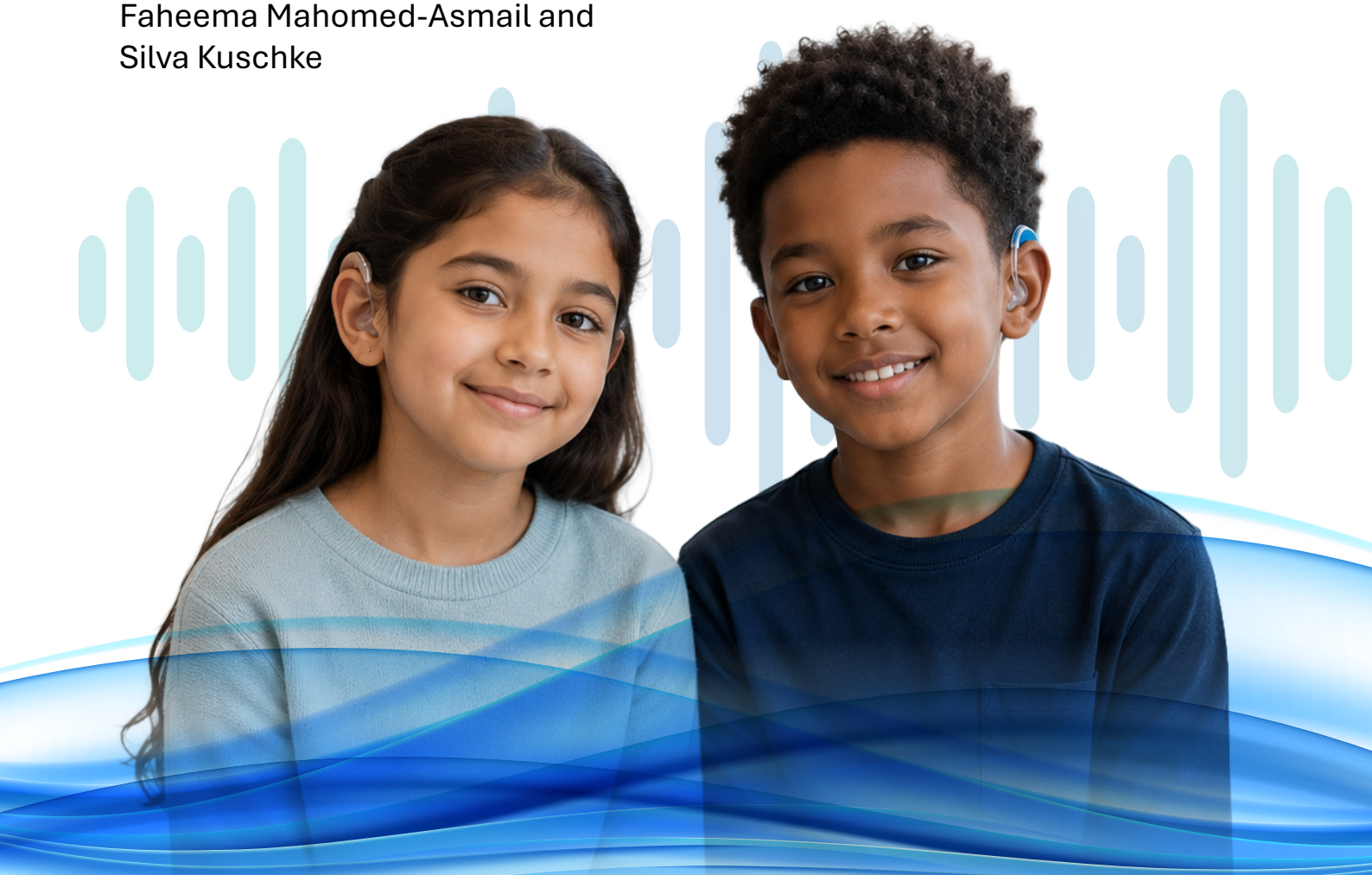
EVIDENCE-BASED PRACTICES IN PAEDIATRIC AUDIOLOGY

Edited by:
Faheema Mahomed-Asmail and
Silva Kuschke



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Foreword

More than 34 million children live with disabling hearing loss, and the majority grow up in low- and middle-income countries where services are scarcest and identification comes last. The consequences of hearing loss left unaddressed in childhood are well documented and lifelong: delayed language, constrained education, narrowed economic opportunity, and reduced participation across the life course. Yet few conditions in child health are as responsive to timely, evidence-based care. Identified early and managed well, most children with hearing loss can develop language, learn alongside their peers, and thrive.

That early identification and intervention matter is no longer in doubt; the harder task has been translating that knowledge into competent, consistent practice, particularly in settings where caseloads are high, resources are limited, and the clinician at the chairside must make sound decisions without the scaffolding of a fully resourced centre. This is precisely the space that Evidence-Based Practices in Paediatric Audiology occupies exceptionally well.

Faheema Mahomed-Asmail and Silva Kuschke have assembled a remarkably complete account of the paediatric audiological journey — from the prevalence and profiles of childhood hearing loss and the anatomy and physiology of the developing ear, through screening, behavioural and electrophysiological assessment, and vestibular and auditory processing evaluation, to surgical, non-implantable and implantable management, aural (re)habilitation, communication and educational options, and the increasingly central roles of telehealth and person- and family-centred care. The contributing authors, drawn from across South Africa and the wider international community, bring both research authority and hard-won clinical experience to every chapter.

What distinguishes this volume is its determination to be useful. The evidence is current and carefully cited, but it is never left abstract; it is consistently translated into practical, real-world guidance that a clinician can apply on Monday morning. The book is honest about context, attentive throughout to the realities of resource-constrained and emerging settings, and grounded in an interdisciplinary philosophy that places the child and family — and the family's choice of language, the language of their heart — at the centre of every decision.

It has been a privilege to watch both editors grow into the scholars and clinicians they are, and a particular pleasure to see them bring this project to fruition. This book will serve students seeking a trustworthy foundation, clinicians seeking a practical reference, and educators seeking a coherent and contemporary text. It will, I am confident, improve the care that children with hearing loss receive — in South Africa, and well beyond it.

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Editor-in-Chief, International Journal of Audiology

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Chapter 1

Introduction To Paediatric Audiology

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'Children are the world's most valuable resource and its best hope for the future. Ensuring their ability to hear and communicate effectively is paramount, for it is through their ears that they will discover the wonders of language, music, and the world around them.'

~Helen Keller

Paediatric audiology

The concept of paediatric audiology began shortly after the development of the discipline of audiology during and following World War II (Tharpe and Seewald 2016). During the twentieth century educators and healthcare professionals across the United States found themselves compelled to participate in the identification, assessment and care of very young children with hearing loss (HL) out of sheer necessity. However, only in the 1950s was paediatric audiology recognised, when Sir Alexander and Lady Irene Ewing introduced some of the fundamental concepts now associated with paediatric audiology: the benefits of early identification and intervention including assistive hearing technology, the importance of caregiver involvement and training at home for the development of speech and language, and the effective approaches for accurate assessment of young children with HL. The profession has developed tremendously since then, with advances in technology for both the assessment and management of HL. However, the goals remain the same: to support children and their families to reach their full potential.

The scope of practice for hearing healthcare professionals (HHPs) includes the prevention of HL, identification and/or screening, assessment and diagnosis of HL and the management thereof through counselling and advocacy (Department of Health 2012). When working with the paediatric population, these aspects need to be individualised for each child based on his or her developmental and/or chronological age (American Academy of Audiology [AAA] 2020). It is thus the responsibility of the HHP to determine the appropriate test procedures to use for each child, after which a diagnosis of and appropriate recommendations for (re) habilitation should be made. The intervention options may be provided primarily by an HHP or in conjunction with an interdisciplinary team of clinicians. Intervention should aim to reduce the impact of the HL and can include non-implantable amplification or implantable amplification devices, communication and language intervention, educational accommodations and support services, auditory training, family-centred intervention and environmental modifications to optimise listening and learning outcomes.

It is also important to note that the journey of (re)habilitation for children is longitudinal, as children are continuously growing and developing. Hearing status and communication needs should thus be re-evaluated every three to six months, depending on the aetiology of the HL. The intervention/management should be adapted accordingly. Furthermore, technology advances rapidly, with new and improved device features being launched on the market regularly. It is thus of utmost importance for HHPs to keep abreast of the latest developments in order to provide the best possible (re)habilitation options.

Hearing loss in the paediatric population

Globally, over 430 million people require rehabilitation to address their disabling HL, of which 34 million are children (World Health Organization, 2021). Hearing loss in children may occur at any stage of development, from the prenatal period through adolescence, and may be associated with syndromic or non-syndromic aetiologies. HL aetiology may be congenital, have a delayed onset or be acquired during one's lifetime.

Hearing loss is considered congenital if identified shortly after birth. Known causes include congenital cytomegalovirus infection, structural abnormalities of the temporal bones, or genetic factors; however, approximately 50% of cases have no clearly identifiable aetiology (Lieu et al. 2020). A delayed onset HL is identified after the neonatal period but is still attributed to any aetiology that was present during birth (Lieu et al. 2020). Acquired HL is attributed to the effects of trauma, infection or ototoxic medications that occur during

childhood. It should also be noted that HL may occur in isolation or be accompanied with other challenges such as vision, cognitive and physical impairments.

The nature of HL, whether permanent or temporary, is usually (though not always) contingent upon its aetiology. If conductive, it is usually temporary in nature and is often associated with conditions such as otitis media (OM) or impacted earwax and is typically addressed through medical interventions. Recurrent conductive hearing loss (CHL) may also mask an underlying sensorineural (SN) component, making it a mixed hearing loss (MHL). It is thus important for clinicians to conduct a full test battery to determine the type of HL and its possible associated aetiology, and to conduct regular follow-up evaluations to ensure that any SN component is not missed. A permanent HL can encompass CHL, MHL and SNHL, and may also include Auditory Neuropathy Spectrum Disorder (ANSD). A breakdown anywhere along the auditory pathway can disrupt the effective use of auditory information. When auditory input cannot be processed effectively, the consequences can be profound if left unaddressed. HL, whether permanent or temporary, and regardless of its nature, requires supportive interventions. Chapter 2 offers an in-depth examination of the prevalence and characteristics of HL within paediatric populations, with Chapter 3 focusing on the auditory anatomy and physiology in particular relation to paediatric HL.

The consequences of untreated hearing loss in the paediatric population are pervasive and multifaceted, affecting the quality and quantity of accessible spoken language, which in turn has downstream effects on speech development, literacy, psychosocial development, mental health, educational attainment and vocational opportunities (Convery et al. 2013). This is particularly relevant given that approximately 90% of deaf and hard-of-hearing children are born to hearing parents and are primarily exposed to spoken language in the home environment (Mitchell and Karchmer 2004). In addition, the effects of paediatric HL extend to the family of the child, often creating a change in dynamics that can subsequently generate tension and additional challenges within the family (Northern and Downs 2014). One of the most effective strategies for minimising or preventing such negative repercussions is the implementation of appropriate and timely intervention (Brodie, Smith and Ray 2018; Paul and Whitelaw 2011; Wang et al. 2019).

Extensive research spanning several decades has described positive outcomes for children whose HL is identified early and who undergo intervention promptly, compared to those with delayed detection and treatment (Antia et al. 2009; Haukedal et al. 2019; Tomblin et al. 2015; Walker et al. 2015; Yoshinaga-Itano et al. 2010). Studies indicate that children who are born

deaf or experience early-onset HL and are identified and receive appropriate interventions within six months of birth demonstrate language development comparable to their hearing peers by age five (Niparko et al. 2010; Yoshinaga-Itano et al. 2010).

There are two main approaches available in supporting the language acquisition of children with HL, which include 1) auditory-verbal (via assistive hearing technology and aural (re)habilitation) and 2) visual-manual (cued speech, signing etc.). A third approach, Total Communication, utilises aspects from both main approaches to facilitate optimal communication on an individual level. Regardless of the approach used, the key outcome for successful development and communication is to give the child and its family the necessary tools and support. The benefit of early intervention will extend to improved social and educational outcomes for this vulnerable population.

Resource constraints

Access to sustainable hearing healthcare resources is a prerequisite for the efficient delivery of hearing healthcare services to address the burden of HL, and improve the quality of life of paediatric patients and their families (Bhamjee et al. 2022; Chadha, Kamenov and Cieza 2021). These resources encompass human resources, equipment, infrastructure and financial assets. In addition, the maintenance and upkeep of these resources are important, for example, the annual calibration of all audiological equipment.

Unfortunately, the vast majority of children with HL live in low- and middle-income countries (LMICs). In these settings, access to services is often limited by barriers such as shortages of HHPs, the high cost of audiological test equipment, limited facilities and restricted access to hearing amplification technologies (Mulwafu et al. 2017; Swanepoel and Clark 2018; Wagner and Fagan, 2013; Waterworth et al. 2022). In South Africa, healthcare is divided between private and public systems, with the majority of the population (approximately 85%) relying on the public sector, where, particularly in low-resourced contexts, hearing healthcare is often not prioritised (Kerr, Tuomi and Müller. 2012; Mayosi and Benatar 2014; Pillay et al. 2020). A study conducted in South Africa focusing on newborn hearing screening (NHS) services within the public healthcare sector revealed that the shortage of HHPs who can conduct hearing screenings, along with limitations in screening equipment, poses significant obstacles to the successful implementation of NHS (Bezuidenhout et al. 2018). As a result, there is limited or delayed early identification of HL in South Africa.

Early identification and intervention are crucial in maximising the language and communication competence, psychosocial well-being and literacy development of children with HL (Joint Committee on Infant Hearing 2019). A large number of infants with HL will thus go undiagnosed and may only be identified later due to parental concerns or only once they enter the school system. Chapter 4 of this book focuses on how hearing screening helps HHPs to identify HL early for optimal developmental outcomes.

Population needs

Besides the lack of resources available, HHP may also face additional barriers such as not being able to communicate effectively with the child and their family due to cultural and linguistic differences. Provision of culturally competent services necessitates understanding the unique and complex set of cultural and linguistic variables that each client brings to the interaction and responding with sensitivity and adaptability to these variables (Lowell et al. 2018). In clinical settings, being responsive to others' needs, having flexible conceptual thinking and adapting traditional approaches are key factors for improving the quality and effectiveness of care across the diverse cultures, languages and ethnicities of our clients. In some settings, telehealth services may be beneficial in providing personalised care and decentralising hearing healthcare services (Chapter 16).

In addition to language barriers, the paediatric population is challenging to evaluate due to their inability to communicate directly with the clinician or their reduced attention span which can make it difficult to obtain reliable behavioural responses. In paediatric audiology, behavioural and physiologic testing methods are used to assess a child's hearing abilities. Behavioural testing relies on measuring the behavioural responses of the child to auditory stimuli. The results depend on the child's co-operation, attention and ability to understand the test instructions. Examples of behavioural testing include Behavioural Observation Audiometry (BOA), Visual Reinforcement Audiometry (VRA) and Play Audiometry. Physiologic testing methods do not rely on the child's behavioural responses. These tests measure physiological responses to auditory stimuli, providing additional valuable auditory information, especially in cases where the child cannot or will not co-operate with behavioural testing. Examples of physiologic testing include Otoacoustic Emissions (OAEs), Auditory Brainstem Response (ABR), Tympanometry and Acoustic Reflexes. There are several behavioural (Chapter 5) and physiologic (Chapter 6) test procedures that can be used as part of a protocol to assess and diagnose the degree and extent of the HL. The contents of this book will support HHPs in selecting assessments and management approaches that align with the child's language, cognitive and emotional development by providing practical recommendations.

Perspectives of this book

This book aims to serve as a comprehensive resource for both aspiring HHPs and practising clinicians. The book focuses on identification, assessment (Chapters 4-8) and (re)habilitation options (Chapters 9-13) for the paediatric population. Information on providing family-centred and interdisciplinary care is also discussed (Chapters 14-17). The figure below depicts the outline of the chapters in this book.

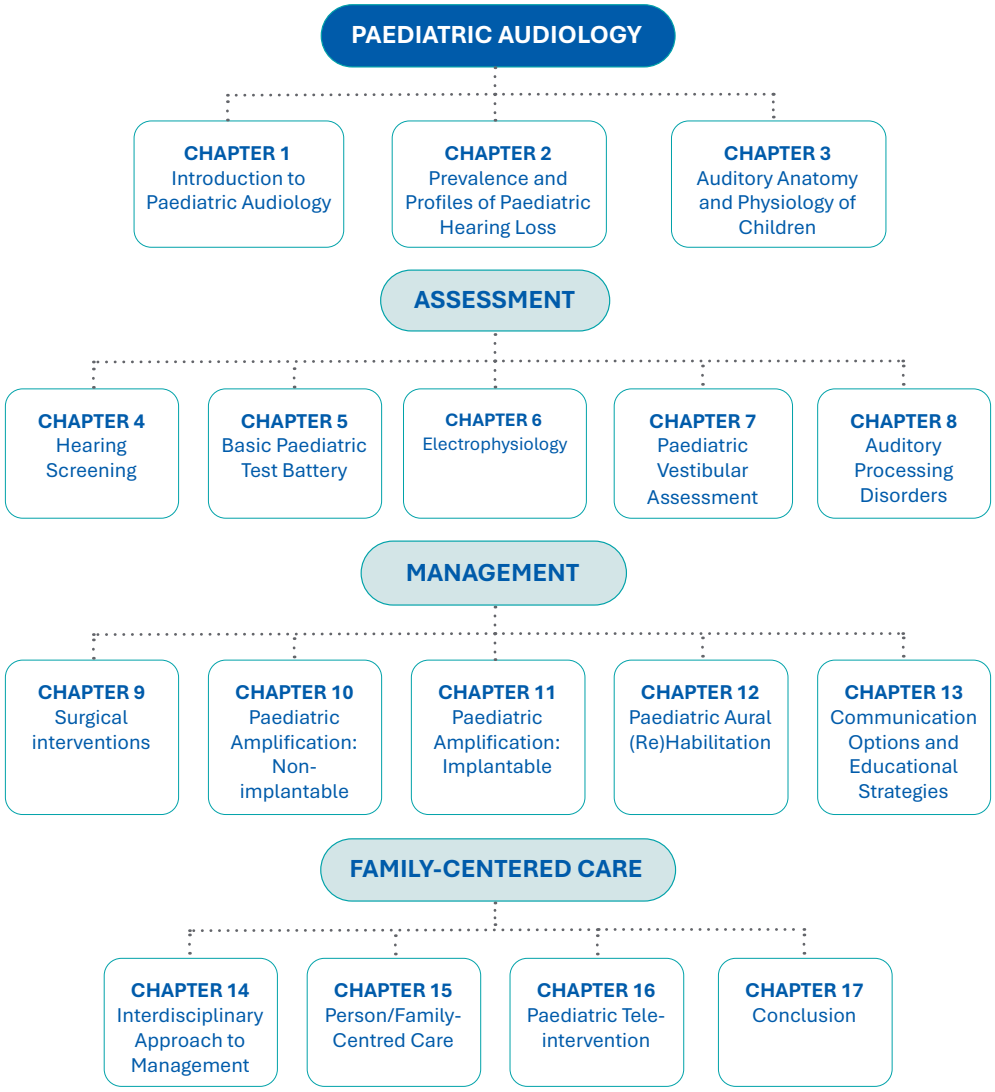

Figure 1.1. Outline of chapters to be included

Written and edited by experienced clinicians, this book aims to offer a unique blend of theoretical foundations and practical insights tailored to the complexities of paediatric audiology. With a focus on real-world strategies, this book strives to enhance clinical decision-making skills and optimise outcomes for young patients and their families.

Conclusion

Hearing care plays a crucial role in the pursuit of universal health coverage, aligning with Goal 3 of the Sustainable Development Goals for 2030 (UN Department of Economic and Social Affairs 2015). This objective aims to provide all individuals, including those with HL, access to services. The realisation of this goal necessitates the adoption of the recommendations of the World Health Organisation (2021), which includes following an integrated, person-centred approach to ear and hearing care (IPC-EHC). Through this book, the reader will obtain theoretical knowledge founded on evidence-based practices and real-world strategies to identify, assess and support children with HL, ensuring accessible and quality care.

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Chapter 2

Prevalence and Profiles of Paediatric Hearing Loss

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"The beginning of wisdom is the definition of terms."

~Socrates

Introduction

Childhood hearing loss (HL) can be described across several domains to build a comprehensive audiological profile. Figure 2.1 illustrates these domains and the potential classifiers within each domain. Certain causes of HL, or aetiology, lead to typical patterns in the resulting audiological profile, which are described later in this chapter.

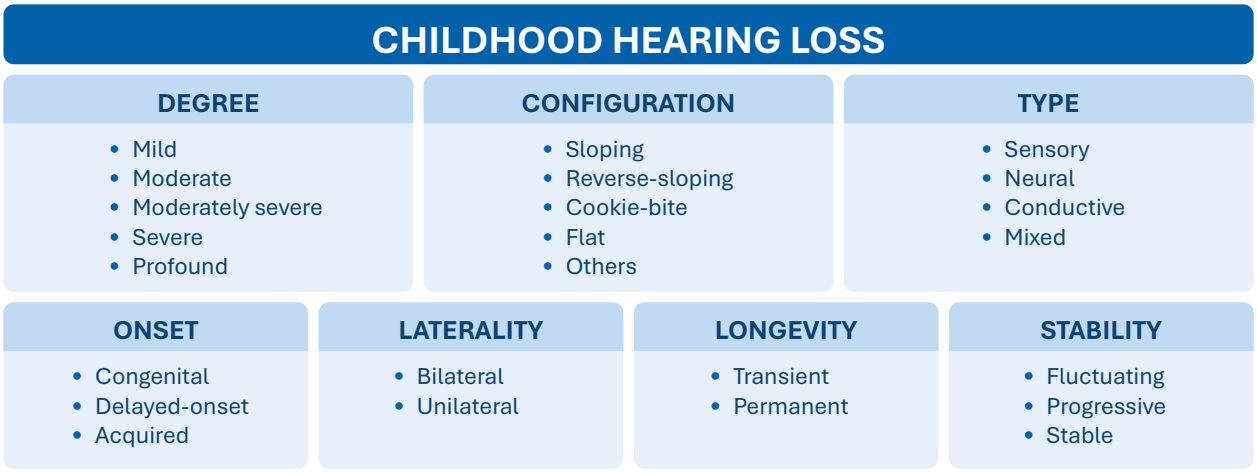

Figure 2.1. Domains of childhood hearing loss, adapted from Mackey (2022)

Describing childhood HL

Firstly, HL can be described according to its degree. Often, the degree is summarised by calculating a pure-tone average (PTA) across key frequencies, which indicates the average degree of HL present. A three- or four-frequency PTA (PTA3 or PTA4) is common, using the frequencies 0.5, 1 and 2 kHz plus 4 kHz for PTA4, though a high-frequency PTA (3, 4 and 6 kHz) may be more appropriate when describing high-frequency HL. In certain configurations, the degree may vary across frequencies. In these cases, it may be more accurate to specify the degree independently for each frequency. There are different upper and lower boundaries for classifying degrees of HL, with the most commonly used criteria outlined by the WHO (2016) (Table 2.1).

Table 2.1. Degree of childhood hearing loss (WHO, 2016)

Degree classification	Range (dB HL)
Mild	20 to 34
Moderate	35 to 49
Moderately severe	50 to 64
Severe	65 to 79
Profound	80 to 94
Complete	95 or more

The onset of HL may be congenital, delayed or acquired. Congenital HL is present from birth, whereas delayed-onset HL appears within the first few weeks, months or years after birth but is associated with a congenital cause. Acquired HL also develops after birth, but results from external causes after birth. Although the exact onset of HL is not always clear during identification and diagnosis, investigating the aetiology may help to clarify its onset and aid in understanding the potential stability of HL. HL stability can be classified based on whether hearing thresholds are fluctuating, progressive or stable. A HL may fluctuate but if it does

not return to normal levels it may result in a progressive HL, which worsens significantly over time. In a study of 330 children, conducted after the implementation of universal newborn hearing screening (NHS), 47.9% of the children were found to have deterioration of thresholds by 10 dB HL or greater in at least one ear, indicating that progressive paediatric HL is a significant issue (Barreira-Nielsen et al. 2016).

HL is further described in terms of laterality, type, duration and configuration. Laterality refers to whether HL is present in one or both ears. Unilateral hearing loss (UHL) affects one ear only, and is typically defined as hearing thresholds of 15 dB HL or better in the unaffected ear, whereas bilateral hearing loss (BHL) affects both ears. The type of HL is classified according to the site of origin, with a sensorineural hearing loss (SNHL) originating either in the cochlea (sensory origin) and/or the auditory nerve and brainstem (neural origin), and conductive hearing loss (CHL) originating in the outer or middle ear. A mixed HL has both a sensorineural and conductive component. The longevity or duration of HL can be classified as either transient or permanent. With transient HL, hearing thresholds return to normal levels over time or with treatment. A transient HL is often conductive in type, whereas permanent HL may either be sensorineural, conductive or mixed. Permanent HL, on the other hand, does not improve, at least with current treatments and technology. Finally, HL configuration is determined by the shape of the HL on an audiogram from low to high frequencies.

Incidence, prevalence and overview of childhood hearing loss

Incidence rates describe the proportion of new cases over a certain time. This is useful to understand how widespread a condition is at certain points in time. Prevalence rates describe the proportion of individuals with a condition at a particular point in time or over a period (for example, one year). Understanding the prevalence of childhood HL is essential for gauging its societal impact, identifying high-risk groups, geographical areas and communities and for effective resource allocation, planning and implementation of prevention, detection, identification and intervention programs. Childhood HL prevalence is typically expressed as a rate per 1000 children or per cent (%), that is, per 100 children. The prevalence of childhood HL varies widely due to the approach used in estimating HL, the geographical region and population and how HL is defined. Childhood HL of any degree and type ranges from three to five per cent according to US data (Su and Chan 2017) and 19 to 30% according to data from

low-and-middle-income countries (LMICs) (Akande and Adeosun 2020; Birhanu et al. 2021; Hussein et al. 2018). The estimated prevalence of HL in African countries is limited due to a lack of audiology services.

Bilateral permanent hearing loss ≥ 40 dB HL (PTA4)

Historically, the criteria of bilateral permanent HL ≥ 40 dB HL (PTA4) have been used to define a 'disabling' HL, despite the fact that mild, transient and UHL can also result in negative effects for children (WHO 2021). Regardless, these criteria are often used when reporting the prevalence of childhood HL. The global pooled prevalence estimates for children from birth to eighteen years are approximately four per 1000 for SNHL alone and nine per 1000 when both SHNL and CHL are included (Wang et al. 2019). After NHS, around one in every 1000 infants has a permanent bilateral HL ≥ 40 dB HL (Bussé et al. 2020). From twelve months to three years old the prevalence increases by approximately 65% (Uhlén, Mackey and Rosenhall 2020), and from three to nine years old it rises by an additional 50% (Ching, Oong and Van Wanrooy 2006). By nine to sixteen years old, the prevalence of permanent bilateral HL ≥ 40 dB HL is at 1.6 per 1000 in high-income countries (Ching, Oong, and Van Wanrooy 2006; Uhlén, Mackey and Rosenhall 2020). In Africa, Asia, the Middle East and Europe, the prevalence of bilateral SNHL ≥ 40 dB HL is higher, estimated at around four to nine per 1000 children from birth to 18 years of age (Wang et al. 2019). This indicates that the growing number of children who pass newborn screening may develop HL during early childhood or school age. Without timely identification and intervention, these children are at risk of delayed speech and language development, educational challenges and reduced social participation, highlighting the need for ongoing hearing monitoring beyond the newborn period, particularly in regions with higher prevalence.

Unilateral hearing loss (UHL)

The prevalence of UHL in infants is 0.78 per 1000 infants based on data from 23 studies, which makes up 25 to 42% of all infants with permanent HL (Bussé et al. 2020). The prevalence of UHL increases by 90% from twelve months to three years of age, a much steeper increase than for children with bilateral HL (Uhlén, Mackey and Rosenhall 2020). From ages three to nine, the prevalence of UHL rises by another 180%. By ages nine to 16 years, the number of children with permanent UHL ≥ 40 dB HL (PTA4) is roughly equal to that of children with permanent bilateral hearing loss ≥ 40 dB HL – around 1.6 per 1000 children in high-income

countries (Uhlén, Mackey and Rosenhall 2020). However, this estimate excludes children with mild UHL who are not eligible for or decline intervention, and children with otitis media (OM) or other forms of transient HL. When considering a broader definition of UHL – including all types and from regions worldwide – the prevalence ranges from approximately two to seven per cent (Wang et al. 2019).

Mild bilateral hearing loss

A mild bilateral HL, from 20 to 40 dB HL PTA4, represents a significant portion of HL among children. Approximately 30% of children with BHL have a mild HL in the better-hearing ear (Mencher and Madriz Alfaro 2000), and the rate of mild HL in childhood may be increasing due to earphone use (WHO 2021). In school-age children in the US, the prevalence of mild bilateral SNHL ranges from one to three per cent (Niskar et al. 1998), and is likely much higher in LMICs, though these data are lacking. Prevalence data for mild HL in infancy are also limited, as most NHS technologies are not sensitive enough to detect HL below 30 to 40 dB HL.

Conductive hearing loss (CHL)

CHL is the most common type of HL in children. Two common causes of CHL – atresia and microtia, and OM. Depending on the cause, CHL may either be permanent or transient. Other causes of CHL in children include wax impaction, foreign objects in the ear canal or malformation of the middle ear ossicles.

Auditory neuropathy spectrum disorder (ANSD)

ANSD is typically a congenital condition characterised by neuropathy or synaptopathy affecting the auditory nerve. The term ‘spectrum’ reflects the variable impact of the disorder. Among children with ANSD, behavioural audiometry often reveals SNHL with auditory thresholds that vary in reliability due to the dyssynchronous firing of action potentials along the auditory nerve. The site of lesion for ANSD varies – dysfunction might exist in the inner hair cells, the synapse between inner hair cells and auditory neurons, different sites along the auditory nerve itself or the myelin sheath (Rance and Starr 2015). A hallmark feature of ANSD is an abnormal auditory brainstem response (ABR) despite functional outer hair cells obtained from intact physiologic measures, including otoacoustic emissions (OAEs) or cochlear microphonics. The main perceptual consequence of ANSD is difficulty with speech perception, particularly speech in noise. Individuals with ANSD have poorer temporal

resolution (that is, detecting onset/offset, changes in amplitude or frequency, and gaps in sounds), which are vital components of speech perception (Zeng et al. 2005). Children with ANSD also have difficulty with localisation of sound that relies on bilateral cues, again due to the de-synchronicity of the signal along the auditory pathway (Zeng et al. 2005).

Prenatal causes of ANSD range from genetic mutations, malformation of the auditory nerve and prenatal infection, such as cytomegalovirus. The otoferlin gene mutation is estimated to be responsible for around 50% of cases of ANSD (Matsunaga et al. 2012). Postnatal associations with ANSD include prematurity, hyperbilirubinemia, hypoxia with mechanical ventilation, ototoxic medications, septicemia and meningitis (De Siati et al. 2020). The association between ANSD and serious complications in infancy is the reason for a higher incidence of ANSD among neonatal intensive care unit (NICU) infants. ANSD accounts for one to ten per cent of childhood HL (as reviewed by De Siati et al. 2020). ANSD in children is primarily bilateral, with 27% showing UHL (Meleca et al. 2020). HL may remain stable or progress, as auditory nerve dysfunction may lead to deteriorating inner hair cell function which can lead to a worsening of hearing thresholds over time (De Siati et al. 2020).

Audiological profiles overview

The following sections outline specific sub-groups of childhood HL and describe their audioThe following sections outline specific sub-groups of childhood HL and describe their audiological profiles according to the domains in Figure 2.1. An exhaustive overview of all categories and aetiology of HL is beyond the scope of this chapter. Instead, the profiles included here were chosen for their particular relevance to the practicing hearing healthcare professional.

Audiological profiles of atresia and microtia

Atresia is the absence or underdevelopment of the ear canal and middle ear structures. As the middle ear develops at about the same time as the outer ear, microtia is almost always accompanied by atresia. Some patients with microtia have what appears to be a normal canal, but which ends in a blind pouch (that is, with no connection into the middle ear). Children with small ear canals can still have normal hearing unless the middle ear is underdeveloped, in which case significant CHL may result. Sensorineural function is usually normal as the cochlea and neural aspects of hearing are unaffected. Chapter 3 provides more details on embryonic development in terms of microtia and atresia.

Microtia and/or atresia occurs in one out of 6,000 to 12,000 births; 90% of cases are unilateral, with the right side being affected more frequently than the left, and with a higher prevalence in males (Hartzell and Chinnadurai 2018). Microtia is seen in higher numbers in Japanese, Hispanic and Native American populations, particularly among Inuit and Navajo children (De la Cruz and Hansen 2004; Murakami and Quatela 2004; Nelson and Berry 1984). Women with four or more pregnancies are at increased risk for having a child with microtia, especially its most severe form, anotia (Harris, Källén and Robert 1996). Parents who have a child with microtia have an increased risk of 5% for microtia in subsequent children. The risk of a child with microtia eventually having a child of his/her own with microtia is also one in twenty.

Thirty-five to 45% of patients have microtia/atresia associated with a syndrome, some of which have an identified genetic mutation (Truong et al. 2022) :

- **Oculo-Auriculo-Vertebral Spectrum,**
- **Goldenhar syndrome and Hemifacial Microsomia,**
- **Town Brocks Syndrome,**
- **Branchial-oto-renal Syndrome,**
- **Treacher Collins Syndrome,**
- **DiGeorge Syndrome (22 q deletion),**
- **Distal 18 q Deletion Syndrome,**
- **Nager Syndrome,**
- **Fibroblast growth factor 3 deficiency,**
- **Lacrimal-auriculo-dento-digital (LADD) Syndrome,**
- **CHARGE syndrome,**
- **Walker Warburg Syndrome,**
- **Klippel-Feil Syndrome,**
- **Meier-Gorlin Syndrome.**

Audiological profiles of otitis media (OM)

OM is another cause of CHL, and poses a major societal burden on school-aged children. OM is defined as inflammation or infection of the middle ear. OM can be divided into different classifications, acute otitis media (AOM), chronic suppurative otitis media (CSOM) and otitis media with effusion (OME). AOM is an ear infection with rapid onset and may be associated with acute pain and fever. CSOM and OME are chronic conditions that can impact the well-being and development of affected children, with consequences for hearing and auditory development.

a) Chronic Suppurative Otitis Media (CSOM)

CSOM is a persistent middle ear infection typically characterised by a perforated tympanic membrane and otorrhea (ear discharge). In children, CSOM typically develops when an AOM infection fails to resolve or when treatment is ineffective. This condition most commonly leads to CHL; however, left untreated, CSOM can also affect the sensory organ, leading to a mixed hearing loss (MHL). While CHL is most common, an MHL is present in 10 to 28% of cases of CSOM (Jensen, Koch and Homøe 2013; Moruskar et al. 2019). Long-term effects of CSOM include ossicular damage, chronic perforated tympanic membrane, cholesteatoma and intracranial infections (Meyerhoff 1988). Globally, there are 4.8 new cases of CSOM per 1000 individuals per year (all ages) (Monasta et al. 2012). The distribution of CSOM cases is concentrated primarily in LMIC (Monasta et al. 2012), as high-income countries have widespread prevention through vaccination and treatment of AOM with antibiotics. Certain ethnic groups, including Indigenous populations in Canada, the US and Australia, are also particularly at risk for CSOM (Bluestone 1998).

A detailed account of surgical intervention procedures is documented in Chapter 9. These options may include topical antiseptics or antibiotics to treat discharge and surgical repair of tympanic membrane perforations. Children with CSOM who do not receive surgery or where surgery did not correct hearing thresholds may benefit from hearing aids (HA). HA may be appropriate if recurrent discharge is not an issue; otherwise, bone-conduction hearing devices may be an effective long-term solution (See Chapter 11).

b) Otitis media with effusion (OME)

OME, also known as serous otitis media (SOM), is characterised by fluid accumulation in the middle ear without an active infection. The defining characteristic of OME is CHL and can occur following upper respiratory infection or AOM. Unlike CSOM, the incidence of OME is high in all countries around the world. The prevalence of OME in eleven African countries was estimated to be eight per cent in a paediatric population in general, and 18% for children under the age of four (Choffor-Nchinda, Bola Siafa and Nansseu 2020), which is similar to the point prevalence found in high-income countries (Martines et al. 2010). Prevalence rates of OME vary depending on environmental and genetic factors. For example, age, season, family size, close contact with other children, exposure to smoke and history of bottle-feeding (instead of breastfeeding) are factors that influence risk of OME (Zielhuis et al. 1989). Down

Syndrome and children with a cleft palate are particularly at risk for OME because of the craniofacial anatomy associated with these conditions.

Not all cases of OME result in HL, and many cases of spontaneous OME may go undetected. Hearing spontaneously returns to levels under 20 dB HL for 50% of cases within three months and for 75% of cases within twelve months (Paing et al. 2025). If an HL is present it is usually conductive in type, with a flat or low frequency configuration, the greatest loss being at 0.25, 0.5 or 8 kHz (Cai and McPherson 2017).

Audiological profiles of hereditary hearing loss

Hereditary HL can be categorised as either non-syndromic (around 70 to 80% of cases) or syndromic (20 to 30% of cases) and accounts for more than 50% of childhood HL (Kochhar, Hildebrand and Smith 2007; Smith, Bale and White 2005). Syndromic HL occurs as part of a syndrome where HL is one of several characteristic symptoms. In contrast, non-syndromic HL results from genetic mutation that causes HL without other symptoms. A comprehensive guide to the genetic origin of HL can be found through a digital resource called GeneReviews®, which is an international peer-reviewed guidebook on inherited conditions such as HL (Adam et al. 1993-2024). The information below provides an overview of some of the most common hereditary aetiology associated with HL.

a) Non-syndromic hereditary HL

Approximately 80% of non-syndromic hereditary HL are congenital, with the remaining 20% having a postnatal onset that may occur months to years after birth (Smith, Bale and White 2005). Non-syndromic hereditary HL can be inherited in autosomal dominant, autosomal recessive, x-linked or rarely, mitochondrial patterns. Autosomal dominant HL typically has a delayed onset, whereas autosomal recessive HL is usually congenital, though there are some exceptions to this trend (Kochhar, Hildebrand and Smith 2007). Both inheritance patterns (dominant and recessive) are associated with mutations in a wide range of genes, leading to SNHL of diverse degrees, configurations and stabilities. Most non-syndromic hereditary HL originates from the inner ear, although certain gene mutations may impact synaptic or auditory nerve function. Despite the heterogeneity in audiological profiles for non-syndromic hereditary HL, if the gene and type of mutation are known, the profile of HL is often quite predictable (Snoeckx et al. 2005).

Around 50% of autosomal recessive HL cases are linked to mutations on the GJB2 gene, which encodes Connexin 26, a protein essential for cochlear function (Estivill et al. 1998). With over 150 possible mutations in GJB2, the resulting HL profiles vary widely. However, the most prevalent mutation of the GJB2 gene mutation leads to a congenital, bilateral, severe-to-profound SNHL in 90% of patients, while the remaining 10% experience mild-to-moderate HL (Snoeckx et al. 2005). X-linked non-syndromic HL also presents distinct audiological profiles depending on the specific mutation. Mutations may lead to a mixed HL with a conductive component due to stapes fixation, a congenital profound bilateral HL or a delayed-onset high-frequency progressive SNHL (Kochhar, Hildebrand and Smith 2007).

b) Syndromic HL

In terms of syndromic hearing loss, there are an estimated 400 syndromes which are associated with childhood HL (Gettelfinger and Dahl 2018). Examples of syndromes which are directly associated with HL include Jervell and Lange Nielsen, Treacher Collins, Brachiootorenal, Pendred syndrome, Waardenburg syndrome and Usher syndrome, and others. These syndromes typically involve HL due to abnormal prenatal development of the auditory system. More rarely, syndromes or other inherited diseases may result in ANSD. Syndromes associated with CHL typically result from outer or middle ear malformations, or secondary to craniofacial abnormalities, such as a cleft palate.

Usher syndrome (Kimberling et al. 2010) affects approximately 11% of children with HL and is associated with vision loss and, in some cases, vestibular dysfunction. Usher syndrome has three types, each with a distinct audiological profile (Table 2.2). Type II is the most common, comprising two-thirds of cases, while Type I accounts for about one-third, and Type III is only frequent in populations of Finnish and Ashkenazi Jewish ancestry (Millán et al. 2011).

Table 2.2. Types of Usher syndrome, audiological profiles, and vision loss

Usher Type	Hearing loss				Vestibular function	Vision loss onset
	Degree	Configuration	Onset	Stability		
1	Severe-to-profound	Flat	Congenital	Stable	Impaired	Childhood
2	Moderate-to-severe	Sloping	Congenital	Stable	Normal	Childhood / adolescence
3	Variable	Variable	Congenital	Progressive	Variable	Variable

Children with Pendred syndrome typically present with bilateral, congenital SNHL, which is often profound at birth or progresses over time to profound. In rare cases, HL is milder or manifests later in childhood (Cremers et al. 1998). Pendred syndrome is often associated with malformations in the inner ear, including an enlarged vestibular aqueduct in 80 per percent of cases (Fugazzola et al. 2000). Besides HL, children with Pendred syndrome have deficient iodine processing which may cause an enlarged thyroid gland (goiter) in some cases and/or hypothyroidism.

Waardenburg syndrome accounts for approximately two to three per cent of congenital SNHL cases (Nayak and Isaacson 2003). HL varies from mild to profound. About ten per cent of children with Waardenburg syndrome do not have HL (Song et al. 2016). Over 90% of children with HL due to Waardenburg syndrome have stable HL, with the remining ten per cent experiencing progressive loss (Song et al. 2016). Besides HL, the syndrome is also characterised by pigmentation anomalies in the iris (for example, heterochromia), a white forelock and dystopia canthorum, where the inner corners of the eyelids are abnormally wide apart.

Down Syndrome, though not inherently including HL as a defining characteristic, is commonly associated with a high risk for CHL. The increased risk is due to a combination of the anatomy of the outer ear, a higher risk of respiratory illness and a weakened immune system in children with Down Syndrome. CHL may result from cerumen impaction, OME or ear disease. Due to narrow ear canals, middle-ear evaluation is challenging, complicating the ability to diagnose and treat OM.

Audiological profiles for congenital infections

Congenital infections are a significant cause of childhood HL. Infections can be transmitted in utero or during birth, passing from the mother to the baby. The severity of symptoms typically depends on the mother's existing immunity level, as well as when and how the virus was transmitted to the baby. Congenital infection is a significant cause of SNHL, particularly in LMICs. Table 2.3. summarises the audiological profiles of children with SNHL associated with congenital infection.

Table 2.3. Typical audiological profiles among children with SNHL associated with congenital infection, and other common co-occurring symptoms

Congenital infection	Degree	Laterality	Onset	Stability / Duration	Other symptoms
Cytomegalovirus	Severe-to-profound (> 70%), otherwise mild, moderate	Commonly bilateral if at birth (> 70%); varies if delayed onset	Commonly at birth (> 70%), delayed onset (18–20%)	Permanent; stable (> 50%), progressive or fluctuating (40–50%).	Varies from none to multiple (microcephaly, seizures, speech and language delay, balance impairment, intellectual disability, vision loss)
Toxoplasmosis	Varies, though most mild-to-moderate	Unilateral or bilateral	At birth; delayed onset possible depending on treatment course, though no cases are reported	Permanent, stable. No cases of progressive hearing loss reported in literature.	Varies from none to multiple (vision loss, intellectual disability, seizures, psychomotor delay, jaundice)
Herpes simplex virus	Varies from mild-to-moderate to profound	Unilateral or bilateral	At birth; delayed onset possible though none reported	Permanent or recoverable. No cases of progressive hearing loss reported in literature.	Varies but can include inflammation, vascular damage, neurologic disability, and hearing loss
Rubella	Varies from mild-to-moderate to profound	Mostly bilateral, some unilateral	Mostly at birth, some delayed onset	Permanent; mostly stable, progressive in 15%.	Varies but can include growth delays, cataracts, deafness, congenital heart defects, problems with the development of other organs, problems with mental development and learning
Syphilis	Varies from mild-to-moderate to profound	Bilateral	Delayed and sudden onset	Permanent; stable or fluctuating	Varies but can include blindness, deafness, neural system problems, deformity of the face
Rubella	Varies from mild-to-moderate to profound	Mostly bilateral, some unilateral	Mostly at birth, some delayed onset	Permanent; mostly stable, progressive in 15%.	Varies but can include growth delays, cataracts, deafness, congenital heart defects, problems with the development of other organs, problems with mental development and learning
Syphilis	Varies from mild-to-moderate to profound	Bilateral	Delayed and sudden onset	Permanent; stable or fluctuating	Varies but can include blindness, deafness, neural system problems, deformity of the face

a) Congenital cytomegalovirus (cCMV)

cCMV is the most common preventable cause of congenital HL worldwide. Approximately 13% of all cases of childhood HL are caused by cCMV (Goderis et al. 2014). The global prevalence of cCMV is around six per 1000 live births, though rates vary from two per 1000 in some countries to as high as 50 per 1000 in some LMICs (Manicklal et al. 2013). The difficulty with cCMV infection is that only ten per cent of children show any symptoms of cCMV infection at birth, and symptoms may develop over time. Among those born positive for cCMV infection, with or without symptoms at birth, around 13% have or will develop HL (Goderis et al. 2014). A significant percentage of children have delayed-onset, progressive or fluctuating loss, making it very important that hearing thresholds for children with cCMV are closely monitored. Children with cCMV may present with a complex constellation of long-term challenges besides HL, and intervention planning within a multidisciplinary team is warranted.

b) Congenital toxoplasmosis

Congenital toxoplasmosis has a global incidence rate of around 1.5 per 1000 births, though rates vary widely by region. Countries in South America, the Middle East, and Africa have incidence rates over three per 1000 births, while in high-income countries, congenital toxoplasmosis is extremely rare (Torgerson and Mastroiacovo 2013). HL occurs in up to 26% of cases of congenital toxoplasmosis and in all reports is present from birth, though theoretically could be delayed onset due to the nature of the illness (Brown et al. 2009). Over a quarter of children with congenital toxoplasmosis show abnormalities in retrocochlear or central auditory pathways with normal cochlear function (de Resende et al. 2010), though it is unclear how these early clinical signs relate to later auditory development.

c) Congenital herpes simplex virus (HSV)

Congenital HSV is a very common in the general population, though congenital infection is rare, with global prevalence around 0.1 per 1000 live births, ranging from 0.03 per 1000 in Southeast Asia up to 0.2 per 1000 in the Americas (Looker et al. 2017). It is estimated that around 23% of children born with HSV have HL (Dahie and McCollister 1988). The audiological profile of children with congenital HSV varies widely, from mild-to-profound and unilateral or bilateral. Similar to other congenital infections, HL is typically present at birth, though theoretically HL may emerge the first few weeks after birth along with other symptoms (Dahie

and McCollister 1988), though no cases of delayed-onset HL have been reported in the literature. Children with congenital HSV often exhibit central nervous system involvement, which may impair auditory brainstem function and central auditory processing systems (Dahie and McCollister 1988).

d) Congenital rubella

Once a common infection, congenital rubella has become much less common due to immunisation efforts. Prior to widespread implementation of the rubella vaccine, the WHO estimated a prevalence of congenital rubella at four per 1000 births, a figure that has since declined by 97% (WHO 2024b). In countries with high vaccine coverage and acceptance, such as the US, congenital rubella is extremely low, approximately one case per five million births (Papania et al. 2014). However, in low-income countries with limited vaccine coverage, congenital rubella remains prevalent, with rates of around one to two per 1000 births (WHO 2024b). As with other congenital infections, the severity of HL varies, though severe-to-profound degrees are most frequently observed, often presenting as a flat or slightly sloping audiometric configuration (Wild et al. 1989). HL is typically present at birth and remains stable, though some reports indicate delayed onset or progressive HL within the first few years of life in around 15% of cases. BHL is the most common presentation, with around 30% of affected children exhibiting UHL (Wild et al. 1989).

e) Congenital syphilis

Congenital syphilis is another congenital infection that is transmitted from mother to fetus before or during birth; however, unlike other infections it can be effectively treated with antibiotics. Pregnant mothers can be screened for syphilis early in pregnancy, and adequate treatment can be provided if the infection is detected. For untreated mothers, infants may be born with congenital syphilis. The incidence of congenital syphilis varies widely across the globe, primarily due to differences in access to prenatal screening and treatment. In countries with available treatment and comprehensive prenatal care, the rates of congenital syphilis are less than 0.05 per 1000 births. In contrast, in some countries in South America and Africa, the rates are two to ten cases per 1000 births (WHO 2024a). HL caused by syphilis is not typically present at birth but develops years afterwards with a sudden onset. Late congenital syphilis is typically associated with a triad of symptoms called Hutchinson's triad – dental stigmata (distinct deformities in the teeth), interstitial keratitis (eye inflammation) and

SNHL. Children identified early with congenital syphilis can start antibiotic treatment which restricts disease progression. However, once symptoms develop, they cannot be reversed.

Other causes of childhood HL

In addition to congenital and genetic causes, children may develop HL from acquired conditions or exposures including bacterial meningitis, exposure to ototoxic medications, such as aminoglycosides and certain chemotherapy agents or recreational noise exposure. These types of HL highlight the importance of monitoring and early intervention to reduce long term auditory and developmental impacts.

Audiological profile of meningitis

Bacterial meningitis is an infection that occurs when bacteria invade the central nervous system. Among children who contract meningitis, the survival rate is approximately 70%, and of those who survive, around 50% develop complications (Hasbun 2022). The most prevalent bacteria causing meningitis differ by age group and vary across countries with high- versus low-vaccination coverage and acceptance. Consequently, the incidence of meningitis in LMICs is about 350 times higher than in high-income countries, where rates in children are very low, around 0.004 per 1000 children (Zainel, Mitchell and Sadarangani 2021; Zunt et al. 2018).

The likelihood of developing HL after meningitis also varies by region. With prompt treatment, the progression of meningitis can be halted, though some survivors still experience HL. The audiological profile associated with meningitis varies but typically presents severe-to-profound SNHL (59 to 74% of cases), and approximately 26 to 41% of cases have mild-to-moderate HL (Koomen et al. 2003; Kutz et al. 2006). HL is bilateral in 31 to 53% of cases and unilateral in the remaining children with HL (Koomen et al. 2003; Kutz et al. 2006). In most cases of HL, the degree is typically stable over time; however, exceptions have been reported. HL onset is during or shortly after infection.

Audiological profile for ototoxicity

Certain drugs used to treat life-threatening illnesses can damage the inner ear, resulting in SNHL. Aminoglycoside antibiotics, such as gentamicin, and certain chemotherapy agents,

like cisplatin, are the most common ototoxic medications. Approximately 60% of patients undergoing such treatment experience effects on the auditory system from ototoxicity (Knight, Kraemer and Neuwelt 2005). The extent of ototoxicity on the auditory system varies according to dose, duration of administration, age of the child and individual susceptibility.

The audiological profile of ototoxicity is typically characterised by bilateral high-frequency SNHL. The loss typically starts with extended high frequencies (above 8 kHz) and may progress to lower frequencies as treatment continues. HL may appear shortly after treatment begins or may be delayed in onset. On average, it takes around 135 days before HL is observed in the standard audiogram frequency range, with a slightly shorter onset period in younger children than in older children (Knight, Kraemer and Neuwelt 2005). In most cases, hearing continues to deteriorate over time, potentially even years after treatment ends (Knight, Kraemer and Neuwelt 2005).

Audiological profile for recreational noise-induced HL

As children reach adolescence, HL from recreational noise exposure becomes a concern, especially in cultures where youths frequently listen to their personal listening devices for extended hours. Prolonged exposure to high levels of music can contribute to a noise-induced threshold shift, characterised by an SNHL at 3, 4 and 6 kHz, with at least 15 dB better hearing in the low frequencies and at least 10 dB better hearing at 8 kHz (Berger et al. 2000). The notched configuration deepens with time and continued exposure. Quantifying the prevalence of SNHL caused by such exposure is challenging due to limited longitudinal studies and the inconsistent findings on a direct link between sound exposure and auditory function. However, data from the US show that around 13% of youths aged twelve to nineteen have high-frequency hearing thresholds of 15 dB HL or greater, and 17% of youths have a notched configuration in their audiograms characteristic of noise damage, though some are still within the normal hearing range (Henderson, Testa and Hartnick 2011). In addition to HL, recreational noise exposure may increase the risk of permanent tinnitus, with a reported prevalence of 15% in adolescents (Gilles et al. 2012).

Audiological profiles for other auditory-related conditions

Besides the profiles of childhood HL represented within the domain framework in Figure 2.1, other types of auditory conditions warrant attention, as they are commonly encountered in

clinical practice. Two of these conditions include: auditory processing disorder (Chapter 8) and non-organic HL.

a) Auditory processing disorder (APD)

Auditory processing disorder (APD) is a condition that affects a child's ability to understand what he or she hears. It is characterised by deficits in the neural processing of auditory information, despite normal hearing sensitivity. Individuals with APD may experience difficulties in accurately perceiving, discriminating and interpreting sounds, especially with speech that is produced in complex listening environments. The prevalence of APD in children has been estimated at two to seven per cent in US, UK and NZ populations (Bamiou, Musiek and Luxon 2001; Chermak and Musiek 2007). When taking into account co-morbidity with other developmental disorders with which APD occurs, APD prevalence in children is approximately ten per cent (Brewer et al. 2016). Higher prevalence rates of nine to eleven per cent have been reported in Poland (Skarzynski et al. 2015), and in 8- to 12-year-old children in Iran (Moloudi et al. 2018).

Early intervention is optimal, and informational counselling should be provided following diagnosis. Management can include dichotic training, listening in noise and spatial stream separation, computer-based auditory training, music training, remote microphone hearing aid systems, audiobooks, phonological therapy, language therapy and communication, cognitive and compensatory strategies (see Chapter 8 for management considerations).

b) Non-organic HL

Non-organic HL occurs when the audiometric thresholds recorded during behavioural audiometry do not reflect true hearing levels. This type of HL may be intentionally feigned, or it may be non-intentional – an unconscious manifestation of HL due to a psychological condition. Regardless of the underlying cause, it is essential to recognize the audiological profile of non-organic HL to avoid misclassifying it as a physiological HL and pursuing inappropriate or even harmful interventions for a child with normal hearing thresholds.

The audiological profile of non-organic HL often appears as a sudden or rapidly progressive HL with a flat audiometric configuration. Behavioural response reliability is frequently poor, and speech audiometric thresholds may not align with those expected based on the

audiogram. Importantly, objective measures, such as otoacoustic emissions and auditory evoked potentials, indicate that auditory function is better than the results suggested by the behavioural audiogram. Management of non-organic HL involves a careful, multidisciplinary approach. Referral to a psychologist or other mental health professional may be appropriate when the HL appears to have a psychogenic component, particularly in cases associated with stress, anxiety, or emotional factors. Education, counseling and reassurance are key strategies, as most children with non-organic HL can regain normal functional hearing once the underlying psychological or behavioural issues are addressed.

Conclusion

Each of the seven domains of childhood HL could affect treatment and outcomes, involving different options or combinations of options, which may also change over the life of the child. Additionally, each domain can affect the reporting of HL in childhood. The chapter includes information about causes of childhood HL, as well as some available treatments. As medical treatments including vaccines are developed, the prevalence of HL and causes of HL in children can change.

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Chapter 3

Anatomy and physiology of paediatric hearing and vestibular structures

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'The human body is the most perfect work of art.'

~Leonardo da Vinci

Introduction

The audiovestibular system is a marvel of biological design, intricately tuned to facilitate the perception of sound and equilibrium from the earliest stages of development. This chapter aims to provide a comprehensive overview of the anatomical and physiological aspects of paediatric hearing and vestibular function, emphasising the complexities of the outer, middle and inner ear. Special attention will be given to the development pathways of the auditory and vestibular nerves. Additionally, the integration of sound processing within the central auditory and vestibular systems will be explored to support the subsequent discussions on assessment methodologies, diagnostic criteria and intervention strategies to support paediatric auditory and vestibular health.

Embryology of the Audiovestibular System

Embryology is a biological discipline dedicated to studying the development of an embryo from the time the ovum is fertilised until it progresses to the foetal stage. A human pregnancy lasts approximately 40 weeks, with the gestation period divided into three trimesters. Each trimester is characterised by distinct developmental milestones and physiological changes. The first trimester spans from week one to week twelve, the second trimester from weeks

thirteen to 26 and the third trimester from week 27 until childbirth. By the conclusion of the first trimester, the foundational structures of the encephalon, or human brain, have begun to take form.

Pharyngeal arches, pouches and clefts are structures that develop in the embryonic stage of human development and are involved in forming various head and neck structures, including the face and ears (Figure 3.1). Inner ear malformations result from developmental arrest at various time points in embryology. Understanding the relationships between anatomical structures is essential for diagnosing developmental disorders.

3.1. (a)



3.1. (b)

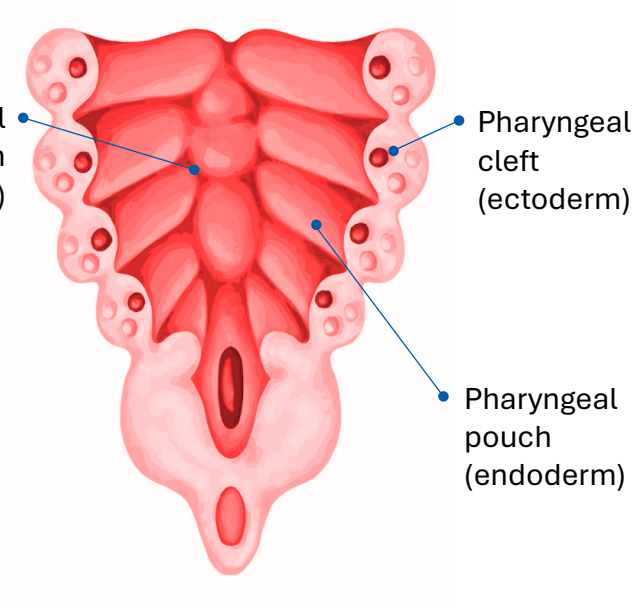


Figure 3.1(a): Five week old embryo
Figure 3.1(b): Pharyngeal arches, clefts and pouches

Temporal bone

The temporal bone is a complex structure of the skull that plays a critical role in housing and protecting the auditory and vestibular systems. It comprises several distinct components: the squamous, petrous, tympanic (cartilaginous) and mastoid parts, each of which has unique embryological origins and follows a specific timeline of development during gestation.

Week 4-8: The temporal bone's development occurs. It comprises several distinct parts, each developing from unique embryonic origins.

Week 5: The petrous part, which houses the inner ear, develops. It encases the auditory and vestibular apparatus, and also houses the internal auditory meatus. Ossification begins around week 20.

Week 6: The squamous part, which originates from the prechordal mesoderm, forms the lateral aspect of the temporal bone. It provides support and protection for the underlying structures of the skull and houses compounds of the cranium.

Week 7: The tympanic part develops from a cartilaginous component of the first pharyngeal arch. It helps to form the external auditory meatus (EAM).

Week 24: The mastoid part arises from the temporal region during the latter half of the embryonic period and develops an air cell system that becomes significant postnatally.

Outer ear

The outer ear comprises the pinna (auricle) and the EAM. By the fourth week, the formation of the auricular hillocks begins. The 'Hillocks of His' are six protrusions from the first and second pharyngeal arches (Figure 3.2a). They include the tragus, helix crux, helix, anti-helix, antitragus and lobule and inferior helix of the pinna which are seen by the sixth week. The hillocks gradually fuse to create the rudimentary structure of the pinna, which detaches from the head by the eighteenth week, achieving adult morphology, not size, by 22 weeks (Figure 3.2b).

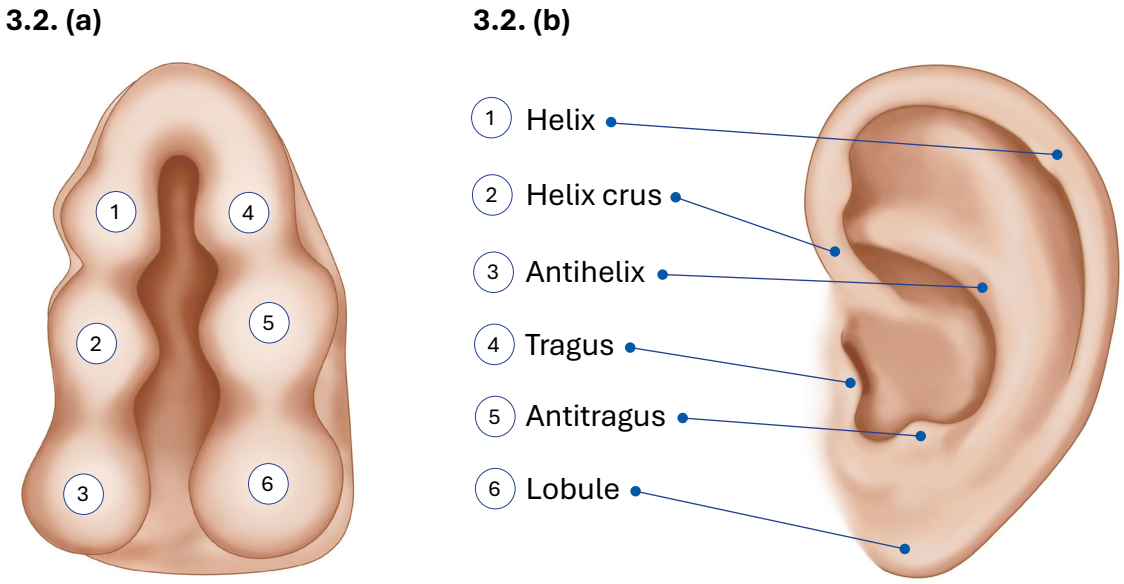
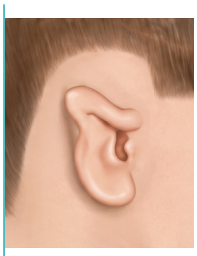
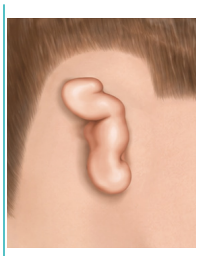


Figure 3.2: (a) The six hillocks of His, (b) The six hillocks of His forms: 1) Helix, 2) helix crus, 3) Antihelix, 4) Tragus 5) antitragus and 6) lobule of the pinna

Microtia, a congenital ear deformity characterised by an underdeveloped external ear, ranges from a small or absent pinna to a partially formed ear structure. Table 3.1 shows how microtia can result from disruptions in development during the sixth to eighth week (Ishinoto et al. 2005). By the sixth week, the EAM develops from the first pharyngeal cleft, which elongates and narrows over time. This canal remains unossified and embedded in mesenchymal tissue during early development. During development, the EAM closes around the eighth week, only to start re-opening in the second trimester. At 21 weeks, it is fully open.

The sebaceous and modified apocrine glands, responsible for the production of cerumen, begin their differentiation at approximately five months gestation, coinciding with the emergence of hair follicles in the outer segment of the external canal.

Table 3.1: Microtia due to disruptions in development, week six to eight

Appearance					
Grade	Normal	*Grade 1*	*Grade 2*	*Grade 3*	*Grade 4*
Description	Normal size of all parts	*External ear is smaller than average. All parts are present. Minor defects.*	*Outer ear is partially formed and small*	*Some of the auricular structures are present but no recognised ear parts*	*Complete absence of the external ear (anotia)*
Time of occurrence	Week 21	*Week 6-7*	*Week 6-7*	*Week 6-8*	*Before week 6*

Middle ear

The ossicles originate from structures derived from the first and second pharyngeal arches. The malleus and incus arise from Meckel's cartilage, which is associated with the first arch. This cartilage is crucial in forming mandibular structures and is a precursor to the malleus and incus. The stapes is derived from Reichert's cartilage, which belongs to the second pharyngeal arch. These structures develop throughout pregnancy, as follows:

Week 4: Formation of the tympanic cavity from the ectoderm of the first pouch.

Weeks 6-8: Formation of the tympanic membrane (TM). Chondrification of Meckel's cartilage,

followed by the ossification that gives rise to the malleus and incus.

Week 7: Development of the Eustachian Tube (ET) – completed by Week 16.

Week 16-17: Formation of the Malleus and Incus.

Week 18: Stapes formed.

Week 26: The ossification of the ossicles is complete (Scheuer and Black, 2000).

Inner ear

The inner ear begins forming at the end of the third week of embryonic development, culminating in the development of the cochlear duct's two and a half turns and the three semicircular canals by the end of the eighth week (Jackler, Luxford and House 1987). It is marked by a thickening of the ectoderm at the cephalic end, leading to the formation of the otic placode. This structure is genetically programmed to develop into the membranous labyrinth. As the process continues, the placode invaginates to create the auditory pit, which later becomes encircled by mesoderm, forming the otocyst by the end of the fourth week (Figure 3.3a shows the Otocyst formation).

The otocyst differentiates into different inner ear structures: The ventral part transforms into the saccule and cochlear duct, while the dorsal part develops into the utricle, semicircular

canals and endolymphatic duct. In the fifth week, the saccule begins to lengthen and coil, forming the cochlear duct. By the sixth week, the semicircular canals begin as folded evaginations from the dorsal side of the otocyst, with the superior semicircular canal forming first. By the end of the sixth week, the vestibular system develops into the rest of the semicircular canals, and the cochlear portion continues to elongate. By the end of the eighth week, the cochlear duct has completed two-and-a-half turns.

Figure 3.3: (a)

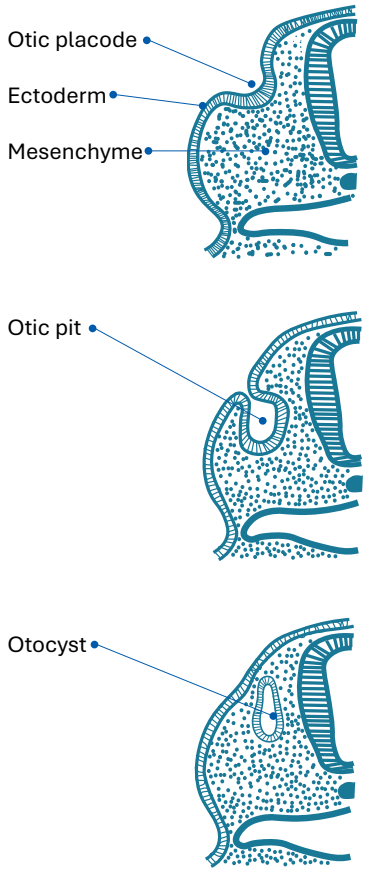


Figure 3.3: (b)

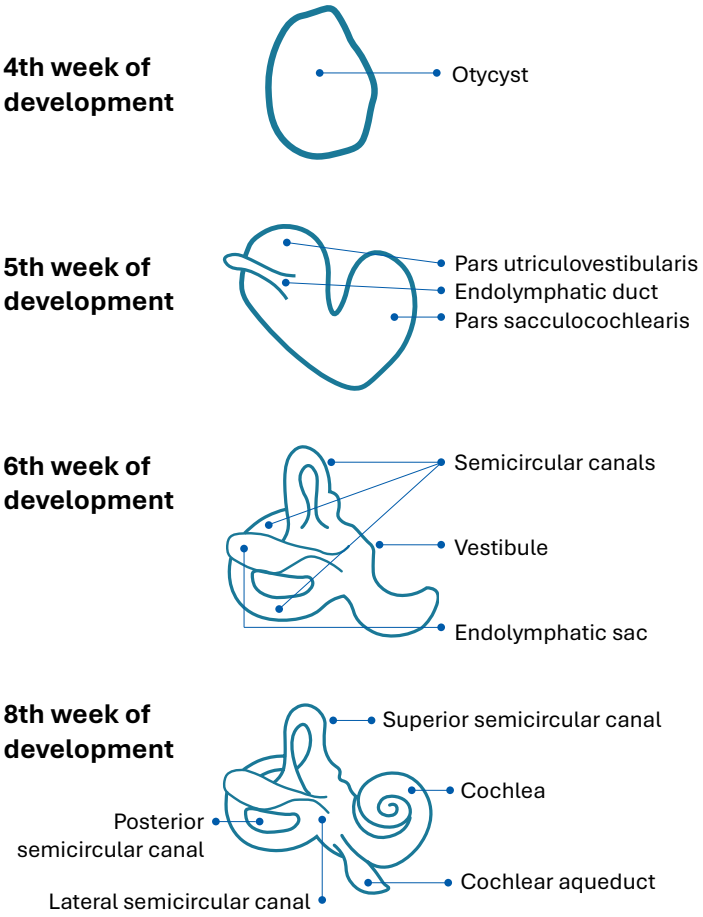


Figure 3.3: (a) Inner ear formation from the 3rd week, developing the otic placode Figure 3.3: (b) The Otocyst further differentiates to form two and a half turns of the cochlear duct and three distinct semicircular canals of the vestibular portion by eight weeks of gestation. Source: Dhanasingh (2022)

In the seventh week, the epithelial cells of the cochlear duct differentiate into the Organ of Corti. Initially similar, these cells eventually form inner and outer ridges, with the inner ridge becoming the osseous spiral lamina and the outer ridge developing into inner and outer hair cells. These hair cells interact with the tectorial membrane, while spiral ganglion cells migrate and form a ganglion along the coiled cochlea. By the tenth week, surrounding mesenchyme transforms into cartilage, which vacuoles to create the cochlea's scala vestibuli and scala tympani. The inner ear reaches near-adult size by the end of the fifth month, achieving total cochlear growth within six months.

The vestibular and cochlear aqueducts arise from the otic placode during the third week of development. The cochlear aqueduct, a narrow channel connecting the cochlea to the subarachnoid space of cranial cavity, forms from the posterior aspect of the developing cochlea, typically emerging around the sixth week of development. The vestibular aqueduct arises from a distinct region of the otic vesicle, connecting the vestibule to the posterior cranial fossa. By the eighth week, this structure is well-formed. Although most of the membranous labyrinth is adult-sized by twenty weeks of gestation, the vestibular aqueduct continues to grow throughout embryonic life (Pyle 2000).

Malformations in the inner ear can vary significantly, depending on the timing of developmental disruptions (Table 3.2). By studying the embryological development of the inner ear, HHPs can identify at which stage differentiation has been interrupted, thus aiding in treatment decisions regarding cochlear implants and other interventions (Dhanasingh et al. 2021) (Figure 3.4).

Vestibulocochlear nerve

The spiral ganglion, which develops from the neuroblasts originating from the otic vesicle, forms the auditory aspect connected to the cochlea. Ganglion cells from the auditory nerve emerge from the otocyst in the fourth week, and their fibres enter the cochlea by the fifth week, spreading parallel to its curvature. Simultaneously, the cristae ampullares in the semicircular canals develop from the neurogenic epithelium of the ampullae (Anniko and Nordemar 1982). By the end of the eighth week, the vestibulocochlear nerve is structurally established and ready to relay sensory information to the brain. Connections to inner hair cells form between the 11th and 12th weeks, while outer hair cells make contact at the apex by the 14th week. Synapses are fully formed by the seventh month, with maturation occurring in the last trimester, allowing the foetus to perceive sound outside the womb by the 28th week (Querleu et al. 1988).

The internal auditory meatus develops through inhibited cartilage formation at the otocyst's medial aspect, a process reliant on the presence of the vestibulocochlear nerve (Amarnath et al. 2016). Cochlear nerve deficiency is the most common cause of single-sided deafness in neonates (Yoshimura et al. 2024).

Brainstem nuclei and tracts of audiovestibular function

The brainstem nuclei and tracts of audiovestibular function refer to the specialised clusters of nerve cells and associated pathways within the brainstem that are responsible for the processing of auditory and vestibular information. Stages of development include:

Week 4: The oculomotor nuclei originate from the mesencephalon and are crucial for controlling eye movements. These nuclei mature as sensory input from the developing inner ear becomes functional. By the end of the week, the hindbrain differentiates into the metencephalon and myelencephalon. It is essential for developing pathways for the vestibulo-ocular, vestibulo-colic and vestibulo-spinal reflexes necessary for maintaining balance and co-ordination.

Week 5: The brainstem's vestibular and cochlear nuclei develop from the rhombencephalon, specifically the caudal hindbrain. The oculomotor nuclei are positioned and increasingly organised, reflecting their specialisation for motor functions. Concurrently, the medial longitudinal fasciculus (MLF), a crucial neural pathway for co-ordinating eye and head movements, begins to develop (Ahsan, Riley and Schubert 2007).

Week 6: The integrity of cranial nerve pathways begins to establish, allowing the vestibulo-ocular reflex (VOR) to take form.

Week 8: The cochlear nuclei, trapezoid body, superior olive, lateral lemniscus and inferior colliculus can be identified, establishing foundational connections for sound processing and vestibular function. The connections between the vestibular system and the spinal cord give rise to the vestibulo-colic and vestibulo-spinal reflexes. These structures mature further, allowing for the refinement of auditory and vestibular pathway function.

Vestibulocerebellum

The vestibulocerebellum is an integral part of the cerebellum. It plays a vital role in maintaining balance and co-ordinating eye movements. Its intricate connections with associated

brainstem nuclei and pathways facilitate vestibular processing and motor co-ordination. The development occurs as follows:

Week 4: The neural tube is fully formed, and will eventually differentiate into the central nervous system. As development progresses, the metencephalon begins to give rise to the cerebellum.

Week 6: Precursors of the cerebellar structures are established, including the vermis and the flocculonodular lobe, which will contribute to the vestibulocerebellum (Amore et al. 2021).

Thalamus, auditory and vestibular cortex

The thalamus, auditory cortex and vestibular cortex are integral components of the brain's sensory processing and motor co-ordination systems. The thalamus serves as a crucial relay station, processing and transmitting sensory information. The auditory cortex is primarily responsible for the perception and interpretation of sound. The vestibular cortex, associated with the vestibular system, is involved in maintaining balance and spatial orientation. Stages in development include:

Week 4: The auditory cortex begins developing when the formation of the inner ear structures primarily drives the auditory system.

Week 5: The thalamus begins its development (Antón-Bolaños, Espinosa and López-Bendito 2018).

Week 6: The otic placode develops into the otic cyst.

Week 10: The thalamus is organised into its future nuclei (Sheridan and Tadi 2024).

Week 10-12: The auditory cortex in the superior temporal gyrus appears as connections are made to the thalamus's medial geniculate nucleus (MGN). The MGN is responsible for processing auditory information, particularly in distinguishing pitch and frequency. Neurons in the MGN project target specific regions of the auditory cortex, allowing for precise localisation of sound sources and decoding of complex auditory stimuli, such as spoken language or music.

Week 16: The central vestibular cortex is formed. The central vestibular cortex, located in the parieto-occipital area, receives input from the thalamus's ventral posterior nucleus (VPN). This development lays the groundwork for spatial orientation and balance by establishing links with visual and proprioceptive systems, stabilising vision during motion and enabling posture and movement co-ordination.

Table 3.2: Inner ear malformations

Condition	Description	Time of development
Michel's deformity	Complete labyrinthine aplasia, absent internal auditory canal arrest of the otic placode.	*3rd week*
Cochlear aplasia (CA)	Absence of the cochlear portion while the vestibular portion may be partially formed.	*Late 3rd week*
Common cavity (CC) malformation	Cystic structure lacking distinct cochlear and vestibular differentiation.	*4th week*
Incomplete Partition Type I (IP type I)	Separable cochlear and vestibular structure but lacking the modiolus.	*5th week*
Incomplete Partition Type II (IP type II)	Cochlea displays cystic characteristics, often accompanied by a dilated vestibular aqueduct, potentially risking fluid leakage during electrode insertion.	*7th week*
Incomplete Partition Type III (IP type III)	*Cochlear structure absent from the central modiolus.*	*Genetically inherited disorder linked to the X chromosome*
Enlarged vestibular aqueduct (EVA)	*Abnormal dilation of the vestibular aqueduct.*	*6th to 12th week*
Cochlear hypoplasia	*Arrest in differentiation of semicircular canals of ves-tibular portion and the elongation of cochlear duct.*	*6th week*

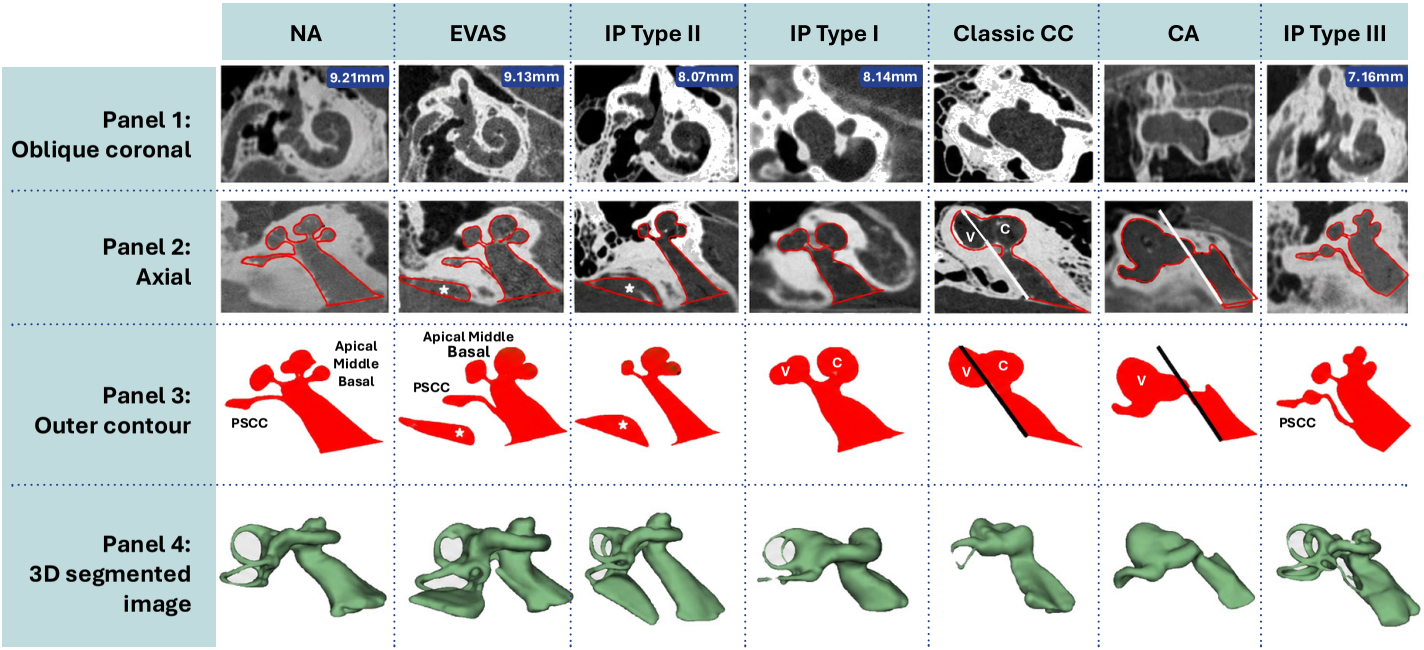


Figure 3.4: Different inner ear malformations. Computed tomography (CT) oblique coronal view (panel 1) and axial view (panel 2). The captured outer contour filled with red (panel 3). The straight line drawn parallel to the posterior edge of the internal auditory canal differentiates the cochlear (C) from the vestibular portion (V). The enlarged vestibular aqueduct (*) seen in enlarged vestibular aqueduct syndrome (EVAS) and the incomplete partition (IP) type II. The 3D segmented images (panel 4). Adapted from: Dhanasingh et al. (2021).

Anatomy and Physiology of the Paediatric Audiovestibular System

Temporal bone

The temporal bone is a paired structure located on the lateral sides of the cranium. It plays a vital role in protecting the delicate structures of the inner ear and houses the auditory and vestibular systems, which are essential for hearing and balance. At birth, the temporal bone is anatomically distinct from its adult counterpart. The newborn's temporal bone comprises smaller, softer, malleable bone structures, accounting for the rapid adaptability needed for the delivery process.

Figure 3.5: (a)

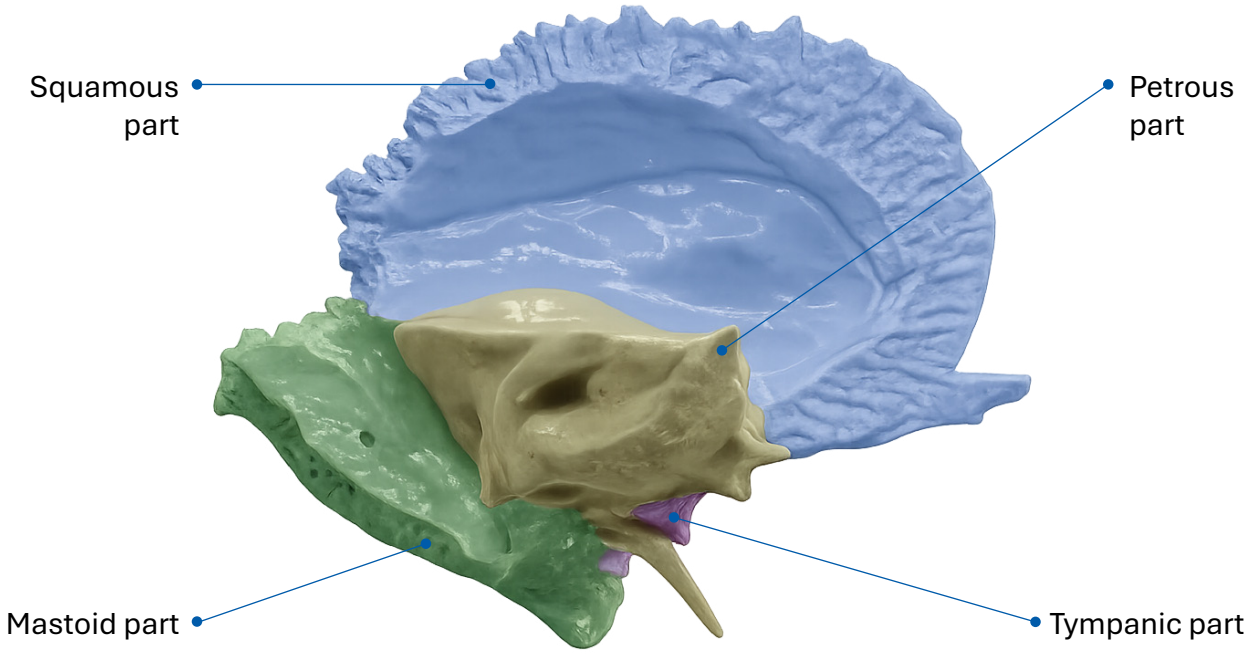


Figure 3.5: (b)

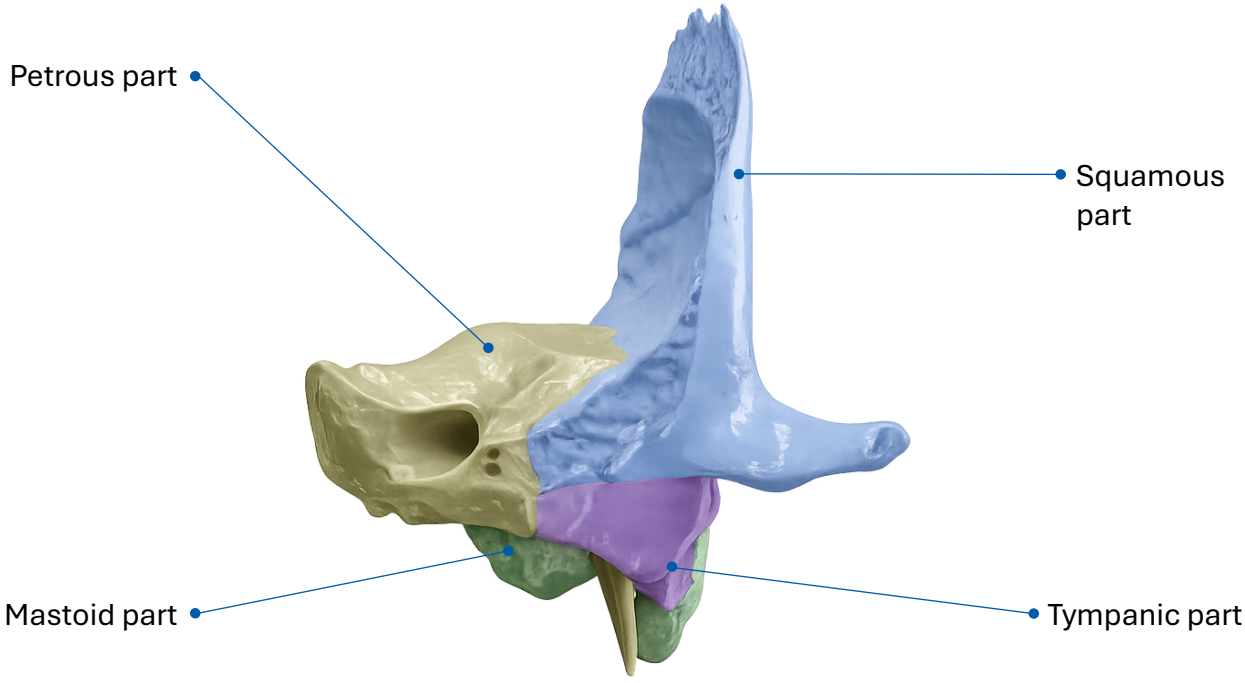


Figure 3.5: (a) The parts of the temporal bone, a coronal view
Figure 3.5: (b) The parts of the temporal bone, a sagittal view/plane

Each temporal bone is anatomically divided into four parts: the squamous, mastoid, petrous and tympanic parts (Figure 3.5). The squamous portion is flat and contributes to the side of the skull, while the mastoid portion houses the mastoid process, which serves as an attachment point for neck muscles. The mastoid process is not fully developed at birth. The petrous part contains the intricate structures of the inner ear, including the cochlea and semicircular canals. The tympanic part forms the external acoustic meatus, allowing sound waves to travel to the eardrum.

The temporal bone also plays a significant role in various functions, ranging from jaw movement to cranial stability. Its association with the temporomandibular joint facilitates the complex mechanics of chewing and speaking. Additionally, the temporal bone's proximity to

vital cerebral structures underlines its importance, making it essential for anatomical integrity and physiological function.

Outer ear

The pinna is the visible part of the outer ear that protrudes from the side of the head. It consists of skin-covered cartilage. In newborns, the pinna is relatively large in proportion to the head, compared to adults. While it is still developing, the overall structure is present at birth. The pinna has a distinctive shape resembling a funnel that helps capture sound waves from the outside world and directs them into the ear canal. This aids in sound localisation, allowing newborns to determine the direction from which sounds are coming. The pinna also assists in amplifying specific frequencies of sound, which helps with auditory localisation. The pinna acts as a natural barrier that helps protect the inner structures of the ear from dirt and foreign objects. Its shape and orientation help minimise the risk of injury or infection.

The external auditory meatus connects the outside world from the pinna to the middle ear space. Anteriorly, it forms the posterior border of the temporomandibular joint, which means movements during feeding or sucking can impact the canal's function. Medially, the canal leads to the TM, and posteriorly, it is bordered by the structures of the mastoid process. The proximity of these structures can influence the spread of infections and the mechanics of sound transmission.

In adults, the ear canal measures approximately two and a half to three centimetres and features a slightly S-shaped curvature. In contrast, the external auditory meatus of newborns is notably shorter, measuring one to one and a half centimetres. Its shape is more linear and horizontally oriented, demonstrating less curvature compared to adults. This anatomical immaturity reflects the ongoing development of the auditory system during infancy. The cartilaginous portion of the ear canal is softer and more flexible in infants, potentially increasing vulnerability to ear infections. This developmental disparity necessitates unique considerations in clinical assessments and interventions for paediatric patients.

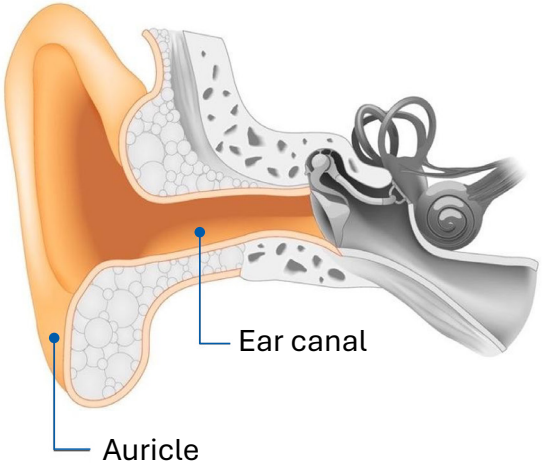
The external auditory meatus is covered with skin-lined epithelium characterised by keratinised and non-keratinised squamous cells. This layered structure aids in the consistent turnover and protection of the ear canal. Ceruminous glands in the outer portion of the meatus are modified apocrine glands that, together with sebaceous glands, are responsible

for the production of cerumen, commonly known as earwax. Cerumen is a natural defence mechanism, trapping dust, foreign particles and microorganisms, preventing them from reaching the TM. The pH of the meatus is slightly acidic, ranging from 4.5 to 5.5. This acidity plays a crucial role in maintaining a healthy environment by helping to inhibit the growth of harmful bacteria and fungi. The natural secretions of the ear, including cerumen, have antimicrobial properties and contribute to the acidic environment, acting as a protective barrier against infections.

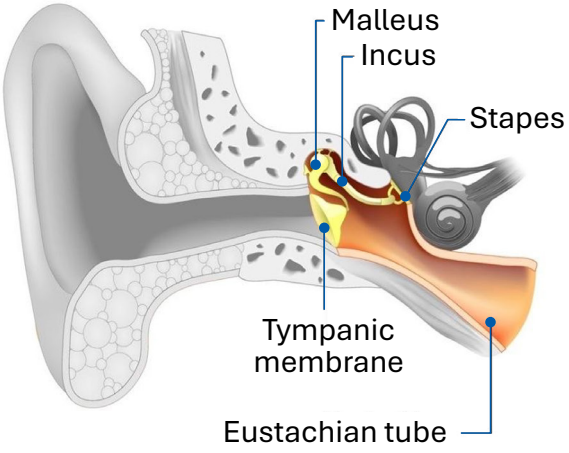
The external auditory meatus has a natural self-cleansing mechanism that helps to keep it free from excess wax, debris, and bacteria. This self-cleaning process involves the production of cerumen, which gradually moves from the inner part to the outer part where it can eventually be washed away or fall out. The movement of the jaw while talking or chewing also aids this process, assisting in the natural expulsion of cerumen.

The resonance frequency of the external ear canal is within the 2 kHz to 3 kHz range, coincidentally aligning with human speech's most essential components. This resonance enhances auditory perception, allowing neonates to perceive and differentiate sounds critical for early language development and social interaction. The small size of the canal, coupled with its specific geometry, accentuates certain frequencies, fostering an optimal auditory environment for the infant.

OUTER EAR



MIDDLE EAR



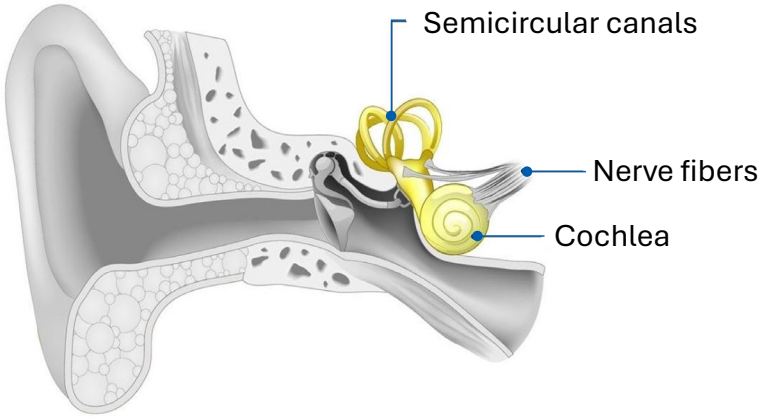


Figure 3.6: The outer, middle and inner ear from the front

Middle ear

The middle ear consists of the middle ear cavity, TM, ossicles, ET opening and the mastoid connection (Figure 3.6). The middle ear cavity is an air-filled space in the centre of the temporal bone. It is situated between the external auditory meatus and the inner ear, functioning as a channel through which sound is transmitted, and air pressure is regulated. The middle ear is well-vascularised and lined by mucosa with secretory properties. Although the ET performs this function primarily, the mucosa also helps with gas exchange in the middle ear. Its blood supply primarily originates from branches of the maxillary and stylomastoid arteries. Innervation is provided by branches of the facial and glossopharyngeal nerves, with sympathetic fibres also present. The TM is the barrier between the external auditory meatus and the middle ear (Figure 3.7). It converts sound waves into mechanical vibrations. In newborns, the size of the TM is smaller than that of older children and adults.

A smaller and thinner TM in newborns influences their auditory capabilities, particularly regarding sound audibility and frequency range. As infants grow, the TM gradually increases in size, paralleling the growth of the surrounding anatomical structures. This maturation process is essential for the enhancement of auditory perception and the ability to process complex sounds.

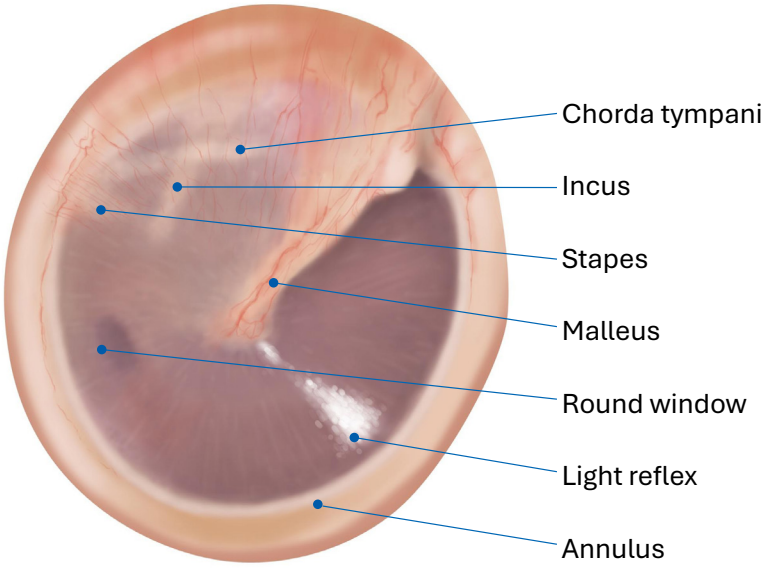


Figure 3.7: Tympanic Membrane

The TM comprises three layers: an outer epidermal layer, a middle connective tissue layer and an inner mucosal layer. In the outer layer, epithelial cells produce keratin, which is shed over time. The keratin migrates outwards to help form the cerumen. Its outer layer provides a protective barrier. The connective tissue in the fibrous middle layer is primarily made of collagen and elastin. It is arranged in an outer radial and inner circular fashion, contributing to the TM‘s tensile strength and vibration capability. The inner mucosal layer protects the interior of the middle ear and connects to the ossicles.

The TM comprises three distinct regions: the annulus, pars tensa and pars flaccida (Figure 3.7). The annulus is a fibrocartilaginous ring that anchors the TM within the external acoustic meatus. It maintains the membrane’s position and ensures it can vibrate effectively in response to sound waves. The pars tensa is the most significant section of the TM, characterised by its tight, fibrous composition. It transforms sound energy into mechanical vibrations. When sound waves reach the membrane, they cause it to vibrate, transferring these vibrations to the middle ear’s ossicles. This mechanical energy is then relayed to the inner ear for further processing. The robustness of the pars tensa also allows it to withstand atmospheric pressure changes and the mechanical forces exerted by sound waves.

The pars flaccida, or Schrapnell's membrane, is located superiorly and lacks the same tension and structural support found in the pars tensa. It comprises fewer fibres and plays a lesser role in sound transmission, primarily serving as a compliant area that accommodates pressure variations within the ear. Branches from the auriculotemporal and vagus nerves innervate the TM and facilitate sensations like pain. In newborns, the nerve supply is often less developed, increasing vulnerability to infections and affecting hearing. The main blood supply to the TM comes from the deep auricular artery and tympanic branches of the maxillary artery.

The ossicles are three tiny bones in the middle ear: the malleus, incus and stapes (Figure 3.8). They facilitate the transmission of sound vibrations from the external environment to the inner ear. The malleus, commonly known as the hammer, is attached to the TM and receives sound waves, transforming them into mechanical vibrations. It then passes these vibrations to the incus, or anvil, which bridges the malleus and the stapes, or stirrup. The stapes, the smallest bone in the human body, connects with the cochlea's oval window and amplifies the sound vibrations before they enter the fluid-filled inner ear.

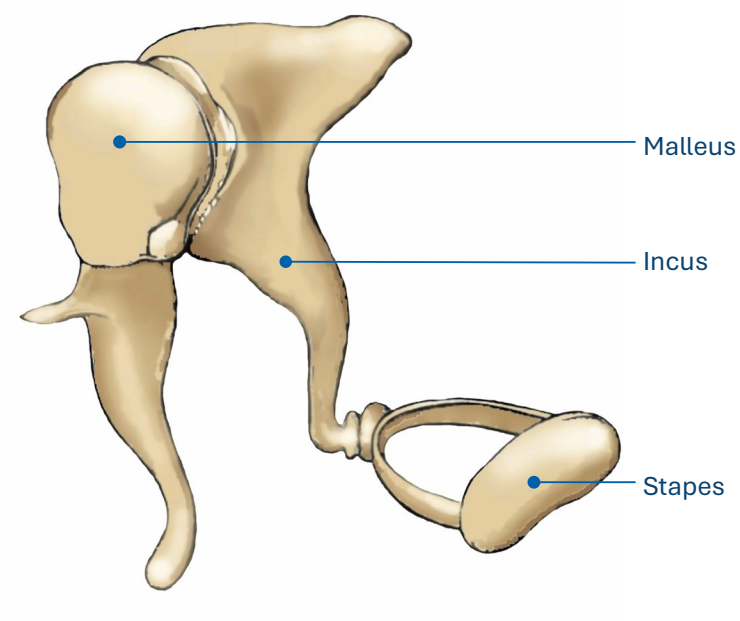


Figure 3.8: The ossicular chain

Amplification occurs in the middle ear through two fundamental mechanisms: the lever action of the ossicles and the differences in surface area between the TM and the oval window. The malleus and incus act as a lever system, magnifying the force exerted on the stapes. Additionally, since the TM is significantly larger than the oval window, the pressure exerted on the oval window is greater than that on the TM, resulting in an effective amplification of sound.

Synovial joints connect the ossicles. They are capable of limited motion, facilitating nuanced changes in sound vibrations. This mobility also enables them to perform a protective function, helping safeguard the TM from pressure fluctuations and maintaining proper ear function during activities such as swallowing or yawning.

The ossicles are further supported by ligaments and connected to two key muscles: the stapedius and the tensor tympani. The stapedius muscle is pivotal in the acoustic reflex, contracting in response to loud sounds, which helps protect the inner ear from potential damage by dampening the vibrations of the stapes. Similarly, the tensor tympani muscle responds to sound and pressure changes, stabilising the TM and reducing the transmission of disruptive vibrations from the throat during swallowing. The tensor tympani muscle is innervated by the trigeminal nerve (CN V), and the stapedius muscle is supplied by the facial nerve (CN VII).

The ET opening lies at the anterior aspect of the middle ear. This opening connects the middle ear to the nasopharynx and facilitates pressure equalisation. It comprises three main sections: the bony, cartilaginous and membranous segments. The bony segment measures about ten millimetres and is found near the middle ear, while the cartilaginous section is the longest segment, connecting to the nasopharynx. The membranous portion is shorter and contains a layer of epithelium, allowing it to maintain patency. The ET is lined with respiratory epithelium, composed primarily of ciliated columnar and goblet cells that produce mucus. This mucus traps particulates and pathogens, while the cilia facilitate the movement of mucus toward the nasopharynx, aiding in the clearance of potential infections. Beneath the epithelium lies a connective tissue layer rich in blood vessels and lymphatics, which plays a role in the immune response.

Figure 3.9: (a)

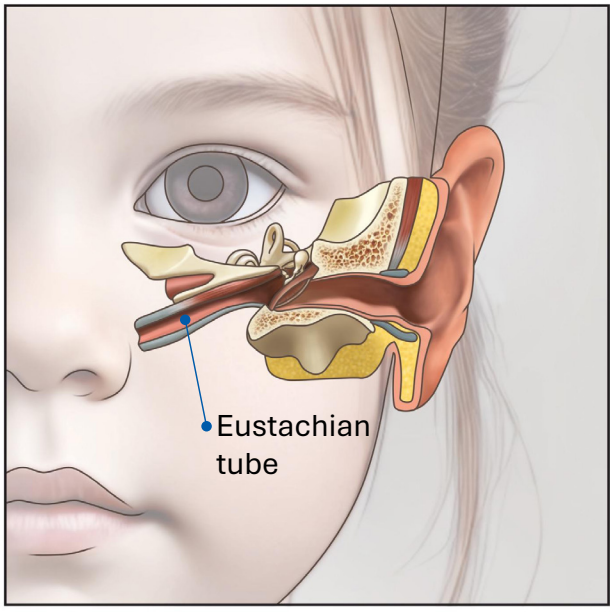


Figure 3.9: (b)

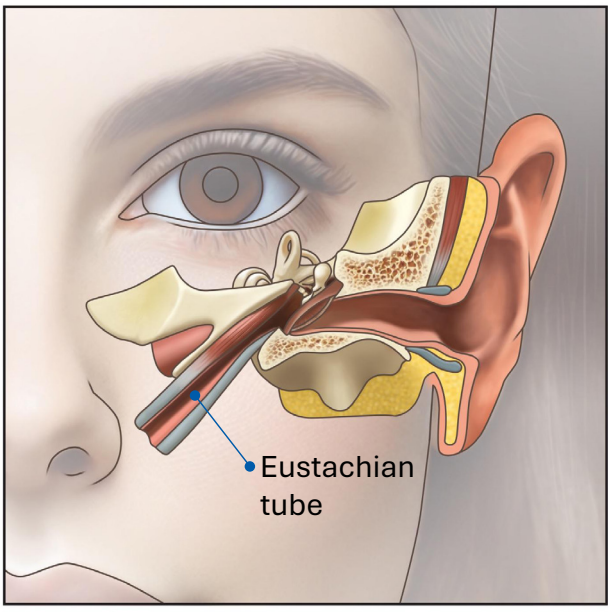


Figure 3.9: (a) The ET in an infant
Figure 3.9: (b) The ET in an adult

Table 3.3: Differences between the infant and adult ET

	Infants	Adults
Size	Smaller	*Larger*
Length	Shorter	*Longer*
Stiffness	Softer	*Stiffer*
Direction into nasopharynx	Horizontal	*More vertical*
Orifice in nasopharynx	Round	*More oval*

Effusion in the middle ear is an exudation formed under negative pressure exceeding 100 millimetres H2.O. This air vacuum is induced when nitrogen, the gas with the most significant diffusion gradient, is absorbed by microvascular circulation quicker than it can be replaced due to ET obstructive dysfunction.

The posterior aspect of the middle ear leads via the aditus ad antrum into the mastoid air cells, the largest of which is the mastoid antrum. The roof of the middle ear forms the boundary with the middle cranial fossa and temporal lobe. The floor of the middle ear is adjacent to the infratemporal fossa, housing the carotid artery and the jugular bulb of the jugular vein.

Mastoid process

The mastoid process is a prominent bony structure behind the ear, forming part of the skull's temporal bone. It is underdeveloped in newborns compared to its formation in adults, as the ossification process remains incomplete at this stage. The mastoid air cells are integral to the mastoid process and vital to maintaining ear health and functionality.

Mastoid air cells comprise a complex network of small, interconnected cavities encapsulated by a mucous membrane. The lining consists of predominantly ciliated columnar cells interspersed with goblet cells responsible for mucus secretion. This specialised arrangement ensures a healthy respiratory epithelium, as it filters and humidifies incoming air. The cilia facilitate mucus movement and capture particles outward, thereby supporting the hygiene of the middle ear and surrounding epitympanic space. Additionally, these air cells harbour dense vascular and neural tissue, contributing to both immune responses and sensory functions.

These air cells are a pneumatic system that helps balance pressure between the middle ear and the external environment. Pneumatisation is when epithelial cells infiltrate the developing bone and create epithelium-lined air cell cavities. This is particularly significant during transitioning from a fluid-filled foetal environment to an air-filled atmosphere after birth, which can introduce pressure imbalances affecting hearing and balance.

The mastoid air cells connect to the middle ear via the aditus ad antrum, thus aiding in the necessary pressure equalisation to ensure proper TM function.

Inner ear

The inner ear or labyrinth is a complex maze consisting of irregular pathways and passages in the inner ear. It contains the hearing and balance organs (Figure 3.10).

The bony labyrinth, also known as the osseous labyrinth or otic capsule, is the rigid, bony outer wall of the inner ear embedded within the temporal bone. The otic capsule reaches its adult size at birth, measuring approximately fifteen millimetres in width. The bony labyrinth is divided into three primary parts: the cochlea, the vestibule and the semicircular canals. The bony labyrinth houses and protects the membranous labyrinth.

Figure 3.10 (a)

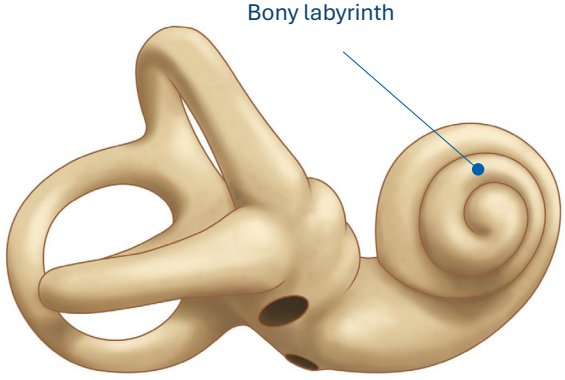


Figure 3.10 (b)

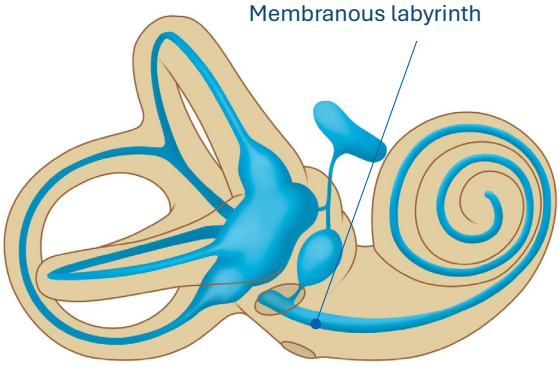


Figure 3.10: The bony and membranous labyrinth.

The membranous labyrinth is a complex system of fluid-filled channels and sacs within the bony labyrinth. It is divided into the cochlear duct, saccule, utricle, three semicircular ducts and the endolymphatic duct and sac.

In adults, the cochlea is a 3.2 to 4.2 centimetre-long spiralled, snail-shaped structure composed of two and a half turns (Figure 3.11). It converts sound vibrations into neural signals. The cochlea comprises three fluid-filled chambers: the scala vestibuli, scala tympani and scala media (cochlear duct). The scala vestibuli and tympani are filled with perilymph, similar to extracellular fluid with a high sodium content. In contrast, the scala media contains endolymph, a unique ionic composition with a high potassium content that supports sensory transduction.

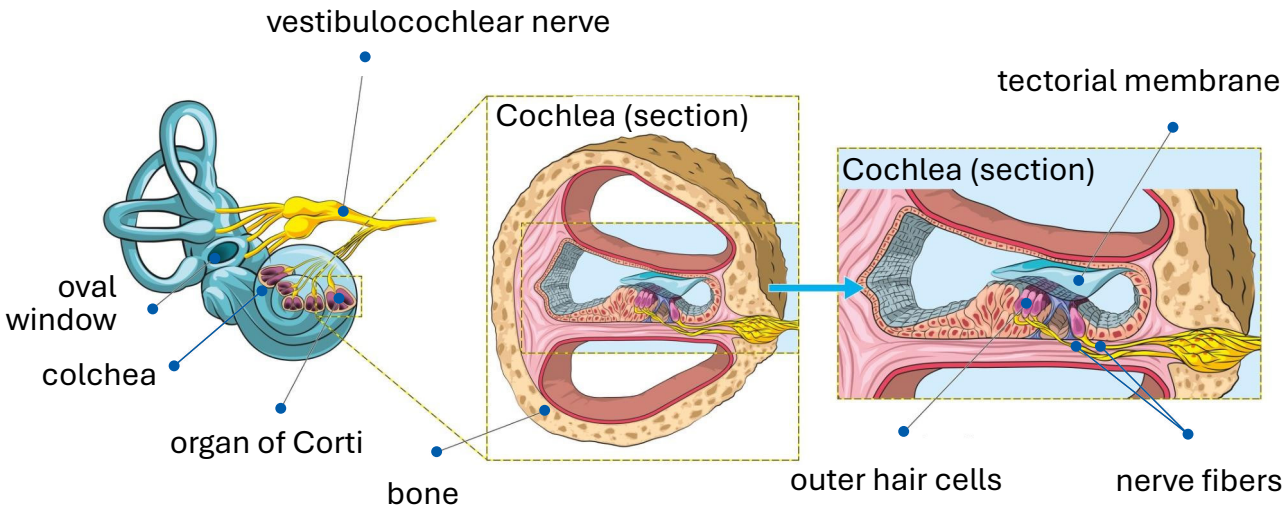


Figure 3.11: The cochlea, scala and organ of Corti

The cochlear duct is separated from the scala vestibuli by Reissner's membrane and from the scala tympani by the basilar membrane (Figure 3.11). The latter supports the organ of Corti, the sensory structure that contains hair cells responsible for transducing sound. Hair cells enable the transduction of mechanical stimuli into electrical signals.

These hair cells are arranged in two rows: The inner hair cells, which mainly convey auditory information to the brain and the outer hair cells, which amplify sound vibrations. The human cochlea has 3500 inner hair cells and about 12,000 outer hair cells. The movement of the basilar membrane in response to sound waves causes the stereocilia on these hair cells to bend, opening ion channels that depolarise and, subsequently, release neurotransmitters.

The cochlea also contains supporting cells and the cells of the stria vascularis at the lateral aspect near the wall, which maintains the ionic composition of the endolymph. Connexins are a family of proteins that form gap junctions between adjacent cells, allowing for direct communication and transport of ions, small molecules and electrical signals.

The cochlear aqueduct is a small channel that extends from the scala tympani of the basal turn of the cochlea to the subarachnoid space. Measuring approximately 0.7 to 1.5 mm in diameter, it plays a role in the maintenance of inner ear fluid and balances the pressure between the perilymph space and the cerebrospinal fluid. It is a potential pathway for the spread of infections, such as meningitis, into the cochlea.

The vestibulum forms the central part of the bony inner ear and is essential for balance and spatial orientation. It encompasses the two otolith organs, the utricle and saccule, which detect linear acceleration, of which gravitation is a constant component. The vestibulum is anatomically located between the semicircular canals and the cochlea.

The utricle and saccule contain maculae, specialised sensory patches of hair and supporting cells. The hair cells in these maculae are embedded in a gelatinous layer laden with calcium carbonate crystals called otoliths. When the head moves, gravitational or inertial forces during acceleration cause the otoliths to shift, leading to a relative displacement of the gelatinous layer. This mechanical distortion causes the bending of stereocilia on the hair cells, releasing neurotransmitters and sending signals to the vestibular nuclei in the brainstem.

The maculae of the utricle and saccule exhibit a highly organised structure. The hair cells' stereocilia are arranged in a specific pattern with taller kinocilia positioned in a defined orientation, enabling the detection of directional movements. The utricle contains approximately 33 000 hair cells and the saccule 19 000 hair cells.

The supporting cells provide structural and functional support. At the same time, the overlying otolith membrane is rich in glycoproteins that contribute to the mass difference necessary for the displacement caused by head movements.

There are three bony semicircular canals: the superior (anterior), posterior (inferior) and lateral (horizontal). Each canal contains a membranous duct and is oriented perpendicularly to the others, thus facilitating the detection of angular accelerations during various head movements. At the base of each canal lies the ampulla, which contains the crista ampullaris, where sensory hair cells reside. The three semicircular canalseach contain 7600 hair cells.. The semicircular ducts are filled with endolymph and surrounded by perilymph.

When the head rotates, the inertia of the endolymph causes it to lag behind the motion of the semicircular canals. This relative movement of the endolymph results in bending of the cupula, a gelatinous structure that overlies the crista ampullaris. Such bending stimulates the hair cells within the crista, leading to their depolarisation and the subsequent generation of action potentials. These electrical signals are conveyed to the central nervous system via the vestibular nerve, thus enabling an organism's perception of its position and movement in three-dimensional space.

The crista ampullaris, which consists of hair cells and supporting cells, is similar to the maculae. However, its unique orientation is specifically adapted for detecting rotational movements. The directional arrangement of the kinocilia in the hair cells within the semicircular canals allows for precise motion discrimination.

The vestibulo-ocular reflex (VOR) is an essential mechanism that stabilises vision during head movements, allowing the eyes to maintain a steady gaze on a target (Figure 3.12). It can be categorised into two primary types: angular (rotational) VOR and translational VOR.

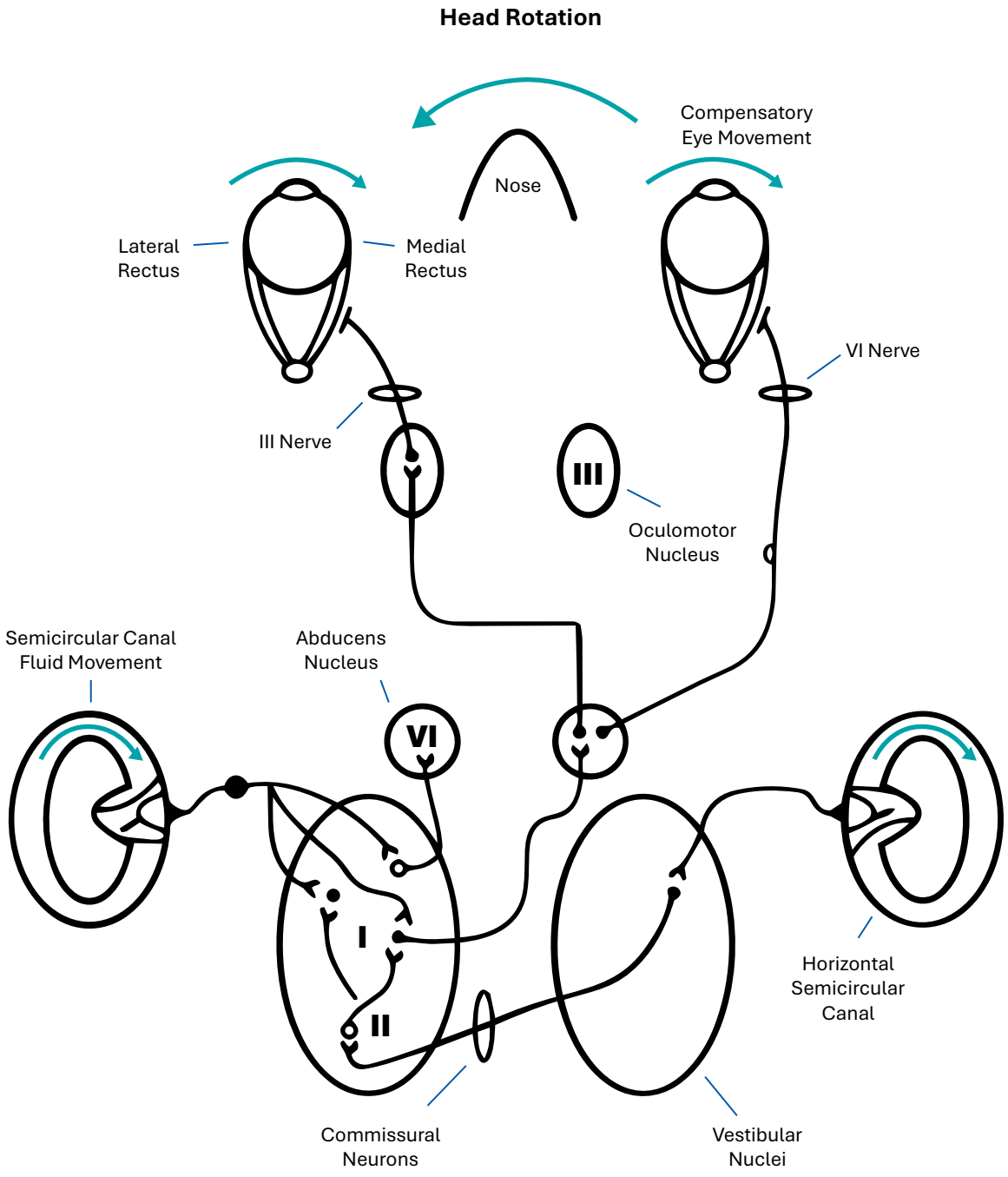


Figure 3.12: The vestibulo-ocular reflex (VOR) of the lateral (horizontal) canal

Rotational head movements activate angular VOR. When the head turns, the semicircular canals in the inner ear detect the angular acceleration and send signals to the oculomotor muscles, resulting in compensatory eye movements. For instance, during a rapid head turn to the right, the right eye will rotate leftward to maintain focus on an object, thus counteracting the head's movement. This reflex is crucial for maintaining visual acuity during high-speed activities, such as sports or driving.

The translational VOR responds to linear accelerations like those experienced during forward or backward head movements. The otolith organs (utricle and saccule) sense gravitational forces and linear motion, eliciting eye movements that stabilise vision in response to shifts in head position. This reflex ensures visual clarity when the head moves up or down or the body is in motion.

The vestibular aqueduct is a bony canal that connects the inner ear with the posterior cranial fossa. The vestibular aqueduct has a variable length averaging one and a half to eight millimetre in the adult. It houses the endolymphatic duct and sac, facilitating the transport of endolymph fluid, which is essential for the vestibular system to function adequately (Figure 3.13). The endolymphatic sac serves as a storage site for this fluid, assisting in pressure regulation within the inner ear and thereby contributing to the homeostasis of the auditory and vestibular systems. It also plays a role in the immunological function of the inner ear.

Figure 3.13: (a)

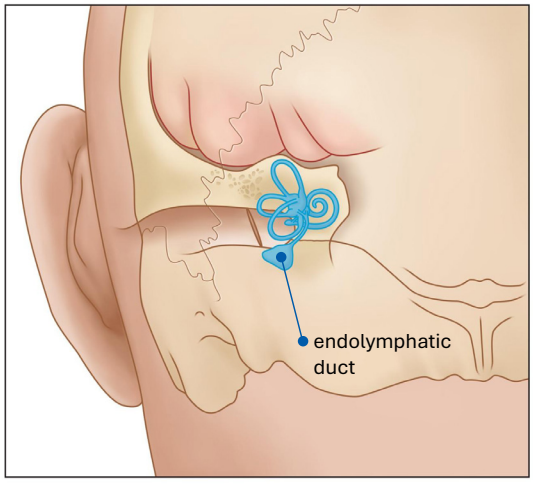


Figure 3.13: (b)

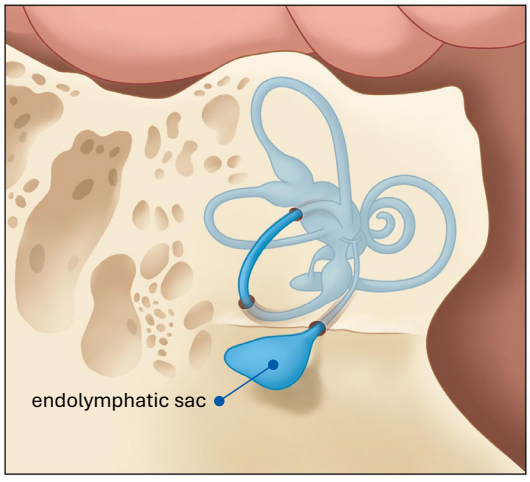


Figure 3.13: (a) The endolymphatic duct
Figure 3.13: (b) The endolymphatic sac

Internal auditory meatus

The internal auditory meatus is a bony canal located within the petrous portion of the temporal bone. It connects the inner ear with the cerebrospinal fluid-filled cerebellopontine angle of the posterior cranial fossa and the brainstem. The average diameter of the internal auditory meatus in neonates ranges from approximately five to seven millimetres, compared to seven to nine millimetres in adults. The internal auditory meatus is nine millimetres long in a baby and eleven millimetres in an adult.

The contents of the internal auditory meatus include the vestibulocochlear nerve (CN VIII) and the facial nerve (CN VII), both of which are essential for facial motor control (Figure 3.14a). The CN VIII consists of the cochlear nerve, which contains roughly 30,000 to 35,000 nerve fibres, and the vestibular nerve, which contains 20,000 nerve fibres.

Additionally, the canal houses the labyrinthine artery, a branch of the anterior inferior cerebellar artery (AICA), which supplies blood to the inner ear (Figure 3.14b).

Figure 3.14: (a)

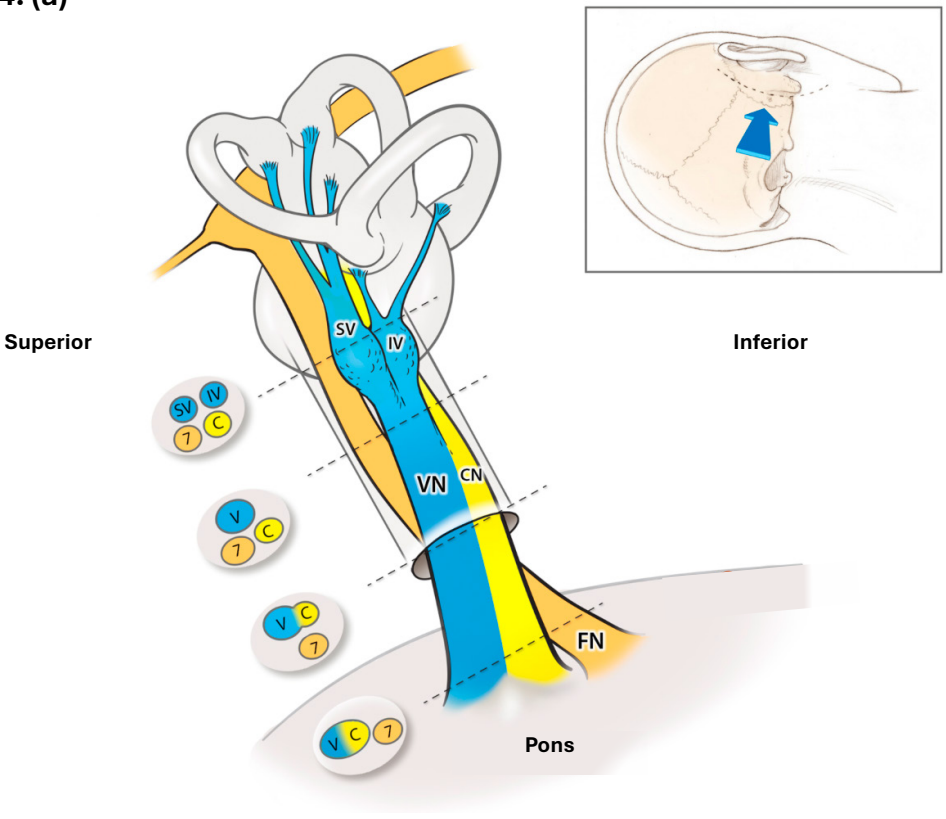


Figure 3.14: (b)

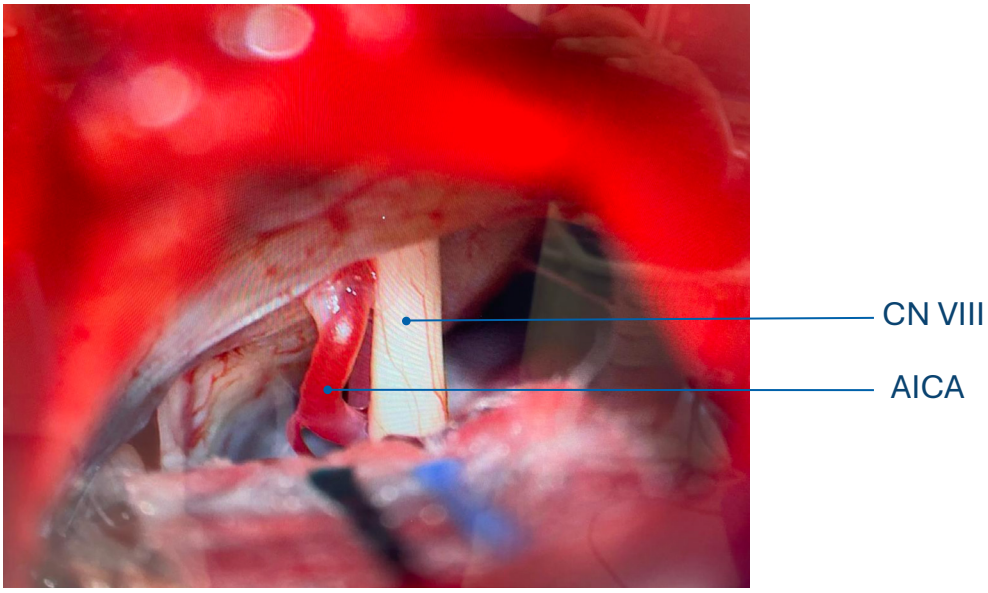


Figure 3.14: (a) Internal auditory meatus with FN and VCN consisting of the VN and CN, with VN further dividing into the SV and the IV
Figure 3.14: (b) Surgical image of CN VIII and AICA

Brainstem nuclei and tracts of audiovestibular function

The brainstem auditory pathway begins in the cochlea, where auditory sensory cells transduce sound vibrations into neural signals, activating the auditory nerve. These signals first converge in the cochlear nuclei, the primary auditory nuclei in the brainstem (Figure 3.15).

The cochlear nuclei have two main components: the dorsal cochlear nucleus (DCN) and the ventral cochlear nucleus (VCN). The DCN processes complex sounds and contributes to sound localisation and discrimination, while the VCN primarily involves sound timing and intensity coding. After initial processing, auditory signals from the cochlear nuclei project to several important brainstem nuclei, including the superior olivary complex (SOC), the lateral lemniscus (LL) and the inferior colliculus (IC).

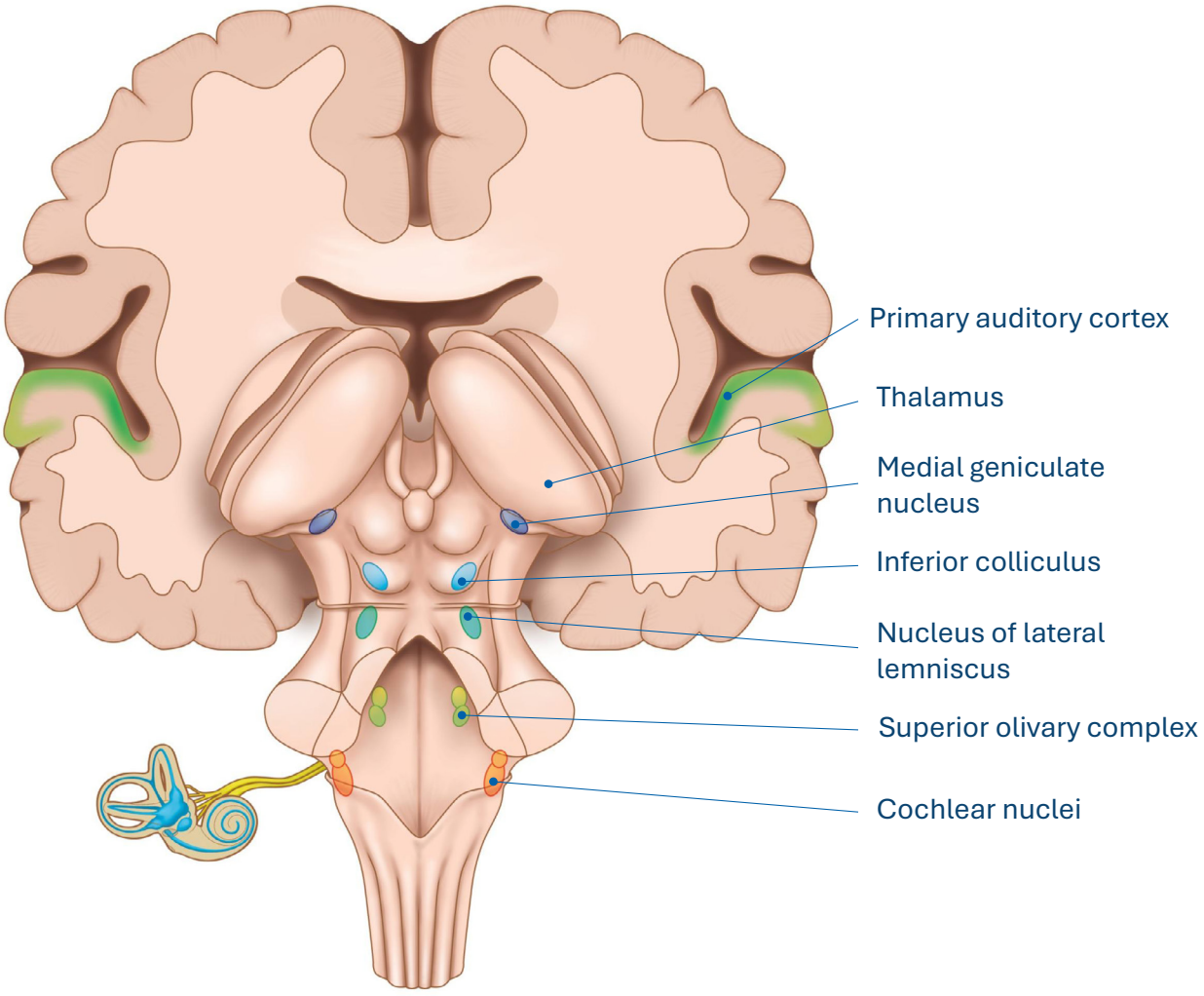


Figure 3.15: Cochlear nuclei, the primary auditory nuclei

The SOC is crucial for binaural hearing, as it receives inputs from both ears and integrates these signals to determine the direction of sound sources. The LL carries information from the SOC and other nuclei to the IC. The IC acts as a relay station that further processes auditory signals, integrating various aspects of sound, including frequency, intensity and timing, before sending information to the thalamus and, ultimately, to the auditory cortex.

The brainstem vestibular nuclei are located in the medulla and pons (Figure 3.16). These nuclei include the superior, lateral, medial and inferior vestibular nuclei. They receive input from the vestibular apparatus in the inner ear. The connections of the vestibular nuclei extend to several pivotal pathways, including the olivary nuclei. From the olivary nuclei, efferent projections travel to motoneurons in the spinal cord via the vestibulospinal tract, facilitating postural adjustments and reflexive head movements.

The medial longitudinal fasciculus (MLF) is the neural pathway that co-ordinates eye movements and integrates sensory information for ocular control. This white matter tract extends longitudinally through the brainstem, interconnecting several cranial nerve nuclei responsible for eye and head movements, notably those of the oculomotor (III), trochlear (IV) and abducens (VI) nerves.

The MLF facilitates communication between the vestibular nuclei and the nuclei of the extraocular muscles, thereby enabling the co-ordination of conjugate gaze and the maintenance of visual stability during head movements. It plays a vital role in the vestibulo-ocular reflex (VOR), stabilising the visual field by adjusting eye positioning in response to head movement.

The vestibular nuclei also connect via the olivary nuclei with the cerebellum, primarily through the inferior cerebellar peduncle, contributing to fine-tuning balance and co-ordination.

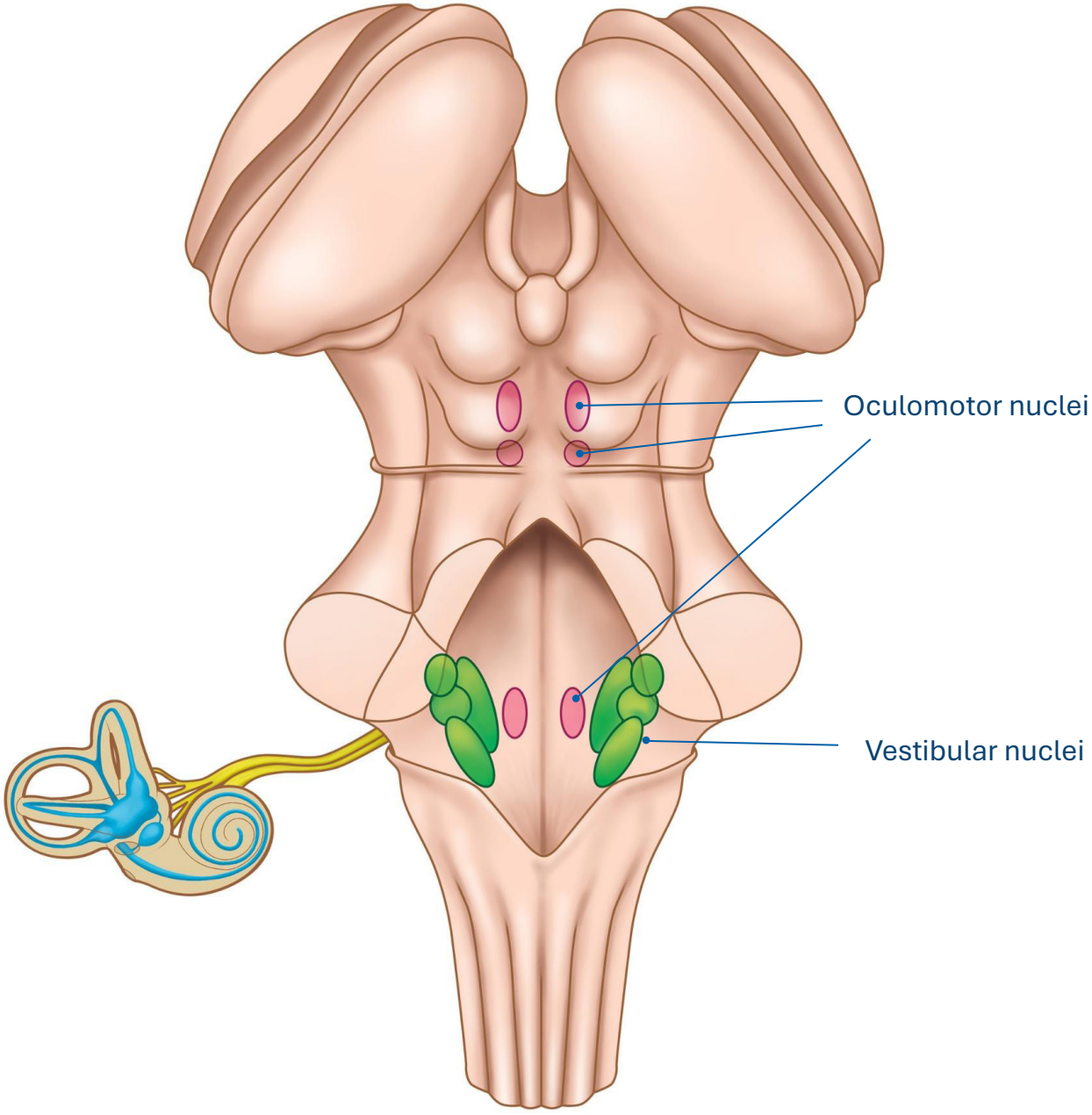


Figure 3.16: Brainstem vestibular and oculomotor nuclei

The vestibulospinal reflex (VSR) and vestibulocollic reflexes (VCR) are two other neural pathways that maintain postural stability and spatial orientation.

The vestibulospinal reflex facilitates balance and posture by transmitting signals from the vestibular apparatus in the inner ear to the spinal cord. This reflex influences extensor muscle activity in response to head and body position changes. This reflex plays a vital role during locomotion, particularly when the body encounters perturbations, ensuring the centre of mass remains stable.

The vestibulocollic reflex is primarily responsible for stabilising the head in relation to the body. It involves bilateral pathways that project from the vestibular nuclei to the neck motoneurons, allowing for co-ordinated head movements that counteract sudden changes in posture or position. Although less important in humans, this reflex is essential during activities such as walking or running, where head stability must be maintained despite the dynamic movement of the body.

Vestibulocerebellum

The vestibulocerebellum is primarily responsible for regulating balance and spatial orientation. (Figure 3.17). It comprises the flocculonodular lobe and is intricately connected to vestibular nuclei in the brainstem, facilitating the integration of vestibular information. Through its modulation of muscle tone and co-ordination of eye movements, the vestibulocerebellum plays a pivotal role in maintaining postural stability and enabling smooth, co-ordinated movements.

The vestibulocerebellum assesses and adapts to head position and motion changes, allowing for real-time adjustments to balance and equilibrium. Dysfunction in this area can lead to disorders such as ataxia, where affected individuals may experience difficulty in maintaining posture or executing coordinated movements. Furthermore, its influence extends to ocular motor control, exemplified by the VOR, which stabilises vision during head movements.

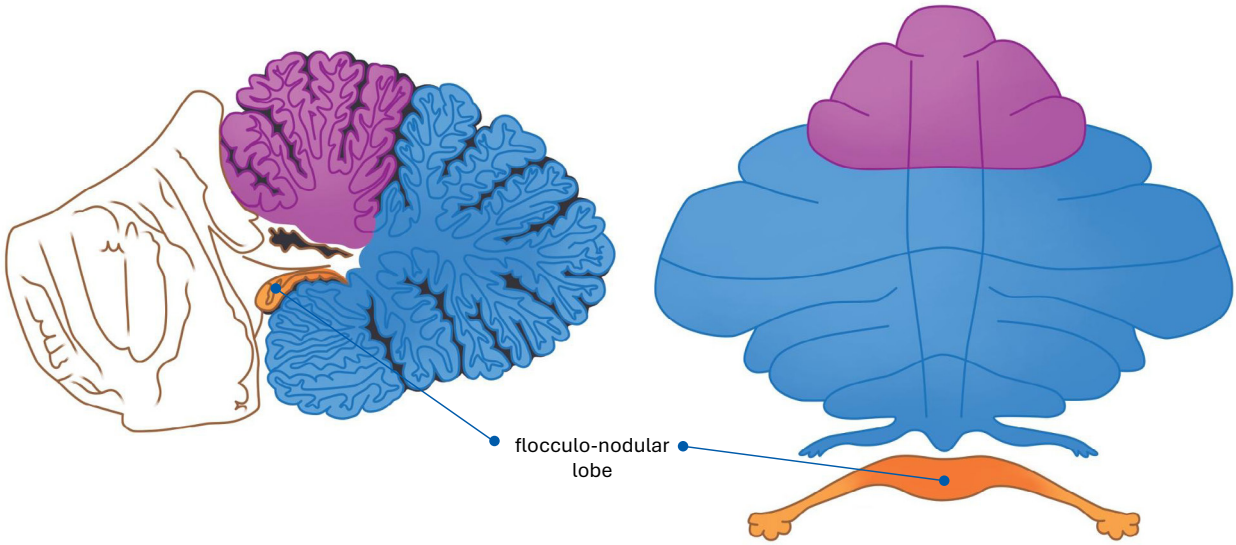


Figure 3.17: The vestibuocerebellum (flocculo-nodular lobe)

Auditory and vestibular cortex

The auditory processing pathway begins with signals originating from the inferior colliculus, ascending to the thalamus's medial geniculate nucleus (MGN). This structure acts as a vital relay station, where auditory signals undergo refinement before transmitting to the auditory cortex. The pathway that connects the MGN to the primary auditory cortex (A1), located in the superior temporal gyrus of the temporal lobe, is called auditory radiation. This neural pathway is essential for auditory perception and localisation, as it integrates information that enables the differentiation of sounds amidst complex auditory environments.

The auditory cortex can be divided into several distinct functional areas, with A1 playing a crucial role in the initial processing of auditory stimuli. The organisation of A1 is tonotopic, which signifies that it spatially arranges different sound frequencies (Figure 3.18). Adjacent regions, such as the secondary auditory cortex, are involved in higher-order processing tasks, such as sound recognition, association and integration of auditory information with other sensory modalities. This hierarchical structure facilitates increasingly sophisticated interpretations of auditory inputs, ranging from basic pitch detection to the complex recognition of patterns inherent in spoken language and music.

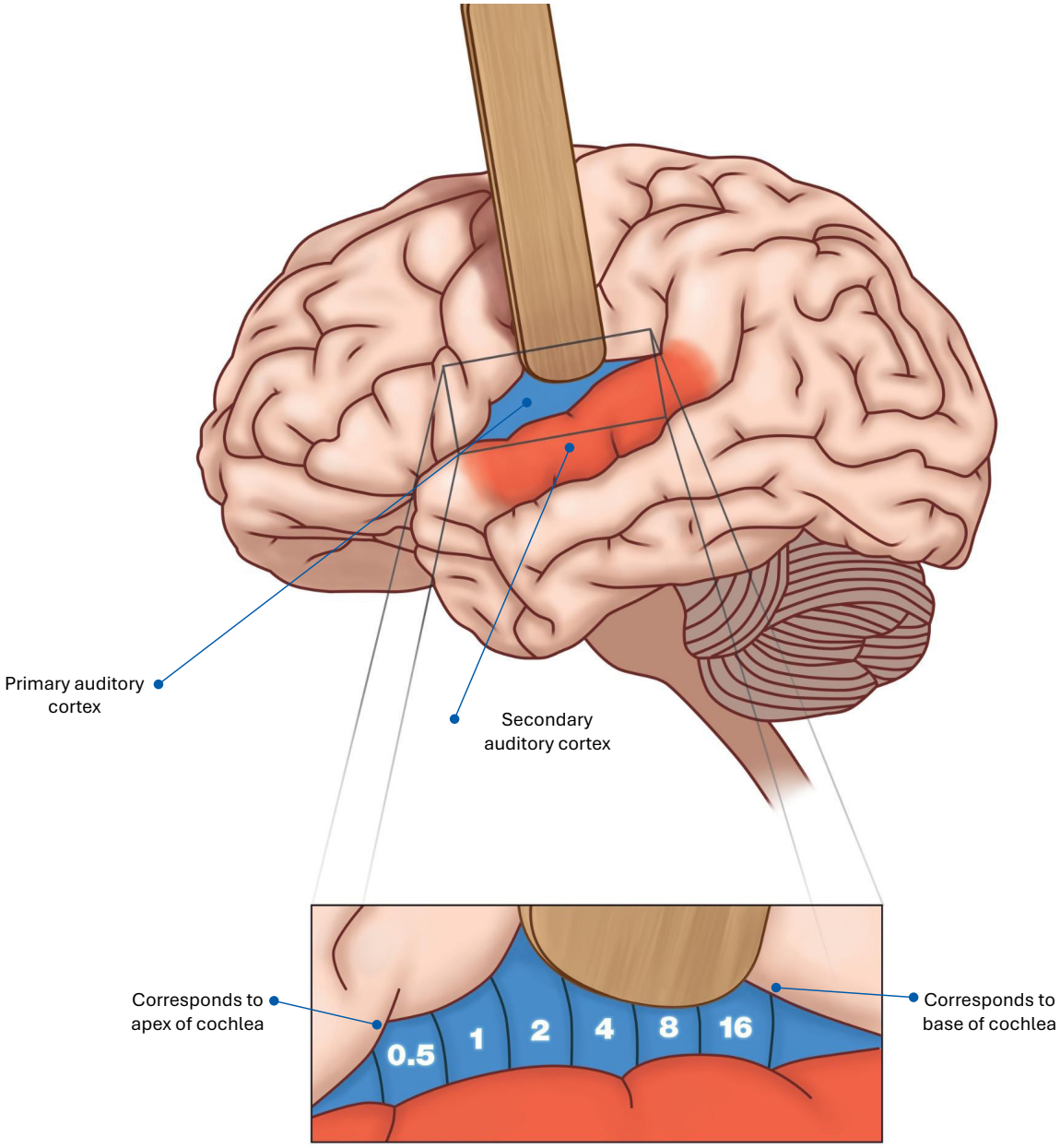
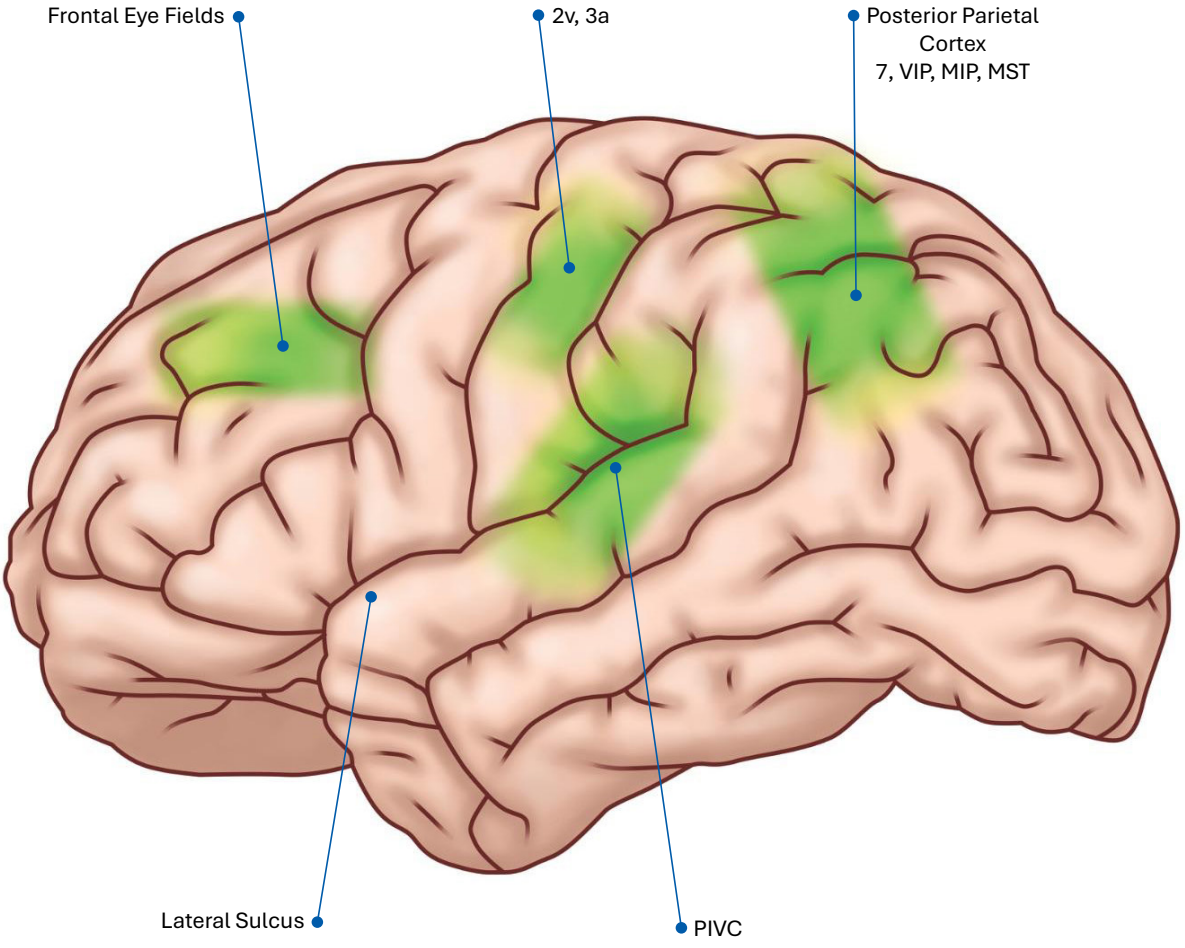


Figure 3.18: The auditory cortex A1 with tonotopic arrangement for different frequencies (kHz)

The vestibular cortex is crucial in processing sensory information related to balance, spatial orientation, head movements and position relative to gravity and motion. The vestibular cortex receives input from the vestibular system, located primarily within the insular and parietal lobes, particularly in areas such as the posterior insula and the parietal operculum.

The vestibular cortex is not isolated in function but works with other sensory modalities, including visual and proprioceptive inputs. Different cortical regions (Figure 3.18) of the brain known to be involved with vestibular processing are summarised in Table 3.4.



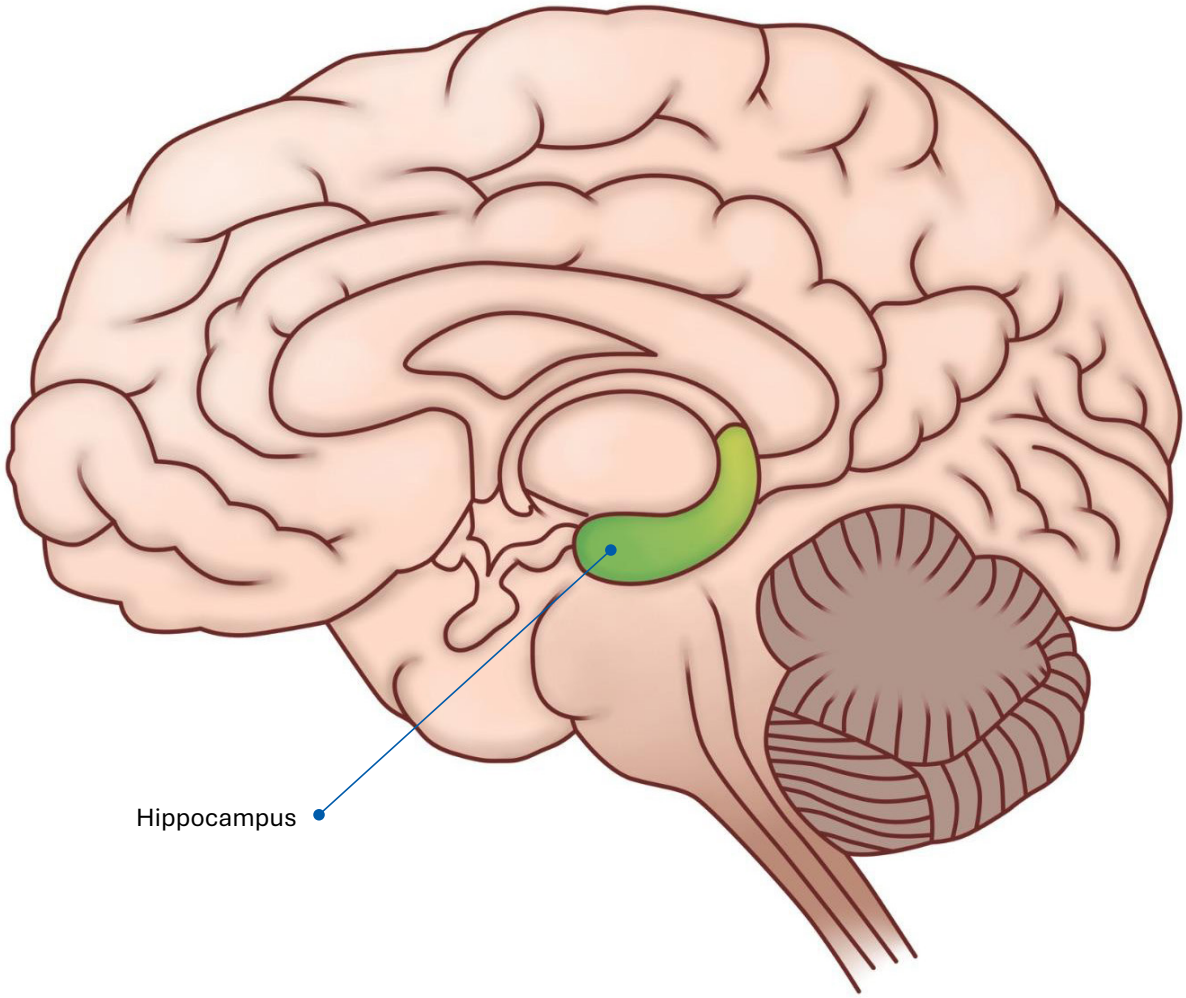


Figure 3.19: The vestibular cortex and associated cortical regions of vestibular processing

This multimodal integration enhances the brain's ability to construct a coherent spatial orientation and movement perception through complex interactions among the vestibular, visual and somatosensory systems.

Table 3.4: Cortical areas associated with vestibular processing. Source: Angelaki and Dickman (2024)

Cortical area	Function
Frontal eye fields (FEF)	Eye movement regulation
Somatosensory areas 2v and 3a	Body location and movement signal mapping
Posterior insular vestibular cortex (PIVC)	Body and head motion processing
Posterior parietal cortex Area 7 *Ventral intraparietal area (VIP)* *Medial intraparietal area (MIP)* *Medial superior temporal area (MST).*	*Motion perception*
Hippocampus and parahippocampal regions	*Spatial orientation and navigation*

Development of the Paediatric Audiovestibular System

Maturation of the temporal bone

The temporal bone in neonates is predominantly composed of cartilage and is not fully developed at birth (Moore, Persaud and Torchia 2018). The squamous portion, exhibiting minimal ossification, initiates its ossification process shortly after birth through intramembranous ossification, vital for flat bone formation. During the early years of life, this area undergoes continuous ossification, progressively obscuring the sutures that connect it to adjacent bones, thereby indicating structural integration. Notable ossification occurs in the formation of the roof of the external auditory canal, with significant development evident between ages eight and twelve, approaching the adult morphology. Full maturation occurs in adolescence.

The mastoid process is also underdeveloped at birth. It experiences significant postnatal growth influenced by surrounding anatomical structures, notably the sternocleidomastoid muscle, which exerts mechanical forces that facilitate its development. During the first two years of life, the mastoid undergoes intramembranous and endochondral ossification and is fully pneumatised by age twelve, albeit with individual size variability.

The petrous part, present at birth as the primary ossification centre, remains largely unossified and is one of the last regions to ossify, primarily through endochondral ossification. Its development, crucial for hearing and balance, progresses significantly through childhood, with substantial changes noted by age twelve. Similarly, the tympanic part is largely cartilaginous at birth. It commences ossification through mesenchymal condensation shortly thereafter, culminating by the end of the second year, reaching its final shape in five years.

Maturation of the outer ear

The pinna undergoes significant growth during early childhood, with the most rapid enlargement occurring within the first two years. By age twelve, the pinna reaches six centimetres, approximately 90% of its adult size. The width dimensions mature earlier, at five and a half to eleven years, whereas the maturity age of length dimensions varies from twelve to sixteen years (Purkait 2013). It eventually acquires a more defined shape that enhances its ability to funnel sound waves into the ear canal, facilitating enhanced sound localisation and resonance, which is vital for effective auditory.

The external auditory meatus gradually elongates and becomes more S-shaped as the child grows, reaching approximately three centimetres by age twelve. Despite their anatomical maturity at birth, the ceruminous and sebaceous glands do not achieve their full functional capacity until puberty.

Maturation of the middle ear

The middle ear space, initially small and less ventilated in neonates, expands as the child grows, reaching its adult size by approximately twelve years (Mishra, Dinger and Renken 2017). This process facilitates better aeration and drainage through the ET. This expansion also enhances the ability to transmit sound waves effectively. Different frequencies follow different maturational timelines.

At birth, the TM is thick and relatively flat and horizontal. It gradually adopts a more oblique position and becomes thinner as the child matures, which enhances sound transmission. The ossicles begin developing in utero and fully ossify by the end of infancy. Their arrangement in the middle ear includes size increases and maturation of their joints and muscles,

improving their mechanical advantage in sound conduction. The majority of the remodelling of the bone in the ossicles takes place in the first months and years of life, while afterwards, only minor reorganisation occurs (Rolvien et al. 2018).

As the child matures, the eustachian tube elongates and angles downward, improving ventilation and drainage of the middle ear. It reaches its adult configuration by age seven (Takasaki et al. 2007).

Maturation of the inner ear

The inner ear structures are responsible for auditory and balance functions, along with the auditory and vestibular nerves that facilitate sensory perception.

The cochlea undergoes significant development and maturation from birth to age twelve. Intrinsic genetic factors and extrinsic environmental influences can influence cochlear development. At birth, the cochlea is anatomically present but functionally immature. The sensory hair cells reach their highest density shortly after birth, with no regeneration occurring after the first week (Heuermann et al. 2022). Auditory thresholds are up to 25 dB higher in neonates, typically around 40–60 dB HL (hearing level), indicating that while some auditory perception exists, it is far from optimal (Sininger, Abdala and Cone-Wesson 1997).

Critical growth and differentiation processes occur during the first few months of life. Cochlear hair cells begin to establish synaptic connections with auditory neurons (Johnson et al. 2019). The maturation of the organ of Corti leads to enhanced sensitivity to sound frequencies, allowing infants to respond to environmental auditory stimuli and facilitating language acquisition. The audiovestibular nerve undergoes substantial development during the first year of life.

The cochlea is adult-sized at birth, although cochlear size variability is present among individuals.

The frequency selectivity of hair cells improves as they mature, which is crucial for understanding speech among background noise (speech-in-noise perception)—a skill increasingly demanded in social interactions and educational environments. Over the subsequent years, typically within four to eight years, this maturation of the cochlea drives the auditory brain toward attaining its optimal performance levels, contingent upon the specific perceptual tasks involved (Pujol, Lavigne-Rebillard and Uziel 1991). Normal maturation of the cochlea, reaching its maximum performance at birth, will slowly (within four to eight years, depending on the perceptual task) drive a normal auditory brain to the acquisition of its maximum performance.

It is essential to consider the impact of auditory exposure on cochlear development. Children frequently exposed to rich auditory environments typically exhibit better auditory processing and language skills (Atılgan, Cesur and Çiprut 2023). Conversely, those with hearing loss may experience delays in cochlear maturation and subsequent language development, emphasising the interactive nature of auditory experiences during this critical developmental window.

The peripheral vestibular system, essential for balance and spatial orientation, comprises several critical structures, including the utricle, saccule and semicircular canals. These components, along with the endolymphatic duct and sac, develop intricately from birth to age twelve, adapting to the sensory demands of a maturing child.

The morphology of the semicircular canals remains largely unchanged following birth; however, a notable tendency for these structures to tilt anteriorly as a cohesive unit emerges with age. The anterior tilt of the semicircular canals may arise from age-related adjustments in the surrounding cranial and cervical structures, effectively altering the orientation of these canals without altering their fundamental morphological characteristics (Lyu et al. 2016).

Maturation of the central auditory system

The central auditory system undergoes a complex process of maturation and development, beginning in the cochlear nucleus in the brainstem and extending to various auditory nuclei, thalamic relay centres, and, ultimately, the auditory cortex and associated speech/language areas. This intricate developmental trajectory is crucial for effective sound processing and the emergence of speech and language capabilities.

In neonates, the cochlear nucleus already exhibits a rudimentary tonotopic organisation—a spatial arrangement where different frequencies are processed in discrete regions. As the child matures into early childhood, the connectivity of the cochlear nucleus with subsequent brainstem auditory nuclei, such as the superior olivary complex and the inferior colliculus, becomes more sophisticated. The neural circuits in these areas demonstrate increased synaptic density by twelve months, and myelination—essential for efficient synaptic transmission—is usually completed in two years (Prado and Dewey 2014).

Electrophysiological measures, particularly Auditory Brainstem Responses (ABRs), serve as critical tools in assessing the maturation of the auditory brainstem in infants and young children (Chapter 6). ABRs provide insights into the integrity and development of auditory pathways by measuring neural activity in response to auditory stimuli. In the first year of life, ABRs undergo significant changes, reflecting the ongoing maturation of both peripheral and central auditory structures. Children often exhibit ABR waveforms that closely resemble

adult patterns by age two (Johnson et al. 2008), indicating the establishment of fully developed brainstem processing capabilities.

While children exhibit uniform neural responses to click stimuli, distinct differences arise when processing speech sounds. Specifically, three—to four-year-old children show delayed and less synchronous neural onset and sustained responses compared to their five - to twelve-year-old counterparts. These findings highlight the critical period of auditory maturation, with significant strides in the neural encoding of speech around the age of five (Johnson et al. 2008).

Auditory signals ascend to the thalamus's medial geniculate body (MGB). The MGB acts as a critical relay centre, further refining auditory information before it reaches the A1. During the late neonatal period and into early childhood, the thalamic neurons that project to the auditory cortex undergo significant maturation. The connectivity between the MGB and auditory cortex strengthens over time, with enhancing synaptic inputs that assist in complex sound processing. The first two years of life are essential for forming these auditory thalamocortical connections. As neural pathways consolidate, the auditory cortex becomes increasingly responsive to various auditory stimuli. Children start to process sounds akin to adults by age three to four, reflecting the substantial myelination and synaptic development that occurred in previous years.

The primary auditory cortex (A1) is the next stage in auditory processing, where complex sound patterns are interpreted. The maturation of A1 is marked by the development of advanced auditory functions, including sound localisation, frequency discrimination and temporal processing—all vital for language development.

The superficial layers of the auditory cortex do not reach full maturity until adolescence. While myelination is observable across various cortical layers by age six, the process of myelination further progresses into adolescence (Hallet and Proctor 1996). This enhanced myelination facilitates more efficient and rapid neural transmission both within hemispheres and across different cortical areas. Additionally, the complexity of dendritic branching structures continues to evolve until approximately twelve years. These maturation processes are crucial, as they contribute to the establishment of effective auditory processing abilities.

In parallel, significant developments occur in central speech and language areas, like Broca's and Wernicke's areas. While these areas do not process auditory information directly, the maturation of auditory cortex functions is foundational for linguistic development. The sensitivity to phonetic contrasts develops rapidly during the first year, influenced by exposure to the ambient language environment (Kuhl and Rivera-Gaxiola 2008). Children begin to exhibit a categorical perception of phonemes, which is critical for developing language.

Maturation of the central vestibular system

The central vestibular system, encompassing the vestibular nuclei, vestibulocerebellum and connections to various brain regions, including the thalamus and vestibular cortex, undergoes substantial maturation from the neonatal stage until approximately eleven to fifteen years of age. This developmental trajectory facilitates increasingly sophisticated reactions to vestibular stimuli.

Following birth, the vestibular nuclei initiate the processing of signals from the fully developed semicircular canals and otolith organs; however, the central integration of these signals remains immature. As infants grow, synaptic connections within the vestibular nuclei expand, notably with ocular motor nuclei, which are crucial for refining the VOR and enhancing visual stability while moving.

2 years of age: The brain's fundamental structural and functional architecture seems to be in place, and the brain maturation that occurs in later childhood slows.

2-3 years of age: Significant balance and postural control improvements are observed, attributed to both vestibular maturation and thalamic integration (Shumway-Cook and Woollacott 2017).

3 years of age: The vestibular system reaches a crucial developmental milestone. The vestibulocerebellum significantly increases connectivity and functional specialisation. This maturation is evident as children engage in more complex activities like walking and climbing. The system continues evolving into school age, improving spatial orientation and postural control, essential for engaging in sports and dynamic activities (Rine 2018).

7 years of age: Vestibular thalamic functions are largely mature.

10-11 years of age: The hippocampus peaks in volume. Connectivity progresses differently in hippocampal subregions, reaching adult levels

15 years of age: The vestibular function reaches adult-like values (O'Reilly et al. 2011). The vestibulo-ocular, vestibulo-spinal and vestibulo-collic reflexes are now fully developed. Children can now perform intricate tasks requiring balance, co-ordination and spatial awareness with increased competence, thanks to the maturation of the vestibular pathways connecting the brainstem and cortex (Figure 3.20).

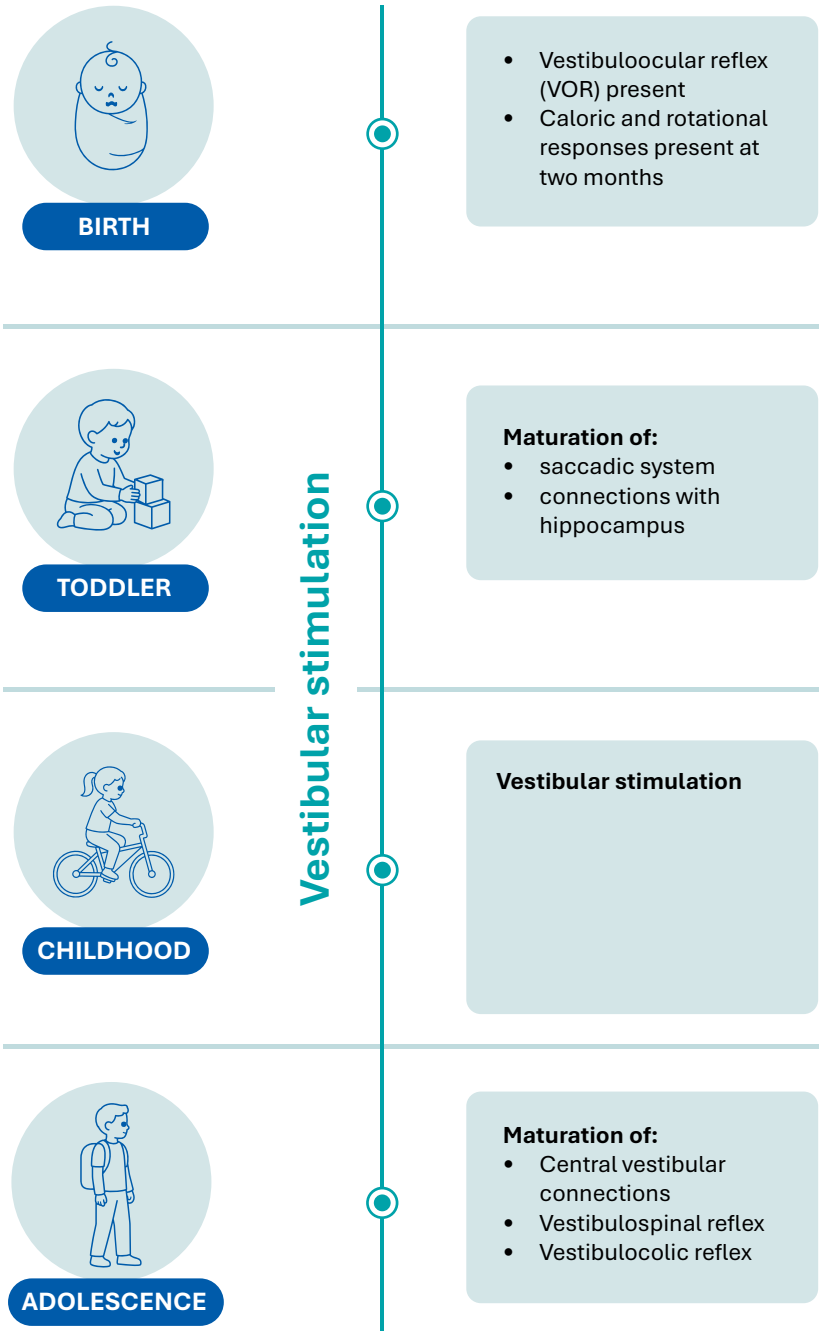


Figure 3.20: Postnatal development of the vestibular system. Source: Urbančič, Battelino and Vozel (2024)

The Human Connectome And The Auditory Cortex

A connectome represents a comprehensive map of effective synaptic connections and neural projections that constitute a nervous system and significantly influence its communicative functions. This intricate network is crucial for understanding the architecture of the brain and the interplay between sensory systems and cognitive processes.

The auditory system plays a pivotal role in the structure and function of the human connectome. Information is processed through auditory pathways, leading to the development of speech and language capabilities. Research highlights the profound impact of auditory experiences on the brain's connectivity, affirming that early auditory input shapes neural circuits essential for communication (Kral et al. 2016).

The implications of childhood hearing loss (CHL) extend beyond mere auditory deficits; they permeate the very fabric of the connectome. Characterising HL as a connectome disease underscores the importance of addressing auditory health holistically. Hearing impairments disrupt the intricate web of synaptic connections, potentially leading to developmental and cognitive challenges.

Neuroplasticity

Neuroplasticity, often referred to as brain plasticity or neural plasticity, is the ability of the central nervous system to adapt and reorganise itself in response to experience, learning and environmental changes. This phenomenon enables the brain to form new neural connections throughout an individual's life, allowing for functional recovery following injury and adaptation to new challenges. Neuroplasticity operates on various levels, from cellular changes (involving individual neurons) to large-scale cortical remapping.

By the end of the first year, the cortex comprises all six layers, signifying the maturation of its structural complexity. In the first three and a half years of life, neurons rapidly multiply in the cortex, especially in the visual, auditory and prefrontal regions. This phase is characterised by creating an overabundance of synaptic connections, laying the foundational architecture for future cognitive functioning.

During this crucial developmental window, myelination of axons also occurs at an accelerated rate, facilitating faster neural communication. Following this period of growth, a significant pruning phase begins around the age of four, during which unnecessary neurons and synapses are eliminated from the visual, auditory and prefrontal cortices. This synaptic

pruning is essential for optimising neural efficiency and functionality, ensuring the remaining connections are robust and relevant to the individual's experiences (Sharma and Cardon 2015).

The Human Balance System

Balance is the ability to maintain the body's centre of mass in alignment with its support base. The balance system reaches its peak maturity around the age of twelve to fifteen. By age two, children exhibit significantly improved balance skills, demonstrating foundational co-ordination in activities such as walking and running. The balance system continues to refine through childhood, with critical developments occurring during pre-adolescence.

This balance function is sustained through a sophisticated interplay of sensorimotor control systems, encompassing sensory inputs from vision, proprioception and the vestibular system. Vision provides essential information about the surrounding environment, while proprioception offers feedback on body position and movement through touch. The vestibular system is critical in maintaining motion, equilibrium and spatial orientation.

Effectively integrating these sensory inputs is crucial for achieving stability, which is then translated into motor outputs that engage the eye and body muscles. When these systems suffer from injury or deficiency, the individual's balance can be significantly compromised. This can manifest in difficulties with co-ordination and stability, ultimately affecting one's ability to perform daily activities safely and efficiently.

Conclusion

A thorough understanding of the anatomy and physiology of paediatric hearing and vestibular structures is paramount for the practical evaluation and treatment of patients. The intricate nature of embryological development in these systems lays the foundation for their subsequent functionality and pathology. As children grow, their auditory and vestibular systems undergo significant maturation, influenced by genetic and environmental factors. This maturation process is critical in identifying deviations that may lead to audiological or balance disorders.

Evaluating the developmental milestones of these structures enables clinicians to detect abnormalities early, facilitating timely interventions that can significantly enhance outcomes. Knowledge of the anatomical variations and physiological norms at various developmental

stages equips healthcare professionals with the insights necessary to tailor assessment and therapeutic strategies to the unique needs of paediatric patients. Moreover, understanding the interplay between anatomy, physiology and maturation can illuminate the reasons behind specific clinical presentations, guiding more effective treatment approaches.

Paediatric practitioners must comprehensively grasp these fundamental concepts to ensure they can provide optimal care and support for children's auditory and vestibular health.

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Chapter 4

Hearing Screening

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'A healthy start leads to a healthy life'

~Ontario Newborn Screening Program

Introduction

Childhood hearing screening programmes can be grouped into three categories: 1) newborn screening, 2) early childhood development screening and 3) school-based screening. These hearing screening services can be provided through nationalised programmes or via targeted and at-risk based approaches.

Successful Newborn Hearing Screening (NBHS) and Early Hearing Detection and Intervention (EHDI) programmes have been implemented in several countries using a variety of screening methods and protocols. It is estimated that nearly one-third of the world's population, primarily in high-income regions, are fully or nearly fully covered by these programmes (WHO, 2021b). These initiatives have been instrumental in the early detection of congenital hearing loss (HL), reducing the average age of diagnosis from 24 to 30 months, to two to three months in high income countries (Harrison, Roush and Wallace 2003).

Despite the increase of NBHS coverage, approximately 36% of children who refer out on a hearing screening do not receive appropriate follow-up care, with this proportion rising to nearly 50% among higher-risk, urban and underserved populations (Harrison, Roush and Wallace 2003). In low- to middle-income countries (LMICs), the lack of relevant policies or

their implementation as well as the lack of human and financial resources and equipment, limited awareness and the associated stigma of HL make the implementation of NBHS challenging (WHO, 2021b).

By the time children reach school age, the prevalence of HL increases, particularly in areas where ear infections and other risk factors are widespread (Refer to Chapter 2 for more information on risk factors for HL). Furthermore, recent trends suggest that the global increase in childhood HL is partly due to rising levels of environmental noise and the use of digital recreational devices which expose children to potentially harmful noise levels at an early age (WHO 2023). These factors emphasise the need for robust and accessible hearing screening programmes, particularly to identify and address HL and reduce the effects it will have on speech, language and cognitive development. This chapter will describe the components of newborn, early childhood and school-based hearing screening and how these screenings can be practically implemented.

Types of Childhood Hearing Screening Initiatives

Hearing screening was conceptionalised in 1957 and by the end of the 20th century NBHS became the industry standard globally (Fisch 1957; WHO 2013). NBHS can be implemented in all facilities where babies are born and/or receive follow-up care. The primary purpose of NBHS is to identify infants who have HL and need further diagnostic testing. In addition, NBHS aims to identify infants with medical conditions that may cause late-onset or progressive HL and to implement a plan for the continued monitoring of their hearing across various phases of childhood development (JCIH 2019). NBHS is still the most effective way to reduce the prevalence and combat the impact of childhood HL (JCIH 2019; WHO 2021a). The lack of NBHS puts children at risk of impaired or limited language development during the prime language acquisition period, and can also lead to associated social, emotional and academic challenges in later life (WHO 2023; Yoshinaga-Itano et al. 2018).

Early childhood hearing screening can be conducted in various settings, including at general practitioners and paediatric consultations, early childhood development centres, well-infant nurseries or healthcare clinics. According to the Joint Committee of Infant Hearing (2019), a paediatrician or another primary health care professional is responsible for monitoring the general health, development and well-being of a child. Hearing screening in these instances is usually conducted using parental questionnaires, observations of the child's ear canal and tympanic membrane using otoscopy and/or tympanometry, or behavioural observations of the child's response to sound (for example, bell-ringing or hand clapping) (Eiserman et al. 2008). While parental concerns are important, studies have shown that parental questionnaires

are often not accurate indicators of childhood HL (Olusanya 2001). Additionally, although otoscopy and tympanometry can help identify anatomical impairments and middle ear disorders, they are not always available in under-resourced areas and do not assess cochlear or auditory nerve functioning. Informal behavioural observations of a child's response to sound are also unreliable for assessing HL in young children (Eiserman et al. 2008). These subjective techniques have shown poor sensitivity for detecting mild and moderate HL and have low specificity, making them unreliable for screening purposes (Downs and Sterritt 1967; Northern and Downs 2002). The effectiveness of early childhood hearing screenings during wellness visits is further compromised by the fact that a significant proportion of children, particularly in lower-resourced countries, do not attend these visits regularly. Therefore, this more informal method of hearing screening should not primarily be relied upon.

School-based hearing screening programmes aim to evaluate the hearing status of children at various educational levels, typically during school entry and at regular intervals thereafter. School (and pre-school) settings present a unique opportunity to conduct universal hearing screening among children, as most children attend school worldwide (Mahomed-Asmail, Swanepoel and Eikelboom 2016). It also provides an opportunity to educate children and teachers about healthy hearing and safe listening practices (WHO 2021a). In countries like South Africa, where there are disparities in access to healthcare services, implementing school-based hearing screening can help bridge the gap for children who may have missed out on early detection and intervention due to socio-economic barriers (Mahomed-Asmail, Swanepoel and Eikelboom 2016). However, it is essential to note that school-based screening cannot replace early detection through NBHS.

While some countries have national NBHS and early childhood hearing screening programmes (Bussé et al. 2021), resource-constrained settings often rely on targeted or risk-based screening due to limited equipment, shortages of trained personnel and broader infrastructural challenges (WHO, 2021a). Targeted approaches prioritise the identification of children of any age who are at increased risk of HL due to factors such as a family history of HL, neonatal intensive care unit (NICU) admission, congenital infections (for example, cytomegalovirus), craniofacial anomalies, syndromes associated with HL, parental concerns and/or exposure to ototoxic medications (see Chapter 2). By concentrating efforts on higher-risk populations, a targeted approach allows for more efficient use of available resources while maximising the likelihood of early detection. Using risk-based approaches can help prevent HL from progressing by allowing earlier interventions, ultimately improving long-term outcomes for children with identified risk factors.

Establishing a Hearing Screening Programme

Whether hearing screenings are conducted as part of NBHS, in early childhood at Early Communication Development (ECD) centres, during home visits or at school age, there are several considerations to ensure their effectiveness. By following a structured approach, programmes can effectively facilitate early identification and intervention for children with HL (JCIH 2019).

a) Collaboration and quality control

Successful hearing screening programmes require collaboration and quality control measures:

1. Collaboration: Facilitate co-operation between public and private institutions, healthcare agencies and individuals involved in the screening, evaluation and intervention processes (JCIH 2019; WHO 2021a).

2. Advisory board formation: Establish an advisory board to support the planning, execution and evaluation of the programme, adapting roles to local needs and resources.

3. Quality control and continuous improvement: Implement processes to maintain and improve the programme's effectiveness through regular quality checks and updates.

4. Identification of care pathways and follow-up mechanisms: Ensure availability of diagnostic audiology and otology services (including medical and surgical services), hearing technology and rehabilitation.

5. Technology: Evaluate the types, availability and validity of screening devices such as OAE, AABR, immittance and pure tone audiometry equipment. These devices should be regularly calibrated and maintained to ensure accuracy and reliability.

6. Availability of personnel: Train and select screeners based on the service delivery setting, including community health workers (CHWs), ECD practitioners, and paraprofessionals.

7. Timing of screening: In a hospital setting, schedule screenings before the infant is discharged to ensure that the necessary follow-up visits can be planned. In community-based settings, schedule screenings during postnatal visits at Midwife Obstetric Units (MOUs) or during the first immunisation visit at six weeks. In the school setting, screen children at preschool/kindergarten, upon school entry and again where possible in grades one, two, three, seven and eleven.

8. Environments: Ensure that screening environments are quiet and free from visual distractions to prevent interference and to ensure accurate results.

9. Follow-up referral criteria: Set clear guidelines for referring children who do not pass the initial hearing screening for further diagnostic evaluations.

10. Information management: Develop a system for recording and managing screening data to ensure accurate tracking and follow-up. (Appendix 4.1 is an example of a screening form.)

b) Reporting and communication protocols
Effective communication and reporting protocols are crucial.

1. Reports: Provide clear, detailed and timely reports on screening results to both physicians and families.

2. Documentation in medical records: Accurately document results in electronic or paper-based medical records.

3. Reporting of results: Ensure results are reported to the appropriate health authorities and databases.

4. Confidential and culturally sensitive communication: Communicate results to families in a manner that respects their cultural backgrounds and is easy to understand.

5. Referral pathways: Identify and define referral pathways to ensure that infants who do not pass screenings have access to appropriate follow-up care either by an audiologist (for diagnostic audiology services), ear-nose- and throat (ENT) specialist or general practitioner.

Audiologists' role in hearing screening programmes

Audiologists play a crucial role in every stage of a hearing screening programme, from planning and implementation to evaluation and follow-up. Their involvement is essential not only at the local level, such as individual hospitals or community clinics, but also at broader levels, including statewide or national initiatives (Mahomed-Asmail, Swanepoel and Eikelboom 2016). Ideally, audiologists guide the clinical and technical aspects of these programmes, ensuring that protocols are meticulously followed and that best practices are upheld.

According to JCIH (2019), during follow-up visits, audiologists should provide comprehensive diagnostic assessments to confirm the presence and degree of HL. They support parents and caregivers in understanding the significance of HL, guiding them through the next steps with empathy and clarity (Chapter 15). Audiologists also evaluate whether the child is a candidate for amplification devices or other sensory and assistive technologies and oversee the fitting and monitoring of amplification devices (Chapters 10 and 11). Their multifaceted involvement in all aspects of hearing screening and intervention ensures that children with HL receive comprehensive, coordinated care, which is tailored to their unique needs and circumstances.

Screening personnel

Personnel should be assigned in accordance with the setting and available resources for the hearing screening programme. The screening personnel may include healthcare professionals (audiologists, speech-language pathologists, ENT specialists, general practitioners), CHWs, ECD practitioners and paraprofessionals (nurses, medical assistants, technicians). Utilising paraprofessionals, CHW and ECD practitioners from the community can enhance the programme by matching the linguistic and cultural diversity of the population, as well as by fostering trust and better communication during the screening process. All screeners should receive comprehensive training in hearing screening methods and referral pathways which must be supplemented with regular monitoring, quality control and ongoing professional development (WHO 2023). Additionally, they should learn the operational aspects of the hearing screening programme, including reporting processes, the specific role of the screener, screening and referral methods, and setting up screening rooms and calibrating equipment properly. They should also be made aware of the use and maintenance of equipment, troubleshooting issues and background noise measurements. Techniques like otoscopy and tympanometry (when available) should also be included so that personnel are fully prepared for their roles. An audiologist or another qualified individual should facilitate this training and monitoring to ensure that high standards are maintained, by utilising existing

resources such as the WHO (2023) training manuals, available here: *https://www.who.int/teams/noncommunicable-diseases/sensory-functions-disability-and-rehabilitation/primary-ear-and-hearing-care-training-manual*.

Setting up Screening Equipment

When setting up to administer hearing screening, routine checks of the equipment are vital to ascertain that the equipment is in working order, that calibration has not noticeably changed, and that attachments, leads and accessories are not defective. A daily listening check should be performed by the screener to rule out distortion and crosstalk and to ensure that there are no defects in major components. It is recommended that the equipment is calibrated at least once a year according to international and national standards (for example, ISO 389-1).

A quiet location should be identified within the hospital or on school or community premises and noise levels should be checked beforehand. Noise levels during hearing screening should not exceed the maximum permissible ambient noise levels (MPANLs) prescribed for the selected tool/headphones. The MPANLs when using TDH39, according to ISO 389-1:1998, are available in Table 4.1 Noise levels can be checked using a sound level meter if available, or by using validated phone applications for noise level measurement (American Academy of Audiology 2011). Examples of phone applications include SLA Lite (iOS), Sound Meter and Noise Detector (Android). If mHealth tools are used for hearing screenings they usually have a built-in noise monitoring system (Swanepoel et al. 2014). Moreover, screening should be paused during transient increases in ambient noise. Ambient noise may occur as a result of ventilation, activity in an adjacent hall or classroom, children moving around in the test room and screening personnel giving instructions. These factors contribute to inaccurate screening and may result in false-positive results. In situations where sound meters are not available, although not ideal, a biologic noise level check can be done by screening personnel, which includes the ability to establish hearing thresholds at least 10 dB HL below the screening level (for example, 15 dB HL for screening conducted at 25 dB HL) at all test frequencies prior to testing. Furthermore, the environment should have minimal visual distractions.

Table 4.1: Maximum permissible ambient noise levels (MPANLs) for TDH 39 headphones in dB SPL according to ISO 389-1: (1998)

Screen intensity	1 kHz	2 kHz	4 kHz
20 dB HL	31.0	37.0	49.7
25 dB HL	36.0	42	54.7

Newborn Hearing Screening (NBHS)

Despite published guidelines from various healthcare bodies in lower-resourced countries (for example, the Health Professions Council of South Africa [2018]) on the importance of EHDI, there is often no standardised implementation of NBHS. Although there is growing awareness of the need for NBHS, clear legislation mandating it is still not fully in effect (de Kock et al. 2016).

The primary goal of EHDI is to make sure that all infants with HL are identified as early as possible and receive timely intervention to optimise speech, language and cognitive outcomes. To guide this approach, the JCIH (2019) established the 1-3-6 rule which outlines three critical and time-sensitive steps: hearing screening by one month of age, a confirmed audiological diagnosis by three months of age, and initiation or appropriate hearing intervention by six months of age. This timeline is based on extensive evidence demonstrating that infants who receive intervention before six months have significantly better language development than those identified later (Yoshinaga-Itano et al. 2018). Consequently, the 1-3-6 rule has become an internationally recognised benchmark for effective infant hearing screening (WHO 2021a; JCIH 2019).

In high-income countries with well-established EHDI programmes, the JCIH now recommends striving towards an even more accelerated 1-2-3 timeline to maximise developmental outcomes (JCIH, 2019). However, in many LMICs, meeting these targets is challenging due to limited equipment, shortages of trained personnel, infrastructure constraints and barriers to follow-up care (WHO, 2021b). As a result, while the 1-3-6 framework remains the global standard, LMICs often adopt a modified or staged implementation.

Types of NBHS

Currently, there are two techniques endorsed for infant hearing screening: OAEs and AABR (HPCSA 2018; JCIH 2019). These tests provide an objective indication of auditory system function but do not estimate frequency-specific behavioural thresholds, and this cannot quantify hearing sensitivity across the hearing spectrum (JCIH 2019). Both screening methods yield a binary result of pass or refer, with an automated decision rule that requires minimal interpretation from the screener (JCIH 2019).

A core component of an effective NBHS protocol is the inclusion of rescreening prior to referral as this reduces false-positive outcomes and unnecessary follow-ups. In larger programmes, using an AABR to rescreen prior to discharge substantially reduces false-positive results (Clemens et al. 2000). This is especially important when screening is performed within the first 24 to 72 hours as there are several transient conductive conditions that can cause an initial refer result. Vernix or debris in the ear canal may interfere with the stimulus delivery and response recording (Shcwarz et al. 2023), while factors such as delivery by caesarean section or vacuum extraction can also lead to higher OAE failure on initial screening (Farlandansky-Gershnabel et al. 2025). Therefore, within the framework, referral for diagnostic testing is indicated when the infant does not pass after the recommended screening attempts (typically two), because indefinite repeat screenings attempts can delay diagnosis and intervention (JCIH 2019).

Otoacoustic Emissions (OAEs)

Otoacoustic emissions (OAEs) (Figure 4.1) are low-intensity sounds generated by the outer hair cells of the cochlea as they actively amplify and fine-tune incoming acoustic signals (HPCSA 2018). This outer hair cell activity produces measurable acoustic energy that travels backward through the middle ear and can be recorded in the ear canal using a sensitive probe microphone. As OAEs reflect the integrity of cochlear outer hair cell function, their presence is a strong indicator of normal peripheral auditory system functioning. There are two primary types of automated OAE measurements used in NBHS: transient-evoked OAEs (TEOAEs) and distortion-product OAEs (DPOAEs). Chapter 6 provides a detailed overview of OAE assessment.

1. TEOAEs: These emissions, also known as cochlear echoes, are produced by the outer hair cells and triggered by response to clicks or tone bursts. These echoes reflect the response of outer hair cells across a broad frequency range and can be recorded quickly and easily without requiring advanced audiological interpretation, making them suitable for NBHS (JCIH 2019). TEOAEs have also been found to have high sensitivity and specificity to identify cochlear dysfunction and can therefore be used as a first stage screen in well-baby programmes (Kemp 2015). However, TEOAEs can be affected by conductive HL, ambient noise and internal noise from the patient.

2. DPOAEs: These emissions are produced when two continuous pure tones are introduced simultaneously into the ear. DPOAEs help predict frequency-specific

hearing sensitivity, particularly in the range of 500 Hz to 8 kHz, allowing more detailed evaluation of cochlear responses in comparison to TEOAEs (JCIH2019). This advantage is not critical for infant hearing screenings. Just as with TEOAEs, DPOAEs are susceptible to vernix, residual middle ear fluid or debris in the external ear canal (Kemp 2015). However, what makes them the preferred choice in NBHS programmes is the fact that DPOAEs perform better in noisy test environments such as NICU, making them a more commonly selected option (HPCSA 2018).

Automated auditory brainstem response (AABR)

The Auditory Brainstem Response test evaluates how sound travels through the auditory system up to the brainstem. It does not rely on behavioural responses and is ideally performed while the baby is naturally asleep. For screening purposes, an automated version, Automated ABR (AABR) is used. This method delivers soft click stimuli to the newborn's ears through earphones connected to a battery-powered device. The electrical responses are recorded using three surface electrodes placed on the head, nape of the neck or shoulder. The device automatically analyses the responses and issues a 'pass' if they match a pre-set response template after a minimum of 1,000 sweeps, or a 'refer' if no match is detected after 15,000 sweeps. If a refer result is obtained, the patient requires further additional diagnostic testing.

AABR devices such as the Vivosonic and Natus screeners are commonly used as they are portable and have simple step-by-step protocols and instructions for use. The Natus in particular is compact and lightweight, easy for use at the bedside and in neonatal wards. Both screeners provide the standard pass or refer results and use disposable electrodes and ear cups that are changed between each patient. The disposable items increase the cost of testing per patient which can be challenging in low-resourced environments.

The MB 11 BERAphone® (Figure 4.1) provides an alternative AABR option to traditional choices (Melagrana et al. 2011). The MB 11 BERAphone® has become increasingly popular as it requires no disposable ear tips or electrodes, reducing the overall cost per screening, which is critical where resources are constrained. This device consists of a handheld headphone with a built-in preamplifier and three fixed electrodes that connects to and are powered by a laptop (Meier et al. 2004). Electrode gel is applied to the electrodes that correlate with the vertex, ground and mastoid sites on a baby's head.

Figure 4.1: (a)



Figure 4.1: (b)



Figure 4.1: (a) A baby undergoing OAE screening
Figure 4.1: (b) A baby undergoing AABR screening

Which screening method should I choose?

AABR screening, although generally more expensive than OAE due to initial capital costs, is recommended for screening infants at risk of auditory neuropathy spectrum disorder (ANSD) before discharge from the hospital (Kezirian et al. 2001). AABR screening can test up to the level of the brainstem, whereas OAE screening only tests up to the level of the cochlea. AABR screening is particularly important for the NICU population, which has a higher prevalence of ANSD due to increased exposure to multiple risk factors for HL. ANSD can only be detected with a neural-based test like the AABR (Sininger 2002). AABR is also suitable for infants who are small for their gestational age or have low birth weight.

However, for infants without risk factors for ANSD, OAE screening is typically recommended. When conducting OAE screening, it is essential to consider ambient noise levels to minimise false-positive results. Additionally, the noise reduction capabilities and signal processing techniques of OAE equipment should be evaluated for different settings, as not all OAE instruments may be suitable for NBHS programmes (Olusanya 2010). Appendix 4.2 can be used when choosing an appropriate screening method. The following steps should be adhered to when conducting OAE and AABR screening:

a) Schedule and environment

- Where possible, post a sign letting others know that hearing screening is in progress.
- Speak to staff and find out if there are quiet times in the wards when screening would be optimal.
- Try to complete screening prior to discharge; if not possible, try and coincide with an immunisation follow-up.

b) Behavioural state of baby

- Speak to caregivers about the baby's next sleep or feeding time; a quiet or sleeping baby is in the optimal state for screening.
- A baby with a clean diaper is a happy, calm baby.
- Swaddle the baby as this keeps it calm and contained. It will also prevent the baby from pulling the OAE probe tip out of the ear during testing.

c) Technique

- For a good probe tip fit on OAE, gently pull the earlobe back to insert the probe. The probe should remain in the ear without support.
- Opt for a slightly larger probe tip as it allows for a tight seal, improving the test accuracy.

- When conducting AABR, put a small amount of electrode gel on each electrode site on the skin so that the baby can get used to the temperature and consistency.

Early Childhood and School-Based Hearing Screening

Early childhood screening is aimed at children post newborn to eight years old and can be conducted in various settings. In resource-constrained countries like South Africa hearing screenings are often conducted in ECD centres (Department of Social Development 2009). ECD centres are established to support the emotional, cognitive and physical growth of children from birth until school age (Department of Social Development 2009). These centres play a role in providing initial access to preventive hearing healthcare for children from underserved communities (Hussein et al. 2018). Research from the US and South Africa have shown that incorporating hearing screenings within early childhood programmes can detect various hearing health issues that may affect language development, literacy, social skills and school readiness (Hussein et al. 2018).

Childhood hearing screenings are generally aimed at children aged four years and older and are primarily conducted within school settings, including pre-school. These hearing screenings may occur as part of an entry assessment or may be risk-based and prompted by concerns raised by caregivers or teachers. In certain high-income countries, school-based hearing screening is mandatory; however, significant variability exists in the protocols, testing methods, threshold criteria and referral guidelines used (Yong et al. 2020).

Early and childhood hearing screenings are recommended on an annual basis; however, the feasibility of implementing annual screening depends on the local epidemiology of ear and hearing disorders, as well as the availability of resources and intervention services within a specific area or country. At a minimum, all children should receive hearing screening in pre-school or kindergarten, at school entry and again where possible in grades one, two, three, seven and eleven (WHO 2023). In addition, targeted screening should be conducted whenever teachers or caregivers express concerns regarding a child's hearing.

The screening should focus on identifying sensorineural, conductive or mixed HL in one or both ears (WHO 2023). The screening referral criteria should be determined based on environmental limitations or health system capacity for referrals. It may include a referral criterion of 20 dB HL for one or both ears, otoscopic examination or tympanometry revealing chronic or acute otitis media, otitis media with effusion, impacted wax, malformations of the ear or

foreign bodies in the ear. The WHO (2023) recommends that early and childhood hearing screening include ear examinations, tympanometry and pure tone air conduction screening.

Ear examinations

Ear examinations include a visual inspection of the outer ear and ear canal (American Academy of Audiology 2011). If any malformations of the outer ear are observed, it should be recorded and a referral to an ENT specialist should be made. Otoscopic examination should be undertaken only where trained staff and decision support for examination and diagnosis of ear diseases are available, either on the spot or asynchronously. Asynchronous otoscopic examinations can be provided using video otoscopy, which necessitates task shifting. There are several available technology options for mobile video-otoscopy, such as HearScope, Firefly, Teslong, Dino-Lite, Otocam and Tyto. Chapter 5 provides details on how otoscopic examination should be performed and interpreted.

Tympanometry

Tympanometry is conducted to determine the middle ear status. As with otoscopic examination, it may only be conducted by trained personnel and interpreted by a clinician either on the spot or at a later time. Chapter 5 provides a detailed overview of how this test should be conducted and results interpreted. Research is underway to create affordable versions of smartphone tympanometers that implement machine-learning algorithms to guide facilitators and layperson screeners to make an accurate diagnosis. Furthermore, in situations where test equipment or clinical support is unavailable, otoscopy and tympanometry may be excluded from the screening protocol (WHO 2023).

Air conduction pure tone screening

Pure tone air conduction hearing screening is used to identify if a child can hear the target threshold across certain test frequencies. This test can only be administered from age four onwards and depends on the child's developmental level and ability to follow instructions. Testing can be conducted manually or using automated sweep tests with mHealth tools (for example, uHear, HearScreen) and should be conducted by trained screening personnel. The target threshold is the intensity at which the tone should be presented and ranges between 20-30 dB HL at frequencies 1-4 kHz. The target threshold selected is based on the test environment and the resources available across the intervention pathway. Using the stricter

criterion of 20 dB HL will identify milder losses but will also result in a significant increase in referrals, with an accompanying increase in healthcare resources. In countries where health resources are limited, alternative screening levels may be necessary to ensure that referral rates do not overwhelm an already burdened health system. A cut-off of 25- or 30-dB HL may provide a reasonable trade-off in terms of identifying HL that is educationally significant. While there is risk of missing some milder losses, this ensures acceptably low referral rates (for example, < 4%) (WHO 2023).

Once the test environment has been selected and MPANLs measured, the equipment should be set up with the child facing the direction of a non-visually distracting space (preferably a wall) and the screening personnel seated behind the child, out of view (Figure 4.2). This set-up ensures that the child does not receive any visual cues during the testing. The child should be instructed to raise his or her hand or place a block into a box (play audiometry, Chapter 5) every time they hear a predetermined tone. The testing should start with a practice round which uses a loud or easily audible tone (for example, 50 dB HL) to condition the child to the screening procedure. Once conditioned, pure tone signals are then presented across the different frequencies, first in the one ear and then in the other. Presentations can occur automatically or manually depending on the device that is being utilised. If a child does not respond at the target threshold at one or more frequencies in either ear, the tester should reinstruct the child, reposition the earphones, and conduct a re-screening within the same screening session in which the initial referral occurred. The presence or absence of a child's response should be recorded for each tone. Children who do not pass the rescreen should be referred for further diagnostic hearing evaluation by an audiologist.



Figure 4.2: Seating position while conducting manual pure tone audiometry screening

Speech-in-noise screening

Speech-in-noise (SIN) screening is recommended for children aged four and older, as younger children typically find speech recognition in noise challenging (Koopmans et al. 2018). The SIN screening test measures the speech reception threshold (SRT), which is the signal-to-noise ratio (SNR) that is required to have a probability of 50% for correct word identification. The SRT is compared to a predetermined age-dependent pass/fail criterion, and a test is either passed (SRT is better than pass/fail criterion) or failed (SRT is worse than pass/fail criterion) (De Sousa et al. 2018). SIN test applications are available, for example the hearing-in-noise test (HINT), QuickSIN Test and HearDigits test. However, for screening purposes and in countries where cultural and linguistic diversity is prevalent, a digit-in-noise test (DIN) may be better suited for example, hearWHO application.

As with pure tone screening, the environment should be quiet with the child facing a non-visually distracting space with the tester seated behind the child. When using an application like hearWHO, the screening personnel should instruct the child to listen to the digits and repeat aloud what they have heard If they are unsure, they should guess the digit they think they may have heard (De Sousa et al. 2018). The headphones are then placed on the child's ears and the screening personnel should start the test, entering the digits provided by the child on the smartphone application to proceed to the next level of presentation. Once the test is completed the application will provide a pass/fail score, with a failure requiring a referral to an audiologist for further evaluation.

Conclusion

The success of hearing screening programmes relies heavily on the collaboration among healthcare professionals, the availability of appropriate technology, and the establishment of clear referral pathways and follow-up mechanisms. In resource-constrained settings, targeted and risk-based screening approaches are essential to maximise the use of limited resources while ensuring that children at higher risk of HL receive timely intervention. Ultimately, the goal of hearing screening programmes is to reduce the prevalence and impact of HL on speech, language and cognitive development, thereby improving the overall quality of life for affected children. Continuous efforts to enhance screening protocols in various contexts, train personnel and raise awareness about the importance of early hearing detection are vital for the success of these initiatives.

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Appendices

Appendix 4.1: Sample Screening Forms

Background information:

Name of school:	**Grade:**
Name of student:	DOB:
Age: _____years_____months	Gender:
First language:	Race:

Otoscopic Examination:

Left ear	**Normal**	**>75% wax in canal**	**Perforation**	**Discharge**	**Other**
Right ear	Normal	>75% wax in canal	Perforation	Discharge	Other

Tympanograms:

	Type	Volume	Pressure	Compliance
Right ear				
Left ear				

Pure tone screening:

	1000 Hz	2000 Hz	4000 Hz	Overall result
Right ear	pass / refer	pass / refer	pass / refer	pass / refer
Left ear	pass/refer	pass / refer	pass / refer	pass / refer

Retest:

	1000 Hz	2000 Hz	4000 Hz	Overall result
Right ear	pass / refer	pass / refer	pass / refer	pass / refer
Left ear	pass / refer	pass / refer	pass / refer	pass / refer

Overall results: Pass/Refer

Refer to: ENT/Audiologist/ Other:_____

Appendix 4.2: Guide: OAE vs AABR

Risk factors for hearing loss	Type of screening	Action
Bacterial meningitis / meningococcal septicaemia	OAE (include extended high frequencies where possible) and AABR	Initial assessment within four weeks of diagnosis. If normal at baseline, follow-up screen at three months, telephonic follow-up at six months. If abnormal at baseline, emergency referral for diagnostic testing, imaging, and ENT referral.
NICU > 5 days	AABR	
Moderate-Severe HIE*	AABR	
Severe Jaundice, NNJ^	AABR	
ECMO**	AABR	
Extremely low birth weight (<1000g) Low/very low birth weight (>1000g-2500g)	AABR	
Maternal infections during pregnancy: Toxoplasmosis, Rubella, Syphilis, CMV	OAE	CMV: Every three months (birth – one year). Every six months (one – three years). Annually (three years and older). Rubella: once-off assessment if normal.

Risk factors for hearing loss	Type of screening	Action
Maternal/child ototoxic medication"	OAE (include extended high frequencies where possible)	Will need diagnostic assessment if the child is old enough (OAE will not pick up high-frequency HL). Monitoring needed depends on length and duration of ototoxic exposure.
Craniofacial abnormalities: cleft palate, Goldenhar, etc.	OAE and AABR	Audiology services with ENT input on a case-by-case basis.
Syndrome related to HL	OAE and AABR	Audiology services with ENT input on a case-by-case basis. If syndrome is related to progressive HL, it will require annual monitoring.
Family history permanent childhood HL	OAE	Once off unless history of progressive HL, then monitor every six months.

***HIE=hypoxic ischemic event; ^NNJ=neonatal jaundice; **ECMO=extracorporeal membrane oxygenation**

Chapter 5

Paediatric Behavioural Testing

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'Children are not little adults. They have unique needs, vulnerabilities, and developmental stages that must be respected and nurtured.'

~Jessica Milligan

Introduction

Children's attention spans are fleeting, and their engagement hinges on creating a nurturing and engaging environment. Recognising the significance of play and alleviating fears and anxieties are pivotal in facilitating a successful assessment process. It is important to use developmentally appropriate assessments, emphasising that chronological age does not necessarily correlate with developmental readiness. By selecting assessments that align with a child's cognitive and emotional development, hearing healthcare professionals (HHP) can ensure a more meaningful and accurate evaluation of auditory function. The assessments described in this chapter are presented based on the usual flow of a paediatric audiology assessment session, but it is important to remember that flexibility is key. The diagnostic test

battery does not necessarily have to follow a specific order, as long as enough physiologic and behavioural information is obtained to make an accurate diagnosis.

A fundamental principle highlighted in this book is the importance of the cross-checking approach. The significance of accurate assessment results for early detection and intervention cannot be overstated. Misdiagnosis or delayed diagnosis can have profound implications for a child's development, speech and language acquisition and overall quality of life. This chapter emphasises the need for thorough and precise assessment protocols to facilitate timely intervention and mitigate potential long-term consequences.

Establishing Rapport

Establishing rapport with a child is an essential step in the assessment process. Before initiating formal assessments, it is crucial to observe the child's general behaviour. Chapter 15 provides more information on ways to establish rapport with the child and their family. Building a trusting relationship with both the child and the caregiver from the outset lays the foundation for effective communication and collaboration throughout the evaluation process, ultimately leading to more accurate diagnoses and personalised interventions for paediatric audiology patients.

This can also be done while taking the case history which is an essential component of any paediatric test battery. A thorough case history involves gathering comprehensive information on prenatal, perinatal, and postnatal development, family hearing history and specific concerns regarding a child's hearing and communication behaviour. This critical initial step helps identify risks for hearing loss (HL) and guides evaluation methods moving forward (JCIH, 2019).

Otoscopy

Otoscopy is used to examine structures of the ear, predominantly the external auditory canal (EAC), tympanic membrane (TM) and the middle ear. It helps HHPs visualise a wide variety of conditions such as acute otitis media (AOM), a perforated TM or a cholesteatoma (Damery et

al. 2019). Delayed diagnosis of various pathologies of the ear can lead to serious conditions or complications (Heah, Soon and Yuen 2016).

While otoscopic examination is important for determining certain pathologies of the ear, there are multiple factors that can lead to its success or failure: poor lighting, a narrow ear canal, poor positioning of the otoscope, and cerumen blockage (Damery et al. 2019). To reduce the challenges involved in performing successful otoscopic examinations, in recent years video-otoscopes have been developed and introduced into clinical practice. The video-otoscope allows the HHP a more controlled and clear view of the TM and surrounding structures by inserting a small camera into the EAC (Tötterman et al. 2019).

Otoscopic examination technique

Children rarely sit still for very long, so when conducting an otoscopic examination the HHP needs to be quick and effective. It is important to first do an external inspection of the outer ear before conducting the otoscopic examination.

An external inspection includes:

1. Inspecting the pinna and surrounding area — the HHP should look for any abnormalities, such as skin tags, preauricular sinuses or atresia.
2. Checking the position of the ears (for example, low set)
3. Noting if there is any tenderness or redness on or around the pinna
4. Noting if there is any sign of otorrhoea or a foul odour

The HHP should also ensure that the otoscopic examination is conducted in a safe manner that accounts for possible movement — this is known as the brace position and helps prevent the otoscope from going too deep into the EAC (Figure 5.1). To obtain the brace position, the HHP should follow the following steps:

1. Hold the otoscope in the hand on the same side as the ear being examined (that is, to inspect the left ear, hold the otoscope in the left hand and use the right hand to manipulate the ear).

2. Hold the otoscope like a pen, between the thumb, first and second finger.
3. Rest the pinkie finger on the child's head to stabilise both the child and the otoscope.
4. Ask the caregiver to assist in stabilising the head if necessary.
5. Manoeuvre the pinna to allow for a clear visualisation of the TM — for children this means gently pulling the pinna down and back.
6. Do the same on the opposite side.



Figure 5.1: Brace position for otoscopic examination

Visualising structures

When conducting otoscopy, the HHP should look for specific identifiers to help determine the status of the EAC and TM. See Table 5.1 for important otoscopic landmarks.

Table 5.1: Otoscopic landmarks

Structure	What to look for
EAC	1. Cerumen impaction 2. Foreign objects 3. Canal oedema (swollen/inflamed EAC) 4. Otorrhoea *Goal: Clear, unobstructed view of the TM.*
TM and middle ear space	1. Colour 2. Bulging or retraction 3. Perforation 4. Presence or absence of expected landmarks *Goal: A grey-coloured, translucent TM, allowing visualisation of the malleus.*

Below, in Figure 5.2, are comparative images of normal and abnormal TMs, including wax, retracted (pulled backward) and perforated eardrums. While a prominent malleus can be an anatomical variant, it is generally associated with retraction or negative middle ear pressure (The Royal Children's Hospital, 2021).

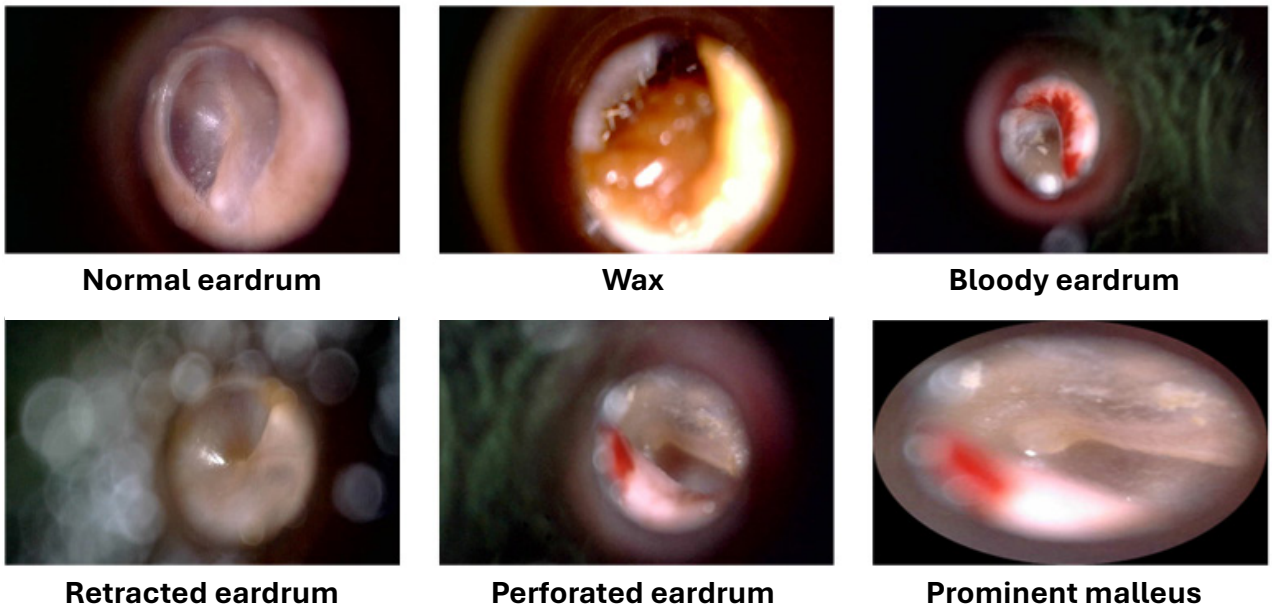

Figure 5.2 Normal and abnormal tympanic membranes

Acoustic Immittance Measures

Acoustic immittance is the umbrella term for tympanometry, acoustic reflex threshold testing and acoustic reflex decay testing (Martin and Clark 2014). These tests are designed to evaluate middle ear functioning and aid in providing a differential diagnosis for various hearing disorders, making them essential for a standard paediatric test battery (American Academy of Audiology 2012; Health Professions Council of South Africa 2012).

Immittance measures are very useful in paediatric assessment as they do not require responses from the child, and they are quick and inexpensive (Hunter and Shahnaz 2013).

These tests are critical in infants and young children as they are most at risk for middle ear problems which typically occur within the first three years of life. Immittance measures are also beneficial because young and vulnerable populations are not always able to express themselves in relation to ear symptoms, and these measures aid in validating the behavioural results obtained.

Tympanometry

Tympanometry measures the mobility of the TM and records middle ear pressure. It is important to remember that tympanometry is not a screening test for HL and cannot identify the presence of HL. It supports the identification of children who have fluid in their middle ear (commonly known as otitis media with effusion [OME]). OME can be asymptomatic in almost 50% of children, can vary in duration and may resolve without intervention (Harlor and Bower 2009).

Tympanometry takes a few seconds to complete and is performed using a soft rubber probe placed at the entrance to the EAC. The probe seals the canal while the machine sends out a soft tone, creating a change in air pressure in the ear canal. The results are automatically recorded as a graph (Tympanogram). A response from the child is not needed for this test. However, patients do need to be quiet and fairly still. Crying, chewing or talking all affect the reliability of tympanometry results. It is helpful to involve the caregiver in calming infants and young children by holding them comfortably on their lap during the tympanometry procedure.

a) Tympanometry classification

The volume, compliance and pressure values of tympanograms are classified into various types based on the shape of the peaks. The primary classifications are A, B and C. Type A indicates normal measurements for all parameters (Table 5.2), while Type B indicates no or limited compliance (<0.1ml), and Type C indicates either a significant increase (> +50 daPa) or decrease (> -200 daPa) in pressure (this is how one can categorise whether the TM is retracted or bulging). Middle ear issues are prevalent in children, making Type B and Type C results common. It is therefore important to acquire bone conduction measures during behavioural testing to identify conductive components. Children who present with abnormal tympanograms should be referred to a physician or ear-, nose- and throat (ENT) specialist to treat any middle ear dysfunction. Chapter 9 describes surgical intervention options for children with CHL or MHL secondary to middle ear dysfunction.

Table 5.2: Guideline for basic paediatric tympanometry norms

Parameter	< 12 years of age	> 12 years of age
Ear Canal Volume (ml)	*0.3 to 0.9 ml*	*0.3 to 1.4 ml*
Compliance (ml)	*0.2 to 0.9 ml*	*0.3 to 1.4 ml*
Pressure (daPa)	*+50 to -200 daPa*	*+50 to -200 daPa*

Source: Minnesota Department of Health (2014)

b) 1 kHz vs 226 Hz Tympanometry

Studies have shown that newborn ears are very resistant to low-frequency tones, likely due to the flaccidity of their middle ear systems (Tharpe and Seewald 2016). For this reason, 226 Hz tympanometry is not recommended for newborns and young infants. Various studies have produced evidence indicating that newborn tympanometry using higher frequencies is more sensitive to changes in middle ear function or status, particularly in infants younger than four months of age (Swanepoel et al. 2007). For this reason, the JCIH (2019) suggests the use of 1 kHz tympanometry for babies that are younger than six months old.

Acoustic reflexes

Acoustics reflexes are useful in paediatric assessment as it can confirm middle ear dysfunction, cochlear HL or retro-cochlear dysfunction. Studies have found that acoustic reflexes in newborns are similar to those of adults, but only if a 1 kHz probe tone is used (Hunter and Shahnaz 2013). Evidence suggests that present acoustic reflexes at normal levels (less than or equal to 100 dB HL) are indicative of a healthy middle ear. When combined with tympanometry, acoustic reflexes are a powerful indicator of middle ear effusion, but alone it is not sufficient.

When performing acoustic reflexes in infants younger than six months, ipsilateral presentation using a 1 kHz probe tone, at 1 kHz and 2 kHz, and no louder than 100 dB SPL, is sufficient

(Katz et al. 2015). In children older than six months, a standard 226 Hz probe tone is suitable and should be paired with tympanometry for accurate measurement of middle ear status.

Reflexes are a quick, easy, cost-effective way to assess the possibility of auditory neuropathy spectrum disorder (ANSD), even in a screening clinic, when combined with OAE or behavioural testing. Patients with risk factors for ANSD, who pass OAEs but have absent or elevated reflexes are at high risk for ANSD and should receive a diagnostic ABR assessment (Berlin et al. 2005).

Visual Reinforcement Audiometry (VRA)

Central to the success of VRA testing is an understanding of children's physical and cognitive milestones. For VRA testing to be effective, children typically need to have reached a developmental stage where they possess adequate motor and trunk control to engage in the behavioural response required, namely turning their heads in response to auditory stimuli (Bonino et al. 2021). During VRA testing, children are conditioned to respond to auditory stimuli by being rewarded with visually reinforcing stimuli, such as animated toys, lights or videos. This conditioning process is based on the principles of operant conditioning, where the desired response (for example, head turned toward the sound) is reinforced through the presentation of a rewarding visual stimulus, which increases the likelihood of the desired behaviour recurring in the future (Norrix 2015). However, children must be cognitively ready to learn to associate the auditory stimulus with a head turn and subsequently with a rewarding visual stimulus. This cognitive readiness to engage in associative learning normally emerges by around six months of age in typically developing children (Tharpe and Seewald 2016). Furthermore, children's cognitive abilities, including attention span, ability to discriminate stimuli and understanding cause-and-effect relationships, play a crucial role in their ability to participate in VRA testing (Tharpe and Seewald 2016).

VRA represents a significant part of the paediatric audiological test battery and offers a reliable and effective method for assessing the hearing abilities of infants and young children. By relying on children's developmental characteristics and utilising conditioning principles, VRA enables the HHP to obtain valuable information about children's auditory function in a manner that is engaging, non-invasive and developmentally appropriate.

VRA equipment and set-up

The testing room should be big enough to accommodate the patient, their caregiver and the distractor, as well as the necessary test equipment and furniture. The testing room should be soundproofed, particularly when testing in the sound field, and should be at least six metres by four metres (British Society of Audiology, 2013).

While there are various ways a VRA room can be set up, Figure 5.3 displays the recommended standard: two testers, two rooms (with the tester operating equipment from a second observation room), allowing for optimal tester view and minimal distraction for the child. This setup is also beneficial for training purposes and for allowing other family members or HHPs to observe the assessment without becoming a distraction. The room setup should also allow the tester to hear what is happening in the test room clearly, ensuring appropriately timed stimulus presentation and communication. With the addition of a two-way headset, the tester and distractor can hear one another and communicate via a more discreet channel.

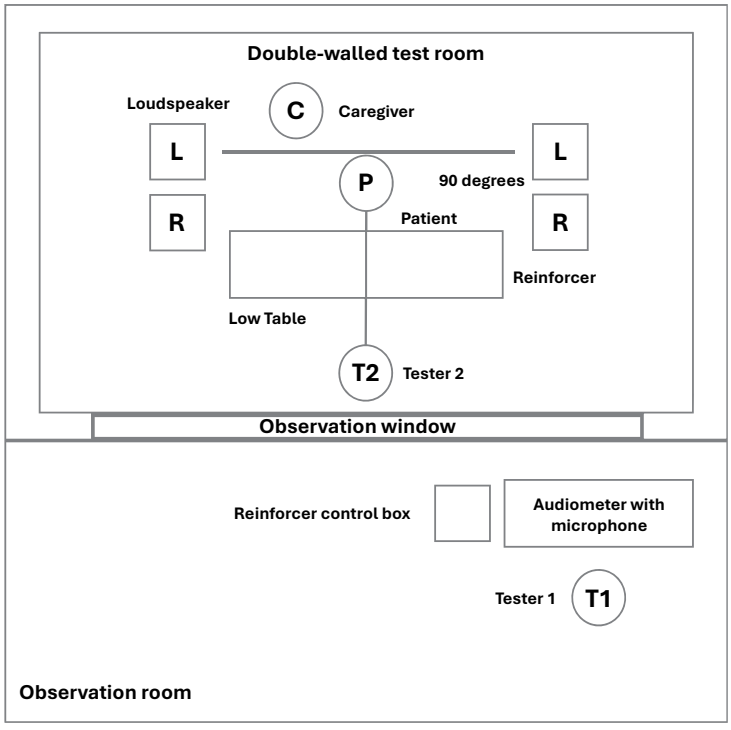

Figure 5.3: Recommended VRA room setup

Loudspeakers should be placed at 90 degrees and at least one metre from the test position at each side (BSA 2013). Ideally, the speaker should be level with the child's head as this allows for the most effective conditioning and establishment of minimum response levels. The reinforcers should be positioned at 90 degrees in order to obtain a clear head turn and should not be more than one or two metres away from the child. There should be minimal distance between the loudspeaker and the reinforcer to ensure the best conditioning when testing using sound field stimuli (that is, one above the other). Adjustable reinforcers are useful as they can be moved closer to the child to assist with better reinforcement. This is particularly useful for children with limited visual ability or reduced developmental age. However, loudspeaker calibration should not be compromised. Additionally, having reinforcers positioned on either side of the child allows them to be rewarded on the side that they prefer (especially when testing with ear-specific transducers or if there are mobility limitations).

Some children prefer videos or Graphic Interchange Format (GIF) over the traditional light-up toys. Children with Autism Spectrum Disorder (ASD) often prefer simple, black line drawings or monotone images. Alternatively, various themed slideshows can be used, such as those focusing on the 'cartoon-of-the-moment'. Observations from initial interactions are especially useful as they may provide insight on whether a child prefers video reinforcers or the 'old-school' light-up reinforcers. Having an array of themed slideshows ensures that the HHP has the correct material for a variety of age and population groups. This way the HHP will be able to switch to more interesting reinforcers if the child loses focus.

Patient seating

Seating required during testing is dependent on a child's developmental age and their level of comfort in the test environment. Regardless of the type of seating used, the child should always be placed in the centre of the test room, so that 90-degree azimuth is used as reference for a clear head turn. Ideally, for testing young children and infants, the use of a highchair is most practical (preferably one with straps to limit movement and sliding out under the table). A highchair will keep the child contained and limit distraction and movement, which is essential when trying to condition and maintain concentration (Figure 5.4). Using a highchair will also eliminate the child's reliance on their caregiver, as well as any possible bias or assistance a caregiver may provide in localising stimuli.

For slightly older children, a small table and chair that are easy to clean but aid in keeping noise levels low are beneficial for distraction and engagement. This setup is also useful in limiting the child's interaction with the various distractors. Children with cognitive or physical limitations may not be able to sit in a highchair or at a table and can in such cases be tested in either their wheelchair or buggy (for example, children with cerebral palsy). Their assistive device can replace the highchair or table in the centre of the room.

Understanding the unique preferences of children, especially those with ASD, is vital. Some may prefer being on the floor rather than in a chair. Getting down to their level can significantly impact session success, as forcing them into an uncomfortable position where they feel caged in may disrupt the testing process.



Figure 5.4: Set up with a highchair

Distractors

Distractors should be both age- and developmentally appropriate. It is equally important to ensure that distractors are regularly disinfected. Distractor toys should be attention-grabbing but also not overly distracting (a fine balance must be struck), while being silent so as not to confuse the child during testing. Light-up toys are often enjoyed the most.

Distractor toys should be kept out of sight by storing them in a locked box or cupboard in the test room. In this way, the child does not get overwhelmed. Having easy access to distractors are essential so that the HHP can quickly switch between toys as needed. The HHP should

try to limit the child's physical interaction with the distractor toys so that they do not lose focus on the task at hand (the VRA assessment).

Managing distractors effectively (in terms of toys and the environment) is essential for accurate results. Maintaining distractor competence and controlling noise levels while avoiding direct cues during stimulus presentation is critical. The HHP's distance from the child should also be considered: Continuous engagement should be maintained without the child being overwhelmed. To maintain focus during VRA sessions, minimising distractions, such as siblings that may have accompanied the caregiver to the appointment, is essential. Siblings may inadvertently provide cues or disrupt the testing environment with noise, affecting the accuracy of results.

Incorporating these techniques and considerations into VRA sessions ensures thorough and accurate assessment of paediatric auditory function, which in turn facilitates timely and appropriate intervention.

Transducers

Often, sound field testing is selected as the first choice when conducting VRA. However, over the years, sound field testing as the first option has been challenged. Ear-specific information is desired, and as such, earphones are required for testing. Testing under earphones will also prevent HHPs from missing a child with a unilateral HL.

Insert earphones can be purchased in various sizes, making it possible to obtain ear-specific responses during testing. If the HHP can insert an OAE probe or tympanometry probe in the child's ear, they should be trying to test behaviourally using insert earphones. In certain cases (that is, microtia or extreme sensitivity to having ears touched) this is not possible, and then the HHP should consider supra-aural headphones. Additionally, bone conduction testing can also be completed using VRA. To ensure comfort during testing, the clinician can switch out the conventional hardband and rather make use of a bone conduction device softband, tightly securing the bone conductor in position (Figure 5.5).



Figure 5.5: Example of bone conduction testing using softband

VRA Test Technique

VRA serves as a pivotal procedure in assessing the hearing abilities of children who are typically aged between six and twenty-four months, and for those with additional disabilities or challenges that may impede more conventional testing methods like play- or standard audiometry. The VRA technique proves particularly beneficial for children who are unable to grasp blocks, toys or a push-button due to physical limitations, or those facing cognitive and attention challenges, such as those with ASD.

a) Frequency selection

When initiating VRA testing, it is essential to select frequencies strategically. One should begin by testing low (500 Hz) and high (4 kHz) frequencies in each ear before filling in the rest of the frequencies. The Ling sounds or LMH 10 are ten specific sounds that cover low, mid and high frequencies of human speech. Some children only respond to speech type stimuli, as such the LMH 10 sounds can be used to check detection at various frequencies (Madell and Hewitt 2021). Some practical resources have been included as appendices at the end of this chapter to show useful ways to capture VRA responses as well as Ling responses.

Starting with speech babble, speech detection, calling the child's name or utilising the Ling sound audiogram (Madell and Hewitt 2021) should be considered to assess various frequency ranges:

- aah /ɑ/ (as in 'car')
- ee: /i/ (as in 'see')
- ooh: /ʊ/ (as in 'book')
- sh: /ʃ/ (as in 'shoe')
- s: /s/ (as in 'see')
- m: /m/ (as in 'man')
- n: /n/ (as in 'no')
- h: /h/ (as in 'hop')
- z: /z/ (as in 'buzz')
- j: /dʒ// (as in 'jump')

b) Response recognition

During VRA testing, discerning a response can be subtle. While an eyeblink or a smile may indicate a reaction to the auditory stimulus, it requires careful observation and judgement. The HHP must be careful not to mistake involuntary responses or unrelated behaviours for a genuine response to the stimulus. Careful recording of repeated, observed responses is essential for accurate assessment results.

c) Team collaboration and caregiver involvement

Successful VRA sessions often involve teamwork, including occupational therapists for regulation support and caregivers for active involvement. Instructing caregivers to participate in the assessment not only aids in maintaining the child's attention but also serves as a valuable counselling tool if the child presents with severe-to-profound HL, with no response being elicited in sound field testing.

d) Stimulus Variation

To sustain the child's interest and prevent false responses, varying the presentation of tones between ears without a predictable pattern is crucial. Balancing the need for thorough conditioning with the child's limited attention span is essential to avoid losing the child's engagement during the session.

e) Avoiding non-verbal cues

Non-verbal cues, such as eye-widening or prompting, must be avoided, as children are highly perceptive and may respond falsely based on these cues. Caregivers should also be

instructed to refrain from providing cues through body language or head movements if the child is sitting on their lap.

f) Bone conduction conditioning

In cases where severe-profound HL is suspected, bone conduction stimulation may be necessary for conditioning. Visual and animated techniques are particularly effective in engaging children with severe-to-profound HL.

g) Microtia considerations

When testing children with microtia, conducting both air conduction and bone conduction tests on the affected ear is crucial for comprehensive evaluation and subsequent selection of assistive technology.

VRA Procedures

VRA is a systematic method that is usually employed to evaluate the hearing abilities of typically developing children (aged six to 24 months) who can sit unsupported and possess good head and neck control. This section outlines the step-by-step procedures involved in conducting VRA, along with strategies to ensure accurate and reliable results. Table 5.3 details VRA normative responses per age group.

a) Brief case history

Prior to commencing the VRA assessment, conduct a brief case history with the caregivers, and observe the child before any formal assessments begin. It is crucial not to miss the opportunity to test the child, and any incomplete information can be gathered after the testing session.

b) Explanation to caregivers

The test procedure should be explained to the caregivers to ensure their co-operation and understanding. They should be informed of the following:

- If the child is seated on a chair, the caregiver should be seated slightly to the left, facing the child.
- Caregivers should be instructed not to talk to the child or provide any cues and to keep the room as quiet as possible.
- The test assistant will sit in front of the child, using a developmentally appropriate toy to maintain the child's focus on the midline. They should not sit too close to the child.

c) Testing procedure

The VRA testing procedure involves the following steps:

- The HHP should always attempt to obtain ear-specific responses under insert earphones or headphones, where possible.
- Initially, appropriately loud enough (typically 65-70 dB HL) warble tones or speech babble should be presented, along with a lighted toy or screen to condition the child.
- After the child turns their head, their attention should be redirected to the midline.
- If the child does not turn their head, the conditioning stimulus intensity should be increased, with conditioning stimulus consistently playing together with the visual stimulus (lighted toy or screen). This should continue until the HHP is confident that the child is appropriately conditioned.
- Once the child is appropriately conditioned, minimum response levels should be obtained using either a descending or ascending approach, depending on the child's response.
- Testing frequencies should start with 500 Hz, then 4 kHz, 1 kHz, and 2 kHz, and if possible, 250 Hz.

**Special Considerations: If testing aided free field thresholds using hearing aids and VRA, warble tones should be used instead of narrow band noise (NBN).*

Table 5.3: VRA normative responses

Age	Minimum response level	Comments
4-7 months	≤ 40dBHL	A localising response (eye or head turn) is sometimes acceptable, but a clear 90-degree head turn should be required as an acceptable and reliable response. Note that, before recording a minimum response level, a response must have been observed twice at a particular level for a specific frequency. If a false-positive is obtained (turning head when no sound was presented), re-check that frequency and intensity.
7 – 16 months	≤ 30dBHL	
16+ months	≤ 25dBHL	

Source: Northern and Downs (2002)

Conditioned Play Audiometry (CPA)

CPA is a dynamic method used to assess the hearing abilities of children through play-based activities. This section outlines the structured procedures involved in conducting CPA, including both conditioning and testing phases, along with techniques for efficient and accurate assessment. Table 5.4 details normative values for CPA per age group.

Conditioning Phase

a) Toy selection and setup

The child should be provided with an engaging toy, such as a stacking toy or blocks in a

container. All components of the toy should be visible and accessible. When placing the earphones it is important not to draw attention to the procedure by telling the child 'It is not going to hurt'. Instead, positive information should be shared, like 'Wow, now you look like an aeroplane pilot! Let's play the game.'

b) Conditioning response

A block should be positioned in the child's hand and they should be instructed to listen carefully. The test should begin in the conditioning phase by presenting a pulsed pure tone at either 500 Hz or 1 kHz at a loud intensity, approximately 50-60 dB HL. The intensity level should be adjusted as needed, especially for children with suspected severe-profound HL, to approximately 80-100 dB HL.

c) Associative learning

A sufficiently loud, conditioning sound should be presented together with exaggerated movement, such as placing the block in the container, to establish an associative response. This procedure should be repeated with assistance for the first two presentations and assistance should gradually be withheld for subsequent presentations to assess the child's ability to respond unaided.

d) Threshold establishment

Once the child is conditioned, the intensity level should be decreased to establish the threshold using the Hughson and Westlake (1944) method, that is,10 dB HL down, 5 dB HL up. The child should be encouraged by using positive verbal or gesturing reinforcement with engagement maintained throughout the conditioning phase.

Testing phase (Threshold determination)

a) Frequency testing

Testing should begin with the right ear or the better ear, if known, and followed with these frequencies:

- 500 Hz Right & Left
- 4 kHz Right & Left
- 1 kHz Right & Left
- 2 kHz Right & Left

It is better to obtain one low and one high frequency in each ear than to have a complete set of frequencies in only one ear and no information on the other ear.

b) Bone conduction testing

Bone conduction testing should be performed as part of the threshold determination process, with masking applied if necessary for older children. Breaks should be provided as needed to ensure the child's comfort and co-operation. The bone conductor can be secured by a softband, if the child does not tolerate the hard, steel headband.

c) Masking procedures

Masking is necessary in air conduction testing when there is a difference in the air conduction thresholds of ≥ 40 dB HL at any frequency compared to the air conduction threshold in the contralateral ear. If using insert earphones are being used, masking is necessary when the air conduction difference between the two ears is ≥ 60 dB HL. For bone conduction testing, masking is required when there is an air-bone gap of ≥ 10 – 15 dB HL (Katz et al. 2015). Explaining the concept of masking may be confusing for the child. Instead of attempting to explain using long, complicated sentences, continuous masking sound be used without drawing attention to it. A pure tone at a level easily detectable above the masking sound should be presented and the child should be assisted in placing the testing object (toy block) in the container as practised in the conditioning phase.

d) Suprathreshold retesting

If the child exhibits inattention or false responses during testing, they should be reconditioned and retested at the suprathreshold level before proceeding with threshold determination. The HHP may switch between ears or different stimuli can be used if the child becomes bored. The type of stimulus used at different frequencies should be recorded.

e) Guarding against patterns

Stimuli should be presented randomly to prevent the child from anticipating the next tone. The HHP can occasionally wait a few seconds before presenting a tone, to maintain the child's attention.

Interpretation of results

a) Threshold acceptance

Responses at 10 dB HL should be accepted instead of spending excessive time attempting to obtain 0 dB HL or 5 dB HL thresholds. It should be indicated on the audiogram that these are minimum response levels.

b) Monitoring responses

If the child's responses suddenly change or deviate from expected levels, the previous thresholds should be repeated to check for reliability. The following factors should be considered: concentration, testing materials or the need for a short break.

c) Abnormal responses

If tympanometry indicates normal middle ear function but the threshold for one or more test frequencies exceeds 15 dB HL for air or bone conduction, a follow-up assessment should be scheduled. Children's attention levels can fluctuate, and more reliable responses may be obtained after rest or at a different time of day (that is, after a nap, or earlier in the morning).

Table 5.4: Norms for CPA

Normative response	Age	Condition
≤ 15 dB HL	*> 24 months*	*Transducer (AC or BC)*
≤25 dB HL	*>16 months*	*Free field*

Source: Northern and Downs (2002)

By following these structured procedures and techniques, HHPs can conduct CPA effectively, ensuring comprehensive assessment of paediatric hearing abilities and appropriate follow-up appointments, as needed.

Standard Audiometry

Standard audiometry is a behavioural hearing test that assesses the hearing sensitivity of the individual through conventional test methods. It typically makes use of pure-tone stimuli with responses via hand raises or the push of a response button. This section highlights the test procedure involved, including speech audiometry.

Preparation phase

a) Instructions

Clear instructions should be provided to the child so that they understand what is expected of them during testing. Children should be informed that they will hear a beep and that as soon as they hear it, they should push the button. They should be informed that the beeps may be loud and soft, and that it is important to push the button when they hear them.

b) Conditioning

The child should be allowed to hold the response button in their dominant hand, and they

should be instructed to listen carefully. The test should begin in the conditioning phase by presenting a pulsed pure tone at either 500 Hz or 1 kHz at a loud intensity, approximately 50-60 dB HL. Make sure that that they hear the sound and are aware of what they should be listening for. The intensity level should be adjusted as needed, especially for children with suspected severe-profound HL, to approximately 80-100 dB HL.

c) Threshold establishment

Once the child is conditioned, the intensity level should be decreased to establish the threshold using the Hughston Westlake (1944) method. The child should be encouraged by using positive verbal or gesturing reinforcement and maintaining engagement throughout. If inconsistent results are obtained, the child should be reinstructed and testing resumed at a louder intensity. Determining thresholds, bone conduction testing, masking indications and interpretation of results for Standard Audiometry is the same as for CPA (see section above).

Speech Audiometry

Speech audiometry is an umbrella term used to describe a series of audiometric tests in which speech is the primary stimulus. These tests can be done both in quiet and in noise. When combined with pure-tone audiometry, they can assist in determining the type and degree of HL (Eggermont 2017). Additionally, pure-tone audiometry is limited in its predictability of an individual's ability to recognise speech. Speech audiometry is an essential tool in a paediatric hearing assessment. Speech testing can help identify the reliability of a child's responses by cross-checking their speech and pure tone audiometry results.

Speech detection testing

During speech detection testing (SDT), the HHP should determine whether the child can hear speech. It is the most basic of speech tests as no level of understanding is required. This is often the mode of speech testing used when conducting VRA testing. It is presented via loudspeaker or earphones.

The speech stimuli should be presented at various intensity levels and an observation of whether the child reacts to the stimulus noted. Frequency-specific sounds (that is, the Ling sounds) should be presented at the level at which the child detects at least 50% of the

speech, which is considered their speech detection level. Starting at 10 dB HL is recommended, increasing intensity in 5 dB HL increments until a response is obtained.

Speech recognition testing

Speech recognition testing (SRT) builds on the foundation of SDT, with the expectation that the child not only detects the speech but is also able to recognise the word/s presented to them. These words are known as spondee words, of which various standardised lists exist. SRT is used to confirm the reliability of the pure tone audiogram. SRT should always be attempted under earphones.

a) Materials

No more than five spondee toys or trochee toys (trochee words have two syllables: the first is stressed and the second is unstressed, which is similar to spondee words) should be used. These are most appropriate for the two-to-three-year-old age group. Spondee pictures boards are most suitable for ages four and older and can include six-to-eight pictures.

b) Procedure

It is essential to make sure that the child is familiar with the words being used. For SRT, testing should commence at 25 dB HL above the child's pure tone average. Their recognition level is the lowest intensity if they are able to identify 50% of the words correctly.

Word identification testing

This form of speech testing is when the child is presented with a list of phonetically balanced words at a single frequency, which they must repeat out loud. They are scored on whether they repeat the words correctly or incorrectly, and the result is expressed as a percentage. Word identification scores can typically be obtained from around four years old(or two to three year old if the child has good language skills).

a) Stimuli

Recorded material or live voice are available. There has been a movement away from live voice testing due to variations in tester tone and pronunciation of words. Most of the newer

audiometers have built-in recorded speech tests available making this more accessible than previous years. The first list should be presented at 50 dB HL or 30 dB HL above the SRT.

b) Material

The most common clinically used word identification tests are the phonetically balanced kindergarten 50-word list (PBK-50), the Word Intelligibility by Picture Identification (WIPI) test and The Northwestern University Children's Perception of Speech (NU-CHIPS) test. These are available in various countries and cover a range of vocabulary ages. Typically, one list (25 words) is presented at each level.

c) Responses

Non-verbal responses or pointing is accepted for closed-set testing, with the HHP providing positive reinforcement. If a child's vocabulary is adequate, then a verbal response is required.

d) Results

To calculate results, the number of correct responses (out of 25) should be multiplied by four to obtain the percentage word score (out of 100%). If the score is between sixteen and 88%, it is recommended to increase the word list to 50 words as this will improve the reliability of the results. The total percentage score at each level is then plotted on the speech audiogram. Once plotted, it is critical to check for retro-cochlear pathology by using the rollover index formula. A rollover index of more than 0.45 is indicative of a retro-cochlear pathology and further referral is indicated (Katz et al. 2015). The child's actual responses should be recorded to allow for phonemic analysis, if indicated.

It is important, as far as possible, to obtain word identification scores for a child who has been referred with the following concerns: Poor school performance, limited attention, auditory memory challenges or a language delay, as these identifiers are commonly associated with auditory processing difficulties.

Conclusion

The need for assessing hearing in young children has been understood for almost a century. During this time, test procedures have evolved, with evidence showing that using developmentally appropriate test techniques allows for an efficient and precise manner of obtaining audiological outcomes. The information that HHPs are able to obtain through behavioural testing is the quantifiable functional hearing of a child. It is essential that all test protocols and recordings are carried out in consistent and repeatable ways to ensure that the authenticity of results is maintained.

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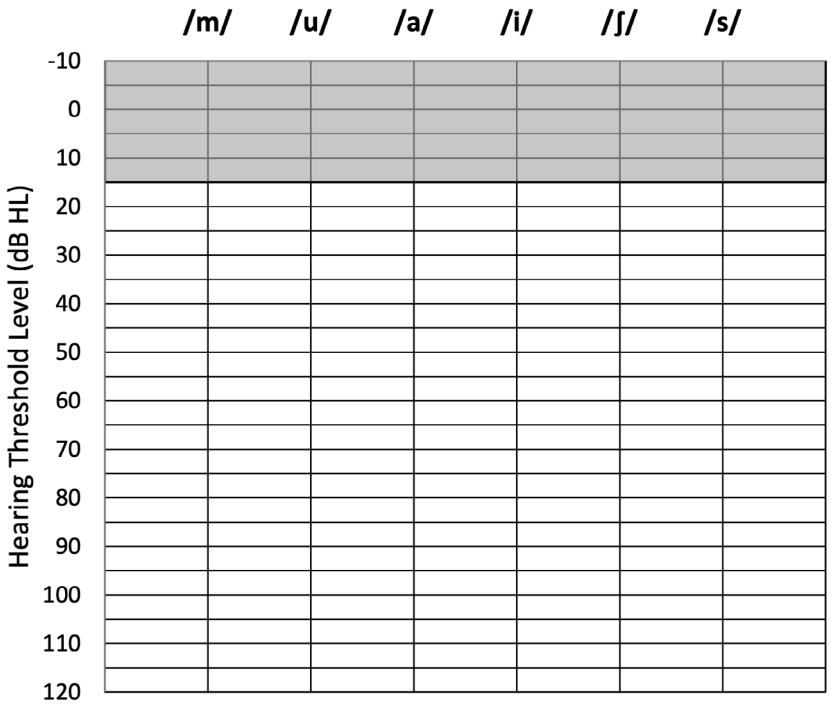
Appendices

Appendix 1. Sample Ling audiogram



Appendix 2. Sample VRA recording audiogram

Patient Hospital Sticker

Referred by: _____ Contact No.: _____ Test Date: _____ Stats No.: _____
Audiologist 1: _____ Test No.: _____
Audiologist 2: _____ Reliability: POOR FAIR GOOD

TRANSDUCER	
Insert Earphones	
Supra-aural headphones	
Speaker / Soundfield	

STIMULUS	
Warble/FM Tone	
Narrowband	
Live speech	

ICD CODES	
RIGHT	
LEFT	

PAEDIATRIC AUDIOGRAM

VISUAL RESPONSE AUDIOMETRY (VRA)				
	RIGHT		LEFT	
	dBHL	Response	dBHL	Response
250Hz				
500Hz				
1000Hz				
2000Hz				
4000Hz				
6000Hz				
SDT				

AGE-APPROPRIATE SOUNDFIELD NORMS FOR NORMAL HEARING		
MONTHS	dBHL	X
0-4	≤ 60	
4-7	≤ 40	
7-16	≤ 30	
>16	≤ 25	

ABBREVIATIONS	
LOC	Localised Response
N	Neutral Response
CON	Conditioning Response
NR	No Response
MR	Minimum Response

TYPMANOGRAM		
PROBE TONE: □226Hz/□1kHz	RIGHT	LEFT
TYPE		
PRESSURE		
COMPLIANCE		
ECV		

IPSI-LATERAL ACOUSTIC REFLEXES				
	500Hz	1kHz	2kHz	4kHz
RIGHT				
LEFT				

SCREENING OAEs		
	RIGHT	LEFT
PASS		
REFER		
NOISE		
CNT		

AABR SCREENER		
	RIGHT	LEFT
PASS		
REFER		
NOISE		
CNT		

NOTES:

HISTORY:

SUMMARY & PLAN:

Chapter 6

Electrophysiological Evaluations

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'Fast is fine, but accuracy is everything.'

~Wyatt Earp

Introduction

Hearing healthcare professionals (HHPs) are trusted by parents and caregivers to evaluate their child's hearing and provide them with answers and guidance. Understanding and respecting the fact that most parents will be very anxious by the time they enter a diagnostic clinical setting will highlight the gravity of the HHP's role in their child's life.

Physiological diagnostic tools like Auditory Brainstem Response (ABR) and Otoacoustic Emissions (OAE) offer valuable insights into the functioning of a young patient's auditory system. It allows the opportunity to cross-check various findings and physiologically confirm the presence or absence of a variety of dysfunctions, especially in 'difficult-to-test' populations. According to the clinical guidance document of the American Academy of Audiology (2020), 'difficult-to-test' is a term that refers to 'children with developmental delays or other conditions that affect communication, and cooperation'.

Physiological tests help to pinpoint the potential site of the lesion, provide guidance to do further testing when needed and support the HHP in decision-making regarding audiological management. This chapter will provide practical insights on optimising the test environment and patient set-up, tailoring the test battery for the specific patient population, prioritising information gathering in an efficient manner and approaching these test procedures with respect and caution.

This chapter is divided into three sections:

- Electrophysiological equipment setup and environmental considerations
- Otoacoustic emissions (OAE)
- Auditory Brainstem Response (ABR) and Auditory Steady State Response (ASSR)

Electrophysiological Equipment Setup and Environmental Considerations

The accuracy of both OAE and ABR evaluations is heavily influenced by the test environment and equipment setup. The room location within the clinical setting will have a significant impact on successful testing when performing diagnostic OAEs and ABRs, and a quiet room with good acoustics will always be preferable. It is thus important to select a location that is further away from high levels of 'foot traffic' and general ambient noise (for example, the reception area, bathrooms and waiting rooms). Whereas a sound-treated room may be ideal for minimising background noise and reverberation, adding carpets and curtains is a simple way to improve the test environment for OAE and ABR measurement.

When conducting OAE measurement, the test space should be arranged in a way to facilitate easy movement between the equipment and the patient. The room should also allow for a comfortable test position for the patient and caregiver while the measurement is in progress. Seating options will differ based on the age of the patient. Smaller infants can be tested comfortably in their parents' arms or in their baby carrier, and a toddler might be more comfortable seated on their parent's lap or independently on a chair. Therefore, the physical space and size of the test room must be a consideration when selecting a room for OAE measurement.

The workspace for ABR measurements is usually carefully prepared to allow for easy access and optimal patient positioning. Parker, Slack and Skoe (2020) found that a simulated home environment delivered comparable quality ABR recordings compared to traditional laboratory settings. However, although this may be true for ABR, ambient noise levels might have a more pronounced impact on the integrity of DPOAE and TEOAE measures, as highlighted by Kanji (2019). Therefore, if the test system allows for both ABR and OAE testing, the ambient noise levels preferable for OAE measurements should be taken into consideration when choosing the location of the test room.

Another critical consideration, more so for ABR testing, is the electrical circuit characteristics of the clinical room. Electrical circuitry might influence the selection of a suitable room within a larger practice or hospital setting. Within a hospital setting, proximity to the radiology department, operating theatres, elevators, communication systems and anaesthesiology monitoring equipment may increase the likelihood of encountering troublesome artifacts on a regular basis (Stegeman and De Weerd 1990). According to Hall and Swanepoel (2010), accurate electrophysiological estimation of hearing thresholds is dependent on the ability to detect a signal in the presence of noise. Noise interferes with the recording in several ways and might arise from multiple varied sources, for example, airborne electrical or electromagnetic energy, electrical circuit interference, rectified radio frequency noise, physiological electrical artifact originating from the patient's nervous system and a myogenic (muscle) artifact (Hall and Swanepoel 2010). Although newer generation ABR equipment can filter out unwanted artefacts and noise, it is recommended to still make every effort to minimise interference where possible. Relying on these filtering technologies alone to improve waveform quality may result in 'smoothing' waveforms to an extent where analysis of the waveforms becomes very challenging and errors occur more frequently.

Various options are available to facilitate a comfortable patient position, even in small clinical spaces. Generally, the ABR clinical setting will require an examination bed, especially if sedation is being used. However, a comfortable couch or 'lazy boy' has proven to be useful in smaller spaces. In some cases, the test procedure can be performed while the patient is resting in a baby carrier or in the arms of the caregiver/parent. This is mainly to ensure that the patient is as comfortable as possible and to ensure that they remain asleep for the duration of the test. It is important to be cognisant of other physical comfort factors such as body temperature (too cold or too warm), movement restriction or even anxiety, as these factors may also influence the patient's ability to remain calm and quiet for the duration of the test.

Before any electrophysiological evaluation appointment, the equipment should be checked for optimal functioning, and all necessary consumables should be made easily accessible to facilitate quick and efficient testing. These steps should form part of the clinical 'biological checks' in general, and are non-negotiable. The last thing any clinician wants to face is faulty equipment or lack of consumables after their patient is already asleep and/or sedated.

Otoacoustic Emissions

OAEs were first described in the 1970s by David Kemp and are considered a well-established test procedure in the general diagnostic test battery. The presence of cochlear emissions was first hypothesised in the 1940s based on complex mathematical models of cochlear nonlinearity, but it was only truly proven in the late 1970s. Only then could low-intensity sounds that are produced by the cochlea could be successfully recorded using highly sensitive microphones (Campbell and Mullin 2023).

OAEs offer valuable insight into the outer hair cell functioning of the cochlea and are particularly helpful in understanding the overall clinical picture of young patients. To employ OAEs effectively in practice, HHPs must understand the mechanism by which OAEs are generated. According to Prieve and Lamson (2014), OAEs reflect the functioning of mechanical processes in the inner ear (specifically the outer hair cells) and are a byproduct of the cochlear amplifier rather than a reflection of the cochlea's sensory or neural functioning. According to Siegel and Kim (1982), OAEs may be present despite a severed acoustic nerve (CN VIII) and are typically absent in persons presenting with cochlear hearing loss (HL) (Siegel and Kim 1982).

OAE testing is a relatively simple test procedure, and technological advancements offer significantly improved ease of application and faster test speed. It is also relatively cost-effective and does not require multiple consumables to administer successfully. OAEs have shown tremendous diagnostic value since the early 1980s, and this technique has been enthusiastically embraced worldwide ever since.

There are four known types of OAEs, namely, spontaneous otoacoustic emissions (SOAEs), transient otoacoustic emissions (TEOAEs), distortion product otoacoustic emissions (DPOAEs) and sustained-frequency otoacoustic emissions (SFOAEs) (Campbell and Mullin 2023). Clinically today, two types of OAEs are mainly utilised in practice: TEOAEs and DPOAEs. The choice of the type of emission will depend on factors such as the patient's age range, the type of evaluation (screening vs diagnostic) or the target frequencies. The major differences between TEOAEs and DPOAEs are summarised in Table 6.1.

Table 6.1: Differences between TEOAEs and DPOAEs

TEOAEs	DPOAEs
• Mainly used in newborn hearing screening procedures	• Offers more frequency-specific information
• Most sensitive at test frequencies between 500 Hz and 4 kHz	• More sensitive to higher frequency regions (>4 kHz), which is particularly valuable for ototoxic monitoring
• More limited frequency specificity: TEOAE response is linked to a larger region of the cochlea. The detection and frequency content are therefore influenced by the status of the entire cochlea.	• Generally assessed as a function of primary tone frequencies. Acoustic distortion products (ADPs) are measured concurrently with the presentation of two pure-tone stimuli (for example, primaries) in one ear. When two primaries are presented to a healthy cochlea, it will result in the production of intermodulation distortion products that are mathematically correlated to the primaries' frequencies.
• Measured using a click (broader band) stimulus	• Measured using narrowband filtering focused on the frequency of interest
• Generally present in cases where behavioural hearing thresholds are better than 30 dB HL without functional or neural components	• Results reflected on a DP-gram that resembles an audiogram, but should not be interpreted as such
• Generally present if cochlear sensitivity in the region of 2 kHz–4 kHz is approximately 20–40 dB HL	• Can be measured in ears with greater levels of HL.

TEOAEs	DPOAEs
• May be reflected on a graph or audiogram, although this is mainly system specific. TEOAE testing generates a waveform that represents emissions produced by outer hair cells at a specific frequency in response to a stimulus. A clearly defined waveform suggests normal functioning of the hair cells, while a weak or absent waveform suggests a possible cochlear dysfunction.	

Adapted from Campbell and Mullin (2023), Prieve and Lamson (2014), Prieve and Fitzgerald (2009) and Tzanakakis et al. (2016).

There are two categories of OAE equipment on the market today: systems predominantly designed for screening OAEs, and systems designed for diagnostic OAE measurement. Screeners are focused on identifying the possibility of HL and do not offer detailed information about the degree and/or nature of an HL. Diagnostic OAEs, however, provide more frequency-specific information for each ear (Prieve and Lamson 2014). A more detailed outline of screening OAEs is presented in Chapter 4.

The interpretation of diagnostic OAEs is beyond the scope of this chapter. Information on the interpretation of OAEs can be found in Otoacoustic Emissions: Principles, Procedures, and Protocols (Dhar and Hall 2017).

Patient state and positioning in OAE testing

Diagnostic OAEs are very sensitive to noise generated internally from the patient. A quiet and calm patient will make measuring OAEs simpler, faster and more effective. With experience, reading a patient's body language and mood will become easier, which will make the planning and the timing of the test battery flow' more successfully. If a patient seems drowsy

during the case history interview with the parents, you may forgo the case history conversation and proceed straight to the OAE measurement. The case history can be returned to at a later stage.

With newborns and young infants, feeding may be useful as a calming strategy to be able to successfully insert the probe tip and initiate measurement. Bear in mind that testing may need to be paused intermittently when sucking, burping, crying or cooing occurs. Keeping the patient calm is the main priority, not only clinically, but also relationally. The HHP should rely on the patient's parent/caregiver to suggest the most comfortable position for the procedure, for example, in their arms, on their lap, in their car seat or in a stroller. It is important to remember that establishing a trusting relationship with the patient and parents/caregivers is just as important as obtaining results. If a patient is physically too restless or upset to obtain reliable results, then a follow-up appointment may offer a second opportunity to try again. OAE measurement is remarkably adaptable when the HHP gains confidence in its application and optimal patient positioning is possible when HHPs get a little creative.

When performing OAEs in toddlers or older children, their ability and willingness to co-operate might improve (although not always in the case of patients with special needs and physical disabilities). Older children should be given the autonomy to choose a comfortable position for the test procedure, for example, sitting independently in a chair or holding onto a parent or caregiver's hand. Offering a form of distraction may also be helpful to keep them calm and quiet for the duration of the test (for example, a muted iPad or tablet, story books, picture books). Whereas performing OAEs earlier in a test battery may be helpful with newborns or young infants to capitalise on their restful state, fatigue may conversely be helpful with older children to keep them quiet and calm for the objective measures. Fatigue caused by behavioural testing (visual reinforcement audiometry and play audiometry) may even result in a quieter and calmer patient. Being flexible in your approach is critical. Figure 6.1 shows examples of patient positions for optimised OAE measurement.

Figure 6.1: (a)



Figure 6.1: (b)



Figure 6.1: (a) Positioning and distraction of a small baby using a small soft toy
Figure 6.1: (b) Independent positioning of an older toddler, with an 'unconventional' distraction tool (calculator)

Measurement and factors influencing OAE interpretation

The first step is to perform an otoscopic examination and to confirm a clear and intact outer ear, ear canal and tympanic membrane (see Chapter 5). This is generally followed by tympanometry to evaluate the middle ear's integrity. Patient factors like occluding earwax, middle ear effusion and the presence of grommets or ventilation tubes might influence the ability to measure emissions reliably, and therefore any external factors that may influence the test must be identified before starting the measurement. The stimulus that evokes OAEs must travel through the middle ear to the cochlea and back. Therefore, any factors influencing the outer or middle ear will impact an OAE measurement (Prieve and Lamson 2014). According to Sanches et al. (2024), the combination of diagnostic OAE measures and tympanometry is valuable for identifying and confirming changes in middle ear integrity as the measured OAE amplitudes are directly related to the integrity of the functioning middle ear and tympanic membrane. It should further be noted that successfully conducting an OAE measurement hinges on a secure probe fit. An unstable probe fit can result in poor repeatability, high noise levels and unreliable results.

Generally, sensitivity for OAEs is lower in the lower frequency and higher frequency ranges and internal and external noise may impact the validity of the measurement (Prieve and Lamson 2014). Therefore, limiting the test frequency range to a spectrum of 2 kHz – 4 kHz may be helpful if a patient is restless and co-operation is limited. Remember that some information is preferable to no information. HHLs are encouraged to perform at least two test runs to confirm repeatability and reliability. Recordings should present with a sufficiently low noise floor across the test frequency range to be accepted as valid and reliable.

The need for age-appropriate normative references has come under scrutiny as most diagnostic OAE systems in the market apply adult-normative ranges to their results by default. According to Blankenship et al. (2018), DPOAEs present with significant age-related differences, and these differences arise mainly from developmental changes that occur in the outer ear and middle ear at specific ages. Using adult normative references in paediatric DPOAE analysis is a risk for misdiagnosis. DPOAE amplitudes in children are generally larger and more robust compared to adult patients. HHPs can easily consider the DPOAE measurements of a paediatric patient that fall within an adult range as present, when they are in fact reduced or absent if referenced against paediatric norms. These normative ranges are generally pre-set on the equipment software and selecting the appropriate normative range for different age groups or even entering the correct date of birth is critical for accurate interpretation of test results.

Auditory Brainstem Response (ABR)

ABR was first introduced to the field of clinical audiology in the 1970s. This discovery has broken boundaries for the diagnosis and early intervention of the youngest infants and most difficult-to-test patients, which would have been impossible otherwise. Also known as Auditory Evoked Potentials (AEP), this measure provides a physiologic method of evaluating the integrity of the eighth cranial nerve (CN VIII) and estimating hearing thresholds electrophysiologically. The success of performing and interpreting ABRs relies heavily on the skill and knowledge of the clinician, particularly regarding the anatomical generators of each of the ABR's components (Hood 1998).

A normal ABR is generated by several anatomical and neural sites (Hood 1998):
Wave I – Distal portion of CN VIII
Wave II – Proximal portion of CN VIII
Wave III – Neurons in the cochlear nucleus
Wave IV – Third-order neurons of the superior olivary complex
Wave V – Lateral lemniscus and inferior colliculus

In this section, both neurological and threshold estimation techniques will be discussed. The interpretation of the ABR is beyond the scope of this chapter. However, information on the interpretation of ABRs can be found in the eHandbook of Auditory Evoked Responses: Principles, Procedures & Protocols by James W. Hall III (2015). Normative references for ABR testing are developmentally specific and HHPs are encouraged to review the normative data of several respected authors in the field of electrophysiology when analysing test results.

Patient state in ABRs

In an ideal world, all young patients are perfectly behaved, calm and quiet for as long as required to complete their electrophysiological tests. However, it is simply not reasonable to expect young patients to sit or lie perfectly still for ten minutes, let alone for the hour needed to conduct extensive ABR testing. Bioelectrical (physiological and myogenic) artifacts are among the most common artifacts HHPs encounter when performing auditory evoked potentials like ABRs (Stegeman and De Weerd 1990). To eliminate the probability of bioelectrical artifacts, the HHP may require the patient to sleep naturally or be sedated when needed.

Natural sleep ABRs

To facilitate an effective natural sleep ABR, it is recommended that the appointment be scheduled around the young patient's best nap time. Sleep deprivation may also help to fatigue the patient enough to fall asleep quickly and remain asleep for an extended period. The HHP should instruct the parent/caregiver to keep their childe up late the night before, wake them up early and keep them awake until they arrive at the test setting. It is also encouraged to perform the test preparations before the patient is allowed to fall asleep. Attempting to prepare the skin and placing electrodes once the patient is already asleep can wake the young patient up.

It is important to plan enough time for preparing the patient, performing the test (which may require the HHP to stop and start several times as the patient progresses through various sleep stages) and providing results, feedback and/or counselling. Natural sleep ABRs may sometimes require several attempts at various appointments to obtain a complete (reliable) test battery, depending on the patient's sleep quality and co-operation on any given day. Children over the age of one and a half years rarely sleep long and peacefully enough to obtain a complete (and reliable) ABR measure in one session, although there are always exceptions to the rule.

Sedated ABRs

If a patient is unable to sleep naturally or co-operate for the duration of an ABR evaluation, then sedation may be required. At this point, it is vital to stress that patient safety must always remain the highest priority, as there will always be risks associated with sedation. The HHP must always weigh the balance between potential sedation risks and the audiological information that needs to be acquired. If the risks outweigh the potential gains, then sedation should under no circumstances be considered an option, and an alternative solution must be explored. Furthermore, an HHP must never attempt a sedated ABR without the assistance and supervision of an anaesthetist, regardless of whether the test is conducted in a controlled setting like a theatre, hospital ward or practice room.

The type of sedation used for ABRs and other auditory evoked potentials may also impact the test reliability and validity. Conscious sedation is an excellent option for this procedure, as it is a lighter type of sedation, where the patient maintains an open airway independently and can be aroused by physical or verbal stimulation (Hall 2007). Intravenous Propofol (also known as Diprivan) is a sedative drug generally used during conscious sedation procedures that typically produces a very modest delay in ABR inter-wave latencies (Hall 2007). This type of sedation has proven to be effective for patients older than three years of age. Hall (2007) also regards *Chloralhydrate* as a suitable conscious sedation option. Clinically, this option is more suitable for younger children (between the ages of one and two years) with a relatively stable daytime sleeping routine. *Chloralhydrate* is rarely effective when administered to children with body weight exceeding fifteen kilograms or when administered outside of their best nap time. Sleep deprivation increases the effectiveness of *Chloralhydrate*.

Certain sedative drugs are, however, contraindicated for auditory evoked potentials as they affect the latency, amplitude or in some cases even prevent potentials from generating altogether (Hall 2007). It is highly recommended that an experienced anaesthetist be consulted about the most suitable sedative drugs for electrophysiological evaluations. Lastly, test duration should be taken into consideartion with sedation. All sedation carries a level of risk to patients, and the longer they remain under sedation the more risk they are exposed to. A skilled and prepared clinician should be able to complete a comprehensive diagnostic ABR evaluation including neurological (click) ABR, threshold estimation testing (air-conduction and bone-conduction) and/or auditory steady-state response (ASSR) testing in under an hour.

Anatomical sites of electrode placement

Electrode placement and test parameter setup must be optimised to facilitate smooth transitioning between different test sets without unnecessarily disturbing the patient. The position of the electrodes is allocated via the international 10-20 system (Figure 6.2) which is typically used in auditory evoked potential measurement and clinical neurophysiology (Hall 2007). Four electrode sites are utilised for the above-mentioned test battery: Inverting electrodes (placed on the right and left earlobes), non-inverting electrodes (placed on Cz, which is located on the vertex or top middle of the head) and ground electrodes (placed on Fz or high on the forehead). The use of a jumper cable or 'connector cable allows the tester to seamlessly alternate measurement between the two ears when obtaining responses and enables the tester to record two-channel recordings.

Figure 6.2: (a)

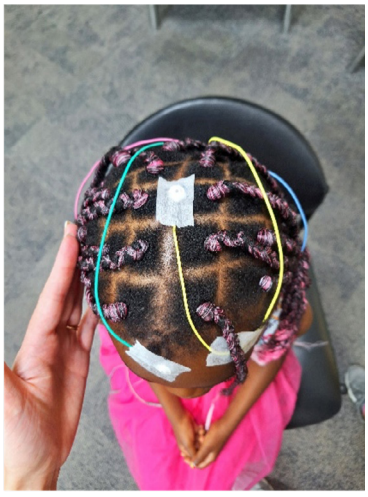

Figure 6.2: (b)



Figure 6.2: (c)



Figure 6.2: (a) Electrode placement and patient positioning non-inverting (yellow) and ground electrode (green) placement
Figure 6.2: (b) Left inverting electrode (blue) on the left earlobe, non-inverting electrode (yellow) and ground electrode (green), which is stabilised on the forehead
Figure 6.2: (c) Patient positioning in relation to headbox/pre-amplifier and insert earphones

The earlobes are considered the superior option for application of the inverting electrodes as they provide the tester with several distinct advantages, as described by Hall and Swanepoel (2010):

- Reduced post auricular muscle (PAM) artifact – if the electrode is placed directly over the postauricular muscle, it is particularly vulnerable to recording bothersome muscle artifact.
- Increased amplitude of wave I – an increase in amplitude of approximately 30% has been noted with electrode placement on the earlobe compared to the mastoid.
- Reduced stimulus artifact – with the placement of the inverting electrode on the earlobe, the bone conduction oscillator can be placed on the mastoid as normal without running the risk of significant electromagnetic stimulus artifact arising from placing the oscillator directly onto an electrode.

Skin preparation, and electrode and transducer placement

The skin must be carefully prepared for electrode placement. The anatomical sites are cleaned using a specialised granular gel or abrasive tape to remove any excess oils and dead skin cells from the surface where the electrode is placed. Care should be taken not to clean the anatomical sites too abrasively, resulting in serous drainage and/or bleeding, which will impact the impedance measurement. In addition, any excess granules should be removed from the anatomical sites before securing the electrodes, as this may also impact the impedance measurement.

When scooping conduction paste from its container, it is recommended that a small amount of the paste is transferred onto a prepared piece of gauze, using a plastic or wooden tongue depressor. Conduction paste should never be scooped directly from the container using an electrode, as conduction paste is known to be an excellent growth medium for microorganisms. Care should also be taken not to use too much electrode paste as protrusion of the paste increases the recording surface area and impacts the quality of the recording. The electrodes should then be secured to their designated anatomical sites using a small piece of micropore tape. A firm though gentle hand helps to secure the electrode for consistent and repeatable recordings and to prevent intermittent artifacts arising from an unstable electrode during testing. For optimal testing, the impedance should measure between 2 – 5 kΩ across the electrode range.

Earphones should be used where a patient's ear canal is available. If the patient ear canal is not available (as seen with atresia, microtia and/or anotia), then headphones might be considered. However, it should be noted that headphones can cause significant stimulus artefacts at high test intensities.

Test parameters

The test parameters for various auditory evoked potentials are system-specific and test-specific. Decisions regarding the selection of the test parameters should be based on the recommendations of respected authors in the field of auditory evoked potentials, manufacturer recommendations, HPP preferences and the test environment. It is important to highlight that any changes in the test parameters will have an impact on the tracings, analysis and interpretation of the test results. Normative references that have been determined for a specific test system and environment, for specific auditory evoked potential evaluations, are no longer valid when any of the test setting parameters are altered and indeed may have a detrimental impact on the analysis of the test results.

When approaching an ABR evaluation, the tester should aim to answer two major questions:

1) Is CN VIII conducting sounds normally up to the midbrain level? (Neurological evaluation)
2) What are the estimated (electrophysiological) hearing thresholds? (Threshold estimation)

Neurological ABR evaluation

Once the patient is sleeping peacefully, either naturally or under sedation, and is appropriately prepared and positioned for the test, the HHP is advised to start with the neurological (click) ABR evaluation. This provides the opportunity to predict what is likely to be encountered later in the evaluation, for example, delayed absolute latencies are suggestive of possible conductive dysfunction, and the presence of cochlear microphonics is suggestive of possible Auditory Neuropathy Spectrum Disorder (ANSD) or retrocochlear dysfunction, etc. In this way, the next steps can be planned more efficiently, to obtain as much information in as little test time as possible.

The goal of the Neurological ABR should be to obtain waves I, III, and V. If the waveforms are unclear or poorly repeatable at first, an increase in the intensity may result in evoking

these waveforms or responses. The use of a two-channel recording will also make analysis and interpretation simpler and more accurate by helping the HHP to confirm the presence of wave III and V. Changing the polarities (rarefaction, condensating or alternating) is also a useful and necessary tool to confirm the presence and legitimacy of a specific waveform. Simply repeating a single polarity is not enough to confirm the legitimacy of a waveform and some artifacts may repeat themselves quite convincingly if the HHP is not careful in their analysis.

An important rule when analysing Neurological ABRs is that with an increase in intensity, the amplitude of the waveforms should increase and the latency of the waveforms should decrease. Conversely, with the decrease in intensity, the amplitude of the waveforms should decrease and the latency of the waveforms should increase.

When analysing Neurological ABRs, it is critical to be aware of the effects that nerve maturity may have on the appearance of the waveforms. The major expected neuronal maturation occurs within the first two years of life, and interpeak latencies (IPL) decrease significantly during this time (Matin et al. 2021). Therefore, reviewing reputable age-appropriate normative data when analysing interpeak and absolute latencies is essential for an accurate diagnosis.

Threshold estimation ABR

Threshold estimation may follow the neurological evaluation. Tone burst ABR should be the preferred test method to physiologically estimate hearing thresholds, as it provides frequency-specific information and has proven reliable. Where possible, it is preferable to obtain both air- and bone-conduction estimated thresholds, since this provides information about the presence or absence of a conductive component and therefore also insight on the nature of the HL. As conductive pathologies are more prevalent in the paediatric population, a potential middle and/or outer ear influence will have to be considered with any intervention for a young patient.

With threshold estimation, the goal is to obtain a repeatable wave V up to the lowest intensity level upon which the wave V tracing disappears. Threshold estimation techniques using tone burst, CE-Chirp or even audiological click evaluations will require the HHP to use unique correction factors that are specific to their test equipment and test environment. To procure

environment and system-specific correction factors, the HHP must acquire normative data for their clinical equipment and setting. This is achieved in the following ways:

- When acquiring a new test system, spend sufficient time customising the test settings and parameters (based on the recommendations of respected authors in the field of ABRs and manufacturer recommendations) for the specific testing needs.
- Perform biological hearing threshold estimations on at least twenty normal hearing individuals, for example, determine the lowest audible hearing level for each test frequency for twenty normal hearing individuals, specifically for each ear, using insert earphones.
- Calculate the average thresholds, at each frequency. The average will be accepted as the correction factor for each frequency. Never use the correction factors of another HHP's system, as this will result in inaccurate interpretation and an unreliable diagnosis.

Threshold estimation hinges on the same principles as the neurological evaluation when considering the latency and amplitude of wave V. The absolute latency of wave V should increase or, at the very least, remain constant with the incremental decreases in intensity. The latency of wave V will never be earlier at a lower intensity compared to a louder intensity. These simple rules will guide the HHP in avoiding mistakes when analysing their waveforms.

Auditory Steady State Response (ASSR)

The ASSR was first introduced in 2001. Before that, the ABR was the only option for objective estimation of hearing thresholds. According to Hall and Swanepoel (2010), it is important for the HHP to understand that the ASSR does not refer to a single response but rather a series of responses that are generated at different locations along the auditory pathway. These responses are directly related to the stimulus characteristics presented to the ear (Hall and Swanepoel 2010).

One of the advantages of the ASSR is its maximum output levels. The ASSR can often produce stimulus levels at much louder intensities compared to the tone burst or click stimulus. This gives the HHP the advantage of being able to test more severe degrees of HL than would

be possible through other ABR test techniques. Although ASSR is a convenient and relatively simple test technique to administer, it is important to be aware of certain limitations:

- ASSR should not be used as a standalone test and must be considered part of a comprehensive test battery.
- The ASSR only provides an estimated behavioural hearing threshold — it cannot distinguish between sensorineural, conductive, or mixed HL.
- The worse the degree of HL, the more accurate the estimated thresholds tend to be with ASSR.
- Minimal, sharply sloping and conductive types of HL tend to deliver more unreliable estimated hearing thresholds with ASSR.
- High levels of internal and external noise during ASSR testing may negatively impact the estimated hearing thresholds.
- Estimated hearing thresholds obtained in cases where ANSD and/or retrocochlear pathology is suspected must be interpreted with great care as the auditory pathway is compromised. The thresholds may be better or worse than indicated.

The Electrophysiological Toolbox – Where OAEs and ABRs Come Together

If we simply consider the intricacies of normal hearing, then investigating abnormal hearing undoubtedly requires a comprehensive diagnostic toolbox in the hands of a skilled and experienced HHP. ANSD and other retrocochlear hearing conditions often present with strange clinical pictures that require an in-depth understanding of each portion of the auditory pathway. ANSD was first described by A. Starr and his colleagues in 1996 (De Siati et al. 2020). ANSD is characterised by the absence of repeatable ABR waveforms and/or altered waveforms and absent or elevated acoustic reflexes, in the presence of Cochlear Microphonics (CM) and/or OAEs (De Siati et al. 2020). Considering that ANSD accounts for up to 30% of all newborns discharged from neonatal ICU (NICU) who present with sensorineural hearing loss (SNHL) (De Siati, et al. 2020), the need for advanced and reliable electrophysiological testing in the paediatric audiology clinic becomes vital. As the outcomes for intervention in this special test population hinge directly on the correct diagnosis of the HL, the margin for error becomes ever smaller.

Correct diagnosis, and appropriate audiological intervention and counselling will have a powerful impact on the young patient's language acquisition, scholastic performance, and the family's expectations and acceptance of the HL. Neural maturation is also frequently overlooked when it comes to ABR analysis. With improvements in technology, our ability to diagnose younger children earlier has improved dramatically. Nevertheless, the push towards earlier detection may in some cases be harmful, particularly in high-risk paediatric patients (as defined by the American Joint Committee on Infant Hearing), 2019). According to Psarommatis et al. (2017), a significant number of high-risk infants with absent ABR waveforms showed complete or partial recovery at re-examination. They also found that the younger the infant was at the initial ABR evaluation, the higher the probability of a false-positive ABR finding (Psarommatis et al. 2017). The presence of OAEs was also found to be a strong predicting factor for the recovery of ABR waveforms despite the presence of multiple other risk factors (Psarommatis et al. 2017). Therefore, early cochlear implantation (generally before the age of one year) should be considered with great caution, especially for children fitting the ANSD profile. Cochlear implantation should only be considered after obtaining stable and reliable electrophysiological and behavioural test results (Psarommatis et al. 2017).

Conclusion

Electrophysiological testing requires regular practice and constant learning. These physiologic evaluation tools are routinely utilised in the most vulnerable age groups where a potential error can have a huge impact on the trajectory of a child's life. A hard but necessary truth to acknowledge in your role as an HHP is that there is no room for error when it comes to children. Efficiency helps, but accuracy is everything.

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Chapter 7

Paediatric Vestibular Assessment

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'Children's developing systems require not only smaller tools, but greater insight.'

~Piaget

Introduction

The vestibular system plays a critical role in maintaining balance, spatial orientation and gaze stability. In children, vestibular disorders can negatively affect both intellectual and physical development, with consequences for motor skills, learning and academic performance (Rogers 2010; Gioacchini et al. 2014). One key mechanism of the vestibular system is the vestibulo-ocular reflex (VOR), which stabilises vision during head movements. Impaired VOR function in children can make it difficult to keep text in focus during reading tasks, particularly in dynamic classroom environments that require frequent head movements, such as copying from the board or shifting attention between peers and materials. This visual instability may lead to impaired reading fluency, diminished comprehension and increased visual fatigue. Vestibular dysfunction can also disrupt spatial and non-spatial cognitive processes, delay gross and fine motor development and contribute to broader learning difficulties (De Kegel et al. 2012; Wiener-Vacher et al. 2013). Likewise, Franco and Panhoca (2008) found a statistically significant association between peripheral vestibular dysfunction and underperformance in school. A systematic review by Gioacchini et al. (2014) reported a prevalence of up to 15% for vestibular disorders in the paediatric population.

Vestibular-mediated reflexes are present at birth (for example, head righting response: an automatic body reaction that keeps the head straight, level, and upright relative to the

ground)(Adamović et al. 2020). Children with impairments that alter the vestibular reflexes are slower than their normal counterparts in reaching motor key milestones (Inoue et al. 2013). In the paediatric population, disruptions in vestibular function can significantly impact motor development, coordination and overall quality of life. Despite its consequences, vestibular dysfunction in the paediatric population often goes underdiagnosed, partly due to the challenges of identifying and assessing vestibular impairments in this age group. Children might present with subtle or non-specific symptoms, such as delayed motor milestones, poor balance or frequent falls, which can easily be attributed to other causes.

Advances in diagnostic techniques have facilitated the evaluation of vestibular function in the paediatric population, enabling early detection and management of vestibular disorders. Paediatric vestibular assessments now encompass a range of assessment methods, including caloric testing, rotational chair testing, vestibular-evoked myogenic potentials (VEMPs) and video head impulse testing (vHIT). These tools, when combined with a thorough case history and clinical observation, provide a comprehensive approach to identifying vestibular dysfunction in children. However, the outcomes of these assessments are significantly influenced by the child's developmental stage, willingness to participate, capacity to co-operate and comprehension of the test instructions. As the vestibular test battery comprises multiple procedures, it is crucial to identify, prioritise and begin with the most suitable tests for each specific patient. This approach aims to maximise the diagnostic information obtained within a limited timeframe, acknowledging the potential for diminished concentration in younger patients. The overall aim of the assessment is to determine if the child presents with a possible peripheral or central vestibular disorder, guiding further intervention and referrals to other healthcare professionals.

Case History

The case history is essential in the paediatric vestibular assessment. It must be conducted at the outset as it provides critical context to guide test selection, interpretation, and clinical management. Children may not be able to describe their symptoms accurately, making a thorough history crucial for identifying potential vestibular dysfunction. Parents and caregivers are essential sources of information when obtaining a comprehensive case history, particularly for non-verbal children. The hearing healthcare professional (HHP) relies on

caregivers to report the child's developmental milestones, general behaviour and observations of balance and gait, as well as any avoidance behaviours, such as fear of heights or reluctance to walk in the dark or on uneven surfaces. To facilitate the case history, HHPs may provide a questionnaire, such as the Vanderbilt Paediatric Dizziness Handicap Inventory (VPDHI) (McCaslin et al., 2015) for caregivers to complete at home with the child or by observing the child before the day of testing. For verbal children, this questionnaire approach also allows the child to share details more comfortably and openly in a familiar environment. In addition, the caregiver can ask these questions in the child's first language. In this way, the HHP can clarify and confirm the information with the child on the day of the assessment. It is important to use age-appropriate language when communicating with the paediatric population and to avoid jargon (Chapter 15). Collecting detailed background information provides valuable context for interpreting vestibular assessment results and tailoring appropriate management strategies.

The case history should focus on family history, medical history, the impact of symptoms on the child's daily life, behavioural observations of the child, as well as the following aspects.

Prenatal and birth history

Understanding a child's prenatal and birth history is essential in identifying potential factors that may contribute to vestibular dysfunction. Maternal infections during pregnancy, such as cytomegalovirus (CMV) or rubella, can impact the developing auditory and vestibular system. Similarly, the use of certain medications during pregnancy may pose ototoxic or neurodevelopmental risks. Birth complications, including prematurity, hypoxia or low birth weight, can also affect the integrity of the inner ear and central nervous system (Chapters 2 and 3).

Developmental milestones

A child's developmental milestones provide insight into their vestibular function and overall motor development. The timing of major milestones, such as sitting, crawling, standing and walking, can indicate whether vestibular or motor delays are present. Delays in achieving these milestones may suggest vestibular hypofunction or other underlying issues affecting balance. Additionally, difficulties with gross motor skills, such as frequent falls or an inability to navigate age-appropriate physical tasks, can further highlight potential vestibular dysfunction. It has been reported that gross motor milestones predict vestibular loss in children with hearing loss (HL) (Janky and Shepard 2019).

Hearing and vision history

A history of HL, tinnitus or recurrent ear infections can signal potential inner ear dysfunction, which may impact both auditory and vestibular pathways. Similarly, pathologies of the visual system, such as strabismus (misalignment of the eyes) or difficulty focusing, can exacerbate balance problems or contribute to sensory integration challenges.

Symptom inquiry

A structured approach, such as the **SOSTONED** acronym—**S**ymptoms, **O**nset, **S**ituation, **T**riggers, **O**thers affected, **N**eurological symptoms, **E**volution and **D**uration—can ensure that a thorough and systematic history is obtained. The HHP should elicit a detailed description of the child's symptoms in their own words, including:

- Type of sensation: for example, room spinning, feelings of falling, nausea, vomiting or imbalance in specific situations.
- Onset and evolution: sudden or gradual onset, changes over time, duration and frequency of symptoms.
- Situational context and triggers: for example, during movement, positional changes, exposure to loud sounds, or visual stimuli.
- Environmental factors: whether others in the environment experienced similar symptoms (to rule out environmental causes).
- Neurological symptoms: for example, blurred or double vision, numbness or photophobia.

Table 7.1: outlines the symptoms that can be expected with a peripheral vestibular pathology versus central pathology.

Table 7.1: Symptoms of a peripheral vestibular pathology versus central pathology

Peripheral Vestibular Pathology	Central Pathology
Sudden onset of symptoms	Gradual onset of symptoms
Intermittent attacks with severe symptoms	Symptoms are constant but mild.
Symptoms are worsened by changes in head position and movement.	Symptoms are unaffected by head position and movement.
Nausea and vomiting are severe and frequent.	Nausea and vomiting are infrequent and mild.
Motor function, gait and coordination are typically intact.	Motor function, gait instability and loss of coordination are more likely to occur.

Vestibular Test Battery

Once a comprehensive case history has been conducted, the HHP should determine which test procedures to prioritise. The vestibular test battery comprises a series of diagnostic tools designed to evaluate the functional integrity of the peripheral vestibular system, as no single test can evaluate all aspects of vestibular function. Each test targets specific components, including the semicircular canals, otolith organs and their associated neural pathways. In the paediatric population, selecting appropriate tests requires careful consideration of developmental stages, co operation levels and the presenting symptoms. By combining physiological and behavioural measures, HHPs can obtain a detailed understanding of the child's vestibular function.

Behavioural test battery

Behavioural vestibular assessments play a critical role in evaluating balance and spatial orientation in paediatric populations, especially when physiologic testing may not be feasible, readily available or tolerated. These tests rely on observing the patient's responses to structured tasks that challenge the vestibular system in age-appropriate and developmentally appropriate ways. The behavioural test battery offers valuable insights into vestibular function by assessing postural control, gaze stability and compensatory strategies through non-invasive and often play-based methods. In children, especially those under the age of five years or with neurodevelopmental differences, behavioural assessments serve as a practical and informative alternative or complement to physiologic vestibular tests. Table 7.2 outlines the components, administration and interpretation of the behavioural vestibular test battery, emphasising its clinical relevance and adaptability across the paediatric age range.

These behavioural vestibular assessments provide a crucial and adaptable approach to evaluating vestibular function in children, particularly when traditional physiologic tests are not practical or tolerated. By using these observation-based, developmentally appropriate methods, HHPs can gather meaningful information about a child's balance, gaze stability and spatial orientation. These assessments are especially valuable in younger children (under three years) and those with neurodevelopmental differences, offering a flexible, non-invasive means of identifying vestibular impairments. When integrated with caregiver input and physiologic clinical findings, behavioural vestibular assessments enhance diagnostic accuracy and support early intervention planning, reinforcing their indispensable role in paediatric vestibular evaluation.

Table 7.2: The components, description and interpretation of the behavioural vestibular test battery

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
Romberg Test	3+ years	Assesses postural stability with eyes open and closed to identify proprioceptive or vestibular deficits.	"Stand like a statue with your feet together. First keep your eyes open, then close them and stay very still."	Normal: Stable with minor sway. Abnormal: Excessive sway or falling with the eyes closed may indicate vestibular or proprioceptive dysfunction.
Modified Clinical Test of Sensory Interaction on Balance (mCTSIB)	4+ years	Evaluates balance under different sensory conditions, including firm and foam surfaces with the eyes open and closed.	"Let's play a balance game! You'll stand still on different surfaces with your eyes open and closed."	Poor performance on foam with the eyes closed may indicate vestibular dysfunction.

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
Head Impulse Test (Bedside vHIT)	5+ years	Assesses semicircular canal function through high-acceleration head impulses while monitoring eye movement.	“Keep your eyes on my nose. I’m going to give your head a quick turn!”	Normal: Eyes remain on the target. Abnormal: Eyes move off the target with a catch-up saccade, indicating peripheral vestibular loss.
Head Shake Test	5+ years	The head is shaken from side to side. The presence of nystagmus after movement may indicate unilateral dysfunction.	“I’m going to gently shake your head from side to side. Just relax and keep looking straight ahead.”	Post-head-shake nystagmus suggests unilateral vestibular hypofunction.

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
Dynamic Visual Acuity (DVA)	6+ years	Compares visual acuity during head movement with static visual acuity to assess vestibulo-ocular reflex (VOR) function.	"Tell me what letters you see on the wall in front of you. Now I'll gently move your head while you keep reading."	A drop of more than three lines in visual acuity during head movement may suggest bilateral vestibular loss.
Oculocephalic Reflex (Doll's Eye Reflex)	0–2 years	Assesses brainstem function by observing compensatory eye movement during passive head turning.	Gently rotate the infant's head from side to side while observing the eyes. No instruction is needed.	Normal: Eyes move in the direction opposite to the head. Absent response: Possible brainstem dysfunction or vestibular loss.

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
Vestibular-Triggered Postural Reactions	0–2 years	Observes vestibular responses, such as head righting, during changes in position.	Observe spontaneously or during postural transitions. No instruction is needed.	Delayed or absent head righting may suggest vestibular impairment in infants.
Gait Observation and Tandem Walking	4+ years	Assesses balance and coordination by observing the walking pattern along a straight line.	“Can you walk to me on this line like you’re on a tightrope?”	Normal: Smooth, stable gait. Abnormal: Ataxia, veering or a wide-based gait may indicate vestibular or cerebellar pathology.

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
Spontaneous and Gaze-Evoked Nystagmus (can also be performed using VNG goggles)	All ages with co-operation	Examines involuntary eye movements that may indicate vestibular or central dysfunction.	Ask the child to look straight ahead and then to the left and right. “Watch this toy or finger move.”	Present at rest: Possible acute vestibular neuritis or central pathology. Direction-changing nystagmus may indicate a central lesion.
Dix-Hallpike Test (can also be performed using VNG goggles)	5+ years	A positional test used to diagnose posterior canal benign paroxysmal positional vertigo (BPPV) by observing nystagmus and vertigo in a head-hanging position.	“I’m going to help you lie back quickly with your head turned to the side. Try to stay still and tell me how you feel.”	Torsional, upbeating nystagmus indicates posterior canal BPPV on the side of the downward ear.

Test	Age Range	Test Description	Instructions (Age-Appropriate Language)	Interpretation
General Positional Tests, for example the Supine Roll Test (can also be performed using VNG goggles)	5+ years	Assesses horizontal canal BPPV and other forms of positional vertigo by moving the child's head into various supine positions.	"We'll play a game where I move your head while you're lying down. Just relax and look at me."	Horizontal nystagmus suggests horizontal canal BPPV. Its direction and duration help differentiate geotropic and apogeotropic variants.

Physiological test battery

The physiologic vestibular test battery encompasses a range of assessments designed to objectively evaluate the functional integrity of the vestibular system. In paediatric populations, physiologic vestibular testing presents unique challenges due to developmental, cognitive and behavioural factors. However, advancements in technology and age-appropriate protocols have improved test feasibility and reliability across a wide age range. Understanding the indications, proceduresand interpretation of the available physiologic tests is essential for accurate diagnosis and effective management of vestibular disorders in the paediatric population.

a) Vestibular evoked myogenic potential (VEMP) testing

The vestibular evoked myogenic potential (VEMP) is a clinical test for assessing vestibular pathologies by evaluating otolith function (Özgür et al. 2015; Walther and Cebulla 2016). It measures an electromyographic (EMG) response elicited by sound, vibration or electrical stimulation (Murofushi 2016). Colebatch and Halmagyi (1994) first described the cervical VEMP (cVEMP) as a short-latency response from the sternocleidomastoid muscle (SCMm), mediated by the saccular macula, inferior vestibular nerve and vestibulospinal pathways. Later, Rosengren et al. (2019) and Todd, Rosengren and Colebatch (2007) identified ocular VEMPs (oVEMPs), recorded from extraocular muscles and reflecting utricular function (Rosengren et al. 2010).

The clinical application of VEMPs in the paediatric population has been proven by the test's ability to aid in the diagnosis of vestibular neuritis, congenital vestibular hypofunction, ototoxicity, vestibular migraine and superior semicircular canal dehiscence syndrome (SCDS) (Cal and Bahmad 2009). Children with syndromes associated with HL, for example, Usher syndrome and Waardenburg syndrome, often exhibit vestibular dysfunction that can be detected using VEMPs.

VEMPs are widely considered as a safe, non-invasive tool that is useful to supplement caloric, video head impulse and rotational chair testing, all of which are limited to the evaluation of the horizontal semicircular canal function and superior vestibular nerve integrity. VEMP testing is generally feasible in children aged three and older. At this age, many children are able to engage in brief periods of co-operation and, in the case of cVEMPs, can generate sufficient tonic activation of the sternocleidomastoid muscle (SCMm) through head-lift or resistance tasks. However, test success improves with age and development. For oVEMPs, children must remain still and fixate their gaze upward, which may be challenging for those under

five years. Modifications, such as using play-based approaches or shortened protocols, may enhance test feasibility in younger children. Table 7. 3 depicts the test set-up for cVEMP and oVEMP.

The test procedures for VEMPs are as follows:

- It is essential to conduct otoscopy and tympanometry (Chapter 5) before commencing with cVEMP and oVEMP testing to exclude an outer or middle ear pathology, as this would most likely result in absent responses. In cases of abnormal outer- or middle-ear functioning, bone-conduction VEMP testing can be done.
- The child can be seated independently or placed on the caregiver's lap.
- The skin should be cleaned with nu-prep to reduce impedance and surface electrodes should be placed according to the test type. The electrode impedance should not exceed 5kΩ.

Table 7.3: cVEMP and oVEMP test procedure and interpretation

Aspect	cVEMP	oVEMP
Electrode placement	Electrodes are placed on the belly of the sternocleidomastoid muscle (SCM), with a reference electrode on the sternum and a ground electrode on the forehead.	Electrodes are placed below the eyes over the inferior oblique muscle, with a reference electrode on the chin and a ground electrode on the forehead (Figure 7.1).
Test position	The patient sits upright or lies supine with the head turned away from the stimulated ear to activate the SCM. Encourage consistent muscle contraction through verbal prompts or engaging activities, such as holding a toy.	The patient sits upright and gazes at a point on the ceiling, or lies supine and gazes upward at a fixed point or toy placed at a 30–40° angle to engage the extraocular muscles.

Aspect	cVEMP	oVEMP
Test interpretation	A positive peak, referred to as P1, is recorded at approximately 13.00 ms, and a negative peak, referred to as N1, is recorded at approximately 23.00 ms, as depicted in Figure 6.2A (Chihara et al. 2007; Todd, Rosengren and Colebatch 2007; Welgampola et al. 2009). Larger amplitudes can be expected in children.	A negative peak, referred to as N1, is recorded at approximately 10.00 ms, and a positive peak, referred to as P1, is recorded at approximately 15.00 ms, as depicted in Figure 6.2B (Chihara et al. 2007; Todd, Rosengren and Colebatch 2007; Welgampola et al. 2009). Larger amplitudes can be expected in children.



Figure 7.1: oVEMP patient set-up

Figure 7.2: (a) cVEMP (A) and oVEMP, (b) results for the right and left ear

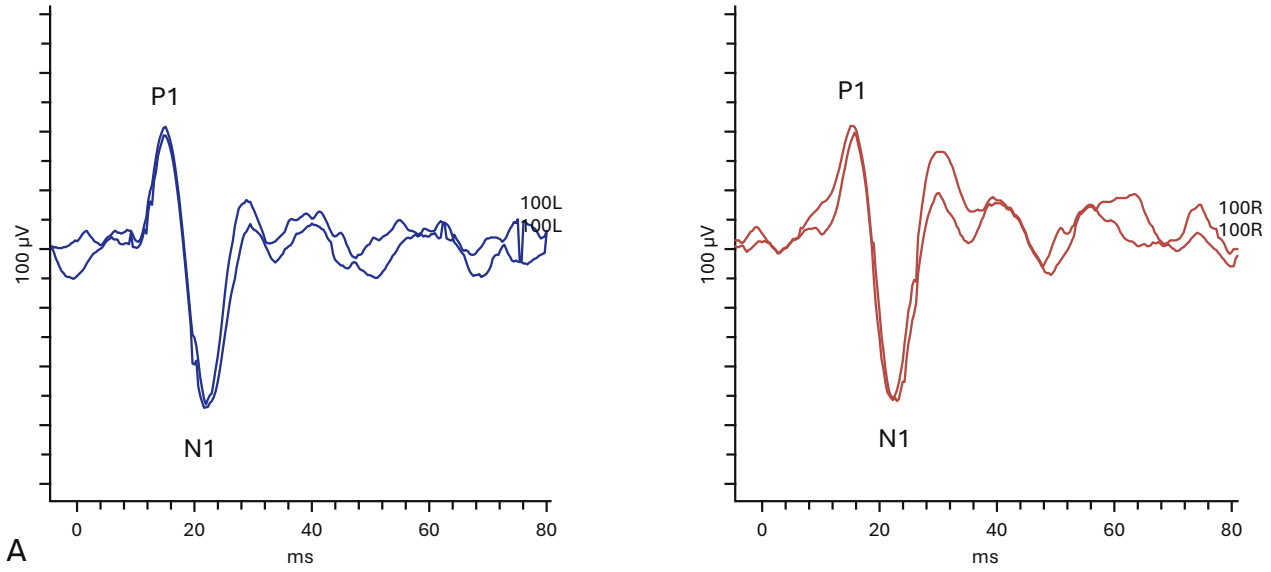

Figure 7.2: (a)

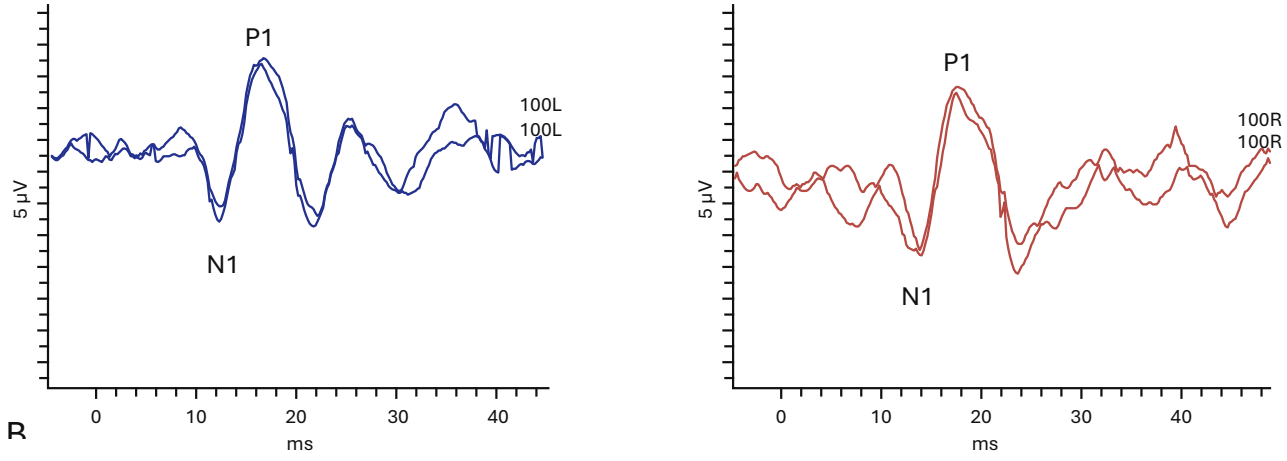

Figure 7.2: (b)

Video head impulse test (vHIT)

The video head impulse test (vHIT) provides an assessment of the function of the vestibular ocular reflex (VOR), which is one of the primary vestibular reflexes. When a child with a normal vestibular system moves their head, the eyes reflexively deviate in the opposite direction so that the image is stabilised on the retina without blurring. This reflexive eye movement is called the VOR and may also be referred to as the angular vestibulo-ocular reflex (aVOR). In recent years, the vHIT has become a valuable tool in assessing all six semi-circular canals individually at higher frequencies (Ross and Helminski 2016). This test is well tolerated by children and can also be done using a remote camera system instead of goggles (Wiener-Vacher and Wiener 2017).

When specifically looking at the paediatric population, Ross and Helminski (2016) observed challenges in the vHIT system. Due to children's smaller physical features, such as smaller head sizes and smaller eyelid openings, measurement errors seem to occur more easily than in adults. These measurement errors are caused by goggle slippage, excessive blinking, etc., which can result in artifacts and inaccurate aVOR gain recordings (Mantokoudis et al. 2014). Another consideration is that the results are dependent on the experience and skills of the examiner to elicit correct and precise impulses. Additionally, a lack of inherent stiffness of the cervical spine is observed in the paediatric population, resulting in difficulty eliciting a head impulse greater than 100°/sec2. Despite these limitations, the vHIT has proven to be a sensitive and efficient vestibular test for the paediatric population and is well tolerated by children aged three to sixteen (Hulse et al. 2015). Success depends on the patient's ability to maintain steady visual fixation and tolerate rapid, passive head movements without excessive blinking or head resistance.

a) Patient and caregiver preparation

Use a child-sized chair or allow the patient to sit on a caregiver's lap if necessary for comfort and compliance. Visual aids, toys or demonstrations may be used to reduce anxiety and gain co operation. Provide the following test instructions:

- To the caregiver: 'We're going to do a simple test to check how your child's balance system is working. It involves wearing lightweight goggles, like a pair of special glasses, which will help us see how their eyes move when we gently turn

their head. The test is quick, doesn't hurt, and will only take a few minutes. You can stay right here to keep them comfortable, and I'll explain everything as we go along.'

- To the patient (for a younger child < six years): 'We're going to play a fun game! You'll wear some cool goggles, like superhero glasses, and all you have to do is keep looking at the dot on the wall (or a fun cartoon). I'm going to gently turn your head—kind of like a little dance move—but don't worry, it won't hurt at all. Just keep your eyes on the dot, and we'll finish super quickly!'
- To an older child (> six years): 'These special goggles help us see how your eyes and balance system work together. All you need to do is sit still, keep looking at the target on the wall, and I'll gently move your head back and forth a little. It won't hurt, and you'll be done before you know it!'

b) Positioning and Testing

- Seat the patient upright in a stable position.
- Align the patient's head in the horizontal plane for testing the horizontal semicircular canals or in the appropriate orientation for vertical canal testing.
- Instruct the patient to fixate on a target (for example, a dot or toy) placed at eye level about one meter away.
- Perform small, quick, unpredictable head movements (10–20 degrees amplitude) in the plane of the semicircular canal being tested, ensuring the movements are within physiological range and not abrupt or excessive.
- Test each ear separately by rotating the head to the left and right for horizontal canals or tilting for vertical canals.

c) Test interpretation and normative values

Normative data for paediatric populations is age-dependent and varies slightly between studies (Table 7.4). Figure 7.3 depicts normal vHIT results and VOR function of the right and left lateral canals, and Figure 7.4 demonstrates abnormal VOR function of the right lateral canal, with reduced gain and covert saccades observed at approximately 100 ms and covert saccades observed at approximately 200 ms.

Table 7.4: Age-based normative vHIT data for paediatric patients

vHIT Parameters	Typical Range/Interpretation
VOR Gain – Horizontal Canals (Infants & Toddlers, up to three years)	Approximately 0.60–0.75
VOR Gain – Horizontal Canals (Preschool-aged, 4–6 years)	Approximately 0.75–0.90
VOR Gain – Horizontal Canals (older children and adolescents, 7+ years)	Approximately 0.90–1.00
VOR Gain – Vertical Canals	Typically lower than horizontal canals; ~0.60–0.85 depending on age and canal
Asymmetry (Inter-ear Gain Difference)	< 7% is considered normal
Latency of Corrective Saccades	Presence of covert or overt saccades suggests semicircular canal dysfunction

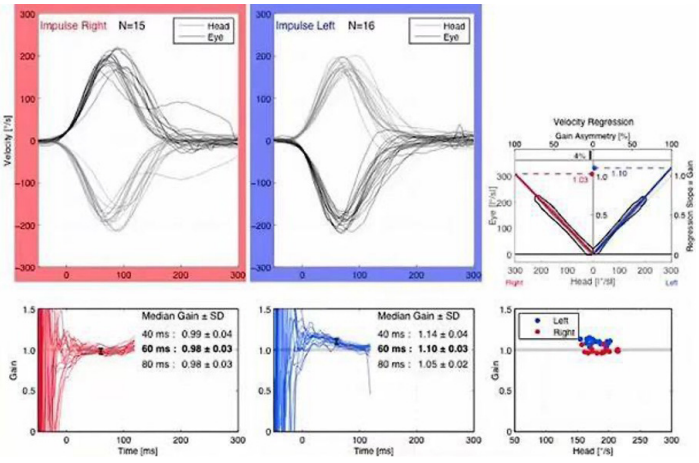

Figure 7.3: vHIT results depicting normal VOR function of the right and left lateral canals (Interacoustics A/S, 2023)

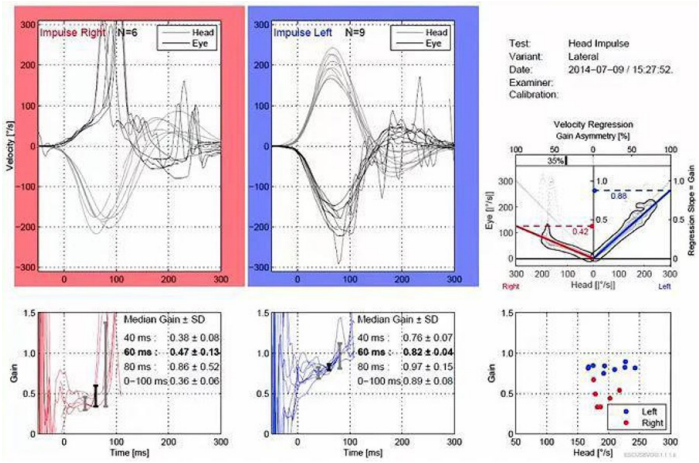

Figure 7.4: vHIT results depicting abnormal VOR function of the right lateral canal (Interacoustics A/S, 2023).

Caloric testing

The caloric test assesses the low-frequency function of the horizontal semicircular canal and the superior vestibular nerve and offers valuable diagnostic information when appropriately tailored to the paediatric population. The caloric test is useful in identifying unilateral or bilateral vestibular hypofunction which may manifest as delayed motor milestones, imbalance or dizziness in children. The test is recorded using Videonystagmography and is conducted by introducing air or water at a specific temperature into the external auditory canal. This process induces endolymphatic flow within the vestibular system, creating a non-physiological stimulus. The resulting temperature gradient across the canal alters the density of the endolymph, leading to measurable vestibular responses. A normal caloric response is characterised by a clear, sustained horizontal nystagmus with an appropriate directional component based on the temperature used, whereas reduced or absent responses may indicate peripheral vestibular hypofunction. Figure 6.5 depicts a right unilateral peripheral vestibular weakness. The procedure and its underlying mechanisms were first described by Robert Bárány in 1907. Bárány proposed that the test be conducted with the patient in a supine position, with the head elevated at a 30-degree angle from the horizontal plane to align the horizontal semicircular canal optimally. As the head is positioned at an angle of 30 degrees with the body in the supine position, the horizontal semicircular canal develops the

largest temperature gradient due to its proximity to the stimulus (Baloh and Kerber 2011). A warm stimulus causes the endolymph to become lighter and move upwards, resulting in an ampullopetal (towards the ampulla) flow. This induces horizontal nystagmus beating toward the stimulated ear. Conversely, a cool stimulus causes the endolymph to become heavier and move downwards, resulting in an ampullofugal (away from the ampulla) flow and horizontal nystagmus away from the stimulated ear (Baloh and Kerber 2011; Barin 2008). This is known as the 'COWS' rule, that is: cool opposite, warm same.

Caloric testing in children presents unique challenges due to developmental, behavioural and anatomical aspects that should be considered when interpreting results. The caloric test may not produce reliable results in very young children (for example, under two years of age) due to underdeveloped vestibular responses or lack of co-operation. Children may have limited understanding or ability to comply with the test instructions. Anxiety and fear of unfamiliar procedures can hinder co-operation. Strategies like explaining the procedure in simple terms or using distraction techniques (for example, toys, videos) can help for older children. However, testing younger children or those with special needs may require mild sedation or alternative vestibular assessments. Therefore, caloric testing is typically reserved for children aged five to six years and older. Water irrigation is often preferred over air in paediatric patients due to its greater reliability and ease of inducing a response. However, the temperature and volume must be carefully adjusted for smaller ear canals. Particularly in the paediatric population, further contra-indications to caloric testing include a significant degree of space-occupying cerumen, otitis externa, middle ear effusion or the presence of a hypermobile tympanic membrane (British Society of Audiology 2010).

a) Patient and caregiver preparation

- It is essential to conduct otoscopy and tympanometry prior to commencing with caloric testing to exclude an outer or middle ear pathology.
- Explain the procedure in simple, child-friendly language.
- Reassure the child and involve the parent or caregiver to reduce anxiety.
- Use toys or videos to explain the process where appropriate.
- The environment may need to be modified (for example, allowing the caregiver to hold the child or shortening the test duration) while focusing on obtaining critical data.

b) Positioning and testing

- When using videonystagmography (VNG), the lights in the room should be dimmed to prevent light from entering the goggles, a common issue caused by the smaller head size of children, which can affect the fit of the goggles.
- The child should be placed in a supine position on an examination bed.
- The head should be elevated by approximately 30 degrees from the horizontal plane to align the horizontal semicircular canal with gravity for optimal stimulation.
- Warm and cool water or air should be used, depending on equipment availability and patient tolerance. For water irrigation, insert water at 30 degrees Celsius (cool) and 44 degrees Celsius (warm) for 30–40 seconds. If water is contraindicated or not well tolerated, use air irrigation by inserting air at similar temperatures for 60 seconds. Proper positioning of the probe is crucial during air irrigation to ensure that the air is directed toward the tympanic membrane. Incorrect positioning, where air is delivered into the ear canal instead, may cause irritation or damage to the skin within the canal.
- Each ear should be stimulated sequentially, alternating between warm and cool stimuli with five-to-seven-minute rest periods between irrigations.

c) Monitoring and observation

- Watch for signs of discomfort, fear or distress.
- Pause or stop the procedure if the child becomes excessively agitated.
- Observe and record nystagmus direction, duration and velocity during and after each irrigation. If testing is incomplete and needs to be stopped due to the child's inability to tolerate the procedure, visually observe the strength and direction of the nystagmus, even if accurate readings cannot be obtained from the equipment. This observation may still provide valuable information on whether vestibular function is present in both systems.

d) Interpretation

- Age-appropriate normative data should be used for interpretation as depicted in Table 7.5.
- Total Response: The total caloric response is calculated based on the sum of the

peak slow phase velocity (SPV) of all four irrigations; left warm (LW), left cold (LC), right warm (RW) and right cold (RC). It is widely accepted that a bilateral weakness is present if the total caloric response is less than 25 degrees per second, or if the total left or right ear response is less than 12 degrees per second.

- Directional Preponderance (DP): DP is a calculated value that compares the nystagmus strengths in the right beating direction with the nystagmus strengths in the left beating direction. The DP is given as a percentage. A value of 30 % or greater is generally used to suggest the presence of a DP.
- Unilateral Weakness (UW): Unilateral Weakness is a calculated value that compares the nystagmus strength induced by irrigation of the left ear with that of the right ear. The UW is given in percentage. A difference greater than or equal to 25 % is indicative of a clinically significant UW in the ear producing the lesser responses.
- Fixation Index (FI): This is a measure of the change in nystagmus intensity when comparing the nystagmus observed during fixation to the intensity of nystagmus immediately prior to fixation. The FI is measured in both directions as suppression of nystagmus may occur in one direction and not the other.

Table 7.5: Age-appropriate normative data for caloric testing in paediatric populations

Parameter	Age Group	Typical Value/ Range	Interpretation/ Notes
Response Magnitude (SPV)	< 3 years	Variable/Often low	Immature responses; difficult to interpret
Response Magnitude (SPV)	3–6 years	≥ 10°/sec	May be lower than adults; variable
Response Magnitude (SPV)	7+ years	20–30°/sec	Approaches adult norms
Unilateral Weakness (UW)	All ages (if tolerable)	< 25%	Suggests unilateral hypofunction if >25%

Parameter	Age Group	Typical Value/ Range	Interpretation/ Notes
Directional Preponderance (DP)	All ages (if tolerable)	< 30%	Increased DP may reflect central asymmetry
Test Tolerance	< 6 years	Often limited	Incomplete testing common; consider using air calorics
Test Tolerance	7+ years	Improved	More complete, reliable data possible

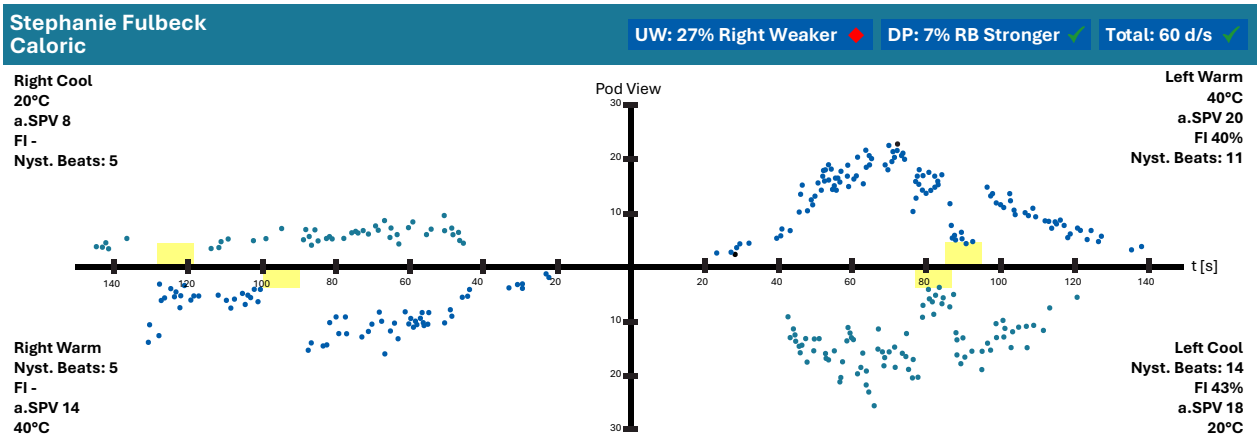

Figure 7.5: Caloric test results depicting a right unilateral peripheral vestibular weakness

Intervention and rehabilitation

Intervention in paediatric vestibular disorders requires a multidisciplinary and individualised approach. Once vestibular dysfunction is identified, timely referral is critical to minimise the impact on a child's motor development, balance, spatial orientation and academic performance. Children exhibiting significant delays in gross motor milestones, persistent imbalance or vertigo should be referred to a paediatric neurologist or Ear, Nose and Throat Specialist for further diagnostic workup and medical management where necessary. In parallel, referral to a paediatric physiotherapist or vestibular rehabilitation therapist is essential for developing an age-appropriate vestibular rehabilitation programme, which may include balance training, gaze stabilization exercises and motor coordination activities. While most evidence-based protocols are derived from adult populations, paediatric vestibular rehabilitation therapy (VRT) is adapted to the developmental level, attention span and motor abilities of each patient. Coordination with occupational therapists, speech-language pathologists or audiologists may also be warranted for patients with coexisting sensory or developmental challenges. The patient's teachers and caregivers should be included in the vestibular rehabilitation plan to ensure that appropriate environmental and educational accommodations are made to support the child in activities of daily living.

Management strategies should be guided by whether the vestibular dysfunction is of peripheral or central origin. In peripheral vestibular disorders, such as vestibular neuritis, bilateral vestibular hypofunction or congenital malformations, intervention typically focuses on promoting compensation through VRT. Activities might include gaze stabilisation exercises (for example, VOR x1 viewing), dynamic balance activities using uneven surfaces or balance boards, and age-appropriate locomotor tasks to improve postural control. In contrast, central vestibular disorders, often associated with conditions such as cerebellar ataxia, traumatic brain injury or multiple sclerosis might require a broader neurorehabilitative approach. This could involve coordination and proprioceptive training, visual motion sensitivity exercises and cognitive-motor integration tasks, often in collaboration with neurology and neuropsychology services. Medication management may be indicated in central pathologies to address symptoms such as migraines or seizures. In both cases, consistent monitoring and re-evaluation are crucial to tailor intervention goals as the child develops and their needs evolve.

Conclusion

The assessment of vestibular function in children is a complex yet essential aspect of paediatric care. Early identification of vestibular disorders through age-appropriate testing, combined with careful interpretation and multidisciplinary collaboration, can significantly improve outcomes in balance, motor development and quality of life. Incorporating both behavioural observations and physiologic tests that are tailored to the child's developmental age ensures a more accurate diagnosis. Equally important is timely intervention, including vestibular rehabilitation, referrals to specialists and supportive educational strategies. As awareness of paediatric vestibular dysfunction grows, HHPs are better equipped to provide comprehensive, family-centred care that supports each patient's full potential.

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Chapter 8

Auditory Processing Assessment and Management in the Paediatric Population

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'If a child cannot hear the language sound – that is, process the sound correctly through their auditory network – they cannot speak it. And if they cannot speak it, they cannot write it.'

~ Dr Anita Collins

Introduction

Auditory processing is vital for making sense of the world, as it involves not just hearing sounds but also interpreting and assigning meaning to them. A breakdown anywhere along the auditory pathway—from the ear to the brain—can disrupt this process, impacting the effective use of auditory and language information. When auditory input cannot be processed effectively, the consequences can be profound if left unaddressed.

The American Speech Hearing Association (2005) and the American Academy of Audiology (AAA, 2010) define auditory processing disorders (APDs) as a deficit in the central auditory nervous system that manages auditory information through neurobiological activity. This definition is widely accepted, but variations have been proposed by authors such as DeBonis (2015), Kraus and White-Schwoch (2015) and Moore (2013). These variations highlight ongoing controversies regarding definitions, diagnosis and intervention. The first part of this chapter aims to provide a foundation on which to build one's approach to auditory processing assessment and intervention, while the second part offers a practical approach to auditory training and management.

Auditory perception relies on the collaboration of multiple brain regions that process, integrate and interpret sound signals. The cochlea converts sound waves into electrical signals, which are refined through brainstem structures like the cochlear nuclei and superior olivary complex for localisation and contrast enhancement. These signals travel through the inferior colliculus and medial geniculate nucleus, integrating sensory inputs before reaching the auditory cortex, where pitch, volume and speech elements are analysed. Higher-level processing in the auditory association cortex, Wernicke's area and Broca's area enables speech comprehension and production, with the corpus callosum ensuring interhemispheric coordination for complex auditory tasks. Table 8.1 describes the processes underlying auditory processing skills, according to ASHA (2005) and AAA (2010).

APD frequently coexists with a range of other conditions, highlighting its complex nature and the importance of a multidisciplinary approach in diagnosis and intervention (Drosos et al. 2024; Gokula et al. 2019). The interplay between APD and these conditions emphasises the need for tailored assessment tools and interventions addressing both auditory and broader developmental challenges (see Chapter 14).

APD is linked to various developmental conditions, including language and speech disorders, learning disabilities like dyslexia and attention deficit hyperactivity disorder (ADHD), due to shared deficits in phonological processing, memory and attention. It also overlaps with autism spectrum disorder (ASD), where auditory filtering and speech comprehension challenges are common. Additionally, APD is associated with working memory and visual attention difficulties, further affecting auditory information processing.

Models of Auditory Processing

To structure an appropriate protocol for an auditory processing assessment, it is imperative that the HHPs adopts a model that will guide their assessment and intervention. Several models provide frameworks for understanding auditory processing and its disorders. Some of the more well-known models include: Erber's Model (Erber 1982), also known as the 'auditory hierarchy', which outlines stages from basic sound detection to comprehension. The Bellis/Ferre Model (2002) categorises APD into sub-profiles linked to different brain regions and emphasises targeted interventions. Katz's Buffalo Model (2019) focuses on key areas such as decoding, memory, organisation and integration, using direct auditory training. The Spatial Processing Model (Dillon and Cameron, 2011) isolates spatial processing disorder

(SPD) as a distinct subtype, highlighting the brain's ability to filter directional sound, whereas Medwetsky's Spoken Language Processing Model (2011) integrates auditory, cognitive and linguistic mechanisms with a pragmatic, real-world approach. Finally, the British Society of Audiology Approach (Moore et al. 2013) classifies APD into developmental, acquired and secondary types, recommending assessments and tailored management strategies. Each model provides a unique framework for assessing and treating APD, highlighting the importance of a comprehensive approach to address various auditory challenges. HHPs can choose and adapt these models to structure their assessment and treatment strategies effectively.

Table 8.1: Processes underlying auditory processing (ASHA 2005; AAA 2010)

Auditory Processing Skill	Sound Localization and Lateralization	Auditory Discrimination	Auditory Pattern Recognition	Temporal Aspects of Audition	Auditory Performance Decrements with Competing Signals	Auditory Performance Decrements with Degraded Signals	Amblyaudia	Spatial Processing Disorder
Description	Localization: Identifying where a sound occurs in space. Lateralization: Perceiving sound within the head (e.g. left or right side) when using headphones.	The ability to differentiate one sound from another, not limited to speech sounds.	Identifying and interpreting patterns in sounds including speech, music and environmental noises.	How we perceive and process the timing of sounds, essential for speech and music comprehension.	Challenges in understanding sounds amidst competing background noises and other sounds.	Difficulty processing incomplete, distorted or time-compressed sounds.	Neural integration disorder characterized by abnormal asymmetry between the ears during binaural integration tasks (Moncrieff and Black 2008).	A type of APD involving difficulty using spatial cues to separate sounds in noisy environments (Cameron et al. 2014).
Key Components	Crucial for navigating and interacting with the environment enabling accurate sound pinpointing and appropriate responses.	Fundamental for recognizing diverse auditory stimuli.	Processing timing, pitch, timbre and sequence for speech, music and sound differentiation. Involves memory, attention, integration and cortex.	Temporal resolution; Temporal Integration; Temporal Ordering; Temporal Masking.	Common in noisy environments; hinders focus and comprehension.	Affects understanding in situations where acoustic signals are compromised (e.g. poor phone reception or muffled speech).	Normal performance in the dominant ear. Requires consistent interaural asymmetry across two or more dichotic listening tests.	Difficulty processing sound timing and intensity differences; affects localization, separation, and speech understanding in noise.

Criteria for the Diagnosis of APD

The criteria required to make a diagnosis of APD as proposed by ASHA (2005) and espoused by AAA (2010) includes the following: APD exists when the results are two or more standard deviations (SDs) below the mean on any two tests or three SDs below the mean on any one test. Dillon, Tomlin and Kelly (2014) suggest focusing on patterns of deficits or weaknesses rather than strict cut-off scores. They argue that using a hard cut-off of -two SD below the mean can exclude children who are close to this threshold (for example, -1.92SD) from receiving services, despite showing a pattern of weakness. To address this, they recommend using a double arcsine transformation to convert scores to age-specific Z-scores that have accounted for the asymmetric distribution of the normative results to allow for a more accurate comparison.

There are certain aspects that teachers, psychologists, occupational therapists, academic support specialists, paediatricians, caregivers and others working with children can use as a checklist or indicators for APD (See Appendix 8.1). In young children, the behaviours which should raise concerns would include difficulty learning rhymes and songs, inattention to the speaker or ignoring a speaker when participating in other activities, sensitivity to sound or noise, and difficulty with complex directions. Currently, HHPs are able to assess for APD from a much younger age (three years and six months), and these warning signs could enable earlier assessment and intervention than was previously thought.

Assessment of APD

Early identification of APD is crucial. While assessment was traditionally recommended from age eight, normative data now support testing as early as three years and six months using tools such as the Auditory Skills Assessment Test (Geffner and Goldman, 2010), Staggered Spondaic Word Test (Katz 1998), SCAN-3C (Keith 2009), Phonemic Synthesis Picture Test and Phoneme Recognition Test (Katz 1998).

An individualised, evidence-based approach is essential, ensuring that HHPs have the necessary training and support to conduct evaluations effectively. The test battery should align with the patient's specific concerns, clinical framework and principles of validity, reliability and efficiency. Given the variability in APD and learning disabilities, determining test

reliability can be challenging; however, tests that successfully identify lesions remain the standard for validity. Assessments should thus incorporate tests for various auditory processes using both verbal and nonverbal stimuli with minimal linguistic bias. Even without normative data, qualitative evaluations can provide valuable insights, establish baselines and guide interventions.

Considering the individual's characteristics enhances assessment accuracy, allowing HHPs to tailor their approach for optimal comprehension and motivation. Awareness of the lowest normed test age is crucial, particularly for electrophysiologic measures, where understanding primary pathway development—especially middle latency responses —is vital. Standardised protocols should be followed, with documented modifications as needed, and session duration should be monitored for patient well-being. Collaboration with other professionals can enhance the assessment process, leading to appropriate referrals for suspected additional deficits.

An effective APD assessment should integrate test results with case history and observations to tailor interventions. The primary goals should include confirming the disorder, identifying patterns of deficits and establishing a differential diagnosis by linking auditory challenges to real-world functional difficulties. Diagnostic accuracy depends on intra- and inter-test pattern analysis across auditory and multidisciplinary evaluations and ensures ecologically valid, individualised intervention plans. Inconsistencies in test results, such as differing ear advantages on dichotic tests or uniformly poor performance, should prompt a review of test specifics and potential confounding factors, including attention, motivation, cognitive abilities or language deficits.

Tools available for APD assessment

The assessment process depends on the chosen model, with each providing theoretical support for test selection, evaluation areas and result interpretation. The HHP is responsible for investigating these foundational elements. Research emphasises the importance of evaluating the influence of cognitive factors, such as memory, attention and intelligence on auditory processing. These variables should be accounted for when analysing the relationship between auditory processing abilities and functional performance in real-world contexts to ensure accurate interpretation and application of findings (Seeto, Tomlin and Dillon 2021). Every clinician, clinic and practice should develop a context-specific protocol tailored to its

demographic, patient age group, co-morbid conditions, referral reasons and approach to auditory processing. Below is a suggested framework:

Screening APD assessment

Referral sources such as speech-language therapists, psychologists, teachers, academic support staff and HHPs might use screening tools to justify a diagnostic, person-centred assessment. Appendix 8.1 lists 'red flags' for referrals. Before auditory processing screening, at least a basic hearing test should be done, and ideally a full diagnostic hearing assessment. Age-appropriate, culturally relevant tests should be selected (see Table 8.2), considering administration time and proper training for accurate interpretation. If a child scores below the expected range, a comprehensive auditory processing assessment by an HHP is recommended to delve deeper into specific auditory skills and formulate individualised intervention plans.

Table 8.2: Screening tools available for APD

Test Name	SCAN-3 (Child)	Auditory Skills Assessment (ASA)	Differential Screening Test for Processing (DSTP)
Subtests	• Filtered Words • Auditory Figure Ground (SNR + 8dB) • Competing Words • Competing Sentences • Gap Detection • Time Compressed Sentences • Competing Words Free Recall • Auditory Figure Ground (SNR +0dB and +12dB)	Speech Discrimination Domain: • Speech discrimination in noise • Mimicry Phonological Awareness Domain: • Blending • Rhyming Nonspeech Processing Domain: • Tonal Discrimination • Tonal Patterning	Level One – Acoustic Subtests: • Dichotic Digits • Temporal Patterning • Auditory Discrimination Level Two – Acoustic Linguistic Subtest: • Phonemic Manipulation • Phonic Manipulation Level Three – Linguistic Subtests • Antonyms • Prosodic Interpretation • Language Organization
Age	5.00 – 50.11 years	3.6 – 3.11 years	6 - 12 years
Areas Assessed	Differentiation of auditory processing disorder from auditory attention and auditory comprehension difficulties.	• Speech Discrimination • Phonological Awareness • Nonspeech auditory processing"	Acoustic Processing Acoustic-Linguistic Processing Linguistic Processing

Test Name	SCAN-3 (Child)	Auditory Skills Assessment (ASA)	Differential Screening Test for Processing (DSTP)
Comments	The SCAN-3 includes screening and diagnostic tests. Screening takes 10-15 mins and diagnostics take 30-45 mins. Not all tests are required. Filtered Words and Auditory Figure Ground tests may be less sensitive. Competing Words and Sentences tests assess attention, binaural integration and separation.	The ASA is a criterion-referenced screening tool that identifies young children at risk for APD. It takes 5-15 mins to administer depending on the child's age and ability and yields a single age-based cut score. It is also useful for monitoring progress in young children.	The DSTP uses a processing continuum from auditory to language processing. It is a screening test only and should not be used for diagnostic purposes. The Pass/ Fail criteria are already one standard deviation below the mean.

The assessment should include pre-assessment, an intake interview and an in-person or online clinical assessment. Each sub-section of the assessment is described below.

Pre-assessment

a) Case History

Caregivers, teachers or the patient should complete a detailed case history form. This will serve as a valuable starting point for discussions during the intake interview. Any available reports (for example, from educators and medical professionals) should be collected to build a comprehensive understanding of the patient's challenges. Having relevant professionals and family members complete questionnaires provides insight into their perception of the areas of difficulty. Commonly used questionnaires include the Buffalo Model Questionnaire (Katz et al. 2019), The Fisher's Checklist (Fisher 1976), eClips (Barry et al. 2015), CHAPPS (Smoski, Brunt and Tanahil 1998) and the Auditory Processing Domains Questionnaire (O'Hara and Mealings 2018).

b) Intake interview

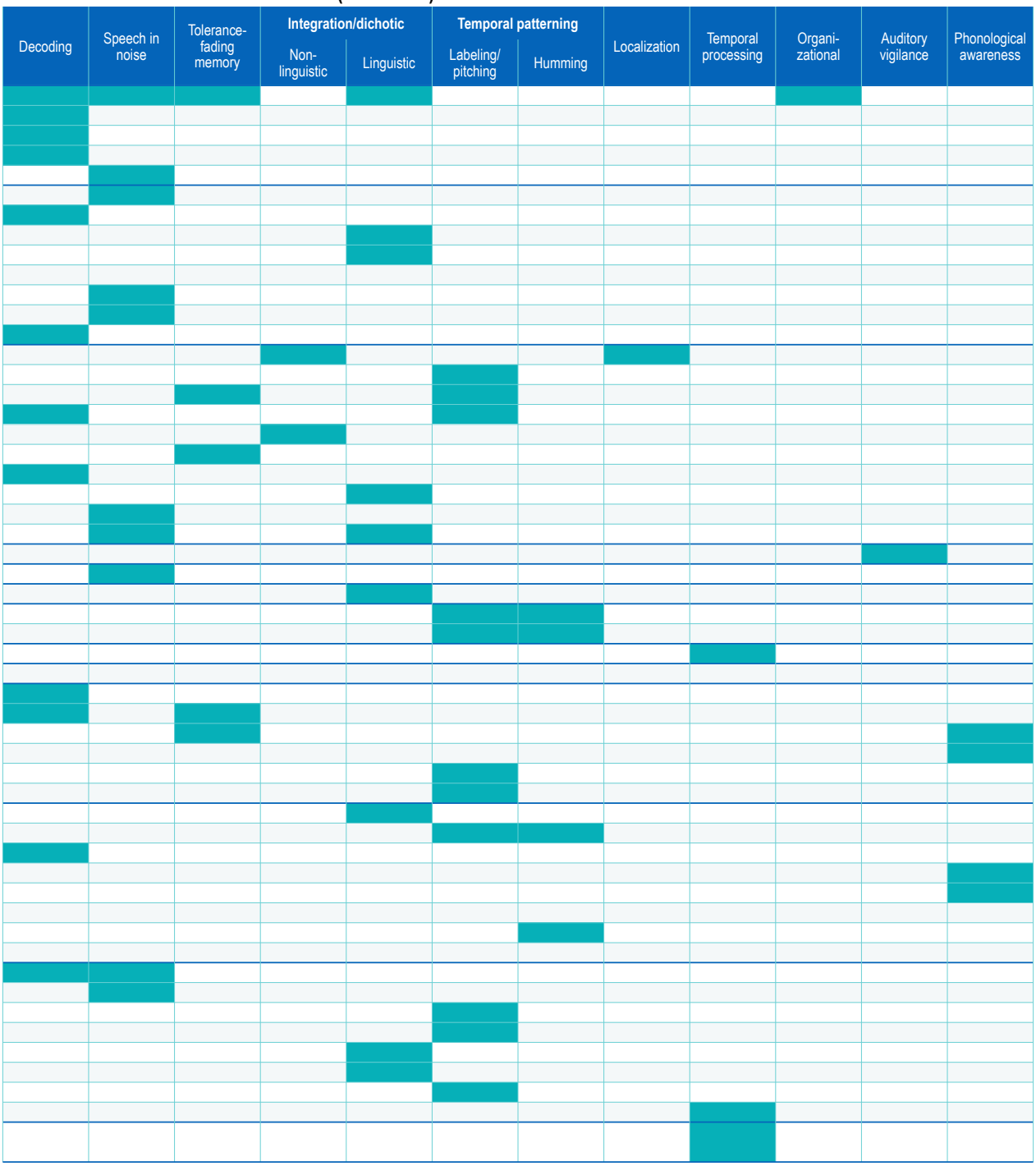
There are various factors to consider when scheduling and preparing for an auditory process-ingassessment as these factors will modify the assessment process. These considerations include the age of the child, time and length of assessment, presenting concerns, cognitive status, emotional and psychological well-being and language and hearing status. An interview with the caregivers (without the child present) should be conducted to encourage open discussion. If resources allow, peripheral hearing assessments should be conducted in parallel by another clinician. The hearing assessment should include pure tone audiometry, speech audiometry, immittance measures and otoacoustic emissions.

The assessment tools selected need to be appropriate to the child as well as to the social and economic environment in which the testing occurs. Not all assessment tools are affordable or appropriate in every situation. In many countries, there is limited access to audiological services for peripheral hearing difficulties, let alone for APD. HHPs working in areas where access to appropriate auditory processing assessment tools is not readily available should not limit their investigation, but should instead use available tools to collect information that will guide intervention. Table 8.3 provides an introduction to some of the auditory processing assessment tools available. More extensive information is available in Appendix 8.2.

APD ASSESSMENT TESTS SUMMARY

TEST	SUBTEST	SUPPLIER	AGE
Central Test Battery	Staggered Spondaic Word Test	Precision Acoustics	5 and above
	Phonemic Synthesis Test		6 and above
	Phoneme Recognition Test		All
	Phonemic Synthesis Test with Pictures		4–7 years
	W-22 in Noise and Quiet		5 and above
SCAN-3	Auditory Figure-Ground +8	Pearson	6 and above
	Filtered Words		
	Competing Words		
	Competing Sentences		
	Competing Words Free Recall		
	Auditory Figure-Ground +12		
	Auditory Figure-Ground 0		
	Time Compressed Speech		
Feather Squadron	Hearing Screening and Lateralisation	Acoustic Pioneer	5 and above
	Tonal Pattern Temporal Processing		
	Tonal Pattern Memory		
	Rapid Tones		
	Dichotic Sounds/Double Sounds		
	Word Memory		
	Rapid Speech		
	Dichotic Double Sounds		
	Speech in Noise (Without Localisation Cues)		
	Speech in Noise (With Localisation Cues)		
Auditory Continuous Performance Test		Auditec	6.0–11.11years
Listening in Spatial Noise –S/U		Soundstorm	6 and above
Dichotic Digits Test		Auditec of St Louis	7 and above
Temporal Patterning Tests	Frequency Patterning	Auditec of St Louis	7 and above
	Duration Patterning		7 and above
Gaps in Noise Test		Auditec	7 and above
Random Gap Detection Test		Auditec	5 and above
Auditory Skills Assessment	Speech Discrimination in Noise	Pearson	3.6–6.11 years
	Mimicry		
	Blending		
	Rhyming		
	Tonal Discrimination		
	Tonal Patterning		
Differential Screening Test for Processing	Dichotic Digits	LinguiSystems	6.0–12.11 years
	Temporal Patterning		
	Auditory Discrimination		
	Phonemic Manipulation		
	Phonic Manipulation		
	Antonyms (Language)		
	Prosodic Interpretation		
	Language Organization (Language)		
Multiple Auditory Processing Assessment	Monaural-Selective Attention Test	Academic Therapy Publication	8 and above
	Speech in Noise for Children		
	TAP Test		
	Pitch Pattern Test		
	Dichotic Digits		
	Competing Sentences		
	Duration Pattern Test		
	Gap Detection		
Frequency FollowingResponse	Experimental		

APD ASSESSMENT TESTS SUMMARY (continued)



Post-assessment

a) Feedback session

A follow-up visit should be scheduled to review findings with caregivers and discuss recommended auditory training and management strategies. To support caregiver understanding and engagement, structured feedback materials (for example, a feedback book or PowerPoint presentation) may be used to guide the session, ensuring that complex audiological information is communicated in clear, accessible language appropriate for non-specialist audiences.

b) Analysis and interpretation

Results should be interpreted in the context of the case history, accompanying reports, observations and assessment outcomes. No assessment task should be considered in isolation; instead, a pattern of responses should be identified to understand what each task assesses. Interventions must be process-based, targeting specific auditory processes identified as areas of deficit (see Table 8.4). These may include auditory training, compensatory strategies and environmental modifications. When patterns are unclear, a broader scope of interventions addressing both auditory and non-auditory factors should be considered to comprehensively support the child.

Table 8.4: Patterns of responses

Test	Deficit	Underlying Process
Staggered Spondaic Word Test	"Decoding Tolerance Fading Memory"	Decoding
Feather Squadron	"Rapid Speech Speech in Noise"	
Scan- 3 (Child)	"Filtered Words Auditory Figure Ground Time Compressed Speech"	

Test	Deficit	Underlying Process
Feather Squadron	"Dichotic Sounds Dichotic Words"	Integration
Scan- 3 (Child)	"Competing Words Competing Sentences"	
Dichotic Digits	Dichotic Digits	
Feather Squadron	Temporal Pattern Temporal processing	"Temporal Patterning (Prosodic)"
Temporal Patterning	Frequency Patterning	
Gaps in Noise	Temporal Resolution	

c) Recommendations

Recommendations should be person-centred and tailored to the specific needs of the child rather than relying on generic suggestions (Chapter 15). Training recommendations must be directly based on assessment findings to ensure targeted and effective intervention. While generalised strategies are not encouraged, certain activities have been shown to enhance auditory processing skills, such as learning to play a musical instrument, listening to audio-books and engaging with songs and rhymes.

d) Referrals

An educational psychologist or paediatrician might be consulted for cognitive, memory and attention difficulties, while an occupational therapist can assist with sensory integration issues beyond auditory challenges. A psychologist may be involved to address anxiety or behavioural concerns, and a speech-language therapist can provide intervention for language-based difficulties. By following this structured and adaptable approach, clinicians can ensure assessments are thorough and person-centred, and lead to actionable insights for intervention and support.

Electrophysiological Assessment

The Frequency Following Response (FFR) is a neurophonic auditory evoked potential that reflects how efficiently speech sounds are encoded. FFR uses the complex syllable /da/ to evoke auditory brainstem response (ABR), which is mainly used to analyse the neural activity of the brainstem to speech stimulus. The FFR is disrupted in various speech and language disorders, making it a valuable tool for testing auditory processing abilities across different languages. Unlike the click-evoked ABR, which identifies hearing loss or auditory neuropathy, the FFR detects real-world auditory processing deficits. For instance, individuals with concussions may have normal ABRs, yet the FFR can reveal subtle deficits with high accuracy (95% sensitivity and 90% specificity). The FFR also shows promise as a potential biomarker for literacy. In children with normal ABRs, atypical FFR results may predict a higher risk for language development issues, making it a valuable diagnostic tool. Although currently expensive, it represents a significant advancement and is likely to shape the future of APD assessment. Its value in countries where English is not the first language will be paramount, as assessment will not be limited to non-linguistic or low-linguistically loaded assessment tools.

Intervention for APD

Intervention for APD should cover three areas (Bellis 2011): a) direct auditory training (changing the individual), b) environmental modifications (changing the world of the individual) and c) compensatory strategies (helping the individual to change their world).

Direct auditory training for APD is grounded in several key principles, as highlighted by leading experts in the field (Barker and Hicks 2020; Bigras et al. 2024; Geffner and Ross-Swain 2026a; Weihing et al. 2015). Tailoring auditory training programmes to address the specific deficits of each patient is crucial. This personalised approach ensures that interventions target the unique auditory challenges faced by the person, leading to more effective outcomes. Auditory training typically incorporates both bottom-up and top-down processing approaches. Bottom-up training focuses on strengthening the neural encoding of acoustic features such as frequency, intensity and temporal cues, whereas top-down strategies engage higher-order cognitive processes, including attention, memory and linguistic knowledge, to support sound interpretation. Integrating these complementary approaches allows

for more comprehensive improvement in auditory processing abilities, particularly in complex listening environments.

Implementing adaptive training methods that adjust the difficulty level based on an individual's performance helps maintain engagement and propels continuous improvement in auditory skills. Additionally, incorporating multisensory elements, such as visual aids or tactile feedback, enhances learning by engaging multiple sensory pathways, thereby reinforcing auditory processing skills. Continuous assessment of progress allows for modifications to training programmes, ensuring that interventions remain effective over time and evolve with the individual's needs. By adhering to these principles, clinicians can design and implement auditory training programmes that effectively address the specific needs of individuals with APD, improving their auditory processing capabilities and overall quality of life.

There has been a resurgence in interest in providing direct, person-centred and evidence-based auditory training for individuals with APD, with a move away from generic recommendations (Katz et al. 2019). The following are commonly used auditory training models:

- Buffalo Model of Auditory Training (BMAT) is aimed at addressing auditory-related deficits in a gradual or incremental manner to change the unformed or incorrect representations imprinted on the auditory cortex. Given the challenges in sub-Saharan Africa related to affordability, geography, language, culture and accessibility, the BMAT (Katz et al. 2019) offers a practical, low-tech solution for addressing auditory processing deficits. The BMAT is designed to gradually reshape incorrect or underdeveloped auditory representations in the auditory cortex through structured training over twelve to fourteen sessions, each lasting 45–60 minutes. While the programme follows a structured framework, it can be adapted to an individual's language without compromising its effectiveness. This bottom-up approach builds foundational auditory processing skills, focusing on phoneme recognition in quiet and noise, phonemic synthesis, words-in-noise training, and short-term auditory memory development. Dichotic offset training further enhances dichotic listening skills. Essential materials include phoneme cards, phonemic synthesis exercises (which can be purchased or created by the clinician), short-term memory tasks and pre-recorded words-in-noise training tracks. By targeting key auditory skills, the BMAT enhances speech perception, auditory memory and overall listening abilities, ultimately improving communication and

learning outcomes for individuals with APD. Additionally, the programme can be adapted for online training, expanding access to those unable to attend in-person sessions while still benefiting from structured auditory intervention. The BMAT should be considered a sound auditory training approach for providing auditory training services that may not previously have been available. Clear, step-by-step guidance to evaluate APDs has been provided by Alexander and Abbas (2026), and the same steps can be used for providing online auditory training.

- Auditory Rehabilitation for Interaural Asymmetry (ARIA) was developed in 2000 (Moncrieff and Black 2008) to address interaural asymmetries identified in children during tests of dichotic listening. The term 'amblyaudia' was coined to refer to these asymmetries. Specific, hands-on training is provided to learn how to administer the ARIA. The ARIA is a training programme presented over four weeks for 25 minutes per session. Materials required to administer the training include an audiometer with two channel capabilities, the ARIA software played through a computer or tablet and normal audiological equipment such as high-quality headphones, a quiet environment and a means for the individual to provide feedback. Despite its reported efficacy in addressing dichotic listening difficulty, the range of materials needed limits its use to more clinical environments.

Numerous computer and tablet applications are available for direct auditory training. However, it is essential to recognise that these programmes cannot replace clinician-driven intervention. Instead, they serve as valuable supplementary tools that can enhance direct intervention or provide an alternative when direct intervention is not feasible. The HHP remains responsible for overseeing the training process, monitoring progress and offering support as needed to ensure effective outcomes. The professional grade auditory training programmes that should be considered include the following:

- *ZooCaper SkyScraper* (Barker and Bellis 2018; Barker and Hicks 2020) is a tablet- or computer-based therapy programme designed to develop dichotic listening skills. It is engaging for all ages and structured for completion over approximately 12 weeks, with two-to-three sessions of 20 minutes per week. The HHP can monitor progress remotely and provide guidance as needed.
- *Insane Earpanes* (Barker and Bellis 2018; Barker and Hicks 2020) is another tablet- or computer-based training programme that strengthens auditory processing skills, including lateralisation, pitch pattern recognition, pitch sweeps

and interaural timing and intensity differences. It incrementally builds these skills based on AAA and ASHA recommendations and is suitable for all ages. Like ZooCaper SkyScraper, it is designed to be engaging and manageable within a structured timeframe.

- *Elephant Memory Training* (Barker and Bourland-Hicks, 2020) is a top-down training programme that is conducted four times per week over an eight-week period. Working memory is a critical component in a dynamic processing system that includes auditory processing as well as language and literacy, and relates to the storage and manipulation of information.
- *Sound Storm* (formerly LiSN & Learn) (Cameron and Dillon 2008) is an auditory training programme designed to assist children with SPD with managing classroom listening challenges. By enhancing their ability to suppress background noise and focus on target auditory stimuli, the programme aims to improve speech perception in noisy environments. The programme creates a three-dimensional auditory experience through headphones on an iPad. The training is structured around an engaging space-fantasy narrative, guiding users through ten progressive levels across nine distinct galactic worlds, making the intervention immersive.
- *CAPDOTS* (Lucker et al. 2021) is a web-based AT programme designed to address binaural auditory integration deficits, commonly referred to as dichotic listening difficulties. The programme requires a stable internet connection and high-quality headphones and can be administered in a clinical setting, school or home environment. According to its developers, CAPDOTS enhances skills such as following multi-step directions, maintaining listening attention in background noise, improving inferential listening, understanding group instructions and strengthening auditory memory. It is also reported to support academic performance, particularly in reading comprehension and spelling. While this auditory training tool allows access for those unable to travel to in-person therapy, it requires the support of a capable adult to assist the individual during training. In addition to these professional-grade auditory training programmes, there are useful applications to support and enhance in-person auditory training (see Table 8.5).

In school settings, educators can make use of a number of environmental modifications, including the use of assistive listening devices (Chapter 10) as well as compensatory strategies (Chapter 13).

Table 8.5: Applications to support auditory training

APP	Broad Area	Specific Area	Link
LACE AI: Listening and Communication Enhancement	Direct Remediation	Figure-ground training; auditory separation; temporal integration; auditory working memory	*https://www.laceauditorytraining.com/*
Functional Listening App	Listening in Noise	Figure-ground listening	*https://apps.apple.com/us/app/functional-listening/id1100839826*
Hear Beyond		Listening in noise; sound localization; sound sequencing; pitch patterning; working memory	*https://www.hearbeyond.world/*
Hearapp/ HearappKids	Listening in noise	Word and phrase identification; identification of words and phrases in quiet and noise	*https://apps.apple.com/us/app/hearapp-kids/id1010536075*
Clearly Audiobooks	Speech discrimination; listening in noise		*https://clearlyaudiobooks.com/*
I-Angel Sounds	Sound discrimination and identification	Sound and speech identification; temporal patterning	*http://iangelsound.emilyfufoundation.org/*

APP	Broad Area	Specific Area	Link
ReDi	Temporal patterning	Sound sequencing	
SpeechBanana	Phoneme/speech discrimination task		*http://speechbanana.jhu.edu/*
Heroes	Sound identification	Environmental sounds; introduction to words; single vowels; common consonants; understanding sentences	*https://www.hearoes.com/*
Listening Adventures	Development of listening and language skills		*https://apps.apple.com/us/app/ab-listening-adventures/id898695069*
Auditory Training App		Dichotic digits; alternated dichotic; non-verbal sound; temporal patterns	*https://apps.apple.com/us/app/auditory-training/id1637321202*
Pocket Phonics/ Pocket Phonics Stories	Phonological awareness and processing	Phoneme recognition and identification	*https://www.readingrockets.org/resources/literacy-apps/pocketphonics*
CVC Train	Phoneme-to-grapheme skills	Letter recognition and sound blending	*https://wordwall.net/en-us/community/cvc-train*

Conclusion

Children with APD often struggle with following verbal instructions, listening in noisy environments and completing language-related tasks. These difficulties can overlap with conditions like ADHD or language disorders, makinan g accurate diagnosis through multidisciplinary assessment essential. Without intervention, APD can hinder academic success and social development, leading to frustration and low self-esteem. Early identification and targeted support equip children with the necessary skills to navigate their challenges, promoting confidence, independence and long-term success. While further research in APD is needed, access to services should not be denied based on research thresholds alone. Addressing APD is not just about mitigating auditory challenges; it is about empowering children with strategies and support to reach their full potential in all areas of life.

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Appendix 8.1: Red flags and concerning behaviours for APD

- Poor listening skills.
- Difficulty learning through the auditory modality; does better with visual stimuli.
- Difficulty following auditory instructions, especially multi-step instructions or instructions embedded in other information.
- Short-term and auditory memory span deficiencies; trouble retaining sequences or retelling stories.
- Difficulty understanding speech in background noise, reverberant environments, or when speech is distorted or rapid.
- Frequently says "what?" or "huh?" and often requests repetition.
- Mishearing or misunderstanding spoken words or messages.
- Delayed or slowed response to verbal stimuli.
- Poor auditory attention and fatigue with prolonged listening tasks.
- Easily distracted by environmental sounds; hears background noise more prominently than the speaker.
- Reduced tolerance to loud environments or hypersensitivity to sound.
- Deficits in auditory integration, including sound blending, auditory closure, phonological awareness, rhyming, and phonic skills.
- Inability to detect subtle acoustic differences and changes in prosody, such as sarcasm or humour.
- Difficulty with speech recognition in noise; figure-ground discrimination deficits.
- Problems understanding and retaining orally presented information, including stories and classroom discussions.
- Problems with spelling, reading, and written language.
- Difficulty with foreign language learning.
- Cannot write accurately from dictation.
- Cryptic or insignificant notetaking; misses key points or facts.
- Struggles with mathematical word problems.

- Takes longer to process and respond to questions or instructions.
- Poor singing ability, music appreciation, or musical memory ("poor ear for music").
- Tinnitus, especially perceived in the head.
- Academic underperformance or learning difficulties.
- History of middle ear infections (otitis media).
- Fidgety or uncomfortable in noisy settings, such as the cafeteria or gym.
- Avoids participating in class discussions or group learning tasks.

Appendix 8.2: Summary of auditory processing assessment tools

Test	Link
Central Test Battery (CTB) (Katz 1998)	*https://precisionacoustics.org/products/central-test-battery-kit*
Feather Squadron (Barker 2017)	*https://acousticpioneer.com/*
Auditory Continuous Performance Test (Keith 1994)	*https://auditec.com/2015/10/26/auditory-continuous-performance-test-acpt/*
Listening in Spatial Noise – U (Cameron & Dillon 2008)	*https://www.soundscouts.com/*
Dichotic Digits Test	*https://auditec.com/2015/10/01/dichotic-digits/*
Temporal Patterning Tests: Frequency Patterning; Duration Patterning	*https://auditec.com/category/temporal/*
Gaps in Noise Test (Musiek 2005)	*https://auditec.com/2015/09/22/gaps-in-noise-gin/*
Multiple Auditory Processing Assessment (MAPA-2)	*https://assessments.academictherapy.com/sku/2211-8*
(Cameron & Dillon 2008)	*https://www.soundscouts.com/*
Dichotic Digits Test	*https://auditec.com/2015/10/01/dichotic-digits/*
Temporal Patterning Tests: Frequency Patterning Duration Patterning	*https://auditec.com/category/temporal/*
Gaps in Noise Test (Musiek 2005)	*https://auditec.com/2015/09/22/gaps-in-noise-gin/*
Multiple Auditory Processing Assessment	*https://assessments.academictherapy.com/i/multiple-auditory-processing-assessment-mapa-2*

Chapter 9

Surgical Solutions for Conductive Hearing Loss in Children

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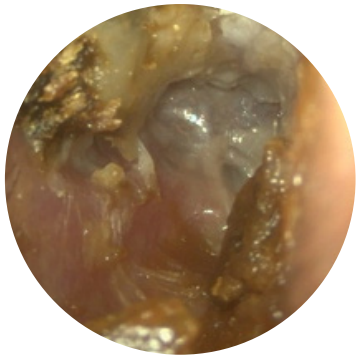
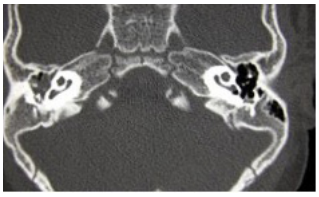
There is something beautiful about all scars of whatever nature.'
~Harry Crews

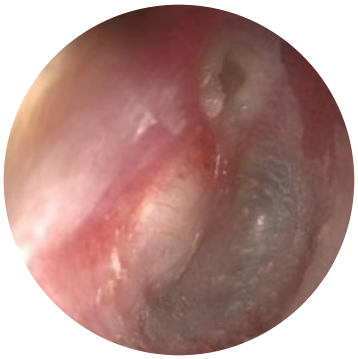
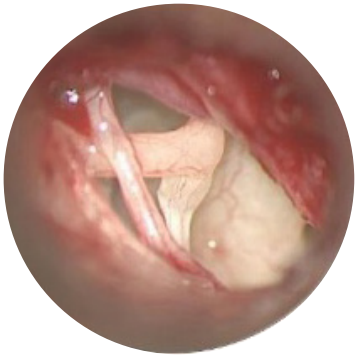
Introduction

Conditions leading to conductive hearing loss often require surgical intervention performed by an ear, nose and throat (ENT) surgeon. While the procedures themselves fall within the ENT surgeon's scope, it is important for hearing healthcare professionals (HHPs) to understand the underlying conditions and the surgical approaches used, and to work collaboratively with the surgeon to ensure optimal patient outcomes. This chapter also addresses the benefits of wax removal, which, although not strictly surgical, can improve hearing and support overall ear health. Table 9.1 lists the medical conditions most commonly associated with conductive hearing loss (CHL).

Table 9.1: Medical conditions commonly causing CHL

Condition causing CHL	Degree of CHL	Depiction of conductive condition *(all images are of Right ears)*
Cerumen *(earwax)*	Variable, depending on the extent of obstruction; typically 5–40 dB HL	
Canal atresia *(image shows CT scan of ear with atresia)*	Typically maximal: up to 60 dB HL	

Condition causing CHL	Degree of CHL	Depiction of conductive condition *(all images are of Right ears)*
Otitis Media with Effusion (OME) *(image shows retraction of tympanic membrane with OME)*	Variable: none to 30 dB HL, typically 20–30 dB HL	
Perforated tympanic membrane *(image shows antero-inferior 'central' perforation of tympanic membrane)*	Variable depending on the size and site of the perforation: less than 30 dB HL	
Ossicular chain discontinuity/fixation *(image shows retracted and tympanosclerotic tympanic membrane with erosion of incus long process which fails to meet the stapes head)*	More than 30 dB HL	

Condition causing CHL	Degree of CHL	Depiction of conductive condition *(all images are of Right ears)*
Cholesteatoma *(image shows pars flaccida/'attic' tympanic membrane defect with cholesteatoma visible behind the posterior-superior tympanic membrane)*	Variable depending on the pathology: none to greater than 30 dB HL	
Tympanosclerosis *(image shows extensive tympanosclerosis of the tympanic membrane)*	Very rarely causes CHL – only if it fixates to part of the ossicular chain	
Stapes fixation *(Image of the middle ear with the tympanic membrane lifted to show the incudostapedial joint being crossed by chorda tympani nerve)*	Variable depending on the fixity	

The surgical interventions for CHL which will be discussed are:

- Cerumen (earwax) removal and desquamated skin removal from the ear canal
- Meatoplasty
- Canalplasty
- Ventilation tubes ('grommets')
- Tympanoplasty/Myringoplasty
- Ossiculoplasty
- Surgical repair of hearing as part of cholesteatoma surgery
- Stapedioplasty (stapedectomy/stapedotomy)
- Eustachian tube (ET) dilation

Cerumen Removal

Total blockage of the ear canal with cerumen and desquamated skin of the ear canal is rare, and soundwaves only need a tiny opening in the ear canal to reach the tympanic membrane (TM). Thus, even a large amount of earwax should not cause noticeable hearing loss (HL). Cerumen plays a protective role in the external auditory canal and is typically moved outward by the canal's natural epithelial migration and jaw movements. However, this self-cleansing mechanism does not always prevent buildup, and cerumen impaction is a common clinical problem.

If the ear canal is plugged tightly with earwax, cerumen impaction or earwax blockage, it can cause CHL of up to 48 decibels (dB) (Narayanan, Raman and Chong 2019). Removal can be achieved by syringing, mechanical removal with a Jobson-Horne probe, suction under microscopic or endoscopic visualisation or a combination of the above. Various syringing systems exist, ranging from a simple 50ml syringe with a luer lock and cannula through purpose-designed syringes to devices which pump water automatically. Removal is useful to provide good visualisation of the eardrum. Cerumen removal can generally be performed by a trained audiologist, nurse, general practitioner or ear, nose and throat (ENT) surgeon.

Syringing should always be performed gently, using body-temperature water (37 degrees Celsius) to prevent discomfort and vertigo from caloric (temperature-related) effects. Prior to syringing, one should ensure that there is no history of a perforation of the TM or grommet insertion – in which case syringing is contra-indicated. One should then establish the

confidence of the child and explain what is going to be done. Prior softening of the wax may be necessary or useful. As wax is water-soluble, ordinary water or dilute hydrogen peroxide (1.5%) is as effective as commercial preparations. The parent or caregiver can be given a dropper bottle and asked to instil the water/peroxide, jiggling the pinna or tragus for about 20 minutes prior to syringing.

The principles of the technique include:

- Having unrestricted access to the ear.
- Providing good light (possibly with a headlamp)
- Pulling the pinna backwards to straighten the ear canal
- Filling the canal gently with water before syringing more firmly
- Patience – allowing the combination of the dissolving action of the water as well as the mechanical water flow to loosen the wax
- Placing a container under the ear to catch the water

Meatoplasty and/or Canaloplasty

A narrowed, closed (stenosed) or undeveloped (atretic) bony ear canal (EAC), or a similarly affected cartilaginous external auditory meatus (EAM), can impede sound transmission to the eardrum directly or indirectly by promoting cerumen obstruction. These ear canal conditions may also make the fitting of a hearing aid (HA) problematic.

When such EAM/EAC narrowing or atresia is congenital, it may be associated with anomalies of the middle ear and/or inner ear which may contribute to HL – conductive and/or sensorineural. Mild or moderate cases of EAM/EAC narrowness can be improved by relatively simple surgical techniques. A narrow EAM can be widened by resection of obstructive cartilage and the transposition or interdigitation of small skin flaps – as, for instance, in the technique of M-Meatoplasty (Mirck 1996). A narrow bony EAC can be widened by drilling of the ear canal, splitting or 'barn-dooring' the EAC skin and grafting the resulting gap with fascia.

Severe congenital anomalies, as are found in atresia or microtia, represent a far more difficult problem for surgical reconstruction (Jahrsdoerfer and Hall 1986). Expert surgeons with significant experience report that even in their hands the results are often unsatisfactory, both in terms of hearing outcomes and in the health and stability of the newly-created ear

canals. It should only be considered by surgeons who are expert in this area and in cases where the radiology (CT and MRI) is favourable (McKinnon and Jahrsdoerfer 2002). A preferable solution is to address the often-associated cosmetic aspects of the pinna with surgery or a prosthesis, and to leave the EAC stenosed in favour of a bone-anchored hearing solution (Chapter 11).

Ventilation tubes ('tympanostomy tubes'/'grommets')

Ventilation tubes (VTs) may be indicated for insertion into the eardrums of children with otitis media with effusion (OME). OME is common enough to be called an 'occupational hazard of early childhood' (Rosenfeld 2004) by the American Academy of Otorhinolaryngology: Head and Neck Surgery (AAO-HNS), as 90% of children have OME before school age (Tos 1984) and they develop, on average, four episodes of OME every year (Mandel et al. 2008). Most episodes of OME resolve spontaneously within three months, but at least 25% of OME episodes persist for ≥3 months (Rosenfeld and Kay 2003) and 5% to 10% of episodes last ≥1 year (Stool et al. 1994; Williamsonet al. 1994). The prevalence and persistence of OME in children with Down Syndrome or cleft palate, however, is much higher, ranging from 60% to 85% (Flynn et al. 2009; Maris et al. 2014).

The HL associated with OME varies considerably (Fria, Cantekin and Eichler 1985), typically ranging from mild to moderate CHL (Moore, Hartley and Hogan 2003), although some children may have no detectable HL. In light of the above, the AAO-HNS has produced guidelines on OME (Rosenfeldet al. 2016) and ventilation tubes (Rosenfeldet al. 2022). These guidelines include the following recommendations:

- Determine if a child with recurrent acute otitis media (AOM) or with OME of any duration is at increased risk for speech, language or learning problems because of baseline sensory, physical, cognitive or behavioural factors (Table 9.2).
- Use pneumatic otoscopy for diagnosing OME in a child.
- Obtain tympanometry measures in children with suspected OME for whom the diagnosis is uncertain after performing (or attempting) pneumatic otoscopy.
- Manage the child with OME who is not at risk with watchful waiting for three months from the date of effusion onset (if known) or three months from the date of diagnosis (if onset is unknown).

- Obtain an age-appropriate hearing test if OME persists for three months or longer or, alternatively, for OME of any duration in an at-risk child.
- Take note that the ENT will offer bilateral tympanostomy tube insertion to children with bilateral OME for three months or longer, and who also have documented hearing difficulties.
- Be aware that tympanostomy tube insertion should not be performed in children with recurrent AOM who do not have middle ear effusion (MEE) in either ear at the time of assessment for tube candidacy. Bilateral tympanostomy tube insertion should be offered to children with recurrent AOM who also have unilateral or bilateral MEE at the time of assessment for tube candidacy.
- Perform an adenoidectomy as an adjunct to tympanostomy tube insertion for children with symptoms directly related to the adenoids (adenoid infection or nasal obstruction) or, alternatively, in children aged four or older to potentially reduce future incidence of recurrent otitis media or the need for repeat tube insertion.

The procedure of insertion of VTs is generally very simple: under general anaesthetic, a small incision is made in the TM. The middle ear effusion is evacuated using suction. The particular VT chosen is then inserted into the defect created in the TM (Figure 9.1). Different types of VTs can be used, and they typically remain in the TM for varying periods before spontaneously extruding. Typically, one chooses a short duration of VT initially, and reserves longer duration tubes for recurrent/resistant cases. Very long-term/permanent tubes (for example, 'T-tubes') should be avoided in children.

Figure 9.1: (a)

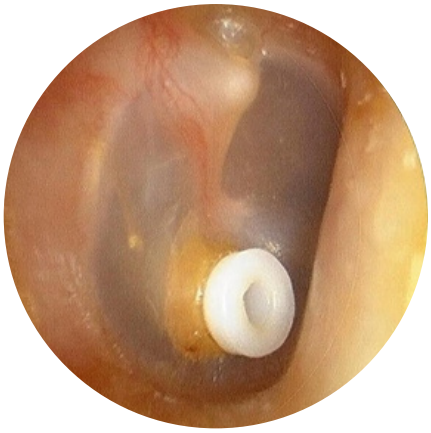


Figure 9.1: (b)



Figure 9.1: (a) ventilation tube in a right TM
Figure 9.1: (b) examples of ventilation tubes (with a paper clip shown for size reference) suitable for use in children include 'Reuter bobbins' – plastic and titanium – and 'Umbra Minor' tubes

The complications of VT insertion are mild and rare. Insertion may be followed by otorrhoea (9%) which is easily resolved with ear toilet and the use of antibiotic/steroid eardrops (for example, Cilodex®). The lumen of the tube may become obstructed with dried mucus or blood. The tubes may be extruded prematurely, and the OME may recur. Rarely, when the tubes are extruded, they may leave a perforation in the TM (2%) (Kay, Nelson and Rosenfeld 2001). It is recommended that the AAO-HNS guidelines be followed, and that the children receiving VTs be followed up six-monthly until extrusion of the VTs.

Adenoidectomy is often performed with VT insertion when there is reason to believe that enlargement or infection of the adenoids may be contributing to ET dysfunction, recurrent otitis media or obstruction of nasal breathing. The adenoidectomy procedure involves a general anaesthetic, the use of a curette to scrape out the adenoids (lymphoid tissue in the nasopharynx above the soft palate) and then visualisation with a mirror endoscope to see they are properly removed and to control bleeding. Adenoidectomy should not be performed on a child with a cleft palate as this may result in palatal incompetence.

Tympanoplasty/Myringoplasty

Correctly-speaking, the term 'tympanoplasty' means surgical repair of the tympanum – the entire middle ear, comprising the TM or 'myrinx'; the spaces of the middle ear, in particular the mesotympanum; and the ossicular chain. 'Myringoplasty' is the repair of the TM alone – but this term is seldom used. In common usage, 'tympanoplasty' is the term used for the repair of the TM. The indications for, and aspects of, tympanoplasty may therefore include any of the following:

- Closure of a perforation of the TM (myringoplasty).
- Repair/reinforcement of a retraction of the TM.
- Repair of the ossicular chain ('ossiculoplasty').
- Removal of disease from the middle ear (for example, cholesteatoma).

Myringoplasty

The procedure 'freshens' the edges and undersurface of the TM and places a graft underneath to support epithelial growth across the defect. Various graft materials have been used, with temporalis fascia, cartilage and perichondrium being most commonly favoured. The surgical techniques used vary too: The approach can be post-aural (behind the ear), end-aural (incision down the ear canal) or trans-canal (incision at the bottom of the ear canal). A microscope or endoscope can be used, and the graft material can be placed and secured in various ways. The site and size of the perforation may determine these choices by the surgeon.

The quoted success rate in the literature for closure of the TM varies significantly, from about 60% to 90%. Factors believed to influence the success rate of closure include the size and position of the perforation, the experience of the surgeon, the age of the patient and the graft material used (cartilage appears to have a slightly better success rate than fascia (Lajdam et al. 2023). There is much debate about a minimum age before performing tympanoplasty. While most surgeons would prefer to wait until six or eight years of age, this is not a hard and fast rule. Considerations affecting the decision include not only the age of the child but also the status of the other ear, whether there is a problem with recurrent otorrhoea, whether swimming is an issue, etc.

In terms of outcomes, successful closure of a perforation alone gives only a modest improvement in hearing – mean improvement of the order of 8 dB HL. Some residual CHL remains as the repaired TM is not normal. Nevertheless, this improvement can make a significant difference to the patient. There are furthermore the added benefits of having a closed eardrum with no/low risk of otorrhoea, and an ear which is more amenable to HA use. Myringoplasty also has a low complication rate.

Myringoplasty for repair/reinforcement of a retraction of the TM

Retraction of parts of, or the entire TM, with consequent loss of the sturdy fibrous layer and atelectasis of the TM, is believed to be the consequence of persistent ET dysfunction. It varies considerably in extent, from mild retraction of a portion of the TM to complete retraction of the entire TM ('adhesive otitis media'). There may be associated erosion of variable portions of the ossicular chain. The HL is also variable, from a conductive loss of greater than 30 dB HL if the ossicular is eroded, to no HL at all – as indeed may be the case even in an adhesive otitis media. There is no conclusive evidence that VT insertion can prevent or reverse TM retraction. Nevertheless, surgeons often insert VTs and thereafter monitor patients closely, provided that there is no ossicular erosion causing HL or TM retraction severe enough to raise concern for cholesteatoma development.

Reinforcement myringoplasty with excision of and/or support of the atelectatic portion of the TM is performed similarly to standard myringoplasty. Additional considerations would include attention to ET function, careful dissection of any adherent retracted TM off the ossicular chain or promontory of the middle ear, repair of the ossicular chain if eroded (ossiculoplasty, see below) and the use of cartilage as a sturdier graft material.

Retraction of the TM can be managed either by observation or by reinforcement of the TM with cartilage. Considerations in this decision include:

- Whether there is visible discontinuity of the ossicular chain
- Whether there is a CHL
- Whether the retraction looks as if it is progressing and is at risk of becoming cholesteatomatous

Ossiculoplasty

Defects of the ossicular chain can be congenital or acquired, and involve fixation, disarticulation due to trauma or discontinuity due to erosion. Ossicular chain defects causing CHL include:

Congenital: (rare)

- Fixation: Ossicular malformations
- Discontinuity: Ossicular malformations, congenital cholesteatoma

Acquired:

- Fixation: Tympanosclerosis (rarely affects the ossicles)
- Discontinuity
- Disarticulation: Head injury, surgical (rare, except when deliberate in cholesteatoma surgery)
- Erosion: Retraction of TM, chronic otitis media including cholesteatoma

The most common site of erosion of the ossicular chain is the delicate lenticular process of the incus, followed by erosion of the delicate stapes superstructure. Extensive cholesteatoma may cause osteitis and erosion of the more robust body of the incus and malleus. The principles of ossiculoplasty involve either re-establishment of the continuity of the ossicular chain or creation of an alternative conductive mechanism from the malleus handle or the TM to the stapes (superstructure (SSS) or footplate). The materials used in ossiculoplasty include the patient's own ossicles, homograft ossicles (ossicles from other human beings), cortical bone, cartilage or manufactured ossicular prostheses (currently usually made of Titanium) and, more recently, bone cement (for example, Ketac Cem®).

If erosion of the lenticular process of the incus occurs, the traditional approach has been the remodelling of the patient's incus to connect from either the malleus handle or the TM to the head of the stapes, or the use of a PORP (partial ossicular replacement prosthesis). A recent innovation is the use of bone cement capable of reconstructing absent bone, to reconstitute the long process/lenticular process of the incus (Demir et al. 2019). One such cement, Ketac Cem®, was created for dental work but is very successful and robust in this application. This adheres to the principle that reconstructing the long process of the incus produces the best results.

Surgical Repair as Part of Cholesteatoma Surgery

In cholesteatoma surgery, the principles of myringoplasty and ossiculoplasty, as described above, generally apply. The difference is that disease removal is a priority prior to the process of reconstruction. In such cases, an intact ossicular chain will need to be disarticulated. When the cholesteatoma is superficial to the various components of the ossicular chain, it may be possible to preserve the ossicularchain. . In this case movement and manipulation of the ossicular chain should be minimised to avoid sensorineural hearing loss (SNHL). If much manipulation is anticipated, it might be wise to temporarily disarticulate the incudo-stapedial joint prior to the operation. There is debate as to whether ossiculoplasty should be undertaken during the initial cholesteatoma surgery or be delayed until the second stage. However, most clinicians prefer to do this during the initial surgery, particularly in children. Reasonable improvement in hearing can be obtained. In a series of cases where reconstruction took place during primary surgery, an overall air bone gap improvement of at least 7 dB was obtained (Wilson, London and Shelton 2013; Eliçora et al. 2017).

Stapedioplasty (stapedectomy/stapedotomy)

Stapedioplasty (also known as stapedectomy/stapedotomy) is a relatively common procedure in adults, owing to the 0.3 – 0.4 prevalence of development of otosclerosis in early adulthood. It is very rarely indicated and performed in children because of the rarity of stapes fixation in the young. Stapedectomy/stapedotomy/stapedioplasty is an operation performed for a fixed stapes bone in the oval window (most commonly, otosclerosis). The crura (arms)/ superstructure of the stapes bone are carefully removed (often using a laser) and a small hole (stapedotomy) is made in the stapes footplate. A piston-like prosthesis is secured onto the long process of the incus so that movements of the ossicular chain are transmitted through the piston to the fluid of the inner ear.

Eustachian tube (ET) dilation

It is generally accepted that most middle ear problems originate from inadequate ventilation due to poor ET function, which is common in children as their ET is of smaller dimension and more exposed to the recurrent upper respiratory tract infections of childhood. The frequency of these problems diminishes with age and generally settles by age eight. Children with a

cleft palate and Down Syndrome, where the ET is negatively affected, have a far higher prevalence of middle ear problems.

Until recently the problem of inadequate ET ventilation of the middle ear was circumvented using ventilation tubes, and the ET was considered too inaccessible for intervention. There is now some evidence that balloon dilation of the ET may resolve middle ear problems attributed to ET insufficiency in a certain percentage of adult patients (Anand et al. 2019), although the improvement is only in the region of 60% (64% improvement in tympanometry), and many experts remain sceptical.

The concept of ET dilation in children is even more controversial, but there are some series showing subjective benefit and some significant objective improvement. Although there are no prospective randomised control trials, a systematic meta-analysis of the literature concluded that ET balloon dilation ± tympanostomy tube placement may produce outcomes that are comparable to tympanostomy tube placement in the treatment of recalcitrant OME in the paediatric population. Furthermore, the procedure was found to be safe, with the most common complication being epistaxis (nosebleed). Acclarent Inc. received clearance in December 2023 from the U.S. Food and Drug Administration (FDA) for use of their ET balloon dilation system in the treatment of children between the ages of eight and seventeen. Nevertheless, this more complex and uncertain procedure should be weighed against the far simpler one of myringotomy and placement of a ventilation tube.

Conclusion

Procedures on, or surgery to, the ear are often required to resolve CHL. If outer- or middle ear conditions leading to CHL are suspected in children, they should be referred to an ENT surgeon for further management. It is important that HHPs establish a referral system and collaborate with ENT surgeons to ensure successful management of children with CHL.

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Chapter 10

Paediatric Amplification: Non-Implantable Devices

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'The miracle of hearing includes that human ears are uniquely tailored to pick up and enhance the sound of a human voice.'

~Bagatto et al. (2023)

Introduction

Children are born with the need and the ability to communicate (either through spoken language or through visual language). This chapter focuses on hearing aid fitting (HAF) to promote optimal auditory access for spoken language development. Measuring the ear canal's response to sound (in sound pressure level [SPL]), referred to as the Real Ear Unaided Response (REUR), has made it clear to researchers that the human ear, through the outer ear canal and the concha, naturally amplifies sounds within the 3 kHz to 4.5 kHz range, which encompasses critical frequencies for speech perception (Bagatto and Scollie 2023).

A hearing healthcare professional (HHP) should always aim to identify hearing loss (HL) as early as possible and, if the parents choose this option, to fit hearing aids (HAs) very soon thereafter. As a mother once said, after an early inconclusive hearing test, two weeks of waiting for the follow-up test might not seem like much to the HHP, but for her baby it is precious time lost. Every passing day matters. It is critical not to delay diagnosing and fitting HAs. Appropriate technology as part of early intervention can change the course of a child's entire life.

Paediatric Hearing Aid Candidacy

The main aim of amplification for children should be to provide audibility of the speech signal, to ensure that the child has access to speech and to allow the child to participate in conversations. With these aims in mind, amplification should be considered for any degree of HL, from slight to severe. There are many factors that will guide and inform HAF, and the management of the process should always be a team effort. The child's parents and family, the paediatrician, ear-, nose- and throat (ENT) specialist, speech therapist, teacher and others form part of this team and should be integrally involved in the selecting and fitting of the HAs (Chapter 14). Managing HL and HAF should be an ongoing process that should be closely monitored to accommodate all variabilities along the way:

- Are there any hidden co-morbidities that were initially missed?
- Is language and speech developing as it should?
- Is the child's environment changing and should the HA or amplification be adapted to accommodate these changes?

All of these questions and more should be asked at the beginning of and throughout the process of selecting HAs. The answers will guide the selection and inform the fitting of HAs. The underlying cause of the HL should be identified where possible; this could give an indication of whether the HL is progressive or not. Even if the aetiology is uncertain, ongoing monitoring to assess progressive HL is essential (Chapter 2). An infant or child with a severe to profound HL might not wear HAs long term as they may be candidates for cochlear implants (CI). Whereas some children with obvious middle ear pathologies (atresia, for example) might enter a bone conduction hearing device (BCHD) programme at once (Chapter 11), most of the paediatric population with HL will be long-term HA wearers.

There are a few important characteristics of the paediatric population that will guide every part of the HA process, from candidacy to verification and fitting of the HAs. These include:

- The etiology of the HL
- The size of the child's ear canal (external meatus)
- The nature of HA sound processing
- The changing communication needs of the developing child
- The sound environment of the infant and child
- The unique and evolving needs of the moving, learning child

Candidacy criteria

Infants and children with HL greater than 30 dB HL across the frequencies of 0.5, 2 or 4 kHz in any ear are considered candidates for amplification (Bagatto and Scollie 2023). Considerations for unilateral and bilateral hearing loss (BHL), as well as for different types and configurations of HL, will be discussed.

a) Unilateral hearing loss (UHL)

Without access to sounds from both ears, children miss binaural cues important for localisation and hearing in background noise. Binaural hearing is important in communication and provides advantages especially in coping with background noise. A child with a UHL is at greater risk for academic failure and difficulty in social situations (McKay, Gravel and Tharpe 2008). Recently, studies have also focused on listening-related fatigue as a contributor to difficulties experienced by children with UHL. School-aged children with UHL are at risk of the same level of fatigue experienced by children with bilateral moderate degrees of HL (Bess et al. 2020). In the case of infants with UHL the same principles of early intervention as with BHL apply. The etiology of the HL will give important clues about the nature of the HL and whether it might be progressive or associated with other co-morbidities. Children with mild-to-severe UHL are candidates for a HA on the affected side. Ongoing monitoring of UHL is critical.

Mild UHL in children, though often overlooked, can significantly impact auditory development and classroom functioning. Bess et al. (1998) found that children with minimal or unilateral sensorineural hearing loss (SNHL) frequently demonstrate reduced speech perception in noise, poorer academic performance and increased listening effort, indicating that such losses are not benign. Bagatto et al. (2019) and Scollie (2005) emphasise that even mild losses can compromise access to soft speech sounds, particularly in complex listening environments. Clinical management should include amplification when appropriate, remote microphone (RM) systems and functional assessments to monitor the child's auditory access and communication development over time.

If the UHL is moderate to severe, a HA is the first choice for amplification and all the same principles as with bilateral HAFs will apply in terms of the selection and fitting process. In the case of a profound UHL or Limited Usable Hearing Unilaterally (LUHU), an HA might not be an option. A contralateral routing of signal (CROS) system might be recommended, where a

microphone is encased in a behind-the-ear (BTE) style HA casing on the side with HL and a receiver on the other side. Sounds are then transmitted wirelessly to the normal hearing ear. CROS systems are recommended, and widely used, as a treatment option for LUHU in adults. There is conflicting evidence of benefit using this option for children. In a quiet environment, speech will be transferred from the side with HL to the normal hearing ear. In the presence of environmental noise, however, interfering noise will also be transmitted to the hearing ear. A CROS system is also unlikely to aid in localisation of sound. This system will require the ability to manage the device and the listening environment and is therefore not recommended for infants and young children. As soon as the child reaches an age where they can trial the CROS system and are able to manage the device, a CROS trial can be considered. This is typically around five to seven years of age, depending on developmental, cognitive and language abilities. The occlusion of the small normal hearing ear of the infant or young child is another risk when fitting a CROS system. An important benefit of binaural hearing is the ability to hear in background noise and a child with LUHU will therefore need additional support in noisy environments such as classrooms. An RM system is the best option for this situation (Oosthuizen et al. 2021).

b) Bilateral hearing loss (BHL)

In the case of mild BHL, the effect of ear canal acoustics on the hearing thresholds of the child should be considered when using the audiogram as the standard for candidacy for mild HL in the paediatric population. Ear canal pressure (ear canal SPL) is higher in a smaller ear than it would be in a bigger ear. This is important to keep in mind since the dial reading of the audiometer (using dB HL) is calibrated using the 2-cc coupler that typically has a lower ear canal SPL than those of infants and young children (using inserts of ear moulds). This difference is called the real-ear-to-dial difference. When the ear canal acoustics are taken into consideration, children may have poorer hearing thresholds than shown on the dB HL audiogram. Another potential negative consequence of this is that the child's ear canal grows over time and the sound pressure in the ear canal decreases. As a result, the hearing thresholds (in dB HL) will seem worse even if the child's auditory function does not change. It might be perceived as a progressive HL, when in fact it is just the result of a growing ear canal. Considering the effect of ear canal acoustics on the audiometric thresholds of infants and young children is important when making decisions about HA candidacy (using the Real Ear to Coupler Difference [RECD] measurement). RECD is also applied during the fitting process to consider the effect of the individual ear canal acoustics with amplification. RECDs will

be covered in the next section of the chapter. Using RECDs to convert dB HL to dB SPL and then using the speech intelligibility index (SII) (American National Standards Institute, 1997) is the recommended approach when applying candidacy criteria for mild HLs. The SII is used to quantify speech audibility either with an HA (aided SII) or without (unaided SII). The SII accounts for the child's ear canal SPL and estimates the audibility of speech sounds and the impact of HL on communication.

Another consideration when deciding to fit a child with mild BHL with HAs is the ability to provide a vent in the ear mould. An occluded ear canal will not allow sound to escape in the low frequency region and might provide sound where there is no need for HA gain. The upward spread of masking, a phenomenon where a lower frequency sound spills over and masks the higher frequency sound (Bess and Tharpe 1986) should be considered. When using an HA, this happens when low-frequency amplification spills over and masks high-frequency speech sounds. Instead of benefiting the child with HL, it will degrade the speech signal, especially in noisy environments. One way of ensuring that the low frequencies do not interfere in this way is through venting the ear mould sufficiently, which is something that might not be possible with the very small ear canal of an infant or small child. Figure 10.1 from Bagatto and Tharpe (2014) can be used to guide the decision on whether to fit the child with a mild bilateral or unilateral HL with HAs.

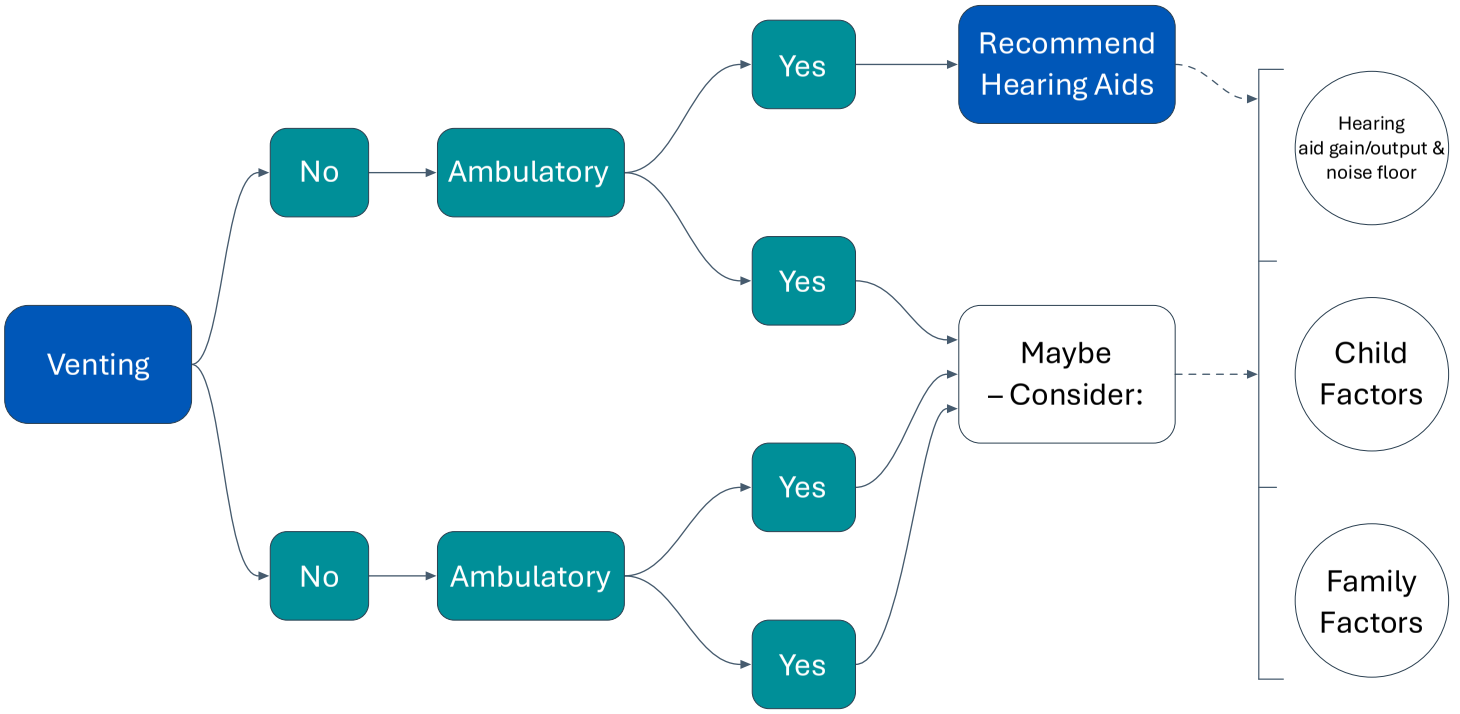

Figure 10.1: Decision support guide for HHPs considering HAs for children with mild BHL. Source: Bagatto and Tharpe (2014)

The term ‘ambulatory’ in this figure refers to the mobility of the child, whether the child is crawling, walking, stationary on the mother’s lap or in her arms. Speech intelligibility is negatively affected when the child moves away from the speaker and the signal-to-noise ratio (SNR) diminishes as the distance from the speaker increases. That should also be taken into consideration when deciding to fit a child with mild bilateral HL with HAs.

In the case of moderate-to-severe BHL, there is no doubt that a child with this degree of HL should be fitted with HAs as soon as possible. Children with severe-to-profound HL will potentially be candidates for HAs even though the HL might be better suited for a CI. In the case of babies and small children, several factors should be considered when fitting a severe-to-profound HL with HA. HA feedback, due to high gain levels and increased ear canal SPL, might be a problem. Depending on the severity of the HL, the aided SII might be too low and therefore not provide enough speech sound audibility, which is critical for language development. It is recommended that these children be fitted with HAs as part of the CI evaluation in conjunction with the CI team (Chapter 11). Candidacy for HAs for children with mild and profound HL will be considered in more detail in the following section.

Types and configuration of hearing loss

In the case of certain types of conductive hearing loss (CHL), such as atresia, an air conduction HA is not an option; rather, the child will be a candidate for implantable bone conduction devices. On the other hand, some types of CHL in children (such as otitis media) can be treated and the ENT surgeon plays a crucial role in diagnosing and treating these pathologies (Chapter 9). However, there are also types of CHL that, despite their apparent ‘treatable nature’, persist and leave the child with a semi-permanent HL. Any child with an HL that could negatively impact their ability to learn language and develop speech or to perform well in the classroom should be considered a candidate for HAs. A recent study (Pienaar 2021) showed that HAs are beneficial for children with CHL. Another example of a CHL for which HAs can be fitted while awaiting surgery is with a tympanic membrane (TM) perforation. It is not uncommon for a child to be left with a perforation in the TM after insertion of grommets (Vlastarakos et al. 2007). Surgery is not always an immediate option after a TM perforation, and it is therefore often associated with HL. HAF is a viable option for providing the necessary amplification in the interim. Infants and young children with SNHL are candidates for air conduction HAs. These infants and young children will show clinical evidence of impaired outer hair cells (such as absent otoacoustic emissions [OAEs]).

Auditory neuropathy spectrum disorder (ANSD) is more complex to manage than typical SNHL and the prognosis for spoken language development is uncertain. Conventional guidelines for SNHL management do not apply to ANSD. As behavioural testing in young children is often unreliable, management of ANSD in the paediatric population is likewise difficult. Controversy still surrounds the recommended management of young children with ANSD, since the benefits of HAs in aiding the development of speech and language are unclear for this population. On the one hand, the HA may be providing amplification to a system that does not require it, and on the other hand the HA may be amplifying sounds that remain unclear and therefore meaningless to the child.

Spectral and temporal processing deficits are associated with ANSD and amplification through HAs cannot overcome these deficits. CI constitutes the major rehabilitative intervention that allows for speech perception in patients with bilateral ANSD who do not benefit from conventional amplification. The clinical HHP is faced with these questions: 1) Should HAs be offered to infants with ANSD? and 2) How long should HAs be trialed before deciding that the outcomes are not appropriate? While HA trials are warranted, children with ANSD must be tracked closely to avoid delays in decision making.

Key Considerations in Hearing Aid Selection

When deciding on the HAs features for children, it is essential to consider four key factors that influence the decision-making process.

- First, the nature and severity of the child's HL must be carefully evaluated.
- Second, their sound environment plays a significant role, including exposure to noise, whispering, sports and music.
- Third, the unique aspects of their developing brains and bodies should be considered, as growing auditory systems require a clearer signal and factors such as ear size, acoustics and other health conditions can impact HA selection.
- Lastly, the family context, including support systems and communication needs is critical in determining the most appropriate solution.

These factors will guide the HHP's decisions about HA features essential for infants and children. Each factor should be considered when making decisions about specific HA selection, to ensure that every aspect of the child's unique hearing needs is considered.

The nature of childhood hearing loss

HL in children differs from adults in meaningful ways. HL in children is more severe, more asymmetrical and more progressive than that of adults (Pittman and Stelmachowicz 2003). Once HL has been confirmed, the HAs chosen should be flexible in terms of gain, output and configuration to accommodate any changes in hearing.

Children's sound environment

a) Noise and reverberant spaces

Once the infant with HL leaves the mother's arms and starts to move, noise will become one of the main challenges when choosing HAs. Children's schools and social engagement are characterised by noise and reverberant spaces. Noise management in HAs is used as an umbrella term for noise reduction, multiple programmes and directional strategies. The concern for HHPs is that noise management in HAs might take away from the speech signal or degrade the speech signal. Over the past decade noise management systems in HAs have evolved and there is good evidence that children benefit from new technology, adaptive gain and noise reduction systems (Pittman and Stewart 2023).

In a recent study investigating the data gathered from 19,201 paediatric HAFs (newborn babies up to children eighteen years of age) through the data logging systems in the HAs, it was found that wearing time of HAs increased with higher levels of technology (Pelosi 2024). Higher levels of technology employ more noise management systems, which makes HAs more comfortable to wear in noisy situations and as a result daily wearing time increases. Directionality is another way of improving SNR for managing background noise. Traditionally, the HHP had to choose the HA programme to receive sounds equally loud from all directions or louder from the front. Directional HA microphones, developed to minimise the negative effect of background noise, however, assumes that the child faces the speaker. Children have a low rate of orienting their face to the speaker (Scollie et al. 2010) and localisation only matures from age six onwards. Toddlers, once they start to crawl, move away from the speaker and overhearing across all directions is important in the early years. Traditionally, omnidirectional mode was the preferred choice for young children's HAF. Adaptive directionality in newer technology instruments is a beneficial strategy to leverage the benefits of both omni- and directional HA features (McCreery et al. 2012).

As soon as toddlers start to move away from their parents or enter noisy educational environments, they will be candidates for assistive listening devices (ALDs), in addition to their personal HAs. ALDs, such as RM systems, have been successful in improving listening environments with poor SNRs, overcoming the negative effect of reverberant rooms and compensating for substantial distances between the child and the teacher or caregiver. The RM system makes use of a remote microphone attached to the speaker, connecting wirelessly to the HA of the child. Even if the child is far away, the speaker's voice will be perceived to be close by, overcoming the background noise, distance and negative effect of the echoes in the reverberant room.

b) Sports

Other sound characteristics of children's environments include their participation in sports. Moisture management systems and HA durability should be diligently considered as well as the distance and reverberation that inevitably play a role in these circumstances. HA compatibility with ALDs and timeous introduction of these systems are important. As with noise, an RM system with a remote microphone attached to the sports coach, for instance could aid participation for the child with HAs. Children who participate in sports also sweat and are exposed to dust, which needs to be considered when choosing HAs.

c) Music

Music training for children with HL should be encouraged as a complementary means of habilitation to improve language and listening outcomes (Lo et al. 2020). HA signal processing for live music and music performance should support music lessons.

Children's brains and bodies

a) Developing speech and language

Since children's auditory systems are still developing, HAs should provide a clear signal with more bandwidth, a better SNR and a higher SII than HAs for adults. Children's speech and language abilities are developing, and access to all speech sounds should be the goal of amplification. Children might not have access to some high-frequency speech sounds, such as /s/ or /sh/ because of HL severity or feedback issues, due to small ear canal sizes. Certain

signal processing strategies aimed at lowering the inaudible frequencies to provide access to these high frequency sounds should be considered. This technology is evolving, has been shown to be efficient and should be implemented when fitting HAs to infants and children (McCreery et al. 2012).

b) Ear canal size

Ear size affects acoustics. When prescribing HA gain and other electro-acoustic measurements for the infant and child's hearing, an individualised prescription should be used, which considers the age and HL of the child. Many infants are born with sufficiently developed pinnae and ear canals to support an HA connected to a custom ear mould. BTE HAs are the appropriate choice for most infants and young children. The pinnae will only be fully grown by the age of ten, and the concha will show rapid growth in the first two years of life. Regular ear mould changes are essential to ensure well fitted HAs. Venting the ear mould will not be possible with a very small ear canal. Soft material (silicon or vinyl) are safe options compared to hard material, such as acrylic. Due to the rapid growth changes and resultant ear mould changes, certain HA types such as in-the-ear or canal (ITE/ ITC) are not practical options for infants and young children. Custom products are also more prone to feedback due to the proximity of the receiver and microphone. Receiver-in-the-canal (RIC) HAs look like other BTE HAs but have the receiver in the ear canal instead of being housed in the HA casing. These HAs are less secure than BTE HAs as the receiver can easily be blocked with wax, needing a daily wax check. For this reason, they are not suitable for discharging ears. The RIC type aids can be considered as an option for older children and teenagers.

Children's family contexts

Supporting the caregiver means supporting the child. Apart from the emotional toll of the HL and possible other disabilities of the child, caregivers will also experience practical barriers. Besides the cost of the devices often being very high further ongoing expenses might be required, such as insurance (in case of breakage or loss), challenges with HA retention, regular ear mould remakes and appointment scheduling. The HHP might need to assist the parents in securing funding, or choose HAs that are less costly, without compromising essential features (Chapter 15). If the child has any other disabilities, inter-professional collaboration will be needed. In that case,the HHP will need to support the caregiver to ensure collaboration. At various points in the journey, caregivers will need additional support in connecting them

to the necessary professionals and other parents of hearing-impaired children and in transitioning the child from childcare to school. The use of ALDs such as RM systems will require training and support to the caregiver and teacher. Some practicalities might seem trivial to the HHP but might be a barrier for the caregiver. For instance, rechargeable HAs might be a safer option for the young child (since loose batteries could be swallowed), but if the family does not have constant and good sources of electricity, the HAs will not be optimally charged. Rechargeable HAs might also be more expensive. Finally, caregivers' expectations should be realistic and should be supported to ensure that the quality of the language input is sufficient for optimal outcomes for the child. Regular contact with the caregiver is of utmost importance. This contact might be through cellphone apps, messaging, calls or visits. The HAF is just the start of the process and not the end goal.

Assistive Listening Devices (ALD)

The most widely used of the ALDs are RM systems. These systems use radio frequency signals to transmit sound directly from a source (like a teacher's microphone) to the child's receiver. The speaker's voice is carried to the child's HAs or CI. Because of the proximity of the speaker's mouth to the microphone, the distance between the teacher and child or the child and parent is minimised. RM systems effectively lower the signal to noise ratio, overcoming distance and reverberation. They are often used in classrooms to help children with HL hear their teachers.

Newer technology HAs offer Bluetooth connectivity and Bluetooth streamers can be used as an alternative to RM systems. Bluetooth capability has also opened the doors for other systems that might be helpful. Smartphone connectivity facilitates the use of applications for listening activities or even text messaging and other communication aids. Activities should be age appropriate, and their usefulness should be investigated by the HHP, the child's family and other team members.

Other types of ALDs include:

- Infrared systems used with the television
- Sound Field Systems that use strategically placed speakers in a classroom to enhance the speech signal for all students, including those with auditory processing challenges or mild to moderate HL

- Loop systems that utilise an electrical current in a wire loop to transmit sound directly to HAs or cochlear implants that have a 'telecoil' feature
- Flashing and vibrating alarms
- Telephones that make sounds louder

Fitting of hearing aids

Electro-acoustic requirements: Fitting to target

The single most important refrain in the HHP's mind when fitting HAs to infants and young children should be:

In the context of early intervention, infants will wear their HAs at fixed, clinician-determined settings for months or years before they are able to clearly express their preferences. Therefore, the validity of the clinician-determined HA settings may be of crucial importance for auditory development in the early years of life. (Scollie 2005).

A child is not just a small adult. When HHPs fit HAs on children, they must use all the tools available to them to ensure that the child will be able to hear speech. A child needs to receive sound consistently and at safe and comfortable levels to hear speech and learn language. Consensus in the field is that HA prescription should be done in an objective way (Bagatto et al. 2023; Pittman and Stewart 2023). A prescriptive fitting formula provides systematic and consistent fitting that ensures that speech is audible and comfortable for children at different input levels. This is done by calculating the amount of amplification that is needed to ensure audibility of the incoming signal. A specific amount of gain is then prescribed per frequency, and this is individualised for each child. Other electro-acoustic parameters, such as output, maximum power output (MPO) and compression ratios, are also prescribed. HA performance is then measured against this target.

There are two methods specifically suited and most widely used as the algorithms of choice when fitting children. These are the Desired Sensation Level (DSL) v5.0a (Scollie. 2005) and the National Acoustic Laboratories (NAL) Non-Linear 2 (Byrne et al. 2001). Numerous studies have shown that school-age children with HAFs that approximate the targets of these two prescriptions achieved high levels of aided speech understanding, audibility and loudness comfort over a wide range of input levels (Pittman and Stewart 2023). These formulas are

preferred above the HA manufacturers' algorithms because they are grounded in empirical evidence specific to the paediatric population. These prescriptions consider the child's hearing thresholds, age, language (tonal or non-tonal) and ear canal size and adjust the target values based on these variables.

DSL v 5.0a or the NAL-NL 2 formula

There are several differences between the two strategies. The most significant difference is that the DSL prescribes more gain than NAL for soft speech for children with the same HL. NAL provides more comfort in noise through higher compression thresholds than DSL. Different studies have shown that children (when they are old enough to express a preference), prefer the DSL for quiet speech and the NAL for noisy situations. Despite these differences, children's early speech and language outcomes and the ability to perceive conversational speech are similar for the two prescriptions (Ching et al. 2013; Scollie et al. 2010). Research indicates that while children can discern differences between the DSL and NAL prescriptions, these variations have minimal effect on overall communication outcomes. That leaves the HHP with the choice to use either of these prescriptions for optimal outcomes.

As hearing instrument technology advances, HHPs should base their decision on a combination of all factors. For instance, if it is available and the child's HAs have adaptive situational gain (ASG) (currently available in higher technology levels that are typically also more expensive), more audibility for soft level inputs can be obtained through ASG. In this case, the HHP might opt to use the NAL formula since the additional input that the DSL might add at soft levels will be achieved through the HA processing (Pittman and Stewart 2023). If the infant has a moderate-to-severe HL and maximum audibility of speech is the aim, then DSL is the formula of choice. The SII values, a numerical estimate of audibility across the frequency range of speech can be used to quantify the audibility of speech. Frequencies are weighted based on their contribution to the average speech; the Long-Term Average Speech Spectrum (LTASS) and percentage value is calculated with 100% representing complete audibility (ANSI 1997). If fitting to the NAL target results in a low SII value, the DSL target should be used to ensure audibility of speech.

Whichever of these two prescriptions the HHP chooses, the fitting should be verified at different input levels and frequent follow-up visits and feedback from the caregivers should be gathered to make sure that speech and language development is optimal.

Rationale for objective verification of the HAF

Prescriptions are defined electro-acoustically (gain, output, compression ratios) and should be verified using specific electro-acoustic measures to ensure that the prescription is achieved (Scollie 2005). Aided sound field measures have previously been used to verify HA settings, but their usefulness is limited to counseling and proving benefit. The electro-acoustic parameters that need to be verified are optimum gain at different inputs and across the frequency spectrum, and the maximum power output of the HA to ensure that it is not too low (which causes distortion) or too high (which causes discomfort). If it is found that audibility in the high frequencies is too low, frequency lowering is recommended, which should be verified using a 's' and 'sh' signal.

During the HAF process, modern HAs are connected to the manufacturer's software directly or wirelessly. The software recommends electro-acoustic settings based on the child's age and HL. These pre-fitting recommendations provide a good starting point for the fitting. It is not uncommon, however, to find discrepancies between the actual performance of the HAs and the prescriptive targets for gain and output during verification measures. Without verifying the HA performance in the child's own ear, the HHP cannot assume that speech will be audible and loud sounds comfortable. It is crucial to account for the increase in real-ear SPL when fitting HAs to infants and young children (Stelmachowicz 2000). The best way to do this is to obtain real-ear measures of gain and output using a probe microphone system. Performing traditional probe-microphone techniques, where the child must sit still for the insertion of the probe in the ear canal, and remain quiet and still with the head in an upright forward-facing direction for a prolonged time, is not always possible with younger children. Therefore, simulated or coupler-based real-ear measures are the verification method of choice because infants and young children cannot sit quietly facing a loudspeaker for the duration of real-ear verification. Furthermore, a controlled test-box environment is an accurate and reliable way to measure the RECD.

The RECD procedure requires minimal co-operation from the child and can be performed in less than five minutes. All subsequent measures can be performed in a test box and the RECD can be applied to predict real-ear gain and output. New RECD measures should be obtained each time the ear mould is replaced. The HA must then be adjusted to best match the targets to ensure optimum audibility of the speech signal. HA performance across input levels in the range of 55-75 dB SPL should be conducted to determine the audibility

and compression characteristics of the HAs. This should be done using a calibrated pre-recorded speech signal. Equipment often incorporates equipment-specific speech passages as a signal for measurements (such as the 'carrot' passage in the Audioscan equipment). This kind of signal, however, incorporates only one language. An internationally used, standardised speech signal that is available in most probe microphone measurement equipment is the International Speech Test Signal (ISTS). It includes the relevant properties of speech and allows for reproducible measurement conditions (Jorgensen and Novak 2022). The languages that were incorporated in the development of this signal is American English, Arabic, Chinese, French, German and Spanish (Holube et al. 2010). The ISTS is shaped by LTASS targets, which are calculated using the prescriptive target (NAL or DSL). This is the signal of choice when verifying HA function for children.

If frequency lowering is employed in the HA, verification procedures should be included for this technology. Frequency lowering can potentially lead to spectral overlap and perceptual confusion of /s/ and /sh/. Using calibrated /s/ and /sh/ sounds, the audibility of the sound with amplification can be determined. If the upper limit of the curve is above the threshold at the particular frequency, the /s/ sound is audible, if not, frequency lowering is activated and adjusted to ensure that the sound is audible, but not overlapping the /sh/ sound. The maximum output of the hearing instrument must be verified to ensure that it does not exceed uncomfortable levels and is not set too low, causing distortion. This is done by using a high intensity signal of 90 dB SPL, such as a broadband stimulus.

Frequent visits to the HHP for HA adjustments are an important part of the infant's ongoing habilitation (every two to four months) to re-assess thresholds and adjust RECDs.

A Step-By-Step Guide to Fitting Hearing Aids

Once the candidacy for HAs has been established (through diagnostic tests and evaluations by the ENT specialist and other relevant team members) and the HAs have been selected, the HHP should immediately proceed with taking ear mould impressions.

Procedure for obtaining ear impressions

The prescription for the ear moulds should include the length of the canal and helix, material (silicon, etc.), tubing type and vent (if possible). The child's ear moulds should be made of

soft material for comfort, safety and retention. BTEs are the HAs of choice for infants and younger children. For older children, RIC HAs can be considered and acrylic tips might be an option, depending on the degree and configuration of the HL. Impressions will also be needed for these tips.

1. It is important to start with an explanation of the process of taking ear mould impressions for the parents. If it is an older child, illustrations might be helpful. Rapport should be built and the child put at ease. If it is an infant, the caregiver could be encouraged to feed the baby to maintain calm. If the child should get upset during the procedure, it might be helpful if the parent holds the child gently but firmly with one ear against their chest. It is important to discuss the expected duration of this support and suggest distraction techniques to keep the child calm until the process is complete. An otoscope, ear light, otoblock, distraction toys, impression material and the syringes should be ready and available.
2. It is also important to adhere to hygiene protocols, like washing hands thoroughly or using gloves. Wearing a mask is compulsory. It is furthermore necessary to keep talking to engage the child.
3. An otoscopic examination should be conducted. The ear canal should be examined to ensure there are no conditions, such as discharge or excessive cerumen, that would prevent taking an ear mould impression. The size and length of the ear canal should be assessed. To demonstrate the process to older children, it is often helpful if they inspect their parent's ear (while the HHP holds the otoscope) or an ear model, if one is available. The HHP could furthermore shine the light of the otoscope on the child's hand.
4. Before the ear light is used, it should be marked to ensure that it is inserted at a safe depth. The guidelines are ten mm from ear canal entrance for children younger than six months and ten-to-fifteen mm from entrance for children older than six months.
5. The otoblock should be inserted using the marked ear light. For this part of the procedure, the mother or caregiver should gently press the child's head against their chest with one hand against the child's cheek (Figure 10.2). For young children, the standard sized otoblock could be halved by pinching off a piece. The otoblock should be gently placed into the ear canal until the mark aligns with the canal entrance. The depth and position of the otoblock should be verified with an otoscope and a string wrapped over the ear to secure it. The same should be

repeated for the other ear if it is a bilateral fitting.

6. With both otoblocks secured, the impression material should be mixed in the amounts specified by the product directions. Once mixed through, the material should be loaded into the syringe, ensuring it is ready for application. For an older child the material could be mixed for them to see and a small amount of material could even be handed to the child to mix and roll, although caution should be taken that it is not swallowed.
7. The child's head should be softly secured and the loaded syringe positioned in the ear canal. It is important not to pull the ear or press against the ear as it will alter its shape and may cause an inaccurate impression. The canal should then be filled by slowly depressing the plunger. The syringe should not be pulled out, but should rather slowly be pushed out by the syringe: The canal should be filled first and then the concha. Inject the material into the angle of the helix using the syringe. The tragus should be gently pressed to confirm it is adequately filled. Gentle jaw movements during the process by sucking or drinking are encouraged. This stepwise procedure should be repeated on the other side if it is a bilateral fitting. It is important to ensure that one does not press against the impression material, changing its shape. It might be necessary to wait until the first impression has hardened before moving on to the other ear.
8. The material is set when pressing a fingernail into it leaves no indentation.
9. The pinna should be gently pulled to loosen the impression. The bottom part of the impression should be lifted by the HHP, using a finger to break the seal, and gently pulled from the canal, concha and then the helix. It is important to ensure that the otoblock has been removed along with the impression material. Otoscopy should be conducted after removal, to ensure that no impression material has been left in the ear canal.
10. If there are no holes or bubbles and the canal seems long enough, the impression can be sent to the manufacturer.
11. Ear moulds should be replaced regularly (Figure 10.3). For the infant this means every two months. As the child grows older, the caregiver should regularly assess the fitting. If feedback occurs or the ear moulds visibly do not fit, they will need replacing. It is important to communicate this with caregivers from the start.



Figure 10.2: Taking the ear impression while the infant is being fed

Procedure for pre-programming hearing aids

Most new technology HAs are connected wirelessly to the manufacturer's software. Choose a manufacturer that supports paediatric fittings through dedicated research and whose HA software includes the DSL or NAL prescriptive target. These HAs will also have default settings based on the age of the child, incorporating changes in the ear canal, and will suggest frequency lowering if possible. If you are unsure, contact the company representative and learn as much as possible about the paediatric settings of the HAs you choose.

The process of pre-programming includes:

1. Once the HAs are received, and before the fitting appointment, the adult size tone hook should be replaced with the paediatric size tone hook. These smaller tone hooks come in a variety of colours. The caregivers should be encouraged to let the child choose the HA and tone hook colours to increase adoption rate and retention.



Figure 10.3: BTE HA with silicon ear mould.

2. All fields should be completed with appropriate software, for example, age, audiogram, etc. (with as much information as possible).
3. DSL or NAL software algorithms should be chosen. Generally, the DSL prescription would be the prescription of choice for infants and young children with moderate to severe HL or for progressive HL. NAL could be considered for milder HL, older children in noisy school surroundings or if the HHP gets feedback that it might be too noisy, and retention becomes a challenge. Outcome measures and feedback from the caregivers are crucial to guiding the decision.
4. The manufacturer's default setting should be used as a starting point for the fitting (including activation of noise reduction systems and frequency lowering).
5. The HHP should make sure that the volume control on the HA is deactivated. Access to these features is possible nowadays through HA applications for most smartphones. That way the caregiver has control over the volume settings, if needed.
6. The settings should be saved and the HAs should now be ready for verification at the fitting appointment.

Procedure for fitting the hearing aid in the child's ear and performing the feedback test

On the day of the fitting, a two-hour session should be scheduled with the caregivers and child. If the HHP does not schedule enough time, anxiety might be created for both the HHP and the caregivers should something unexpected happen (a crying infant, programming glitches, tears, etc.). A story picture or written information sent ahead might be helpful to prepare caregivers for all the steps in the process.

1. The ear mould should be placed in the child's ear to ensure that it fits well, and to cut the tube to the correct length. Even though the first fit is part of the preparation for the fitting and not for the purposes of instructions to the parents yet, the HHP can talk through the insertion of the mould. Repetition is key. Insertion in the ear canal should be demonstrated. The HA should fit snugly behind the ear. After the correct length of the tube is marked, the HA can be attached to the ear mould.
2. The HAs should be connected to the software and the instruments muted.
3. The HHP should ensure that the child is comfortable and occupied (or feeding) and should then insert the HAs in both ears. Due to the small ear canal size and increased ear canal SPL, HA feedback often occurs with paediatric HAFs. The child should be kept as quiet as possible and the feedback test for both ears should be run through the manufacturer's software. Often these loud sounds cause the infant or young child to cry, so it may be helpful to keep a distractor close by. The feedback test requires a quiet baby. If the baby is asleep, they can be kept in a sleeping state for as long as possible.
4. The HAs should then be removed and prepared for the RECD measurement.

Procedure for verification of the hearing aid fitting

Verifying the match to prescribed electro-acoustic performance is the next essential step in a scientific approach to paediatric HAF. This should be done using RECD measurements and the HA test box coupled to the HA-2 coupler (Figure 10.4). RECD and the coupler is the verification method of choice for infants and young children. Once the child can sit upright facing the speaker for as long as it takes to verify the fitting and programme the HAs, REMs are possible. This is usually after the age of six. If it is not possible to measure RECDs (the child cries too much or moves, for instance) age-related predicted RECD values can be used. These can be incorporated into the probe microphone system and can be chosen as an option. If RECDs have been measured previously (recently), it can be entered as values and the HHP will not need to measure it again. RECD values from the one ear can be used for the other side as well, if there are no obvious significant differences in the anatomy of the ears. RECDs should be remeasured regularly as the child's ears grow continually. RECDs should also be remeasured every time the ear moulds are replaced. Instructions on how to perform the RECD is equipment specific.

Procedure for measuring the RECD

1. The coupler microphone of the equipment should be connected to the transducer to measure the coupler. The coupler should be connected to the coupler microphone of the equipment and the transducer should be coupled to the other end of the coupler.
2. In equipment software, the HHP should select the option to measure the coupler. A broadband or sweep frequency, generated by the system, is delivered to the coupler and its response is measured by the microphone.

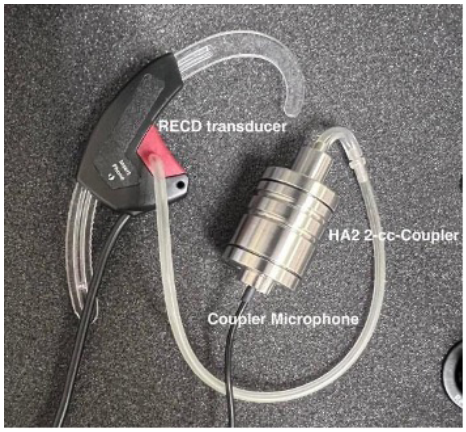

Figure 10.4: RECD equipment

3. Following this measurement, the real ear SPL of the child should be measured. The coupler and ear coupling parameters should be entered correctly into the verification system so that accurate conversions can be applied. The same stimulus should be presented and the real ear response measured and used to obtain the RECD values.
4. For the RECD measurement, a probe tube is inserted in the ear canal (Figure 10.5) together with either the ear mould or the insert foam tip, coupled to the transducer. The probe tube and ear mould or tip should be inserted into the child's ear canal. Distraction techniques can be used to draw the child's attention away from the procedure. The probe should be placed so that it extends two-to-four mm past the tip of the ear mould or foam tip. After six months corrected age, the

probe tube should extend five mm past the probe tube. If the tube is not inserted deep enough into the ear canal, it can lead to inaccurate results, especially at the high frequencies. Ear mould lubricant can be used for easier insertion. The probe tube can be secured to the tip with plastic wrap or it can be placed separately from the tip and ear mould.

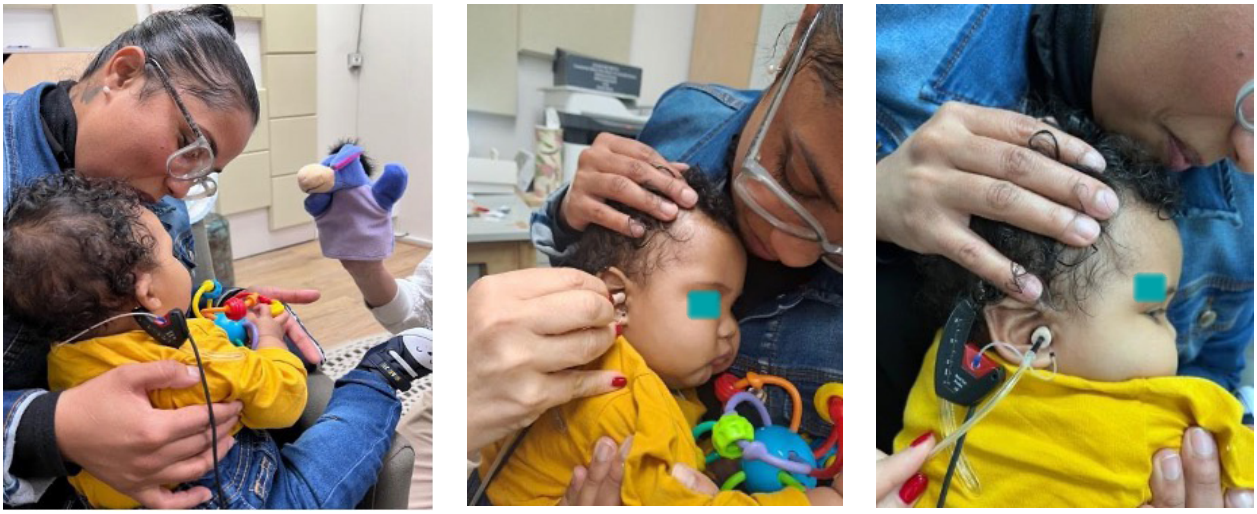

Figure 10.5: Inserting the probe tube and RECD transducer (with a foam tip) in the infant's ear

5. Next, the real ear should be measured. The same sweep tone that was used for the coupler measurement will be transmitted through the transducer. As a child's ear is smaller, the resulting ear canal SPL is higher. Therefore, ear canal measurements typically show positive RECD values in children (there are instances, however, such as with a perforation in the TM where the child's ear canal SPL will not be higher than the coupler values). Figure 10.6 demonstrates this difference in ear canal SPL.

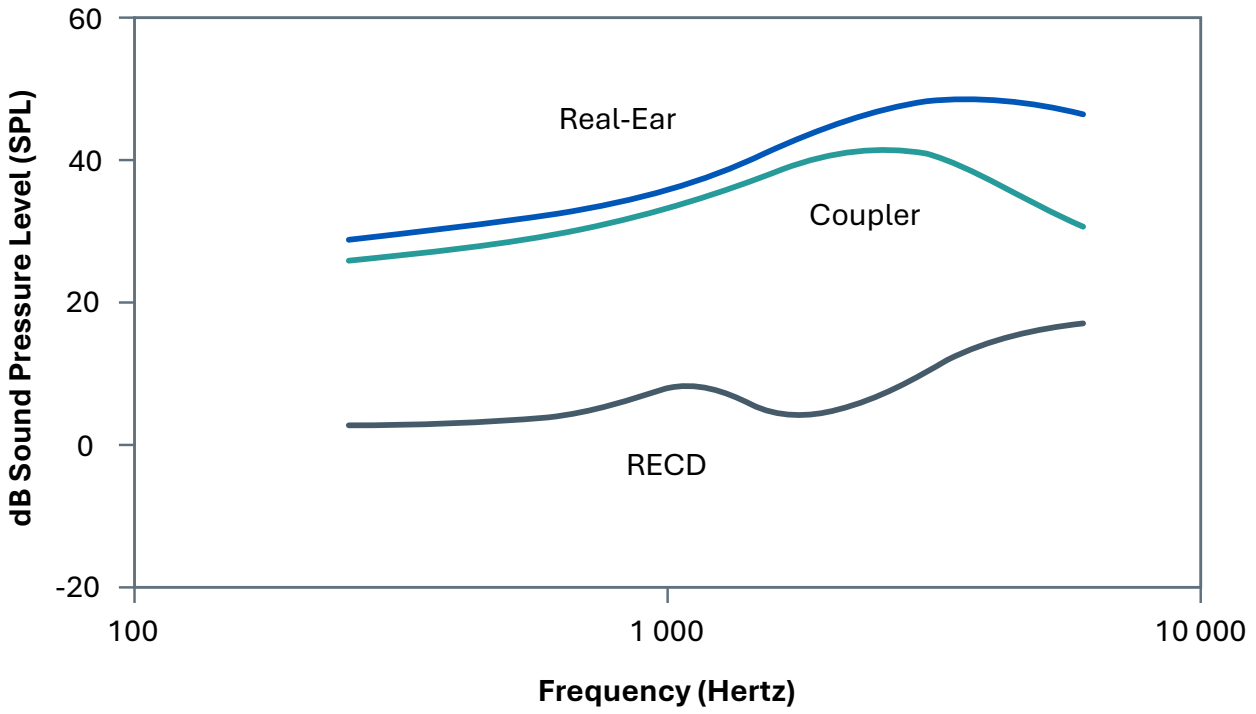

Figure 10.6: Higher ear canal SPL for a typical child's ear, compared to coupler values

If the values are not positive, it is important to make sure the probe tube is inserted deep enough. If the probe tube is blocked or pinched against the canal the real ear values will be very low. If an ear mould is being used it should be changed to a foam tip. If there is a drop off in the high frequencies it is possible that the probe tube is not inserted deep enough, or that there is leakage. The HHP should attempt insertion again or should replace the tip with a larger tip or else with the ear mould. If the child has a perforation in the TM or myringotomy tubes or if the ear mould vents are larger than one mm, the ear canal volume will be larger, with lower ear canal SPL and negative RECD values will be expected. This will result in more prescribed gain, which is appropriate for the child at this time. As the ear canal grows, RECD values will change. RECD values should be repeated regularly. As a guideline, the RECD should be remeasured when the ear mould is remade. RECDs from one ear can be used for the other side.

6. If, after inspection of the real ear curve, the HHP is satisfied that the real ear SPL values are valid, the probe tube and ear mould should be removed. The HHP can now continue programming and verifying the fitting in the test box, using the RECD values.

Procedure for verification of the hearing aid fitting by performing real ear measures (in the test box, figure 10.7)

1. Once the RECD has been measured, the HA should be placed in the test box coupled to the 2cc coupler. The test box should not be closed yet. One HA should be coupled (most systems can only test oe HA at a time), and one HA should be outside the box.
2. Both HAs should be connected to the manufacturer's software. The procedure should be repeated for the other ear after the fitting of the side is verified. The HAs should have already been pre-programmed to the manufacturer's default settings as a starting point.
3. If it was possible to measure RECDs, these values should be entered into the measurement equipment (some equipment is computer software-based, while other types of equipment will need the values to be entered manually). Age-appropriate predicted values can be used if RECDs could not be measured. If it is a follow-up visit, or RECD values have been measured before (this could have been done with the baby asleep after ABR testing, or during any other visit prior to the fitting) these values should be entered into the measurement equipment system or software.
4. The prescriptive target used to pre-programme the HAs should be chosen in the software (DSL or NAL) to generate targets in the system.
5. In the simulated (test box) real-ear section of the system, the HHP should choose a calibrated speech stimulus (ISTS if that is available). Noise reduction and frequency lowering should be disabled in the software for the verification process.
6. A level of 65 dB SPL should be entered to measure a simulated real-ear aided response. The HA should be adjusted to provide a close match to the average speech targets (LTASS) for 65 dB SPL. In general, these measured values should be within ±5 dBs of the prescription target. In the case of a steeply sloping loss or a severe loss, fit-to-target might not be possible. The HHP should use the upper limit of speech (the target line above the LTASS line in the middle) to gauge if these sounds (especially /s/) are audible. If not, frequency lowering should be activated.

7. Next, the HHP should choose a high-level (85 – 90 dB SPL) narrowband stimulus or EUHA MPO stimulus and adjust the HA so it approximates the prescriptive targets and does not exceed the predicted uncomfortable levels.
8. Step six should be repeated for soft (55 dB SPL) and loud inputs (75 dB SPL) to evaluate compression characteristics of the HA. The fit for average speech and MPO targets should not be compromised to obtain a better match for soft or loud speech. A close match to average conversational speech and maximum output targets of the HAs should be given priority when verifying HAs for infants and young children (Bagatto et al. 2023).
9. Once the HHP has adjusted the HAs at 55- and 75 dB SPL, the process for 65 dB SPL and MPO should be repeated.
10. The SII (ANSI S3.5-1997) values are available on most real ear measurement systems. During this verification process of HAs, the aided SII values should be displayed together with the DSL targets and HA verification curves. These values give a valuable indication of the audibility of speech.
11. If the HHP has determined that the child is a candidate for frequency lowering, it should be activated at this stage. The HHP should select the signal simulating /s/ and fine tune the frequency lowering setting until the upper shoulder of /s/ falls within the audible area (that is, above the threshold SPL at that frequency). It is recommended that the chosen setting employs the weakest possible frequency lowering setting, measuring the upper shoulder of the /s/ sound above the upper limit of speech. Associated SII values do not need to be recalculated based on the fitting with frequency lowering activated.
12. In the case of binaural aid fittings, all the steps should be repeated for the HA on the other side. The HHP should make sure to separate (unlink) left and right HAs in the programming software so that the programming that was done for the first HA is not lost.

Procedure for the verification of remote microphone systems

If a toddler or child has been fitted with an ALD such as an RM system, it should be verified. There are two objectives: To ensure that the active microphone does not change the HA gain or output when it is on, and to ensure that the microphone transmits sound to the HA.

1. After the HA verification is completed and fitted to DSL/NAL targets, the HA should be attached to the 2 cc Coupler and the response measured to the LTASS at 65 dB SPL, using the ISTS or calibrated speech.



Figure 10.7: Hearing aid set-up in the test box

2. If the HA has an RM receiver attachment, it should be attached now. Most new technology HAs have an integrated receiver. The RM transmitter should be activated (Figure 10.8a), and the 65 dB SPL measurement repeated with the microphone on, but muted. The curve should not change from the curve without the RM system attached, indicating that the amount of amplification is not affected (less) when the RM system is in use. If the SPL measurement is different, the RM gain should be adjusted in the HA software.

Figure 10.8: (a)



Figure 10.8: (b)



Figure 10.8: (a) RM transmitter outside the HIT, switched on, (b) RM transmitter inside the HIT, switched on

3. Next, the HA and the coupler should be removed from the test box (Figure 10.8B). The RM microphone should be placed in the test box and the signal presented. The microphone will now transmit the signal to the receiver in the RM system active in or attached to the HA. Again, the output curve should be the same as without the RM attached, indicating that the microphone is transmitting speech as it should.

Follow-up sessions and outcome measures

First visit

After the fitting and verification of the HAs, caregivers should be shown how to insert, clean, conduct a listening check, replace batteries or charge the HAs, and should be given advice on retention. It often works best to distract the child after initially inserting the HAs first thing

in the morning. Keeping the HAs in for the first two weeks will be the most important task for the caregivers. It is important to ensure all safety aspects are discussed, such as the danger of ingesting small batteries. All the other small parts should be put out of reach of the child and other curious siblings. Make sure the caregivers are comfortable with the HAs before they leave the session. If the child reacts to sound or benefit can be demonstrated, the caregivers will be more motivated to keep the HAs on. The HHP should give the parents a call the next day to ensure that the first day of wearing the HAs went well. The first follow-up should be scheduled for no longer than two weeks after the initial fitting.



Figure 10.9: A child with hearing aids being placed in his ears.

The following recommendations are made for follow-up visits and outcome measures:

Scheduling of visits

- First two years after the initial fitting: Every three months
- Thereafter: Every four to six months until the child reaches school age
- School-age children: annually or as needed

The following procedures or components should be part of every visit:

- Caregivers should be interviewed to address concerns or troubleshoot problems.

- Otoscopy should be performed.
- Tympanometry should be repeated.
- HAs should be tested in the test box/Listening check should be done by the HHP.
- Ear mould fit should be checked and replacement of the ear mould done if necessary.
- Unaided hearing evaluations (including speech perception testing) should be done regularly.
- Data logging should be reviewed to ensure regular use of HAs.
- RECDs should be re-measured as necessary and always if the ear moulds have been replaced.
- HAs should be re-programmed and re-verified with revised thresholds and RECDs.
- Aided hearing evaluation (including speech perception testing) should be done.
- Caregivers should complete validation questionnaires to ensure optimal audibility.
- Detailed notes should be made in the child's file regarding setting changes.
- Referral to other professionals should be made as necessary.
- Reports should be written after each visit and sent to the child's school and/or the professionals involved with the child.

Objective Validation Measures

HAF objective validation measures include aided sound field testing, as well as parent/teacher questionnaires. Once the aided SII is completed with the values obtained during real ear measures (if it was not done automatically by the measurement system [with the 65 dB SPL and 55 dB SPL input signals]), the HHP must determine if the HAF is electro-acoustically acceptable for the child's pure tone average.

Aided sound field testing: (when age appropriate)

- The child should be positioned so that loudspeakers are at head height and +/- one meter from the child.
- Narrowband noise or pulsed tones should be used.
- Ear specific results should be obtained at least at 2 kHz and 0.5 kHz (take into consideration that the degree of HL, auditory awareness, duration of HL and quality of HAs will affect aided thresholds).

- Aided speech perception: The same hierarchy as with unaided speech perception testing should be used. Speech should not be presented louder than 60 dB HL as loudspeakers will tend to distort sound. Ear specific speech perception scores should be obtained with and without lipreading.

Outcomes questionnaires

Developmentally-appropriate outcomes questionnaires should be used to provide complementary information on auditory development in addition to measures of objective gain and output. Two examples of parental outcomes questionnaires are described below.

a) LittlEARS Auditory Questionnaire

The purpose of the LittlEARS Auditory Questionnaire is to assess the auditory behaviour of infants with permanent childhood hearing impairment who wear HAs or CIs (Coninx, et al. 2009; Tsaikpini, et al. 2004). It is recommended that administration of the LittlEARS questionnaire occurs at some point prior to HAF and at regular follow-up visits. The 35 questions in the LittlEARS questionnaire assess auditory development during the first two years of hearing in the real world and provides insight into receptive and semantic auditory behaviour as well as expressive-vocal behaviour. The questions are listed in an age-dependent order and are in a yes/no format. The total of all 'yes' answers provides a score that can be compared to average and minimum age-dependent values. These values are provided in one month age categories based on normative data (Coninx, et al. 2009). The LittlEARS questionnaire is designed to be answered by caregivers and is not affected by how it is administered (that is, under professional guidance or independently). The total 'yes' score is entered on the score sheet at the point where age and score meet. A child with a score in the shaded region is not meeting auditory milestones for their age. On the other hand, a child with a score above the shaded region is meeting auditory development milestones for their age.

When a minimum score of 27 or better is achieved, the child's performance is at a ceiling score. If ceiling is reached, the LittlEARS questionnaire should no longer be administered. Instead, the clinician can begin to administer the Parent's Evaluation of Aural/Oral Performance in Children (PEACH), either at that appointment or at the follow-up visit.

If a child is not wearing HAs but has an identified HL, the questionnaire can be useful for monitoring auditory development and tracking progress over time. In this case, the LittlEARS questionnaire should be administered at every follow-up visit.

b) Parent's Evaluation of Aural/Oral Performance in Children (PEACH) (Ching and Hill 2007).

The PEACH Rating Scale may be administered to children with normal hearing as well as to children with HL who may or may not wear HAs. The instructions ask caregivers to observe their child's behaviour in everyday life and rate their child's hearing performance over the previous week across a range of hearing and communication scenarios. The nature of the rating scale allows it to be answered by the caregiver during an appointment with guidance from the clinician. Administration of the PEACH should occur at regularly scheduled follow-up visits when the child has reached a score of 27 or greater on the LittlEARS Auditory Questionnaire and is 24 months of age and older. This prerequisite should ensure that the child's auditory skills are within the range of the PEACH. The overall score is summed, along with summed scores for the quiet and noise sub scales. Each sum (overall, quiet, noise) is converted to a percentage. The unshaded and shaded regions can be used as benchmarks against which to interpret individual scores. The current score ranges can also be used as benchmarks with which to compare individual scores. A child who scores well on the Quiet sub-scale but not in the Noise sub-scale would warrant further review of technology, or the use of an RM system in situations other than the classroom can be considered. These decisions should be made on a case-by-case basis and in conjunction with the needs of the child and family.

Conclusion

HAF in infants and children is a dynamic, lifelong process that demands careful consideration of candidacy, technology, verification and family involvement. Any type or degree of HL with the potential to hinder speech and language development qualifies a child for HAF, provided there are no contraindications for air conduction hearing devices. The preferred device for most infants and children is a behind-the-ear (BTE) HA with a silicon ear mould, paediatric ear hook, rechargeable or tamperproof battery, broadband amplification, noise reduction features and RM system compatibility. Verification of the fitting must include RECD values to confirm audibility of speech at comfortable and safe levels. These RECD values should be remeasured frequently and whenever new ear moulds are issued. Output at various input

levels must be verified using a calibrated speech signal, and MPO must be measured using a sweep tone. All children who are candidates for HAs are also candidates for RM systems, which should be verified in a HA test box. Outcome measures should include the use of validated questionnaires. Regular follow-up appointments are essential to monitor the stability or progression of HL, assess the impact of ear canal sound pressure levels on thresholds and HA performance and to replace or modify ear moulds as necessary. Crucially, the family should be involved at every stage of the HAF process to ensure optimal support and outcomes for the child.

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Chapter 11

Paediatric Amplification: Implantable Hearing Devices

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'Every day, with appropriate technology and intervention, children with hearing loss are learning to listen, talk and thrive'

~Hearing First, 2025

Introduction

Advances in implantable hearing solutions, including bone conduction implants (BCIs), cochlear implants (CIs), auditory brainstem implants (ABIs) and middle ear implants (MEIs), offer effective options for managing HL, especially when traditional air-conduction hearing aids (HA) are insufficient. Implantable hearing devices provide direct stimulation to different parts of the auditory system, allowing better sound perception when HAs fail to meet the child's needs (McCreery 2014). Throughout this chapter, we reference several devices currently offered by various companies, including links to their respective websites. Please note that as technology rapidly evolves, device names and specifications may change, and some links may become outdated or may no longer be functional.

When choosing amplification, the following factors need to be considered (Bagatto and Scollie 2023):

- Age and timing: Early intervention is critical to maximise spoken language and auditory outcomes. Children with congenital HL should be treated as soon as possible — the sooner the intervention, the better the potential hearing outcome.
- Family-centred care: Involvement of parents and caregivers is essential for success (Chapter 15).
- Site of lesion and type of HL: For different types and sites of HL, different implantable solutions are available. In choosing an implantable hearing device, it is important to understand that the choice of the appropriate device depends on the site of the lesion.
- Long-term sustainability and upkeep of chosen hearing technology: Especially within resource-constrained environments, it is important to consider whether the family has access to the necessary resources to maintain the device and provide the necessary support. Implantable hearing devices are a costly intervention, and so is their maintenance, as well as auditory rehabilitation (Chapter 12) and other services like occupational therapy and educational support (Chapter 13).

Bone Conduction Hearing Devices (BCHDs)

BCHDs are used for children with conductive (CHL) or mixed hearing loss (MHL) or with limited usable hearing unilaterally (LUHU). BHDs transmit sound vibrations through the skull bone directly to the inner ear, bypassing the sound conductive mechanism of the outer and middle ear, and transmitting sound directly to the cochlea by creating vibrations in the bone of the skull (Reinfeldt et al. 2015). The device typically consists of an internally implanted component and an external sound processor that captures and converts sound into vibrations that are transmitted to the skull.

Category 1: Non-implantable BCHDs

The first category is non-implantable BCHDs, which can include traditional body-worn devices, soft band BCHDs and adhesive BCHDs (Table 11.1). Figure 11.1 shows an example of a soft band BCHD.



Figure 11.1: Child with a BCHD on a soft band. Source: Alder Hey Children's NHS Foundation Trust (2026)

Table 11.1: Non-implantable BCHD options

Non-implantable BCHD options	Description	Example
Traditional BCHDs	Body-worn devices or bone-conduction spectacles	Body-worn devices/ spectacles: *www.heartronix.co.za*
Soft-band BCHDs	Devices mounted on a soft headband that transmit vibrations through the skin	Cochlear BCHDs on a soft band/SoundArc: *www.cochlear.com/us/en/professionals/products-and-candidacy/baha-start*; Oticon soft band: *www.oticonmedical.com/solutions/bone-conduction/softband*; MED-EL soft band: ADHEAR can be worn on a soft band.
Adhesive BCHDs	Adhesive devices that stick to the skin and transmit sound vibrations	ADHEAR™ by MED-EL: *www.medel.pro/products/adhear*

Category 2: Implantable BCHD options

The second category is implantable BCHDs. There are currently two main types of BCHD implants available on the market: active BCHDs and passive BCHDs (Table 11.2). Passive BCHDs are further categorised into transcutaneous and percutaneous devices.

Table 11.2: Implantable BCHD options

Category	Description	Examples
Active BCHDs	The implanted device actively vibrates within the bone, stimulated by the external sound processor, which is held in place by a magnet.	MED-EL Bonebridge: *www.medel.com/hearing-solutions/bonebridge* Cochlear Osia: *www.cochlear.com/us/en/home/products-and-accessories/cochlear-osia-system* Oticon Sentio: *www.sentio.oticonmedical.com*
Passive BCHDs: Percutaneous	A titanium abutment is implanted into the skull and protrudes through the skin; the sound processor clips onto the abutment.	Cochlear Baha System: *www.cochlear.com/us/en/home/products-and-accessories/cochlear-baha-system/baha-implant* Oticon Ponto: *www.oticonmedical.com/solutions/bone-conduction/ponto-system*

Category	Description	Examples
Passive BCHDs: Transcutaneous	This is a fully implanted disc under the skin; the external sound processor is held in place by a magnet over the skin.	Cochlear Attract: *www.cochlear.com/au/en/home/products-and-accessories/cochlear-baha-system/baha-implant*

Active BCHDs

An active BCHD (Figure 11.2) is an implantable hearing solution designed to directly stimulate the bone, bypassing the outer and middle ear to deliver sound to the inner ear (Reinfeldt et al. 2015). Unlike passive systems, active BCHDs contain an implanted transducer that vibrates within the bone itself, improving sound quality and reducing energy loss. All active bone conduction devices are transcutaneous because they incorporate a built-in actuator that actively generates vibrations (Reinfeldt et al. 2015). They are designed to provide a more robust sound output by converting electrical signals into mechanical vibrations directly stimulating the bone. Many manufacturers offer this type of device (Table 11.3). The introduction of active transcutaneous devices (BONEBRIDGE™ and Osia™) offers electronic signal transmission to optimise audiologic gain.

Table 11.3: Active BCHDs

Device	Manufacturer	Implant technology	*Key Features*	*Coupling*
Bonebridge™	MED-EL	Bone Conduction Floating Mass Transducer (BC FMT)	The implanted transducer vibrates the bone directly; excellent sound transmission; cosmetically appealing.	External sound processor held by a magnet
Osia System™	Cochlear	Piezoelectric Actuator	Offers a powerful and stable performance; designed for conductive/ mixed HL; fully implanted transducer.	External sound processor held by a magnet
Sentio™	Oticon Medical	Skin-friendly transcutaneous design with an implanted vibrator	Ensures effective bone conduction while preserving skin integrity; discreet and reliable.	External sound processor held by a magnet.



Figure 11.2: Illustration of an active BCHD. Source: Hearing Loss Association of America (2026)

Passive BCHDs

In passive BCHDs, a portion of the implant embedded in the skull 'passively' receives the vibrations from an externally worn vibrating device – the sound processor (Reinfeldt et al. 2015). Table 11.4 details the key differences between transcutaneous and percutaneous passive BCHDs.

a) Transcutaneous BCHDs

These devices have an external sound processor placed against the skin, typically held in place by a magnetic or adhesive mechanism, with no part of the device passing through the skin. Sound vibrations are transferred through the skin to the underlying bone, stimulating the cochlea. The implanted portion of the device is entirely buried under the skin, with nothing penetrating through the skin to connect to the exterior component (Reinfeldt et al. 2015). Active devices are all transcutaneous, while some passive devices are transcutaneous with the external and internal components held in communication with magnets. In passive devices, some sound attenuation occurs because of the intervening skin. Figure 11.3a below illustrates a transcutaneous and percutaneous passive BCHD.

Figure 11.3: (a)

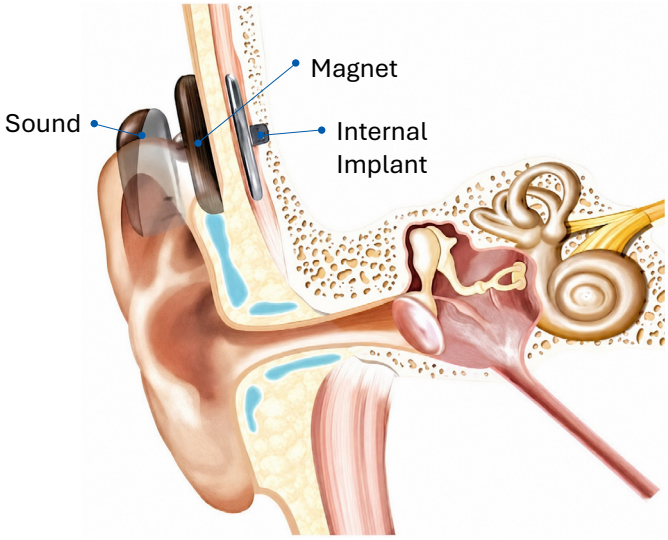

Figure 11.3: (b)

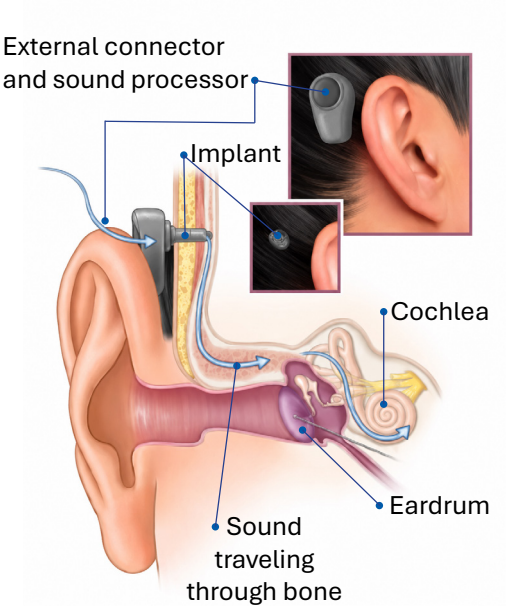

Figure 11.3: (a) Transcutaneous passive BCHD. Source: Questions and Answers in MRI (2026).
Figure 11.3: (b) Percutaneous passive BCHD. Source: Cleveland Clinic (2023).

b) Percutaneous BCHDs

A small titanium implant (an 'abutment') is surgically placed in the skull bone. The external processor connects/clips onto the abutment where it protrudes through the skin. This avoids sound attenuation, but the abutment traversing the skin may give rise to infections and requires good care. Sound vibrations from the external processor are transmitted directly through the abutment to the bone, providing efficient sound conduction (Reinfeldt et al. 2015). Figure 11.3b illustrates a percutaneous passive BCHD.

Table 11.4: Summary of key differences between transcutaneous and percutaneous BCHDs

Feature	Transcutaneous devices	Percutaneous devices
Skin Penetration	External processor placed on the skin	Through an abutment
Sound Transmission	Through the skin to the bone	Directly to the bone
Active or Passive	Primarily active	Primarily passive
Risk of Complications	Lower risk due to no skin penetration	Higher risk of skin infections
Maintenance	Generally easier maintenance	Requires care of the abutment area

Indications for BCHDs

Children with CHL can be treated through surgery that addresses the cause of the CHL (Chapter 9). Where the CHL cannot be corrected with reconstructive middle ear surgery, or the outcome of middle ear surgery is poor or unsatisfactory, conventional HAs or BCHDs can be a solution to provide a child with access to sound. BCHDs are considered in the following cases:

- Congenital microtia and/or atresia (absent or narrow ear canal).
- Ear canal stenosis.
- Syndromes including Down, Treacher-Collins and Goldenhar.
- Chronic or recurrent otorrhoea with chronically draining ears preventing use of traditional HAs.
- Chronic suppurative otitis media not (yet) resolved by surgery.
- Mastoid cavity problems where feedback and dizziness occur using HAs.
- Severe skin conditions preventing the use of traditional HAs.
- LUHU: Profound sensorineural hearing loss (SNHL) and/or poor word recognition in one ear, with normal or near-normal hearing in the other ear.

Additional considerations

- Age of the child: Younger children may benefit from non-surgical options (for example, soft band) until they are old enough for surgery.
- Children with Down Syndrome who often have fluctuating CHL and very narrow ear canals must be considered for BCHDs on a soft band.
- Medical contraindications to surgery: In some cases, BCHDs are the best alternative to traditional air conduction aids, especially when occluding the ear with a conventional air conduction HA might exacerbate the occurrence of middle ear infections.

Contraindications to BCHDs

These include:

- Scalp dermatologic problems due to the propensity for skin infection or skin breakdown related to the device.
- Patients with bony anatomy not amenable to osseointegration.

- The need for ongoing MRI evaluation, especially of the ipsilateral temporal lobe.

Assessment candidacy

Comprehensive audiological and medical evaluations are required to determine BCHD candidacy. The assessment process typically involves:

a) Audiological assessment (Chapter 5):

- Pure-tone audiometry to measure hearing thresholds.
- Masked and unmasked bone conduction testing to confirm conductive or mixed HL.
- Speech recognition testing in quiet and noise to evaluate speech perception.
- Bone conduction ABR testing to confirm CHL in young children (Chapter 6).

b) Medical assessment:

- Otoscopic examination will be needed to evaluate outer and middle ear status (Chapter 5 and 9).
- Imaging (for example, CT or MRI) may be necessary to assess anatomical structures. In a passive percutaneous BCHDs routine, a preoperative CT scan is not essential. It might be helpful in children, patients with congenital aural atresia or in patients with an irradiated bone to determine the ideal location for implant placement. In contrast, in active BCHDs, a preoperative CT scan is obtained routinely to determine if the patient's anatomy is adequate for placement of the device and for suitable device positioning.
- Consultation with an ENT specialist is necessary to determine if surgical implantation is appropriate.

As sound vibrations can easily travel through the skull, the BCHD will stimulate the cochlea (inner ear) with the best hearing, whether it is on the same side as the device or the opposite side (Samra 2018). This means that when fitting a BCHD, it should be based on the masked bone-conduction hearing levels or the hearing levels measured with the device in place (in situ thresholds). Each manufacturer of BCHDs has their own bone conduction fitting range. It is thus important to consult the different fitting ranges to select the ideal implant for the patient's HL. The specification sheets can be obtained directly from the manufacturer. Figure 11.4 provides the maximum bone conduction thresholds that can be fitted with an implantable bone conduction device (65 dB HL).

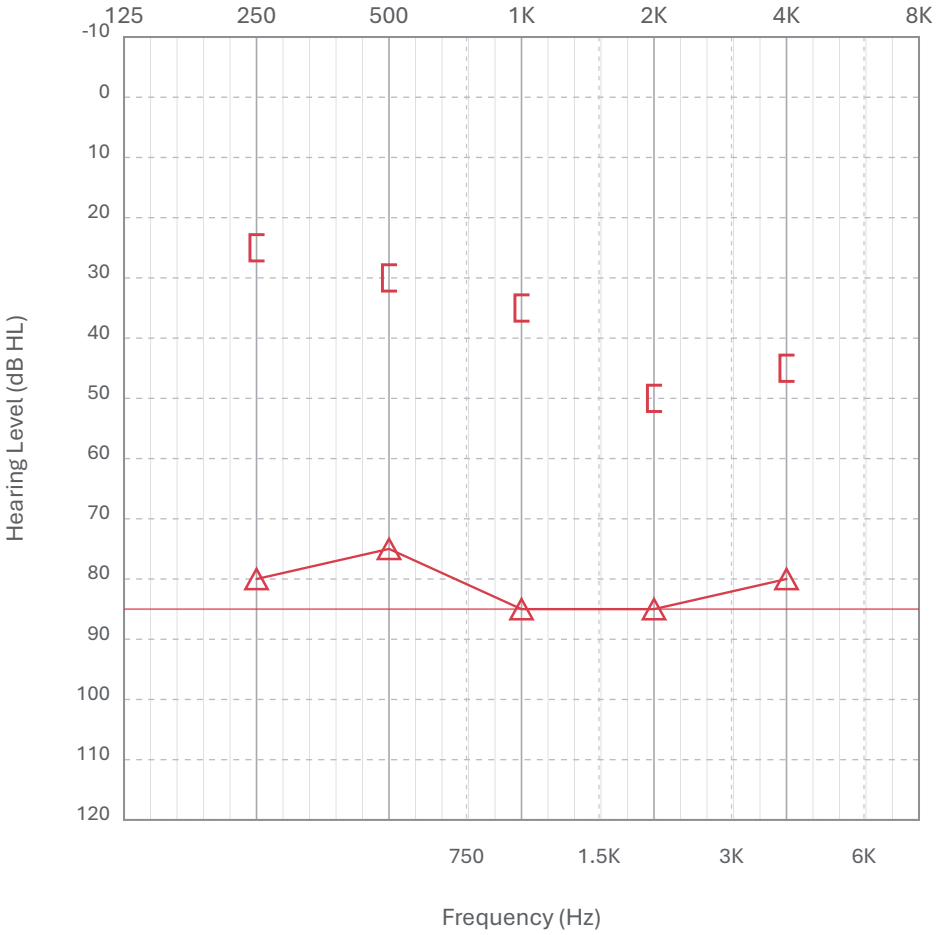

Figure 11.4: Maximum bone conduction thresholds considered for BCI selection for the right ear ([= masked bone conduction thresholds; Δ = masked air conduction thresholds)

Trial with BCHD

The child's family should be informed about the different types of BCDHs available, and the selected device should be explained in terms of its function and effectiveness in addressing the child's HL (Cleveland Clinic 2023). The longevity, reliability, upgrade options and MRI compatibility should be included in the discussion. The advantages and disadvantages of the different systems available on the market should also be included. Written information about the devices should be made available to the family.

A trial with a bone conduction soft band or sound processor allows the clinician to assess the benefits and the family to assess the suitability of a BCHD. Both aided pure tone testing and speech perception testing should be conducted to determine the benefit. It is important when choosing a device to consult the different bone conduction fitting ranges of devices available on the market. It should also be determined whether the child needs a BCHD on a soft band or if surgical implantation must be done. Children under the age of five years may not qualify for bone conduction implantation (Roman et al. 2011). These children should be fitted with a device on a soft band until they are old enough to receive an implant. Children with temporary CHL that will improve in the future either with surgery or medical management should be fitted with a BCHD on a soft band.

Surgical technique

The principle of the surgery is the suitable and secure placement of the implanted portion in the skull/mastoid bone where it is fixed and can achieve integration into the bone ('osseo-integration') (Wazen et al. 2007). The surgery is carried out under general anaesthetic and takes between forty minutes and one hour. The standard surgical approach consists of a postauricular incision with placement of the implanted component in or around the mastoid bone. The location and size of the incision will vary based on the device chosen, anatomic structures, planned implantation location and considerations of external ear reconstruction.

The ENT surgeon considers multiple factors before deciding on a surgery plan. In children with microtia who are planning or may be candidates for otoplasty in the future, the abutment should be positioned carefully in co-operation with the otoplasty surgeons to allow for microtia repair without compromising blood supply (McKay-Davies et al. 2024). Failure of osseointegration is more likely to occur when bone quality is immature or abnormal. Paediatric bone has lower mineral content and is softer and thinner than mature adult bone. To compensate for these factors, younger children may need to undergo two-stage surgery. Some devices are also not suitable for young children because thicker bone density is required.

Depending on the device chosen and surgeon preferences, the skin flap over the device may be thinned. The bony area immediately surrounding the implant location will need to be flat or drilled until it is flat to facilitate the firm placement of a passive transcutaneous device

(McKay-Davies et al. 2024). Active transcutaneous devices might require the drilling of a recess, depending on the particular device.

Post-operative care and complications

Patients are monitored postoperatively for wound care and soft tissue healing. Intraoperative complications of bone conduction implant (BCI) surgery are rare but can include damage to nearby structures such as the dura or sigmoid sinus, with associated risks of cerebrospinal fluid (CSF) leak or bleeding, respectively (McKay-Davies et al. 2024). If these complications occur, CSF leaks are typically managed with careful intraoperative repair and postoperative bed rest, whereas significant bleeding requires immediate hemostasis and, in rare cases, surgical revision.

Postoperative complications are most commonly soft tissue-related and include local skin numbness or pain, infection, excess granulation tissue, and hypertrophic scarring. Management strategies include:

- Local skin numbness or pain: Usually resolves spontaneously; symptomatic management may involve topical analgesics or short-term oral pain medication.
- Infection: Mild infections are treated with topical or systemic antibiotics; more severe infections may require debridement or, rarely, temporary removal of the implant.
- Excess granulation tissue: This can be treated with topical corticosteroids, silver nitrate cauterisation or surgical removal.
- Excess scarring: Excess scarring is managed conservatively or with minor surgical revision if it interferes with device function or skin integrity.

Late complications such as skin breakdown over the implant or chronic infection may necessitate more extensive intervention, including revision surgery or, in severe cases, explantation of the device.

Bone-related complications, though infrequent, include failure of osseointegration, technical fractures or issues related to trauma, chronic soft tissue infection or inadequate bony recipient sites. Management may involve surgical revision, removal and re-implantation,

depending on the severity and underlying cause. Osteomyelitis is exceedingly rare and if it occurs, it requires systemic antibiotic therapy and surgical debridement.

Activation, programming and (Re)habilitation post-surgery

a) Activation and initial fitting:

- Once the BCHD is surgically implanted or the soft band is fitted, the device is activated.
- During the activation, the clinician adjusts the sound processor's settings to meet the child's hearing needs.
- Aided sound field warble pure tone thresholds are obtained with the BCHD.
- Speech perception testing is carried out with the device, and if possible, speech perception in noise testing.

b) Programming:

- The device must be programmed to the child's audiogram, ensuring appropriate amplification across different frequencies.
- Some companies' BCHDs offer in-situ measurement or direct hearing thresholds of bone conduction thresholds. This means that when the processor is placed on the implant or soft band, the clinician can perform bone conduction threshold testing through the processor (Samra 2018). This is valuable for ensuring that accurate bone conduction thresholds are used to set the gain settings of the device across the frequency spectrum.
- Fine-tuning may be required in subsequent visits based on feedback from the child and caregivers.

c) (Re)habilitation:

- Following activation, auditory rehabilitation is crucial to maximise the benefits of the BCHD.
- Speech-language therapy and auditory training help children develop listening and communication skills.

- Ongoing audiological monitoring ensures that the device continues to meet the child's hearing needs.

Outcomes

The success of BCHDs in paediatric patients largely depends on early intervention, consistent device use and access to auditory rehabilitation services. Some reported outcomes include:

- Minimal surgical complications: BCHD surgery has demonstrated low rates of surgical complications, with successful healing in most patients and minimal postoperative issues.
- Improved speech perception in quiet and noise: BCHDs significantly improve speech understanding, especially in noisy environments. This is particularly beneficial for individuals with conductive or mixed HL and LUHU, helping them perform better in challenging listening situations.
- Improved sound localisation/spatial awareness: Children with CHL will experience improved sound localisation, although the accuracy will be lower than children with normal hearing. In children with LUHU, it will not be true sound localisation but rather improved spatial awareness.
- Improved hearing thresholds: BCHDs help close the air-bone gap, particularly in conductive and mixed HL, improving overall hearing thresholds.
- Better quality of life: Users of BCHDs report significant improvements in quality of life, citing enhanced social interactions, communication abilities and reduced listening effort, all of which are particularly important for children in educational settings.
- Effective treatment for LUHU: For individuals with LUHU, BCHDs provide significant auditory benefit by transmitting sound from the deaf side to the better-hearing ear, improving spatial awareness and overall hearing ability.

Cochlear Implants (CIs)

CIs are a well-established and effective treatment for children with severe to profound SNHL. By providing a child with access to sound, a CI enables the potential for the acquisition of spoken language.

CI team

A CI programme is made up of a dedicated, specialist, multidisciplinary team that functions within the context of a CI Programme (Dettman et al. 2016). The team consists of the following core hearing healthcare practitioners (HHPs): audiologists, otologists, radiologists and specialists in rehabilitation (speech and language therapists and/or auditory verbal therapists) (Warner-Czyz et al. 2022). At a minimum it should consist of an ENT surgeon with specialist experience in Otology and training in CI surgery, an audiologist and a speech-language therapist. Audiologists in the team should have received specialist training in CI MAPping. The speech-language therapist should be trained to work with children who have HL and are learning spoken language through technology. The team should have specialist knowledge and skills to work in the field of CI to manage, assess and work with children and adults with a range of complex needs, additional to their HL. The specialist CI team must be able to service all future needs of the child. CI involves a lifelong commitment by the family and the CI team. The teams should develop partnerships with local services where appropriate (Warner-Czyz et al. 2022). In South Africa there are thirteen CI programmes, details of which can be found here: https://www.sacig.org.za/patients-corner/implant-programs/. CI team members should belong to their local CI organisation that should have a set of guidelines to ensure the highest realistically possible standards of care for patients.

CI functioning

The principle of the CI system is that it bypasses the dysfunctional hair cells in the cochlea to directly stimulate the cochlear nerve with electrical current (Figure 11.5) (Carlyon and Goehring 2021). The auditory nerve, which is still functional, then takes these signals to the brain where they are interpreted as sounds and speech.

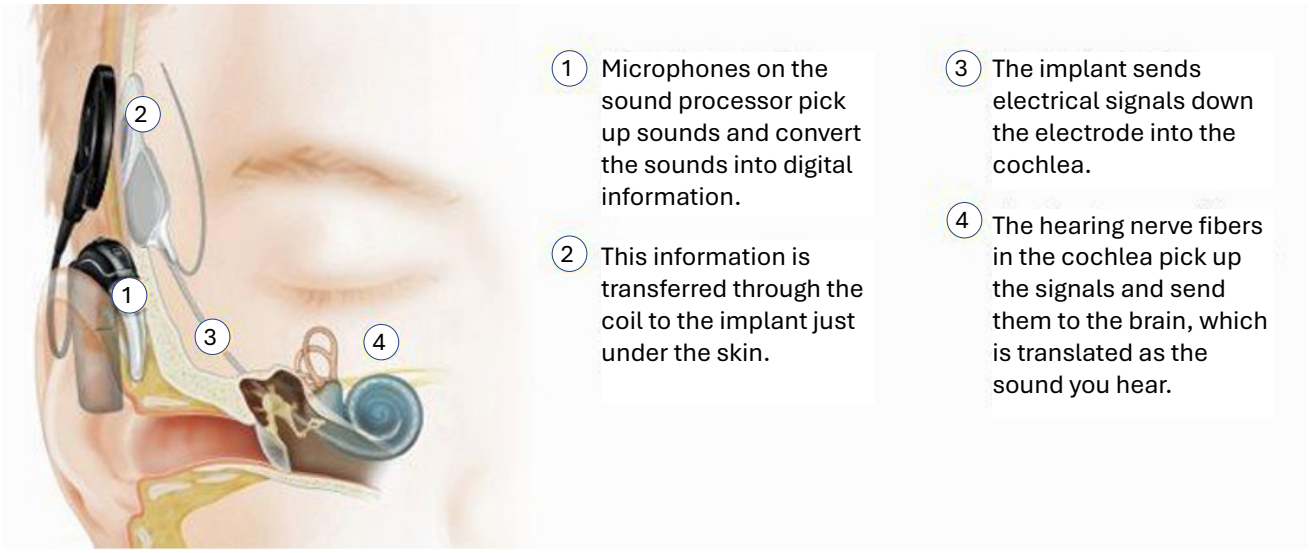

Figure 11.5: The components and functions of a CI. Source: ENT & Audiology Associates (2026).

A CI device consists of two primary components: an externally worn sound processor and an implanted receiver-stimulator and electrode array (Carlyon and Goehring 2021). The sound processor consists of a microphone and transmitter. The microphones on the processor pick up (acoustic) sounds from outside the ear. The speech processor converts the sounds into digital signals which it sends through a coil to the internal receiver that has been placed under the skin behind the ear. The signal is sent through the skin via a radio frequency signal. The receiver-stimulator sends the electrical signals to the multi-channel electrode array that has been inserted into the cochlea. The electrical signals activate the spiral ganglion cells to stimulate the cochlear nerve, which sends the signals to the brain. The brain interprets the electrical signals as sounds.

The coils of the external sound processor and internal receiver are held in apposition to send and receive the electrical signal by an external and internal magnet respectively. The sound processor is worn either behind the ear (Figure 11.6) or off the ear. The behind-the-ear processor includes a cable that connects the processor to the external coil and magnet directly over the magnet of the internal coil. The off-the-ear processor contains the magnet, the processor and the coil and is therefore placed directly over the internal coil.

There are three FDA (USA Conformity) and CE (European Conformity) approved manufacturers of CIs:

- Cochlear: www.cochlear.com
- Medel: www.medel.com
- Advanced bionics: www.advancedbionics.com



Figure 11.6: Behind-the-ear speech processor

Referrals for CI evaluation

Referral for CI by the HHP is based on both physiological and behavioural audiological information, within the context of access to sound with optimally fitted HA technology for each ear (Bagatto and Scollie 2023). The foundation of optimal outcomes is primarily based on early and appropriate intervention during the period of cortical plasticity (Ching et al. 2018). Should the family desire a spoken language outcome for their child, the recommendation is made for evaluation for candidacy and the child should be referred to a local (or accessible) CI team.

When making a CI referral, consideration should be given to each ear as well as the degree and configuration of the HL to determine whether a child is receiving appropriate access to sound through technology. Sixty-six per cent of speech sounds are heard above 1 kHz. To hear the softer sounds of speech (for example, /s/, /th/, /k/, /p/), which is necessary for acquiring spoken language, HAs should provide adequate access to sounds across the frequency spectrum (Figure 11.7).

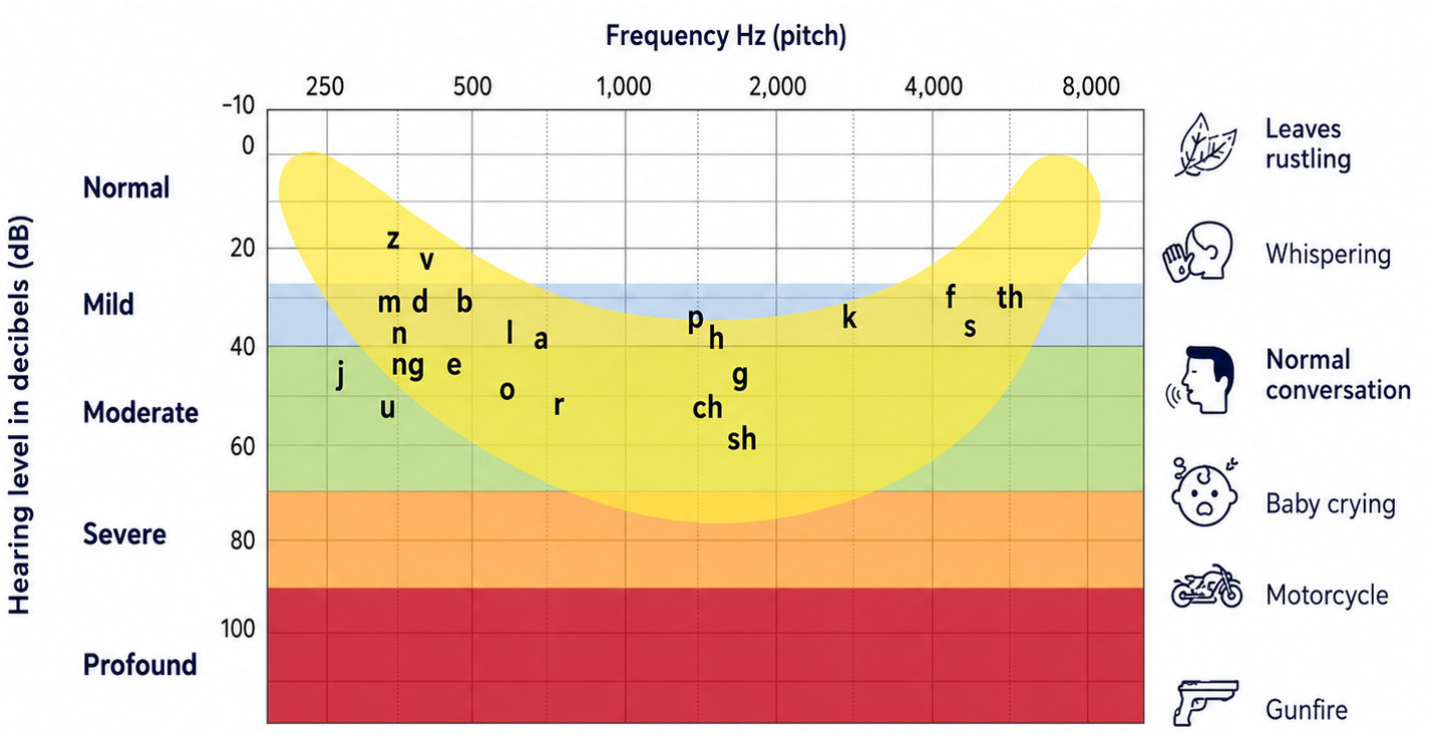

Figure 11.7: The 'speech banana' shows where speech sounds occur across the hearing range for pitch and loudness

The Speech Intelligibility Index (SII) is a standardised measure designed to evaluate speech audibility across different frequencies during the HA verification process (Bagatto and Scollie 2023). Information about the aided audibility of sounds for the child can be obtained from an aided audiogram or through the SII obtained from the HA verification. McCreery (2014) recommends that children who have an aided SII of less than 0.65 should be considered for CI regardless of (unaided) pure tone thresholds. If aided audibility is 0.65 or less for average conversation in quiet, it is likely to be much lower across realistic listening situations where there is noise and reverberation, and/or the talker is more than one meter away. Even sloping configurations with three Frequency Pure Tone Threshold (FPTA) as low as 43 dB HL can result in an aided SII of less than 0.65 (Leal, Marriage and Vickers 2016). About 90% of what children learn is through incidental hearing and learning (Madell, 2016). This means they need to be able to access soft speech easily through their technology.

Recommendations for evaluation for CI (Figure 11.8) should maintain flexibility and consider a child's skill progression (month-by-month progress in speech, language and auditory development) and quality of life with appropriately fitted HAs and under the management of an appropriately trained speech-language therapist. Children with HL should be carefully monitored on at least an annual basis to ensure their hearing levels and HA benefit remain stable. Should hearing deteriorate in either ear, referral for CI evaluation should be guided by their SII, aided thresholds, speech perception benefit or functional performance in that ear. This should ensure configuration of appropriate technology for each ear. Assessing functional outcomes based on parent and clinician feedback on a child's auditory awareness, auditory responsiveness and progress in acquiring language skills should become fundamental considerations for implant candidacy. Children with confirmed Auditory Neuropathy Spectrum Disorder (ANSD) should be considered as potential candidates for CI due to their disproportionately poor speech perception in relation to their audiogram.

Key Considerations for CI Candidacy

HHPs should refer paediatric patients to a CI team for evaluation for CI if they meet any of the following criteria (in either ear):

- Appropriately selected (aided) word recognition scores ≤ 50% correct
- Unaided pure-tone thresholds ≥ 60 dB HL

- Poor functional auditory performance, limited progress in language or auditory development or poor quality of life.

Parental and clinician information on functional outcomes should guide the primary managing audiologist regarding timely referral for cochlear implant evaluation. Consideration of the family's lifestyle, goals and expectations should be considered as part of the holistic care of the child. Importantly, in cases of deafness resulting from meningitis or an inflammatory condition of the cochlea where there is radiological evidence of ossification, CI should be considered an emergency and (bilateral) implantation should be recommended (Birman, Elliot and Gibson 2012). This may need to be prior to the age of six months.

Process of implantation

Once candidacy is confirmed by the team, the steps towards implantation and longer-term management are determined (Figure 11.8).

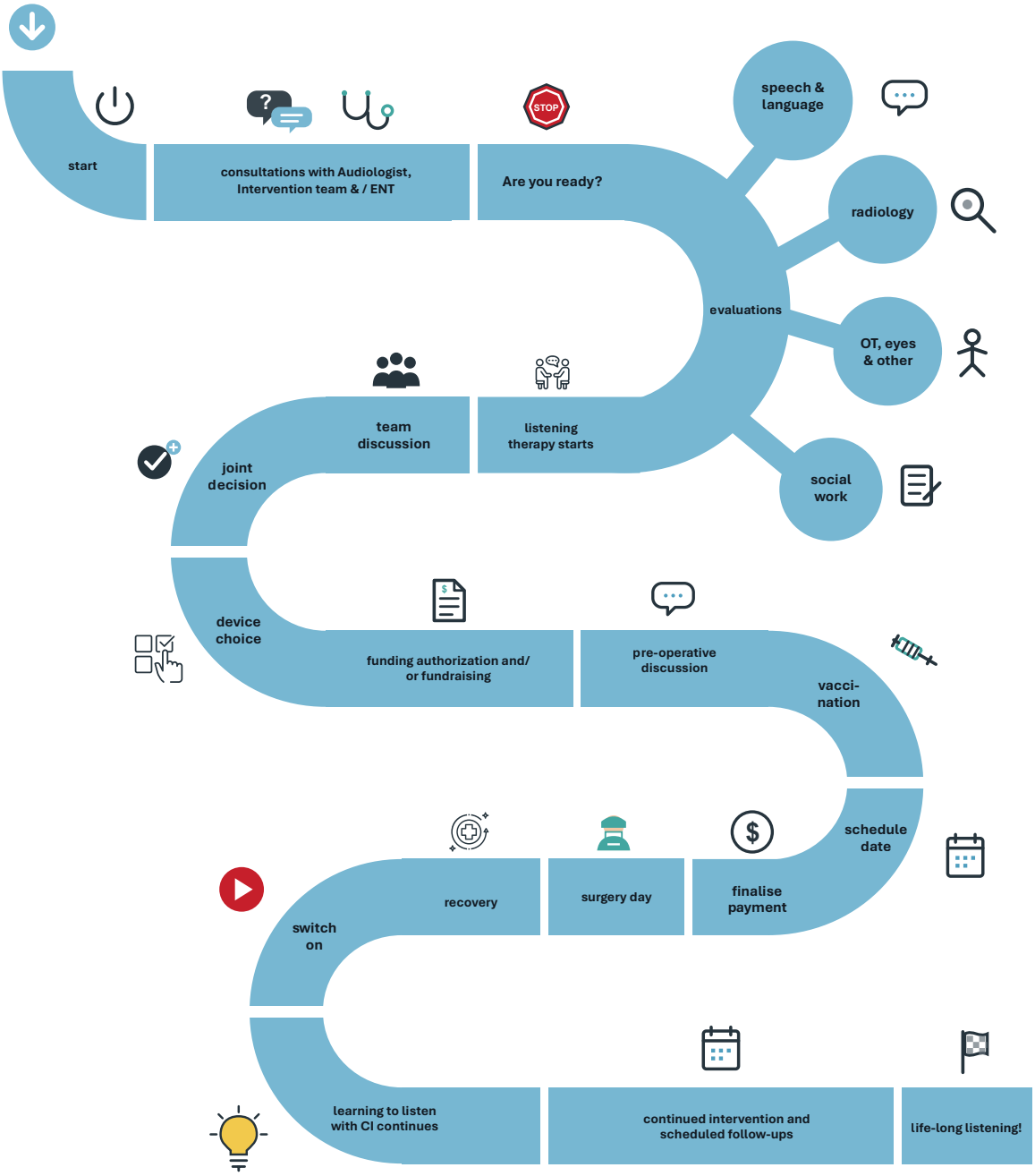

Figure 11.8: Roadmap for parents outlining steps in assessment and activation period of CI. Source: Surida Booysen

Assessment for candidacy

Assessing candidacy is an important part of the CI process. Considerations are given to the following factors to determine appropriate candidacy:

- Audiological criteria.
- Radiological criteria: The anatomy of the cochlea and auditory nerve should be suitable for insertion of the electrode array.
- Developmental factors:For example, in the case of other associated conditions, such as developmental delay, visual impairment, sensory processing disorders.
- Age: Implantation should ideally take place within the first three years of life for children with congenital bilateral severe to profound SNHL).
- Family participation.
- Financial factors: In countries with limited resources for long-term maintenance costs, consideration should be given to the ability of the family to maintain the sound processor and components, as well as to access appropriate educational support and facilities.

CI surgical implantation

After consideration and confirmation of suitable candidacy by the CI team, surgery should be scheduled. Surgery is done by the CI team's ENT surgeon and usually lasts two to three hours. Pneumococcal vaccination is given at least two weeks prior to the operation. Absolute sterility is essential to avoid the risk of infection. Implantation cannot be done on an ear that is infected or that holds risk of future infection. In such a case, preliminary subtotal petrosectomy and blind-sac closure of the ear must be considered (Hellingman and Dunnebier 2009). Infection risk following CI surgery is reduced by the use of an antibiotic that crosses the blood-brain barrier.

CI surgery comprises the following steps:

- Creation of a pocket/recess/bed for the body of the receiver-stimulator under the scalp (the periosteum) posterior-superior to the ear
- Creation of a pathway for the electrode array from this bed to the cochlea via:
- A cortical mastoidectomy

- A posterior tympanotomy exposing the round window
- Opening the cochlea
- Insertion of the electrode into the cochlea

The description provided is of the most commonly used, standard approach. Other techniques exist and anatomical variations might call for different approaches. The creation of the bed for the implant body has been much simplified in recent times. Historically a large incision was used, a deep bed drilled and ties used to hold the implant down. Currently, partly because the implants are far slimmer, a smaller incision is adequate to elevate a limited subperiosteal pocket which holds the implant in position (Carlyon and Goehring 2021). Surgeons may elect to drill a limited wedge-shaped recess at the anterior edge of the implant to prevent it from migrating anteriorly. In this case a small tunnel is made forward of this bed for the electrode array.

The surgeon must then provide a pathway for the delicate electrode array towards the cochlea. Generally, this is done by drilling an opening through the mastoid air cells located posterior-superior to the ear canal, down to the mastoid antrum – the largest of these air cells (this procedure is called a 'cortical mastoidectomy') (McKay-Davies et al. 2024). The surgeon then exposes and identifies the lateral semicircular canal and the short process of the incus in the fossa incudis. This guides the next step – the careful drilling of a 'posterior tympanotomy'. This is a slot drilled into the posterior recess of the middle ear, deep to and behind the eardrum and the chorda tympani nerve and superficial and anterior to the facial nerve. Through this posterior tympanotomy opening the surgeon can visualise the basal turn of the cochlea and the round window (Warner-Czyz et al. 2022; Wu et al. 2023).

The surgeon is then in a position to open the cochlea and insert the electrode array. The ideal is to create, as a-traumatically as possible, an opening for insertion of the electrode into the scala tympani. Two options exist: to drill a cochleostomy into the basal turn of the cochlea, or to insert the electrode through the round window after incising its membrane (Wu et al. 2023). As electrodes have become thinner and more delicate and implants are being performed increasingly on patients with hearing to preserve, most surgeons currently practise 'soft surgical techniques' and favour round window insertion (McKay-Davies et al. 2024).

Insertion of the electrode into the cochlea should, again, be performed as a-traumatically as possible to preserve hearing and the fine structure of the cochlea. The choice of electrode

type, use of corticosteroids both systemically and locally, very slow and even insertion and correct angle of insertion to avoid damage to the basilar membrane and transposition into the scala vestibuli are some of the principles of 'soft surgery' (McKay-Davies et al. 2024). During the surgery, the CI audiologist tests the integrity of the electrode function and response of the auditory nerve to electrical simulation via CI software after the surgeon has inserted the electrode array.

The operation is completed by sealing off the opening into the cochlea with fascia, securing the electrode in the mastoidectomy to mitigate against extrusion, and using a layered closure of the skin incision.

The device is tested for function using impedance measurements and electrically evoked neural response (eCAPS) telemetry is performed. A modified Stenver's view mobile x-ray is performed to confirm that there is no misplacement or tip fold-over of the electrode (device testing may also be used to exclude this). A firm bandage is used for a few days and the patient is seen by the surgeon within a week to identify and treat possible early complications.

Activation, programming and (re)habilitation

a) Activation and MAPping

Activation of the external sound processor occurs about two to three weeks after surgery by the audiologist on the CI team. Activation involves determining both soft and comfortably loud levels of sound through electrical stimulation, a process known as 'MAPping' (Fu and Shannon 2002). This should include both behavioural and physiological measures (Electrical Compound Action Potentials [eCAPS] or Electrical Stapedial Reflex Thresholds [eSRT]) to guide the fitting process. Within or at least by three months post-activation a hearing test should be performed in the booth to confirm MAPping levels and whether appropriate access to sound levels across the frequency spectrum is obtained. Aided thresholds should be 20-30 dB HL across the frequency spectrum (Fu and Shannon 2002).

b) Family coaching

Family coaching by the speech-language therapist is essential for the development of appropriate listening and spoken language throughout the immediate and longer-term (Chapter15). The family (caregivers) play a crucial role in this process.

c) Follow-up

After an appropriate MAP is established, the intervals when the child should be seen by the audiologist are three-monthly for the first two years, then twice a year for the next two years and annually thereafter (Wu et al. 2023). Once the child starts formal schooling, the audiologist should establish and maintain contact with the educators to ensure optimal access to sound in the classroom. This should include the use of assistive listening devices (for example, RM systems) so that the child has optimal access to sound within the adverse listening conditions of a typical classroom.

Outcomes

Ongoing, holistic care involves a range of factors, including continued accurate MAPping (programming) of the CI, verification of the programme provided, multidisciplinary rehabilitation, consistent monitoring and tracking of listening and communication skills and family compliance. Feedback and teamwork among immediate core team members as well as external role players form the fundamentals of optimising the child's outcome.

Important factors influence the outcome of CI in children:

a) Age at implantation

Early fitting of hearing devices is key to achieving better speech, language and functional performance outcomes by five years of age as shown in the LOCHI study (Ching et al. 2018). Early binaural fitting of appropriate technology reduces the negative effects of deafness on the developing auditory system and captures the remarkable neuroplasticity of early brain development. This includes implantation of the deaf ear in cases of LUHU.

Implantation prior to twenty-four months of age can promote speech understanding, and ideally even before twelve months, can support speech intelligibility and language outcomes on par with normal-hearing peers (Dettman et al. 2016). Children implanted ≤ twelve months of age have better auditory outcomes than older children (Wu et al. 2023). Children who receive a cochlear implant at nine months of age have a better opportunity for reaching typical language development milestones by the time they enter school. Early CI should therefore be considered for infants with HL. The FDA (2020) has approved cochlear implantation in infants

from nine months of age. It is recommended that congenitally deaf children should receive bilateral CIs before three years of age.

b) Special populations

Sixty per cent of children with HL have a secondary disability, most commonly an intellectual disability, a learning disability or developmental delay (Birman, Elliot and Gibson 2012). These conditions may result in slower rates of skill acquisition and greater variability in communication outcomes; however, the presence of additional disabilities should not automatically preclude cochlear implantation. When considering CI in these special populations, clinicians should carefully assess:

- Overall health and medical stability: To ensure the child can safely undergo surgery and general anesthesia
- Cognitive and developmental profile: To set realistic expectations for post-implant progress and tailor rehabilitation goals
- Motivation and support systems: Family involvement and access to rehabilitation services are critical for successful outcomes.
- Communication environment and resources: Including availability of specialised therapy, assistive technology and inclusive educational settings
- Potential for auditory skill development: Even if outcomes are slower or unpredictable, early implantation may still provide meaningful benefit in receptive and expressive communication.

By systematically evaluating these factors, clinicians can provide individualised recommendations and maximise the likelihood of functional benefit from cochlear implantation, even in children with complex needs.

c) Consistent device use

As with HAs, consistent CI use has been shown to be a vital factor contributing to better outcomes (Warner-Czyz et al. 2022).

d) Resources

Financial resources, access to care and therapeutic support should be accounted for and considered in ensuring a holistic perspective on the child's outcomes.

Auditory Brainstem Implants (ABIs)

ABIs provide direct electrical stimulation to the brainstem: the electrode paddle is placed directly on the cochlear nucleus in the brainstem. ABI would be considered for children for whom CI is not possible due to radiological evidence of an absent auditory nerve or cochlear anatomy where insertion of an electrode array would not be possible. This is usually a last resort when CIs are not viable. ABIs provide meaningful auditory sensations and environmental sound awareness, often improving quality of life for affected children (Colletti et al., 2005). Outcomes vary and the ability to achieve accurate speech perception is less predictable than in the case of CIs.

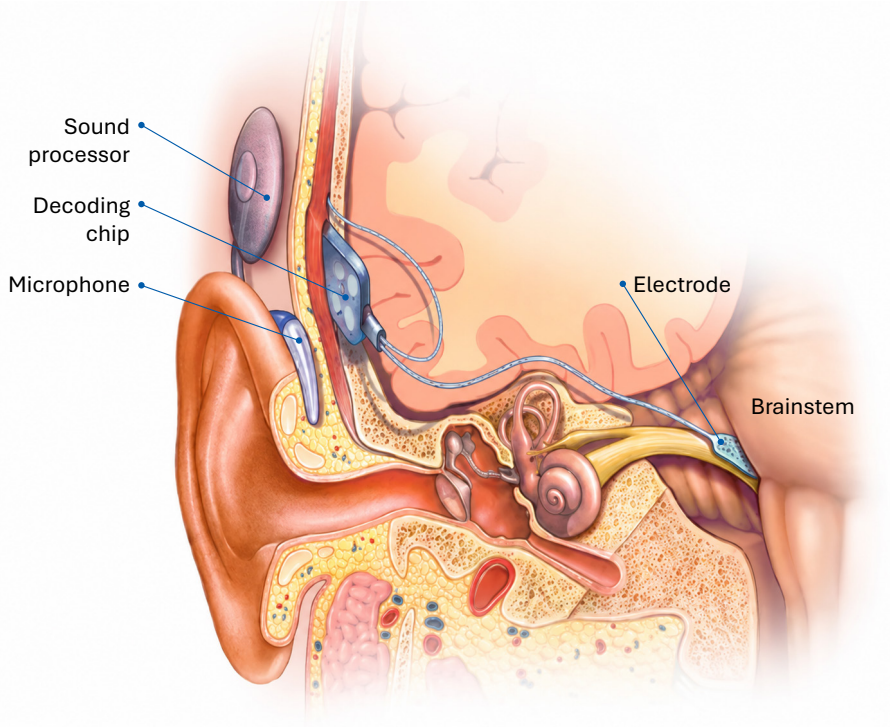

Figure 11.9: ABI components including external sound processor, cable and coil and internal receiver and electrode array. Source: Mayo Clinic (2026)

Middle Ear Implants (MEIs)

MEI are suitable for children with CHL or MHL who cannot wear traditional HAs due to chronic ear infections, ear canal issues or personal preference (Beutner and Huttenbrink 2011). These devices mechanically stimulate the ossicles (middle ear bones) to transmit sound (Figure 11.10). MEI is considered for mild to severe SNHL or MHL, chronic middle ear conditions and unilateral or bilateral HL.

This section does not cover MEI in detail, however, key factors to consider when evaluating candidacy are listed below:

- The child's age and craniofacial anatomy (suitability for implant placement)
- Middle ear status (active infection or ossicular chain abnormalities)
- Degree and type of HL
- Rehabilitation potential and family support

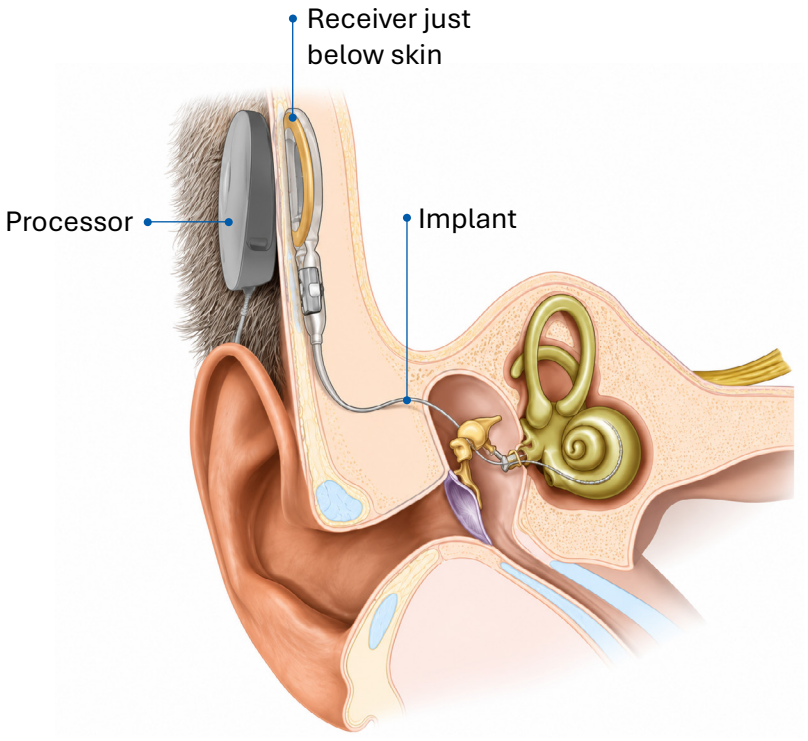

Figure 11.10: Illustration of a middle ear implant. Source: Hearing Loss Association of America (2026)

Considerations for Establishing an Auditory Implant Programme in a Low-Resourced Country

Auditory implants provide an opportunity for a child to learn spoken language, enhance educational opportunities and to ultimately be part of the hearing world. However, auditory implants in low-resourced countries come with many challenges and should be carefully considered when establishing new programmes.

Some considerations are:

- Available, appropriately trained core team members where international support is available when required
- Regular, appropriate parent coaching by suitably specialised speech-language/ Auditory-Verbal therapists within the team with the aim of developing listening and spoken language
- Sustainable programmes able to provide life-long support for auditory implant recipients
- Appropriate educational options for children once they start school-going age
- Available and sustainabley financial resources for both initial and long-term costs (including lifelong maintenance costs of the external speech processor).
- Careful device choice, considered against the critical role the company will play in lifelong support (for training, logistics and technical device support)
- Good sanitary conditions to avoid potential infections, as some auditory implants require a percutaneous connection between the implant and the processor

Considered against a backdrop of financial and infrastructure challenges in countries with limited financial, educational and professional resources, implantable hearing devices can potentially be highly cost effective for society, reducing educational and other costs (Bond et al. 2009). However, the establishment and maintenance of auditory implants in low-resourced countries should be approached in a responsible, calculated manner with careful regard to ensuring ethical, cost-effective sustainable and evidence-based standards of practice to recipients over their lifetimes.

Conclusion

Professionals working with children with HL are uniquely positioned to facilitate timely referrals for implantable hearing devices that are critical for optimal auditory brain development. This, in turn, supports the development of listening, speech and language skills — foundational abilities that significantly influence later educational achievement and employment opportunities. A team-based, multidisciplinary approach, which brings together audiologists, speech-language pathologists and ENT specialists, ensures comprehensive assessment and decision making around candidacy. With appropriate device fitting and consistent (re) habilitation, auditory implants can meaningfully enhance hearing outcomes and improve the overall quality of life for paediatric patients.

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Chapter 12

Paediatric Aural (Re)Habilitation

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'Excellent listening and spoken language outcomes are probable for children with hearing loss when we do what it takes'

~Mission Probable: Hearing First, 2019

Introduction

Changes in the paediatric hearing healthcare landscape have allowed for unprecedented possibilities and extraordinary outcomes for children with hearing loss (HL). The ability to screen for HL shortly after birth, together with changes in hearing technology, underpin these possibilities and outcomes. There is a wide range of intervention approaches (Figure 12.1) ranging from an only visual mode of communication to a Listening and Spoken Language (LSL) approach.

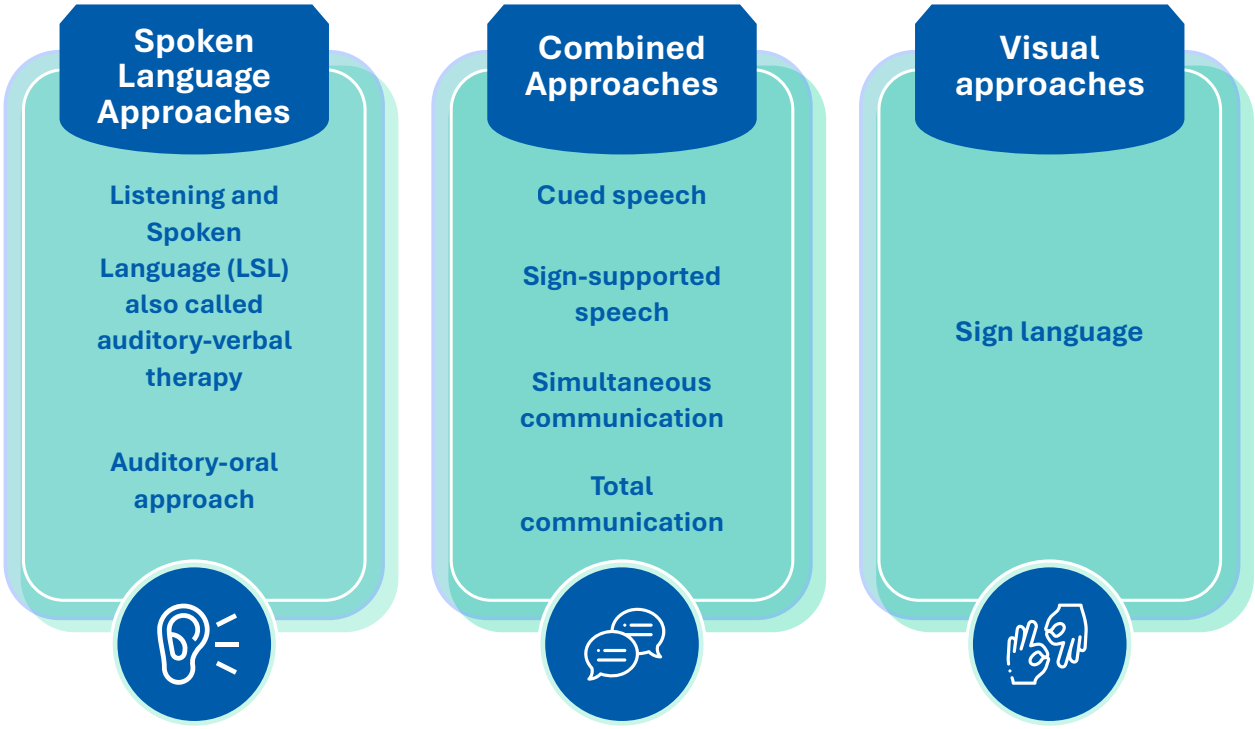

Figure 12.1: Communication approaches from most to least auditory

Within any of these intervention approaches, certain aspects may be more or less feasible for the child or the family, particularly in resource-constrained contexts and when children are identified later than recommended. In many emerging contexts, delayed diagnosis remains common due to the absence of mandated universal newborn hearing screening (NBHS) programmes. At the system level, healthcare services may be limited by constrained access to technology and inconsistent service delivery. In some instances, even after HL has been identified, hearing technologies may not be offered by governments, or the options offered may fall short of current standards of care. At the family level, competing priorities such as meeting basic needs may limit the ability to cover the costs associated with hearing technologies, therapy or travel to services. In addition, services may be unavailable or inaccessible in the family's area of residence across all levels of care. In such contexts, hearing healthcare professionals (HHPs) should strive to promote equity by recognising and responding to the

time, financial and emotional demands placed on families. Where professional-led decision making predominates, careful attention to the principles of informed consent is essential to ensure that each family's autonomy and right to self-actualisation are respected.

Madell and Hewitt (2022) highlight important questions that can be used to guide the decision-making process on which communication approach to follow:

1. What is the family's desired outcome?
2. If the caregivers choose a language with which they are unfamiliar (for example, Sign Language), can they commit to becoming fluent?
3. Will all family members be able to communicate in the selected mode?
4. Will the communication approach allow the child to feel part of the family?
5. Is the selected communication approach appropriate for that particular family and child?
6. Will the child be ready to start school with the selected communication approach?

These questions are intended to support families and professionals in reflecting on how different communication approaches align with the child's needs, the family's goals and the realities of their context. Rather than directing families toward a single option, these questions highlight the importance of informed, values-based decision-making. Regardless of the communication modality chosen, successful language development depends on sustained caregiver commitment, consistent participation and access to appropriate support. Families play a central role in creating rich language environments and their ongoing engagement is a critical determinant of developmental outcomes across all approaches.

Listening and Spoken Language (LSL) approach

This chapter focuses on supporting families who choose a Listening and Spoken Language (LSL) approach and on building a listening brain as the foundation for spoken language and higher-order cognitive skills.

The LSL approach is based on the ten principles of Auditory Verbal Therapy (AVT) (Estabrooks et al. 2016):

1. Promote early diagnosis of HL in newborns, infants, toddlers and young children, followed by immediate audiologic management and AVT.
2. Recommend immediate assessment and use of appropriate, state-of-the-art hearing technology to obtain maximum benefits of auditory stimulation.
3. Guide and coach caregivers to help their child use hearing as the primary sensory modality in developing listening and spoken language.
4. Guide and coach caregivers to become the primary facilitators of their child's listening and spoken language development through active, consistent participation in individualised AVT.
5. Guide and coach caregivers to create environments that support listening for the acquisition of spoken language throughout the child's daily activities.
6. Guide and coach caregivers to help their child integrate listening and spoken language into all aspects of the child's life.
7. Guide and coach caregivers to use natural developmental patterns of audition, speech, language, cognition and communication.
8. Guide and coach caregivers to help their child self-monitor spoken language through listening.
9. Administer ongoing formal and informal diagnostic assessments to develop individualised auditory-verbal treatment plans to monitor progress, and to evaluate the effectiveness of the plans for the child and family.
10. Promote education in regular schools with peers who have typical hearing and with appropriate support services from early childhood onward.

Within the LSL approach, caregivers are acknowledged as the primary agents of change in their child's life. They need to be empowered with skills and strategies that will support developing listening as the foundation for later talking and thinking. HHPs need to be able to support both adult and child learning during each session. The AG Bell Academy in the USA is a non-profit organisation responsible for the certification of LSL professionals. The current gold standard for therapists working within this approach is certification as an LSL Specialist, certified Auditory Verbal Therapist (LSLS Cert.AVT®).

Using an LSL approach requires significant shifts in thinking and practice. Below is an introduction to ten topics that will support this shift.

Starting early changes everything

The LSL approach holds that starting early is fundamental. Starting early means that the 1:3:6 guidelines have been met – having screened, diagnosed, fitted technology and enrolled the family in a supportive, developmental programme with a skilled therapist by the time the baby is six months old. Starting early not only changes everything for the family and infant, it also changes everything for the HHP. Supporting preverbal babies requires HHPs to gain expertise in caregiver guiding and coaching, enabling them to support their child's mastery of the next developmental skill set according to typical developmental norms. The caregiver needs to recognise what their child has mastered across many domains and have the skills and strategies to support the next developmental phase. The HHP needs to be able to guide and coach this process.

Early attachment and preverbal skills that focus on the broader aspects of brain development become important areas of focus in this early stage. Being able to hear during the development of early skills building ensures that listening is part of the neural networks that are maturing in the infant's brain. The earliest role of the HHP is to protect and impact the space between carer and infant. Hearts for Hearing refer to this as 'the heart space'. This space lays the foundation for a secure attachment, warm loving relationship and a regulated brain ready for optimal learning.

The effects of not having access to a coherent language model go beyond clear speech and good language. By starting early, the downstream effects of HL that build over time can be mitigated. When starting early there is more developmental synchrony and less of a 'gap' between chronological age and functional age (Ching et al. 2018).

It's all about the brain

The overall, foundational, neural architecture of the brain is built, primarily, within the first year of life, a critical time during which brain pathways are connecting and interlinking to form neural highways that will support cognitive, emotional and social abilities. Sensory exposure and repetition within a social relationship are what influence the wiring of this intricate connectome. The earlier a baby's brain has access to sound, the more probable the

outcome of spoken language. Conversely, the later a baby has access to sound, the less probable it becomes that functional listening can be wired into the superhighways of the brain. This is referred to as 'the use or lose it' principle. The first three and a half years of life are the critical period for language development and a child needs optimal access to sound for robust spoken language outcomes. The longer the period of hearing deprivation, the greater the chance of cross-modal reorganisation.

Importantly, delayed identification and intervention remain common in many low- and middle-income contexts, and meaningful auditory and language development is still possible when access to sound occurs later in childhood. Children who receive hearing technology or cochlear implants beyond the optimal window may follow different developmental trajectories, often requiring more intensive, prolonged and individualised intervention to support listening and language acquisition. In these cases, progress is more variable, rates of learning may be slower, and expectations must be aligned with the child's hearing age rather than chronological age. Nevertheless, neuroplasticity persists beyond early childhood, and with consistent auditory access, caregiver engagement and skilled intervention, children identified later can continue to develop functional listening and spoken language skills.

Hearing versus listening

There is a difference between hearing and listening. Hearing activates the primary auditory cortex while listening involves more whole-brain work. Listening is trying to understand what one hears. This auditory attention forms the basis of all auditory learning and progresses through a hierarchy of developmental skills discussed earlier. Factors that influence listening include:

- Age at which HL was acquired
- Age of diagnosis
- Cause of HL
- Degree of HL/amount of residual hearing
- Effectiveness of chosen technology to provide full auditory access
- Previous auditory experience

- Presence of other learning challenges
- Health of the child
- Effectiveness of audiological management
- Emotional state of family
- Degree of family involvement
- Skill of the therapist and the emphasis placed on auditory learning
- Skill of the parent
- Learning style of the child and family
- Innate learning potential of the child
- Level of sensory regulation for optimal learning: 'just right' zone

When looking at the ten guiding principles that inform the approach of AVT, six of them begin with 'Guide and Coach the parent' (caregiver). Within the LSL approach, HHPs need to collaborate with the caregiver, acknowledging that both parties bring expertise that ultimately benefits the child. HHP knowledge and experience of typical development and expertise with HL partners with the caregivers knowledge and expertise of their unique child and family. HHPs need to actively include caregivers at every stage and in every session, so that they can observe and practice the skills they need to include in their everyday interactions with their child with HL.

Adults employ a variety of learning styles. These are informed by their world knowledge, life experience, and level of motivation. As coaches, HHPs need to understand adult learning styles so that information and practice is presented in a way that best fits how each caregiver learns. The learning and coaching style of the HHP also impacts the therapy process. HHPs furthermore need to be sensitive to cultural differences that may impact the various relationships in the room: caregiver-child and caregiver-professional. Caregivers need to be motivated to implement changes. Interactions between caregivers and child should always occur in the family's home language.

Learning how to focus on adult learning, while also keenly observing the developmental needs of the child, requires focus and skill from the HHP. Seeking out both knowledge, in the form of courses or qualifications, as well as guidance from a mentor can support growth as a clinician in the LSL approach.

Caregivers need to be empowered and skilled. As Harvard University's Centre on the Developing Child states: a parent can 'provide(s) what nothing else in the world can offer - experiences that are individualised to the child's unique personality style; that build on his or her own interests, capabilities, and initiative; that shape the child's self-awareness; and that stimulate the growth of his or her heart and mind.' (National Scientific Council on the Developing Child 2004).

Listening is the foundation

Many caregivers, when starting intervention with their child with HL, are anxious to know when their child will start to talk. HHPs may fall into the trap of focusing on the production of first words from the very beginning of therapy. It is important to remember that if spoken language is the goal for a child with HL, then the initial focus needs to be on building a listening brain. Hearing is a passive, physiological process. It refers to the automatic detection of sound waves by the ear, which are then sent to the brain. Listening, however, is an active process that involves paying attention, interpreting and understanding the sounds that are heard (Cole and Flexer 2007).

Listening develops over time and follows a well-established auditory hierarchy, beginning with basic awareness of sound and progressing through several stages to reach auditory comprehension (Madell and Hewitt 2022). There are several well-known, evidence-based hierarchies of auditory skill development which can be referenced when working with children with HL:

- The Pollack Hierarchy of Auditory Skills Development
- Auditory Learning Guide
- Functional Listening Index – Paediatric (Shepherd Centre)
- Track a Listening Child
- Cottage Acquisition Scales for Listening, Language and Speech (CASLLS)

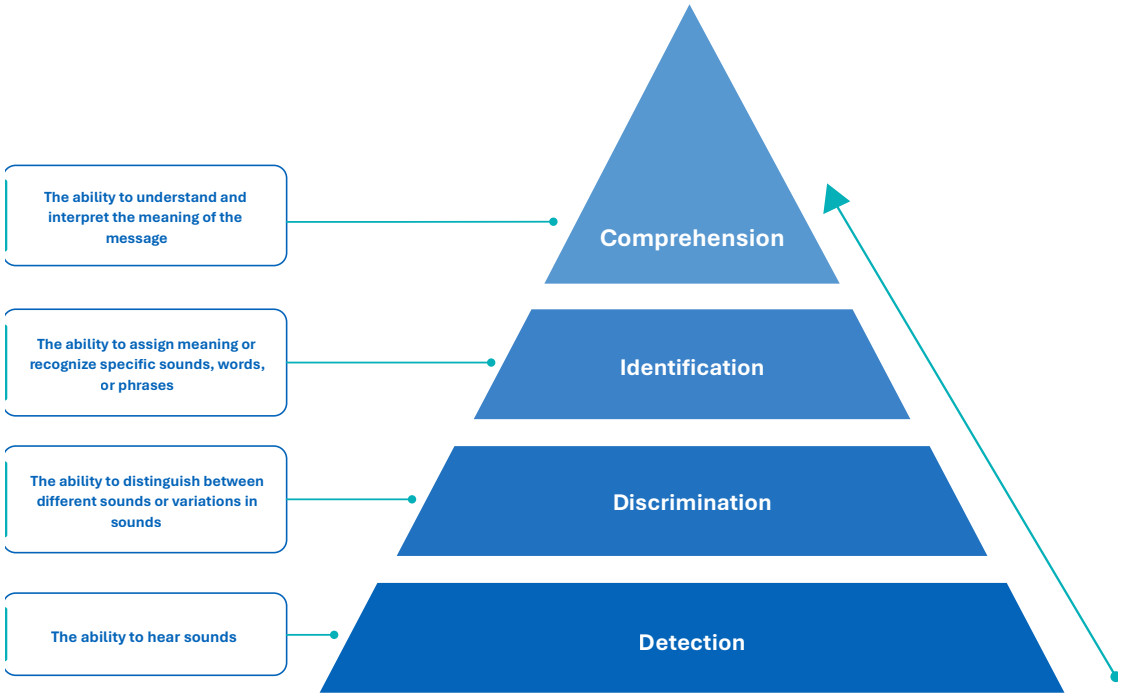

Figure 12.2: Erber's Hierarchy of Listening Skills (adapted from Erber 1982)

Erber's hierarchy (Figure 12.2) starts with basic listening skills and moves towards more complex listening. This framework can guide intervention to ensure a strong listening foundation for all children with HL. Although these auditory skills are set out in a hierarchy moving from basic to complex skills, the development is not strictly linear, with development occurring across levels. It is important seek auditory comprehension from the earliest stage possible.

Before auditory detection skills can be addressed, it is essential that children with HL have access to sound via appropriately fitted hearing technology (Chapters 10 and 11). It is necessary to ensure that the technology provides the child with full access to the speech spectrum at soft levels. The HHP needs to coach and guide caregivers on the importance of day-long listening through hearing technology. One of the first goals in intervention is to ensure the technology is being worn for all waking hours – eyes open, ears on! This sounds like an easy, straightforward process but is often very difficult to achieve. Babies, toddlers and young children are often resistant to something on or in their ears and find the hearing technology very interesting to play. HHPs need to ensure that caregivers understand the vital role that daylong

listening plays in brain development (Gagnon et al. 2021). Prioritising this issue and working on the problem together will help device retention on little ears. Caregivers should be made aware of the many retention options available to ensure that this essential goal is achieved.

Once a child has access to sound through technology, detection of sound can begin. Children need to show awareness of sounds around them, and caregivers need to support the skill of tuning into sounds by talking around their little one. Caregivers need to be coached to provide the child with rich auditory experiences in everyday routines. Babies and young children will show detection of sound in a variety of ways. Initially, a baby might look at their caregiver, stay still or widen their eyes when they hear a sound. Later, they will learn to turn towards a sound (localisation). HHPs should coach caregivers to identify their child's detection responses- These detection responses reflect the beginning of auditory attention.

Without auditory attention, little or no auditory learning can occur. If a child is not responding as expected, further investigation is necessary to evaluate sound access, wear time and the listening quality. It is also important to consider a child's level of sensory regulation, which may contribute to their readiness to listen.

The following activities can be used to facilitate the detection of sound:

- Noisemakers can be used as babies explore different sounds. It is important to connect the sound the toy makes to the toy itself (for example, 'You heard the rattle go shake-shake.'). Noisemakers should be about play and experimenting rather than acting as a constant test from which babies will soon disengage.
- Interesting sounds in the environment can be brought to the child's attention by pointing to one's ear and saying, 'I hear that', showing the child where the sound is coming from and labelling the sound (for example, 'I hear the dog barking woof-woof').
- Ling sounds and Ling-Madell-Hewitt low-mid-high (LMH) sounds can be used during play to determine detection of specific speech sounds, using their technology.
- Learning to Listen sounds (Table 12.1) can be incorporated into play to associate sounds with specific objects. These sounds will capture a baby's listening attention and help build meaning from an early age.
- Songs can be sung to encourage detection of different intonation patterns.
- Parentese (infant-directed speech), with exaggerated vowels, a slower rate and pitch variation, can be used to hold attention.

Table 12.1: Early Learning to Listen sounds

Action sounds	Animal sounds
Crying – waaaaah Sleeping – shhhhh Eating – mmmmmm Smelling – mmmmm	Dog – woof woof Cat – meow Cow – moo Duck – quack quack Mouse – ee ee ee Pig – oink oink Sheep – baa Horse – neigh or tongue clack Lion – roar Monkey – oo aa oo aa
Transport sounds	
Car – brrrm Aeroplane – aaah Ambulance/police – wooooowooowooo Train – choo choo Fire truck – neenaaneenaa *Hooter – beep beep*	

The next auditory level in the hierarchy is discrimination, where the child begins to differentiate between sounds rather than merely detecting their presence. For example, children start discriminating between sounds of different duration, pitch and intensity.

The following activities can be used with babies and young children to facilitate the discrimination of sound:

- Using music to encourage dancing and movement. The child's response to the presence of music versus speech sounds should be observed, for example, they may start moving or clapping in the presence of music.
- Turning music on and off to observe the child's response to the presence or absence of sound.
- Using Learning to Listen sounds that vary in duration, intensity and pitch to encourage discrimination, for example 'ahhhhh' for an aeroplane vs. 'ee-ee-ee' for a mouse.

Auditory identification develops when babies and young children can attach specific, unique meaning to words and sounds. This occurs prior to the ability to follow instructions or answer questions. At this level they also demonstrate the ability to imitate speech sounds. Once a child can associate a sound with a specific object or copy a speech sound, they have mastered the skill of identification.

The following activities can be used to facilitate the identification of sounds:

- Encouraging imitation of the LMH sounds and other Learning to Listen sounds. The HHP or caregiver should wait patiently for the child to take their turn.
- Using pictures or objects with the LMH sounds to see if the child can point to the correct picture when the sound is made.
- Integrating Learning to Listen sounds into daily routines and observing the child's ability to identify the sound-object pair (for example, if you say 'woof-woof', do they point out the dog).

Comprehension develops when children can follow chunks of information, including following instructions, answering questions or retelling a story. From an early age, comprehension is demonstrated through nonverbal responses, such as, blowing a kiss when prompted. It is critical that caregivers and professionals distinguish between auditory and visual responses. Children often use all available sensory input for learning; however, for diagnostic purposes, it is essential to determine whether comprehension is occurring through listening alone.

The following activities can be used to facilitate the comprehension of words and phrases:

- Using the predictability of daily routines to support understanding of common, everyday words and phrases (for example, 'Wave bye bye', 'Clap your hands', 'Hoorah',; 'Blow a kiss', 'Give a cuddle', 'Close your eyes', 'High five', 'Don't touch', 'Wait').
- Linking words and sounds together to capture listening attention and support understanding (for example, 'Let's go up-up-up and down – wheeeeeee'/'Blow a kiss – mwah'/'Give a cuddle – aaah').
- Allowing the context to support understanding in the beginning (for example, nappies always go in the bin, so 'throw it away' is supported by the predictability).
- Using action songs and rhymes daily without providing visual cues. Coaching caregivers to start singing and playing music without any accompanying actions

or movement ensures that an auditory rather than a visual response is observed.

- Sharing a book daily to practice the understanding of labels and routine phrases (for example, 'Turn the page', 'Knock-knock', 'Where's the ___?').

Talking takes time

When a developmental model of intervention is followed, the first words are typically expected after approximately twelve months of listening experience. Focusing on words without adequate listening time sabotages later language development when children need fine listening skills to hear differences in grammatical structures (for example, the difference between [s] or [z] plural markers or a [t] to mark past tense). Time spent building a strong and robust listening foundation supports later language development.

In low-resourced contexts where children with HL are typically identified late, there can be a tendency to prioritise functional language at the expense of good listening. Even when starting late it is important to move through typical development (Madell and Hewitt 2022).

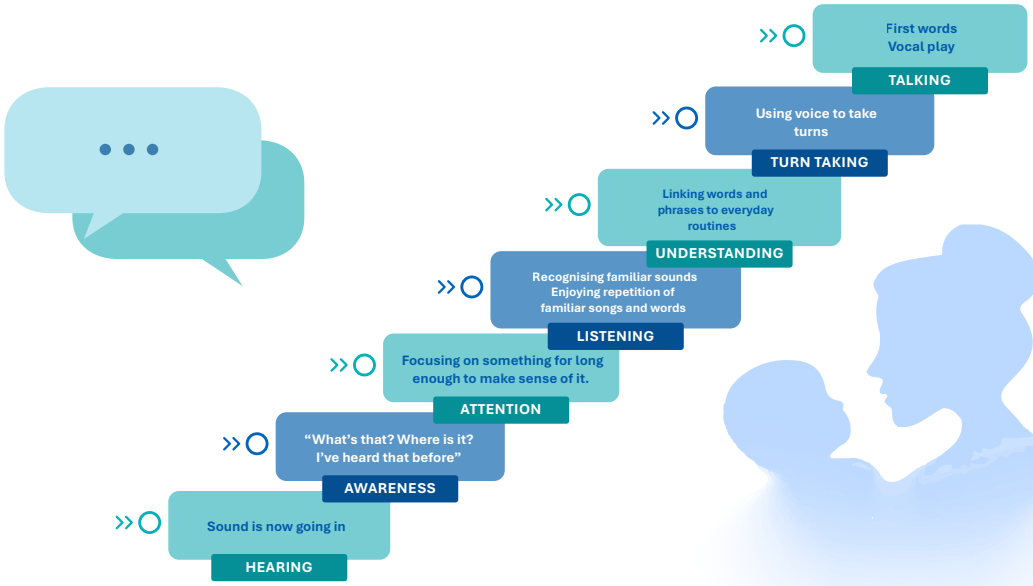

Figure 12.3: Talking takes time

Figure 12.3 highlights the steps from hearing to talking. Each step is necessary to build a rich, robust listening and spoken language system.

Focus on Stage Rather Than Age

In low-resourced contexts where many children may be identified late, a significant discrepancy often exists between their chronological age and their hearing or language age (for example, a five-year-old child with a severe HL who was diagnosed and fitted at four years of age will have a hearing age of one year). Even in high-resourced contexts, it is estimated that 15% to 40% of children are identified outside of the 1:3:6 EDHI guidelines (Madell and Hewitt 2022). Given this discrepancy, intervention should be based on a child's hearing age and current language level. After appropriate hearing technology is provided, the HHP must assess listening, speech and language skills to determine the child's developmental stage. Various free assessment resources are available that are applicable across languages and contexts:

- Functional Listening Index – Paediatric *https://shepherdcentre.org.au/how-we-help/programs-and-services/download-functional-listening-index-paediatric/*
- Integrated Scales of Development *https://www.cochlear.com/au/en/home/ongoing-care-and-support/rehabilitation-resources/scales-of-development*
- A Child's Journey (app) *https://play.google.com/store/apps/details?id=com.medel.childsjourney&hl=en_US&pli=1*

Where available and appropriate, standardised assessments can also be used to determine a child's speech and language level. Recommended tools include:

- Preschool Language Scale 5th edition (PLS-5)
- Clinical Evaluation of Language Fundamentals 5th edition (CELF-5)
- Diagnostic Assessment of Articulation and Phonology (DEAP)

Following a developmental approach ensures that no critical skills are missed and allows for the establishment of a comprehensive listening and language system. When children begin intervention later than ideal, intensive support is necessary to help them progress through developmental stages at an accelerated rate. Rotfleisch and Martindale (2023) identify six developmental stages that can guide intervention:

1. Prelinguistic
2. Singe-word
3. Emerging word combination
4. Communication with child-like errors
5. Competent communicator
6. Advanced communicator

Strategies to Teach Caregivers

LSL specialists aim to coach and guide families in developing their child's listening and spoken language through the implementation of specific strategies (Fickensher, Gaffney and Dickson 2016). A strategy is a specific plan used to achieve a goal. When working with a child with HL, it is essential that the therapist knows and is able to coach specific, evidence-based strategies (Table 12.2). These strategies are intentional methods used to achieve specific goals in auditory development. Although weekly intervention sessions with a skilled therapist are essential for children with HL learning to listen and speak, it is often said that therapy begins as the family leave the session. Caregivers need to be equipped with essential strategies to carry over the therapy targets at home and make them part of their everyday lives. Christina Prodinger and Jayne Simpson Allen from Medel highlight fifteen essential strategies that caregivers need to be taught during the sessions (Med-EL Medical Electronics 2025). Sessions are divided into three broad categories, namely getting ready, listening and talking. Strategy cards can be downloaded at *https://blog.medel.pro/rehabilitation/15-strategies-to-develop-listening-skills-speech-at-home-with-children/*

1. GETTING READY: Strategies to get ready to listen each day:	
1.1 Eyes open, ears on	Ensure that a child's hearing technology is on and in working order during all waking hours.
1.2 Come close to me	Stay close to the child while speaking to them to ensure the best access to sound. This may mean going down to the child's level.

1. GETTING READY: Strategies to get ready to listen each day:	
1.3 Reduce background noise	Make the room as quiet as possible by turning off any background noise.
1.4 Same thinking place	Follow the child's lead and talk about what they are focused on.
2. LISTENING: Strategies to develop a child's listening skills	
2.1 Talk, talk, talk	Narrate life. Talk about what you see, hear, do and think to surround the child with language.
2.2 Acoustic highlighting	Make certain words or phrases stand out using emphasis by speaking softer/louder, pausing, longer or using a different pitch.
2.3 Wait, wait, wait	After asking a question or giving an instruction WAIT to allow the child time to process and respond.
2.4 Auditory hooks	Get the child's attention through listening by using interesting sounds like 'wow', 'ah', 'look'
2.5 Listening first	Give the child information through spoken language first before adding any visual cues or gestures.
2.6 Say it again	Repeat or rephrase part of the message if the child has not heard or understood.

3. TALKING: Strategies to develop a child's spoken language development	
3.1 My voice matters	Recognise every communication attempt and acknowledge every sound or word they make.
3.2 Auditory closure	Use familiar sentences or rhymes and then leave off the last part for the child to complete (for example, 'ready, steady....').
3.3 Expansion and extension	Repeat what the child says but add more information or correct the sentence structure or grammar.
3.4 Use choices	Ask questions with choices so the child can use their language to make decisions.
3.5 Sabotage	Create situations where the child needs to ask for help or to solve a problem.

Monitoring Progress Is a Professional Responsibility

LSL practitioners need to supply regular and ongoing evidence of how the child and caregiver is progressing across a range of skills. This monitoring helps to understand the child and family's rate of learning and identify the level of support needed to master new skills. Knowing progress over time also allows for timely referrals to other professionals if needed. Rossetti's Patterns of Change monitor how the gap between chronological age and functional age is shifting over time: is the gap closing, widening, staying constant or fluctuating? Each interaction with the family provides diagnostic information, which, when collated over time, shows the progress made. It is recommended that each child have a baseline assessment with six-monthly follow-up evaluations. Having a protocol that guides assessment intervals aligns with international standards of care. Indicators that a child may need more support include:

- Insufficient calm-alert moments
- Disengagement in the presence of warm, loving relationships
- Lack of spontaneous orienting to face-to-face interactions
- Poor visual attention
- Disengagement in play and communication interaction
- Lacking in curiosity about the world
- Not acquiring nonverbal intentional communication behaviours
- Overemphasis by adults on establisihng joint attention in play
- Not developing nonverbal skills of turn taking, imitation and matching
- Not persisting in getting needs met or in overcoming appropriate challenges
- Poor sensory regulation: impulse control or emotional control
- Delays in innate vocal development
- Global developmental delays
- Plateaus or regression in progress

When children show slower than typical progress with listening, collaboration with the HHP who fit the hearing device is essential to ensure that the child has access to all sounds across the frequency range. Both threshold levels to tones and responses to speech are important. Data logging, or wear time, plays a significant role in how quickly or well children build auditory attention that supports understanding. Families who struggle to attend regular sessions, who are challenged by integrating new skills into their parenting styles or who do not have access to specialised intervention services may progress more slowly. This also holds true for children with additional needs beyond HL.

The impact of HL extends beyond the ability to speak clearly. Research proves that the presence of an HL can have an impact on higher-level cognitive skills, including theory of mind and executive function skills. Theory of mind refers to the ability to put oneself in the mind of another and is essential to develop empathy and maintain healthy relationships. Executive function refers to a set of brain-based processes responsible for the active management and control of cognitive resources, emotions and behaviours. Executive function allows for planning, organising and achieving goals (Barkley 2012). These skills also include aspects of working memory, time management, impulse control, emotional regulation and flexibility. When working with children with HL, it is essential to incorporate activities that develop theory of mind and executive function skills from an early stage.

Conclusion

HHPs may hope for sessions to unfold flawlessly, but true growth occurs through continuous learning and reflection. Each family provides new insights and opportunities to enhance clinical practice. By staying open to change, nurturing curiosity and maintaining high standards, professionals can ensure that children with HL receive the best possible support. When HHPs do what it takes, children thrive.

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Chapter 13

Visual Communication Options and Educational Strategies for Children with Hearing Loss

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"The ever-growing issue of public, paediatric hearing loss is a barrier to quality education for deaf and hard of hearing children'

~United Nations Sustainable Development Goal 4, 2015

Introduction

In social and cultural settings amongst children and adults with hearing loss (HL), individuals are defined as either being hard of hearing (HoH), small deaf or big Deaf. Individuals who are HoH usually have sufficient hearing with or without their hearing aid (HA) or cochlear implant (CI) to cope with daily verbal communication. Individuals with HL who define themselves as being deaf embrace technology such as HAs, often use speech to communicate and mingle mostly in the hearing world, though some are also comfortable in the Deaf world. Deaf individuals are those with HL who are totally immersed in the Deaf culture, embrace sign language (SL) as their home language and do not see the need to use HAs or CIs – though some do. In this chapter, the use of HoH/deaf/Deaf will be used only if it is warranted according to the visual modality discussed. Otherwise, the generic term of deaf will be applied to all children with HL.

Irrespective of the language or communication modality chosen, the role that caregivers and family play in the life of a child with an HL is critical. The difference between a caregiver's involvement and their commitment can have a big impact on the child's language development, especially in the critical early formative years. Caregivers who opt for either the Oral/Aural route or the Visual Communication route (SL, Cued Speech, Signed Exact English, Total Communication, etc.) need to be aware that any communication modalities require effort on the caregiver's part to support intact language development in their child if they wish them to keep up with their hearing peers in terms of language, social-emotional and academic milestones. A significant challenge for deaf children born to hearing parents is not only the potential language delay but also the stress experienced as they try to select appropriate communication modes based on how those around them react and respond to their speech only, sign only or sign and speech combined (Lillo-Martin 2016).

A fascinating finding is that both auditory and visual language is processed in the same area of the brain – the auditory cortex – previously thought to be the place of processing sound only. Languages have distinct patterns, whether expressed through the lips or hands, that deaf children must consistently access to develop the internal language skills necessary for reasoning, thinking and communication (Petitto 2000).

Visual Communication options are often only explored when it becomes clear that the child with HL cannot cope effectively in a listening and speaking environment, despite HAs or CIs, making spoken language acquisition challenging and frustrating. For some families it is the only option available to them due to factors like late identification of HL or a family's financial situation. Visual Communication implies that the eyes are used during communication, just as auditory communication suggests that the ears play a major role. There can be a grey area between where both senses are used or needed to communicate effectively – think about a hearing person trying to have a conversation in a noisy environment – it is highly likely that both ears and eyes are being used to help make sense of input. Many may be surprised at how often they unconsciously lip-read while listening, even though they may hear very well.

Each of the different Visual Communication options will be discussed in terms of supporting language development and use in education. All the options have their strengths and weaknesses, which is why some children will do better in one of these languages or modalities as opposed to another, depending on the child's learning style and degree of HL. Essentially, caregivers and therapists need to be encouraged to explore and find the modality that is best suited for the child and the family.

Manualism: South African Sign Language (SASL)

The National Institute on Deafness and other Communication Disorders (NIDCD) defines SL as a complete, natural language that has the same linguistic properties as spoken languages but with grammar that differs from English or other spoken languages. SL is not universal. American Sign Language (ASL) differs in many signs to SASL (South African Sign Language). However, some signs may be universal. SLs are also regional just as some spoken languages have different dialects and different slang words.

The parameters of any language are those features used in its production. For example, spoken English has long and short vowels, hard and soft consonants and diphthongs. SLs are soundless and manual; as such, their five parameters (handshape, palm orientation, placement, movement and non-manual features) define the use of the hands and the body to express the different functions. For example, English speakers raise the pitch of their voice when asking a question. In SASL the question is asked by raising of eyebrows, widening of eyes and slightly tilting forward.

SLs were birthed out of the need to communicate in a world of silence. For centuries, SL was not officially recognised as a language and was stigmatised. Today SLs are recognised around the world. In South Africa, SASL was recently adopted as the 12th official language.

Benefits

There are potentially no communication barriers for a Deaf child since communication is not dependent on hearing, providing that communication partners are also proficient in visual or signed language. The Deaf community has a strong identity in their deafness, their culture and their language, and does not regard deafness as something to be fixed. For the Deaf community, SLs create social networks and relationships between Deaf people across nationalities and cultural communities, binding the Deaf community together in a common language of familiarity, sociability and communality (London 2020). SLs and are also an identity marker for Deaf people from diverse backgrounds. Thus, the ability to use South African Sign Language (SASL) as a mode of communication provides Deaf learners with several important benefits (McIlroy 2008):

- Access to the Deaf community and cultural identity.
- A means of communication both socially and in the classroom.
- Increased confidence and a sense of belonging as valued learners.
- Improved communication with teachers, which enhances learning outcomes.
- Strengthened relationships between hearing teachers and family members.
- Support for a bilingual-bicultural identity, reinforcing both SASL and literacy development.

Deaf children born into families where either one or both parents are Deaf (and fluent in SL) are exposed to the natural SL of the home. For the most part, these children reach similar milestones as their hearing peers and arrive at school with age appropriate language skills.

Not all caregivers who choose to use SASL as a communication modality ever reach an appropriate level of SL skill to meet the ever-growing language needs of their deaf child. Those who do are determined to acquire SL as quickly as possible. Some do so naturally, but for many parents, learning a new second language as an adult is not easy and requires tremendous self-discipline and effort. Those parents who master SL enjoy good communication with their children and are involved with supporting them both academically and socially-emotionally as they learn from each other. Their children, for the most part, reach the same expressive language milestones as their hearing peers, the difference only being the modality. They are able to think, reason and enjoy the same benefits that language has for intellectual and cognitive development.

Challenges

Parent guidance support is needed to encourage first language development and teach strategies to caregivers to ensure all Deaf children arrive at school with a language age on par with their hearing peers. Not only is learning another language at a later age a challenge for parents, but they also need to be supported in how best to accelerate learning and use SL effectively with their baby or young child. Despite the ease of communication that can stem from using SL, the complexity of Deaf Education is highlighted by Deaf learners who come from diverse visual, signing, literacy or even oral backgrounds (Garcia and Cole 2014). McIlroy (2008) emphasises that there is no singular, universal deaf narrative, but rather a multiplicity of personal journeys that affirm the diversity and complexity of the struggles that different deaf individuals experience. It also highlights the possibilities available to Deaf learners to overcome difficulties regarding language and literacy learning (Böhringer 2021).

The limitation of caregiver proficiency in SL means their child may not receive the appropriate level of communication and increased language input in the home environment (Pontecorvo 2024). In addition, family members and friends need to learn SL too, sincethe emotional well-being and social network of the deaf child depend on their willingness to do so. However, acquiring SL may prove to be a challenge for them. The issue is compounded further in that most schools for the Deaf are residential, meaning that new learners' SL skills develop rapidly and effortlessly through being immersed in a hostel with Deaf hostel parents, Deaf peers and those older Deaf learners who are already fluent and able to model conversational signs. Furthermore, many learners do not go home every weekend; thus, the signs they learn at school do not necessarily carry over into the home. It is for this very reason that learners who are unable to communicate effectively at home say they feel more at home at school because they cannot communicate with family members and are isolated during the holidays. This can cause tremendous emotional stress on the caregivers, siblings and the deaf child.

Academic support

In South Africa, most of the Schools for the Deaf fall under the purview of the Department of Education and follow the national Curriculum Assessment Policy Statement (CAPS) curriculum. The Home Language SL document, for the most part, mirrors that of spoken home language documents with some semantic changes to accommodate the difference between a Signed and Spoken language (for example, 'Listening and Speaking' becomes 'Viewing and Signing'). Provided that the Deaf learner has access to early intervention prior to coming to school, it is assumed that they will progress at the same rate and level of hearing peers. However, due to the severe language delay that many Deaf learners present on arrival to school, the necessary academic and language support required to make allowance for the gaps in language development are not necessarily covered in the CAPS curriculum. While the content of subjects and concepts can be readily accessed through signing, the need to address low world-wide literacy levels amongst the Deaf remains the most urgent challenge.

Most schools for the Deaf teach SASL as a Home Language and English as a First Additional Language through writing. This presents its own set of challenges as learners have to express themselves in a language they struggle to access, internalise and express in a written format. It furthermore causes barriers to academic development as it depends heavily on the skills and ability to master reading and writing. However, with concessions like SASL interpreting or

adapted question papers and extra time allocation, some learners go on to achieve amazing results.

Auditory–Visual Approach: Cued Speech/Cued Language (CS/CL)

Cued Speech is a visual system of communication that uses eight handshapes in four locations near the mouth, in combination with the information visible on the lips, to make a spoken language clear through vision alone (https://cuedspeech.org/). It supports visual and auditory access to a spoken language. Used in any setting, Cued Speech/Cued Language (CS/CL) is a lesser-known visual communication tool that supports a spoken language. It was designed to aid lip-reading by removing the confusion of similar-looking speech sound patterns or phonemes on the lips, resulting in accurate visual receptive access by the deaf child to the intact, complete and meaningful units of a spoken language in all its forms, without needing speech or hearing (LaSasso 2010).

Benefits

Using CS/CL provides visual access to the spoken language for children with a severe to profound HL who struggle to access speech sounds despite HAs or CIs. It takes two weeks to six months to master the cue-sound associations for the forty-four sounds used in the English language. Caregivers are not learning a new language – it is merely a tool for making the language they already know easily accessible to the deaf child. A child consistently exposed to a cued language visually internalises the language in its intact form in the same manner a hearing child does through audition, and benefits from being able to better lip read than those who do not cue (or sign).

Challenges

As with all communication options, the benefits of CS/CL are dependent on those willing to master it and use it optimally for the benefit of the deaf child. It is not widely known and many in the Deaf community feel threatened by it, fearing that SL will be replaced by CS/CL. CS/CL was designed to support visual receptive access to a spoken language like English. As with all communication options, not all deaf children may benefit, especially older learners set in

their ways and those with barriers to learning. It can take up to three years for full benefits to be seen. As with all languages, the earlier the exposure the better.

Academic support

Using CS/CL with a deaf child is versatile in that it can be used in any setting – be it in the home, in a classroom with hearing children, in a classroom with deaf learners as manual bilingualism or in therapy sessions. Being able to communicate both at home and at school has positive emotional-social implications. The use of the word 'speech' in CS does not imply that the deaf child has to speak. If the child wishes to learn to speak, they will still need speech therapy since the cues do not show how to produce the speech sound, but simply which sound is being said in a motoric sequence of sounds that make up a word. However, the speech therapist can cue the target sound clearly and teach the child how to make the targeted sound, leading to natural speech sound – known as sound-cue association. Some deaf children do, in fact, start talking voluntarily when they have the confidence of knowing what sound to make and when to say it in a word, as there is clear reception and no guessing.

The cues prime the brain on what to listen for and takes away the guesswork and confusion of lip reading alone. We constantly encode what we write (from speech sounds to letter-sound association) and decode what we read (from letters to speech sounds) and thus the words and phrases are said aloud. It is a skill desperately needed by young deaf learners to support them on the ladder to literacy to aid phonological awareness, syllabification, fluency, vocabulary and comprehension (Kyllo 2010).

Bilingualism: Bi-Bi (Bilingual-Bicultural)

The Bilingual–Bicultural (Bi-Bi) approach incorporates the use of SASL as the Language of Learning and Teaching (LoLT) in the classroom (Böhringer 2021). English or another spoken language is taught through reading and writing print. The deaf culture, its history, values and customs are also taught. Strong emphasis is placed on developing a healthy sense of self through social contact with other Deaf individuals.

Research on bilingualism has birthed different approaches and new terminology. Bimodal Bilingualism is bilingualism involving two modalities, such as SL and a spoken language. This

generally refers to a CODA (a Child of Deaf Adult/s) raised in a family with one or more deaf parents but is hearing, not deaf. A CODA can code-switch and/or code-blend. It can also refer to a Deaf child who is fluent in both signing and in a spoken language like English through writing or speaking. Emergent Bilingualism is when learners continue to develop their home language while also learning a second language. Simultaneous Bilingualism applies to very young children who are exposed to two or more languages at the same time and become fluent in both. More recently, the phrase Translanguaging has come to the fore. This is a process by which multilingual speakers use their various languages at different times to convey meaning in a conversation, and use the most appropriate word in any of their known languages to communicate.

Benefits

Relevant for Deaf Education is the Bi-Bi approach, which stresses learning two languages – SASL and English/Afrikaans, and two cultures – the Deaf and the hearing world. A bilingual learner has a better opportunity of integrating into the mostly hearing world. Being able to communicate in both SL and in the spoken language – either through speech or writing – is a benefit and essential to be employable. Being able to communicate in the language of the home will also go a long way in breaking communication barriers and providing the emotional-social support a deaf child desperately needs.

Challenges

The SASL CAPS document encourages educators of the Deaf to fulfil the mandate that 'SASL and English is to exist independently but to be parallel in status' (Department of Basic Education, 2014). Currently, the two languages do not exist independently, since SASL is used to support teaching in English. The patterns embedded in English are not accessed easily as they are overshadowed by the structure of SASL. Teacher skills in SASL (a different language to Signed English) and teacher understanding of the cultural identity of the Deaf community impact the success of the programme. The when and how of introducing English is still a controversial topic. Teaching English through reading and writing is not conducive to natural language acquisition.

Hearing children come to school with an intact language that they use for the task of learning to read and write. This is not usually the case for the severely delayed language Deaf learner (Kyllo 2010). These children do not benefit from the usual crossover that a Home Language learning of phonics and phonological awareness offers to a second spoken language, as SASL is a soundless, spatial language. Sadly, most Deaf learners do not achieve age-appropriate literacy skills as they struggle to master a language they cannot hear and understand, but are required to read and write without first being able to internalise the language through natural face-to-face interaction during pre-school years.

Total Communication (TC)

TC is an approach (or philosophy) to communicating that aims to make use of all means of communication, such as using hearing technologies to encourage access to a spoken language, including the use of listening, speechreading, speech, natural gestures, body language, fingerspelling, signing, written and visual aids, and in some programmes, CS/CL, depending on the particular needs and abilities of the person. The selection of specific communication modalities is to support the unique needs of the child and their family. A child may require more of an auditory/oral approach to listening and speaking, while another may benefit more from watching and signing. The teacher could also vary input that is at times only oral and at times only sign-supported.

Benefits

TC rests on the premise that every deaf/Deaf child's communication needs and receptive access to a spoken language are different and may require different tools to develop language and literacy competence. TC is also used to address communication difficulties associated with a range of additional conditions like neurological problems, autism, learning difficulties and physical disabilities. Today, hearing technologies are much more advanced so that more deaf children are able to access a spoken language effectively and are less dependent on sign systems. An increasing number are even able to cope in mainstream schools.

For natural language acquisition using any of the modalities discussed thus far, the language used needs to be accessible, and the parents and other members of the deaf child's social circle should be able to use the language effectively in face-to-face communication. Only then will there be the relevant volume of interaction needed for language, cognitive, and

emotional development to occur (Mayer and Trezek 2023). Caregivers need to have a clear understanding of the difference between a language and a modality, that is, to choose the language they want to communicate in before choosing the modality that supports access to that language. It is important to help caregivers understand that access to a spoken language can be supported visually.

Challenges

Hearing technologies still limit access to effective auditory input for profoundly deaf learners and most TC programmes lean more to the visual-manual component (Mayer and Trezek 2023). The success of these programmes also depends heavily on the teaching skills of the teachers in the classroom – some make more use of simultaneous English-based signing, and some use more of the oral component, both of which may omit access to the critical grammatical structures of English because of less-than-optimal hearing. Unless it is clearly stipulated that English is the desired language and signing is used as a modality – namely, signing some key words or signing everything – the integrity of both languages can be corrupted. Neither is it then made accessible in its intact form.

SASL has its own grammatical structure that is very different to English. Additionally, there are concerns for exposing learners to a form of simultaneous English-based signed communication that may not match their level of reading or language readiness, especially in the early formative years. It is important to determine whether the programme offered matches the needs of the deaf child.

Educational strategies

Historically, educationally significant HL ranged from a moderate to profound degree, but more recent research shows the vast impact of minimal and mild HL as well as unilateral HL on academic performance (Winiger, Alexander and Diefendorf 2016). Please note: In this section, the term ‘caregiver’ or ‘family’ is used as many children are raised by carers who may not be their biological parents, due to cultural, financial, health or employment choices.

To address the diverse communication needs of children with HL, several education/schooling options should be available, including inclusion in mainstream, facilitation in mainstream or special educational needs. In an ideal world, these schools would provide an academic

curriculum using SL, spoken language and TC. However, realistically these options are not widely available, and if they are available, they are often not accessible to those who need them. In struggling education systems, particularly in low-middle income countries (LMICs), the availability of various educational options may not be a reality. This often results in special needs boarding schools for deaf learners, which are scattered across the larger cities in these countries. Listening and spoken language (LSL) schools, traditionally known as oral/aural schools, are often dependent on the availability of technology, while schools for the Deaf, primarily using SL, may struggle to find educators proficient in SL. In either situation, family commitment to language learning and education is pivotal for the child's success. These pitfalls can result in limited access and poorer education levels for children with HL.

Inclusive education

Inclusive education is the integration of learners with disabilities into general, mainstream education, with suitable support and assistance, to meet their different abilities, learning styles and backgrounds (Li and Potmesil 2022). Mainstreaming usually refers to the physical placement of a child with special needs in regular classrooms with typically developing children. Inclusion is a broader term referring to a more complex process that involves adaptation to each child's needs within the regular classroom (Armstrong, Armstrong and Spandagou2011).

Special needs

A school for learners with special needs caters for children with learning, physical and behavioural difficulties. Generally grouped with children of similar needs, this education system should be adapted to suit the unique learning styles required while customising the curriculum. Special needs schools are known to foster a sense of belonging. It can be beneficial to place a child with HL who is learning to listen and speak in a special needs LSL school for their foundation years and, if appropriate, transition them to a mainstream school for primary/middle and/or high school education. However, this is dependent on careful consideration and support from the family, healthcare and education teams.

Individualised Education Programme

An Individualised Education Programme (IEP) serves as the foundation for providing a high-quality education to every child with a disability. Regardless of whether a child with HL is educated in a mainstream or special needs education system, an IEP is essential to keep the team accountable and working towards appropriate, current, shared goals for the benefit of the child's development. The IEP should be reviewed at least every six months for reflection on successes and challenges and for planning ahead. This meeting should be open to all team members and must always include caregivers.

Factors Affecting Choice of Education

Age of diagnosis

The age of diagnosis has a significant impact on the deaf or HoH child's education options – most notably for the congenitally deaf child. Newborn hearing screening (NBHS) is the first step to early hearing detection and intervention (EHDI) which is 'a vital predictor of future language and educational outcomes' (Moeller and Tomblin 2015) (Chapter 4). Before age three years, the brain will expertly form neural pathways to establish a communication system, be it visual or auditory. However, should the brain be exposed to visual stimuli only (because an infant is born or acquires HL early in life) it is difficult to alter the mode to auditory communication after age three (Kral and Sharma 2012). Children with early-onset HL living in countries where NBHS is not implemented have a later age of diagnosis. This, in turn, affects their audiological management opportunities and, of course, success in spoken language programmes. Some CI programmes have limits for the age at which they will implant a child with congenital or early-onset HL (anything between two and five years of age) due to resource constraints and proven poorer outcomes with later implantation. These strict candidacy criteria are particularly prevalent in LMICs (WHO 2018). Along with technology, (re)habilitation is key for deaf or hard-of-hearing children whose families have chosen spoken language as their communication mode.

Home language

Home language may differ from the language of education for learners with HL who attend special needs schools. The limited number of special needs schools for deaf children means

that the language of choice is generally the most established language in that country. For families who speak a minority language, this may mean educating their child in a second or even third language or perhaps a language unknown to the caregivers. This language barrier has implications for communication between educators and caregivers, while also posing a difficulty for caregivers in assisting learners with homework and carryover. In these cases, schools may need to offer homework opportunities after contact time at school, provide correspondence to caregivers in alternative languages and have interpreters available for meetings between families and educators.

Caregiver support

Due to the frequency and length of a child's school career, it is essential that caregivers and teams have realistic, achievable plans in place for the child. These plans are primarily the caregiver's responsibility and may include reliable, safe transport arrangements, after-school care, adult supervision for homework, and availability to meet teachers and team members. Caregivers may need to join therapeutic sessions for effective carry-over at home and engage in additional homework in speech-language therapy or occupational therapy. They may also need to consider longer, more expensive transport options if the school is far away from home (in the case of attending special needs education). These additional requirements necessitate financial supplementation— it is expensive to educate a child with HL on top of maintaining the expensive hearing technology. In education systems where special needs support is lacking, caregiver involvement is key to the academic success of the child. Incidental learning, through overhearing, accounts for up to 90% of what children learn (Madell 2016). However, for children who are deaf or hard-of-hearing, incidental learning is not guaranteed, even with appropriate hearing technology.

Ecosystem around the Child with HL

Team

From the time of the HL diagnosis, a team should develop around the child. Caregivers are central to this team and must be empowered to manage their child's hearing and educational access, so that the child's abilities are promoted and their needs met at each stage of their academic career. This therapeutic/healthcare-based team usually includes some, if not all, of the following professionals: An audiologist, speech-language therapist, early

interventionist/teacher of the deaf, paediatrician and ENT specialist (Chapter 14).

Some children may require additional assessment and management by an occupational therapist, physiotherapist, paediatric neurologist, ophthalmologist or geneticist. The advisory team should not be so big that responsibility is diluted and management not carried out, but also sufficiently informed that all aspects of the child's development are considered. The child's team will change over time due to their developing needs. Due to the wide range of academic, linguistic and social-emotional needs of a school-age child with HL, no one discipline can provide all the required input (Gustafson, Newsome, Pilling and Segura 2024). The term 'educational audiologist', which is not widely used, refers to an audiologist based in the education sector who ensures a child with HL has optimal and consistent auditory access in class through acoustic accommodations and remote microphone technology. Hearing Resource teachers, Itinerant Teachers, Teachers of the Deaf or Peripatetic Teachers refer to teachers with additional training who are involved in the education of children with HL (Estabrooks, MacIver-Lux and Rhoades 2016).

Acoustic accessibility

To maximise the learning potential for a child with early onset HL who is learning to listen and speak in a special-needs or mainstream education environment, optimal acoustic accessibility must be provided. This can only be made possible through hearing technology for children who are deaf or hard-of-hearing. The reciprocity in the teacher-child relationship is vital for the success of children with HL in the mainstream environment. Children and their families will affect teacher expectations and the teacher's beliefs and knowledge will affect the child's success of inclusion. Special needs classrooms for deaf and HoH children require established daily routines, time-based schedules and low child-to-staff ratios. In either education system, the child's level of communication should be acknowledged, understood and accommodated by everyone working with the child.

Children with either unilateral or mild HL can experience similar educational barriers to those with bilateral HL. Unilateral or one-sided HL affects the ability to localise sound in in the presence of background noise. This lack of localisation has a negative impact in the classroom, which is a naturally occurring noisy environment, and on the sports field where direction of sound is important (Winiger, Alexander and Diefendorf 2016). Learners with mild HL face comparable risks of poor psychoeducational outcomes to those with moderate and severe permanent HL (Shafer, Kirby and Miller 2020).

The acquisition of routine and play-based skills promotes academic success as children move through the stages of schooling. Language and social skills spill over into academic skills. For example, early childhood maths skills are used in solving simple story problems in a real-world context, and in understanding specific times for particular routines. Therefore, these skills need to be taught to the child with HL who may have missed the language cue.

Listening fatigue

Listening fatigue is caused by the increased effort in cognition required to process and understand speech. Contributing factors to listening fatigue are the degree of a child's HL as well as background noise. For a child with HL to understand and process speech in the noisy classroom setting, there will be a greater demand on cognitive resources. Over the duration of an academic day, this increased mental effort can have a cumulative effect. According to McCreery (2015), 'These challenges can place children with HL at a greater risk of academic difficulties and even long-term health problems'.

Remote microphone technology

Distance, noise and reverberation negatively impact everyday listening situations. The ideal distance for good speech understanding is approximately 1.5 metres from the hearing technology microphone to the speaker's mouth (Wagener, Vormann, Latzel and Mülder 2018). Maintaining this distance is not feasible for children on the move and certainly not realistic in an education setting. To ease complex listening situations for deaf children using hearing technology, a remote microphone (RM) is an essential accessory that improves speech understanding and educational performance in the classroom. The signal-to-noise ratio (SNR) is used to measure the level of a speaker's voice in relation to the background noise. With technology improvements, RM is available for many listening requirements. Even children with typical hearing but additional listening needs (for example, auditory processing [Chapter 8] and attention deficit disorders) can benefit from RM technology.

Classroom environment

The classroom is a noisy environment for all children, and it creates significant listening challenges. An immature auditory system struggles to process spoken language from teachers and peers. Classroom noise is caused by learners' movements and speech, environmental

sounds outside the classroom (traffic, construction or gardening machinery) and operational equipment (air-conditioners, projectors, fans, computers). High levels of background noise will mask the teacher's voice and degrade the speech components. It is well known that 'children with HL perform more poorly in noise than children with typical hearing' (Estabrooks MacIver-Lux and Rhoades 2016). Increasing the SNR is the ultimate goal, but reducing background noise sources is the most cost effective and practical. Table 13.1 presents environmental modifications and instructional strategies that can be helpful in the classroom.

Table 13.1: Classroom modifications and instructional strategies

Classroom modifications	Instructional strategies
Turn off equipment that is not in use to reduce low-frequency background humming, for example, projectors and fans.	Rephrase questions or answers from both teachers and peers for clarity.
Close windows, schedule maintenance outside contact hours and move children with hearing technology away from sound sources.	Ask open-ended questions to clarify whether the learner heard correctly, for example, "What did you hear?"
Reduce reverberation—the persistent sound energy that continues after the initial sound source has stopped—by using sound-absorbent materials.	Provide new vocabulary before the lesson, for example, before learning about different angles, provide the child with words like 'obtuse', 'acute', 'reflex' and frequently repeat new vocabulary items.
Install curtains, carpeting and tennis balls or soft pads on chair and desk legs.	Use a natural vocal loudness to reduce distortion caused by overly loud speech.

Classroom modifications	Instructional strategies
Add acoustic panelling, sound tiles, corkboards or even egg boxes to ceilings, walls and shelves.	Reiterate information presented in videos or clips using visual or auditory formats, or ideally both.
Plan strategic seating arrangements for children with HL. Place them away from noise and as close as possible to the person who speaks most frequently—generally the teacher.	Give visual examples of new sounds, words or concepts, for example, a written word with a picture or symbol.
Begin each day with a listening check, as this is essential for children using hearing technology. Ear-specific assessment of each device ensures that the child can detect and/or identify speech sounds across the frequency spectrum, for example, the Ling–Madell–Hewitt 10-sound test.	Gain the child's attention, make eye contact and avoid speaking from a distance or with your back to the learner.

Distance is a critical factor for children using hearing technology. The distance that sound energy needs to travel from a speaker's mouth to a listener's ear influences the loudness of that message. Thus, the greater the distance, the less intense the sound, the poorer the speech perception. This can be difficult to control and consistently monitor, but in the absence of RM technology it becomes one of the single most important acoustic modifications that can be made in the classroom. Putting the child as close to the teacher (or speaker) as possible will limit speech energy loss and promote speech understanding (Estabrooks, MacIver-Lux and Rhoades 2016).

Instructional strategies

Acoustic and classroom modifications cannot entirely solve the difficulty of speech understanding in the classroom. Strategies implemented by teachers, assistants, coaches, friends, parents and the child themselves can greatly improve the ease of listening for a child with HL and/or hearing technology. It is the responsibility of the child's team to ensure all those working with the child understand their hearing levels with and without technology, their receptive and expressive speech and language ability as well as additional difficulties.

Preparatory strategies are essential in inclusionary style learning for peers to understand the needs of the deaf or HoH child. Peers should be encouraged to support deaf or HoH learners by speaking one at a time, raising their hands or saying their own names for the deaf learner to localise the sound source, avoiding interrupting, using visual support to explain new topics, making lips and faces visible when speaking and being prepared to repeat themselves when required. These strategies can be facilitated through awareness and training from class inception, for example, through a demonstration of the senses and how they work, discussions about disability and how we all differ, reading books and stories about children with HL and even simulating degrees of HL to foster understanding and compassion.

Self-advocacy

Self-advocacy is an essential skill for children with HL. There will be times in their lives when those around them are unaware or apathetic of their needs. For the child to hold a strong self-concept and belief, they need to observe advocacy from their community from a young age. Like with all learned human behaviour, the way in which their caregivers respond to questions or reactions about their HL will determine their own response to it. All children must possess basic self-worth before feeling sufficiently secure to respond to questions or actions regarding their HL and/or technology.

It is imperative that children participate in social and academic opportunities at school — that is in fact inclusion. A child's success in social situations will be largely determined by their language skills as well as soft skills, which rely on the effective development of executive function and non-verbal social communication. These social skills required for interpersonal relationships often need to be taught in children with HL as they are not intrinsic.

It is essential that a child who is deaf or HoH takes increasing responsibility for their technology and RM in education settings at an age-appropriate stage. Depending on the developmental ability and technology history of the child this can be as early as four years of age but is probably more typical around the formal education age of five-to-six years. Self-learning is best facilitated through problem solving and role play which is the responsibility of the caregivers, educators and therapeutic team. Improvising situations and then using those practiced skills in real-life supported situations will lead to effective self-advocacy in the future.

Conclusion

Following the diagnosis of HL in children, families must make critical decisions regarding mode of communication as this influences educational placement. These decisions have significant implications for auditory, linguistic, cognitive and psychosocial development, and should be guided by a multidisciplinary team through evidence-based, family-centred and individualised intervention planning.

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Chapter 14

Interdisciplinary Approach to Management of Childhood Hearing Loss

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'Alone we can do so little; together we can do so much'.

~Helen Keller

Introduction

Children with hearing loss (HL) have a myriad of needs that must be met in a timely manner by the interdisciplinary team. The critical period of neuroplasticity in the first years of life is where the foundation of cognitive, linguistic, social, emotional and motor development is laid (Çelik et al. 2021). While HL may directly lead to delays in language and listening development without appropriate intervention, approximately 40% of children with HL have additional disabilities beyond the HL (Núñez-Batalla et al. 2023). The impact of additional disabilities also affects children with mild or unilateral HL. The presence of additional needs may delay the diagnosis of HL and/or intervention, and impact communication and educational decisions. These additional needs may include: visual impairment (4-57%), autism spectrum disorder (ASD) (2-4.2%), speech language disorder (61-88%), cerebral palsy (CP) (2-13%), intellectual or developmental disabilities, specific learning disabilities, vestibular and balance disorders, multisensory processing and auditory processing difficulties, as well as additional health concerns (Núñez-Batalla et al. 2022; Alkhamra and Abu-Dahab 2020).

Decreased vestibular function is also common in children with HL (see chapter 7), with teachers reporting poor co-ordination, balance issues and clumsiness (Wiener-Vacher, Quarez and Le Priol 2018). Vestibular dysfunction may impact gaze stability, which is essential for reading, standing and walking. Children with HL are also more likely to have difficulty with praxis, visual processing, spatial integration, sustained attention and executive functioning, with an increased incidence of Sensory Processing Disorders in children with HL (Alkhamra and Abu-Dahab 2020). Fifty percent of congenital or early childhood HL is genetic in origin. For many children, HL is one of many sequelae of a genetic syndrome. Many of these syndromes result in additional learning impairment, progressive visual impairment and other medical challenges. Other aetiologies include illnesses such as meningitis, congenital Cytomegalovirus (CMV) and congenital rubella (see Chapter 2), which may also result in visual impairment, vestibular dysfunction, cognitive impairment and other medical challenges. All of these have a significant impact on the child's development as well as on the burden to the family and their ability to attend and implement intervention plans.

According to Çelik et al. (2021), the presence of an additional disability is the strongest variable impacting on developmental outcomes in children with HL. Another critical predictor of outcomes is parent-child interaction, which emphasises the need for family-focused intervention (Holzinger, Fellinger and Hofer 2022). For those children with additional needs, a fluid interdisciplinary collaboration including multiple professionals and the family is essential (Núñez-Batalla et al. 2022). Case study 1 highlights the additional needs of a child with HL and suggests the type of interdisciplinary team that can be formed to meet the needs of the child and family.

Interdisciplinary Collaboration

Interdisciplinary collaboration is a partnership in which a diverse group of professionals develop a collective identity and shared responsibility, with the aim of providing comprehensive patient and family-centred services (Golom and Schrek 2018). An interdisciplinary approach which results in enhanced person-centred care (PCC) and leads to improved outcomes is recommended when addressing the multifaceted needs of children with HL (WHO 2010). This approach boosts productivity by using time and human skills optimally (Dinh et al. 2020) which is critical in a resource-constrained context like South Africa (see Chapter 15). Professionals working within this model of care engage in joint problem solving, which relies on the expert opinion of each member (Bendowska and Baum 2023). A collaborative, synergistic system is created in which professionals work together to establish goals and priorities with the families of children with HL.

Effective communication

Interdisciplinary collaboration is dependent on effective communication within the team. To achieve this, professionals need to establish open communication, understand and respect each member's contribution, develop a shared language and coordinate their approach as a team (Newhouse and Spring 2010). Failure to communicate effectively can directly impact the outcomes of intervention programmes for children with HL. Diaz et al. (2023) state that effective interdisciplinary communication must be clear, brief and include all the required information to make treatment decisions. Formal communication systems, for example, team meetings, should be set up to ensure that recommendations are followed through. Having these formal communication systems ensures that patient privacy and confidentiality are protected (HPCSA 2019). Informal communication, such as a meeting during lunchtime, also increases trust among team members (O' Reilly et al 2017), which is imperative for achieving successful patient outcomes. Trust can be built by taking time to establish and build relationships, especially if the team members have not worked together previously. A team-building programme that is integrated into already-existing spaces and programmes is more likely to succeed.

Role definition

Diaz et al. (2023) identify clear role definitions and the standardisation of formal processes to be key factors in achieving effective interdisciplinary communication. Hudson et al. (2017) emphasise that understanding team roles is essential for fostering trust and mutual respect. Improved awareness of team members' roles enhances role clarity, enabling the optimal utilisation of professional expertise and leading to better patient outcomes and more cost-effective healthcare systems (Ganann 2019). The varied organisational and reporting structures related to interprofessional teams highlight the need for clear policies and procedures to guide collaboration and communication. The use of technology can furthermore support continuity of care and decision making in interprofessional teams. Social media applications like WhatsApp are being used successfully in many settings as they allow for instant communication between several team members. Interprofessional teams need to be aware of policies and ethical guidelines that govern the use of these platforms, especially due to the risk they pose to patient confidentiality (HPCSA 2019).

Care co-ordination

Due to the involvement of multiple providers and services within a complex healthcare system, coupled with frequent transitions across the care continuum, families are at risk of care being uncoordinated, inconsistent and challenging to navigate (Leach, Stack and Jones

2021). In addition, communicating with multiple professionals within varied hierarchical structures can be daunting for parents of children with HL. The lack of co-ordination can result in increased costs to families, fragmented care from multiple service providers, and duplication of services for children with complex needs (Geese and Schmitt 2023; WHO 2018). Identifying a key person who coordinates appointments and information with families could increase the continuity of care and improve child and family outcomes. Adopting the use of a primary carer or a primary care practitioner (WHO 2018) is recommended to assist with improved communication and co ordination between families and professionals (Geese and Schmitt 2023). This will ensure a consistent and continuous flow of information through a constant point of contact. Care co ordination could also address the gap between diagnosis and timely management (Flindlen 2019) which is critical in the South African context where the burden of HL is disproportionate to the services available (Kuschke et al. 2021). Co-ordinated care is integral to interprofessional collaboration as well as to increased access to services for children with multiple disabilities. Co-ordinated care works best within a family-centred model of care (Dunst 2016), which incorporates family perspectives and goals. A study by Adugna et al. (2020) found that when interprofessional care was provided, families were more motivated to attend health care services.

Caring for a child with multiple disabilities within a low-to-middle income country (LMIC) is very challenging for families who cannot afford the transport costs to access services, and who sometimes put their employment at risk to attend multiple appointments (Adugna et al. 2020). Coordinated care is critical for transitioning children between different levels of health to ensure continuity of care (Kuschke et al. 2021). The National Referral Policy for South African Health Services and Referral Implementation Guideline (NDOH 2020) maintains that for the referral system to work optimally, each facility must clarify its roles and responsibilities, and establish protocols and effective communication systems between health care providers. This remains a longstanding challenge in LMICs like South Africa, where different levels of healthcare are involved in the care of children with complex needs (Amri, Chatur and O'Campo 2022). Brooks, Winship and Kuzel (2020) recommend collaborative visits so that families of children can be connected to appropriate services at a single location.

Interdisciplinary Team

Due to the complex needs of children with HL and the relatively high rate of related medical, social and developmental challenges, it is essential to build a supportive team around the child and family (Findlen, Malhotra and Adunka. 2018). As described in Table 4.1, the team includes (but is not limited to) audiologists, speech-language therapists, otolaryngologists, psychologists, social workers, occupational therapists, physiotherapists, neurodevelopmental paediatricians, geneticists, community health workers, educators and community-based organisations.

Table 14.1: The interdisciplinary team

Team Member	Role
Audiologists	• Audiologists manage early identification programmes (screening), ensure accurate diagnosis of HL, co-ordinate timely audiological services and deliver effective family support and counselling. • They ensure that amplification devices recommended are delivering the intended benefits by verifying and validating their performance. • They regularly monitor the child's hearing status, manage hearing technologies to ensure proper use, care and functionality, and provide training, troubleshooting and guidance as needed for adjustments. • They play a key role in establishing the referral system for early intervention services and possibly serve as the care co-ordinator within the interdisciplinary team. • Some audiologists are AVT trained and can provide these services to children.

Team Member	Role
Speech-language therapists	• Speech-language therapists play a vital role in screening, assessing and providing speech, language, literacy and cognitive-communication services for children with HL. • They provide families and caregivers with information about various language modalities (such as signed, spoken and other forms, including AAC), communication tools, strategies and programmes to help them make informed decisions that align with their child's and family's goals (Chapter 13). • For those families who choose to work towards spoken language outcomes, AVT provided by a speech-language therapist helps children with HL thrive in a mainstream learning environment, becoming independent, active and contributing members of society. • AVT promotes natural conversations and the use of spoken language for communication, emphasising a parent-centred approach. For those families where Sign Language (SL) or Total Communication (TC) is selected (Chapter 13), it is essential that the speech language therapist assists the family in accessing services to support this decision.

Team Member	Role
Deaf Mentor/Deaf Role Model	• A deaf mentor provides families with lived experience perspectives on deafness and hearing loss, supporting informed decision making and realistic expectations regarding communication, identity and participation. • The mentor offers role modelling for children who are deaf or hard of hearing by promoting positive Deaf identity, self-advocacy and social-emotional development. • The mentor supports families in understanding and accessing Deaf culture, sign language and community resources, where appropriate, and complements clinical input provided by healthcare professionals.
Otolaryngologists (ENTs)	• ENTs play a key role in the diagnosis and management of HL in children. • They identify the underlying causes of hearing loss, and assess and manage conditions that contribute to HL. • ENTs perform various surgical interventions such as placement of ventilation tubes, tympanoplasty, cochlear implantation, ossicular reconstruction and implantable hearing aids (Chapter 9 and 11). • They work closely with audiologists in monitoring the progression of HL.

Team Member	Role
Neuro-developmental Paediatrician	• The neuro-developmental paediatrician plays a vital role in diagnosing neurological causes of HL, managing associated conditions and ensuring the child receives appropriate therapies for optimal development. • They identify underlying causes: HL in children may have neurological causes, such as congenital infections (for example, CMV), genetic syndromes or brain abnormalities (JCIH 2019). • They assess and monitor neurological function: Since HL can be part of broader neurological disorders (for example, auditory neuropathy and neurodegenerative diseases), a neurologist evaluates brain function. • Imaging and tests: They can orderi MRI or CT scans to check for structural brain abnormalities, auditory pathway issues or inner ear malformations.

Team Member	Role
Paediatrician or Primary Care Provider (PCP)	• The paediatrician or other primary care provider is responsible for monitoring the overall health and development of the child. • The PCP plays a critical role in ensuring that newborn hearing screening has taken place, following up on the EHDI process, and in monitoring middle ear status and developmental milestones (Chapter 4). • The PCP's role includes partnering with other specialists to facilitate co-ordinated and comprehensive care for children who are deaf or hard of hearing (JCIH 2019).
Geneticist	• At least two thirds of childhood HL have a genetic cause. • An aetiologic diagnosis forms the foundation for accurate genetic counselling and precise assessment of the likelihood of HL recurrence within families.

Team Member	Role
Psychologist	• Parents of children who are deaf or have a hearing loss have rated psychosocial development as one of their main concerns (Wong et al. 2018). • Children with HL are at risk for behavioural problems, increased stress and depression (Dougherty and Day 2022) and therefore a psychologist is required on the team. • Parents of children with deafness and HL require additional support and counselling to cope with the complex needs of their children as well as their own emotional needs.

Team Member	Role
Social Worker	• Children who are deaf or have a hearing loss have complex disabilities, with socio-demographic challenges which may require support from social services. • This support can include informational and emotional counselling to help families better understand their child's disability (JCIH 2019). • Social work services may also include a parent support group to help parents navigate the challenges of raising a child with HL. • These groups foster a supportive environment where parents can share experiences and access valuable information. • Social workers also provide guidance on suitable educational placements and access to social grants.
Physiotherapist	• HL in children is closely linked to challenges in maintaining balance. • Physiotherapy is necessary for children with HL that have underlying vestibular challenges.

Team Member	Role
Occupational Therapist (OT)	• Occupational therapists can assist children with HL who present with various co-morbidities, such as fine and gross motor delays, visuo-motor integration disorders, sensory processing disorders and difficulties within the vestibular system (Alkhamra and Abu-Dahab 2020). • They also assist with preparedness for school.
Community based Organisations and community Health Workers	• Community based organisations and community health workers assist with providing support to families of children with HL. • Organisations like HI HOPES offer home-based support and information to families with an infant or toddler with HL (Chapter 13). • Community Health Workers are a key component of the ward-based community intervention (Khuzwayo and Moshabela 2017). They are in a prime position to help families identify early HL by referring them to their closest health facilities.

Team Member	Role
Educators	• Educators play a crucial role in supporting children with HL, fostering their academic, social and emotional development. • They support the child's mode of communication, for example Sign Language, and support the optimal use of assistive devices within the learning environment. This may include tasks like battery checks (Chapter 10). • It is critical for educators to collaborate with the child's family and intervention team to ensure carry over into the classroom (Chapter 13).

Working Within a Systems Approach

Holistic intervention for children with HL, particularly those with multiple disabilities, needs to account for the complex needs of the child within a Bioecological Systems Approach (Bronfenbrenner 1986). The child's microsystem should be described by identifying everyone who is involved in their care and interaction. This could be achieved by completing an ecomap with the family. An ecomap allows insight into the mesosystem, which encompasses the interactions between the child's microsystems (Sadownik 2023). This process will help the family and the primary care practitioner to identify methods of increasing the support available to the child and family and strengthening the collaboration between the interdisciplinary team and the community involved with the child. The ecomap in Figure 14.1. shows the strong relationship between the speech therapist, who is the care coordinator, with the family and the team based at the hospital. The relationship is moderate for team members outside of the hospital. This could be due to time constraints or large caseloads, which could contribute negatively to collaboration. This ecomap could also be used to motivate more regular communication and consultation between team members. A systems approach also accounts for the ecosystem which refers to the broader formal and informal social structures that are not directly linked to the child's intervention, but impact on the child's well-being and development (Sadownik 2023).

A community resource map could build on the team's understanding of resources available to the family, thereby strengthening their informal support systems and facilitating transitions between different microsystems. Engaging in developing a community resource map could assist the family in increasing their opportunities for learning outside the home and increase the family's informal support system.

Case study 1: Nsuku's story

Nsuku is a 16-month-old child who presents with syndromic features, including a bilateral profound sensorineural hearing loss (SNHL) diagnosed at four months old, as well as cortical visual impairment, severe learning difficulties, low muscle tone and abnormal appearance of the hands and feet. He has abnormally formed cochleae and auditory nerves. He has difficulty with sensory registration and regulation. Nsuku received binaural hearing aids (HA) at the age of five months. After extensive engagement between the ENT surgeon, paediatric neurologist, psychologist, audiologist, speech therapist and the family, it was determined that he was a candidate for cochlear implants (CI), which he received at one year of age. He has attended weekly speech therapy with a Listening and Spoken Language (LSL) Therapist (certified in Auditory Verbal Therapy) since the age of eight months, alternating between face-to-face and telehealth sessions from his home. In the past months, he has shown increased engagement with his environment, an understanding of cause and effect and anticipation of preferred activities. He turns inconsistently to loud sounds, is vocalising an increased range of sounds, and occasionally imitates the suprasegmentals/rhythm of another person's voice or singing.

Nsuku has been on the waiting list for physiotherapy at his local clinic for over a year, and his family has recently managed to find a private physiotherapist to assist twice a month. He can now sit independently, pull to stand and is starting to cruise. He attends occupational therapy once a month at his local clinic. He has limited hand function but has started to grasp and release objects. A paediatric neurologist facilitated appointments for scans and referred the child to a geneticist for diagnosis of a possible syndrome. He was assessed at the Genetics Clinic, but the required tests to confirm his diagnosis are currently not available in the country. His vision has been assessed by an ophthalmologist and an occupational therapist, who report that he can see light and dark and can occasionally focus on a nearby object. His mother disagrees with the results of the visual assessment and feels that his vision is an area of strength for him.

His parents are divorced, and he alternates weeks between the two homes, travelling with a full-time caregiver between houses. The parents consider each other to be a significant source of practical support to one another, and they communicate and co-parent effectively. His mother was recently retrenched from her employment. She is attending psychology services for the anxiety she is experiencing. Nsuku's father understands that his child may not develop language but wants to give him access to hearing through a CI and multimodal communication to offer him the best possible quality of life. His mother believes that a cure may be found but would like to explore South African Sign Language (SASL) to help facilitate his academic journey until then. His family have elected to follow a Total Communication Approach, communicating with him in a combination of gestures, tactile cues, SASL and SeSotho.

His family attends a monthly caregiver support group for families of children with HL and reports that this has been extremely helpful in reminding them that they are not alone on this journey. They report that his speech-language therapist has provided them with a space to process Nsuku's challenges and to experience hope and joy as his parents. His maternal grandmother, who was his mother's primary support, has passed away. His mother reports that she avoids taking him to church and public spaces because she feels he becomes overwhelmed and is happier when home with family.

Application of interdisciplinary principles to Case Study 1

a) Identifying the team

There are many professionals involved in Nsuku's management team, including the team at the hospital that made his initial diagnosis, his occupational therapist at the local clinic, his physiotherapist in private practice and his CI interdisciplinary team, which includes ENT, radiology, speech therapy, audiology, occupational therapy and child and adult psychology. This places him at risk of duplication and fragmentation of services. Unfortunately, hospital catchment areas (in healthcare, a catchment area refers to the geographical region that encompasses the population served by a particular health facility), as well as the fact that his father lives three hours from the hospital, make it impossible to bring all his management under one medical roof. His CI programme has been established as his medical home.

b) Coordination and communication

The CI multi-disciplinary team (MDT) meets monthly to discuss challenging cases and develop care plans for patients. Factors that facilitate successful case management include regular communication between team members, an annual plan with meeting dates, a meeting agenda and minutes that are circulated monthly, with feedback shared with the families involved. Family meetings take place between the relevant professionals and the family to ensure that there is shared understanding of progress and joint goal setting with the family. The team has policies and protocols that guide processes and clarify roles for each member of the team and case management responsibilities in preparation for team meetings. This ensures that case management is done effectively and that the family is involved in all aspects of decision making for their child.

c) Case management within the MDT

Interdisciplinary management within the CI unit is facilitated by clear roles and responsibilities for each member and a shared online booking system with access to online notes from some of the professionals. The team works from one institution and facilitates regular meetings as well as impromptu discussions and joint sessions as the need arises. It is important to have a structured programme for strategic planning, including increased collaboration in service planning and teambuilding to strengthen interpersonal relationships between professionals. Nsuku's speech therapist takes the role of case manager in flagging concerns to the team and in helping the family to advocate for Nsuku with the service providers at other institutions.

d) Communication platforms

A WhatsApp group was created that includes Nsuku's parents, nanny, audiologist and speech therapist. To facilitate the success of the WhatsApp group, all members of the group agreed to clear boundaries regarding the content, times and expected turnaround times for communication on the group. His speech therapist shares take-home messages from their sessions, the family shares videos of his progress in other therapies and their implementation of

strategies (See Chapter 16). The WhatsApp group is also used for telehealth troubleshooting of his device, co-ordination of appointments and sharing of family triumphs and concerns.

e) Facilitating access

Attendance at multiple appointments has proven challenging for the family, and this has resulted in role release, with the occupational therapist and physiotherapist providing programmes and strategies to the family and speech therapist for sensory regulation, visual therapy and motor development. The team has worked to develop open and robust relationships between professionals at different sites and within both public and private healthcare sectors. Telehealth has allowed the speech therapist to observe Nsuku in his natural environment and to work around his nap times to facilitate more successful sessions.

f) The family and informed choice

The team has supported Nsuku's family in making decisions regarding the type of amplification they select for their child, the language they choose and the modality in which he learns language. It is important that the team working with the child collaborates so that the family can make decisions based on their child's profile of strengths and abilities, their values and beliefs, social circumstances, cultural influences and personal choice (Harris, Hemer and Chur-Hansen 2021; Findlen, Malhotra and Adunka 2018). Nsuku was referred to services that support SASL and provided with therapy that utilised Makaton, spoken language, gestures and the introduction of Alternative and Augmentative Communication (AAC). The caregiver support group has facilitated contact between Nsuku's parents and other families of children with additional needs. This has helped them to further develop their own views and goals for his future.

g) Educational placement

A functional communication method is imperative in the deaf-blind learner's establishment of relationships, facilitation of social participation and independence, access to social activities and, most importantly, in their progress across their education trajectory. Finding the right educational environment for Nsuku will be critical in ensuring that he reaches his academic and communicative potential (see Chapter 13). This will require intersectoral interdisciplinary teamwork and communication throughout his educational journey.

e) Ecomap

Figure 14.1 shows Nsuku's interdisciplinary ecomap, providing which provides a visual display of the members of his interdisciplinary team and the strength of their relationships with one another as well as with Nsuku and his family. This tool will guide the team in terms of improving the communication and collaboration between members in Nsuku's ecosystem.

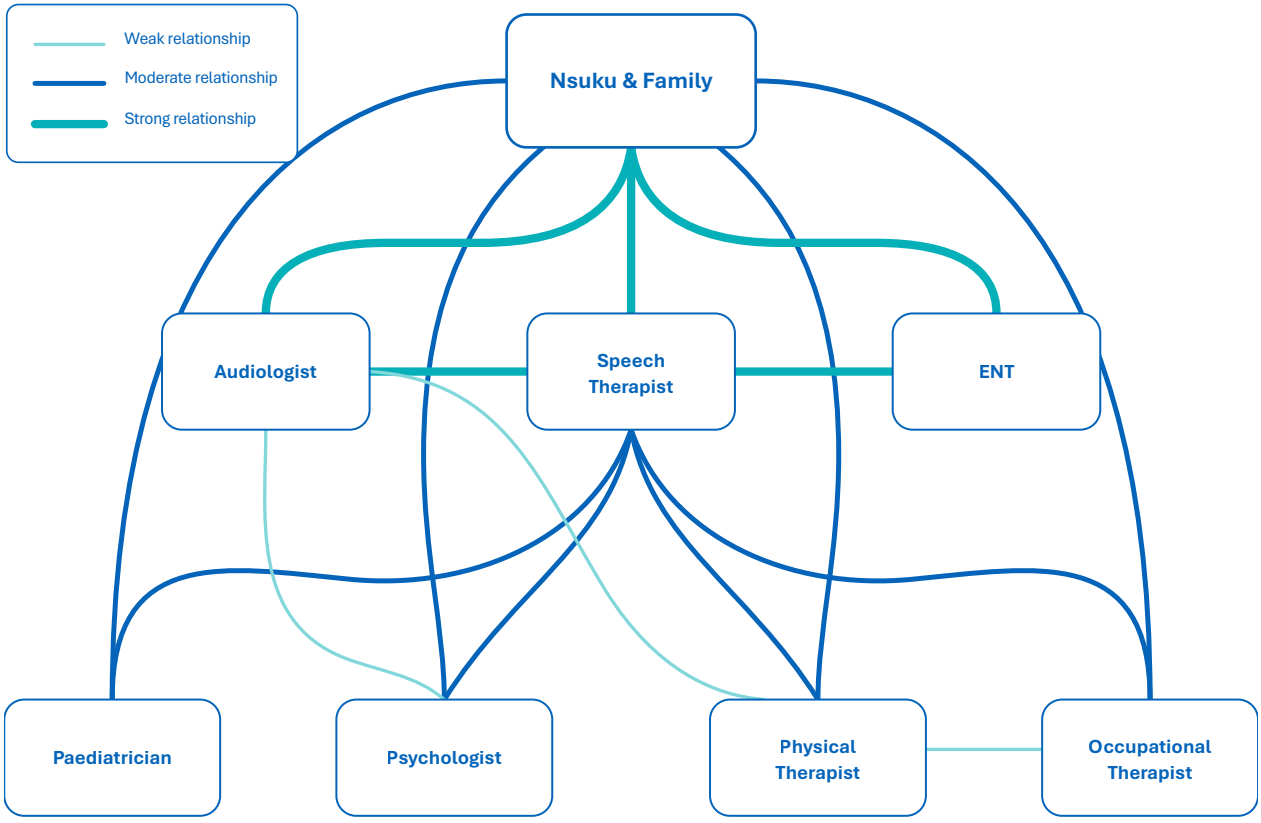

Figure 14.1: Nsuku's interdisciplinary teams ecomap

Conclusion

The importance of interdisciplinary teamwork in the intervention approach with children with HL and their families has been identified as key to achieving positive outcomes. Various practical tools can be utilised to improve communication, such as establishing regular meeting dates, developing protocols and guidelines, setting up communication systems such as WhatsApp groups, and keeping the patient and their families at the centre of all decision making. A systems approach allows for solution-based thinking to improve child and family outcomes by strengthening the relationships between all involved in the child's wellbeing.

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Chapter 15

Person and Family Centred Care

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'Can you see me? All of me? Probably not. No one ever really has.'

~Jeffrey Eugenides

History and Context of Person Centredness

Person-centredness (PC) has historical roots dating back to Florence Nightingale, who emphasised the patient's experience over the illness itself. Psychologist Carl Rogers (1961) significantly contributed to PC theory in the 1940s, advocating for a client-centred approach grounded in respect for individual potential and resourcefulness. Over time, PC spread across healthcare disciplines, inspiring a more holistic model of care delivery known as Patient-Centred Care (PCC), which gained prominence in the late 1960s. Initiatives like the Picker-Commonwealth Programme of 1987 defined key dimensions of PCC, such as respect, coordination, communication and emotional support. Similarly, South Africa's Batho Pele principles (Department of Public Service and Administration 1997), which was launched in 1997, aligned with PC values, emphasising public service improvement through access, transparency and respect.

By 2000, PC become globally recognised, with the World Health Organisation advocating for person-centred health systems. Subsequent academic work, including Mead and Bower's five-dimensional PC framework, further defined its practical aspects: biopsychosocial care,

shared responsibility, therapeutic alliances, and the recognition of both patient and provider as individuals (Mead and Bower 2000).

Person and Family Centred Care (P/FCC)

Although related, Person Centred Care and Family Centred Care are not conceptually the same. Both focus on the person; however, there are key differences. PCC is focused on the person, as an individual, seeking care, while FCC is focused on the family system of the person seeking care (Coyne, Holmström and Söderbäck 2018). See Table 15.1. below for the description and foundations of P/FCC.

Table 15.1: Description and foundations of P/FCC

	Description	Foundations
Family-Centred Care	FCC is a philosophy of care that fundamentally supports and respects the family's participation in their child's care through a partnership approach. It involves shared healthcare decision-making between the family and the healthcare provider.	The family is both the focus and the unit of care. FCC is based on the belief that child and family well-being are best achieved when the service system supports the family's ability to meet the needs of their child. Partnership among families, children and healthcare providers is a core component of FCC (Coyne, Holström and Söderbäck 2018).

	Description	Foundations
Person-Centred Care	PCC is a holistic approach to care that is respectful and individualised. It allows for the negotiation of care and offers choice through a therapeutic relationship in which the person receiving care is empowered to participate in healthcare decisions to the extent they desire (Institute for Patient- and Family-Centered Care 2023).	The person's needs and goals should be the focus of care, rather than paternalistic and biomedically oriented healthcare. This requires organisational vision and commitment, effective leadership, motivation to change, feedback and organisational learning, shared governance and appropriate personal qualities among staff (Institute of Medicine 2011). Collaboration within the team and competencies such as leadership, clinical excellence and critical thinking are also required.

In paediatric care, FCC values enable and prioritise the contribution of parents/caregivers of the children receiving care, as they are considered expert partners concerning their child's health and holistic needs. Both PCC and FCC are important to ensure that the child and their family unit are collaboratively considered within decision-making. Some scholars also highlight the need for CCC, where the voice of the child, their assent and their safety should be taken into consideration in healthcare decision making, planning and outcomes (Coyne Holmström and Söderbäck 2018).

Informed Consent: This is a legal and ethical requirement by means of which a parent or legal guardian provides permission for a child to undergo medical evaluation or treatment. The parent or guardian must be given sufficient information about the purpose, risks, benefits and alternatives of the proposed intervention, and they must voluntarily agree, without coercion (Beauchamp and Childress 2019). In paediatric settings, children generally cannot provide legally binding consent due to their developmental stage, so the responsibility lies with the parent or guardian.

Assent: This is the child's agreement to participate in healthcare decisions or interventions, appropriate to their age, maturity and understanding. While it does not replace legal consent, it respects the child's developing autonomy and encourages engagement in their own care (American Academy of Pediatrics 1995). Assent involves explaining procedures in age-appropriate language, checking that the child understands and ensuring that they are willing to proceed.

Consent vs Assent: The key difference between these two concepts is that informed consent is legal authorisation provided by the parent or guardian, whereas assent is an ethical practice that acknowledges and encourages the child's developing autonomy and willingness. In practice, involving children through assent can improve co-operation, reduce anxiety and foster a long-term positive attitude toward healthcare.

What is the anticipated impact of P/FCC in hearing healthcare?

Audiology is a profession with a focus on the diagnosis and management of hearing loss (HL), which has lifelong limitations and restrictions on communication, functional health and overall quality of life. Management of HL globally has been marred with challenges, and chief among them is low patient adherence to hearing health interventions such as hearing aid (HA) use, as well as the impersonal, acute and one-size-fits-all approach to audiological rehabilitation. This indicates alignment with a 'biomedically focused, device-centred, and clinician-led' (Convery et al. 2019) management approach instead of P/FCC (Figure 15.1). In the traditional biomedical approach to health and hearing care, there is a top-down approach from the care provider to the patient. The focus is on diagnosing the condition and providing a 'quick' cure (for example, prescribing HAs to treat HL), where the health professional is seen as the expert who directs treatment and goal setting. Research has shown that the 'biomedically focused, device-centred' audiology service approach is failing up to 83% of those affected by HL and, in South Africa, up to 85%, because of the lack of access to hearing amplification devices (Orji et al. 2020; Hlayisi and Ramma 2019). In recent South African research on factors influencing PCC, audiologists and speech-language pathologists (53.9% of respondents) identified linguistic and cultural differences between the health professional and patient as one of the top barriers against PCC (Mahomed-Asmail et al. 2025). Leplege et al. (2007) have long posited that, in agreement with PCC and FCC principles, rehabilitative interventions should tackle the difficulties considered most relevant by the person with the disability.

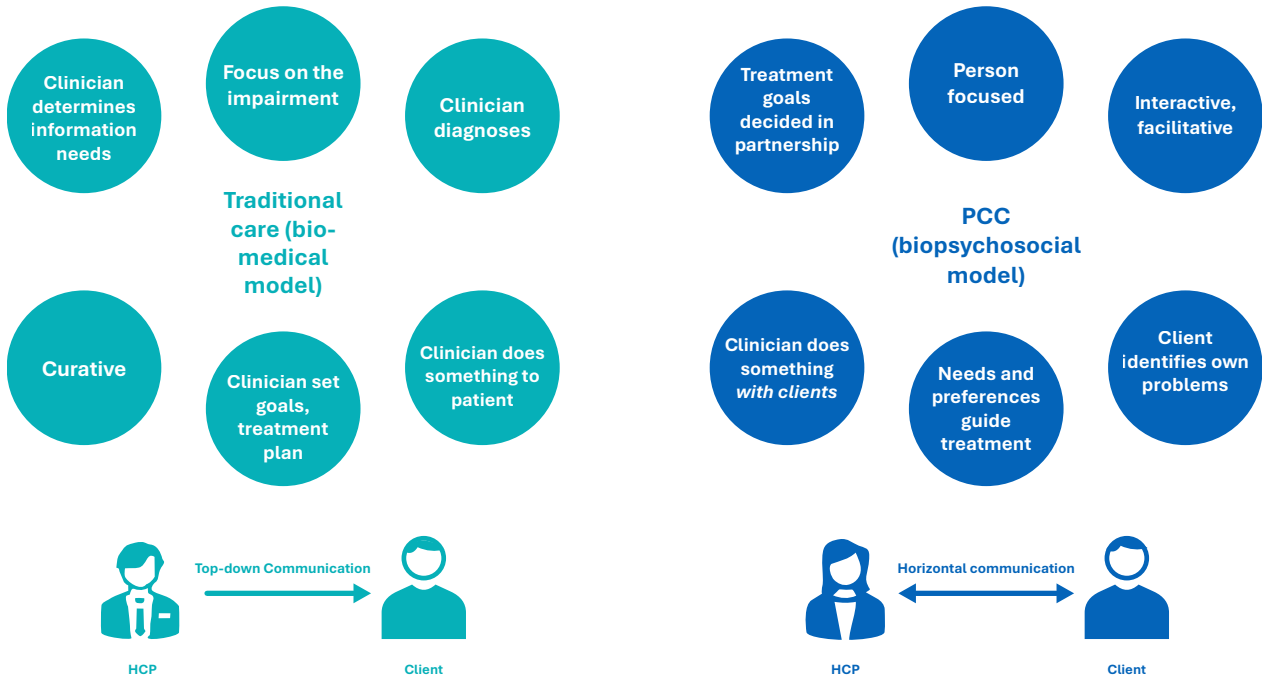

Figure 15.1: Differences between a PCC and traditional, biomedical care approach

Proposed Strategies for Implementation of P/FCC in Audiology

In audiology, P/FCC implementation has been propelled mostly by the Ida institute, a Danish non-profit organisation that was established to focus on the human side of HL management (Kirkwood 2008). The Ida Institute was driven to global importance by a collective of experts from ethics, audiology, anthropology, education and social responsibility backgrounds (Kirkwood 2008). Over time, the Ida Institute has, with its partner experts in hearing care, curriculum development, research and clinical practice, put together various tools to operationalise P/FCC in audiology. Most of their tools were developed to make the key aspects of P/FCC tangible and has resulted in what they term ‘the six elements of PCC’. Parallels can be drawn between these principles and the seven PCC dimensions of by the Picker-Commonwealth programme, the eight Batho Pele principles and Mead and Bower’s five dimensions of PC. The principles include:

1) Active listening
2) Open-ended questions and reflective conversations
3) Empathy
4) Involvement of family and friends
5) Shared goal setting
6) Decision making, and understanding of individual preferences

Practical clinical applications of each P/FCC element are described below with suggested tools (paper-based) to facilitate and enable implementation. Some of these tools are freely available on the IDA website (*www.idainstitute.com/tools*) for clinicians, students and families. While the use of these tools is not essential, doing so could be useful in getting the process started. It is important that P/FCC should be understood as an overarching approach to hearing healthcare that informs all interactions with the child, person or family rather than being reduced to a discrete tool, technique or isolated component of a hearing care appointment.

Elements of P/FCC and clinical applications

a) Active listening

It is crucial for HHPs to pay full attention to the person speaking during a consultation to determine their needs, values and preferences. In paediatric sessions, active listening can be applied across all aspects of care, for example, whenever a parent or caregiver shares their concerns, gives feedback or asks a question. Active listening includes paying attention to verbal and non-verbal cues of the parents and/or caregivers, as well as the child. It is important that active listening strategies be used when communicating with the child to ensure that they are involved in clinical discussions, decisions and goalsetting where possible and in an age-appropriate manner. See Table 15.2 for some practical examples.

b) Open-ended questions and reflective conversations

HHPs should ask questions that encourage detailed responses. This helps with identifying emotions, experiences, needs and so much more than a list of symptoms. Open-ended questions may seem unstructured at first, but asking one open-ended question can frequently provide more information than several closed-ended questions. Reflective conversations are often facilitated through open narratives that build a relationship. In paediatric hearing care,

the child and family will most likely meet with the HHP over an extended period of time, so it is important to have continued investment in understanding each other. The purpose of open-ended and reflective conversations is to encourage the child and family to 'hear' their own thoughts, to let them know that the clinician is trying to understand and to encourage them to keep talking.

c) Empathy

An empathic HHP can help the person or family feel understood and seen, and build a therapeutic relationship. There are three types of empathy that may be useful to apply as a clinician in paediatric care, and include the following:

- Cognitive empathy: This is the ability to mentally understand another person's perspective/experience and impacted aspects of life. Here, a clinician is advised to be non-judgmental and open-minded, and have the capacity to engage others through their lens (Areas of difference, such as culture, religion, traditions, practices and beliefs, test cognitive empathy).
- Emotive empathy: This is the ability to feel with another person through active listening, reflective conversation and openness to engage emotions in clinical settings.
- Empathic action: This involves putting empathy into action. For example, an HHP can put empathy into action by showing compassion and holding space silently so that the parent of child they are assisting can feel heard.

Another important aspect to consider as an empathic HHP is building rapport. Building rapport in paediatric care involves developing a trusting and collaborative relationship with both the child and their family, fostering open communication and a sense of safety (Brown, Mulhern and Hardy 2020). Rapport serves as the foundation for person- and family-centred care, where children and families are active participants in decision making and management (Kuhlthau et al. 2011). Rapport is achieved through empathy, respect and active listening, enabling understanding of the child's needs and the family's concerns (Levett-Jones, Cant and Lapkin 2017). Coyne, Holmström and Söderbäck (2018) describe some of the steps that can be used in practice:

- Adapting communication to the child's developmental level
- Using play or non-threatening interactions to reduce anxiety
- Validating the experiences of caregivers

Establishing rapport is vital for the HHP to enhance co-operation during clinical encounters and to strengthen intervention adherence, which is critical for long-term health and developmental outcomes in paediatric care.

d) Involvement of the parents and family

In paediatric care, it is important that decisions be made in collaboration with both the caregiver and/or parents and child (where appropriate). The contribution and participation of the family in paediatric hearing care is advantageous for continuity of care, compliance and overall long-term success. The HHP can encourage involvement of the family and siblings of the child by:

- Inviting the family and siblings to appointments with the intention to involve them in the session by encouraging the siblings to provide feedback and make observations about play and encouraging each member of the family to take on roles and responsibilities, such as: for the dad or co-parent to make notes for home activities, for the mom to fill out the questionnaire.
- Adding pre-planned tasks for family participation during appointments:

    - Pre-planning certain tasks for each family member to place family at ease and guide their participation at appointments. For example, during assessments, one parent can be tasked with the role of play audiometry response reinforcer (Chapter 5). This will enable their active participation during assessment and place the child at ease inside a booth with a familiar face. Furthermore, a sibling can be tasked with bringing a familiar toy that can be used during play audiometry.
    - Discussing Third-Party disability which describes the functional difficulties and psychosocial challenges that may be experienced by the parents and siblings as a consequence of a family member's health condition, despite not having the impairment themselves (Scarinci, Worrall and Hickson 2008). In the context of paediatric HL, parents and siblings may experience emotional distress, social withdrawal, financial strain, and increased caregiving responsibilities, all of which can negatively impact their quality of life (Scarinci, Worrall and Hickson 2009). For example, parents often report heightened stress related to communication breakdowns, hearing device management and educational or healthcare systems navigation (Hétu 1996). Recognising third-party disability in paediatric audiology is essential,

as the well-being of family members directly influences the child's developmental and rehabilitative outcomes. Integrating counselling, education and psychosocial support during an appointment can helps mitigate third-party disability and strengthen the overall effectiveness of intervention.

e) Shared goal setting and decision making

HHPs can facilitate collaborative goal setting and develop a care plan based on the individual's needs and preferences as well as those of their family. Applying shared decision-making and prioritising individualisation can seem tricky to balance. Some practical tips to consider are:

- Understand the child and family context.
  - Ask about the child's daily routines, communication environment and family priorities.
  - Consider cultural, linguistic and lifestyle factors that may affect hearing care choices.
- Present options clearly and collaboratively.
  - Explain intervention choices (for example, the use of HAs and therapy strategies) in age-appropriate language for the child and accessible terms for caregivers.
  - Discuss the benefits, risks and realistic outcomes of each option.
  - Tailor strategies to the individual.
  - Adjust therapy techniques to the child's attention span, learning style and communication abilities.
  - Modify follow-up plans and home strategies based on family feedback and observed progress.
- Encourage ongoing collaboration.
  - Encourage caregivers to share observations, concerns and successes.
  - Revisit goals regularly to ensure care continues to meet the evolving needs of the child and family.

Successful application of shared goal setting is linked with active and attentive listening to the needs of the child and family. Furthermore, the goals should follow a solution-based approach in line with the shared concerns, needs and preferences. It is also important to take note of the power dynamics in decision-making and establishing equal decision-making

capacity. Moreover, it is crucial to understand that in paediatric care the goals can shift and evolve as the child develops. Some key aspects to consider in facilitating shared goal setting and decision making include:

- Create an enabling space for all to contribute their goals.
- Be clear about how goals will be prioritised, for example, where there is a difference of opinions and ideas.
- Remember that the HHP's role as the knowledge bearer clinically is to guide the parent or caregiver, who, in turn, is the knowledge bearer in the child's life. Therefore, the goals that are set should interweave these two worlds.
- Be mindful of the ethical considerations for and differences between consent and assent, where appropriate.

f) Understanding individual preferences

HHPs are encouraged to recognise and respect the unique needs and desires of the child (where applicable) as well as of their families. Individualisation of hearing care depends on the HHP's ability to identify the unique needs and particularities of the child and the family. Determining their needs and preferences is an ongoing and critical aspect of P/FCC. Individual preferences should be considered in conjunction with all other aspects, as previously discussed, that need customisation. Practical tools that may assist patients and their family/caregivers in this regard include:

- The Communication Rings (a tool to help identify the social network of the child and family, and to assess how HL may affect relationships, that is, 'who I communicate with', available from https://idainstitute.com/tools/communication-partners--communication-rings.
- Living Well (a tool to help identify the most important communication situations and strategies to optimise communication in these situations, that is, 'where I communicate', available from https://idainstitute.com/learning-hall.
- Question prompt lists for parents (Available from *chrome-extension://efaidnbmnnnibpcajpcglclefindmkaj/http://www.heartolearn.org/tutorials/transcripts/QPL_Parents_ENGLISH.pdf)*

Table 15.2: Elements of PCC and practical examples for implementation

Element of PCC	Examples of implementation	Tools
Active listening	Verbal cues to indicate full attention: • Inviting the person to say more, that is, 'Tell me more about that/please elaborate on …' • Acknowledging what they are saying through paraphrasing, asking clarifying questions and reflecting emotion and affirmation: - *Paraphrasing*: 'Just to make sure I understand, you're saying your child doesn't respond to soft sounds at home?' Reflecting emotion: 'You mentioned that you've had to push quite hard to get a referral — that must have been exhausting.' -*Clarifying*: To the parent: 'Can I just pause to check — I think I heard you say the school has noticed issues in the classroom too?' To the child: 'You're saying that your HAs are okay at school, but they feel 'itchy' after a while?' • Where appropriate, expressing appreciation for the sharing of vulnerable or personal information. This can be useful to build rapport, for example, 'thank you for sharing that with me, I value your trust.' • Acknowledging body language and emotions. This could include statements such as 'I see you are tensing up/I see you are feeling sad.' • Responding with soft affirmations, such as 'Mmm,' 'I see…' • Taking brief notes and explaining why, for example, ' I'm just writing this down so I don't miss anything — it's important.' Non-verbal cues to indicate full attention: • Maintaining eye contact with the person speaking or facing the child when they are engaging. • Nodding gently or using facial expressions that show engagement. • Using warm, open body language, for example, arms relaxed, slight forward lean. • Minimising distractions during case history/counselling, for example, making notes or records afterwards, if possible. • Pausing and giving time for both parent and child to speak — avoid interrupting.	

Element of PCC	Examples of implementation	Tools
Open-ended questions and reflective conversations	• *Asking What/When/Why/How questions*. These give context to and show the impact of the symptoms. Examples include 'Can you tell me When you feel most impacted by XYZ in your normal day?', 'How does XYZ affect your day-to-day activities?', 'Why do you think XYZ is a concern for you?' • *Asking for real life examples* which provides details on lived experiences. This could include 'Can you share the most recent incident of this?', 'When do you first remember noticing this?', 'What can you give as a real-life basic example of this?' • Facilitating reflective conversations and interactions between the HHP and the child and parents by: - Asking the parent/child to tell you how they got to you today. - Summarising what the person is saying and asking them to confirm that you have understood them. - At the end of the session, asking them how they feel about everything shared. • Other practical ways to offer reflective comments that can strengthen connections include: - *Reflecting feelings* such as 'Sounds can feel too loud sometimes, can't they?' or 'It sounds like noisy places aren't your favourite.' - *Acknowledging perspective*, which could include 'You've done a great job telling me what it's like for you.' Or 'You're trying really hard to hear in school - that must take a lot of focus!' - *Building confidence* by using phrases such as 'I can see how much you've learned about your hearing already.' or 'You're the expert on what works best for you!'	a) Living Well and b) Motivation Tools

Element of PCC	Examples of implementation	Tools
Empathy	Some practical ways to express emotive empathy to caregivers/parents include the following statements: - *When they're feeling worried*, 'It's completely understandable to feel anxious when something doesn't seem quite right with your child.' or 'This is a lot to take in — many parents feel the same way when they hear this for the first time.' - *When they have had a long journey*, 'It sounds like you've been trying to get help for a while — that must have been exhausting.' or 'You've been through a lot just to get to this point. I really admire your persistence.' - *When they express guilt*, 'It's very normal to question yourself — but please know this isn't your fault.' or 'You care deeply about your child, and that comes through in everything you're doing.' - *When they are unsure*, 'It's okay to feel unsure — this is a big decision, and we'll take it one step at a time.' or 'You don't have to have all the answers today — we're here to support you through it.' Practical ways to apply empathic action when interacting with the child include the following; - *When they are nervous or shy*, 'It's totally okay to feel a little nervous — lots of kids feel that way when they come here.' or 'You're being really brave just by coming in today.' - *When frustrated*, 'That must feel really frustrating when you can't hear things clearly.' or 'It makes sense that you don't like loud sounds — they can be really uncomfortable.' - *When engaged*, 'You've done such a great job today — I can tell you've been trying really hard.' or 'You're really good at telling me what works for you — that's so helpful!'	
Involvement of the family	• Actively seek input and participation from each member through asking open ended questions like 'What would YOU like to add/contribute?' or 'How can YOU best support in this?'	a) My Hearing Explained b) The Communication Rings

Element of PCC	Examples of implementation	Tools
Shared goal setting and decision making	*Clear, person-centred communication* • Use everyday language to explain test results and hearing conditions, for example 'This graph shows how soft or loud a sound needs to be for your child to hear.' • Explain pros and cons of each option in the family's context: 'One option is HAs, which can help with soft speech in quiet and noisy places. Another option is monitoring for now, depending on how things go at home or school.' • Use visuals or handouts when helpful: 'Here's a simple chart that compares the options — we can go through it together.' *Eliciting parent values and priorities* • Acknowledge lived experience by making statements such as: 'You know your child best — what have you noticed works well for them?' • Summarise what the parent has said, for example 'So, you're saying you'd like to explore options that don't feel too overwhelming for your child right now?' • Reflect both facts and emotions, for example 'It sounds like you're feeling a bit torn — you want to support your child but also want to make sure they're not uncomfortable with the solution.' *Involve the child (when appropriate)* • Include the child's voice in age-appropriate ways such as 'You've said it's hard to hear your teacher — what would help make that easier?' or 'Let's look at some colours and styles of HAs. Which ones do you like?' • Empower the child in choosing or using technology where possible. *Giving space to decide* • *Allow time to process*, for example, 'You don't have to decide today. I can give you some information to take home, and we can talk more at the next visit.' • *Offer follow-up support*, such as 'If more questions come up, just call or email — we're here to support you.'	a) The Goal-sharing for Partners Strategy (GPS) b) Partner Journey c) My Turn to Talk for Parents

Element of PCC	Examples of implementation	Tools
Understanding individual preferences	*Structured Conversations with Caregivers* • Ask open-ended questions about the family's routines, values, and goals for their child. Example: 'What activities does your child enjoy most at home or school?' or 'What is your main priority for your child's hearing this year?' Discuss past experiences with healthcare or devices to identify preferences or concerns. *Observation of the Child in Natural Settings* • Watch the child interact in play or during routine tasks to notice communication style, attention span, and responsiveness. Example: Observe how the child reacts to background noise or group settings to guide hearing device recommendations. *Use of Visual Aids and Choice Boards* • Present options (HAs, therapy activities, or communication strategies) using pictures or models so children and parents can indicate preferences. Example: Let a child choose between two colourful HA styles or select which therapy game they want to try first. *Incorporating Questionnaires or Preference Scales* • Use short forms to capture caregiver priorities and child comfort levels with devices or therapy approaches. Example: A Likert-scale sheet asking parents how important aesthetics, comfort, or durability are in hearing devices. *Collaboration with Other Professionals* • Consult teachers, speech therapists, or early intervention specialists who observe the child in different contexts. Example: Ask the teacher how the child participates in classroom discussions to guide amplification choices.	a) My World b) My Hearing Explained c) Living Well for Teens and Tweens d) The people I talk to

Practical Application of Person- and Family-Centred Care (P/FCC) across Developmental Stages

Preschool-aged children with amplification

At this age, family input and early learning settings are central to the child's development. Parent observations, combined with input from early childhood educators and clinicians, provide a holistic view of the child's listening, language and participation across daily routines. These observations help guide both device management and intervention strategies in ways that are meaningful to the family. Some examples of these checklists and questionnaires include:

- LittleEARS Auditory Questionnaire (parent-reported auditory behaviours in infants/toddlers), available at *https://hearlife.org/data/en/med-el/LittleEARS_Auditory_Questionnaire.pdf*
- Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (structured parent interview about spontaneous listening behaviours), available at *https://www.unm.edu/~lkrulik/it-mais.pdf*
- Auditory Skills Checklist (tracking early auditory milestones in everyday contexts), available at *https://www.chchearing.org/wp-content/uploads/2018/09/Auditory-Skills-Checklist.pdf*
- Ages and Stages Questionnaire (ASQ-3) (parent developmental screening tool with items relevant to communication and social engagement), available at *https://agesandstages.com/products-pricing/asq3/*
- Parent coaching strategies (clinician-guided observation and support for embedding listening and communication practice into daily play and routines)

School-aged children with amplification

The practical application of P/FCC can be found in the management of a school-age child with hearing amplification. Classroom observation by the audiologist, combined with observation checklists from parents, educators, and the child, can provide valuable input about a child's performance in the school and home setting, and offer a more holistic perspective about the impact that a CI has on the life of the child and family. Some examples of these checklists and questionnaires include:

- Managing Listening Conditions Checklist (observations of the educator and/or child)
- Observational Record of Behaviour of Deaf or Hard of Hearing Students by audiologist observation (Landrud and Anderson 2011)
- Teacher Evaluation of Aural/Oral Performance in Children (TEACH), available at *https://www.outcomes.nal.gov.au/teach*
- Parent Evaluation of Aural/Oral Performance in Children (PEACH), available at https://www.outcomes.nal.gov.au/peach
- Quality of Life (QoL) Cochlear Implant Questionnaire, available at *https://pubmed.ncbi.nlm.nih.gov/33974778/*
- Vanderbilt Fatigue Scales (child, parent and teacher versions), available at *https://www.vumc.org/vfs/pediatric-scales-and-user-guide*

Adolescents with amplification

The practical application of P/FCC for adolescents should focus on supporting both the young person's growing independence and the family's role in scaffolding self-advocacy skills. At this stage, P/FCC involves engaging the adolescent as an active decision-maker while ensuring parents, educators and clinicians work together to address academic, social and emotional needs. Examples of strategies include:

- Self-Advocacy Checklist for Teens with Hearing Loss (student self-report of advocacy and independence skills), available at *https://successforkidswithhearingloss.com/product/self-advocacy-checklist-for-teens/*
- Listening Inventory for Education – Revised (LIFE-R) (student and teacher questionnaires about listening effort and classroom participation), available at Listening Inventory for Education – Revised (LIFE-R)
- Vanderbilt Fatigue Scales – Adolescent version (student and parent report of listening-related fatigue), available at *https://www.vumc.org/vfs/pediatric-scales-and-user-guide*

Conclusion

This chapter aimed to equip the HHP with fundamental knowledge of P/FCC theory and guide the practical application of PCC principles throughout every appointment in the hearing care journey. When these principles are routinely incorporated into everyday consultations, the benefits of patient-centred healthcare are realised for both patients, families, clinicians and health systems alike.

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Chapter 16

Telehealth in Paediatric Audiology

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'Digital technologies are now integral to daily life, and the world's population has never been more interconnected. Even so, its application to improve the health of populations remain largely untapped, and there is an immense scope for use of digital health solutions.'

~World Health Organisation, 2025

Introduction

Telehealth, the provision of specialised services at a distance using information and communication technology, has become an important aspect of modern healthcare delivery worldwide. Telehealth reduces the geographical and logistical barriers that frequently limit access to timely diagnosis and intervention, especially for families in remote or underserved regions, and aligns closely with global priorities of equitable healthcare and person/family-centred care (P/FCC). In audiology, telehealth (also referred to as teleaudiology, tele-therapy and telepractice) has been adopted in a variety of hearing care services, including screening, diagnostic assessment, hearing aid (HA) fitting, cochlear implant (CI) mapping and auditory-verbal therapy (AVT). For children with hearing loss (HL), telehealth services are delivered through synchronous video consultations and asynchronous data exchange. The feasibility and effectiveness of telehealth in the identification and management of HL have been demonstrated across different contexts (including remote and resource-constrained

environments), and its use has been recommended by the latest World Health Organisation's World Report on Hearing (World Health Organisation 2021).

Benefits of Telehealth in Paediatric Audiology

Telehealth addresses longstanding challenges related to access, continuity of care and equity of service provision. It offers families more flexible and responsive care options and aligns closely with P/FCC. The following section explores the key benefits of telehealth in paediatric audiology.

Efficacy

Studies have shown no significant differences between telehealth and in-person hearing screening methods, including the use of distortion product otoacoustic emissions (DPOAE) (Ameyaw et al. 2019) and automated auditory brainstem response (AABR) (Hatton et al. 2019; Sithi, Govender and Ntuli 2024). Oremule et al. (2024) reported that mobile audiometry closely aligns with conventional audiometry in measuring hearing thresholds. Similarly, other studies have demonstrated the effectiveness of telehealth in diagnosing otitis media (OM) (Habib et al. 2023). In intervention, telehealth can be effective for delivering listening, spoken language and speech intervention, with outcomes comparable to traditional in-person therapy (Constantinescu et al. 2014). However, other authors have recommended that telehealth be used as a second option due to yielding better results in-person (Ebrahimi et al. 2024). Remote CI programming and HA monitoring have also shown positive outcomes (Muñoz et al. 2021; Schepers et al. 2019).

Cost effectiveness

For families, telehealth reduces costs associated with travel, childcare and time away from work, alleviating common financial barriers to accessing care. Providers similarly benefit from decreased expenses related to travel for outreach services, clinic space, utilities and administrative overhead, while the flexibility of telehealth scheduling reduces missed appointments (McCarthy et al. 2019). Research has demonstrated the cost reduction for patients and providers associated with reduced travel-related expenses, with these savings rising with increased service intensity (for example, Hatton et al. 2019).

Improved accessibility and convenience

A major benefit of telehealth is improving accessibility and convenience in paediatric hearing services. In rural and remote areas, access to hearing healthcare is often limited due to a shortage of qualified hearing healthcare professionals (HHPs) and the high costs associated with establishing local services or travelling to urban centres (Swanepoel et al. 2010). Through telehealth, families in these areas can access services without the burden of long-distance travel. Telehealth may also be used to provide better access for Indigenous populations (for example, Habib et al. 2023). Similarly, in low-resource settings like Sub-Saharan Africa, telehealth can be a feasible method of providing access to screening and diagnostic assessment (for example, Sithi, Govender and Ntuli 2024).

Telehealth also enhances access for families in metropolitan regions by overcoming barriers such as traffic congestion, limited public transport and long waiting lists. It allows flexibility in scheduling appointments around school, work, care of children with additional needs and other commitments (McCarthy et al. 2019). Beyond benefiting families, telehealth also enables HHPs to manage their caseloads more flexibly, increasing service efficiency and reducing travel demands.

Enhanced continuity of care

Telehealth also plays an important role in reducing loss to follow-up and enhancing continuity of care. By enabling ongoing remote monitoring and check-ins, telehealth minimises the risk of families disengaging from care pathways (Ameyaw et al. 2019). Furthermore, telehealth supports continuity when families travel or relocate, or when HHPs move practices. During the COVID-19 pandemic, telehealth proved invaluable in maintaining care continuity despite restrictions (for example, Mardinli et al. 2023).

Increased caregiver involvement

As HHPs are not physically present to be ‘hands-on’ during sessions, caregivers naturally assume greater responsibilities, such as assisting with assessments and implementing intervention strategies. For instance, in assessment, studies have explored the feasibility of parents performing otoscopy using smartphones (Shah et al. 2018; Erkkola-Anttinen et al. 2019). In intervention, telehealth could facilitate a coaching environment where professionals

guide parents in mastering skills, promoting FCC (McCarthy et al. 2019). This engagement also extends to other family members, with siblings often participating in telehealth sessions, further strengthening the support system around the child with HL (McCarthy et al. 2019).

Implementation of Telehealth Services in Paediatric Audiology

Models of telehealth

Telehealth in audiology (that is, teleaudiology) includes various models designed to deliver comprehensive hearing care to families of children with HL aged zero to thirteen years. These models can be used flexibly, taking advantage of digital technologies to provide a range of hearing-healthcare services, ensuring that children receive timely and effective hearing care regardless of their geographical location. Four main models of telehealth in audiology are described in the next section.

a) Synchronous teleaudiology

Synchronous teleaudiology involves real-time interactions between HHPs and clients through video conferencing platforms. This synchronous model (Figure 16.1) allows professionals to conduct live assessments, provide immediate feedback and engage in interactive consultations and interventions with both children and caregivers. For example, procedures like remote video-otoscopy, pure-tone audiometry, speech-in-noise tests and immittance audiometry have been found reliable when performed remotely, with the audiologist guiding an on-site facilitator (for example, a community healthcare worker [CHW] or parent/caregiver) on how to position the child, use the equipment and complete the procedure (Rushbrooke, Waite and Huston 2020). The synchronous model of teleaudiology care is beneficial for conducting not only screening and assessment procedures but is also optimal for follow-up appointments, intervention and counselling sessions where children and families are supported throughout the hearing care continuum (Rushbrooke, Waite and Huston 2020).

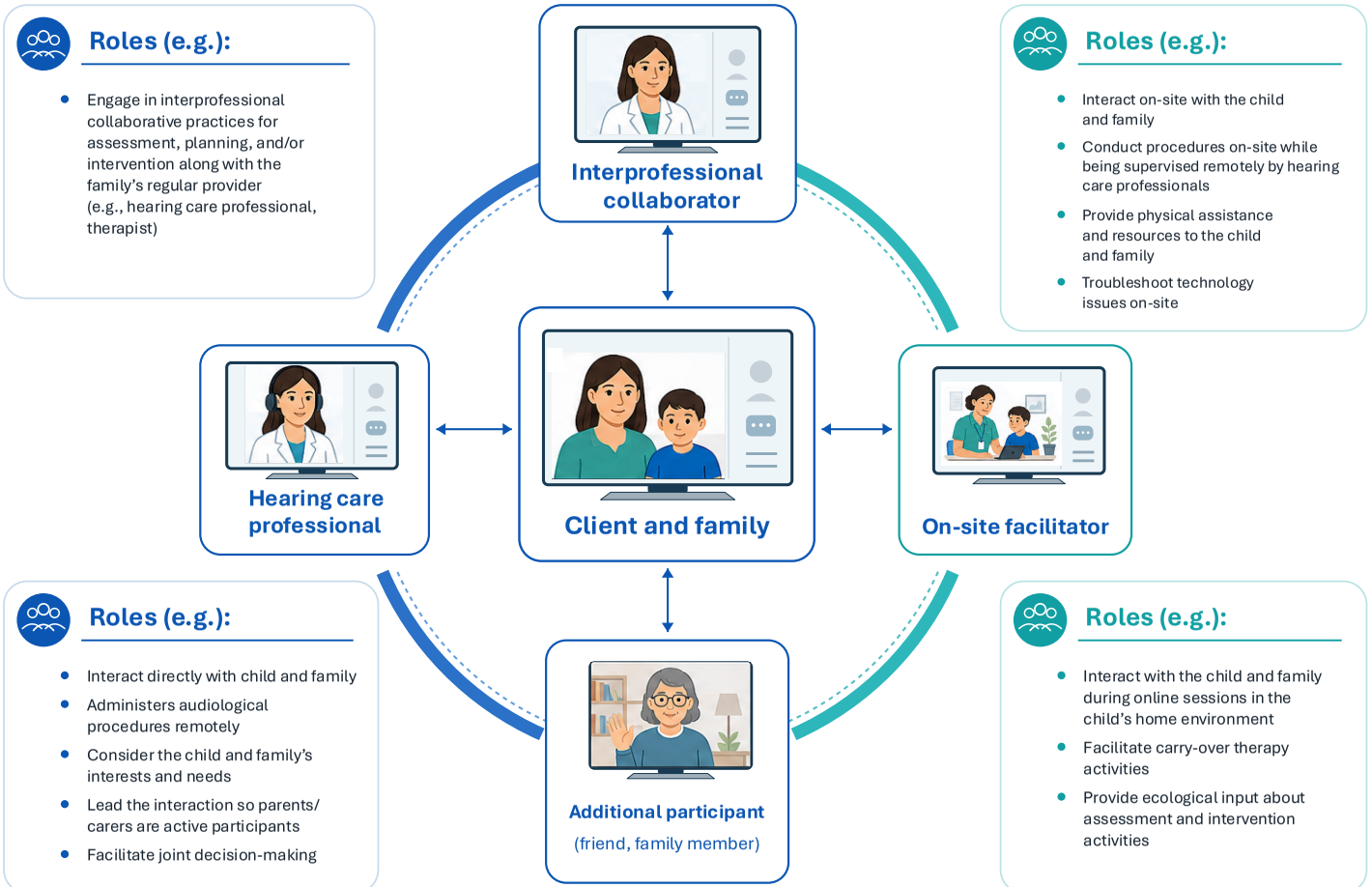

Figure 16.1: Synchronous paediatric teleaudiology model with a description of potential roles of each participant

b) Asynchronous teleaudiology

Asynchronous teleaudiology, also known as 'store-and-forward', involves the collection and transmission of audiological data to be reviewed by the HHP at a later time. The asynchronous model (see Figure 16.2) is useful for initial screenings and diagnostic evaluations where immediate interaction may not be needed or be possible. For example, on-site facilitators (CHWs, allied-health assistants) can use portable devices such as smartphones, tablets or iPads to conduct hearing screening tests at home or in primary care settings, and the results are uploaded to a secure cloud-based platform for the HHP to review later. Altamimi et al. (2024) used a store-and-forward approach to improve access to hearing care services for children with OM in Perth, Australia. Children with OM were assessed by audiologists and the data reviewed later by a multidisciplinary team (MDT; that is, otolaryngologist registrar, clinical nurse specialist, audiologist) which then provided individualised care plans to caregivers via phone calls. The asynchronous approach enabled early access to hearing care and reduced the cost associated with initial and follow up appointments (Altamimi et al., 2024).

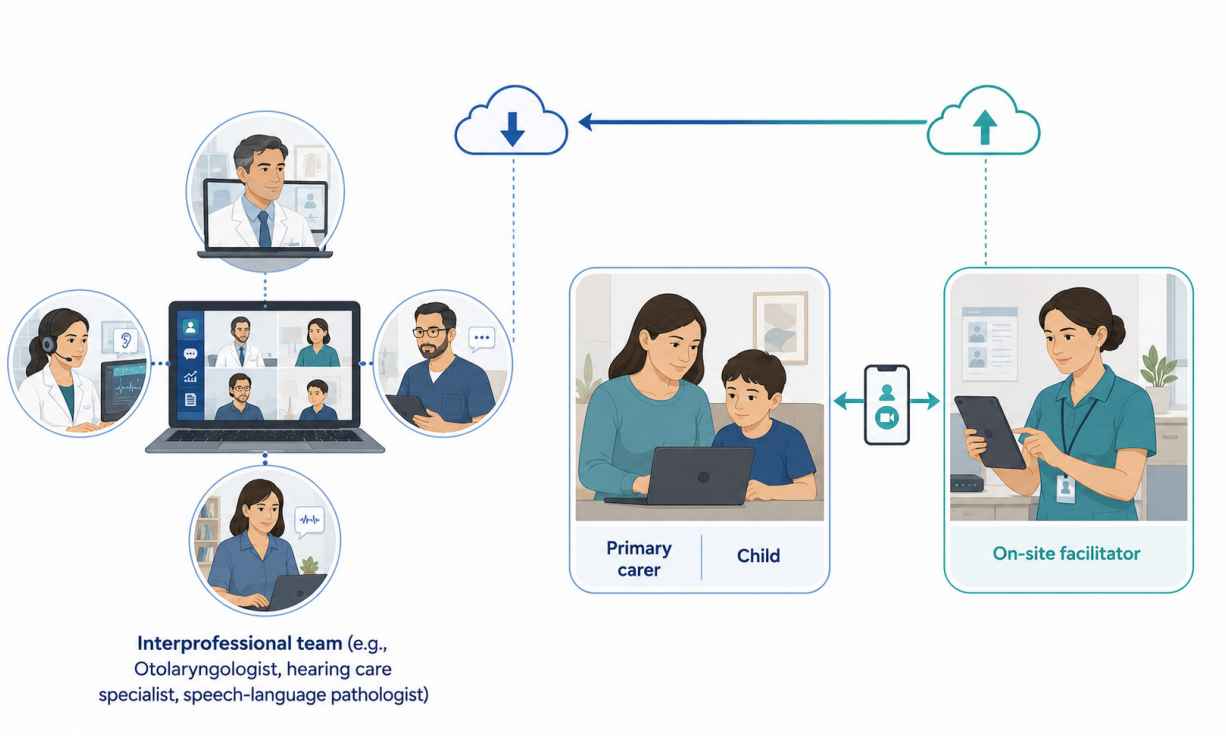

Figure 16.2: Asynchronous model of paediatric teleaudiology assessment

c) Hybrid teleaudiology

As the name suggests, hybrid teleaudiology combines elements of synchronous, asynchronous and in-person models to provide a comprehensive and holistic approach to hearing care. This model allows for initial data collection and preliminary assessments to be conducted asynchronously, followed by synchronous consultations for detailed assessment and intervention, or an initial synchronous audiological intervention followed by in-person sessions. For example, a child's hearing test results are uploaded to a cloud-based platform, and the audiologist reviews the data before a scheduled synchronous video consultation to discuss the findings and plan further interventions (Rushbrooke, Waite and Huston 2020). Another example is a telehealth consultation combined with an in-person assessment, which then includes intervention provided via in-person and telehealth sessions. This integrated approach ensures thorough, tailored, and person-centred hearing care while maximising the efficiency of telehealth services (see example in Chapter 15).

d) Mobile Health (m-Health) applications

m-Health applications (herein m-Health apps) have increasingly been used in teleaudiology to facilitate remote hearing assessments and interventions. These apps can be installed on smartphones and/or tablets/iPads, allowing HHPs and trained workers to conduct hearing screenings and monitor children's hearing health across different contexts and clinical settings. Schools can partner with audiologists to conduct remote hearing screenings and follow-up assessments using portable audiometers and video conferencing tools (see example in Chapter 4).

The four models of teleaudiology described in this section provide flexible and effective solutions for delivering hearing care to children with HL and families. It is important for the HHP to consider which model(s) of telehealth are the best fit for the clinical services they wish to undertake and will best meet the needs of their clients. Some practical insights into the technology and set-up for telehealth assessment and intervention, including best practice principles are provided later in the chapter.

Integration of telehealth into hearing care systems

Integrating telehealth into existing service delivery systems in paediatric audiology is

essential for creating a seamless care pathway. This may involve the consideration of how telehealth could be used to complement existing in-person services in a hybrid model of service delivery. Audiology clinics could adopt a hub-and-spoke model where more specialised services such as diagnostic testing are provided in-person from a central location (the 'hub'), and satellite sites (the 'spokes') are used to provide basic or intermediate care services, with telehealth used to connect back to specialists at the hub as needed. Integrating telehealth into existing systems would have the benefit of leveraging established healthcare infrastructure, such as electronic health records and information technology systems. This approach enables HHPs to maintain continuity of care and collaborate remotely with the MDT to develop comprehensive care plans (Al-Worafi 2024).

Telehealth Assessment in Paediatric Audiology

For assessment, telehealth allows HHPs to conduct hearing screenings, diagnostic evaluations and follow-ups remotely. Based on the purpose of the assessment, HHPs will need to determine the technological requirements to successfully complete each procedure.

As discussed in Chapter 4, remote hearing screening can be conducted using portable audiometers connected to smartphones or tablets based on the available resources at each site. These smart devices are particularly effective in community-based settings such as daycares or schools, allowing for the early identification of HL.

Diagnostic assessment using telehealth

Diagnostic assessments, including pure-tone audiometry and tympanometry, can also be conducted remotely with trained facilitators supporting on-site equipment use and using asynchronous or hybrid methods. Ramkumar and Krumm (2023) established a viable telehealth model for remote diagnostic testing, involving remote control and access of audiology equipment and videoconferencing software. Their diagnostic test battery included video-otoscopy, tympanometry, DPOAE and AABR tests. Using a mobile telehealth van with satellite connectivity, an HHP conducted the diagnostic assessment synchronously via telehealth. Despite some challenges related to ABR assessment accuracy, the telehealth model was associated with higher follow-up rates suggesting the potential for enhancing patient adherence and continuity of care in contexts where resources are limited and HHPs unavailable (Ramkumar and Krumm 2023).

As mobile technology continues to evolve rapidly, the integration of this technology presents a promising and cost-effective approach for detecting middle ear conditions, particularly in low-resourced regions where the availability of specialised professionals remains scarce.

Teletherapy for Children with Hearing Loss

The delivery of telehealth intervention (that is, teletherapy) services for children with HL has gained popularity in recent years. Teletherapy can provide access to specialised AVT and (re) habilitation services to support listening, speech and language development while adhering to the principles of FCC. Teletherapy leverages digital technologies to deliver high-quality, personalised care, overcoming geographical and logistical barriers that often prevent access to traditional in-person (re)habilitation services. By using telehealth, HHPs can offer timely and effective support to children and their families, while ensuring that critical developmental milestones are met, and children are able to reach their full potential.

Principles and outcomes of AVT in teletherapy

AVT focuses on developing listening and spoken language skills in children with HL through active participation by caregivers (Chapter 12). In teletherapy, auditory-verbal therapists (for example, certified listening and spoken language therapists and educators) guide families to implement strategies that maximise auditory input and encourage their child's spoken language and communication. There is growing research that describes the effectiveness of telehealth (re)habilitation services for children with HL after they have received appropriate hearing amplification (that is, HAs and/or CIs) across diverse contexts and age groups (for example, Grigsby et al. 2024). Recent studies have demonstrated that telehealth interventions are not inferior to traditional in-person rehabilitation when measured by intervention outcomes, such as language outcome measures (Grigsby et al. 2024).

Best practice principles for telehealth delivery for children with HL

Telehealth continues to have huge potential to bridge gaps in service delivery, yet it is still underused and underexplored in paediatric audiology care. As such, delivering hearing care services remotely requires some adjustments to traditional approaches that suit the telehealth model of service delivery. Table 14.2 below presents key considerations for HHPs interested in setting up a successful telehealth service for children with HL. These

considerations include a series of best practice principles for telehealth delivery, including assessment and intervention sessions, and cover the technology setup, environment and telehealth approaches. Table 16.1 presents foundational evidence-based principles that must always be considered when implementing telehealth for hearing care, including the assessment and intervention of children with HL.

Table 16.1: Best practice principles for telehealth assessment and intervention

Physical Setup
Location and Environment (for example, participants, furniture, lighting, sound, background) • Choose the most appropriate location for telehealth sessions. This entails setting up a quiet, well-lit space free from distractions, with the camera positioned to capture every session participant (for example, the child, parents/caregivers, HHPs, siblings, pets). • Determine how many people will participate in each session and define their role. • Consider whether the room environment will be appropriate for the equipment needed (for example, furniture on wheels, soft surface tabletops). • Ensure that the room is away from high-traffic areas with excess environmental noise. • Check the flooring, wall covering, lighting and room dimensions. All these will impact the environment for telehealth sessions. • Make sure both the HHP and client(s) have an appropriate background so people and objects can be clearly seen on the screen. The HHP can ask clients to remove any visual/auditory distractions. • Include aural variety (for example, voice pitch and tone, videos, music) so clients can access diverse sources of auditory stimuli. **Professionalism** • Dress professionally to present a clear image to clients and avoid distracting accessories or clothing items. • Look and sound professional on the screen. Telehealth is as good as in-person services, so the HHP must maintain the same high-quality standards.

Resources (hardware and software)

Hardware

- Where possible, use high-definition cameras (preferably external webcams with high definition, wide viewing angle and auto-focus). Image quality needs to be clear for clients to see objects, images, facial expressions and body language clearly.
- Use high-quality microphones , as crisp and clear audio is more important for client engagement than a high-definition image. Clients need to hear conversation and stimuli without any interference or noise.
- Use dedicated headphones (for example, with active noise-cancelling) and a microphone (for example, headset, lapel mic, desktop mic) to ensure clear audio transmission.
- Optimise the audio settings by reducing background noise and adjusting microphone sensitivity to ensure clear, effective communication.

Videoconferencing software

- Ensure that clients' data is safeguarded from any breaches. Although the Health Insurance Portability and Accountability Act (HIPAA) regulations apply to providers in the United States, it is highly recommended that videoconferencing software is HIPAA compliant.
- Always choose a secure software that suits both the HHP and the client, considering pricing options, features and internet bandwidth.
- Keep children engaged during sessions by using videoconferencing platforms that include screen-sharing tools supporting interactive activities, such as storybooks, digital flashcards, visual promptsand rewards.

Planning and preparation

Preparing the client and setting expectations

- Always prepare clients ahead of the first session. Provide basic information about the telehealth appointment and discuss logistical practicalities to ensure a successful first experience.
- Conduct technical tests to troubleshoot connectivity or audio issues before the first formal session.
- Familiarise parents/caregivers and clients with platform features to ensure a smooth experience (for example, mute/unmute, turn camara on/off, screen sharing).
- Take some time to warm up the session, as there is no 'catch-up' time while walking to the room.
- Set clear expectations for the first session. This includes meeting time, date, duration of the appointment and platform to be used; resources needed for the first session and an agenda of topics to be covered; participants who will join the session; and troubleshooting procedures if needed.
- Talk clients through the first telehealth session as 'seeing the HHP' online may still be foreign to the clients. Only if they understand why and how a telehealth session is to take place, will they be at ease and engaged during the interaction.

Shared decision making and collaborative practice

- Ask for the client's input to make them feel part of the interaction, as their personal values, wishes and expectations for the intervention will be integrated into decision making and goal setting processes, enhancing their engagement.
- Consider each family's strengths and needs and discuss their short- and long-term goals as a result of the intervention process. The delivery of child- and FCC via telehealth has reported increased client engagement and outcome achievement for families of children with communication difficulties (for example, HL, speech and language disorders, autism) among others.

Planning and preparation

Setting roles and expectations

- Set out the roles and expectations for all participants early on and revise them as the nature of the appointment changes. This is important because of the lack of physical contact during telehealth, so that parents/caregivers are expected to have a more hands-on role during telehealth sessions.
- Slowly build on the parents'/caregivers' confidence and skills so that they can take a more active role as the telehealth intervention progress.
- Adopt the role of a parent/caregiver coach during telehealth intervention sessions for young children with HL and other communication difficulties, guiding families on how to best support their children. .
- Ask the family who they would like to involve during telehealth appointments (especially for intervention sessions). One of the advantages of telehealth is the ease with which additional family members and other persons besides primary caregivers can be involved.
- Remember that the success of teletherapy depends on strong and effective collaborative partnerships with families. Parents and caregivers are coached to use intervention strategies and techniques during daily activities, so it is the HHP's responsibility to actively involve them in sessions.

Materials and activities

- Planning therapy activities with clients and families via telephone, email, or videocall ahead of telehealth sessions will help both HHP and families to gather and prepare appropriate resources and materials.
- Although specific equipment and devices are needed for telehealth assessment (for example, remote control of devices), telehealth intervention typically requires less expensive and specific resources and more creativity from both professionals and clients.
- Using telehealth for intervention does not mean that all resources must be digital, complex and sophisticated (for example, videos, websites, Apps, online resources). Simple, tangible and concrete materials can also be used effectively and may even be better for younger clients with HL.
- Although not entirely unique for children with HL, embedding therapy goals into family routines enables natural opportunities for language development and practice. Storytelling and shared book reading also strengthen listening comprehension and narrative skills.
- For children with HL, auditory information is particularly important, so both input and output must be clear for children and families for them to access to and practice listening and communication skills.
- Clients may benefit from having the same materials as the HHP (for example, duplicate objects), including technology, information, and resources.
- Resources and materials should be clearly seen on the screen. Always consider the complexity, colour/contrast, size, handling and meaning of these resources to increase their effectiveness.

Communication and Interaction

Communication skills

- The most important skill during telehealth interaction is effective and clear communication. Clarity of speech, eye contact, turn-taking and listening skills must be mastered to compensate for the lack of physical contact and cue reading.
- Clear, specific, open and honest communication will facilitate and support effective interaction with clients and other participants during telehealth.
- Use modelling and imitation and then observe. First, HHPs must think about the concept they want to introduce and create a way of demonstrating it through the screen. Once they have shown the client/family this concept, the HHP can ask them to imitate and replicate it immediately so they know that clients/families have fully understood this new concept/skill. Once they have understood and demonstrate this new learning, the HHP can sit back and observe a family's interaction while providing timely support and guidance.

Client and family engagement

- Many engagement-building strategies used in in-person settings can also be used in telehealth interactions. However, there are several unique features of engagement that the HHP may need to deliberately demonstrate while interacting with children and families during telehealth sessions. For example, the way in which therapeutic relationships are built remotely, the communication style used during the interaction, the telehealth environment and the use of resources available from the family's home are all aspects relevant to telehealth engagement (see Retamal-Walter, Waite and Scarinci 2023a, 2023b).
- Telehealth sessions with children often require creative problem solving to maintain their engagement. Strategies to keep children engaged include:
  - Incorporating movement-based activities, such as dancing to music, to balance sitting and active time.
  - Scheduling breaks or using shorter session durations for children with limited attention spans.
  - Using visual supports, such as visual schedules, pictures and/or props, to capture their interest and assist their understanding.

Flexibility

- HHPs must be prepared to adjust intervention goals dynamically based on session flow, child and family feedback and outcome achievement. This flexibility ensures that therapy remains effective and family centred.

Telehealth etiquette

- Maintain privacy and confidentiality as in any other appointment. HHPs must introduce themselves and the other people present 'in the telehealth room', set clear expectations for the assessment/intervention process and set boundaries for the therapeutic relationship to grow and remain professional.
- Adjust interactions to suit the individual cultural and personal characteristics of the client and family.
- Follow up on pending items from previous interactions and outline an agenda for each session, so all participants are aware of the topics to be covered.
- End each session formally, making sure that enough time is allocated to review and plan for the next session. The end of a telehealth session includes asking for feedback, answering questions, reviewing key points, discussing follow-up plans, setting goals and confirming appointment details, including referrals and action items.

Staffing and training

Personnel training

- HHPs will need to be upskilled in using the equipment and procedures effectively via telehealth. This may be in the form of self-paced and/or specific courses delivered by telehealth experts. Many organisations include telehealth training during onboarding processes and continuing professional development.
- Skills needed for telehealth include research and implementation, and effective communication.

Troubleshoot and support

- HHPs must remain calm when things do not go as planned. Ensure there are support mechanisms available to troubleshoot technology issues that will inevitably arise during telehealth sessions.
- Best telehealth practices include discussing internet breakdowns and backup plans before starting any audiological procedure via telehealth.
- Clients continue to see telehealth as a 'second best option' after in-person services. Therefore, customer experience is essential in keeping clients accountable for, motivated by and engaged in telehealth services.

Reflective practice and creativity

- HHPs should regularly review telehealth sessions and reflect on how sessions go. When reviewing sessions, HHPs must see them through a provider and client lens to identify areas for improvement during future interactions.
- Advice from more experienced professionals in telehealth delivery allow HHPs to refine telehealth skills and service delivery.
- HHPs should remain flexible and creative while delivering assessment and intervention via telehealth. HHPs must be able to transfer knowledge, expertise and skills from previous in-person interactions into a different model of service delivery.

Overcoming Challenges of Telehealth in Paediatric Audiology

Implementing telehealth services in paediatric audiology presents multifaceted challenges, particularly in resource-constrained settings where systemic barriers can impede service delivery.

Limited technology access and digital literacy

Access to technology remains a significant barrier to the implementation of telehealth in paediatric audiology, particularly in resource-constrained settings where reliable internet connectivity and access to devices such as smartphones, tablets and computers are often unavailable. This challenge is particularly prevalent in rural and underserved areas where digital infrastructure is underdeveloped or prohibitively expensive, limiting families' ability to engage with telehealth services and exacerbating disparities in healthcare access (Al-Worafi 2024; Swanepoel, Olusanya and Mars 2010). Studies have highlighted the lack of high-speed broadband networks, unreliable internet connections, and limited information technology support as key barriers to telehealth implementation in such regions (McCarthy et al. 2019). Limited digital literacy among families poses additional challenges (Alami, Gagnon and Fortin 2019). Families who are unfamiliar with digital tools or virtual communication platforms will potentially struggle to navigate telehealth systems, reducing their ability to access care effectively.

Targeted and practical solutions should be considered to address the issues of reduced technology access and digital literacy. For example, community resources, such as public libraries, schools and health centres, can serve as hubs for families to access the internet and telecommunications technology in secure and supportive environments. Such hubs could include staff or volunteers trained to assist families in navigating telehealth platforms, ensuring that technology use becomes less intimidating. Mobile health units equipped with telehealth technology offer another practical solution, bringing services directly to remote and underserved populations. Finally, asynchronous telehealth methods could also be used. For example, preloaded instructional videos, USB drives and other offline resources can support families who are without reliable internet connections, thereby ensuring continuity of care.

Financial constraints

Financial constraints may also significantly impact the implementation and accessibility of telehealth services in paediatric hearing care, particularly in resource-constrained settings. Families may face prohibitive costs associated with purchasing devices, maintaining internet connectivity or traveling to community-based telehealth hubs. In low-income contexts, even minor expenses such as transportation or data costs can limit access to services (Bhamjee et al. 2022). Additionally, resource-limited healthcare systems encounter challenges in funding the infrastructure, equipment and training required for telehealth implementation, with high initial setup costs for items such as video conferencing systems, further exacerbating the issue (Bennett et al. 2023). While some countries have established structured reimbursement models, inconsistencies and gaps in reimbursement often hinder equitable access and long-term sustainability (McCarthy et al. 2019). Without clear legislation and regulations to support sustainable funding, financial barriers continue to restrict access to telehealth for both families and healthcare systems (Bennett et al. 2023).

Workforce limitations

Workforce limitations, including shortages of skilled HHPs, present significant challenges to the consistent delivery of telehealth in paediatric hearing care, particularly in rural and underserved areas. In many countries, HHPs are concentrated in urban centres, with retention in rural areas remaining a persistent issue (Al-Worafi 2024). Furthermore, HHPs in rural settings often lack specialised expertise in paediatric audiological practice, exacerbating gaps in care. These limitations are particularly pronounced in resource-constrained settings, where insufficient healthcare workforce capacity and inadequate training further hinder the implementation of telehealth services (Bhamjee et al. 2022).

Building engagement and relationships

A key challenge in implementing telehealth for paediatric audiology is fostering engagement and the therapeutic relationships necessary for effective care. The virtual nature of telehealth can make it more difficult for professionals to build rapport with families (Retamal-Walter, Waite and Scarinci 2023a), particularly when non-verbal cues are limited or when technical issues, such as audio or video lag, disrupt the natural flow of communication. Furthermore, young children may demonstrate variability in engagement in telepractice sessions

(Retamal-Walter, Waite and Scarinci 2023b). Additionally, support for families may be difficult due to the limited ability for the HHP to provide hands-on guidance typical of traditional appointments (Retamal-Walter, Waite and Scarinci 2023a). Preferences for in-person care are often reported, as parents and caregivers value the direct, physical presence of clinicians for reassurance and troubleshooting, particularly when managing complex hearing devices or learning new intervention strategies (Mardinli et al. 2023). These challenges can lead to lower levels of engagement, reduced trust, and, ultimately, a potential impact on the effectiveness of audiological interventions delivered remotely.

To address these challenges, a family-centred and creative approach to telehealth delivery is critical. HHPs can enhance engagement by incorporating interactive, child-friendly activities that hold a child's attention and enable connection with the hearing care professional. For example, telehealth sessions can include play-based learning strategies, familiar toys or games to encourage participation and comfort (Retamal-Walter, Waite and Scarinci 2023b). Allowing additional time during interactions and enhancing verbal communication using facial expressions, vocal variations and physical cues can further increase children's engagement (Retamal-Walter, Waite and Scarinci 2023b). Within a family-centred model of care, audiologists can use telehealth to actively coach caregivers in supporting their child during assessment, device management or intervention, empowering families to take on an active role in their child's care. Clear and intentional communication is also critical, demonstrated through the setup of expectations, using warm and personable language, and maintaining consistent follow-ups to demonstrate reliability and support (Retamal-Walter, Waite and Scarinci 2023b).

Furthermore, hybrid models of care can address preferences for in-person interactions by allowing for periodic in-person sessions combined with telehealth services. Hybrid models provide opportunities for clinicians to develop rapport during in-person visits and then maintain engagement remotely (Retamal-Walter, Waite and Scarinci 2023a). Additionally, strategies such as pre-session preparation and personalised follow-up emails, videos or additional planning sessions conducted via telehealth can help families feel more connected and supported throughout the telehealth process (Retamal-Walter, Waite and Scarinci 2023b).

Ethico-legal concerns

Ethico-legal considerations are critical to the successful implementation of telehealth in

paediatric hearing care. Ensuring the privacy, security and confidentiality of patient data presents unique challenges, particularly if families rely on shared devices or public internet access. Differences in legal frameworks across countries or regions further complicate service delivery and accountability, as providers may need to navigate regulatory requirements outside their own jurisdictions (Swanepoel, Olusanya and Mars 2010). Additional concerns include the absence of comprehensive policies, protocols or guidelines that govern telehealth practices (Bhamjee et al. 2022) and fears surrounding the misuse of patient data by governments or for commercial purposes (Alami, Gagnon and Fortin 2019). Insecure telehealth systems are also vulnerable to cybersecurity threats, which can lead to data breaches and legal repercussions. Addressing these challenges is essential to maintain trust, ensure ethical care and prevent inequities in telehealth delivery.

Proactive measures are needed to address these ethico-legal concerns and ensure that telehealth practices meet global standards. Implementing secure telehealth platforms with robust encryption, firewalls and secure protocols for data storage and access is essential for protecting patient data. Further, clinicians must make sure they adhere to national and regional privacy laws that provide a framework for responsible data handling. Informed consent processes should be adapted to telehealth, using culturally relevant and simple language to explain patient rights and responsibilities. Addressing licensure barriers is equally critical. International and national agreements are needed to standardise telehealth practices across jurisdictions, and to enable providers to work effectively under differing regulatory frameworks. Training clinicians in the ethical issues and responsibilities in telehealth further supports the delivery of respectful and secure care. The checklist in Appendix 16.1 can be used to ensure that HHPs are considering ethico-legal considerations in their telehealth practice.

In addition to the checklist for ensuring compliance, Table 16.2 provides some examples of professional guidelines and regulatory frameworks for ensuring ethical, secure and effective delivery of telehealth services in audiology for various regions.

Table 16.2: Examples of telehealth guidelines and regulatory frameworks

Country	Professional Guidelines	Data Security Legislation/ Standards
Australia	Australian Teleaudiology Guidelines (Audiology Australia)	Australian Privacy Principles/ Privacy Act
Canada	SAC Position Paper on The Use of Virtual Care in Speech-Language Pathology and Audiology Services (Speech-Language and Audiology Canada)	Personal Information Protection and Electronic Documents Act (PIPEDA)
South Africa	General Ethical Guidelines for Good Practice in Telehealth (Health Professions Council of South Africa)	Protection of Personal Information Act (POPI Act)
United Kingdom	A guide to remote working in Audiology (British Academy of Audiology)	The UK General Data Protection Regulation (GDPR)
United States	Position Statement: The Use of Telehealth for the Delivery of Audiological Services (American Academy of Audiology)	Health Insurance Portability and Accountability Act (HIPAA)

Implementation Frameworks for Telehealth in Paediatric Audiology

Several frameworks can be considered in the development of telehealth services, to guide effective and sustainable implementation. These frameworks consider aspects of access, training and acceptability, and therefore can be used to mitigate the challenges to telehealth implementation discussed in this chapter. Two relevant frameworks are outlined in this section.

Access framework

While not designed for telehealth, Penchansky and Thomas' Theory of Access (1981) conceptualises access to healthcare as a multidimensional construct. The five elements of this theory could be used as a framework to guide the development of telehealth services by ensuring each dimension is addressed in the planning and implementation phases:

1. Availability: The service is available to those needing it. Assess the resources required to deliver telehealth services, such as trained HHPs, technology and infrastructure, ensuring they are sufficient to meet the demand in the target population.
2. Accessibility: The user has access to the service. Evaluate geographical and technological barriers that might prevent families from accessing services and implement solutions like internet subsidies or telehealth hubs in schools or community centres.
3. Adequacy: The service responds to the needs of the community. Design services that are flexible and user-friendly, accommodating the needs of families with varied schedules and caregiving responsibilities.
4. Affordability: The cost of the service is considered. Develop funding models to reduce cost barriers, such as partnering with government programmes or NGOs to subsidise telehealth technology and services.
5. Acceptability: The service is considerate of the community's cultural and attitudinal needs. Engage stakeholders to ensure services are culturally sensitive, trusted, and aligned with the preferences of families and HHPs.

I2I-4-Telehealth

The innovation to implementation for telehealth, or I2I-4-Telehealth (Theodoros et al., 2016), is a knowledge translation (KT) tool adapted to help start telehealth services. It begins by establishing a foundation for change, which includes clearly defining the telehealth innovation, identifying key stakeholders and developing a comprehensive plan to communicate with and engage these stakeholders. The process of implementing telehealth innovation operates alongside this foundation, requiring a coordinated change management strategy that complements the KT activities. It comprises seven steps guided by key focus points:

1. State the purpose of the KT plan: Clearly define the main goal of the KT plan, focusing on what needs to change and what success would look like. This helps establish the rationale for implementing telehealth and secures buy-in from stakeholders.
2. Select an innovation: Identify a specific, feasible telehealth innovation that addresses the purpose of the KT plan. This includes ensuring the innovation has a strong evidence base and aligns with organisational and community needs.
3. Specify key players and actions: Identify all stakeholders involved in the implementation process and outline the specific actions they need to take. This ensures clarity about roles and responsibilities.
4. Identify agents of change: Determine influential individuals or groups who can advocate for and drive the adoption of the telehealth innovation. These could be opinion leaders, champions or early adopters.
5. Design the KT Plan: Develop a comprehensive plan for implementing the innovation, including the methods, materials and strategies to engage stakeholders and encourage adoption.
6. Implement the KT plan: Put the plan into action, either at once or in phases. Use feedback from stakeholders to adjust the implementation as needed to improve acceptability and feasibility.
7. Evaluate success: Assess the impact of the telehealth innovation and the KT plan based on specific metrics such as reach, effectiveness, adoption, implementation and sustainability.

These systematic steps, together with an example relevant to paediatric audiology, are presented in Figure 16.3.

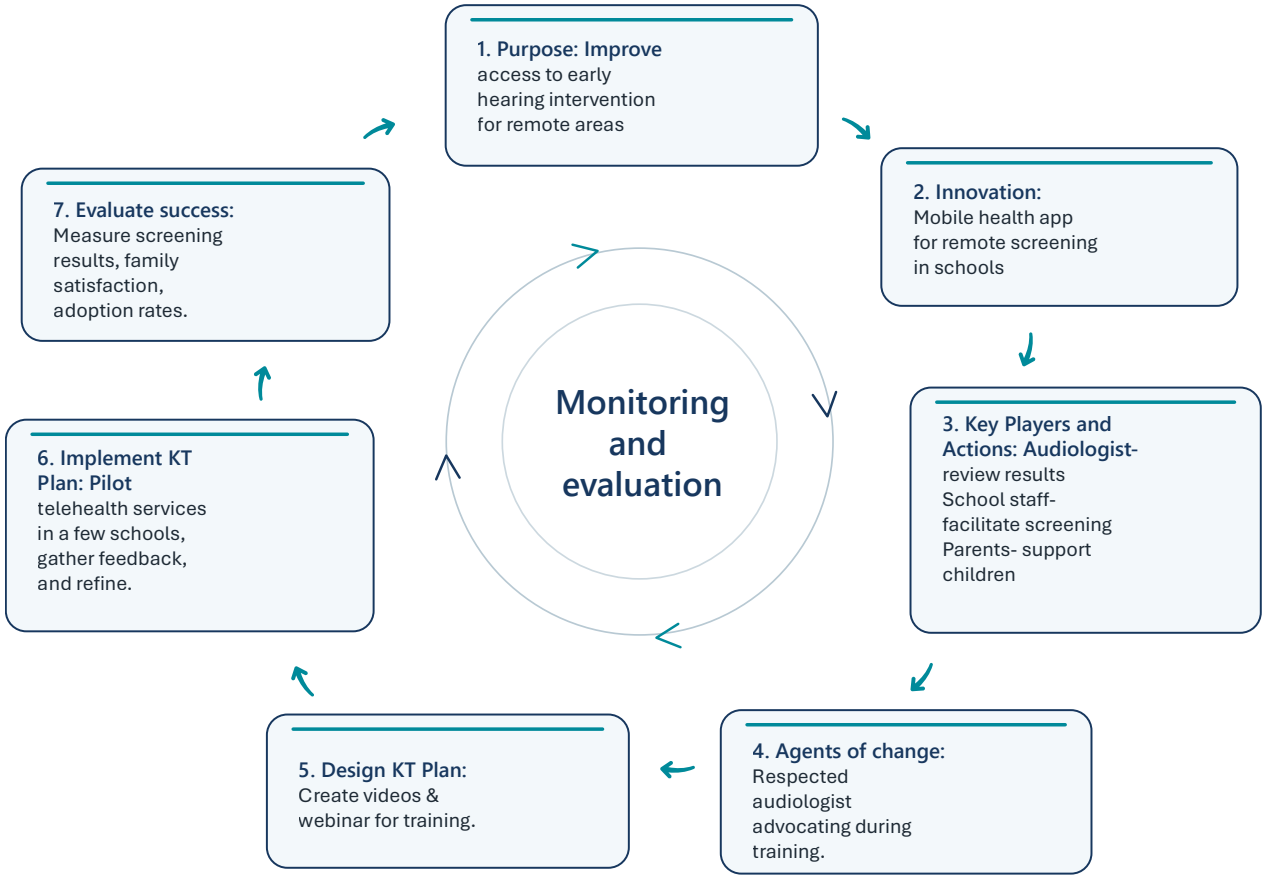

Figure 16.3: Application of the I2I-4-Telehealth in paediatric audiology

By addressing these dimensions systematically, the framework ensures telehealth services are equitable, sustainable and capable of meeting the diverse needs of the community.

Continuing professional development in telehealth audiology

Staying up to date with best practices in telehealth is essential for HHPs supporting children with HL. As service delivery models continue to evolve, so too must the skills and confidence of the HHP in using technology to engage families and deliver effective intervention. Ongoing professional development helps ensure that HHPs remain responsive to emerging evidence, with the knowledge and skills to provide high-quality, evidence-based and family-centred services across a range of settings.

Examples of reputable online platforms that offer comprehensive continuing professional development opportunities in telehealth for HHPs are listed below:

- Audiology Online (*www.audiologyonline.com*) has collated a number of professional development courses for HHPs to prepare for telehealth assessment and intervention. The course catalogue for Telehealth (*https://www.audiologyonline.com/audiology-ceus/telehealth/*) offers over 40 introductory, intermediate and advanced courses for professionals interested in offering audiology services at distance both for children and adults.
- For speech pathologists, there are over fifteen dedicated courses for professionals working via telehealth with diverse populations and communication difficulties (*https://www.speechpathology.com/slp-ceus/all/#/category_id:telehealth-and-telepractice*).
- It is important to check what training and courses are available to you as part of your affiliation and/or membership to professional associations at the national and international levels.

Conclusion

Telehealth presents an exciting and transformative opportunity for HHPs to expand their reach and enhance service delivery in paediatric audiology. By addressing barriers such as geographic isolation, accessibility and cost, telehealth ensures equitable and timely care for children and families who might otherwise struggle to access vital hearing services. With

growing evidence demonstrating comparable outcomes to in-person care, telehealth empowers the HHP to deliver high-quality, family-centred interventions. Embracing telehealth with a commitment to best practices and continuous professional development enables HHPs to overcome challenges and leverage technology to make a meaningful impact on children's auditory and communication development. The integration of telehealth into clinical practice enhances efficiency and outcomes and also highlights the profession's adaptability and innovation in meeting the diverse needs of families worldwide.

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Appendix 16.1: Checklist for ensuring compliance and best practices in telehealth

a) Licensure Requirements and Professional Guidelines

- ☐ Ensure licensure or certification in the jurisdiction where the client is located.
- ☐ Verify compliance with professional guidelines from relevant organisations.
- ☐ Follow evidence-based standards for telehealth to provide care equivalent to in-person services.

b) Patient Consent

- ☐ Obtain informed consent from clients before delivering any telehealth services.
- ☐ Clearly explain the nature of the telehealth visit, including potential benefits, risks, and deviations from in-person care.
- ☐ Inform client of their right to withdraw from the service at any time.
- ☐ Document the consent appropriately for legal and ethical compliance.

c) Privacy and Data Protection Regulations

- ☐ Ensure compliance with relevant regulations or legislation.
- ☐ Ensure the use of telecommunication tools that provide encryption and firewalls to ensure data protection.

- ☐ Implement access controls to ensure only authorised personnel can view client information.
- ☐ Conduct regular risk assessments to identify and mitigate potential vulnerabilities in data security.
- ☐ Train staff on relevant regulations and secure handling of client data.

Chapter 17

Conclusion

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'Children require the highest standard of care, where efficiency cannot compromise precision.'

~World Health Organisation, 2025

Overview

Integrated, person-centred and evidence-based approaches are advocated throughout this book to ensure accessible and equitable hearing services for the paediatric population, especially in diverse settings.

This book provides Hearing Healthcare Professionals (HHPs) with insights into the multi-faceted nature of paediatric hearing loss (HL), its varied causes, evolving prevalence and developmental implications (Chapter 2). A deep dive into the anatomical and physiological underpinnings of the auditory and vestibular systems further reinforces the importance of clinical precision (Chapter 3). These insights are linked to broader programmatic efforts, such as hearing screening protocols (Chapter 4) and behavioural (Chapter 5), electrophysiological (Chapter 6) and vestibular assessments (Chapter 7). Auditory Processing Disorder (APD), because it shares certain symptoms and characteristics with other neurodevelopmental

conditions, presents a distinct diagnostic and therapeutic challenge (Chapter 8). These assessment findings provide a quantifiable foundation for diagnostic and rehabilitative decisions and should be approached with both technical expertise and ethical responsibility.

Accurate identification requires a multidisciplinary approach involving collaboration among audiologists, speech-language therapists, ear-, nose- and throat (ENT) specialists, psychologists and educators. The importance of such teamwork is reinforced throughout the book, highlighting how structured and solution-focused interdisciplinary collaboration (Chapter 14) leads to more effective intervention outcomes. This collaborative ethos extends to the management of conductive hearing loss (CHL), where surgical intervention may be required (Chapter 9). Seamless referral systems and strong interprofessional relationships, especially between audiologists and ENT specialists, are critical to ensuring timely, coordinated and effective care for children requiring medical or surgical treatment.

It is of utmost importance that HHPs prioritise early, individualised and family-centred intervention in managing the identified hearing and/or vestibular pathology. Hearing aid fitting (HAF) is not a one-time event but a dynamic process requiring precise verification, routine monitoring and close collaboration with families (Chapter 10). When hearing aids (HAs) are insufficient to provide quality auditory access, timely referral for implantable devices becomes crucial (Chapter 11). These interventions, when implemented early and supported through comprehensive (re)habilitation, lead to optimal auditory brain development and enhance long-term communication, and educational and social outcomes for the paediatric population.

The single most important intervention decision after the diagnosis of a child's HL is the family's choice of language — the language of their heart. This decision has lifelong implications, influencing not only how a child communicates but also their educational access, social identity and developmental trajectory. The Auditory Verbal Therapy (AVT) approach is one possible pathway. This approach acknowledges caregivers as the primary agents of change in their child's life and involves coaching by the HHP (Chapter 12). Building on these principles, other communication options, such as Cued Speech and Signed Language, as well as the broader educational landscape for children with HL, are discussed (Chapter 13). There are multiple schooling options, such as mainstream inclusion, facilitation or special education. However, across all educational settings, the consistent thread is the critical role of family commitment and support.

The holistic nature of paediatric audiological care is driven by collaboration, empathy and innovation in order to ensure better outcomes. Establishing structured communication systems, regular meetings and shared protocols improves clinical efficiency, and also places the child and family at the heart of decision-making (Chapter 14). This theme is deepened in Chapter 15, which explores Person and Family-Centred Care (P/FCC) as a guiding philosophy that recognises the lived experiences, values and voices of both children and their families.

Acknowledging the diverse nature and availability of resources across the globe, Chapter 16 offers guidelines on the use of telehealth to overcome geographical and resource-related barriers, supporting equitable, high-quality care that aligns with both P/FCC principles and the growing demand for innovative, flexible service delivery. By integrating theoretical foundations with applied clinical practice, the book aims to equip readers with the tools necessary for effective assessment and intervention.

Conclusion

In conclusion, paediatric audiological care demands a balance of evidence-based precision, contextual awareness and family-centred compassion. This book has illustrated how theory and practice intersect, showing that early, individualised intervention supported by strong interprofessional collaboration leads to the best outcomes for children with HL. By integrating diverse perspectives, case studies and global approaches, the content reinforces the adaptability required in real-world clinical contexts. The consistent emphasis on empathy, innovation and partnership with families reflects the broader vision of person-centred care aligned with global health goals. Ultimately, readers are encouraged to carry forward these principles in their practice to ensure that every child has equitable access to effective and life-changing hearing care.

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Louis Murray Hofmeyr is a qualified medical doctor (MBChB) and practices as an Otolaryngologist in Western Cape (South Africa). He has managed more than 800 patients with vestibular schwannomas (acoustic neuromas), one of Africa's most extensive case series and has performed over 400 cochlear implants. He has also described and managed the first patient in South Africa with superior canal dehiscence syndrome (SCDS) in 2006 and has since treated and operated on more than 200 patients with SCDS. Dr Hofmeyr currently

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Silva Kuschke is a clinical audiologist and researcher based in the Cayman Islands, and also holds an adjunct Assistant Professor of Audiology appointment at Vanderbilt University (Tennessee, USA) and an honorary Senior Lecturer position at the University of Cape Town in South Africa. Before her relocation to North-America and the Caribbean, she was Senior Research Audiologist at the hearX Group (South Africa), the Chief Audiologist and Head of the Audiology Department between April 2016 – February 2023 at the Red Cross War Memorial Children's Hospital in Cape Town (South Africa); the largest dedicated academic paediatric hospital in sub-Saharan Africa. She enjoys working with children, providing support to families with hearing-impaired children, clinical teaching, and lecturing. Her research interests are conducting context-specific and clinically relevant research into childhood hearing loss, with a particular focus on decentralising hearing healthcare to make it more accessible in low- resourced contexts.

Susan Strauss is an audiologist and the owner of Stellenbosch Audiology, a well-established private practice in Stellenbosch, South Africa. She has been in the profession since 1995, with clinical and academic experience across a broad range of audiological services. Her areas of special interest include paediatric audiology, hearing aid fitting, and evidence-based hearing technology. She holds a PhD in Audiology, with her research focused on noise-induced hearing loss. Passionate about quality care and professional standards, she remains actively involved in leadership and ongoing development within the field.

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List of abbreviations

AABR: Automated Auditory Brainstem Response
ABI: Auditory Brainstem Implant
ABR: Auditory Brainstem Response
AEP: Auditory Evoked Potential
ALD: Assistive Listening Device
ANSD: Auditory Neuropathy Spectrum Disorder
AP: Auditory Processing
APD: Auditory Processing Disorders
ASG: Adaptive Situational Gain
AOM: Acute Otitis Media
AUC: Area Under Curve
AVT: Auditory-Verbal Therapy
aVOR: angular Vestibulo-Ocular Reflex
BCHD: Bone Conduction Hearing Devices
BCI: Bone Conduction Implant
Bi-Bi: Bilingual-Bicultural
BTE: Behind-The-Ear
CAPS: Curriculum Assessment Policy Statements
CCC: Child Centred Care
CI: Cochlear Implantation
CHL: Conductive Hearing Loss
CHW: Community Healthcare Workers
CMV: Cytomegalovirus
CODA: Child of Deaf Adult/s
CROS: Contralateral Routing of Signal
CS/CL: Cued Speech/Cued Language
CSOM: Chronic Suppurative Otitis Media
dB HL: Decibel Hearing Level
DPOAE: Distortion Product Otoacoustic Emissions
DSL: Desired Sensation Level
EAC: External Auditory Canal
EAM: External Auditory Meatus
ECD: Early Communication Development

EHDI: Early Hearing Detection and Intervention
eTOT: Engagement in Telepractice Observational Tool
FCC: Family Centered Care
FFR: Frequency Following Response
GPDR: General Data Protection Regulation
HA: Hearing Aids
HAF: Hearing Aid Fitting
HHP: Hearing Healthcare Professionals
HIPPA: Health Insurance Portability and Accountability Act
HL: Hearing Loss
HoH: Hard of Hearing
HZ: Hertz
IEP: Individualised Education Plan
IPA: International Phonics Alphabet
ISTS: International Speech Test Signal
ITE: In-The-Ear
ITC: In-The-Canal
I2I-4 Telehealth: Innovation to implementation for telehealth
KMC: Kangaroo Mothercare
KT: Knowledge translation
LMICs: Low- and Middle-Income Countries
LoLT: Language of Learning and Teaching
LSL: Listening and Spoken Language
LUHU: Limited Usable Hearing Unilaterally
LTASS: Long-Term Average Speech Spectrum
MDT: Multidisciplinary Team
MEI: Middle Ear Implant
MPO: Maximum Power Output
MPANL: Maximum Permissible Ambient Noise Levels
NAL: National Acoustics Laboratories
NBHS: Newborn Hearing Screening
NICU: Neonatal Intensive Care Unit
NIHL: Noise-Induced Hearing Loss
OAE: Otoacoustic Emissions
OC: Ossicular Chain
OME: Otitis Media with Effusion

PCC: Person-Centered Care
PC: Person Centeredness
PEACH: Parents' Evaluation of Aural/Oral Performance in Children
PHC: Primary Health Care
PIPEDA: Personal Information Protection and Electronic Documents Act
POPI Act: Protection of Personal Information Act
RECD: Real Ear-to-Coupler Difference
REUR: Real Ear Unaided Response
RIC: Receiver-In-Canal
RM: Remote Microphone
ROC: Receiver Operating Characteristic
SASL: South African Sign Language
SCDS: Semicircular Canal Dehiscence Syndrome
SCMm: Sternocleidomastoid muscle
SD: Standard Deviation
SFOAE: Sustained-Frequency Otoacoustic Emissions
SII: Speech Intelligibility Index
SL: Sign Language
SLP: Speech-Language Pathologist
SNHL: Sensorineural Hearing Loss
SMS: Short Message Service
SNR: Signal-to-Noise Ratio
SOAE: Spontaneous Otoacoustic Emissions
SPD: Spatial Processing Disorder
SPL: Sound Pressure Level
SRT: Speech Reception Threshold
TC: Total Communication
TEOAE: Transient Otoacoustic Emissions
UNHS: Universal Newborn Hearing Screening
UHL: Unilateral Hearing Loss
vHIT: Video Head Impulse Test
VEMP: Vestibular Evoked Myogenic Potential Testing
VOR: Vestibulo-Ocular Reflex
VT: Ventilation Tubes
WHO: World Health Organization

SUMMARY

Evidence-Based Practices in Paediatric Audiology is a comprehensive text designed for students, educators and researchers in audiology and hearing healthcare, as well as practitioners seeking to ground their work in current evidence.

Paediatric hearing healthcare is not a peripheral concern; it is central to child development, educational equity, and human potential. This book critically synthesises the research underpinning the identification, assessment and management of hearing loss in children, from birth through adolescence.

Each chapter examines the strength and quality of the available evidence and the theoretical frameworks that inform practice. Central to the book is the evidence that children identified with hearing loss and treated within the first six months of life can achieve language development on par with hearing peers by age five.

Throughout the book, the gap between international evidence and its application in resource-constrained contexts is critically examined. The book considers how evidence can be meaningfully interpreted and adapted to diverse linguistic, cultural and health-system contexts, and offers practical, evidence-informed solutions that support equitable and context-relevant services.

Ultimately, the book equips readers to critically appraise and apply evidence in paediatric audiology. In doing so, it is both a foundational academic resource and a call to educators, practitioners and policymakers to translate evidence into accessible, context-relevant services and ensure that no child's potential is diminished.

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