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An Insight into the Causes of Hearing Loss in Neonates and Children

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Abstract

World health organization estimates around 32 million children suffer from hearing loss. Hearing loss significantly affects the speech and language development of children. Onset of causes for hearing loss can occur as early as at embryonic development. Understanding hearing loss and their causes facilitates early intervention.The article provides an intuition into the reversible and irreversible causes for hearing loss at different stages through childhood.

Keywords: Conductive Hearing Loss; Sensori-Neural Hearing Loss; Syndromic; Non-Syndromic; Otitis Media; Pre-Natal; Peri-Natal; Post -Natal.

Introduction

Hearing loss is a partial or total inability to hear sounds. Hearing loss profoundly affects development, comprehension, production and maintenance of language, specifically speech. The effects are proportional to the degree and the type of hearing loss. Degree of hearing loss varies from minimal to profound. Depending on the cause and site of lesion, hearing loss can be of conductive (outer and/or middle ear), sensorineural (inner ear/ auditory nerve), and mixed (outer/ middle and inner ear) type. An estimated 32 million children suffer from hearing loss in the world. The prevalence of disabling hearing loss in children is greatest in South Asia, Asia Pacific and Sub-Saharan Africa [21]. As the incidence and prevalence of hearing loss is high in pediatric population, it is essential for the health professionals to be aware of its causes. This article would provide a fair insight into the causes of hearing loss in neonates and children.

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Causes Based on the Anatomical Ear Structure Involved

Outer Ear

Approximately 5% of the hearing impaired population has some sort of ear malformation. Congenital anomalies are the product of errors in embryogenesis (malformations) or the result of intrauterine events that affect embryonic and fetal growth (deformations and disruptions).The formation of the ear involves fusion of ectoderm, endoderm, and mesoderm. Defects in formation may lead to a wide variety of dysfunctional or malformed structures. Some of the abnormalities are microtia (abnormally small pinna), anotia (completely absence of auricles), atresia (absence of an external auditory meatus) and aural stenosis (abnormal narrowing of canal opening) [2].

Middle Ear

The external auditory canal, middle ear, and bulk

of the ossicular chain develop from the first branchial groove, first and second branchial arches, and first pharyngeal pouch. Embryologic development of these structures is complex. Rarely there are two anomalies identical. Common structural deformities of the middle ear are gross malformation or absence of middle ear cavity and antrum, ossicles may be abnormally formed (malleus and incus are often conglomerated into a unit), absent middle ear bones, and/or not attached to abnormally formed middle ear cavity either directly or via bony ridges. Tympanic membrane may be rudimentary or absent [2].

Inner Ear

Development of the inner ear structures occurs independently of external ear structures, and concomitant involvement is unusual. Some of the common inner ear deformities are Michel's aplasia (total absence of inner ear structures; occurs even with a normal conductive mechanism), Mondini's dysplasia (incomplete anomalies and developmental failure of bony and membranous ducts of inner ear), Scheibe's aplasia (dysplasia of membranous ducts of cochlea and saccule), Alexander's aplasia (congenital abnormalities of cochlear duct, especially affecting basal turn of cochlea) etc. [15].

Genetic

Hereditary deafness is genetically a highly heterogeneous disease with many different genes responsible for auditory dysfunction. It is estimated that 50% to 75% of all childhood deafness is due to hereditary causes. There are two main forms of genetic hearing loss, syndromic and non-syndromic. Children with syndromic hearing loss have other clinical features in addition to the hearing loss. About 15-30% of the hereditary hearing loss is syndromic, while the vast majority is non-syndromic (70%). Genetic heterogeneity has been observed repeatedly in improved understanding of syndromic hearing loss. Mutations of several different genes can cause the identical clinical phenotype. On the other hand, different mutations in one gene can cause variable phenotypes [5,14].

Syndromic Hearing Loss

There are over four hundred described syndromes with hearing loss [18]. The Online Mendelian Inheritance in Man has comprehensive descriptions of the clinical features and molecular genetics of these syndromes as well as an all-inclusive list of references. Some of the syndromes which are commonly known to be associated with hearing loss are Alport syndrome, Branchial-oto-renal syndrome, Jervell and Lange-Nielsen syndrome, Pendred syndrome, Stickler syndrome, Treacher Collins syndrome, Usher syndrome, Waardenburg Syndrome, Hurler Syndrome & Hunter Syndrome and Down's syndrome [7].

Non-Syndromic Hearing Loss

There are autosomal dominant, autosomal recessive and X-linked forms of non-syndromic hearing loss. In general, recessive inheritance shows pre-lingual onset of hearing loss. And the severity is severe to profound with all frequencies affected. In autosomal dominant forms, the phenotype is less severe and the onset is usually post-lingual. The factors associated with non-syndromic hearing loss are genes for homeostasis, transcription factors, cytoskeleton proteins, extracellular matrix components, mitochondrial hearing loss to name few [19, 20].

Based on Birth History

Prenatal Causes

TORCH syndrome, fetal alcohol spectrum disorders, malnutrition, physical trauma and exposure to radiations in the mother are some of the prenatal causes [13].

Perinatal Causes

The perinatal causes of hearing loss include asphyxia, hypoxia, ischemia, physical injury (forceps delivery, etc), contracted infections during birth, prematurity and/or low birth weight, severe hyperbilirubinaemia, sepsis etc [4].

Postnatal Causes

Childhood infections (meningitis, encephalitis, jaundice, etc.), traumatic brain injury, lead poisoning, allergic reactions to medications, exposure to toxins or other environmental conditions [10] are some of the post natal causes.

Based on Type of Hearing Loss

Conductive Hearing Loss

Conductive hearing loss occurs when the conducting sound waves are attenuated anywhere along the route through the outer ear, tympanic

membrane (eardrum), or middle ear (ossicles). The conductive hearing loss is most often treated medically or surgically. Prolonged conductive hearing loss or in certain conditions sensorineural hearing loss co-exists with conductive hearing loss and is known as mixed hearing loss. Causes of conductive hearing loss include malformation of outer ear/ear canal/middle ear structures, fluid accumulation in the middle ear, impacted wax etc. [12].

Sensori-neural Hearing Loss

Sensorineural hearing loss (SNHL) occurs when there is damage to the inner ear (cochlea), or to the nerve pathways from the inner ear to the brain. Most of the time, SNHL cannot be medically or surgically corrected. This is the most common type of permanent hearing loss. Some of the important causes for sensorineural hearing loss are exposure to loud noise, head trauma, virus or disease, autoimmune inner ear disease, hereditary, aging (Presbycusis), malformation of the inner ear, tumors etc [12].

Major Causes of Hearing Loss in India

WHO (survey4) have listed some of the major causes of hearing loss and ear diseases in India. Ear wax (15.9%) being the most common cause of reversible hearing loss is followed by middle ear infections such as chronic suppurative otitis media (CSOM) (5.2%) , serous otitis media (3%) and dry perforation of tympanic membrane (0.5%). On the other hand 50% of congenital hearing loss causes is attributed to environmental factors (congenital hyperbilirubinemia, ototoxic medication exposure, neonatal hypoxia, viral infections and meningitis) and the rest 50% to inherited (genetic). Approximately 30% of these hereditary cases are classified as syndromic and the other 70% as non-syndromic. The small subset (syndromic) of hearing loss patients (15% of all patients with hearing loss) is the group most readily diagnosed by physicians due to recognizable features other than hearing loss [3, 8].

Urban Vs. Rural India

Rural children in India more often suffer hearing loss than children living in urban areas. A study on Indian school children aged between 12 and 14 years found that 33% of rural children suffered from some sort of hearing loss in comparison to 6% of urban living children. The difference was also observed in the degree of hearing loss where 27 % of the children from rural areas suffered from moderate hearing loss as compared to approximately 3 % among the urban children. On the other hand 6 % of the rural children and about 2 % of the urban group had mild hearing loss. The significant difference in the prevalence is attributed primarily to the lower socio-economic status of the rural population often leading to malnutrition. Secondary reasons such as poorer health education and inadequate medical facilities increase the risk of hearing problems. Otitis media was the most common cause of hearing impairment in both the groups [16].

Major Reversible/Preventable Causes of Hearing Loss

Following are some of the preventable or reversible causes of hearing loss.

Impacted Wax

Impacted cerumen is when earwax (cerumen) builds up in the ear and blocks the ear canal; it can cause temporary hearing loss and ear pain. Earwax produced by glands in the ear canal helps protect the ear by trapping dust and other foreign particles that could damage the ear. At times the extra wax can build up and harden in the ear canal and would become difficult to remove. Earwax also can become impacted when, during ear cleaning, the wax is accidentally pushed deeper into the ear canal. Along with symptoms of pain, transient hearing loss, itching, it can also increase the risk of outer ear infection. Impacted cerumen needs intervention by an otolaryngologist so as to avoid damaging the ear. Hearing usually returns completely after the impacted earwax is removed [1].

Otitis Media

Otitis media is an inflammation in the middle ear and is usually associated with accumulation of fluid. The fluid may or may not be infected. The Symptoms vary depending upon the severity, frequency, and duration of the condition. In the early stage, thin, clear, non-infected fluid without any pain or fever is observed with a slight decrease in hearing ability. If left untreated, the symptoms worsen and the accumulated fluid is infected and turns into a thick "glue-like" fluid and possible complications such as perforation of tympanic membrane or permanent hearing loss is observed. Hearing loss is nearly always fluctuating in otitis media depending on the extent of infection [6]. Otitis media is considered to be the most common cause of hearing loss in young

children and is attributed to the shorter eustachian tube in children than adults which allows easy entry of bacteria and viruses into the middle ear. Until the eustachian tube changes in size and angle as the child grows, children are more susceptible to otitis media. Wrong feeding postures are one of the major causes of otitis media.

Signs you can look for that may indicate impacted wax or recurring fluid in the ear:

1. Inattentiveness
2. Wanting the television or radio louder than usual
3. Misunderstanding directions
4. Listlessness
5. Unexplained irritability
6. Pulling or scratching at the ears

Drug Induced Hearing Loss

Certain drugs known to be ototoxic induce hearing loss. Most often the hearing loss observed is temporary in nature, unless prolonged usage of drugs. The toxic effect is seen on the sensory and the neural system, inducing sensorineural hearing loss. Hearing loss observed is symmetrical and bilateral. Some of the ototoxic treatment includes antibiotics such as vancomycin, amikacin, and tobramycin. Prolonged use of non-steroidal analgesic anti-inflammatory drugs, chemotherapeutic drugs, quinin and loop diuretics also cause hearing loss [11].

Noise Exposure

The American Academy of Pediatrics note that the infant's exposure to noise in the neonatal intensive care unit (NICU) may result in cochlear damage and may disrupt normal growth and development in premature children. Some evidence indicates that noise exposure and the use of ototoxic drugs such as aminoglycosides are synergistic in producing an auditory damage [17].

Awareness about Causes for Hearing Loss

Among the causes studied, more awareness was present for family history (67%), noise exposure (62%), and ear discharge (61%). Less than 50% of the participants were aware of other causes. 63% were aware that hearing loss could be congenital. Interestingly, 38% of the participants believed that the hearing loss was caused by bewitchment and 31% believed it was because of ancestral sins. Only 20% were aware that hearing loss could be identified at birth, 75% thought that intervention is possible for hearing loss, and 33% believed that hearing-impaired children could attend regular school, if intervened [9]. The statistics are a warning sign and brings forth the myths and misconceptions among the community to be addressed. In addition, it is evident that the early identification is still not given importance.

Conclusion

Hearing loss can thus occur due to various causes. It is well recognized that hearing is critical to speech and language development. Earlier the hearing loss, serious is the effect on the child's life. However, this effect can be minimized by early identification and intervention. Hence, it is essential for the health professionals, parents and caretakers of the children to be aware of the causes of hearing loss. This knowledge would aid in the early identification of hearing loss. The priority place held by primary health care center (PHC) in the Indian health care delivery system, demands more emphasis in educating the professionals at PHC. Further, measures should be taken to clear misconceptions about causes of hearing loss, as it may impede the identification of hearing loss.

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