CHAPTER

02 Prevalence and Profiles of Paediatric Hearing Loss



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

~Socrates

Summary

This chapter describes childhood hearing loss according to seven parameters: 1) degree, 2) configuration, 3) laterality, 4) type, 5) stability, 6) longevity and 7) onset. The prevalence of childhood hearing loss is described according to a selection of these domains, as well as hearing loss caused by auditory neuropathy spectrum disorder. The chapter also includes information regarding the audiological profiles associated with various causes of hearing loss, such as atresia and microtia, otitis media, non-syndromic and syndromic hereditary hearing loss, congenital infections, ototoxicity, recreational noise-induced hearing loss, auditory processing disorder and non-organic hearing loss.

Learning objectives

From this chapter, the reader will be able to:

1. Define childhood hearing loss across multiple domains.
2. Explain the commonality of childhood hearing loss and how its prevalence varies across age and geography.
3. Identify audiological profiles of childhood hearing loss based on the aetiology of hearing loss.

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.

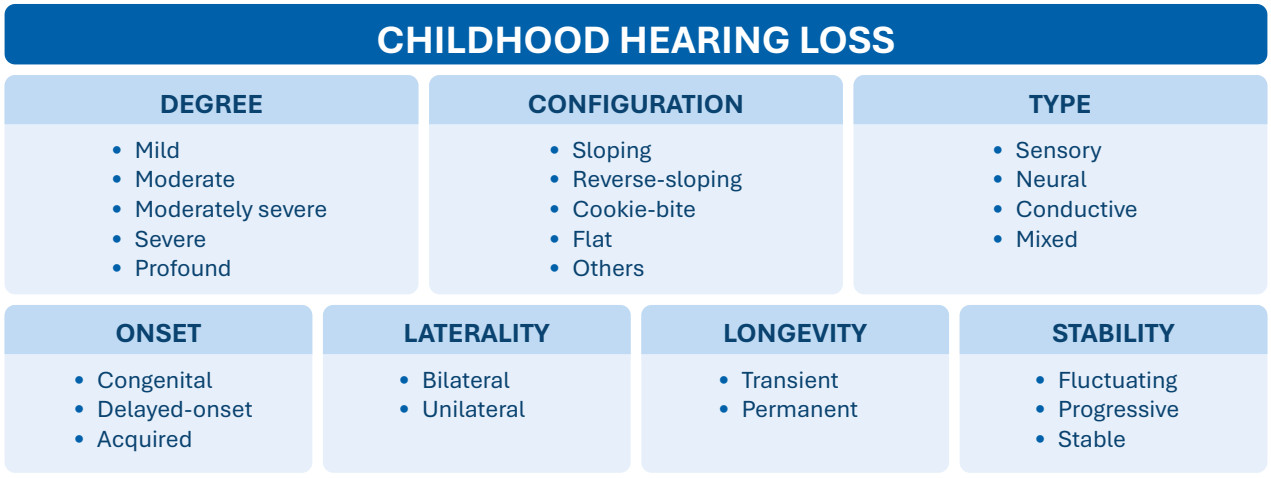


Figure 2.1. Domains of childhood hearing loss, adapted from Mackey (2022)

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).classifying degrees of hearing loss, 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.



Treasures of Wisdom

Description of HL

- In certain configurations, the degree of HL may vary across frequencies, specifying the degree according to frequencies may be better suited.
- Knowing when the HL likely began and its stability over time is critical for developing an individualized plan for monitoring hearing thresholds and planning intervention.

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



Treasures of Wisdom

Prevalence of HL

- Prevalence highly depends on the criteria used to define HL revalence will be much higher if slight, mild and transient HL are included.
- Since HL can develop at any stage of childhood, the prevalence increases from birth onward.

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.



Treasures of Wisdom

Atresia, microtia, OM

- For children younger than five years of age with microtia, non-surgical audiological options include a variety of hearing devices and should ideally be fitted by four months of age; children over five years of age are eligible for a surgically placed osseointegrated implant retention system.
- HL associated with OME may recover over time, however, the auditory deficit experienced during vital years for central nervous system development and language acquisition could have a functional impact later in childhood.

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

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.



Treasures of Wisdom

Audiological profiles for hereditary HL

- Usher syndrome is particularly relevant to audiologists due to its high prevalence among children with HL, and the dual impact of both HL and vision loss.
- In some cases of Pendred syndrome, HL progresses after a triggering event, such as minor head trauma, respiratory tract infection or sudden pressure change.
- Most children with Waardenburg syndrome have a stable HL, though the degree varies from mild to profound.
- Children with Down Syndrome have a higher risk of CHL because of wax impaction, OME or ear disease.

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 hearing

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



Treasures of Wisdom

Audiological profiles for meningitis, ototxicity and noise induced HL

- Quick audiological assessment and intervention is necessary to receive optimal outcomes for children with HL caused by meningitis.
- Children who have received ototoxic medications should continue receiving regular follow-up audiological testing for some years even if hearing is normal when treatment ends.
- There are several campaigns and programmes aimed at educating school children on HL prevention to improve knowledge and behaviours (for example, Dangerous Decibels).

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.



Treasures of Wisdom

Audiological profiles for other auditory-related conditions

- Children with APD have normal hearing thresholds but they struggle to understand what they hear. This occurs when the brain and ears don't fully coordinate, making it difficult for the brain to interpret sounds, especially speech.
- Non-organic HL emphasises the importance of using the cross-check principle in pediatric audiology.

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.

Test your knowledge:

Based on the content covered answer the following questions:

1. Describe the differences between congenital, delayed onset and acquired hearing loss and provide an example of a condition or cause for each type.
2. Explain why cCMV requires ongoing audiological monitoring, even in children who pass newborn hearing screening
3. Explain how hereditary non-syndromic and syndromic hearing losses differ in terms of onset, audiological profile, and predictability based on genetic information?

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