Section A -Research paper

Pediatric Hearing Loss Updates

Ahmed Mohamed El-Sayed Khater (1), **Ola Abdallah Ibraheem (2)**, **Dina Mamdouh Zein Elabdein (3)** and **Fatma Ahmed Salem Eljetlawi (4)**

(1) Professor & head of Audio-Vestibular Medicine, E.N.T. Department, Faculty of Medicine - Zagazig University

(2) Professor of Audio-Vestibular Medicine, Faculty of Medicine - Zagazig University

(3) Lecturer of Audio-Vestibular Medicine, Faculty of Medicine - Zagazig University

(4) (M.B., B.C.H), Faculty of Medicine-Misurata University-Libya

Corresponding Author: Fatma Ahmed Salem Eljetlawi

E-Mail: fgetlawy@gmail.com

Article History: Received: 11.06.2023 **Revised:**08.07.2023 **Accepted:** 19.07.2023

Abstract

Speech communication is uniquely exclusive to humans. With hearing, children have full access to the language of their home environment. If they have hearing loss or impairment, they are deprived of experiencing the sounds of nature and humans around them. Children who lose their hearing or have trouble hearing may struggle to learn and understand spoken language. This can make it difficult for them to communicate with others and navigate their environment. Hearing loss can impact a child's ability to develop age-appropriate skills in speech communication, language, and other activities. Pediatric hearing loss is a broad category that covers a wide range of pathologies. Early detection and prompt management are essential, as the development of language and psychosocial skills are significantly influenced by pediatric hearing loss. Additionally, early detection may identify potentially reversible causes or other underlying problems that can be managed. This informative article provides valuable insights into the latest research and advancements in the area of pediatric hearing loss. It explains how to assist children with hearing loss and enable them to lead a better life. The article keeps the readers informed about the latest developments in this field.

Keywords: children; hearing loss, speech communication

DOI: 10.53555/ecb/2023.12.1160

INTRODUCTION

Hearing loss (HL) in children is a broad category that covers a wide range of pathologies. Early detection and prompt management are essential for the development of normal language and psychosocial functioning, as well as for identifying potentially reversible causes or other underlying problems. There are three main types of hearing; conductive, sensorineural, and mixed. The former typically occurs due to a problem transmitting sounds at the level of the external or middle ear. The major cause of conductive HL in children is otitis media with effusion (**Dimitrov and Gossman, 2023).**

Sensorineural hearing loss (SNHL) results from a disruption of the auditory pathway at any point from the cochlea of the inner ear through the retrocochlear pathway. Despite being relatively uncommon in children as a whole, it is the primary cause of permanent HL in the pediatric population.

Etiology of Hearing Loss

HL can be broadly characterized as congenital or acquired in the pediatric population. It can be further classified in pre-, peri- and post-natal, considering its period of occurrence (**Ciorba et al., 2017).**

1. Congenital Causes

Congenital HL can be classified as genetic and non-genetic (environmental) in etiology. Genetic causes are often further subdivided into syndromic versus non-syndromic categories based on whether the patient suffers from an underlying genetic syndrome. Approximately 30% of the genetic causes of HL are syndromic. The non-syndromic category is responsible for greater than half of genetic causes and can be due to either an autosomal dominant, recessive, or sex-linked mutation **(Chari and Chan, 2017).** The most common cause of congenital HL is autosomal recessive, non-syndromic HL **(Morton, 2002).**

Environmental causes of HL could be maternal or after-birth complications. Maternal causes can be bacterial infection, viral infection, and bleeding. TORCH organisms (toxoplasmosis, rubella, cytomegalovirus [CMV], and herpes) have been identified as key infective causative agents. CMV is the most common cause of congenital non-genetic HL in the developed world. After birth complications may involve respiratory distress, hyperbilirubinemia, and perinatal asphyxia (**Katz, 2015).**

2. Acquired Hearing Loss

Otitis media with effusion is the number one cause of acquired HL in children. It is beyond the scope of this article to cover this in detail, but it classically has a bimodal peak at 2 and 5 years of age. It is characterized by a conductive HL associated with a flattened tympanogram. It typically resolves without intervention as the eustachian tube matures or following the insertion of a ventilation tube in the middle ear **(Rosenfeld et al., 2016)**. Adenoidal hypertrophy can contribute to this clinical picture. Infections also present another major category for acquired HL, with a particularly strong link with bacterial meningitis, mumps, and measles. Other reasons include primary otological pathologies such as cholesteatoma, impacted wax, and otosclerosis, as well as trauma **(Skoloudik et al., 2018)**.

3. **High-risk factors in neonates:**
    - Congenital infections
    - Family history
    - Craniofacial anomalies
    - Hyperbilirubinemia
    - Birth weight less than 1500 g
    - Low Apgar score
    - Bacterial meningitis
    - Prolonged intubation **(Chari and Chan, 2017)**.

Epidemiology

HL occurs in 1-3 newborns per 1000 births, with 1-2 per 1,000 suffering from permanent childhood hearing impairment. There is a slightly increased prevalence of HL in boys compared to girls, with a ratio of 1.16:1.0. Around 45,000 children with HL in the UK, half of which are congenital in origin **(Lieu et al., 2020)**.

Disabling HL refers to HL greater than 40 decibels (dB) in the better hearing ear in adults and HL greater than 30 dB in the better hearing ear in children (**WHO, 2015).** The **WHO (2016)** reported that about 60% of HL is due to preventable causes.

During the past three to four decades, the incidence of acquired SNHL in children living in more developed countries has fallen, because of improved neonatal care and the widespread implementation of immunization programs. The overall decrease has been

accompanied by a relative increase in the proportion of inherited forms of SNHL **(Shave et al., 2022)**.

SNHL is the most common sensory deficit in more developed societies. In Egypt, congenital SNHL occurs about three times more frequently than Down's syndrome, six times more frequently than spina bifida, and over 50 times more frequently than phenylketonuria (**Taha et al., 2010)**.

An estimated 4000 infants are born each year with bilateral severe to profound SNHL and another 8000 are born with unilateral or mild to moderate bilateral SNHL (**Sidenna et al., 2020).** Generally hearing losses affect educational achievement, the likelihood of future employment, future learning, the use of healthcare systems, and life expectancy (**Sheffield and Smith, 2019; Sidenna et al., 2020).**

Pathophysiology

Any condition that lowers the transmission of sound from the external space to the cochlea will cause conductive HL. This includes cerumen, abnormalities of the helix or auricle, effusions, and fixed ossicular chain. Besides cholesteatoma, other masses include glomus tumors, schwannomas of the facial nerve, and hemangiomas. SNHL is due to interruption of sound transmission after the cochlea. This may be due to damage to the hair cells or damage to the 8th cranial nerve. Even mild distortions in the hair cells can result in severe HL **(Wrobel et al., 2021)**. SNHL could be:

Cochlear HL* is often associated with damage to hair cells within the cochlea, this damage can lead to an increased hearing threshold in two ways:

1. Outer hair cells (OHCs) act as a biological amplifier /compressor and modify the signal. The basilar membrane (BM) of the cochlea is highly frequency-specific and tonotopically organized. Damage of OHCs impairs the active mechanism in the cochlea, resulting in reduced vibration of the BM for a given low sound level (**Sone et al., 2023)**.

2. Inner hair cells (IHCs) in the cochlea transduce the energy of the traveling wave to an electric action potential and synapse with auditory nerve fibers that form the auditory nerve. Damage of IHCs can result in less efficient transduction. As a result, even with a greater amount of BM vibration, speech recognition has been distorted to a degree that coincides with the extent of IHC damage (**Habib and Habib, 2021).**

Neural or retrocochlear HL results from damage to the fibers of the auditory nerve and the following auditory pathway. This damage may affect the initiation of the nerve impulses in the auditory nerve or the transmission of the nerve impulses along the auditory pathway. Retrocochlear auditory dysfunction is quite uncommon in both children and adult patient populations (**John et al., 2012).**

There are several pathophysiological mechanisms by which damage to the inner ear results in SNHL;

1. **Structural abnormality of cochlear components:** e.g., trauma or congenital conditions.
2. **Aberrant metabolic activity:** Cochlear function is determined by the transport of ions. Genetic or acquired conditions that interfere with this transport can lead to changes in the endolymph and affect hearing **(Lammers et al., 2019)**.

Impact of hearing loss on children

Proper hearing during early life is critical for language development to prevent life-long disability. HL in children represents a great challenge, especially for infants and toddlers who cannot express their difficulty. If HL in children is not managed early and adequately, it could have a significant impact on their development and daily life (**WHO 2023)**, including:

1. *Delayed language and speech development*: Children with HL may experience delays in learning to speak and understand language. They may have unclear speech and difficulty communicating effectively.
2. *Difficulties in social interactions*: HL can make it harder for children to communicate and engage with others, leading to social isolation and difficulties in making friends.
3. *Poor academic achievement*: Children with HL may have poor performance in school, particularly in reading and math. They may also have difficulty following instructions and understanding classroom discussions.
4. *Emotional and behavioral issues*: HL can impact a child's self-esteem and emotional well-being. They may exhibit behavioral problems due to frustration and difficulty in understanding and expressing themselves.

5. *Auditory processing disorder (APD):* Children with peripheral SNHL are considered to be at high risk of APD. Early identification of APD may have important implications for the management of children with HL (**Musiek and Chermak, 2015**).
6. *Limited access to auditory information*: A child with HL may miss out on important sounds, such as alarms, doorbells, and verbal instructions, which can affect their safety and independence.
7. *Reduced participation in activities*: HL can limit a child's ability to engage in age-appropriate activities, such as music, sports, and group conversations. They may feel left out or excluded from these experiences.

Early identification of HL in children is crucial for their language, cognitive, social, and emotional development. It allows children to receive appropriate interventions and amplifications that minimize the effects of HL. Detecting hearing disorders in children at an early age, ideally by three months, is important. Starting appropriate interventions by six months of age is also recommended **(Moeller and Tomblin, 2015)**.

Assessment

1. History Taking

A thorough history taking is required, including pre-, peri- and postnatal history **(WHO, 2023)**. Prenatal history involves asking for exposure to intrauterine infection, teratogenic drugs, and /or trauma. Perinatal history could include hyperbilirubinemia, low birth weight, birth asphyxia, and other perinatal morbidity. In addition, postnatal history may involve chronic ear infections (chronic suppurative otitis media), collection of fluid in the ear (chronic nonsuppurative otitis media), meningitis, and other infections.

2. Physical examination

The examination involves inspecting the ear, including the appearance of the pinna, particularly for any deformities such as microtia or anotia. Otoscopic examination of the external auditory canal and tympanic membrane is crucial, with special attention on the attic for cholesteatoma. A full neurological examination should also be performed with the assessment of cranial nerves and balance, based on the child's age (**Kılıç et al., 2021**).

Moreover, the general examination is important to evaluate the presence of other abnormalities, such as skin dyschromia, eye irregularities, and renal dysfunction, particularly when suspecting syndromic forms and in case of familiarity with HL (**Ciorba et al., 2017**).

3. Audiological evaluation

A. Universal neonatal hearing screening (UNHS):

The international organizations recommend the implementation of **UNHS**. All newborns should have access to hearing screening using a physiologic and objective measure. This includes:

*All newborns or infants who require neonatal intensive care.

*All newborns who receive routine care.

*Newborns in alternative birthing facilities, including home births, should have access to and are referred for screening before one month of age. With the implementation of the UNHS program, nowadays, most patients are identified within a few months after birth, with intervention starting at six months old (**Chorath et al., 2021**).

The Joint Committee on Infant Hearing (JCIH, 2007) indicators for screening are for newborn infants who have failed initial screening and failed any subsequent rescreening before comprehensive audiological evaluation. The JCIH has identified the neonatal audiological risk factors that UNHS programs must consider in their implementation.

Auditory brainstem response (ABR) and otoacoustic emissions (OAEs) are appropriate physiologic measures for screening the newborn population. Both are noninvasive and easily utilized by trained hospital staff. They are available in automated versions that determine pass/fail or pass/refer and do not require interpretation.

B. Basic audiological evaluation:

Behavioral observation audiometry is used in infants aged 0–6 months. However, because it is highly examiner-dependent, it has been supplemented by ABR, OAEs, and auditory steady-state response (ASSR) testing **(Chari and Chan, 2017)**.

Visual reinforcement audiometry is used in children aged 6 months to 2.5 years and can be used to generate a reliable, complete audiogram, although results depend on the child's maturational age and the skill of the examiner **(Belcher et al., 2021)**.

Conditional play audiometry is used in children with 2-5 years of age. The child is conditioned to perform a task in response to an auditory stimulus such as placing a ball in a cup. Once the task is learned the sound volume is reduced to determine their hearing threshold **(Chiaburu-Chiosa, 2020).**

Conventional audiometry: Most children aged 5 years or more can undergo pure-tone audiometry. Hearing thresholds are determined by presenting sounds of various frequencies and at various intensities until the quietest sound is reliably detected 50% of the time. This test requires a higher level of attention and therefore is rarely done below the age of 5 years **(Sone et al., 2023).**

C. Advanced audiological evaluation:

I- Electrophysiological tests:

1) OAEs can be used in the assessment of SNHL as a cochlear and retrocochlear disorder. If the cochlea is disordered, OAEs are expected to be abnormal or absent (**Sommerfeldt and Kolb, 2022).**

2) ABR can be used for the assessment of cochlear disorder and differentiate (together with OAEs) a cochlear from a retrocochlear HL. The ABR is a sensitive indicator of the integrity of the auditory nerve (**Ciorba et al., 2017).**

3) ***ASSR*** can be used to estimate hearing sensitivity. The advantages of ASSR testing are better frequency specificity than the ABR and patients do not need to remain awake (**Alamanda and Hohman, 2023).**

II- Central Auditory Processing (CAP) Assessment

The purposes of central auditory assessment involve: 1) identify the presence of abnormalities or dysfunction of the central auditory nervous system (CANS) and to diagnose central auditory processing disorder (CAPD); and 2) determine the nature and extent of the disorder to develop management and intervention programs for affected individuals (**Hamed et al., 2012).**

There is a variety of tests that assess CAPD:

***Questionnaire*s** are common tools for individuals who exhibit functional behavioral limitations in their communication, language, and learning. These questionnaires involve Fisher Auditory Problems Checklist (**Fisher, 1985**), Children's Auditory Processing Performance Scale **(Smoski et al., 1992**), Screening Instrument for Targeting Educational Risk (**Anderson, 1989**), and Buffalo Model Questionnaire—Revised **(Katz and Zalweski, 2013**).

Behavioral tests include auditory discrimination, temporal processing, dichotic listening, low-redundancy speech recognition (monaural), and binaural interaction testing. Baseline audiological assessments such as standard pure-tone audiometry, speech-in-quiet audiometry, otoacoustic emissions, and ***electrophysiological*** measures are also important when assessing for CAPD to control for the presence of peripheral auditory impairment and complement behavioral auditory processing tests **(Chowsilpa et al., 2021**)**.**

4. Further investigations

Additional investigations should be performed to obtain an accurate diagnosis:

A. Systemic investigations

Congenital cytomegalovirus (CMV) infection was confirmed by viral culture of urine or saliva specimens. Urine analysis contributed to the diagnosis of Alport syndrome. Ophthalmology evaluation is often recommended for neonates with newly diagnosed HL to assess for syndromes that may result in concurrent visual and hearing impairment, such as Usher syndrome, Moreover, it is important because all children with

HL will be more dependent on their vision for speech and language development (**Chari and Chan, 2017).**

B. Genetic testing

Comprehensive genetic testing is an important part of the diagnostic algorithm for congenital HL. Approximately 50% of congenital HL is reportedly genetic, with the vast majority of genetic cases being non-syndromic. Syndromic cases can be divided into autosomal dominant and autosomal recessive cases.

Genetic testing for connexin-26 is recommended by some authors as it is considered a marker for SNHL. New advances in high-throughput sequencing, such as next-generation sequencing and massively parallel sequencing, allow the sequencing of multiple genes simultaneously. Children with bilateral SNHL or auditory neuropathy spectrum disorder may benefit from a genetic workup **(Brewer and King, 2022).**

C. Radiological evaluation

Imaging consisted of computed tomography of the temporal bone and/or magnetic resonance imaging of the inner ear, cerebellopontine angle, and brain. The decision to obtain imaging and the choice of the imaging modality was individualized per patient and made by the multidisciplinary team **(van Beeck Calkoen et al., 2021).**

Management

Appropriate plans include improving audibility, follow-ups, and any modifications through daily and long-term living. There are many different types of communication options for children with HL and their families.

1. Technology to help with communication

With the rapid development of technology, hearing devices appear as a promising approach for the treatment of SNHL. Children can partially or even greatly benefit from hearing devices, including hearing aids, middle ear implants, and cochlear implants. Hearing aids were widely used to amplify the signal of sound for patients suffering from mild or moderate SNHL, whereas middle ear and cochlear implants are suitable for

children whose SNHL cannot be compensated by hearing aids with a severe or profound loss of hearing (**Ren et al., 2022).**

Hearing aids: They work by converting sound detected by a microphone into digital signals, which can then be amplified and re-converted into audible sounds that are transmitted to the ear. They can be classified based on whether these key parts are housed in an earpiece that sits behind the ear, inside the canal, or further inside the canal.

Bone conduction hearing aids are typically used in conductive HL. Bone-anchored hearing aids (BAHA) are fitted surgically under general anesthetic over two stages. A titanium implant is fixed into the temporal bone. Through this setup, a sound is conducted directly to the inner ear by way of the bone, bypassing the middle ear. Typically, the BAHA is fitted from four years of age once the temporal bone has developed **(Chen and Oglhalali, 2016).**

Contralateral routing of sound (CROS): hearing aids are used when there is unilateral SNHL. The sound in the problem ear is diverted to the better-hearing ear without amplification. In cases where neither ear has normal hearing but one side is significantly better, a variation on this can be used called a BiCROS **(Chen and Oglhalali, 2016).**

A cochlear implant is a device that is surgically implanted into the cochlea to deliver electrical stimulation to the auditory nerve. It provides speech perception to children with severe to profound SNHL for whom hearing aids do not benefit. Usually, the sound is converted from mechanical to electrical energy via the hair cells in the cochlea **(Luu et al., 2023)**. Cochlear implants bypass the damaged hair cells and provide direct stimulation to auditory neurons (**Xu et al., 2019).**

Auditory brainstem implant (ABI) is a surgically implanted central neural auditory neuroprosthesis that provides a safe and effective auditory rehabilitation for patients with profound SNHL who are not candidates for a cochlear implant due to abnormalities of the cochlea and the cochlear nerve. The Food and Drug Administration approved the use of ABIs in patients diagnosed with neurofibromatosis type 2 (NF2) **(Kanowitz et al., 2004)**. Recently, it has been reported that the use of ABI is not restricted to only patients with NF2 but also expanded to non-NF2 patients. The ABIs differ from cochlear implants in that the electrical impulse–derived sounds can directly

stimulate the nucleus through a soft silicone paddle that is placed along the surface of the brainstem **(Ren et al., 2022**).

2. Speech and language therapy

This can help children with HL develop clearer speech and improve their language skills **(Wood et al., 2021).**

3. Auditory training

It is used to improve central auditory abilities such as auditory figure-ground to improve hearing in noisy situations **(Hassaan and Ibraheem, 2016).**

4. Family support services

The family plays a vital role in the evaluation and habilitation process in children with HL and is regarded as an integral part of the team. For many parents, their child's HL is unexpected. Parents sometimes need time and support to adapt to the child's HL. Support is anything that helps a family and may include: advice, information, getting the chance to get to know other parents who have a child with HL, locating a deaf mentor, finding child care or transportation, and giving parents time for personal relaxation **(Erbasi et al., 2018).**

5. Educational support*:*

Children with HL may benefit from accommodations in the classroom, such as preferential seating, captioned materials, and frequency modulation systems that amplify the sound and decrease the ground noise **(Wood et al., 2021**).

Future perspectives in hearing loss assessment and management

The assessment and management of HL in children require a coordinated and multidisciplinary approach. Healthcare providers should work together as part of a team to ensure a prompt diagnosis and appropriate interventions. Communication between specialists is crucial to ensure that each child receives the right care and does not become lost to follow-up. Without a team approach, efforts to improve the developmental potential of children with HL may fail **(Sommerfeldt and Kolb, 2022)**.

Conclusion

HL is a prevalent health-related issue among children worldwide. Many possible factors can cause HL in children, which can affect their speech, language development, communication, and learning. Parents and caretakers of children with HL are also affected, as they often experience increased stress and costs associated with seeking treatment for their child's well-being.

To diagnose HL in children, an audiologist will take a detailed medical history and order appropriate tests to determine the cause of the hearing loss, including objective and electrophysiological measures. Management of HL in children requires a multidisciplinary team-based approach that includes counseling for parents and caregivers.

References

Alamanda M., and Hohman M.H. (2023): Auditory Steady-State Response. [Updated 2023 Oct 28]. In: *StatPearls* [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK597346/

Anderson K. (1989). SIFTER: Screening Instrument for Targeting Educational Risk in Children Identified by Hearing Screening or Who Have Known Hearing Loss. Available from: Educational Audiology Association-800/460-7322-EAA@L-TGraye.com

Belcher, R., Virgin, F., Duis, J., and Wootten, C. (2021): Genetic and non-genetic workup for pediatric congenital hearing loss. *Frontiers in Pediatrics,* 9, 536-44.

Brewer, C. C., and King, K. A. (2022): Genetic hearing loss: the audiologist's perspective. *Human genetics*, *141*(3-4), 311-314.

Ciorba, A., Corazzi, V., Negossi, L., Tazzari, R., Bianchini, C., and Aimoni, C. (2017): Moderate-Severe Hearing Loss in Children: A Diagnostic and Rehabilitative Challenge. *The Journal of International Advanced Otology*, 13(3): 407-413.

Chari, D. A., and Chan, D. K. (2017): Diagnosis and treatment of congenital sensorineural hearing loss. *Current otorhinolaryngology reports*, *5*, 251-258.

Chen, M. M., and Oghalai, J. S. (2016): Diagnosis and management of congenital sensorineural hearing loss. *Current treatment options in pediatrics*, *2*, 256-265.

Chorath, K., Garza, L., Tarriela, A., Luu, N., Rajasekaran, K., and Moreira, A. (2021): Clinical practice guidelines on newborn hearing screening: a systematic quality appraisal using the AGREE II instrument. *International Journal of Pediatric Otorhinolaryngology*, 141, 110504.

Chowsilpa, S., Bamiou, D. E., and Koohi, N. (2021): Effectiveness of the auditory temporal ordering and resolution tests to detect central auditory processing disorder in adults with evidence of brain pathology: a systematic review and meta-analysis. *Frontiers in Neurology*, *12*, 656117.

Dimitrov, L., and Gossman, W. (2023): Pediatric hearing loss. In: *StatPearls [Internet]*. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 30855869.

Chiaburu-Chiosa, D. (2020): Optimization of early diagnosis and auditory rehabilitation in children with sensorineural hearing loss. Ph.D. Thesis in Medical Sciences. Available at: http://repository.usmf.md/handle/20.500.12710/7376.

Erbasi, E., Scarinci, N., Hickson, L., and Ching, T. Y. (2018): Parental involvement in the care and intervention of children with hearing loss. *International Journal of Audiology*, *57*(sup2), S15-S26.

Fisher, L. I. (1985). Learning disabilities and auditory processing. *Administration of speech-language services in the schools*, *7*, 231-292.

Habib, S. H., and Habib, S. S. (2021): Auditory brainstem response: An overview of neurophysiological implications and clinical applications-A Narrative Review. *The Journal of the Pakistan Medical Association*, 71(9), 2230-2236.

Hamed, S. A., Youssef, A. H., and Elattar, A. M. (2012): Assessment of cochlear and auditory pathways in patients with migraine. *American journal of Otolaryngology*, *33*(4), 385-394.

Hassaan, M. R., and Ibraheem, O. A. (2016): Auditory training program for Arabic-speaking children with auditory figure-ground deficits. *International Journal of Pediatric Otorhinolaryngology*; 83, 160-167.

John, A. B., Hall, J. W. 3rd, and Kreisman, B. M. (2012): Effects of advancing age and hearing loss on gaps-in-noise test performance. *American Journal of Audiology*, 21(2):242-50.

Kanowitz, S. J., Shapiro, W. H., Golfinos, J. G., Cohen, N. L., and Roland Jr, J. T. (2004). Auditory brainstem implantation in patients with neurofibromatosis type 2. *The Laryngoscope*, *114*(12), 2135-2146.

Katz, J. (2015): Integration Signs-New Addition-Further Support. *SSW Reports*; 37(2): 1-7.

Katz, J., and Zalweski, T. (2013): Buffalo Model Questionnaire – Revised (BMQ-R). Denver, CO: Educational Audiology Association. *Educational audiology handbook*. Plural Publishing.

Kılıç, S., Bouzaher, M. H., Cohen, M. S., Lieu, J. E., Kenna, M., and Anne, S. (2021): Comprehensive medical evaluation of pediatric bilateral sensorineural hearing loss. *Laryngoscope investigative otolaryngology*, *6*(5), 1196-1207.

Lammers, M. J., Young, E., Fenton, D., Lea, J., and Westerberg, B. D. (2019): The prognostic value and pathophysiologic significance of three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) magnetic resonance imaging in idiopathic sudden sensorineural hearing loss: A systematic review and meta-analysis. *Clinical Otolaryngology*, 44(6), 1017-1025.

Lieu, J. E., Kenna, M., Anne, S., and Davidson, L. (2020). Hearing loss in children: a review. *JAMA*, 324(21), 2195-2205.

Luu, K., Shaffer, A. D., and Chi, D. H. (2023): Practice trends in pediatric sudden sensorineural hearing loss management: An unresolved diagnosis. *American Journal of Otolaryngology*, 44(4)**,** 103845.

Moeller, M. P., and Tomblin, J. B. (2015): An introduction to the outcomes of children with hearing loss study. *Ear and Hearing*, 36 Suppl 1(0 1):4S-13S.

Morton, C. C. (2002). Genetics, genomics and gene discovery in the auditory system. *Human molecular genetics***, 11(10), 1229-1240.**

Musiek, F. E., and Chermak, G. D. (2015): Psychophysical and behavioral peripheral and central auditory tests. In: Michael, J., A, François, B., Dick, F., S. (eds.), *Handbook of clinical neurology*, vol. 129, chapter 18. Elsevier, pp: 313-332.

Ren, H., Hu, B., and Jiang, G. (2022): Advancements in prevention and intervention of sensorineural hearing loss. *Therapeutic Advances in Chronic Disease*, 13, 20406223221104987.

Rosenfeld, R. M., Shin, J. J., Schwartz, S. R., Coggins, R., Gagnon, L., Hackell, J. M., Hoelting D., Hunter LL., Kummer AW., Payne SC., Poe DS., Veling M., Vila PM., Walsh SA., and Corrigan, M. D. (2016): Clinical practice guideline: otitis media with effusion executive summary (update). *Otolaryngology–Head and Neck Surgery*, 154(2), 201-214.

Shave, S., Botti, C., and Kwong, K. (2022): Congenital sensorineural hearing loss. *Pediatric Clinics of North America*, 69(2)**,** 221-234.

Sheffield, A. M., and Smith, R. J. (2019): The epidemiology of deafness. *Cold Spring Harbor Perspectives in Medicine*, 3;9(9): a033258.

Sidenna, M., Fadl, T., and Zayed, H. (2020): Genetic epidemiology of hearing loss in the 22 Arab countries: a systematic review. *Otology and Neurotology*, *41*(2), e152-e162.

Skoloudik, L., Kalfert, D., Valenta, T., and Chrobok, V. (2018): Relation between adenoid size and otitis media with effusion. *European annals of otorhinolaryngology, head and neck diseases*, 135(6), 399-402.

Smoski, W. J., Brunt, M. A., and Tannahill, J. C. (1992): Listening characteristics of children with central auditory processing disorders. *Language, Speech, and Hearing Services in Schools*, *23*(2), 145-152.

Sommerfeldt, J., and Kolb, C. M. (2022): Hearing Loss Assessment in Children. In *StatPearls [Internet]*. StatPearls Publishing.

Sone, M., Kobayashi, M., Yoshida, T., and Naganawa, S. (2023): Pathophysiological analysis of idiopathic sudden sensorineural hearing loss by magnetic

resonance imaging: A mini scoping review. *Front Neurol,* 14**,** 12-20.

Taha, AA., Pratt, SR., Farahat, TM., Abdel-Rasoul, GM., Albtanony, MA., Elrashiedy, AL., Alwakeel, HR., and Zein, A. (2010): Prevalence and risk factors of hearing impairment among primary-school children in Shebin El-kom District, Egypt. *American Journal of Audiology*. Jun;19(1):46-60.

The Joint Committee on Infant Hearing. (2007): Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs. *Pediatrics, 120*(4), 898-921.

van Beeck Calkoen, E. A., Pennings, R. J. E., Smits, J., Pegge, S., Rotteveel, L. J. C., Merkus, P., Verbist, B. M., Sanchez, E., and Hensen, E. F. (2021): Contralateral hearing loss in children with a unilateral enlarged vestibular aqueduct. *International Journal of Pediatric Otorhinolaryngology, 150*, 110891.

Wood, J. W., Shaffer, A. D., Kitsko, D. and Chi, D. H. (2021). Sudden Sensorineural Hearing Loss in Children—Management and Outcomes: A Meta-analysis. *Laryngoscope,* 131**,** 425-34.

World Health Organization (2015): Prevention of blindness and deafness: estimates. (http://www.who.int/pbd/deafness/estimates/en/; accessed 11 December 2015).

World Health Organization (2016): Childhood hearing loss: strategies for prevention and care. (http://www.who.int/iris/ handle/ 10665/ 204632).

World Health Organization (2023): Deafness and hearing loss: https://www.who.int/news-room/fact-sheets/detail/deafness-and-hearing-loss

Wrobel, C., Zafeiriou, M. P., and Moser, T. (2021). Understanding and treating paediatric hearing impairment. *EBioMedicine*, 63:103171.

Xu, M., Jiang, Q., and Tang, H. (2019): Sudden sensorineural hearing loss during pregnancy: clinical characteristics, management and outcome. *Acta Otolaryngology,* 139 (1)**,** 38-41.