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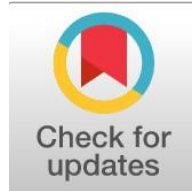
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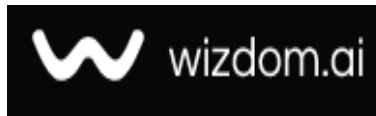
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Hearing Loss in Children: Prevalence, Causes, and Management

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Abstract

General Background Hearing loss is a common neonatal abnormality where early detection is crucial for normal speech and language development. **Specific Background** Children referred to audiology clinics often present with diverse impairments that can be objectively evaluated using auditory brainstem response testing. **Knowledge Gap** The specific incidence, severity distribution, and anatomical location of auditory deficits within this referred pediatric population require continuous clinical documentation. **Aims** This study aims to determine the incidence, severity, and location of auditory impairment in youth referred for assessment. **Results** Among 309 patients aged 1 to 17 years undergoing testing, 91.3% exhibited impairment, whereas 8.7% demonstrated normal bilateral function. Within the impaired cohort, 56% were male and 44% were female. Profound and severe degrees of impairment occurred most frequently, with both ears being predominantly affected. **Novelty** The documented burden of profound bilateral deficits in this specific clinical cohort highlights critical sex-based and severity distribution trends. **Implications** These findings strongly support the urgent need for universal newborn screening and early targeted rehabilitation, including cochlear implantation, to prevent irreversible language delays.

Highlights

- A vast majority of referred patients exhibited significant auditory impairment.
- Profound bilateral deficits frequently presented during clinical assessments.
- Prompt cochlear implantation prevents permanent language development delays

Keywords: Auditory Brainstem Response, Cochlear Implantation, Universal Newborn Screening, Language Development, Pediatric Audiology

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Introduction

Hearing loss is one of the most common abnormalities occurring in newborns and children, with prevalence rates of approximately 1-3 per 1000 healthy infants and 4-5% among high-risk neonates. Approximately 1 in 1000 children are born with permanent hearing loss, while 50-90% acquire hearing loss in early childhood. Early detection and intervention are crucial for normal speech and language development.

Hereditary conditions play a large role in childhood hearing loss. Autosomal recessive mode of inheritance results in 70% of all genetic cases of hearing loss and Autosomal Dominant cases resulting in 15% of all genetic hearing loss cases. Pediatricians and audiologists alike need to be familiar with the cause, categorization and management plan of childhood hearing loss.

Types and Severity of Hearing Loss

Hearing loss in children presents with diverse characteristics based on type, severity, and pattern of involvement. The major types include conductive hearing loss, sensorineural hearing loss (SNHL), and mixed hearing loss. Severity ranges from mild to profound, with functional impact varying based on bilateral versus unilateral involvement and symmetric versus asymmetric patterns.¹

Classification by Pattern

Hearing loss can be classified as congenital or acquired, genetic or non-genetic, syndromic or non-syndromic, and bilateral or unilateral. Congenital or early-acquired bilateral moderate to profound hearing loss significantly affects both receptive and expressive speech abilities in children. Unilateral hearing loss is typically discovered later than bilateral hearing loss but can still impact language development, particularly when mild degrees are involved. Asymmetric hearing loss is defined as a difference in loss greater than 15 dB HL between ears at 0.5, 1, and 2 kHz, or greater than 20 dB HL at 3, 4, and 6 kHz on audiometry. Even mild unilateral hearing loss may affect language development in children. ²

Genetic Inheritance Patterns

Hearing loss with a genetic origin can be inherited in a recessive, dominant or sex-linked way or occur due to mutations in cell or mitochondrial DNA/RNA. Environment and ageing can influence the expression of genetic hearing loss, i.e. Noise induced hearing loss (NIHL) and age related hearing loss. ³

Causes of Hearing Loss

The majority of permanent hearing loss present at birth is sensorineural in nature, with genetic factors accounting for approximately half of all childhood sensorineural hearing loss. Acquired causes include various perinatal, infectious, and environmental factors. ⁴

Acquired Causes

Acquired causes of sensorineural hearing loss include hypoxia and respiratory problems, diaphragmatic hernia, hyperbilirubinemia, ototoxic medications, low Apgar scores, meningitis, trauma, loud noise exposure, otitis media, and viral infections. These factors can cause damage to the inner ear and auditory pathways. ⁵

Cytomegalovirus (CMV) Infection

Cytomegalovirus (CMV) infection is an important cause of unilateral or bilateral sensorineural hearing loss in children, accounting for approximately 9% of childhood SNHL. Notably, 90% of infected infants show no signs of congenital infection at birth (asymptomatic presentation). Progressive and late-onset SNHL are common presentations in CMV-infected children. Early diagnosis within the first two weeks of life is achieved through virus isolation from urine or saliva. Later-onset SNHL in CMV-infected children is diagnosed by detection of CMV DNA in infant blood or umbilical cord blood using polymerase chain reaction (PCR). ⁶

Otitis Media

The development of conductive hearing loss in children with otitis media is common, secondary to the anatomical factors in the paediatric Eustachian tube which is shorter and more horizontal than in the adult and therefore easily occluded by adenoid hypertrophy and infectious processes. Impedance in the middle ear space occurs, vibration in the ossicular chain is limited, leading to loss of acoustic energy and hearing loss which is typically mild to moderate with sounds being 'muffled or distorted'. Children may present with damage to the tympanic membrane, ossicles, or even auditory nerve in cases of recurrent otitis media, this will lead to permanent hearing loss. In otitis media with effusion there may be a lack of acute infection which causes hearing loss that is prolonged and painless and that affects speech and language development during the critical early childhood period.⁷

Investigation and Screening for Hearing Loss

Screening for hearing loss in all newborns, regardless of risk factors, is essential for early detection and intervention. Universal newborn hearing screening (UNHS) is a critical public health initiative. 8

Screening Technologies

Otoacoustic emissions (OAE) and auditory brainstem response (ABR) are the two most commonly used protocols in newborn hearing screening programs. OAE assesses the integrity of outer hair cells of the cochlea, while ABR evaluates the integrity of the auditory pathway from the cochlea to the brainstem. The presence of OAE with negative ABR results indicates auditory neuropathy spectrum disorder, highlighting the diagnostic complexity of childhood hearing loss. Hospitals routinely perform hearing screening tests on infants within the first 24-48 hours after birth. 9

Management of Hearing Loss

Treatment of hearing loss depends on the underlying cause and may involve medical or surgical intervention. Management strategies range from conservative approaches with hearing aids to surgical implantation of auditory devices. 10

Hearing Aids and Speech Therapy

Mild to moderate sensorineural hearing loss is typically managed with hearing aids combined with speech therapy. Hearing aids and cochlear implants for children with hearing loss must make speech audible comfortably and provide adequate acoustic cues without over-amplification of loud sounds. Audibility of soft speech is particularly important for incidental speech and language learning. Rehabilitation is crucial for speech and language development at age-appropriate levels. 11

Cochlear Implantation

Bilateral profound sensorineural hearing loss can be treated with cochlear implantation (CI). Cochlear implants are indicated for severe to profound hearing loss in selected patients to restore hearing, particularly during the critical period of rapid language acquisition in childhood. CI can restore auditory function to children with previous hearing who have lost it, or provide hearing for children with congenital hearing loss. A cochlear implant consists of external components (processor and headset) and internally implanted receiver-stimulator units. The headset contains a microphone and transmitter that picks up sound and converts it into electrical signals via a speech processor. The signal is transmitted transcutaneously to the receiver-stimulator via a transmitter coil, which electrically stimulates the spiral ganglion cells of the cochlear nerve, resulting in auditory perception.

Cochlear implants can make use of the existing residual hearing, and with rehabilitation, can lead to near-age level of speech and language development. Cochlear stimulation causes the activation of central auditory pathways which leads to the sensation of hearing and development of speech perception abilities. 12

Bilateral Hearing Benefits

Bilateral hearing provides superior benefits compared to monaural hearing. Bilateral hearing improves speech perception, sound localization, loudness perception through binaural summation, and listening performance in noisy environments. The use of bilateral hearing aids or bilateral cochlear implants in children with bilateral or unilateral hearing loss plays an important role in speech perception and attention development. 13

Materials and Methods

The present investigation was performed on 309 children referred to the audiology centre in our institution, as part of routine hearing screening/investigation for speech delay, parental concern about the hearing, or if there was a family history of congenital hearing loss. 14

Study Population

A total of 309 children aged 1-17 years were included in the study. They were referred either due to delayed speech development, parental suspicion of hearing loss, or positive family history of congenital hearing loss. 15

Auditory Brainstem Response Testing

ABR testing was performed using a CORONA instrument (manufactured in Germany) with rarefaction polarity signal, which is the most commonly used polarity in ABR testing. Test parameters included a window opening of 10 ms with low and high-pass filters at 100 Hz and 2500 Hz, respectively. Testing was conducted in a quiet environment, with each test requiring 30-60 minutes depending on the child's cooperation and degree of hearing loss. Normal hearing was defined as presence of wave V for acoustic stimuli at 25 dB HL or lower. Hearing loss was defined as presence and persistence of wave V for acoustic stimuli at 30 dB HL or greater. The presence and persistence of wave V at specific intensity levels formed the basis for interpretation of ABR results. 16

Hearing Loss Classification

Degree of hearing loss was categorized as: Mild (30-40 dB HL), moderate (50 dB HL), moderately severe (60-70 dB HL), severe (80-90 dB HL) and profound (100 dB HL the maximum the device could produce). Type of hearing loss was determined as sensorineural, conductive or mixed based on the overall findings from tympanometry and ABR testing. 17

Ethical Issues: The study was performed in accordance with the Declaration of Helsinki. The authors should have institutional approval confirmed prior to submission.

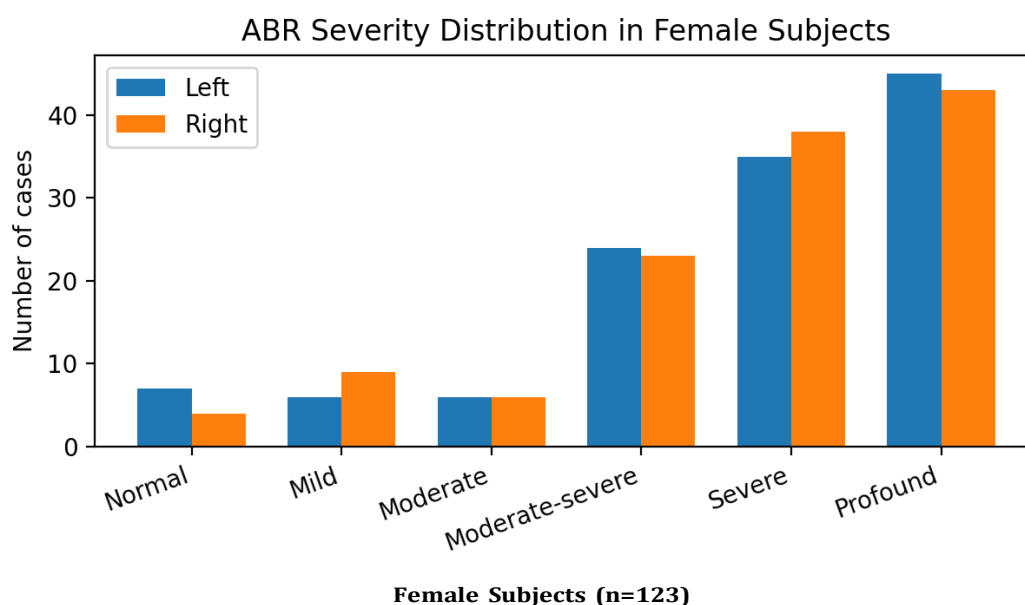
Written informed consent from the parents or legal guardians of the children should be provided prior to audiological evaluation.

Clinical Trial Registration: Not applicable as this was not a randomized clinical trial.

Results

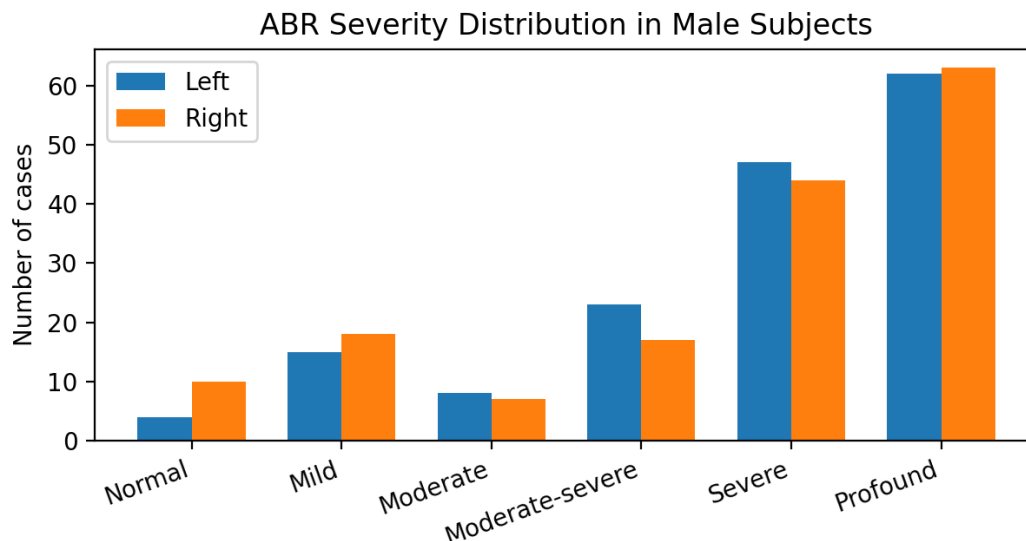
A total of 309 children underwent ABR testing. Of these, 27 children (8.7%) had bilateral normal hearing (16 males, 11 females), while 282 children (91.3%) had either unilateral normal hearing with contralateral hearing loss or bilateral hearing loss (159 males, 123 females). The overall male-to-female ratio among children with hearing loss was 56% males and 44% females.

Figure 1. ABR severity distribution by ear among female subjects.



Among the 123 female patients with hearing loss, the ABR severity distribution is summarised in Figure 1:

Figure 2. ABR severity distribution by ear among male subjects.



Male Subjects (n=159)

Among the 159 male patients with hearing loss, the ABR severity distribution is summarised in Figure 2:

Severity Distribution Tables

Tables 1-5 summarise the distribution of hearing loss severity by sex and ear.

Table 1: Sex Distribution of Mild Hearing Loss.

Severity Level	Male	Female
Mild HL	33	15

Table 2: Sex Distribution of Moderate Hearing Loss.

Ear	Male	Female
Right	7	6
Left	8	6

Table 3: Sex Distribution of Moderate-Severe Hearing Loss.

Ear	Male	Female
Right	17	23
Left	23	24

Table 4: Sex Distribution of Severe Hearing Loss.

Ear	Male	Female
Right	44	38
Left	47	35

Table 5: Sex Distribution of Profound Hearing Loss.

Ear	Male	Female
Right	63	43
Left	62	45

Unilateral Normal Hearing

Among the 309 children studied:

- Males with unilateral normal hearing: 10 cases with right ear normal, 4 cases with left ear normal
- Females with unilateral normal hearing: 4 cases with right ear normal, 7 cases with left ear normal

- Children with bilateral normal hearing: 16 males and 11 females

Discussion

Our study focus was the use of childhood hearing loss as a primary public health issue in all newborns, both the full term as well as the newborns of high risk. Our findings emphasize both prevalence of and difficulty associated with childhood hearing loss.

Prevalence and Genetic Factors

Our results are similar to those observed for hearing loss in general. Hearing loss may occur even in newborns with no apparent risk factors, and there is a genetic cause of hearing loss in 50% of cases, of which 80% are caused by autosomal recessive disorders and 20% by autosomal dominant disorders. The elevated rate of hearing loss (91.3%) in our cohort reflects referral biases; children referred to our clinic suspected of hearing loss undergo testing.

Clinical Presentation

The nature and degree of hearing loss ranges from conductive to mixed to sensorineural from mild to profound and hence the range of clinical presentations varies from uni or bilateral and can be symmetric or asymmetric. The finding that even mild unilateral hearing loss was associated with a language deficit may emphasize the importance of adequate screening and early diagnosis. The severe and profound end of the severity spectrum was most common in our series which might be due to the referral of children with a clinically evident hearing loss.

Aetiology: Genetic and Acquired Causes

Causes Genetic etiologies can be broadly classified as congenital and acquired, which include infectious agents (such as CMV), trauma, and infections such as Otitis Media. Congenital Cytomegalovirus (CMV) has become the leading known etiology for acquired hearing loss in infancy, and subclinical and unnoticed conditions can become evident as they appear, worsen, and progress during time, necessitating a strong screening process and monitoring. Otitis Media (middle ear infection) are common and potentially preventable causes of acquired hearing loss in childhood, owing to the ear anatomy that favors infection. Persistent OME and inadequate treatment can result in irreversible hearing loss, underscoring the importance of effective preventive strategies and treatments during early development.

Screening and Diagnosis

Screening and early diagnosis form the cornerstone of hearing loss management in children. This study employed ABR testing, one of the most objective and reliable methods for detecting sensorineural and retrocochlear pathology. OAE and ABR testing protocols represent the current standard for newborn hearing screening, with their complementary abilities to differentiate cochlear from retrocochlear pathology. The identification of auditory neuropathy through discordant OAE-ABR results highlights diagnostic complexity. Universal newborn hearing screening, ideally within 48 hours of birth regardless of risk status, is the recommended approach to ensure early detection. For CMV-related hearing loss, PCR testing is recommended for confirmation.

Management and Intervention

Different therapeutic approaches are suited to different degrees and types of hearing loss. Mild to moderately impaired hearing loss can usually be well treated using hearing aids coupled with speech therapy. Bilateral sensorineural hearing loss that is severe enough to affect speech development warrants cochlear implantation. A cochlear implant works by electronically stimulating the auditory nerve, effectively bypassing the impaired cochlear mechanisms, in order to facilitate the development of hearing and speech. The reliance on bilateral rehabilitative methods (either hearing aids or cochlear implants) can be seen as reflecting the evidence for the benefit of bilateral auditory stimulation for improved speech discrimination, localization, and understanding speech in noise. This is a critical factor during early childhood.

Study Findings and Sex Distribution

Our study indicated high prevalence of hearing loss among the population surveyed. The severity of the hearing loss were severe and profound with percentages of 38.5% and 42.1% of the population surveyed respectively. Among the affected, 75.6% of them had hearing loss in both ears while 24.4% of the population surveyed had unilateral hearing loss. With respect to the sex distribution, 56% were men and 44% were women with hearing loss. There was a significant difference in the distribution of severity among sexes and a trend towards more women having moderate-severe hearing loss and more men with profound hearing loss, the explanation of which requires further study and investigation in factors related to screening, etiology or referred patients

Clinical Implications and Future Directions

These results reinforce the value of universal newborn and routine childhood hearing screening and underscore that, when this testing leads to early intervention and optimal rehabilitation, good speech and language outcomes can be achieved.

More research is necessary into the specific genetic factors associated with nonsyndromic hearing loss and into environmental modifiers or strategies for effective rehabilitative treatment. These may help lead to wider availability of screening programmes to populations at risk, and genetic counselling to all patients and families with hearing loss.

Conclusion

In our audiology centre, our data shows there is a high burden of hearing loss among our children. 91.3% of the children referred to our audiology clinic during the period of study had identifiable hearing loss. Severe and profound bilateral hearing loss is prevalent and there is a requirement for regular screening and early management. Genetic causes (autosomal recessive) as well as acquired causes of the disease such as CMV infection and otitis media cause a significant burden of disease. Modern management such as cochlear implantation has revolutionized management of children with profound hearing loss. Newborn hearing screening, thorough diagnostic assessment and prompt rehabilitation of hearing loss are keys to effective management of hearing loss in children. Medical professionals should remain aware of potential hearing loss across the whole spectrum of childhood and ensure that all children have access to effective evidence-based diagnostics and therapeutics in order to promote the optimum development of speech and language.

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Conflicts of Interest Disclosure

The authors declare no actual or potential conflicts of interest related to this manuscript.

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