Available online at http://www.journalijdr.com

IJDR

International Journal of DEVELOPMENT RESEARCH

ISSN: 2230-9926

International Journal of Development Research
Vol. 06, Issue, 11, pp.9929-9932, November, 2016

Full Length Research Article

DIAGNOSIS AND CARE OF PEDIATRIC HEARING LOSS

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ARTICLE INFO

Article History:
Received 22nd August, 2016
Received in revised form
17th September, 2016
Accepted 21st October, 2016
Published online 30th November, 2016

Key Words:

Hearing loss,
Childhood,
Deafness,
Education.

ABSTRACT

Hearing loss and deafness can develop during infancy or childhood or adolescence or during old age. Congenital hearing loss is relatively frequent and has serious negative consequences if it is not diagnosed and treated. Worldwide 360 million people living with disabling hearing loss. In this 32 million of these are children (<15 years of age). Children whose hearing loss is > 90 dB are considered deaf for the purposes of educational placement. Children cannot express what they can and what they can't hear and clear observation is the best method used for assessing their hearing acuity. Hearing loss is affect the educational attainment, future employment, future earnings etc. Impairments in hearing can occur in one ear or sometimes it can be in both ears. This information may be useful to understand causes, various types of hearing loss, to give better treatment of the hearing loss, and predict the chances that future children will have hearing loss.

INTRODUCTION

The term "hearing impairment" is used generically to describe a wide range of hearing losses, including deafness. Hearing impairment is defined by Individuals with Disabilities Education Act (IDEA) as "an impairment in hearing, whether permanent or fluctuating, that adversely affects a child's educational performance." Deafness is defined as "a hearing impairment that is so severe that the child is impaired in processing linguistic information through hearing, with or without amplification" (Nichcy Disability Fact act Sheet, 2014). Hearing loss and deafness affect individuals of all ages and may happen at any time. It can develop during infancy or childhood or adolescence or during old age. Children whose hearing loss is > 90 dB are considered deaf for the purposes of educational placement. Sound is measured by its loudness or intensity in decibels (dB) and its frequency in hertz (Hz). A child's hearing acuity is classified as normal if it is within 20 dB. Severity of hearing loss described as mild (20–40 dB), moderate (41–55 dB), moderately severe (56–70 dB), severe (71–90 dB), or profound (when hearing level is >90 dB) (Smith *et al*., 2004). Disabling hearing loss refers to hearing loss greater than 40 dB in the better hearing ear in adults (15 years or older) and greater than 30 dB in the better hearing ear in children (0 to 14 years).

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(WHO global estimates on prevalence of hearing loss, 2012). Congenital hearing loss is relatively frequent and has serious negative consequences if it is not diagnosed and not treated during the first few months of life (Karl, 2003). The impact of hearing loss has been shown to affect the language and reading (Delage *et al*., 2010; Moeller *et al*., 2007), particularly the vocabulary development (Davis, 1981; David, 1986 and Wake *et al*., 2004), and also pragmatic and social skills (Most *et al*., 2007; Wolters *et al*., 2011). Hearing loss is affect the educational attainment, the likelihood of future employment, future earnings, the use of health-care systems, and life expectancy (Mohr *et al*., 2000 and Carvill, 2001).

Prevalence and Etiology

Worldwide 360 million people living with disabling hearing loss. In this 32 million of these are children (<15 years of age). Common Causes for hearing loss include Maternal infections such as Rubella, Meningitis, measles and mumps, Severe jaundice following birth, Birth anoxia, Chronic ear infections, Use of ototoxic medicines, Low birth weight and Noise (WHO; 2011 Estimates) Hearing loss that is presumed to be late onset and at least moderate in severity is diagnosed in 1·2–3·3 per 10 000 school-aged children (Fortnum *et al*., 1997). The most common cause of conductive hearing loss in infants and young children is acute otitis media or otitis media with effusion. Other causes may include tympanic membrane perforation, narrowing of the auditory canal, ear wax, foreign

body etc. Perforations of the tympanic membrane cause hearing loss by reducing the surface area available for sound transmission to the ossicular chain. The main causes of tympanic membrane perforations are chronic otitis media and trauma. Traumatic perforations of the tympanic membrane can occur because of swimming or diving, barotrauma, explosions, penetrating injury, and temporal bone fractures. There are many reasons for sensorineural hearing loss, that include genetic factors, infections during pregnancy, premature birth, complications at birth and childhood illnesses, drugs, physical trauma and structural causes etc. Hearing impairment also occurs in structural damage. It can be congenital or also happens due to Head injuries or Exposure to loud noise. Structural abnormality of hearing loss includes Absence or malformation of the external ear, auditory canal, tympanic membrane, ossicles, and Inner ear malformations i.e. enlarged vestibular aqueduct. By aetiology, more than half of neonates with SNHL have inherited hearing loss (Richard *et al*., 2005). Inherited SNHL generally (70%) appears as an isolated physical finding (non-syndromic SNHL).

In this case, the person only has hearing loss. About 30% of cases are syndromal (i.e., associated with other health problems, such as hearing loss with Kidney problems (Alport), Thyroid gland enlargement (Pendred), Vision impairment (Usher), Heart problems (Jervell and Lange-Nielsen). Genetic testing may help determine if the hearing loss is genetic. Ear infections during pregnancy include TORCH infections (toxoplasmosis, others, rubella, cytomegalovirus, and herpes simplex viruses). After birth infections includes Infections caused by bacteria (bacterial meningitis, syphilis), Infections caused by viruses (measles, mumps). Some drugs and other chemicals can affect the fetus, while others cause hearing loss after birth. Some of these include during pregnancy consumption of alcohol, Medications and (e.g. Accutane, Dilantin), therapies (Chemotherapy etc). Ototoxic drugs (e.g. Aminoglycoside antibiotics, Diuretics, Cisplatin). The world-wide burden of SNHL secondary to congenital rubella syndrome remains high, and in countries without a rubella vaccination programme, congenital rubella syndrome continues to rank as the most important cause of acquired congenital SNHL (Banatvala, 2004). Acquired SNHL in infants and children is most commonly caused by bacterial meningitis. Altogether, bacterial meningitis accounts for about 6% of all cases of SNHL in children (Fortnum, 1993).

Types of hearing loss

Hearing loss is categorized as congenital hearing loss, sensory neural hearing loss and mixed hearing loss. Conductive hearing loss (CHL) means that sound cannot pass efficiently through the external and middle ear to the cochlea and auditory nerve are caused by obstructions or diseases in the external or middle ear. Sensorineural hearing loss (SNHL) is comes under permanent hearing loss. It results from abnormality of the cochlea, auditory nerve pathways, or higher aspects of central auditory area. It includes damage to the delicate sensory hair cells of the inner ear. SNHL is the most common sensory deficit in more developed societies (Davis, 1989; Wilson *et al*., 1999). Most patients with this type of hearing loss are adults, children also can be affected. Hereditary and nonhereditary congenital hearing loss is the two major pediatric classifications. The majorities of hereditary losses is autosomal recessive and are frequently associated with other systemic findings. More than 100 congenital syndromes are associated with sensor neural hearing loss. The consequences of delayed detection can be significant. Neonates considered at high risk for congenital hearing loss (Joint Committee on Infant Hearing, 2000). A mixed hearing loss refers to a combination of conductive and sensor neural hearing loss.

Table 1. Etiology of Hearing loss: (Parthasarathy, 2013)

Type of deafness	Period	Specific cause
Conductive	Any age	Cerumen Foreign body Otitis media and Otitis media with effusion Congenital malformation
Sensory neural	Prenatal	Genetic - Klippel – feil syndrome - Waardenburg syndrome - Pendred syndrome Non Genetic - Disease during pregnancy Toxemia Diabetis Nephritis Measles Other viral infections - Drugs during pregnancy Streptomycin Quinine Salicylates
	Perinatal	Birth trauma - Anoxia Hemolytic disease - Kernicterus Prematurity
	Postnatal	Genetic - Otosclerosis - Femilial perceptive deafness Non genetic - Infectious diseases - Trauma - Otitis media - Ototoxic drugs

A central hearing loss results from damage or impairment to the nerves or nuclei of the central nervous system, either in the pathways to the brain or in the auditory area. Depending upon the cause hearing loss may be temporary or permanent, genetic (hereditary) or non-genetic (environmental) and either stable or progressive. A temporary hearing loss is more common and comes under conductive deafness. Permanent hearing losses are usually sensorineural, but may also be conductive type. Time of onset is established as either congenital or acquired. Other types include Impairment in both ears (Bilateral deafness) or in one ear (Unilateral deafness). Impairment is similar in both ears (Symmetrical deafness) or different in each ear (Asymmetrical deafness). Systemic disease should produce bilateral rather than unilateral ear symptoms (Ottaviani, 1999).

Diagnosis

Children cannot express what they can and what they can't hear and clear observation is the best method used for assessing their hearing acuity. Review all medical records, audiologic records and case history information for risk factors associated with congenital hearing loss and/or late-onset or progressive hearing loss. Inspection and palpation of the auricle and periauricular tissues are done to rule out the local tenderness and other pathology. An otoscope should be used to examine the external auditory canal for wax, foreign bodies, and abnormalities of the auditory canal. The mobility, color, and surface anatomy of the tympanic membrane should be determined. Hearing acuity can be measured with either objective or subjective tests in childrens (Richard, 2005).

Objective Tests

Objective hearing tests do not require responses or co-operation from a child. Physiological tests objectively assess the functional status of the auditory system can be done at any age. These tests include brainstem auditory evoked response, otoacoustic emissions (OAE), auditory steady state response (ABR), and impedance testing (Tympanometry). ABR testing measures the degree of hearing and neurologic function. In this small electrodes are placed behind the child's ears and a ground electrode is placed on top of the head. Very fast click sounds are sent through earphones and then the electrodes pick up the response from the brainstem and send it to a computer screen. It measures the response to sounds from the brainstem. In OAE, Otoacoustic emissions (sounds generated from the cochlea) are measured by a probe which is placed in the ear canal, and the response is displayed on a computer screen. This test is useful to detect the defect present in the cochlea or in the nerve.

Subjective tests of hearing acuity

Subjective testing does require a behavioral response. These tests are done in the test booth by watching how a child processes auditory information and responses to sounds. Subjective tests include behavioural and pure-tone testing. From birth until approximately nine months of age, a child is tested by behavioral observation. From nine months of age until approximately three years, visual reinforcement is used. And after three years of age, play testing procedures are used. In behavioral observation, sounds produced through earphones or through loudspeakers, and observe his response by looking carefully at his face for eye widening or eyebrow scrunching, fast sucking or stop sucking. In visual Reinforcement Audiometry, consists of reinforcing the child's response to sound by visual stimuli. When a tone is presented and the child looks in the direction of that sound, the box is lit and the child sees a character beating on a drum. In play Audiometry, a child can be taught to dropping a toy into a bucket or Building blocks or cups or dancing/Shaking in the chair or poking the child's nose or clapping etc. every time he hears a sound and hearing capacity is noted.

Method of examination hearing loss in young children's

In young children's whispering test and watch test can be helpful in making a gross evaluation of hearing. Next tuning fork test is performed to diagnose the hearing loss. Tuning fork wave length is varying from 128 Hz to 2056 Hzs. It consists two types of tests, one is rinne's test and another one is weber's test. In the Rinne's test air conduction is comparing with bone conduction. The vibrating tuning fork is placed on the mastoid bone and asks the patient about the hearing. When the patient no longer can hear the sound, the tuning fork is placed adjacent to the ear canal. In the presence of normal hearing or sensorineural hearing loss, air conduction is better than bone conduction. Therefore, sound is still heard when the tuning fork is placed adjacent to the ear canal. In the presence of conductive hearing loss, bone conduction is better than air conduction, and the sound is not heard when the tuning fork is placed adjacent to the canal. Weber's test is performed by striking tuning fork and placing it midline on the patient's scalp, or on the forehead. If the hearing loss is conductive, the sound will be heard best in the affected ear. If the loss is sensorineural, the sound will be heard best in the normal ear. The sound remains midline in patients with normal hearing.

Treatment

Condition	Management
If wax is present in the auditory canal	Cerumenolytics
If ear contained any foreign bodies If the object is not impacted or hygrostatic If the child is cooperative	It can be removed by irrigation or with a curette or with endoscope Warm water irrigation or with endoscope It can be removed manually
Otitis externa or Chronic otitis media with tympanic membrane perforation	Ototopical antibiotics and oral antibiotics Surgical correction is advisable in long standing perforations.
Acute otitis media Recurrent acute otitis media	Aural toilet, Antibiotics Myringotomy tubes are recommended
Cholesteatoma, Otosclerosis	hearing aids or surgical repair by stapedectomy
Sensory neural Deafness If not benefitting with aid	Hearing aid Cochlear implant

It is estimated that in developing countries, about 20% of people who have hearing loss require hearing aids, suggesting 72 million potential hearing aid users worldwide. However, current production of hearing aids meets less than 10% of the global need. In developing countries, less than 3% of people who need a hearing aid are thought to have one. (WHO 2011 estimates).

Conclusion

Early detection and intervention is the most important factor in minimizing the impact of hearing loss. Immediate reasonable management such as communication therapy, parent support, hearing devices hearing aids and cochlear implants etc. should not be delayed.

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