and the goal for all oral approaches is for deaf children to acquire spoken language. Since the majority of deaf children are born to hearing parents (Mitchell and Karchmer[5] suggest a figure of 96%), advocates of oral approaches regard themselves as assisting deaf children to communicate both with their family, using the spoken language of the family, and with the hearing world.

Natural auralism

Natural auralism is the oral approach most widely used. The approach was given its name following the formation of the National Aural Group in the early 1980s, when a number of Teachers of the Deaf and lecturers involved in the training of Teachers of the Deaf got together to discuss what they saw as a new and distinct approach to oralism.[6] This group of professionals realised that they had moved away from using structured language teaching approaches, for example Guidelines[7] in favour of seeking to assist deaf children to develop spoken language in a similar way to hearing children by engaging them in meaningful interactions as hearing parents do with young hearing children. They were exploiting the use of aided hearing, afforded by advances in hearing aid technology, including radio aids, and encouraging deaf children to utilise aided hearing rather than relying heavily on lip-reading. Initially, they used the term *natural oralism*,[8] which emphasised the fact that the approach focused on the development of language following the usual pattern for hearing children (thus *natural*), later they adopted the term *natural auralism*, which was felt to encompass the two facets of emphasis on encouraging language development following the pattern of hearing children and the reliance on the use of aided hearing. The Natural Aural Group (NAG) was formed to promote the use of this approach. The group later changed its name to Deaf Education through Listening and Talking (DELTA) as this was considered to reflect more clearly the distinctiveness of the approach. Further information is available on their website.[9]

In keeping with the emphasis on the use of hearing, deaf children following a natural aural approach are 'thought of and treated as children who can hear'. Whilst not denying that the children are deaf, proponents of the approach consider that it is important that they are expected to be able to gain information via aided hearing. Accommodations will be made for the fact that they are listening via hearing aids or a cochlear implant. Thus, background noise will be kept to a minimum, for example by attending to the acoustic environment of the home and not allowing other sounds such as the television to compete with conversation. Parents will be encouraged to speak to their deaf child within the range of the hearing aids and to ensure that the aids are in optimal working order and worn consistently throughout the day. They will be expected to gain the child's attention auditorily, by calling the child's name and anticipating that they will respond. They will be discouraged from using visual or tactile means of attention-getting (e.g. tapping the child or waving to attract their attention). They will bring sounds in the home and environment to the child's notice and anticipate that the child will learn to hear them, but are unlikely to introduce visual or tactile aids such as a flashing light to denote that the doorbell is ringing. The deaf child's responses to sound will be monitored very closely and there will be an expectation that the child will learn to respond to a greater range of sounds, and at lower intensity as they develop their listening skills. If this is not the case, then the reasons will be sought in the hearing aids or implant, which will be checked and adjusted if necessary, in the acoustic environment that the child experiences and in the opportunities that the child is given for developing their listening skills. Lip-reading will not be encouraged or fostered, but neither will it be denied. There is recognition that some deaf children gain information through vision and that this supports their listening, but it is noted that many deaf children using a natural aural approach do not rely on lip-reading, except in poor acoustic conditions. Providing an appropriate listening environment can be a challenge for some parents, as can the necessity to ensure that their child's hearing aids or cochlear implant are maintained, but professionals working with these parents provide help and support.

The introduction of the newborn hearing-screening programme in the UK means that there is a possibility to fit hearing aids and to refer the child for assessment for a cochlear implant at a very young age. Parents of deaf children and professionals, aware of research on neural plasticity, can feel that very little time is 'lost' and that early diagnosis can be extremely beneficial in assisting the child's listening development and hence his or her development of spoken language through hearing.[10]

Spoken language development is promoted by interaction. Deaf children are not expected to say anything until they are ready to do so, but all their contributions (vocal or gestural) are accepted and regarded as having meaningful intent. Just as hearing babies spend some months assimilating sounds and speech prior to beginning to talk themselves, deaf children are given the same space to learn to listen and receive spoken language. Adults are encouraged to talk to them using lively intonation and short, but meaningful, utterances. However, adults are discouraged from slowing down their rate of utterance or exaggerating their lip patterns as these are considered to make it more difficult for deaf children to understand. (For further discussion of the complexities of interaction with deaf children see Gallaway and Richards.[11]) The emphasis, and key to success within this approach, is always on meaningful exchanges of spoken conversation between adult (or more mature language user) and child. The deaf child's utterances are accepted as their contribution to the interaction and they are not corrected, although a more adult model of the articulation or syntax may be given in the adult's reply. There is no expectation that the deaf child will repeat or practise what the adult has modelled. Natural gesture may form part of the exchanges, as it does with hearing children,

but the use of sign (either to accompany speech or as a language) does not form part of the approach. It is anticipated that, having learnt to speak, deaf children will be in a position to make communication choices when they are older. Proponents of the approach therefore consider that it provides deaf children with the opportunity to choose for themselves when they are older, using their knowledge of spoken language to assist them in learning sign language if they wish.

The basic tenets of this approach are therefore maximum use of audition, aided as appropriate by hearing aids or cochlear implants that are maintained to the highest standard, and an emphasis on language development that follows the pattern for hearing children, facilitated by a high level of meaningful interaction between the deaf child and a more experienced language user (adult or child). It can be seen as a holistic approach in which accommodations for the child's deafness are incorporated into the life of the family.

Critics of natural auralism consider that it is a form of 'normalisation' (e.g. Beattie[4]), which denies that the child is deaf and seeks to force deaf children to conform to a hearing model. Since, for some deaf children, the process of learning spoken language is much slower than for hearing children, critics argue that it risks impeding their social and emotional development and retarding their development of concepts and thus their ability to access the curriculum when they start school. However, proponents argue that it affords deaf children the opportunity to access the curriculum in a similar way to hearing children and to communicate with, and thus engage on equal terms with, mainstream hearing society.

Auditory verbal therapy (AVT)

This is an approach that aims to enable deaf children to use technologically assisted hearing to listen, to process verbal language and to speak with the goal that they should attend mainstream education and become independent, participating citizens in mainstream society.[12] It differs from natural auralism in several respects. There are 10 guiding principles of AVT.[13] The main points of these are summarised as follows:

1. There is a need for the child with hearing loss to receive *highly enhanced* auditory and language input.
2. Parents are trained by a qualified auditory verbal therapist to participate in the programme and integrate its practices into daily living.
3. Each session of therapy is diagnostic and leads to highly specific individualised goals for the child and family.
4. There is recognition of the need for the provision and maintenance of the best possible technology to optimise the child's access to sound.
5. Understanding through listening is actively promoted, with no special emphasis on other sensory cues such as lip-reading.

Whilst AVT is not widely available in the UK, certain of its techniques, in particular the practice of *acoustic highlighting*, whereby emphasis is given to certain aspects of a word or phrase, and the *hand cue*, a practice that consists of the adult covering their mouth briefly when the child is looking at them in order to encourage listening rather than lip-reading,[14] are used by some other professionals. This emphasis on listening and restricting access to lip-reading is an inherent part of the approach, which has its roots in the acoupedic approach. Some cochlear implant programmes offer AVT for children who have received a cochlear

implant, and there is also a centre (Auditory Verbal UK). It is probably best described as an intensive programme for parents that emphasises ways to give precedence to listening. It must be delivered by a qualified auditory verbal therapist, who works with parents. Each session is diagnostic and short-term language goals are set that enable parents and therapists to monitor progress. Advocates claim that parents can be trained to incorporate the techniques into their everyday family life and that the highly organised way in which therapists and parents document and evaluate the child's progress ensures careful and systematic monitoring of the child and assists in goal setting. Critics consider that the emphasis on setting and monitoring goals resembles the structure of 'old' oral approaches and dislike some of the techniques, for example the hand cue just mentioned.

There are clear differences between these two oral approaches, although also some similarities. Whereas AVT may be considered too 'structured' for some, natural auralism can be seen by others as laissez-faire. The truth is possibly that with AVT there are many times during the day when the exchanges between parents and children are just as 'natural' as those advocated by natural auralists and that those following a natural aural approach are constantly monitoring the child's understanding and adjusting their input to fit, thus they are highly skilled in regulating their own language and not leaving the child's language development to chance. The use of acoustic highlighting and the hand cue and the setting of specific goals do set AVT apart from natural auralism, as natural auralists would not emphasise any particular word or sound and leave the decision regarding the necessity to lip-read to the child, rather than the adult at times actively restricting access to it. Proponents of natural auralism would monitor progress carefully, using, for example, the Monitoring Protocol for Deaf Children,[15] but would be unlikely to set specific short-term language goals. Another difference lies in the views on educational placement. Whereas deaf children following a natural aural approach may be educated in mainstream or special educational settings, the goal for children using AVT is that they should be educated in a mainstream setting. This may have other consequences, particularly in regard to finding a peer group and developing their identity. If it is accepted that all children need a peer group, then for deaf children their communication approach can be one factor in providing this. Deaf children who use spoken language may find their natural peer group is with hearing children or with other deaf children. Since a natural aural approach is seen as fitting with any educational placement, this can facilitate the development of a peer group at school amongst either hearing or deaf peers. However, within an AVT approach with its emphasis on mainstream school placement and citizenship in mainstream society, providing a deaf peer group would be more of a challenge and might not be considered so important.

Total communication

The second broad approach to be considered is total communication (TC). People use this term differently, so clarification may need to be sought to ensure mutual understanding. Denton[16] used the term in relation to communication with deaf children. This early definition exemplified the ambiguity in use of the term insofar as he initially described a philosophy that included gesture, sign, speech, speech reading, fingerspelling, reading and writing, then went on to say that in practice this meant talking and signing simultaneously. In the UK, when signing was first introduced into educational settings for deaf children (either schools for the deaf or units/resource bases for deaf pupils), the practice was to talk and accompany the spoken message with simultaneous signs for key words. The signs were taken from BSL, but did not include any other BSL features. This approach is known as Sign Supported English (SSE). In some

settings, staff signed every word and feature of English, using invented signs or fingerspelling to represent grammatical features of English, for example word endings such as 'ing' or English words that have no equivalent in BSL such as the definite article. The term for this approach is Signed English (SE). In both these approaches, the emphasis is on supporting the development of spoken English and the use of sign is to facilitate understanding of spoken language. Signed English is not widely used, and when people refer to TC to mean spoken language accompanied by simultaneous use of sign they usually mean SSE. When TC was first introduced, practitioners and parents who used it viewed it positively as it eased communication[17] and it is relatively easy to learn the signs for keywords. It does not necessitate learning BSL as a language with its own grammar and structure, just a list of mostly nouns and verbs. Parents can use the signs they know to accompany speech as they are learning to sign. Some parents and professionals view the use of TC in this way as the easiest way into communication for deaf infants, arguing that it leaves open the options for deaf children to choose whether they prefer spoken language or sign, and they can then follow the child's preference. In the research on communication approach for deaf children who receive cochlear implants discussed later in the chapter, parents reported that they used TC in this way (spoken language with signs for keywords) prior to implantation and in the early post-implant period and then their children discontinued the use of sign as they gained more benefit from their cochlear implant.[18]

However, TC is not without its detractors, who cite several criticisms. The first criticism is that deaf children exposed to SSE do not receive a full language. Since they are being offered only single signs to accompany speech, they are not receiving BSL as a language, only some signs taken from it. There is also a danger that they are not receiving an accurate version of spoken language, with the features that are essential for understanding, such as normal speed of presentation and intonation patterns. Adults who are addressing deaf children using SSE may modify their spoken language in order to accommodate to their own signing ability. Adults who are in the early stages of learning to sign may deliberately phrase their spoken language to match the signs that they know, thus restricting the breadth of spoken language that they use; they may slow down their rate of utterance to allow themselves time to think of the signs, thus making the spoken language more difficult to understand; or they may present spoken language that does not follow the standard grammatical structure of English. Instead of using the normal English subject–verb–object structure, they may begin to use the subject–comment structure associated with BSL. So, for example, they may say 'Your birthday, when is it?' rather than the more common 'When's your birthday?'. This can be done inadvertently, what Watson[19] termed the 'BSL drift', or it can be done deliberately to aid understanding by cueing the child into the topic at the beginning of the sentence.

A second criticism of the use of TC in this way if it is intended to support the development of spoken language is that it presents deaf children with the auditory and visual signals simultaneously. Whereas some would argue that this is beneficial since young babies (hearing or deaf) are presented with visual and auditory symbols simultaneously and they integrate the two, others suggest that in the case of young deaf children, they find it difficult to assimilate both signals simultaneously.[20] This being the case, the argument runs, deaf children are likely to attend more to the visual signal (the signs) that are perhaps easier for them to access and not attend to the auditory signal since this requires them to listen and may not be fully accessible. The situation is made more serious by the fact that the words and word endings that are less acoustically salient and therefore more likely not to be heard (e.g. function words and 'ed' endings) are also words and elements of speech that will not be signed.

Thus, with TC used in this way, deaf children are in danger of not receiving a full version of English. The basic question here, which remains unresolved, is whether the different modes support each other or compete with each other. It used to be the experience of practitioners that once SSE was introduced then deaf children did not stop using it and signing became their preferred mode of communication. However, evidence suggests that there is a shift for children who receive cochlear implants, and it remains to be seen whether the new generation of digital hearing aids also offer an acoustic signal that is so salient that deaf children come to rely more on their hearing than their vision.

A different criticism is levelled by proponents of BSL, namely that TC does not offer deaf children full BSL. Since they are only presented with single signs to accompany speech, deaf children do not receive the linguistic features of BSL that are essential language components. Therefore, TC in terms of its use as SSE is criticised by those who are in favour of oral approaches since it is seen as distracting from the spoken language signal and there is a danger that the English may start to follow BSL word order; it is criticised by those who support BSL since it is not BSL but simply uses single signs taken from BSL. Thus deaf children may not receive an accurate version of either language. However, it must be recognised that there has been little formal research into the use of TC.[21] It is possible that it should not be viewed as negatively as the preceding paragraphs would suggest.

The term TC is used slightly differently in relation to children and young people with learning difficulties, where it means the use of several different communication approaches simultaneously (a TC website[22] lists signs, gesture, body language, facial expression, objects of reference, photographs, drawings, symbols, written words, vocalisation, intonation, verbalisation, access to modern technology). This list is much broader than simply the use of spoken language and sign simultaneously. The same criticism can be levelled with this practice that the onus is on the child or young person to integrate the incoming signals to gain meaning; again some practitioners consider that this is extremely difficult for these children, given that they have learning difficulties.

The other use of the term TC is to refer to the practice of offering each individual deaf pupil the language or communication mode that they prefer or that is deemed to be most appropriate. When the term is used in this way, a local authority service for deaf children or a resource base may describe their communication policy as offering TC and mean that they offer the full range of communication approaches. Thus in one setting, spoken language will be used with some pupils, SSE with others and BSL with yet others. Given the fact that the numbers of deaf pupils who are educated in integrated settings has increased to over 90%, it is no longer viable in many geographical areas to provide separate specialist provision for each approach for those pupils who are not integrated, so an individual specialist setting can be expected to offer the full range of communication approaches and thus be seen as capable of accepting pupils requiring any particular approach. This has the advantage of being able to sustain provision for deaf pupils with viable numbers and provide the flexibility to vary the communication approach used with any individual pupil as their needs change. However, it can make it more difficult for pupils to receive spoken language exclusively, since SSE or BSL will be used with other pupils. Indeed, there are examples of parents who have moved their child away from such a setting as they perceived that it was having an adverse effect on their spoken language development. Similarly, it can be difficult for pupils who are focusing on BSL as their main communication approach since it is not easy to provide BSL all the time in a mixed setting. From the pupils' perspective, it can be a positive experience to be educated with deaf pupils using the full range of communication approaches as this is what they will

experience in the outside world. Alternatively, the pupils may mix solely with other deaf pupils using their chosen communication approach and thus have a very restricted peer group. Since this practice is relatively new, research into both the linguistic and social effects of these mixed settings would be timely.

Sign bilingualism

The third approach to consider is sign bilingualism. This term refers to the use of the sign language of the indigenous Deaf community (e.g. BSL) and the spoken and/or written language of the hearing community (e.g. spoken or written English or Welsh). It was first used in educational settings in the UK in the late 1980s. Its introduction was fuelled by several factors, including:

- Concerns about under-achievement of deaf pupils, who had been educated in mainly oral programmes
- Criticism of the use of TC (in terms of spoken language plus SSE)[23]
- The relative academic success of deaf children from signing deaf families[24]
- The recognition of sign languages as full languages in their own right[25]
- A changed definition of bilingualism away from its traditional use to mean native speaker competence in two languages towards native competence in one language and the ability to use another in certain circumstances[26]
- Evidence from bilingual programmes for hearing minority ethnic language users[27]
- Evidence from the success of bilingual programmes for deaf children in Scandinavia.[28]

The approach as used in the UK was originally defined by Gregory and Pickersgill.[29] Since then, sign bilingual practice has developed and is implemented in different ways, varying with the educational setting. For descriptions and discussion of current bilingual practice, see Swanwick and Gregory.[30] Their definition of a sign bilingual child is one who 'uses two or more languages in their daily life, at least one of which is a sign language' (p. 9). There is an emphasis on the deaf child's development as a deaf individual. Whereas formerly it was seen that sign bilingual deaf children would have BSL as their first language, with more deaf children being educated in integrated environments – and the advantages afforded by cochlear implants or digital hearing aids – some bilingual deaf children may have English as their first or equal language (p. 18), enabling them to move between the deaf and hearing worlds. A key aim of sign bilingualism is for deaf children to acquire age-appropriate language levels in BSL prior to school entry. Whilst this is perfectly possible for deaf children of Deaf parents with native sign language competence, it is much more difficult to achieve for deaf children in hearing families where the parents are in the throes of learning BSL.

Proponents of this approach refer to sign language as the 'natural' language of deaf people, and therefore of deaf children, since they consider it to be the easiest language for deaf children to learn, as it fits with a visual orientation to language. A sign bilingual approach adopts a very positive view of deafness, which is seen in terms of a difference rather than a disability. Deaf people are seen as a minority group with their own language, history and culture. This means that if a deaf child of hearing parents is brought up and educated within a sign bilingual communication approach, then they are likely to become part of the Deaf community, a fact that parents need to recognise when making a decision regarding communication approach. On the birth of a deaf child, some hearing parents will wish to introduce

their child to the Deaf community and expect that their child will find their identity as part of that community, whilst others will expect their child to fit into their hearing family and to find their identity within the hearing community. The reality for many deaf children is actually more complex.

The choice of a sign bilingual approach to communication has implications not only for the deaf child but also for the family, who will need to concentrate on learning sign language and gaining an understanding of Deaf culture. This includes much broader issues than simply sign language, such as strategies for attracting attention, acceptable use of touch and subtler issues like Deaf sense of humour. The family are likely to want to interact with Deaf adults and invite them into their home to share their language and culture with the family. The implications extend beyond the immediate family to embrace the extended family, as, if the deaf child is to be included, then the whole family will not only need to learn to sign but be committed to using it when the deaf child is present, whether they are addressing the deaf child or another hearing member of the family. Research in the past[31] has shown that typically it is the mother who learns to sign best, with other members of the family finding it more of a challenge. The questions of whether to encourage the child to use hearing aids and whether to refer the child for assessment for a cochlear implant will need to be addressed. For many Deaf adults, hearing aids are not regarded as useful.

With regard to education, there will be decisions regarding educational placement. There are also wide variations in the implementation of a sign bilingual communication approach between education settings. Sign language is normally used as the first language for communication by and with young deaf children in this approach. English will usually be introduced and taught as a second or additional spoken language, along with any other spoken language used in the home, although again there is variation. Within school, the important point is that the use of the two languages (BSL and English) is planned. Some subjects may be taught through BSL and others in English, or there may be discussion of the lesson content in BSL, followed by support for written work being offered in English.

Once deaf children have become fluent in BSL, they have acquired a language; then they can apply their knowledge of one language to another language, using their metalinguistic awareness to assist them in understanding the difference between the two languages. Thus, it is stated that the fact that deaf children have learned BSL, rather than interfering with their ability to learn spoken English, will actually enhance it. Proponents argue that this approach enables deaf children to gain a language quickly, since they find BSL accessible, which means that they have a language to use for thinking and developing concepts. It also means, they argue, that if there is a critical period for language development, then deaf children will not miss it. Critics would contend that it is not as straightforward to transfer knowledge of BSL to learning a spoken language since it also involves a change of mode.[32] A second criticism, one that is levelled by those who consider that the easiest way for deaf children to learn spoken language is via aided hearing, is that deaf children who do not use their aided hearing as early as possible will have missed the window of neural plasticity. This argument contends that deaf children are disadvantaged by the delay in introducing spoken language, the effects being most apparent in the intelligibility of their spoken language, since this is most easily learned via audition.

The sign bilingual approach fits with current thinking around bilingual education for hearing children from homes where English is not the first language of the home. Linguistic theorists (e.g. Cummins[33]) argue the advantages of recognising a child's language of the home (L1), in which they are proficient, as the primary language for communication and education, at least

in the early stages of education. They further contend that it is not essential for children to become orally proficient in their second language in order to become literate in that language. This has important implications for sign bilingualism where, with BSL as first language, some would suggest that it is not necessary for deaf children to learn spoken English. The important point is that they need to become literate in English, and, if this can be achieved without learning spoken English, then, they would argue, there does not need to be any emphasis on deaf children acquiring spoken English. However, learning to read and write in English without learning spoken English has proved to be very challenging for many deaf children brought up and educated in a sign bilingual approach.

Since hearing aids are seen as being prescribed in order to give access to the sounds of spoken language to aid its development, then hearing aids assume a far less important place. Instead of focusing on teaching deaf children spoken English, the focus will be on teaching them 'live English', i.e. how to use lip-reading, mime/gesture, writing/drawing in order to understand and make themselves understood in situations where they need to interact with hearing people who do not sign (e.g. in a shop) and there is no interpreter present.

DISCUSSION

The aforementioned descriptions may suggest that the situation is clear-cut and that for any deaf child it is possible to fit the communication approach used by and with the child into one of the three categories just described. However, since deaf children are individuals, with their own unique characteristics and needs, and since deaf children live in families with differing dynamics and behaviour patterns, the reality for many deaf children may be quite different. Whereas some families may hold a strong philosophical view and be determined to follow one particular approach (usually this is either a strong commitment to an oral approach or a strong commitment to the use of BSL), others take a more pragmatic view, changing between approaches as occasion demands or as their child's needs change. If the emphasis within the family is on communication, then parents and deaf children may use a mix of spoken language, gestures and signs. It is possible that the arguments over communication approach and the clear distinction between the different approaches may still rage between professionals who hold strong opinions or advocates of particular approaches, but within many families pragmatism takes precedence over philosophy.

There are commonalities and differences between the various approaches. The commonalities relate to an increased understanding that, whatever approach is used, it is important that facilitating *communication* should be the overriding consideration with young deaf children. In discussing family-centred practice in working with families with young deaf children, Brown and Nott[34] cite the following elements as fundamental: 'working alongside parents, quality of interaction, amount of interaction, ability to capitalize on everyday experiences and effective guidance based on parent–child interaction' (p. 146). Whilst they were writing with particular reference to families using an oral approach, the list of fundamental components would hold for all families with deaf children.

The difference between approaches may be as much a question of attitude, which accompanies the view of deafness held by parents or professionals. Beattie suggested, using a quote that was originally cited by Connor: 'oralism is as much an attitude as it is a method' (p. 337). The underlying conception of deafness that is held by parents, professionals and deaf children themselves will have a profound influence on their approach to communication.

Communication approach and cochlear implants

The increasing use of cochlear implants with young deaf children raises questions concerning the communication approach that should be used with the child prior to implantation and in the period following implantation. Cochlear implants give increased access to the sounds of speech, with the aim of promoting the use of spoken language in deaf children. How best to achieve this aim then becomes a question for consideration. Not surprisingly, given the history of differing views around communication approach and deaf children, there are different opinions about communication approach to use with deaf children who receive cochlear implants. One view holds that spoken language should be used exclusively with these children, both prior to and following implantation. There is research that demonstrates that deaf children with cochlear implants in oral settings outperform those in TC settings in several aspects.[35–37] However, this research suggests that deaf children's communication approach is fixed, when in reality this may not be the case. Recent research[18,38] has demonstrated that in many cases children who receive cochlear implants change their communication approach following implantation towards greater – or exclusive – use of spoken communication. The reason given by parents for this change was that it was child-led and driven by increased audition. Tait, Lutman and Robinson[39] found that the quality of communication pre-implant, by whatever means (i.e. speech, gesture and/or sign) was correlated with performance post-implant. This suggests that, paradoxically, it may be the case, at least for some deaf children, that, if their parents are seeking a cochlear implant with the aim of assisting their child towards the acquisition of spoken language, then they may best achieve this aim by using sign in conjunction with speech (SSE) pre-implant and in the early stages post-implant and then the child will drop the use of sign as the acoustic signal via the implant becomes the more salient one. However, not all deaf children with cochlear implants in the study did change towards the use of spoken language. Therefore, parents with a strong commitment to encouraging their deaf child to use spoken language exclusively may well decide to use it exclusively from the outset rather than start off by using sign in the expectation that their child will change later. Recent research,[40] which comprised interviews with parents and deaf children who had received cochlear implants, found that for some children there was a 'communication journey': pre-implant and in the early stages post-implant they used spoken language with sign support (TC); then, following a period of adjustment to the cochlear implant, they stopped using sign in favour of using spoken language alone, but later, maybe in recognition of their status as a deaf person with both a hearing and deaf identity, they chose to learn BSL. They were then able to vary their communication to use whichever means was most apposite for the situation, becoming what Spencer and Tomblin[21] refer to as 'facile code switchers' (p. 187). This finding contributes a useful perspective to the discussion around choice.

It is possible, then, that more deaf children will become bilingual, in English and BSL. Some will commence, and probably continue, in sign bilingual programmes, whilst others will commence in oral or TC programmes and learn BSL when they are older.

Whose choice is it?

The situation that used to obtain in the UK was that professionals often decided the communication approach to be used with a deaf child. It has been the practice for pre-school deaf children and their families to be supported by a Teacher of the Deaf who visits the home from diagnosis to advise the family on how to foster their child's language and communication and

other aspects of their development. The Teacher of the Deaf would belong to a team known as the Service for Deaf Children (or similar title), employed by a local authority to serve their geographical area. Some services advocated, and only made provision for, one particular approach. Thus a service might espouse an oral approach. Teachers of the Deaf who worked for that service would be expected to advise parents to use exclusively spoken language with their deaf child and would use the same approach themselves in communicating with and educating deaf children. The service would be likely to attract Teachers of the Deaf who were in favour of an oral approach and to be led by a Head of Service with similar views. Another service might favour the use of TC, or have a sign bilingual approach, and therefore advise the use of sign and use it themselves in communicating with and educating deaf children. The only choice for parents who did not agree with what was being offered was to move house to another geographical area, or perhaps to educate their child privately. Whilst this may sound restricting for parents, many were happy with the communication approach and education offered to their child.[41]

In addition to Teachers of the Deaf, other professionals would also offer strong opinions to parents regarding the communication approach to use with their deaf child, speaking from their own perspective and experience. These professionals would include speech and language therapists, doctors and ear nose and throat consultants. The opinion of professionals might have been based on an adherence to a particular philosophical position as discussed in relation to services for deaf children, or it might have been based on degree of deafness – a view that all children with a hearing loss equal to or greater than a certain level would benefit from the use of sign. Thus parents were either given little choice or were strongly advised by professionals. On occasion, the advice they were given by one professional contradicted that given by another, resulting in conflict between the professionals and confusion for the parents.

The current view is that parents of newly diagnosed deaf children should be given full information regarding the different communication approaches and then be expected to make the decision for themselves. This fits with the prevailing political and policy climate in the UK that encourages informed choice. Increasingly, in respect to medical or surgical interventions, patients are being provided with information and then making a choice for themselves. With respect to parents of deaf infants, materials have been produced to assist them with their decision making.[42] Implicit in this publication is recognition of the need to empower parents. Thus parents are expected to make the decision and professionals are there to support them in the decision that they have made and to ensure that appropriate provision is made for the deaf child in accordance with the parents' wishes. Tensions can arise; for example, there can be an inherent tension between an individual as chooser and the wider social consequences of any particular choice, which may be hard to reconcile for parents and professionals (p. 12).

Parents are expected to make an informed choice when they are ready. In Colorado, a professional is employed to support the parents whilst they make this choice. Some areas of the UK have chosen to employ a person to fulfil a similar role. There is evidence that some parents find the choice of communication approach difficult, but there is also some contradictory evidence. Parents may want professionals to make the decision, or at least to advise them, but current thinking suggests that this should be resisted and that it is the family that needs to make (and live with) the decision. The decision regarding communication can be seen as 'a big "one-off" decision that will be difficult to change in the future' (p. 27).

The advent of the newborn hearing screening programme means that diagnosis and confirmation of hearing loss occur when the baby is very young. The pressure for the introduction

of this screening programme came from parents who regretted that their child had not been diagnosed at an earlier age, and from professionals who were keen to commence intervention (in relation to language and communication development and possible use of amplification) as early as possible. However, there is concern to ensure that parents are enabled to consider all options equally. Whilst it may be the case that many hearing parents are anxious for their deaf child to learn to use the spoken language of the family, other hearing parents see signing as the way forward for their deaf child and the family. Young and Tattersall[10] report the findings of interviews with 45 parents, representing 27 infants whose deafness was identified as a result of the newborn hearing-screening programme in England. They found that in ten interviews parents expressed the expectation that their deaf child would have normal speech development and 'be able to manage successfully in mainstream education on a par with their hearing peers' (p. 215). This shows that hearing parents may have very high expectations of their child's ability to develop spoken language following early diagnosis.

Ultimately, the choice of communication approach lies with the deaf child himself or herself. The decision may be presented to the family as a one-off decision that will be difficult to change in the future,[41] but this is not necessarily true. As has been discussed earlier, many deaf children who have cochlear implants decide to change their communication towards greater or exclusive use of spoken language, and this may prove to obtain also for deaf children who have digital hearing aids that give them access to a wide range of speech sounds. Thus, communication approach is not necessarily fixed. Other deaf children who are raised and educated using an oral approach will decide when they are older that they wish to embrace the Deaf community and prefer to use BSL.

CONCLUSIONS

Looking to the future, there are different possible scenarios. On the one hand, with NHSP deaf children should routinely be diagnosed shortly after birth. Following diagnosis, the majority of deaf babies will be fitted with hearing aids at a very young age and, where appropriate, referred for cochlear implants. It is already the case that in some areas of the UK, the majority of profoundly deaf children have cochlear implants. This early diagnosis enables intervention to begin immediately, with the result that more deaf children are likely to acquire intelligible spoken language and to be educated in mainstream settings alongside hearing peers. Thus, more deaf children will have access to spoken language and use it in their daily lives.

On the other hand, the level of signing competence within the profession of education of the deaf has increased, on account of the fact that teachers of the deaf are required to obtain a basic level of signing skill as part of their training, and indeed many achieve a much higher level. In addition, more teachers who are themselves Deaf, and fluent in BSL, are training to be teachers of the deaf. Thus, for those families who choose to use sign, either BSL or TC, with their deaf child, there is support from staff who are knowledgeable about the use of BSL for communication and in education. Early diagnosis can also enable the family to learn to become skilled in the use of BSL and in accommodating to other aspects of Deaf culture.

Deaf children themselves may for the first time have a genuine choice. Research carried out with teenage users of cochlear implants[43] shows that they use whichever communication approach the occasion demands, switching between spoken language and sign language. If it proves possible to give more deaf children this choice, then professionals and parents will be serving them well.

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24 Delay and disorder in speech and language

G. Baird and T. Loucas

INTRODUCTION

The typically developing (TD) child shows remarkably rapid acquisition of speech, language and communicative competence. Communication commences in the first year of life with reciprocal vocalisations and non-verbal social exchanges of smiling, looking and gestures. First words develop between ages 10 and 12 months. The typical 2-year-old has a spoken vocabulary of several hundred words and is beginning to combine words, and his or her speech will be intelligible at least 50% of the time. At age 3 years the child can talk fluently in sentences and uses grammatical elements such as prepositions, plurals and verb endings; complex sentence forms are emerging and speech is 75% intelligible. By the time he or she enters school at age 5, the child has mastered the fundamental aspects of his or her native language. For some children, however, the acquisition of speech and language is impaired even though their development in other areas is typical.

CHILDREN WITH SPEECH AND LANGUAGE DIFFICULTIES

Delay and disorder

Theoretically, difficulties with speech and language development may reflect a simple delay in the rate of an otherwise typical pattern of development or may reflect a disordered or atypical pattern of development. However, in practice delayed and disordered development are commonly found together and 'impairment' is the preferred term. A distinction can be made between a primary speech and language delay (also known as a specific language impairment, SLI), where speech and language skills are delayed relative to the child's other skills in the absence of a clear aetiology, and a secondary delay, where speech and language delay is the result of a known aetiology.

Subtypes of specific speech and language impairment

The International Classification of Diseases (ICD-10)[1] identifies three subtypes of primary speech and language delay, which are termed specific developmental speech and language disorders.

Specific speech articulation disorder

Speech sound production may be the only area affected. A child's understanding of spoken language (i.e. receptive language) and expressive language are within normal limits, allowing

them to formulate meaningful and grammatical utterances at the same level as any other child of their age, but they cannot articulate speech clearly. Impairments of speech production can be broadly divided between those that affect articulation (i.e. an inability to produce particular speech sounds) and those that affect phonology (the mental representation of the speech sound system).

Expressive language disorder

Expressive language alone may be affected. These are children who are impaired in their syntactic and morphological skills but their receptive language is within the average range. Children with expressive language impairments may not have any marked difficulties with speech production, but some do.

Box 24.1 Rapin and Allen's[3] subtypes of developmental speech and language impairments.

- Verbal Auditory Agnosia (also known as 'word deafness')
  The child is unable to comprehend spoken language, but is able to do so visually (e.g. sign and written language). The children are mute or highly dysfluent, typically with poor speech articulation.
- Verbal Dyspraxia
  The child's speech is effortful and utterances are short with disordered phonology. Comprehension is adequate or normal. Syntactic structure is difficult to evaluate because of the severity of the speech deficit. The child appears to know what he/she wants to say but has extreme difficulty formulating words.
- Phonological Programming Deficit Syndrome
  The child is more fluent than in verbal dyspraxia and utterances are typically longer. The child also knows what he/she wants to say, but speech is poorly intelligible. Comprehension is intact. Expressive syntax and grammatical morphology are affected by phonological deficits.
- Phonologic-Syntactic Deficit Syndrome
  In this most common form of SLI, the child speaks in short utterances lacking grammatical morphemes with some dysfluency. Unlike pure speech disorders, comprehension is affected, but less than expressive language. Phonology is also affected.
- Lexical-Syntactic Deficit Syndrome
  Phonology is age-appropriate. The child has a severe word retrieval deficit and difficulty formulating sentences, particularly within the constraints of conversational demands. Expressive syntax is immature. Comprehension is generally more impaired than in phonological-syntactic deficit.
- Semantic-Pragmatic Deficit Syndrome
  The child's expressive language is fluent and well-formed with adequate articulation. Some children are verbose and have sophisticated vocabularies. Circumlocutions, semantic paraphasias and a lack of semantic content are typical. Comprehension is poorer than expressive language. Understanding of questions is particularly impaired and often messages are interpreted quite literally. Pragmatics are impaired, for example conversational turn-taking and maintaining a topic in discourse.

Receptive language disorder

Finally, some children have impaired receptive language. In the majority of cases expressive language is also impaired and speech may or may not be affected to some degree.

Several attempts have been made to further refine the subtyping of speech and language impairments using clinical[2,3] or statistical approaches.[4,5] One widely used by speech and language therapists was suggested by Rapin and Allen,[3] who identified six subgroups of SLI on the basis of clinical description (see Box 24.1).

Rapin and Allen's[3] SLI subgroups, which were based on clinical observations, received support from Conti-Ramsden, Crutchley and Botting[6] using statistical clustering. Van Weerdenburg, Verhoeven and van Balkom's[5] factor analyses of language test results from children with SLI produced four distinctive linguistic domains: lexical-semantic abilities, auditory conceptualisation, verbal sequential memory and speech production. Subsequent cluster analysis suggested four distinct SLI profiles based on different levels of impairment in these linguistic domains.

CHARACTERISTICS OF SPEECH AND LANGUAGE IN SLI

Whilst late talking is the first indication that a child is experiencing a problem acquiring language, children with SLI can present with difficulties at all levels of language. Usually, several components of language are affected: pre-school children who present with poor lexical skills and deficits in syntax and morphology typically show phonological difficulties as well.[7] Evidence from an epidemiological sample suggests that by school age the co-morbidity between speech delay and language impairment is just under 2%, with 5–8% of children with persisting SLI presenting with speech delay and 11–15% with a persisting speech delay presenting with SLI.[8]

Linguistic description

Phonology

Children may show typical, but delayed, patterns of development (often combined with oromotor immaturity) or atypical patterns. Thus sounds that are acquired early (e.g. /m/, /n/, /b/, /w/) emerge before those acquired late (e.g. /s/, /z/), and the phonological processes found in typically developing children, such as cluster reduction (e.g. 'snake' produced as 'nake') and the omission of final consonants, are also the most prevalent processes in the speech of children with SLI.[7] In atypical patterns of development, errors can be consistent, where the child uses non-developmental phonological rules consistently, or inconsistent, where the child uses non-developmental phonological rules variably.[9] For example, a child who is still fronting /k, g/ to /t, d/ at age 4 years would be showing a phonological delay, whereas a child who was backing /t, d/ to /k, g/ would be showing deviant phonology as this is not a typical developmental process.

Lexicon

Children with SLI are often first identified because they are delayed in first words or phrase speech. In SLI first words come at age 23 months rather than 11 months and phrase speech at age 37 months rather than 17 months.[10] Children with SLI show differences from TD children in the pattern of their lexical development and the learning and retaining of new words. They

use a more limited range of verbs,[11] relying on general all-purpose verbs (GAPS), such as 'do' and 'get'.[12] Children with SLI show poorer learning of action words but similar learning of object words when compared with TD children matched on language age[13] and difficulties retaining newly acquired words.[14]

Syntax and morphology

Although delayed, early word combinations are similar to those of typically developing children. All the major syntactic categories are evident. For example, pre-school children with SLI show typical use of nouns with determiners (e.g. in subject position before a verb, '*The man* going home'; in object position, after a verb, 'I get *the toys*'). However, some differences are reported. Children with SLI did not use major syntactic elements in as many different sentence contexts as language-matched TD children,[15,16] and they use the same types of question as TD children but show difficulties with the syntax of questions; for example, failing to invert the subject and auxiliary verb (e.g. 'What we can make?' versus 'What can we make?').[17]

The area of grammatical development that has received most attention in recent years is grammatical morphology. Morphemes which are verb-related appear to be disproportionately affected in SLI, such as past tense '-ed', third-person singular '-s', auxiliary 'be', copula 'be'. These elements are associated with the grammatical property of finiteness. Other aspects of grammatical morphology, such as plural '-s' and verb-related morphemes not associated with finiteness, such as progressive '-ing', do not seem to be so affected. Children with SLI will omit verb-related morphemes more often than younger TD children at the same level of language, but do not omit other morphemes (e.g. plural -s or progressive -ing), which have already been mastered.[18]

Pragmatics

Leonard[7] suggests that pragmatics is an aspect of language that is relatively spared in children with SLI who may show difficulties with pragmatics compared to TD children of the same age, but not relative to TD children at the same language level. For example, their conversational turn-taking is more adjacent than younger children, but less well-timed than older children and they attempt to repair by changing utterance, but less effectively than age-matched controls.

However, for some children, pragmatics (and semantics) are more impaired than the structural (grammar and phonology) aspects of language (see Box 24.1). Bishop[19] has argued semantic impairments do not always co-occur with pragmatic impairments and has suggested pragmatic language impairment (PLI) as a preferable term for a distinct diagnostic category lying on a continuum between SLI and autism spectrum disorder (ASD).[19–21] In PLI, pragmatic ability is disproportionately impaired compared with SLI and is dissociated from the social impairments and repetitive behaviours which define autism spectrum disorders.[19,20] Pragmatic skills can be assessed using the Children's Communication Checklist.[19]

THEORIES OF COGNITIVE UNDERPINNINGS OF SLI

Impairments in language representation

For one set of theories, SLI is a specific deficit in grammatical representation.[22] Hence, children with SLI have more difficulty understanding passive sentences, such as 'The lorry is hit by the car', than active sentences, such as 'The car hit the lorry', because in passives the word order does not reflect the meaning and so grammatical knowledge is required for a correct interpreta-

tion. Pinker[23] hypothesised a set of rules which help map the grammar of a sentence to its meaning. These linking rules can operate both from semantics to syntax (forward linking) and from syntax to semantics (reverse linking). If a child can form a representation of a verb's semantics from experience of it being used, he or she can use forward linking to infer the verb's syntactic properties. Van der Lely[24] found that children with SLI were able to use knowledge of forward linking to learn the meaning of a nonsense verb but were impaired on reverse linking, which suggested an intact ability to use semantic cues and a specific inability to use syntactic cues.

Evidence from a single family with a form of inherited SLI (see below section on 'Single gene defects') has been used to argue that the underlying impairment in SLI is a genetically linked inability to encode grammatical regularities.[25,26] Children with SLI are said to show 'feature-blindness' – their grammars lack syntactico-semantic features. Feature-blindness does not resolve with age, although there may not be any overt impairments in the language of an individual with feature-blindness as they can learn to produce correctly inflected forms by rote. Rice, Wexler and Cleave[27] account for these deficits with tense morphology in terms of a stage found in normal development in which children treat tense marking as optional rather than obligatory. SLI children, they argue, remain at this stage for longer, and may never leave it. They call this the extended optional infinitive (EOI).

Impairments in language processing

Over 30 years ago, Tallal and Piercy[28] proposed that the language deficits in SLI were the result of an impairment affecting ability to process acoustic information rapidly. This theory has been the subject of experimentation and debate ever since. Tallal and Piercy demonstrated that children with SLI subjects were impaired in their ability to discriminate between non-verbal tones presented rapidly, but their performance was normal if the tone were presented more slowly. Receptive language was highly correlated with temporal processing measures.[29] The finding of auditory processing deficits has proven to be robust, but the relationship between these deficits and language impairment remains unclear.[7,30] Bishop[30] notes that the evidence for the relationship between auditory processing deficits and language impairment is correlational and so open to several interpretations.

An alternative auditory processing account explains the grammatical impairments found in SLI in terms of a difficulty processing linguistic elements characterised by low phonetic substance.[31] These are non-syllabic consonants and unstressed syllables which are acoustically brief, low frequency and quiet. Thus English-speaking children with SLI have problems with the low phonetic substance forms plural -s, past -ed, possessive -s, articles, copula 'be', modal 'will', infinitive particle 'to', complementiser 'that', but relatively few difficulties with irregular past tenses formed by vowel changes (e.g. give/gave) which are not of low phonetic substance. Accordingly, Montgomery and Leonard[32] found that children with SLI were less sensitive to low phonetic substance morphemes than age- and language-matched controls in word recognition and grammaticality judgement tasks.

Another influential processing account of SLI was proposed by Gathercole and Baddeley,[33] who argued that children with SLI have poor phonological short-term memory (PSTM). PSTM stores verbal input temporarily, allowing other cognitive tasks such as verbal comprehension and the transfer of phonological information to long-term memory.[34] Thus, deficits in PSTM can potentially affect both comprehension and lexical development. PSTM can be measured by non-word repetition (NWR), a task requiring the child to repeat nonsense words consisting of different numbers of syllables.[35] Children with SLI are very much worse at repeating non-words of more than two syllables than children of the same age and non-verbal intelligence

Table 24.1 Proportions of individuals with SLI who have a co-twin with SLI in three twin studies from Bishop.[42]

	MZ	DZ
Lewis and Thompson[43]	0.86	0.48
Bishop et al.[44]	0.7	0.46
Tomblin and Buckwalter[45]	0.96	0.69

and younger children matched on language level.[33] NWR has come to be seen as a robust clinical marker for SLI which differentiates children with SLI from those without.[36,37]

Risk factors associated with speech and language impairment

Epidemiological studies[38–40] have identified a number of risk factors associated with SLI. These tend to be psychosocial rather than biomedical and include the following: low maternal education, bilingual home, poverty, minority status, parenting stress, low family expressiveness (an index of family distress), impaired parental mental health, and significantly, parental concern about language development. In the children, low prosocial behaviour, imitation and play are associated with an increased risk of SLI.[38] No risks in these studies were found from selected pre- or peri-natal events, such as maternal history of poor pregnancy outcomes, smoking, drug and alcohol use, gestational diabetes, sexually transmitted diseases (STDs) and urinary tract infections (UTIs).[39]

Genetics

SLI appears to recur in families. The possible genetic aetiology has been studied using different approaches: aggregation, twin and adoption studies, pedigree analysis and molecular genetics. Studies of family aggregation suggest an increased rate of language impairment in relatives of individuals with SLI: 48% positive family history in SLI relatives, compared with 18% in controls.[41] This evidence can suggest genetic influence but there are other possible explanations, such as cultural transmission through social learning. Twin studies rely on the fact that identical, or monozygotic (MZ), twins and fraternal, or dyzygotic (DZ), twins differ in genetic relatedness. In twin studies a consistently higher concordance rate is found in MZ as compared with DZ twins (see Table 24.1).

It is thought likely that it is the interaction of several genes that influences language development. Molecular genetic approaches in the UK[46,47] and the USA[48] have identified genes on chromosomes 2, 13, 16 and 19 which are associated with SLI.

SECONDARY SPEECH AND LANGUAGE PROBLEMS: AETIOLOGICAL FACTORS

Hearing loss

Sensory neural hearing loss

Persistent hearing loss has a significant impact on speech and language development, dependent on the degree of loss.[49] Early identification of hearing loss via newborn screening followed

by appropriate aids lead to marked benefits for speech and language development.[50] Consideration also needs to be given to sensory neural hearing impairments that are gradually progressive and to acquired causes, e.g. pneumoccocal meningitis, where deafness occurs in approximately 7% of children following infection.[51]

Otitis media with effusion (OME), or glue ear

Otitis media with effusion and associated hearing loss is extremely common. However, in a meta-analysis of published studies, Roberts, Rosenfeld and Zeisel[52] found little or no association with children's speech and language development. But they noted that most studies did not adjust for factors such as socio-economic status and concluded that, for otherwise healthy children in language-rich environments, the clinical relevance of OME is uncertain. However, for some children OME may be important when combined with other risk factors. It is important to distinguish cases where OME is associated with cranial-facial or other neurological or sensory neural deficit, since they will be at far greater risk of impairment from additional hearing loss or are likely to have very persistent OME. Clinical practice guidelines recommend intervention specifically for this group.[53]

Chromosome anomalies

An increase in chromosome abnormalities, particularly of sex chromosomes, has been reported in children with SLI.[54] Children with Down syndrome (an additional copy of chromosome 21) present with impairments in speech and expressive language relative to non-verbal ability.[55]

Single-gene defects

Certain single-gene defects are associated with particular patterns of strengths and weaknesses in language and speech acquisition; many but not all are usually associated with learning difficulties to a greater or lesser degree. In Angelman syndrome (deletion on chromosome 15) expressive language is severely impaired relative to non-verbal ability.

The combination of palatal dysfunction and speech impairment, occurring with or without learning difficulties, is associated with velo-cardio facial syndrome (a specific deletion of chromosome 22q). The core eight clinical features are as follows: cardiac defects, non-visible/hypoplastic thymus or infection problems, hypocalcaemia, feeding difficulties, cleft palate/speech-language impairment, developmental delay/learning difficulties, characteristic dysmorphic features (i.e. structurally abnormal body parts) and other malformations and deformities. This disorder is often missed in the pre-school years in children who present with a combination of many of the core features, with the median age at diagnosis being 6.7 years.[56] Of those diagnosed after age 2 years, the majority presented with speech–language impairment, developmental delay or learning difficulties and recurrent infections. A high proportion had no cardiac defect, leading to a risk of diagnostic delay; however, characteristic mild dysmorphic features were noticed in all children.

One family with marked speech and language impairments with oro-motor problems and some mild learning difficulties has been found to have a deletion affecting part of the *FoxP2* gene.[57] Although there were initial expectations that this genetic defect might provide an explanation for other forms of SLI, further analysis of the *FoxP2* gene in a large SLI cohort did not find this to be the case.[58]

Pre-natal exposure to environmental hazards

Anti-epileptic drugs taken in pregnancy can have an adverse effect on the foetus, including prematurity, low birth weight, congenital malformations and developmental delay. Foetal effects are dose- and polypharmacy-related. Sodium valproate poses the highest risk, as high as 14%.[59] Dysmorphic features and oro-facial defects are among the major malformations associated with the drug; neurodevelopmental delays, particularly communication delays and autism spectrum disorders, have also been reported. Exposed children had a significantly lower verbal IQ in one study.[60]

Foetal alcohol syndrome (FAS) is a continuum ranging from mild intellectual and behavioural impairments to an extreme that often leads to profound disabilities or premature death.[61,62] It is likely that in milder form it is commoner than is usually diagnosed.

Structural deficits affecting speech – cleft palate and midline submucous clefts

Cleft palate is associated with speech impairment and palatal dysfunction, for example food coming down the nose; a bifid uvula is another physical sign. Examination by palpation of the soft palate can reveal the abnormality. Children with cleft lip and palate also experience increased middle-ear problems and middle-ear ventilation is often warranted. Furthermore, even in the absence of other neurodevelopmental abnormalities, children with cleft lip and palate may show significantly lower scores on tests of cognition, comprehension and expressive language abilities than matched control children at ages 12 and 24 months.[63]

Structural brain abnormalities

In SLI without additional impairments, it is uncommon to find evidence of brain lesions. However, a number of structural abnormalities of the brain can be associated with language delay and speech impairments. Polymicrogyria (literally many small folds on the surface of the brain) syndromes, which are uncommon and result from genetic and non-genetic causes,[64] in the perisylvian regions of the brain result in a broad spectrum of speech and language impairments depending on the extent of cortical involvement. Learning difficulties, cerebral palsy, and seizures are found as well as problems using the muscles of the face, throat, jaws and tongue. When mild, this may lead to speech impairment or a tendency to drool but if more severe leads additionally to difficulties with feeding in infancy. The abnormality may be seen on magnetic resonance imaging (MRI). Polymicrogyria syndrome is now thought to be responsible for the congenital suprabulbar paresis first described by Worster Drought.[65] Features of this syndrome include severe speech impairment, history of feeding problems, drooling inappropriately, delay in gross motor function, learning impairment, pyramidal features on examination, seizures (in one-third of cases) and electroencephalogram (EEG) abnormalities (commonly).

Speech impairment, often with more general learning difficulties, may also be found in conditions causing cerebellar hypoplasia including various forms of Joubert syndrome, a rare developmental disorder that causes coordination and movement problems, mental retardation and speech impairment.

Acquired neurological damage

Acquired causes of speech and language impairment include infections such as meningitis, trauma such as head injuries or other intracerebral problems such as strokes affecting general

brain function or localized areas of speech and language processing. Recovery from unilateral brain injury affecting the speech and language areas, as for example a middle cerebral artery thrombosis, depends upon the age of the child. Those under 5–7 years of age will usually show no difference in language competence compared with other children, even though the damage affects the normally dominant left hemisphere language areas. This illustrates the plasticity of the developing brain, which enables the other hemisphere to take over language functions in the first 5 years.[66]

Cerebral palsy may affect the bulbar apparatus, causing dysarthria. The most severe impairment usually occurs with quadriplegia. A number of disorders involving degeneration of areas affecting motoric speech output may present with dysarthria, for example, cerebellar tumours and Friedreich's ataxia. Disorders affecting the cranial nerves may present with both structural and functional oro-motor impairments, including speech abnormalities. Myopathies may affect the face, eating and speaking functions. In rare cases, acute cerebellar damage from infection or following surgery may present as acquired mutism accompanied by irritability and other features lasting days or months. Outcome to some extent depends upon the underlying pathology.[67]

Epilepsy

Localised epilepsy, especially in the perisylvian region, which may or may not present as overt seizures, can have a devastating effect upon language development. Termed Landau Kleffner syndrome (LKS), this most commonly occurs in children aged 4–7 years where parents may gradually or suddenly notice loss of language accompanied by a profound receptive language impairment. In the most severe cases children do not respond to environmental sounds. Overt seizures are frequently not part of the initial presentation but can be. Sleep EEG shows a continuous spike wave activity, in some definitions more than 80% of the time, and recovery of speech and language is associated with the abolition of the interference this causes. Treatment to stop the epilepsy is essential and might include surgery if there is no response to medication.

More common than a clear epileptic syndrome is the finding of epileptiform EEG abnormalities in sleep in children with SLI.[68] However, their significance is uncertain and they are likely to be epiphenomena rather than aetiologically significant.[69]

Autism spectrum disorders

Autism spectrum disorder (ASD) is an important differential diagnosis in a child presenting with speech and language delay. ASD is characterised by the following: a qualitative impairment in sociability, empathy, the communicative use of language, creative and imaginative play; a restricted range of interests and activities; limited cognitive and behavioural flexibility. There may also be altered sensory responses to the environment and unusual mannerisms.

ASD is more common in children identified as having SLI compared with the general population.[70] This may represent earlier misdiagnosis or a changing clinical picture, or both. Verbal children with ASD who have normal non-verbal abilities can be divided into three groups on the basis of standardized language tests: normal, impaired and borderline. The latter two groups have language profiles similar to those found in SLI.[71] Rapin and Allen[3] found that all of their language impairment subtypes (see Box 24.1) could be found in children with autism, but receptive and pragmatic impairments are particularly marked. The social impair-

ments in ASD do not appear to be a consequence of language impairment. Longitudinal studies indicate that language may improve without an improvement in social interaction. This is especially the case if there are other features of ASD present.[72]

Selective mutism

Selective mutism is a form of anxiety disorder characterised by a failure to speak in social situations in the context of normal comprehension, sufficient expressive language for social communication and evidence of speaking normally in some situations. Children with selective mutism have greater social anxiety and other internalising symptoms compared with controls, but compared with children with social phobia only, they may have subtle speech and language impairments.[73] Reported prevalence rates vary but are approximately 1%[74] with onset at age 3–4 years.

Overt symptoms may improve considerably over time. However, in adults there are significantly higher rates of phobic disorder and any other psychiatric disorders compared with controls, and high levels of individual psychopathology and family psychopathology predicted poorer outcome.[75] Treatments shown to be effective are both behavioural (e.g. cognitive behaviour therapy) and pharmacological (e.g. fluoxetine/Prozac).[73]

PREVALENCE AND OUTCOME OF SPECIFIC SPEECH AND LANGUAGE IMPAIRMENT IN CHILDREN

Primary speech and language impairments are a very common developmental problem. Law et al.[76] reported a median prevalence of 5.9% from 11 studies carried out between 1974 and 1997. However, there was great variability in the estimates produced by different studies (0.6 to 33.2%) which was the result of the different definitions of speech and language delay used by different studies. Using a cutoff of −1 SD on a language development screening assessment, Rescorla et al.[77] reported a rate of expressive language delay amongst 2-year-olds of 19%. In contrast Paul et al.[78] reported a rate of only 1.35%, but this study used clinical judgement rather than standardised assessment to ascertain cases of speech and language delay. A rate of 6% is consistent with a large epidemiological study by Tomblin et al.[39] which produced a prevalence of 7.4% for a population of monolingual English-speaking nursery-aged children in Iowa, USA. Epidemiological studies suggest that SLI is more common in boys than girls at a rate that ranges from 1.25 : 1 to 2.3 : 1.[76]

Law et al.[76] conclude that expressive-only delays are likely to resolve spontaneously in the pre-school period, with up to 60% resolving without input between the ages of 2 and 3 years. However, predicting who is going to resolve spontaneously is difficult because the profile of a child's language skills may fluctuate over time. For example, Silva et al.[79] reported that whilst some children failed at each of three assessment points, 3, 5 and 7 years, others failed at only one or two. Children with combined receptive and expressive delays were the most likely to show persistent difficulties, whereas those with speech-only impairments were most likely to resolve spontaneously.

Paul[80] followed up thirty-six children identified as having slow expressive language development (SELD) at age 20–34 months. At age 7–8 years, 84% of the original SELD group now had language scores above the 10th percentile. Of the measures taken at age 20–34 months only socio-economic status and a parental report measure of expressive language (the

Vineland Adaptive Behaviour Scales, Expressive Language score) predicted expressive syntax at age 7–8 years. Paul[80] concluded that middle-class children with SELD have a good prognosis without intervention and that intervention is not necessary for pre-schoolers if language is the only concern, receptive language is within normal limits, significant progress is made in expressive skills and the child is understood by family, friends and peers.

Associated conditions

Children who enter school with speech and/or language impairments are likely to have continuing problems with speech and language and many also have impairments and dysfunction in other areas, including but not restricted to academic performance,[81,82] and those impairments are often in more than one domain.

Many children with SLI have problems with both literacy and numeracy. Young et al.[83] followed up a population-based cohort of children with SLI from age 5 to 19 years. Individuals were divided into those with a speech impairment and those with a language or speech and language impairment. The outcomes of the individuals with speech-only impairments did not differ from a group with no speech or language impairments. Those with SLI were at nearly eight times greater risk of difficulties with reading, spelling or maths (20% of the SLI individuals compared with 2.6% of the control group showed significant difficulties).

In a prospective study, Shevell et al.[84] found that almost half of a cohort of pre-school children, diagnosed with developmental language impairment at age 3 years and reassessed at age 7 years, had functional impairment in at least two domains of the Vineland Adaptive Behaviour Scales.

It is common to find associated impairments in motor skills, cognitive function, attention and reading in children who meet criteria for specific language impairment.[85] Developmental coordination disorder is a common co-morbidity.[86]

Social-emotional and behavioural difficulties with SLI are particularly common. Cantwell and Baker[87] assessed psychiatric difficulties in 600 children referred to speech and language therapy clinics and found 50% met criteria for psychiatric diagnosis. Girls had an eleven times greater risk of emotional disorders and boys a two times greater risk of attention deficit hyperactivity disorder (ADHD) than children without language impairments. Those with receptive problems were most at risk (81%) and those with speech problems least at risk (30%). Longitudinal studies confirm poor psychiatric outcomes for children with receptive language impairments and lifetime social and adaptive impairment, with concomitant effects on employment prospects.[88]

SCREENING, ASSESSMENT AND MANAGEMENT OF SPEECH AND LANGUAGE IMPAIRMENT

Screening

Significant delay in language development is a symptom which needs evaluation and a differential diagnosis. However, the advisability of population screening for language problems has been a matter of debate. A systematic review considering the value of introducing universal screening for speech and language delays in the UK concluded the evidence did not support formal screening and less formal approaches should be favoured.[76] Central to these was the

role of the parent in identifying a problem in the child and the role of primary care workers in eliciting and acting on parental concerns. In regard to formal measures, both parent-focused measures and assessments of child behaviour performed adequately in terms of their productivity, but instruments were not directly compared and so it was difficult to judge between different approaches. In general, specificity was higher than sensitivity, suggesting it was easier to identify children without speech and language delays than those with a problem. Screening was further complicated by the lack of agreement about cutoffs and 'gold-standard' measures, indicating there was little agreement on what counted as a 'case'. A more recent review in the United States came to similar conclusions.[89]

A systematic approach to the assessment and investigation of speech and language problems

Most children with speech and language impairments will be referred to and managed by the speech and language therapist. If, however, a more wide-ranging developmental delay is suspected or if there are concerns about social and communicative skills, or a severe motoric speech problem, the child should be referred to the child development multi-disciplinary team.

History taking

This should include the following: family history; pre-, peri- and post-natal problems; developmental milestones; current behaviour and perceived problems. Parents provide reliable information if asked the right questions and can usually provide an accurate estimate of the overall functioning age of their child. Parents may be less good at remembering particular milestones (although the age of walking is usually recalled) but they do remember whether a child was delayed or not. Asking parents to bring the parent health record to an appointment is a good aide memoire provided the information has been documented.

Pregnancy

Enquiry should be made about any rashes and fever during pregnancy, which may indicate exposure to a congenital viral infection, and about pre-natal exposure to toxins or drugs, including AEDs and alcohol (see above section on 'Pre-natal exposure to environmental hazards').

Birth and neonatal history

Parents are frequently concerned that any difficulty at birth is the cause of subsequent developmental problems; hence it is important to establish the gestational age at birth, the birth weight and whether the baby was 'small for dates' (i.e. below 10th percentile at birth). Although a difficult birth may be significant, it can also be a marker of pre-existing foetal difficulties. A persistent Apgar score below 5, with any symptoms of neonatal encephalopathy (e.g. seizures), early imaging abnormality, general metabolic disturbance, requirement for breathing support and the period for which that was needed would all be relevant in considering whether a difficult birth was a risk factor for subsequent developmental problems.

Family history

Relevant family history is any speech, language, reading or spelling problem in parents or siblings (see section on 'Genetics'). A family history of other developmental problems may also be relevant, such as ASD, which is reported more commonly in families where a child has a developmental language problem.[39]

Post-natal history

Key features are any major illness, trauma or accident involving head injury, or other event such as a seizure which could indicate a reason for neurological dysfunction.

Environment

A bilingual home environment is unlikely to be a sole cause of significant continuing delay, but it may exacerbate another causative problem. Parental mental health problems may impact on parental responsiveness (see section on 'Risk factors associated with speech and language impairment'). Nursery placements with frequently changing staff and children and with poor staff-to-child ratios are also environments that predispose to constant colds or ear infections.

Developmental assessment

Much developmental assessment is carried out by informed observation in a child-friendly environment; however, it is difficult even for the very experienced professional to accurately gauge a child's comprehension without formal assessment. The more general assessment of play will include gathering information about the degree of sociability, organisation, complexity and variety of play ideas demonstrated by the child. Pragmatic problems may be much more apparent in open conversation and play than in formal tests. Some children with language and communication problems are able to achieve skills in a structured situation with a helpful adult which they cannot achieve 'in real life', especially with peers, and the gap between elicited behaviour and actual behaviour is very informative.

During the assessment of any speech and language problem, in view of the frequency of co-morbidities, a systematic search for other co-existing conditions is important.

Specific observation should be made of the following: the communicative environment and any social interaction problems between mother and child; behaviour, remembering that for speech and language problems accompanied by behaviour problems the outcome generally is less good; motor and coordination skills and any signs of general developmental delay, especially behaviours that should have disappeared such as mouthing. Many non-verbal cognitive tasks may be mediated by internal language affecting non-verbal as well as verbal performance. Problems of attention with impulsivity, physical hyperactivity, poor persistence or attention to task and distractibility need to be distinguished from joint attention deficits where the child does not share interests but can sustain excellent attention for their own interests.

A developmental history and examination specifically looking for features of autism should be part of any assessment, using screening instruments such as the Social Communication Questionnaire (SCQ) and/or structured parent interviews (see Le Couteur and Gardner[90] for

an overview). Structured assessment of social communicative skills may be performed using the Autism Diagnostic Observation Schedule.[91]

General examination

This encompasses watching the child moving about and playing as well as any specific physical examination. Most information about motor difficulties can be gained from observation, including watching feeding. Physical examination then confirms any suspicions and should encompass alertness to signs of neglect or abuse. Head circumference, height and weight should be plotted on appropriate centile charts. Single measures, unless at extremes, are seldom diagnostically helpful but are essential for plotting trajectories, which may be more diagnostically indicative. Dysmorphic features, especially of the face, may indicate specific diagnoses especially in a child with learning difficulties. Physical examination should also include an inspection of the skin for café au lait patches indicative of neurofibromatosis and for white patches (best elicited by the use of Wood's light) indicating tuberous sclerosis. Oral examination should be undertaken in a child with severe speech impairment but may be best left until the end! Specific neurological examination without clear indications of abnormality is unlikely to add to diagnosis, and soft signs are non-specific of neurological immaturity.

Regression

Although rare, regression of speech and language at any stage in a child's development should prompt referral for further investigation. The commonest time for regression to occur is when the child has fewer than ten words in his or her repertoire, usually between the ages of 1 and 2 years. Regression is more likely if the child also shows symptoms of autism; between 15 and 30% of cases of autism report language regression. In one of the largest studies of language regression, over 90% of children who regressed when younger than 3 years of age received diagnoses of autism.[92] In contrast, language regression is very rare in SLI (Pickles et al.[93]).

Specific tests

The purpose of any medical assessment, including physical examination and laboratory or other tests, is to identify causative, associated or exacerbating medical problems. The guiding principle is that treatable conditions need to be identified and a high priority given to any condition with genetic implications for the child or other family members.

The type of speech and language problem can be a guide to investigations. *Dysfluency* in the absence of any other speech, language or developmental concern may require no further medical investigation. A voice problem may require an ear, nose and throat examination. Absence of dysmorphic features and dysarthria make the yield of investigations low in speech impairments. Receptive and expressive language impairments are less specific guides to investigation. Following examination and laboratory tests (metabolic, cytogenetic, imaging), an aetiological diagnosis was made in only 4% (3/72) of a referred sample of children with SLI under age 5 years seen in a tertiary assessment service.[94] In special schools for children with speech and language impairments, aetiological diagnoses are more common, reflecting the obvious point that more severe and persistent impairments are more likely to have identifiable causes. Robinson[95] described a special secondary school population of eighty-two speech-

and/or language-impaired children where aetiology was established in 26% of the total (11% pre-natal, 12% post-natal).

EEG and neuroimaging

Routine EEG in speech and language impairment is not indicated. Clinically the clue is regression or marked fluctuation of language comprehension and speech, which should trigger the request for a sleep EEG. Routine neuroimaging for speech or language impairment in the absence of physical signs is not recommended.

Karyotype and cytogenetic tests

There is a limited evidence base for judging the value of routine karyotypic estimation in language impairment, and much depends upon the population; however, certain features will increase the pre-test probability of finding an abnormality. Speech and language impairment in association with a general global learning problem (mental retardation) may provide the first indication. The routine request for karyotype in non-dysmorphic developmental delay/mental retardation continues to be questioned because of positive findings in less than 1% of cases (but of fragile X, 3%). New techniques such as array comparative genomic hybridisation may alter both the yield and advice about tests.[96] Any motor delay (or problem with running) would prompt a creatine kinase in the young pre-school boy, serum ferritin if there are marked dietary restrictions and more specific genetic tests if there are particular dysmorphic features.

Shevell and colleagues[97] examined the role of clinical features in predicting the identification of an underlying cause for a child's global developmental delay. Over a 10-year inclusive interval, the case records of all consecutive children less than 5 years of age referred to a single ambulatory practice setting for global developmental delay were systematically reviewed. An underlying cause was found in 37% (96/261) of children. Commonest aetiological groupings were genetic syndrome/chromosomal abnormality, intra-partum asphyxia, cerebral dysgenesis, psychosocial deprivation and toxin exposure. Factors associated with the ability to identify an underlying cause included the following: female gender; abnormal pre-natal/peri-natal history; absence of autistic features; presence of microcephaly; abnormal neurologic examination; dysmorphic features. In children without any abnormal features identified on history or physical examination, routine screening investigations (karyotype, fragile X, molecular genotyping, neuroimaging) revealed an underlying aetiology in only 16%. They concluded that the aetiologic yield in an unselected series of young children with global developmental delay is close to 40% overall and 55% in the absence of any co-existing autistic features. Readily apparent clinical features increase the likelihood of an identified aetiology.

Autism spectrum disorders

Currently, ASD as an additional impairment to speech and language problems does not indicate any further medical investigations or tests since the yield is low.

Management of a child with speech and language problems

Every parent faces a difficult adjustment when a child has significant developmental problems, but there are especial difficulties in coping with communication disorders. After every assess-

ment, parents should be given a written report and informed of the voluntary association relevant to their child's needs (e.g. Afasic formerly the Association for All Speech-Impaired Children for those with speech and language impairments). Co-existing conditions, having been systematically elicited, may be helped by strategic intervention. It is particularly important that behaviour problems are given a very high priority. The poorer outcomes of children with comprehension problems academically and behaviourally suggest that this is the group (with its several co-morbidities) which should be identified early.

Intervening for speech and language

Following assessment of the child's speech and language abilities, an intervention programme can be designed which aims to both target the linguistic impairments and increase functional communication. Three main approaches to interventions may be identified: didactic/clinician-directed; naturalistic/child-centred; hybrid approaches, combining elements of the other two.[76,98] Clinician-directed approaches are didactic and usually based on behaviourist principles geared to teaching specific behaviours through drill. For example, the structure 'NP is [verb]+ing' may be targeted (where NP is a noun phrase, the subject of the sentence). The child is shown a picture to describe and is prompted to imitate a model: 'Say, the boy is running'. Child-centred approaches, in contrast, provide more naturalistic settings for the development of the child's communication skills. Typical of this approach is the Hanen Early Language Parent Program,[99] which aims to modify parents' interaction with their language-impaired child by teaching them to follow their child's lead in play, to provide simple language which is focused on the child's current activity and to add to the child's utterances by imitating, expanding and recasting what the child says. For example, if the child said 'doggie', the adult might expand by adding an adjective 'big doggie'; or if the child says 'there doggie', the adult might provide a corrective recast of the child's utterance, 'there is a doggie'. Hybrid approaches, to some extent, combine didactic and naturalistic elements. For example, Fey and Proctor-Williams[100] argue that the most effective way of intervening to develop grammatical abilities is to introduce new grammatical structures using didactic approaches, such as imitation. This can be followed by focused stimulation in natural conversational settings, using corrective recasts when the child produces the new target form incorrectly, to facilitate generalisation to spontaneous language.

Reviews of the effectiveness of speech and language therapy for children with SLI suggest that a range of approaches can be effective.[89,100,101] In a recent systematic review of 33 different randomised controlled trials, Law, Garrett and Nye[102] concluded that there was evidence for interventions targeting expressive phonology and vocabulary being effective. The evidence for the effectiveness of therapy focused on expressive syntax was mixed, with some suggestion that it may only be effective if children do not have severe receptive language difficulties. For children with the worst outcomes, those with receptive language difficulties, there was little evidence that therapy targeting receptive language was effective. However, there were only four studies that met the criteria for inclusion in the review. Evidence from less well-controlled studies suggests comprehension can be improved through intervention.[89]

For some children spoken language may not be the focus of intervention. Children with speech, language and communication needs have been shown to benefit from the use of sign language and/or symbols, which facilitates communication and the development of spoken language.[103] For the severely motorically impaired child for whom achieving comprehensible speech may not be possible, referral for augmentative and alternative communication may be needed.

CONCLUSION

In addition to interventions that target the child's linguistic impairment, management needs to take account of the whole child and how the child functions communicatively in everyday settings. To this end evidence-based and best-practice guidelines recommend collaborative approaches to management that include both parents and educators in the context of the child's environment, culture and language.[104] For the school-aged child, the aim of speech and language therapy management is to help the child become an active participant in his or her learning and to use language effectively; thus the purpose of involving speech and language therapy is to facilitate acquisition and maintenance of communication skills which support learning and life skills.[104]

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25 Psychological effects of deafness and hearing impairment

R. Kentish and J. Mance

INTRODUCTION

The experience of hearing loss brings many challenges throughout the course of childhood and adolescence for both the individual and their family, over and above those one typically expects during these fast changing years. These challenges can be stressful and overwhelming, but can also bring opportunities for learning and personal growth. Overall, however, levels of psychological problems are higher in this group compared with hearing children and young people. Although the psychological consequences of hearing loss result in a range of experiences, anxiety, low mood and low self-esteem are among the more common. These may well underlie presenting difficulties such as refusal to wear hearing aids or to cooperate with medical and audiological investigations, poor attendance or progress academically and non-organic hearing loss.

In order to understand some of the causes of psychological difficulties in deaf and hearing-impaired children, there are some key questions to consider. What are the factors within the child, his or her social and educational environment, that place the child at greater risk of psychological problems? Equally, what factors may be protective? Family response to the child's hearing loss and their ability to meet the demands of their child's hearing loss are important. Finally, what support do children and families need to meet these additional challenges?

PREVALENCE OF PSYCHOLOGICAL DISORDERS

Research upon the prevalence of psychological disorders in the deaf and hearing-impaired population presents a number of challenges. Assessment must be carried out by clinicians experienced in deafness who are able to communicate fluently with deaf children. The limited number of assessment measures standardised and suitable for use with the deaf population adds to the complexity. For clinicians unfamiliar with deafness and hearing impairment, psychological difficulties may be wrongly attributed to the deafness, or conversely the psychological impact of deafness may be minimised or misunderstood.

Nevertheless, a number of studies suggest that levels of psychological problems are higher amongst children with hearing loss, in comparison to the general child population. Rates range from 15.4 to 60%, two to five times that found in comparison groups. One well-conducted study[1] found that 50% of children with moderate to severe hearing loss met criteria for psychological problems, as opposed to 25% of their hearing counterparts. Anxiety disorders were found to be the highest single-diagnostic group. The rate of psychological problems in deaf

teenagers has been found to be 2–3 times higher than that of the normative sample, with up to 39% of young people falling within the clinical range.[2]

It is, however, important to note that there is some evidence to suggest that deaf children of deaf parents do not have higher levels of psychological difficulties than hearing children of hearing parents. They also attain better cognitive and emotional outcomes than deaf children of hearing parents,[3] have better self-image and have less difficulty with impulse control.[4] Although more research is needed in this area, it seems likely that this difference can be attributed in part to the better language and communication skills shared by deaf parents and their deaf child. The deaf child will most probably learn to sign fluently with their deaf parents from a young age, whilst hearing parents may well be learning to sign alongside their child. This highlights the importance of language and communication for emotional and psychological development from the very earliest years. Furthermore, deaf parents are more likely to share with their child a positive view of deafness and Deaf culture, which supports the child's developing sense of identity and self-esteem.

RISK FACTORS FOR PSYCHOLOGICAL DIFFICULTIES

When considering factors that contribute to psychological well-being in children with hearing loss, it must be remembered that deaf and hearing-impaired children are not a homogeneous group. General factors which influence psychological well-being in any young person will continue to have a role: the child's personality; additional disability or illness; and life experiences such as trauma or family breakdown.

We will briefly examine in turn factors within the child, the family and school that can potentially increase the risk for psychological disorder and those that are more likely to promote healthy psychological development.

FACTORS WITHIN THE CHILD

Additional disability

Perhaps surprisingly, the level of hearing loss itself does not appear to be a major risk factor; indeed, research seems to suggest that there is little or no correlation between psychological disorder and level of hearing loss. Rather, lack of communication skills in the child, and between child and parents, has been identified as being of far greater importance.[5]

It is known that the presence of an additional disability increases the risk for psychological disorder. The prevalence of an additional disability is around three times higher in hearing-impaired children than in the general school population. Causes of this include the syndromes with which the deafness is associated, and illness or neurological insults that also caused the hearing loss. Hearing-impaired children are approximately three times more likely to have additional difficulties such as cerebral palsy, pervasive developmental disorders, epilepsy and learning disabilities.[6]

Attention deficit hyperactivity disorder (ADHD)

Hearing-impaired children are commonly thought to be more impulsive than hearing children. However, the incidence of ADHD in hearing-impaired children seems to be on a par with the

general child population, with the exception of children with acquired hearing loss and/or additional difficulties.[3,7] Assessment of ADHD in hearing-impaired children presents a number of challenges for the clinician and it is important to take into account how the child's hearing loss will affect their attention span and control. There are important differences in the way hearing-impaired children attend to their environment. For example, questions such as 'Is the child easily distracted by extraneous stimuli?' will have a different meaning for the hearing-impaired from those with normal hearing. Where there are significant concerns about a child's attention span, a comprehensive assessment by an experienced clinician is very important. Deaf and hearing-impaired children are particularly vulnerable to the detrimental effects of ADHD. The compounded effects of both deafness and ADHD can have a disastrous effect upon the child's overall learning, development and social communication skills, as well as increasing the risk of the child becoming socially isolated as a result of both conditions.

Hearing loss, cognitive abilities and educational attainment

Within the child population, there is evidence to suggest that lower IQ not only places a child at greater risk of lower educational achievement, but also increases the risk of psychological disorder. Severe hearing loss has often been associated with educational disadvantage and lower performance on cognitive tests, particularly on the verbal tests. However, it is important to note that there is some evidence to suggest that even mild hearing loss has been associated with educational and cognitive disadvantage. There may be a number of reasons for this educational disadvantage such as difficulty in following classroom teaching, which in turn leads to poorer academic motivation and attainment.[8]

It is commonly thought that many deaf children have enhanced spatial skills, related to their use of sign, but this is not always the case. Where visual perceptual deficits are present, it is particularly important that these be identified. For a child whose primary modality for learning and communication is visual, any deficit in spatial cognitive skills will have a devastating effect upon the child's ability to comprehend the spatial components of sign language; thus language, cognition and social development will be affected.

THEORY OF MIND

Theory of mind refers to the ability not only to reflect upon one's own thoughts and feelings, but also to understand that other people have different thoughts, feelings and views from one's own, or in other words that other people have different 'inner worlds', which direct their behaviour. These skills are generally considered to develop during the pre-school years, the time when language skills are typically developing most rapidly. Research has shown that by the age of 2 or 3 years, young children have begun to develop an understanding that other people see and experience things differently from themselves. By the age of 3–4 years, young children have usually progressed to understand that things are not always what they seem, and that individuals may hold different beliefs about the same thing, depending upon what information they have been given. Pretend play, where different roles are acted out and the partners in play must adjust their actions to accommodate those of their play partner, is an important part of developing perspective taking. It is through social interaction that a child learns about others' mental states and how these relate to behaviour and the sequence of events.

The research in this area is highly complex but there is now a body of evidence to suggest that, in comparison with hearing children, theory of mind skills are delayed in deaf children of hearing families. However, the same delay is not found in deaf children of deaf parents.[9] This finding gives us information about the crucial experiences necessary for developing theory of mind skills. In contrast to deaf children of hearing parents, deaf children of deaf parents typically develop language and communication skills at a normal rate and have the more usual opportunities for conversational experience, which in turn gives children access to other people's mental states. Thus, early exposure to language, be it signed or oral, facilitates performance on theory of mind tasks.[10,11] As already mentioned, opportunities for social interaction and social communication are essential; here again, the deaf and hearing-impaired child of hearing parents is frequently at a disadvantage, with more limited communication opportunities. The finding highlights the likely significance of peer interaction and early fluent communication with peers and family, whether in sign or speech, in order to facilitate the growth of social cognition and language.

Not only do theory of mind skills develop as a result of language, communication and social interaction, but inevitably as the child grows older, the ability to understand and empathise with others' thoughts and feelings will lead to more successful social interaction and engagement with others; this in turn has implications for the social, behavioural and cognitive development of the child.

THE FAMILY

Parental beliefs and attitudes towards hearing loss will play an important part in determining how parents respond to the challenges that arise from their child's hearing loss – the extent to which they view this as an overwhelming trauma and source of grief which negatively impacts upon them and their ability to parent their child, or as a challenge to which they can accommodate.

The influence of deafness on families has been shown to be far-reaching. Since 90% of deaf children are born to hearing parents,[12] the majority of deaf children are raised in families who have no prior experience of deafness. Issues of identity will also be influenced by the child's experiences of their family – many children will be the only deaf person in the family and may feel very 'different' in a way that children from other minority groups would not experience in a family setting. Parental beliefs and attitudes towards hearing loss, either positive or negative, will be transmitted and internalised by their child, as part of their self-identity.

Diagnosis of deafness

In culturally Deaf families, the diagnosis of a deaf baby may be a positive and longed for event.[13] However, given that most deaf children have hearing parents, the news that a child is deaf is usually both shocking and unexpected. The child's deafness may be detected shortly after birth, by newborn hearing screening programmes (NHSPs), or may only be confirmed after several years as in the case of a progressive hearing loss. The experiences that parents have prior to the diagnosis of deafness, the timing of the diagnosis and the way that the news is conveyed to the parents may vary immensely. These factors have been found to affect parental grieving and coping, as they adjust to the news and to having a deaf child in the family.

Receiving the news that a baby is deaf has been likened to being bereaved.[14] Hearing parents are unlikely to have expected that the child would have a hearing loss, and they may be very unfamiliar with what this might mean for the child's life. When parents receive this difficult news, they are likely to feel extremely shocked and may be disbelieving or denying of the deafness. As the natural grieving process continues, anger is frequently expressed. Parents may come to a stage of questioning why the deafness occurred and may blame someone for the deafness, possibly themselves, the child or medical professionals. Feelings of guilt may ensue, if parents perceive themselves to be at fault, perhaps having unwittingly passed on a genetic vulnerability to the child, or having consented to a medical procedure which had untoward side effects. Sadness will also be prominent, as the parents come to terms with the deafness and what this will mean for their child's life.

The bereavement model proposes that eventually, having moved through the stages of grieving, the family are able to accept the deafness and form an attachment with the deaf child. It should be noted that whilst all the stages of this kind of reaction are usually transient, in some cases they may be interrupted and may take years to resolve. For example, a lengthy court case over medical negligence may result in a protracted 'anger' stage, delaying acceptance of the deafness. Some parents may find it difficult to move through the stages of disbelief and denial, which may result in unrealistically high expectations, or over-investment in interventions such as having a cochlear implant which they hope will 'fix' the deafness. Some authors suggest that the grieving model is rather simplistic, since there may be a complex range of feelings involved. Feelings of shock may be tempered with relief that the child survived serious illness in the peri-natal period,[15] or that long suspected deafness will finally be taken seriously.

However, for most families, feelings of loss will predominate. Indeed it has been suggested that, rather than a bereavement model of grieving, a model of grieving and coping more similar to that of chronic illness may be more appropriate. The deafness does not resolve,[16] and it is something to be accepted and lived with, rather than grieved as a discrete event.

Impact of newborn hearing screening

Universal newborn hearing screening has been found to be enormously beneficial in the early identification of deafness and provision of services. Parents also seem to be in favour of newborn screening, whether their child was screened or not.[17] However, it should be noted that early diagnosis might bring its own challenges to parents. Whereas in previous years there may have been a gradual recognition that the child showed difficulty with hearing, parents are usually unprepared for a diagnosis of deafness, now received shortly after birth. The diagnosis now takes place at a time when the family is already managing the transition of a birth, and at a time when parental mental health can be vulnerable. Some parents report that the early diagnosis, and the associated grieving, makes it hard to bond with their baby,[18] though this was seen as being outweighed by the practical benefits that early diagnosis brings.

The way in which the news is broken to parents has not always been reported as sensitive, with some parents finding the delivery of the news rather cold and blunt, which they felt made it hard to accept. There may also be a risk of 'medicalising' the child from the very beginning, or creating high expectations of language attainment which then become a source of anxiety for the parents if they are not met.[17] Overall, though newborn hearing screening has great practical benefits, clinicians should be aware that it may result in a range of reactions from parents.

In some cases, parents have long suspected that their child has a hearing loss but felt that they struggled to obtain a diagnosis and were falsely reassured by professionals along the way, possibly in the case of a progressive hearing loss. When the diagnosis is finally made, possibly several years later, parental reaction to the diagnosis may at first be a sense of relief but is often mixed with feelings of shock, grief and also anger. Clinical experience has frequently shown that late diagnosis of deafness can result in parents having even greater difficulty in adjusting to the child's hearing loss. Professionals are often subsequently perceived as less trustworthy for failing to detect the child's hearing loss at an earlier stage. Any delay in language development, or behavioural or emotional difficulties in the child are attributed to the delay in diagnosis, and parents express a sense of loss and anger that 'things could have been different' if the hearing loss had been detected and hearing aids fitted at an earlier stage. Parental grief and anger can continue for years, and with ongoing difficulty in accepting their child's hearing loss. This sense of loss is inevitably transmitted to the child, who in turn will have difficulty accepting their deafness with concomitant poor self-esteem. It is not unusual to find that late diagnosis of deafness results in poor parental adjustment, rejection of deafness by the child and poor acceptance of hearing aids in these children.

Effect of deafness on bonding and attachment

Newborn babies are completely dependent on their caregivers to meet their needs; they are unable either to meet physical needs, such as for food and warmth, or to regulate their own internal emotional arousal. Attachment theory, developed by Bowlby,[19] conceptualises the main caregiver (usually, though not always, the mother) as a secure base from which the baby is able to gradually explore and experience the world. In an optimal or 'good enough' scenario, the baby signals its needs to the mother, and the mother is responsive to the baby and able to meet its needs, whether this be for physical contact, feeding, changing and so on. The degree to which the mother is sensitive to and responsive to the communications of the baby is known as 'attunement', and this is gradually built up over a series of interactions. In turn, attunement leads to the development of an attachment between the mother and baby.[20] Research has identified that approximately 55% of children show a 'secure' pattern of attachment, in which they experience caregivers as positive figures with whom they feel secure and can separate from happily to explore their environment.[21] They return to the caregiver for comfort when distressed and experience parents as responsive to their needs. The remainder of children are understood to show 'insecure' patterns of attachment, which may be predominantly 'ambivalent', 'avoidant' or 'disorganised'.[22,23] Children with these kinds of attachment patterns have been found to be at risk of developing emotional and behavioural difficulties,[24] though these classifications are not immutable and later positive experiences with attachment figures can help a child to develop a more secure attachment style.

Bonding is not only a function of the baby's needs being met, but is affected by the perceived responsiveness of the baby to the mother – so in this way, the mother and baby affect one another in a reciprocal way. For example, if the mother talks to the baby, and the baby makes eye contact with her and gurgles in response, this is mutually reinforcing for the mother and the baby. If the baby has a hearing loss, however, this can have a significant effect on these early processes. Hearing babies are able to hear their mother's heartbeat whilst in the womb, and show a preference for the mother's voice shortly after birth.[25] Hearing a voice, particularly that of the mother, has been found to have a soothing effect, decelerating the heartbeat of babies.[26] However, a deaf baby may be unable to access this comforting experi-

ence. If a parent approaches the baby from outside its line of vision, the approach may not be heard, and the baby may be startled at the parent's sudden arrival and may cry or respond in a way that seems negative. Parents in this situation may feel rejected or inadequate. This may, in turn, lead parents to give less stimulation to the baby, feeling that the baby 'does not give much back' – when in reality, the baby may need increased sensory input through visual and tactile modalities.

With support and understanding of how they may need to adapt their behaviour with a deaf baby, parents can facilitate the development of secure patterns of attachment. Indeed, these issues may be dealt with at an earlier stage with the advent of NHSPs, which identify deaf babies at a much earlier stage. It should be noted that these kinds of difficulties with attachment do not seem to be prevalent amongst deaf parents of deaf children, with deaf children being rated as having similar patterns of attachment to hearing peers.[27] However, in the case of late diagnosis or temporary conductive losses, as already described, it can be seen that auditory input has an important role in the parent/child bonding and development of an attachment relationship.

Parental stress

Parents of children with a disability are commonly at greater risk of stress, and this in turn has been linked to insecure attachment, and more negative interactive patterns between parent and child. Parents of the hearing-impaired child may well feel social isolation, exacerbated by the child's limited language skills, or by the use of sign language which others outside the home do not know.

The use of social support systems is a well-known factor in family coping and adjustment, and again NHSP should bring this to families at an earlier stage. However, support is measured more usefully in terms of its quality rather than quantity and the extent to which families perceive it to be supportive and helpful. Indeed, some families find professional support in itself stressful, having to cope with a large number of helping professionals, each with their own set of advice. For the parent of the hearing-impaired child, there is much to be learnt about their child's hearing loss and how this will affect their child's development in the early years. There are undoubtedly new skills to be learnt in terms of furthering their child's development, which will be new and potentially overwhelming to them. Whilst welcome, advice can be experienced as undermining of parents' feelings of competence in managing their own child, can reinforce feelings of inadequacy at parenting a child who has special needs and can increase feelings of helplessness.

Parenting style and the hearing-impaired child

As we have already seen, the early interaction between parent and child forms the basis of communication and a loving relationship between the parent and child, together with an emotional bonding. Interactions between the hearing-impaired child and their caregiver are inevitably affected by the child's loss of hearing. After the early baby years and as the child enters the toddler years, successful communication between child and parent remain as important as ever; for this, the parent and child must share a common language, whether spoken or signed. Language is usually the means by which mothers can attract and hold their child's attention. Consequently, mothers of hearing-impaired toddlers have been found to spend less time in, and have less complexity of joint attention than those with hearing toddlers.[28] The consequences of these features of mother/child interaction have not been investigated, although

Calderon & Greenberg[29] hypothesised that low levels of communication between mother and child can lead to less secure attachment relationships and be associated with later behavioural difficulties, such as non-compliance and aggression.

As children get older, linguistic interaction becomes even more important for social and emotional development. For parents, this is the primary means of communicating more complex information about why some behaviours are acceptable and others not, the reasoning behind social norms, behavioural rules and the links between social, behavioural and emotional events. Lack of effective communication between parent and child has been shown in several studies to be related to impulsive behaviour, poorer self-image, more disruptive behaviour and an external locus of control on the part of the deaf child.[30]

Lack of good language and communication between parent and child has further implications. In observational studies, hearing parents have been described as more controlling of deaf children than hearing children.[31] Lederberg[32] noted that, as a result of the child's language delay caused by the hearing impairment, parents use simpler and more controlling communication and 'take over' the conversation more frequently. She hypothesised that this would increase passivity in children with hearing impairment and increase the risk of emotional problems.

Impaired language and communication between parent and child will also affect parents' style of behaviour management. Mothers of deaf children are more likely to select physical responses to their child's misbehaviours and for the discipline to escalate if the child persists with the transgression.[33] If a deaf child misbehaves, they are more likely to be withdrawn from the situation rather than verbally admonished. The consequences of this style of parenting for the child are that they have fewer opportunities to learn from their behaviours, or to learn substitute behaviours that would be more acceptable. In addition, parents are modelling to their child that avoidance and physical action are acceptable ways of solving difficulties. In contrast, a less intrusive and authoritarian style of parenting in early childhood is more likely to result in better adjustment in deaf adolescents.[29]

Children need freedom to explore and to learn from their own mistakes, but they also need to know the boundaries around acceptable behaviour, which in turn gives the child a sense of security that there is some predictability to their lives. Inconsistent discipline on the other hand has been associated with more behavioural problems.

Parental expectations and boundary setting

Many factors will influence parenting behaviour and style, but amongst them are parental views and attitude towards hearing impairment and to disability. The prevailing societal view of disability is one of tragedy and is largely negative.

One consequence of viewing deafness as a disability is that parents are at risk of perceiving emotional and behavioural problems as being an inevitable and unalterable part of deafness. The implication of this is that parents have no choice but to tolerate their child's difficult behaviour as being 'part and parcel' of deafness.

Parents of disabled children frequently feel a responsibility to compensate for their child for the 'tragedy' of being deaf. It is not uncommon to find parents whose natural feelings of protectiveness have become magnified and who seek to compensate their child for any distress or hardship he or she experiences as a result of deafness. Parents often acknowledge that they make extra allowances for their child and that they find it harder to instil appropriate boundaries around behaviour and are reluctant to discipline or upset their child as perhaps they would do with their other children.

Over-protectiveness

When a child's language skills are delayed, parents often feel that their deaf child needs more supervision than a hearing child. As a result they may seek to shelter their child, both socially and emotionally, from situations that could potentially cause difficulty for their child. They may feel a strong need to act as interpreter for their child, or as negotiator with the outside world. Whilst often helpful, such a stance may result in parents finding it more difficult to separate from their child and feeling anxious about the child when they are not with him or her. Such over-protectiveness is understandable and helpful in the short term, but limits the child's opportunities to learn independence and to develop his or her social interactions and communication skills. It may also result in a child who is passive, dependent and less mature socially.

Social development of the child outside the family

We may observe that children who attend our clinics find social settings very difficult, or that they do not seem particularly at ease with other children. When encountering the outside world, the hearing-impaired child whose language skills are delayed, or who does not share the same language as the other children, will be at a distinct disadvantage. The social development of deaf children is strongly related to their use of language. For example, hearing loss can lead to difficulties in learning the rules of interaction with other children, such as the instructions for playing a new game, rules and turn taking,[34] which in turn affect the child's relationships with others and place in the social world. Though hearing adults who know the child may be able to understand their communication, other hearing young children may not persist in developing relationships with a deaf child if they find it difficult to communicate with them. (Perhaps the current trend for teaching sign language to hearing babies may influence these relationships in future!) Studies of deaf children in a range of contexts show that those with more severe losses tend to have more deaf friends and to play more with children of similar hearing status.[35]

For older children who communicate orally, the transition from primary to secondary school may place heightened demand on spoken language skills. The broadening of the curriculum, and the emphasis on negotiating the school day more independently, may be particularly challenging. Adolescents often use slang and may talk in what seems like 'code' to outsiders.[36] This may present a particular challenge to a young deaf person who relies heavily on lip-reading or context to understand what is said. Socialising out of school may become increasingly difficult in adolescence, where the focus of social life is away from the home. Young people often socialise in poor listening environments such as echoic halls, or pubs and clubs with variable lighting and high levels of background noise.

THE IMPORTANCE OF SCHOOL SETTING FOR SOCIAL DEVELOPMENT

School setting plays an important role in any child's life, and deaf and hearing-impaired children may find themselves in a range of school settings: a school for the deaf; a hearing-impaired unit; or a mainstream school possibly with additional support.

Interestingly, in Hindley's study,[1] rates of psychiatric disturbance in children in hearing-impaired units attached to a mainstream school were one and a half times higher than in children who attended a deaf school. Mainstreamed adolescents have been found to have higher levels of social anxiety and more negative self-perceptions compared to those in deaf schools.

The reasons for the higher levels of psychological disturbance found in children in deaf and mainstream schools are probably complex. It may well be that hearing-impaired children with additional learning and psychological difficulties are more likely to be placed in specialist provision. However, it is very likely that the social aspects of being in a mainstream school with predominantly hearing peers is more challenging for hearing-impaired children. In a mainstream school, children with hearing impairments are in a minority and typically face more teasing and bullying centred on their deafness.[37] Hearing-impaired children are therefore at particular risk of isolation and low self-esteem. Manfredi[38] hypothesised that mainstreamed children were more aware of the constraints derived from their impairments and that realising 'the extent of their differentness' accentuated feelings of loneliness. In contrast, deaf schools offer the protective effects of a similar peer group, the use of sign and a deaf cultural identity.

IDENTITY, SELF-ESTEEM AND PSYCHOLOGICAL WELL-BEING

Background information

As established earlier, the notions of self-esteem and identity seem to be particularly important in the context of the psychological well-being of young people with hearing loss. Self-esteem can be understood as the evaluative and affective dimension of self-concept[39] – i.e. not only how the person sees themselves, but the degree to which they view themselves positively or otherwise in relation to aspects of themselves that they consider to be important. We commonly think of self-esteem as being a global factor and tend to describe people as having high or low self-esteem. However, global self-esteem is derived from an evaluation of abilities or successes in a number of domains that we personally consider to be important to us, in the areas that we have aspirations to succeed. Success in areas of little importance is not likely to affect our sense of self-esteem very much. We all vary in the extent to which we prize being good at sports, successful academically, popular or attractive. If we do not prize these things, then our lack of knowledge or success in this area will not matter. In addition to a global sense of self-esteem, we may have a positive sense of self-esteem in some areas, but a negative self-esteem in others. What matters to us are the areas that we *do* consider important, and low self-esteem has been described as arising from a mismatch between what we hold important, and what we achieve. For a hearing-impaired child, being a member of the hearing world may result in low self-esteem when they find themselves struggling to cope when communicating with hearing friends, to hear in classrooms and so on.

Children's appraisals of themselves is one of the strongest predictors of emotional and behavioural problems in paediatric conditions. Low self-esteem is known to be a feature of a range of psychiatric disorders,[40] such as depression, anxiety and eating disorders. Low self-esteem has been found to be causally linked to psychiatric disorders such as depression,[41] though it may also be an effect of other wider problems such as social difficulties or isolation.

Adolescence is a time when the young person struggles to form a coherent sense of identity and to define themselves, so the issue of identity can become particularly prominent for young deaf people. Social identity theory[42] suggests that people have both a sense of personal identity (derived from personal characteristics, such as beliefs or skills) and social identity (derived from being a member of certain groups, such as ethnic group, culture, profession). Research on self-esteem and identity suggests that there are important links between these concepts; for example, Phinney[43] describes a number of studies, which investigate the link between a consolidated sense of ethnic identity and psychological well-being.

This brief introduction to identity and self-esteem is aimed at providing a framework for understanding some of the psychological difficulties which are prevalent amongst young deaf people, such as isolation, emotional problems and low self-esteem. Much of the research in this area suggests that the psychological well-being of young deaf people must be considered in the context of identity and group membership.

DEAFNESS AND GROUP MEMBERSHIP

The experience of deafness can be understood in a range of ways and may cause a deaf person to feel part of different groups and communities. A 'medical' model would suggest that deafness is an impairment, which leads to handicap and disability. This implies that deafness should be corrected, for example by using hearing aids to maximise available hearing, and to integrate into the 'hearing' world. The term 'Deaf' with a capital 'D' has come to be used for people who regard themselves as culturally Deaf and part of the Deaf community, as described by Lane.[44] Deafness is viewed positively, and any disability is seen as arising through social, linguistic and cultural phenomena, rather than being located in the Deaf person. The hearing loss itself is not seen as a disability, and sign language allows Deaf people to communicate as freely as hearing people do when using spoken language.

There are long-standing debates[45] about the value of hearing aids to allow integration into the hearing world, versus the use of sign languages to allow assimilation into Deaf culture. Factors such as the school attended, language used, family background and personal choice will influence whether the young person considers themselves to be part of the Deaf community, whether they feel that they are more comfortable identifying with hearing people or whether they are at ease in both communities. More worryingly, it may leave a child feeling left out and alienated from any community, a member neither of the 'hearing world' nor of the 'Deaf world'.

Identity and difference

There is evidence that a well-established sense of identity is particularly important for young deaf adolescents. Research by Israelite et al.[46] found that themes such as fitting in with mainstream society and acknowledging how one is different formed a major part of adolescent experience of orally educated young deaf people. The shared experience of deafness is also one of the factors which contribute to the cohesiveness of the Deaf community.[47] Studies have found that feeling 'different' can be problematic for some young deaf people; Bat-Chava and Deignan,[48] Bat-Chava[49] and Farrugia and Austin[50] found that deaf children in mainstream schools had lower self-esteem than those in Deaf schools. Bat-Chava[49] suggests that this arises because mainstreamed deaf children are a minority group in a hearing school, and this may

cause them to have a heightened sense of being 'different'.[51] Leigh and Stinson[52] found that marginalisation, ambivalence and isolation were prominent in deaf adolescents who attended mainstream schools. Few deaf adolescents in mainstream schools have deaf peers[53] and at a time when self-awareness and difference are so important, they may also experience bullying or teasing from hearing peers.

Being in a mainstream school has not been universally found to result in reduced self-esteem[54] and should not necessarily be considered to be a damaging option for a young deaf person. However, the research above does suggest that it is important to consider the 'fit' between the young person and the chosen school in each case, given their preferred way of communicating, view of self, additional difficulties, family hearing status and so on. Given the influence of peer relationships on psychological well-being,[50] out-of-school groups such as support groups or deaf clubs may be helpful for some children to maintain social links with other young deaf people, or with young hearing people, as appropriate.

Diverse identities

There is a long history of comparing Deaf identity to ethnic identity.[45] In the same way that we might speak of a young person being able to relate to two ethnic groups, it can be helpful to consider the extent to which a young person can identify with both Deaf and hearing people – such as being able to use both sign language and spoken language. Bat-Chava et al.[55] suggest that difficulties in communication are a major contributor to experiences of alienation from hearing peers – for example, a deaf young person with good oral communication skills may integrate with hearing peers much more successfully than a young person who struggles with speaking and listening. Wald and Knutson[56] used the Deaf Identity Development Scale[57] to compare the cultural identity of orally educated deaf adolescents with and without cochlear implants, and found both groups to be similar in endorsing items relating to a bicultural identity, rather than exclusively Deaf or exclusively hearing. Mance[58] found that, for young people with cochlear implants, psychological well-being was strongly associated with attaining a desired identity, whether it be related to being deaf or to being hearing. However, there was a trend for similarity to hearing peers to be highly valued amongst this group, most probably as a result of the oral emphasis on education in this group of young people. Overall, research suggests that young people who are deaf may construct their identity in a range of ways.

CONCLUSION

In summary, we know that successful interaction between parent and child lays the foundation not only for the strong emotional bond between them, but also for developing the child's social interaction skills. From very early on, the baby's lack of response to auditory stimuli, and reliance primarily upon visual input, will influence the way the parents interact with their baby, and vice versa. As the child grows older, language becomes increasingly important for the child in their task of learning about their social world and about how to understand other people. It is clear that language and communication skills play a central role in the social and emotional development of the hearing-impaired child.

Families lay the foundations for the development of healthy, happy children. For parents of the hearing-impaired child, the fundamental task of parenting will be the same as for a

hearing child. However, parents will need to adapt their parenting style and the child's hearing loss will inevitably influence the way that they parent their child. As they endeavour to teach their child the skills needed to integrate into the social world, parents will need to take into account the way that the child's hearing loss and possibly delayed language development will influence family interaction and their style of parenting and behaviour management.

A strong and positive sense of identity and self-esteem as a deaf or hearing-impaired person is important for psychological and emotional well-being. As the child enters school, issues of language use, communication, the type of schooling and the peer group become more prominent influences in the child's life. These become even more important during the formative adolescent years when self-image and self-esteem are key developmental tasks to be negotiated. Attitudes towards deafness within the family, and within society at large, will influence the child's sense of identity and self-esteem. A positive construction of identity as a deaf or hearing-impaired person provides one of the most important protective factors against psychological disorder for the child.

Psychological and emotional factors play a part in the experience of hearing. The psychological impact of hearing loss can be far-reaching. It is important for professionals to hold in mind that psychological issues may well underlie difficulties encountered in the audiology setting, such as: poor behaviour, high anxiety, limited compliance with audiological testing, or non-organic hearing loss. Refusal to wear hearing aids may well reflect a poor self-image and difficulty accepting the deafness on the part of the child and family. Some of these children and families will need additional help to maintain good levels of psychological well-being, and psychological support may be indicated in some cases. An enhanced understanding of psychological well-being in this group will support us in enabling children and their families to experience positive well-being and for young deaf people to grow up into happy and resilient deaf adults.

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26 The education of deaf and hearing-impaired children

I. Tucker

INTRODUCTION

In the first edition of this book, there was a real sense of optimism that the future was exciting to anticipate and that there were real possibilities on the horizon of many more deaf children making much more educational progress. There was the near certainty that universal neonatal screening (UNS) would, at least in the developed countries, and with associated structures, have a major impact on the detection of childhood deafness and ultimately child development. It was Kemp's[1] work that had led the way for the development of screening devices for the identification of hearing impairment in neonates and young children. However, concern was expressed amongst educational professionals that without better educational structures and appropriate training of staff, the potential benefits to child development coming out of early detection would not be taken full advantage of.

There was also the potential for a large-scale switch from analogue to digital signal processing (DSP) hearing aids and the possibility of a rapid increase in the number of children who would be fitted with cochlear implants at ever earlier ages. This earlier fitting of such aids to hearing would therefore stimulate children at a period in life when they were most biologically receptive.

In the first edition, we saw a field still riven by the centuries' old controversy between those who favour the use of manual sign systems as the principal medium through which the curriculum should be delivered and those who favoured an auditory/oral approach and spoken language. But the argument has changed to one which is essentially political rather than educational. Former claims that deaf children educated in systems which use signs will reach a higher level of educational attainment and literacy than those following sign-free approaches are now less widely heard. The most recent evidence, certainly in the UK, shows that by the time they reach the end of their period of compulsory education, deaf pupils educated in a system that focuses on the use of the spoken word, backed up by appropriate and consistent use of the latest technology and with regard to the acoustic environment, are the highest achieving group of deaf pupils.

MEDICAL AND TECHNICAL ADVANCES INFLUENCING SPECIAL EDUCATION

Universal neonatal screening

In the first edition of this text, we signalled ahead the huge potential of UNS programmes to open the door to much earlier intervention with children in terms of both amplification and

the training of parents. Indeed, studies even in the 1980s[2] and before had shown that it was early diagnosis, the fitting of hearing aids and the support for families that was likely to lead to improved linguistic and educational outcomes for hearing-impaired children.

Supported by data from an excellent monograph,[3] and after ministerial approval, a UNS programme was rolled out in phases and became countrywide by the summer 2006. Early data from the UNS programme indicate a median identification age of 2 months with low false positive rates of 1%, similar to those reported in the USA.[4] The coverage reported in the Universal Neonatal Screening Programme (UNHSP)[5] was very high and supportive USA evidence[6,7] suggests only normal levels of parental stress in the process. Neonatal screening for hearing impairment is rapidly becoming just one of the important health tests carried out on all babies.

The effects on language acquisition have also been reported. In two separate studies, the evidence is positive with pre-2-month-old groups scoring significantly higher on both the expressive and receptive language tests than all other age groupings.[8,9] Now, in Colorado, the average age of identification of deafness in children is 2 months, support is state-wide and hearing aids are fitted, where required, by an average of 4 months.[10] We now need to see a range of longitudinal studies post UNHS focusing on amplification, support mechanisms and communication modality.

When Lewis, in a paper on reading achievement,[11] wrote about the underlying tenets of natural aural approaches, she focused on two key features – the maximal use of residual hearing and the need for meaningful input – and yet we see, too frequently, pupils placed in facilities where the support is by relatively untrained staff and where the differentiation of delivery required of the teachers is not embedded in their understanding and practice. Similarly, little understanding of the complexities of the requirements of the acoustic signal needed to reach children's hearing aids, provides a recipe for the linguistic underdevelopment of the deaf child.[12] This suggests that there is a real danger that such children may not achieve sufficient 'intake' of the language around them to reach the necessary thresholds for language acquisition. The publication *Deaf Friendly Teaching*[13] is certainly a good starting point for approaches teachers should consider and for the required infrastructure.[14]

HEARING AID SERVICES AND DSP AIDS

An essential follow on from early detection is the need for appropriate intervention, and along with the UK roll out of the national UNHSP the government funded an NHS modernisation programme for both children's and adults' hearing aid fitting Modernising Children's Hearing Aid Services (MCHAS) and Modernising Hearing Aid Services (MHAS), respectively).

Most professionals in audiology would argue that there appear to be self-evident benefits from the fitting of DSP hearing aids as opposed to analogue versions, particularly in the possibilities of frequency shaping and the signalling processes that enable better performance in noise and offer degrees of control over signal leakage and its resultant acoustic feedback. However, there has been, to date, only a modest amount of research into these benefits. In 2000, the UK NHS National Institute for Clinical Excellence (NICE)[15] concluded 'There is not enough robust scientific evidence to support the nationwide introduction of digital hearing aids at this time.' More recent evidence has been positive though attention has been drawn to the real need for debate and a fresh look at the methodology of research in this area.[16–18]

When service modernisation was undertaken in the NHS – a significant accomplishment – a key decision was to fit appropriate DSP hearing aids to all newly identified children in the MCHAS programme. Children identified and fitted with hearing aids within the first few weeks of life will require new earmoulds every 2−3 weeks during the first year of life; hearing aids should be adjusted using real ear measures for both children[19] and adults.[20]

COCHLEAR IMPLANTS

Cochlear implantation is now a very well-established procedure for providing hearing access in children in the UK. Implantation has proved to be a safe[21,22] and cost-effective[23,24] procedure. More recently, O'Donoghue and Nikolopoulos[21] pioneered a minimally invasive approach and the hearing aid part of the device has also been miniaturised, so that it can be worn as a post-auricular device. Processing strategies are continually being upgraded, FM systems are increasingly linked to implants and an increasing number of children being fitted bilaterally to benefit from the binaural advantages.

Many UK schools do not meet UK standards on reverberation because there is no retrospective enforcement, so only new buildings must comply with recent regulation. However, there remains the problem that implants are as affected by adverse acoustic conditions as any other aid to hearing.

A key development has been ever earlier implantation, with many children now being fitted in the first year of life, taking advantage of greater brain plasticity and a more natural early-language learning environment. This has resulted in implanted children being more likely to be 'educated as severely rather than profoundly' deaf children.[25]

WORKING IN PARTNERSHIP WITH PARENTS AND PROFESSIONALS

It has long been accepted that parents have an important role to play in the education of their children. Parent guidance programmes that had been pioneered by Professor Sir Alexander Ewing and his first wife Lady Irene Ewing at Manchester University were the first to bear fruit, demonstrating that, provided they were given regular support and appropriate training, parents were more effective 'teachers' of their children as far as language development was concerned than professionals.

Formal recognition of the vital role of parents came with the publication in 1978 of the landmark government 'Report of the Committee of Enquiry into the Education of Handicapped Children and Young People'.[26] Paragraph 5.3 states 'In the earliest years parents rather than teachers should be regarded wherever possible as the main educators of their children.' Furthermore, Paragraph 9.1 states that parents are 'equal partners with professionals'. However, the report recognised that most parents would not have all the necessary skills and suggested ways in which they could be helped to develop them. Professor Ian Taylor, then in the chair at Manchester, introduced the first such specialist courses for teachers of the deaf to work with families.

Following Warnock's Report, the Education Act 1981[27] laid down that the special educational needs of children with handicapping conditions that resulted in learning difficulties should be recorded in a 'Statement of Special Educational Needs'. The process was refined

by the Education Act 1993,[28] which also established the Special Educational Needs Tribunal (SEN Tribunal), an independent body to which parents in dispute with their local education authority (LEA) in respect of their child's special educational needs could appeal.

The 1993 Act required the Secretary of State for Education to issue a 'Code of Practice'[29] and revised the Special Educational Needs Code of Practice[30] to provide guidance to all LEAs and governing bodies of schools on their responsibilities to children with special educational needs. In summary, the code advises that a child with special educational needs should have their needs met, that the views of the child (wherever possible) should be sought and taken into account, that parents have a vital role in supporting their child's education, that the special educational needs of children will be normally met in mainstream schools or settings and that children with special educational needs require the greatest access to a broad and balanced education, including the National Curriculum.

One advantage to families in the revised version of the code is that where an LEA decides unilaterally to make a change to provision described in an existing statement of need, parents can insist on the status quo until the decision of the LEA is tested at an SEN Tribunal.

The Statement of Special Educational Needs, which has legal status, is drawn up following a multi-disciplinary assessment of the child's special needs. Parents are involved throughout this process, contributing as they feel appropriate. The Statement has several parts but the key ones are parts 2, 3 and 4, which are the description of the child's learning difficulties and special educational needs, the educational provision that should be made to ensure that these identified needs are met and the school or educational establishment where this provision can be made. Parents have a right to state a preference for the school that they wish their child to attend, and the LEA must comply with that request unless:

1. the school is unsuitable to the child's age, ability, aptitude and special educational needs;
2. the placement would be incompatible with the efficient education of the other children with whom the child would be educated; and
3. the placement is incompatible with the efficient use of resources.

Readers should note that ultimately parents in the UK do not have the right to *choose* a school for their child, only a right to state a preference. If the LEA do not agree and refuse to place the child at the school chosen by the parents, parents may then appeal to an SEN Tribunal. Parents also have a right to appeal against parts 2 and 3 of the Statement if they believe that their child's strengths, weaknesses and special needs have not been properly described, or if they believe that the provision set out to meet those needs is not appropriate. It is a matter of concern to many families that investigations and reports on their child are inadequate and lack the necessary detail in educational psychology reports so important in identifying strengths, weaknesses and needs.

There have been attempts to persuade the government to abandon the use of Statements of Special Educational Need, and the specific support detailed in them, but this has been fiercely opposed and even though appeals to the SEN Tribunal have risen significantly over the years, it is seen by parents as a bulwark against inadequate or inappropriate provision offered by the LEA.

The current government see children with special educational needs as allies to other socially excluded groups and have moved forward to develop measures that would lead to 'inclusion' and equal rights.[31] From the year 2000 onwards, the government have issued many guidance documents in the area of support for children and families, and practical help for professionals.

'Together from the Start' gives practical guidance for professionals working with disabled children (from birth to 3 years old) and their families,[32] and was jointly issued by both the Department for Education and Skills (DfES) and the Department of Health (DoH). It is concerned with the delivery of effective intervention services, good practice and partnership in working to meet the needs of children with disabilities and their families. Two other significant publications put flesh on the bone of this guidance regarding support for families, 'Guidelines on Working with Deaf Children Under Two and Their Families'[33] and 'Developing Early Intervention/Support Services for Deaf Children and Their Families'.[34] These publications came out an opportune moment in terms of supporting the role subsequent to the UNHS programme.

The 'Together from the Start' guidance included supportive information, both child- and family-focused. For families, this information was aimed at support for bringing up the child, understanding the child's situation, enabling informed choice and accessing sources of help. It was the precursor to the 'Early Support Programme' (www.earlysupport.org.uk) which was the implementation vehicle of the Together from the Start programme. It has subsequently become one of the government's most influential and important investments in children with newly identified disability. Indeed the programme, which was very well funded and managed, trained professionals, provided supportive materials for families and utilised very useful tools for support services to audit themselves against. The All Party Select Committee on Special Education called the work 'one of the Government's great success stories'. The Audit Commission[35] however, whilst critical of services overall, saw early support as one of the key levers for delivering service to meet need, to support inclusion and for ongoing improvements.

Every Child Matters[36] and Every Child Matters: Change for Children[37] are a key series of documents underpinning child- and family-related issues. They will influence changes in the UK for many years to come and every minister of relevant departments of state was involved and was required to sign up to the programme for his or her department. This series of documents brought together all the themes that had been emerging over a number of years under the banners of 'Be healthy, Stay safe, Enjoy and achieve', 'Make a positive contribution' and 'Achieve economic well-being'. High-profile child abuse cases and pressure from families of children with disabilities, highlighting poor professional support, have been key drivers for reform.

The Children Act[38] provided the legislative spine to support the programme for the wider strategy of improving the lives of children with special educational needs. The key elements were integrated planning, commissioning and delivery of services, improved multidisciplinary working and improved accountability by inspection.

All of the major developments in the UK in the period since the last edition of this book demand greater professionalism and the availability of more professionals. For example, it is inconceivable that there would not be a need for more teachers to carry out the work effectively, especially when early years are so labour-intensive. Sadly, the numbers of teachers being trained is declining from 153 in 1989 to 91 in 2004. In the year 2000, there were 1,766 full-time equivalent teachers of the deaf.[39] What is perhaps more worrying is that the age profile of the profession is also rising with 72% over the age of 40 and 31% over the age of 50. With an ambitious plan to deliver as much of the educational programme as possible to children with special educational needs placed within the mainstream, the aforementioned data do not bode well for the educational progress of deaf children.

In Removing Barriers to Achievement,[40] the government indicated their support for early intervention, their determination to embed 'inclusive' practice into every school and early years setting, to raise teachers' skills and strategies with children with special educational needs,

and to deliver improvements in partnerships to support them. There is no doubt that much of the strategy was commendable, but the failure of the government to properly enforce staffing levels and specialist training to meet their goals, and also the belated attempts to engage the special school sector in this low-incidence disability, was a serious error of judgement. The outcry from many parents who were feeling that their child's special educational needs were not being met in mainstream schools had MPs and Lords speaking up in the Houses of Parliament about outstanding special schools in their constituencies faced with closure because of the policies of local authorities in 'not sending them children who needed to be there'. Even Baroness Warnock entered the fray with a monograph[41] with statements such as 'There is increasing evidence that the ideal of inclusion, if this means that all but those with the most severe disabilities will be in mainstream schools, is not working.'

Warnock believes that abolishing the old category of the ineducable child has resulted in a failure to arrive at clear definitions of need and the educational requirements to meet them. In the case of deafness, because there is a low incidence and it is fundamentally a communication disability, without necessarily being an intellectual disability, this has led to enormous problems and has undoubtedly led to mainstream schools with insufficient resources to cope with the range and complexity of this special need. Warnock also believes that the concept of inclusion (formerly known as integration, p. 22) has led to confusion over disability need and provision via statements and appropriate funding.

Other reports[35] have also shown disparities and confusion in the current system with many parents failing to get what their child needs, wide variations between LEAs and special schools uncertain of their role.

SEGREGATION, INTEGRATION, INCLUSION

In developed countries, there has been pressure to educate children with disabilities in ordinary schools, and with respect to deafness, this gathered pace in the 1950s with the setting up of special units sited within ordinary schools.

In education, the political definition of 'inclusion' and 'integration' refers to the right of all children, regardless of level of disability, to attend mainstream schools. However, the whole issue is closely linked to human rights: special schools then become defined as 'segregated', and thus second-class. The Centre for Studies in Inclusive Education[42] states 'Inclusion is a fundamental human right for every child and young person' and they regard placement in a special school as a violation of that right.

The implication is clearly that denial of a mainstream place would be equivalent to denial of a human right. Farrell[43] argues that the aforementioned does not sit easily with the 'right' of parents to: 'Express a preference for a special school'[29] and to promote 'inclusion within mainstream schools where parents want it and appropriate support can be provided.'[44]

Parents are far more pragmatic than politicians and would wish to choose the mode of educational delivery they believe would offer the child the greatest opportunities for making educational and social progress and the best chances, either now or in the future, of inclusion into mainstream society. There have been many attempts to establish a definition of inclusion which reflects this. One that comes close to achieving this is that of the National Association for Special Educational Needs (NASEN), who define 'inclusion' as follows:

> Inclusion is not a simple concept, restricted to issues of placement. Its definition has to encompass broad notions of educational access and recognise the importance of catering for diverse needs. Moreover,

> inclusive principles highlight the importance of meeting children's individual needs, of working in partnership with pupils and their parents/carers and of involving teachers and schools in the development of more inclusive approaches. Inclusion is a PROCESS not a state.

Warnock[41] reminds ministers that inclusion is not a matter of where you are geographically, but of where you feel you belong. 'There are many children, and especially adolescents, identified as having special educational needs, who can never feel that they belong in a large mainstream school. We all need to work together to change that perception.'

SPECIAL SCHOOL VERSUS MAINSTREAM

Whilst the government recognise a continuing need for some children to be educated in special schools, legislation and guidance published in recent years have all stressed the desirability of children with special educational needs being educated alongside ordinary children in mainstream schools. The result has been a thrust by LEAs to place children with hearing impairment in ordinary schools and this thrust has been strongly reinforced by financial considerations. Interestingly, in 1994, UNESCO issued a statement on special needs education that has become known as the Salamanca Agreement.[45] The British government of the day was a signatory to that agreement which strongly promotes the concept of inclusion, *except for deaf and deaf/blind persons*: 'Owing to the particular communication needs of deaf and deaf/blind persons, their education may be more suitably provided in schools for such persons or special classes and units in mainstream schools.' As in the Warnock Report, the World Conference recognised the unique nature of deafness as a fundamental obstacle to education.

The low incidence of deafness means that overall numbers are small, approximately 525 severe/profoundly deaf children per year in Britain. In 2000, the British Association of Teachers of the Deaf[39] carried out a survey on the education of hearing-impaired children, with returns approaching 100%. This survey indicated that at that time there were 28,184 children receiving educational support in the age range 0−16+ years. Those in mainstream were 20,081 (71.3%); those in a special school for the deaf were 2,115 (7.5%). Thus, the great majority of hearing-impaired children in the UK *are* educated in a mainstream facility. Comparative figures are over 80% placed in mainstream in Australia[46] and 82% in New Zealand.[47]

In the UK, there is very poor information on the educational outcomes of deaf children educated in the mainstream. Powers[48] reported on a survey of deaf children in LEAs in the UK. He collected information on 1995 and 1996 General Certificate of Secondary Education (GCSE) results. The results achieved by deaf children were poor in comparison with their normally hearing peers.

The government collect education data based on a unique pupil identifier − the 'PLASC' number. When Lord Ashley asked for information about the performance in national examinations of deaf children whilst in mainstream education, the results shown in Table 26.1 were released.

There has clearly been a very significant improvement on the results from the earlier Powers study, which were very low indeed, but it would not be unreasonable to assume that because the majority of these pupils are placed in the mainstream, they have, in general, less disability than those in special schools (see Lewis and Hostler[49] for LEAs that buck the trend). Taken in the round, they still lag behind normally hearing children in the mainstream, and in comparison with a large group of hearing-impaired children in a special school.

Table 26.1 Mean outcomes at GCSE[1] for hearing-impaired pupils in mainstream, for hearing pupils, and for hearing-impaired pupils at Mary Hare School in Newbury (% achieving 5+ A* to C grades).

	2003/2004	2004/2005	2005/2006
H.I. – LEA schools	31.9%	37.9%	? Not reported
Overall national data	53.4%	56.5%	? Not reported
Mary Hare School for the Deaf	87.1%	81.5%	83.3%

[1] General Certificate of Secondary Education, the national examination at 16 years.
Source: Data from DfES.

THE SPECIAL SCHOOL

The type of provision made by special schools for the deaf is, on the face of it, fairly straightforward. Children are educated in small classes of usually no more than 10 pupils, taught by specialist teachers of the deaf, which is a government requirement. Regardless of the communication approach being used, the classrooms are well treated acoustically, with reverberation and noise levels carefully controlled so that hearing aids can be used effectively. The use of personal hearing aids or implants is also frequently supplemented with the use of radio hearing aid systems or headphone amplification systems such as auditory training units or group hearing aids. The latter provide the best form of amplification available since all children are able to hear themselves and every other child in the class at optimum listening levels. There is also considerably less distortion with these systems. Because of the low incidence of severe and profound deafness, most special schools for the deaf have residential provision, and consequently, there is the problem of being segregated from society at large. However, special schools are aware of this danger and some go to great lengths to combat it. This is clearly more difficult where some form of manual communication is adopted by the school for its teaching. Only large cities are able to run a viable day school for the deaf, and even where this is the case, the numbers on roll are small. Several special school closures have occurred during the past decade as a result of falling rolls; in 1998 there were 40, and by 2005, there were only 30 special schools for the deaf in the whole of the United Kingdom.

Schools for children with low-incidence disabilities, such as vision and hearing, have been under severe pressure to even survive, unlike some other special-needs schools which have been growing. It took significant pressure from parents and professionals to gradually shift the wholly 'integrationist' views of the government. This prompted at least a partial rethink on approach resulting in the government setting up a working group to consider the future role of special schools.[50] As a result, some supportive changes were made, and, as a key recommendation of the report, special schools were to be included in 'the full range of new policy initiatives coming from the Department', and 'policy initiatives will be specifically tailored for special schools'. Some of these changes have been rolled out, and leading special schools have received funding to become 'Training Schools', sharing their expertise to raise the skill levels of those working in the mainstream and to actively work in partnership with mainstream schools. Some of these 'Training Schools' are chosen to be 'Specialist Schools' in a particular disability. This has been a significant step forward.

The Mary Hare school in Newbury, already a training school, was invited to become a 'trailblazer' for the Specialist School (Sensory) area. The team there have developed courses specially for mainstream learning support assistants in deafness and vision, and have prepared and run three Masters' degree modules in Early Years and Deafness to focus on newborn to 3-year-olds and their families. The topic areas are Audiology, Family Support and Language Development. Teachers are also trained to become teachers of the deaf, achieving the Teacher Training Agency's mandatory qualification, an MA in Hearing Impairment and an MSc in Educational Audiology. Alongside this, there is a significant amount of advising, mentoring and auditing work in the mainstream. All of the courses, apart from those with assistants, are in association with Oxford Brookes University. Consultancy is provided for the Specialist School Trust managing the roll out of specialist schools in the sensory strand. However, on their own such measures will not ensure that key special schools will be around for the future, since the finance to support the new arrangement is minimal. Any idea that modern special schools are islands of segregation rather than inclusion are incorrect. In the field of deafness generally it has not meant that parents no longer need to appeal to an SEN Tribunal to establish a place for their child at a special school.

Primary special schools have been hit the hardest by the integrationist approach, with falling numbers and closures. On the one hand, it seems somewhat understandable in terms of residential arrangements for younger children. However, here there is a mismatch with early diagnosis and the view that substantial and effective early intervention can mitigate the effects of deafness and potentially allow a more mainstream approach later. The Audit Commission[51] indicates that the reverse is happening and that special school placement grows in the secondary school years (see Figure 26.1).

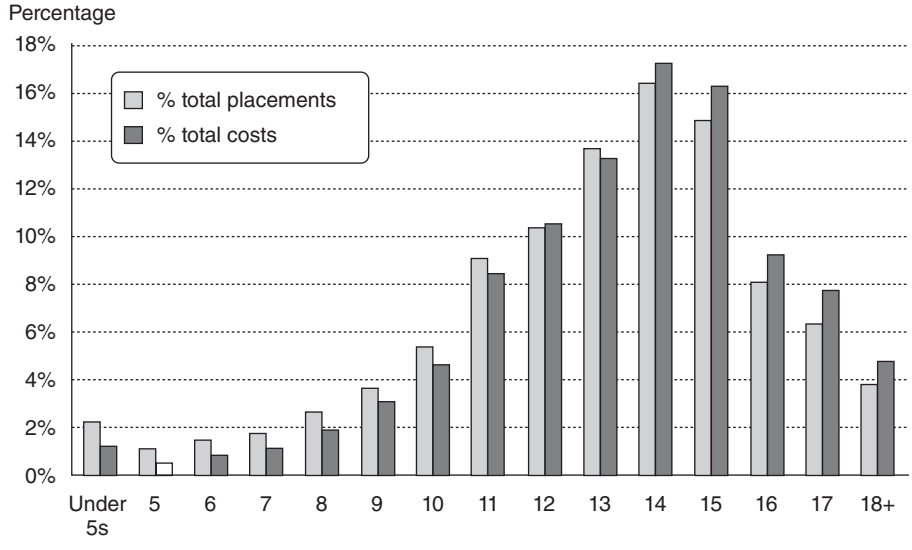

Figure 26.1 Percentage of pupil numbers and costs by age. Pupils aged between 13 and 15 are most likely to be in out of authority placements. *Source*: Regional Partnerships survey of out of authority placement 2006. Reproduced with permission.

MAINSTREAM

Types of mainstream placement are more varied. There are a range of options, for example:

1. Complete integration or mainstreaming within the ordinary class without any supportive help.
2. Mainstreaming with varying levels of individual support.
3. Basing children in a resource room or base unit and integrating them on a part-time basis into the ordinary class. The subjects for which they integrate can also vary.
4. Team teaching by the ordinary teacher and a teacher of the deaf of an integrated class into which is placed one or more hearing-impaired children.
5. Reversed mainstreaming is where normally hearing pupils become part of a class of hearing-impaired pupils. This is less common, but seems to be done more frequently at the nursery end where the educators of the hearing-impaired place high value on the influence of ordinary children and set up a nursery to encourage children with normal hearing to mix with the hearing-impaired. Nursery places are difficult to find in many areas and this seems to be a successful route to integration of such children.
6. Self-contained classes or units from which pupils go to ordinary classes for one or more specific academic subjects.
7. Self-contained classes or units from which pupils go to ordinary classes for one or more non-academic activities.
8. Completely self-contained classes or units with little or no contact with normally hearing peers.

Of these options, mainstreaming with varying levels of support is by far the largest group, four times as many as those in a ‘unit’, a ‘resource’, or a ‘base’. Each type offers a level of flexibility that enables an individual programme to be changed as the individual child’s special needs change. It also allows planners to be more flexible in their use of resources. However, on occasion authorities have found provision difficult because of the low numbers of deaf children; a unit that was once originally thriving may now, because of demographics, have become unsustainable. Conversely, need may arise elsewhere where it is not yet addressed.

Another problem presented by resource units is that often the child has to make a considerable journey away from his or her home area in order to attend. This usually entails a taxi journey and can make it difficult for the child to participate in after-school extra-curricular activities. Also, of course, when he or she gets home, there are no school friends to play with. This can lead to social isolation in the home area, unless positive steps are taken to overcome this.

The advantages are that there is usually a teacher of the deaf on the premises who is able to provide intensive help and follow-up work with language development and in areas of the curriculum with which the child is having difficulty. This teacher is also able to check and ensure that the amplification equipment used by the child is in good working order, or if not arrange for it to be repaired. The specialist resource-base classroom is also often acoustically treated, making it easier for children to use their hearing aids more effectively. As I have mentioned earlier, mainstream classrooms, especially at secondary age, are generally not acoustically treated, and even when using a radio aid system the child may have difficulty in following if background noise levels are not very carefully controlled by the teacher.

Where there is no specialist resource base in school, it may be difficult for a teacher of the deaf supporting a child to find an area that is sufficiently quiet for intensive work to take place. Usually the support from a teacher of the deaf comes from a peripatetic teacher who visits the child in school on a regular basis. The frequency and duration of these visits will vary according to a child's individual needs (and resources available!). Some children will receive daily visits and some only one per term. Part 3 of a child's Statement of Special Educational Needs should be 'specific, detailed and quantified' and should record the frequency and duration of such visits, or the total time allocation per week or per term.

Most children who spend the majority of their time in mainstream classrooms will be provided with some form of individual support, either from teachers who are not qualified to teach deaf children or, more usually, from classroom support assistants. A number of different nomenclatures are used to describe such personnel, from 'teaching support assistants', 'learning support assistants', 'special needs assistant' to 'human aids to communication', but their role is essentially the same. It is to facilitate access to the curriculum for the child. The method of facilitation will vary from child to child and may take the form of simple note taking, pre-tutoring, lip-speaking or a signed commentary on the content of the lesson. The support assistant will also provide differing degrees of help in completing the tasks that children are set to perform. The problems inherent in this system are that the child may have more difficulty developing independent learning skills and may become overly dependent on his or her support assistant. Also, it can mean that the child does not access the curriculum directly but through an intermediary who may know little of the subject matter being taught, which in the secondary school curriculum can be very wide-ranging − anything from Spanish to physics to economics.

Most severely and profoundly deaf children place considerable reliance on their lip-reading ability, but during classroom discussion sessions they may have difficulty in locating the speaker since the amplification equipment they use provides very little directional information. Once they have located the speaker, the moment may have passed. But even if the speaker can be located in time, lip-reading may be difficult because of distance, lighting or angle of view. Also, if a child's own speech is sometimes difficult for his or her classmates to understand, the child may be discouraged from participating actively in the proceedings. Teacher strategies may help to overcome this problem, but they tend to interfere with the pace of the lesson and to destroy its spontaneity.

A big advantage of provision in the neighbourhood mainstream school is that the child has more opportunity to be part of the local community. If he or she has been successful in establishing friendships at school, these friends will probably be around after school. Participation in after-school activities is likely to be less of a problem, and local events that so frequently figure in playground chatter will be more meaningful. *However, this does depend on the attitudes of the mainstream schools towards inclusion*. The focus for educational provision in the UK is the local authority, and moves in recent years to make several unitary (small) authorities from large county authorities can make range of provision and management difficult.

COMMUNICATION APPROACHES

Chapter 23 provides a detailed and excellent evidence base for making choices in communication methodology. However, I would be remiss not to briefly consider this area and interpret it in the light of current approaches being used in the practice of education with deaf children.

There are now three broad categories of communication approach used with young deaf children: the auditory/oral approaches, sign bilingualism and total communication (TC). Each of these groups has variations within it.

TC, which advocates refer to as a philosophy rather than an approach or method, is a loose conglomerate of practices which vary from school to school, class to class and child to child. However, 'TC as generally practised involves the use of speech simultaneously with a sign version of all or part of the spoken utterance'.[52] TC is intended to foster the development of spoken English, with the aim to provide additional information that the child can use to decode the message and to provide a crutch that will facilitate language acquisition. However, unfortunately, the general goal of TC has not been realised possibly due to the interference of the signing that accompanies speech with the spoken pattern, and speech interfering with the fluency and synchronicity of the signing. The vital acoustic patterns of speech are distorted and the comprehensiveness of the sign is lacking. A large survey[53] in the USA demonstrated that in spite of the huge amount of energy that went into the TC programme, the standard of education achieved by students completing their education was no higher than it had been a decade or so earlier, before the introduction of TC.

The BATOD survey[39] does not include TC; it is called Monolingual Bimodal (speaking and signing) and whilst it is now largely discredited as an educational solution to the problems of educating deaf children, it is still used in more than a third of specialist units. Very few TC-educated pupils have successfully developed intelligible spoken language, and their reading ability and educational attainments as indicated by public examination results are poor.

The alternative that is growing in numbers is bilingualism, or now more commonly 'sign bilingualism'. Under this system, a natural sign language, such as British Sign Language (BSL) is used in the early stages of language development, until a measure of fluency is achieved. Then English is taught as a second language. It is however conceded that it may be extremely difficult for a severely or profoundly deaf child who has developed sign language as his or her first language to be given sufficient opportunity to develop hearing well enough for spoken English to be a realistic goal. Therefore, for many deaf children, written English, rather than spoken English, becomes the second language aim. Of course, competence in written English is an essential educational goal, since sign language itself does not have a written form and the ability to read is crucial if the school curriculum is to be accessed effectively.

Some studies[54,55] have indicated that many normally hearing parents have problems in learning sign language and interacting effectively with their children through its means. Young children who attend specialised facilities where sign language is used may quickly come to outstrip their hearing parents in sign language competence. How important this is for the linguistic and more general development of the child has yet to be shown. The figures currently available indicate that 7.5% of severely and profoundly deaf children are following either BSL or sign bilingual programmes in the UK. This number is growing and the political thrust with which bilingualism is being promoted makes it likely that it will continue to do so.

The great majority of congenitally deaf children (95%) are born to normally hearing parents and the great majority of these parents would prefer their children to be able to talk and understand spoken language. It not surprising therefore that the most commonly used communication approach in the UK is natural auralism, which is followed by 44.2% of the severely and profoundly deaf population of schoolchildren. It is used almost exclusively with children with more hearing than this.

It is now widely held that if, from the moment of diagnosis, deaf children are given consistent and appropriate amplification, and a facilitatory linguistic environment where there is

frequent exposure to meaningful spoken language, they will become very competent English-language users by the end of their period of compulsory education. This view is borne out by the evidence of the educational achievements of children educated in this way. In terms of their literacy, they outperform pupils educated through other approaches;[11] for pupils in special schools, the attainments in public examinations, such as GCSE, of those being educated in auditory/oral schools are markedly better than those attending schools that use TC or bilingualism;[56] and a study by the Ewing Foundation in five LEAs[49] has shown that severely and profoundly deaf pupils who have followed a natural aural approach in mainstream schools achieve GCSE examination results that are at par with those of normally hearing children, and that their reading levels are very close to their chronological ages. As well as the educational case, there is a strong sociological argument that if a deaf child can be educated in a way that enables him or her to use spoken English fluently as a means of communication, then social, educational and vocational opportunities are greatly increased.

PRE-SCHOOL EDUCATION

Regardless of the communication approach that parents choose to use with their offspring, the help and support that they receive during the pre-school years are crucial. In many LEAs parents of young deaf children are visited regularly in their homes by peripatetic teachers of the deaf who seek to help parents create a language environment from which the child can benefit. Parents are shown how to check and maintain hearing aids, how to talk to their child, what to say and how to involve all members of the family so that as much interaction as possible takes place. Such work demands a high level of skill. (See Clark[57] for what makes quality interaction.) Once the child is of nursery age, he or she might be placed in a nursery class supported there by peripatetic teachers of the deaf and special-needs support assistants. In many cases, visits to the parents' home would still continue during this period since it is recognised that parents have a vital role to play at this phase.

THE NATIONAL CURRICULUM, CONSTRAINTS, OPPORTUNITIES AND ACCOUNTABILITY

The National Curriculum, which was designed to help raise education standards, was first introduced into schools in England in 1989. It lays down the subject matter that must be covered for all ages of schoolchildren and the standards that they are expected to reach in each. In addition to requiring that the curriculum be broad and balanced, it recognises the importance of three core, now called 'entitlement', subjects: English, Maths and Science. The attainments of all pupils in these core subjects are regularly assessed by the government-administered Standard Assessment Tests (SATs). All schoolchildren have a right to the National Curriculum and this includes children with all degrees of deafness.

In 2004, a modern foreign language, which was initially compulsory became part of an *entitlement area*, i.e. it must be offered, but not necessarily chosen. The government have moved to more pro-actively introduce foreign language into primary schools (compulsory by 2008 to encourage it to be chosen at secondary level). Now compulsory in secondary education, along with English, Maths and Science, are Information and Communication Technology, Religious Education, Citizenship, Work-Related Learning, and Personal, Social and Health Education.

The UK has been criticised and has indeed criticised itself in respect of its vocational education. This has restricted opportunities for students, so in 2005, the government announced a programme of Vocational Diplomas to be offered to 14- to 19-year-olds to try to redress this imbalance.[58] All students, including those who are hearing-impaired, will be able to choose between pursuing general qualifications through a new General Diploma or another diploma with a mixture of general and applied occupationally related skills. The first diplomas will be available in 2008, and by 2010, there will be a total of 14 in a wide variety of vocational areas and designed with the support of businesses.

Standards in education are carefully monitored, so in addition to the specially designed SATs, results of the performance of the pupils in individual schools are published annually to ensure accountability. 'Value added data', national comparisons weighted by social deprivation data and also by a nationally computed 'value added' figure measuring the progress from tests carried out at age 11 years to those at age 16 years, will also be used. From 2002 to 2006, Mary Hare School for deaf children was the top school in the country for 4 of the 5 years for all schools whether special or hearing mainstream.

With the creation of the Office for Standards in Education (OFSTED) (HMSO 1992), all schools, including special schools, are now regularly inspected and reports published as part of a rolling programme. In the opinion of most commentators, the National Curriculum and the 'accountability' measures being undertaken are contributing to raising standards generally. Regrettably, the process frequently highlights the degree of underachievement of some deaf children. This may be uncomfortable knowledge, but at least it raises awareness of the shortcomings of individual schools. There have been significant improvements in 'weak' schools and failing schools are put in 'special measures' with support and further inspection. A poor OFSTED report is a frightening prospect for heads whether mainstream or special school.

POST-COMPULSORY EDUCATION – COLLEGE AND UNIVERSITY

In spite of the lead given in the early part of the 20th century by luminaries such as Thomas Arnold and his pupil Abraham Farrar, the entry into higher education by deaf people was slow in coming. It was really the vision of a famous teacher, Miss Mary Hare, who, having developed a very successful private school, bequeathed in her will all her property to be used for the establishment of a national grammar school for deaf children, that provided the breakthrough. Following her death, a school bearing her name was established in 1946. Since that time, Mary Hare School for the Deaf has striven for academic excellence and today the great majority of its students gain places at British universities for undergraduate courses of study. Many take higher degrees and other post-graduate qualifications. With the growth of numbers of deaf people with such qualifications, the opportunities for careers of a professional nature have greatly increased.

There are also now students from around the country with severe and profound losses who have attended mainstream schools and then carried on to university for a degree course. Support varies according to student need and the quality of the universities provision, but includes grants for equipment and human communication aids such as note-takers, lip-speakers, and sign interpreters.

Whatever the drawbacks of this system, there can be no doubt that the number of deaf people graduating each year from British universities is increasing steadily year by year, and we are moving into an age where more and more well-qualified and well-educated deaf people

are starting to occupy more prominent positions, in spite of what may seem at times a glass ceiling against promotion to the highest levels of business and industry, not only in the workplace but in society as a whole. There is certainly sufficient upward occupational mobility to warrant national research in this area.

CONCLUSION

A recent issue of *New Scientist* (November 2006) began with a headline 'Ear implant success sparks culture war' and a first line 'Could the end of sign language for deaf children be in sight?' It was 'summarising' research showing deaf children implanted pre-one year achieving normal language by age 4−5. Could it be the end of sign language? Not really, since some deaf people regard deafness as a cultural difference rather than a disability and would not have their child implanted, reflecting their informed choice. However, they are a tiny minority since most deaf babies are born to hearing parents who may well never have come across a deaf child before and are likely to choose the implant, if offered.

The relatively small studies on cochlear implants are showing early gains in language, reading and literacy levels, but those with implants show increasing difficulty in these areas as they become more complex. These studies should now become large-scale to enable us to fully understand why some implanted children are very successful in the areas mentioned and others are not. The upward trajectory is only possible with appropriate support strategies involving both parents and professionals. We must pay attention to all the language-related variables, such as language-level-appropriate input and be fully aware of the effect of audiological variables, such as reverberation and background noise, and the effects of teachers not necessarily having clearly defined and embedded practices that aid deaf students. However, structures are gradually being put in place and we can anticipate that these will have the effect of continuing to raise standards.

So, yes, there is good reason to be optimistic as far as the education of deaf children is concerned. The many developments of the past half century and the more rapid development of recent times have changed the education of deaf children out of all recognition. Our growth in knowledge of the language-acquisition process, of acoustic phonetics and how even very tiny amounts of residual hearing can be effectively harnessed; medical advances in the prevention of childhood deafness caused by the rubella virus, rhesus incompatibility, and difficult births, and a greater awareness of ototoxic drugs; the exciting new work on genetic structures and our better understanding of the impact of technology with the development of sophisticated signal processing amplification systems; the micro-surgical developments of implants in the first year of life; our understanding of the critical importance of the acoustic environment and the crucial importance of the parental role, especially in the early years and our absolute determination to better support the hearing-impaired have all contributed to that optimism.

Spoken language, used in conjunction with the latest technology, linguistic knowledge and the latest educational practices, is asserting itself as a practical and effective way of educating deaf children. The future for deaf children today is potentially brighter than it has ever been.

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Index