Pediatric hearing loss – causes and management

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Abstract
Introduction: Around 34 million children have disabling hearing loss. The causes leading to impairment in the hearing analyzer are classified as congenital and acquired.
Purposes: Early diagnosis and hearing rehabilitation are essential for the child's development. The purpose of this article is to assist the approach of detection and management of this condition, therefore to highlight the syndromes and diseases that lead to disturbance in hearing, making them more recognizable.
Methods: The information in this article is based on our clinical experience with children with hearing disorders as well as on the literature review.
Results: Children in whom diagnosis and rehabilitation were performed timely show better development and more effective results than those where the delay has led to disabilities in linguistic and social abilities.
Conclusions: The London Dysmorphology Database has listed 396 syndromes that include hearing disorders, with 30% of congenital deafness being part of syndrome complex. Screening programs in newborns allow early diagnosis and rapid intervention. Some of the causes that lead to hearing loss are treatable, while others are overcome by hearing aids.

Introduction

Hearing impairment is one of the most prevalent chronic conditions in children. Around 360 million people – 5% of the world's population – live with hearing loss which is considered disabling; of these, nearly 32 million are children. Hearing loss can be a congenital pathology, caused by genetic factors or by complications during pregnancy and childbirth. It can also be acquired later in life, at any age.
The types of hearing loss depending of the physical location of the problem within the ear are conductive, sensorineural, mixed and auditory neuropathy. Classified by its onset hearing loss can be congenital which is always prelingual, and acquired which can be prelingual or postlingual. According to the American Speech-Language-Hearing Association (ASHA), normal hearing occurs in the range of −10 to 15 dB, with a slight HL if the range of loss is within 16 to 25 dB. Mild HL occurs when the hearing loss ranges between 26 to 40 dB, moderate HL is when the hearing loss ranges between 41 to 55 dB and moderate to severe HL ranges between 56 to 70 dB.

Epidemiology and risk factors

The estimated prevalence of permanent bilateral hearing loss is 1 per 1,000 live births as it increases to 4 per 1,000 live births in the developing countries. (1)

The Joint Committee on Infant Hearing (JCIH) endorses early detection of and intervention for infants with hearing loss. The goal of early hearing detection and intervention is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. The JCIH Year 2007 Position Statement identified eleven indicators associated with hearing loss. Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood:

1. Caregiver concern regarding hearing, speech, language, or developmental delay.
2. Family history of permanent childhood hearing loss.
3. All infants with or without risk factors requiring neonatal intensive care for greater than 5 days, including any of the following: ECMO,* assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion.
4. In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplamosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fractures that requires hospitalization.
11. Chemotherapy.

According to the World Health Organisation (WHO) a number of factors determine what the impact of hearing loss is on an individual. These include: age of onset, degree of hearing loss, age of identification and intervention, environment. WHO estimates that about 60% of hearing loss is due to preventable causes.

Over 30% of childhood hearing loss is caused by infections, such as rubella, cytomegalovirus, mumps, meningitis, measles and chronic ear infections. Meningitis and rubella together are responsible for over 19% of childhood hearing loss. Most of these infections can be prevented by immunization and good hygiene. Ear infections and glue ear can be prevented through good ear care and general hygiene, and can be treated by prompt medical and surgical interventions.

Complications at birth, such as lack of oxygen, low birthweight, prematurity and jaundice, account for 17% of childhood hearing loss. Such complications can be prevented through improved maternal and child health practices.

Use of ototoxic medicines in pregnant women and children is responsible for 4% of childhood hearing loss, which could potentially be avoided.

Diagnosis and management

Early identification of hearing impairment in children when followed by timely and appropriate interventions can minimize developmental delays and facilitate social development, education and communication. The implementation of national newborn hearing screening programs has improved the management of affected children by rehabilitating them at early stages. (2)All infants should have access to hearing screening using a physiologic measure no later than 1 month of age. Such measures include OAE and automated ABR testing. Both OAE and automated ABR technologies provide noninvasive recordings of physiologic activity underlying normal auditory function, both are easily performed in neonates and infants. (3)

The audiologic assessment algorithm includes the following: case history, otoscopy, behaviour assessment, physiologic assessment, genetic testing. All infants with confirmed permanent hearing loss should receive early intervention services as soon as possible after diagnosis but no later than 6 months of age. Severe to profound bilateral hearing loss can be managed by cochlear implantation (unilateral or bilateral), if picked up at early age while mild to moderate bilateral hearing loss is easier to manage with conventional hearing aids. (4)(5)

Approximately 30% of the genetic cases of hearing loss are considered to be syndromic. The London Dysmorphology Database has listed 396 syndromes that include hearing disorders. Among the well-known syndromes are Alport syndrome, Brachio-oto-renal syndrome, Down's syndrome, Hunter's syndrome, Pendred's syndrome, Sickler's syndrome, Treacher-collins, Usher's syndrome, Waardenburg's syndrome. (6)

Once the underlying cause of hearing loss is established, management options include specific antimicrobial therapies, surgical treatment of craniofacial abnormalities and hearing aids.

Summary and conclusion

Hearing is the key to learning spoken language and is important for the cognitive development of children. Hearing loss is the most common neurosensory disorder in humans. It can be a congenital pathology due to genetic factors or caused by complications during pregnancy and childbirth. It can be also acquired later in life. Key elements in ensuring the best outcomes for children with permanent hearing loss are: early identification; appropriate hearing technology; professional support for communication, learning and education; and a family-centred approach.

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