Hearing impairment among children

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Abstract

Hearing impairment is present in 3-6 per 1000 live born babies. Early diagnosis and adequate treatment can improve the overall development of the child. Reliable screening tests and appropriate treatment are available at present even during early infancy. The high incidence of hearing impairment, availability of screening tests and appropriate treatment fulfill all the criteria for compulsory screening for all babies during early infancy.

Key words: Hearing impairment, Otoacoustic emission, Auditory brainstem response, Cochlear implant

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Introduction

Speech perception is one among the most sophisticated human function. True speech perception is unique to human beings. Impairment in hearing is the most common congenital disabilities in childhood. Hearing impairment in children can be either acquired or congenital. The incidence of congenital permanent hearing loss is 1.5 to 6 per 1000 live births. [1] Recent study done in India reported 5.6 per 1000 live birth. [2] Genetic factors contribute to 50% of the childhood hearing impairment. Prenatal, perinatal and postnatal environmental factors account for 20-25% and etiology is uncertain in 25-30%of cases [3]. Unless screened, hearing impairment remains undetected during infancy.

Early screening for hearing impairment is very important. The incidence of hearing loss is much higher than the usually screened conditions like congenital hypothyroidism (0.25 in 1000 live births) and phenylketonuria (0.08 in 1000 live births) [4]. Significant hearing loss may lead to poor language development, communication skill, cognition and social-emotional development. This in turn results in lower education level and employment opportunities. Early detection and intervention can lead to normal language development and cognition because of plasticity of developing sensory system. The critical period of speech and language development is 0-3 years [3, 5-9]. Cost effective screening tools like otoacoustic emission (OAE) and auditory brain stem response (ABR) are readily available. These tests have high sensitivity (92%) and specificity (98%) in early neonatal period itself [3]. There is easy availability of cost effective intervention which, if utilized appropriately, can correct the disability. It comprises of amplification or sensory devices such as hearing aids, frequency modulator and cochlear implants [3,5,7,10].

The timely information provides acceptance by parents and public and improves the readiness for interventions.

Auditory system and speech development:

Development of the auditory system starts early in the fetal life. The first sign of expansion of auditory brain- stem pathway occurs by 16 weeks of gestation. First appearance of the synapses corresponds to the fetal hearing. It starts by 28th week of gestation. Auditory brainstem responses can be recorded by this age. Development of the inner ear and functional auditory brain stem pathway is complete several weeks before the birth. A normal newborn can process the sound and accurately analyze the loudness and pitch of the sound. Localization of the sound and discrimination of speech occurs in early infancy. Infants with hearing thus learn to process speech and understand language by one year of age. These infants are linguistically sophisticated by one year of age. When there is deprivation of the sensory input, morphological and func- tional properties of neuron breaks down. Reintroduction of the sensory stimulation during this period can ameliorate the deleterious effect [11].

In the present scenario most of the children with hearing impairment are unidentified during their early infancy. It goes undetected until as late as two years of age without a specialized screening test [8, 11]. Most of the cases are identified by the parents once the children do not start achieving language milestone for the corresponding chronological age. The mean age of presentation of hearing impairment without screening is approximately thirty months and the critical period of the language development starts before six months of age [8]. This time lag can be reduced by early screening.

Hearing evaluation during early infancy:

Newborn screening for hearing loss evolved over decades. In 1896 Thomas Barr introduced sound bells and sharp voice for evaluation of hearing. There was high false positivity for these tests and mild and moderate hearing loss could not be detected. These methods were used till 1940. During the period of 1940-1950, electro-dermal response audiometry was used. In 1974, Blair Simmons developed an automated technique of behavioral screening called Crib-o-gram. He utilized motion sensitive transducers attached underneath the infant's bassinets, with a strip to record motion, an automatic timer and a loudspeaker. A high frequency signal of 92dB presented in scheduled interval and baby's movement was charted. The incidence identified was about 1 in 1000 in well-baby nursery and 1 in 52 in intensive care problems with Crib-o-gram. It depended upon baby's behavioral state, optimal stimulus, and a very low ambient noise. It had a high false positivity and negativity with poor reliability [3, 4].Later two tests namely;otoacoustic emission (OAE) and auditory brainstem response(ABR) were found to be most useful for screening of hearing defects in infants.

Who should be screened?

In early 70s National Joint Committee on Infant Hearing Screening (JCIH), composed of representatives of academy of pediatrics, academy of otolaryngology and American speech and hearing association recommended the high risk register for hearing evaluation [3]. The five categories included were:

1. A history of hereditary childhood hearing impairment.
2. Rubella or other nonbacterial intrauterine fetal infections
3. Defects of ear, nose or throat.
4. Birth weight < 1500gms
5. Any free or indirect serum bilirubin concentration judged to be potentially neurotoxic.

Prevalence of hearing impairment in high risk group was 14 times greater than the general population and this concept assumed that one can identify small group of children possessing high chance of having the handicap. Although there was increased incidence of hearing impairment in this group, around 50%of the children with significant hearing loss did not have risk factors. In 1993, National Institute of Deafness recognized the above issue and recommended universal hearing screening within first three months. A two stage screening protocol with Otoacoustic emission (OAE) followed by Auditory brainstem response (ABR) for infants who fail OAE were recommened. In 1994 JCIH recommended identification of the impairment by three months and intervention by six months [3].

In 2000, JCIH recommended the following [12]:

1. All babies should be screened before one month and all babies who required admission in NICU should receive screening before discharge.
2. All infants who do not pass birth admission screen should be screened before three months of age.
3. All infants with confirmed permanent hearing loss should receive services before six months of age.
4. All infants who pass screening and have risk factors should receive ongoing audiological and medical surveillance and monitoring for communication development.
5. Infants and family rights are guaranteed through informed choice, decision-making, and consent.
6. Infant hearing screening and evaluation results are afforded the same protection as all other health care and educational information.
7. Information system used to measure and report the effectiveness of early hearing detection and intervention (EHDI) services.

In 2007 JCIH updated 2000 statement. These included definition of targeted hearing loss, hearing screening and re-screening protocol, diagnostic audiological evaluation, medical evaluation, early intervention, surveillance and screening in medical home, communication and information infrastructure. The definition was expanded to include neural hearing loss (auditor dysnchrony) in infants admitted in NICU [13].

Recently, in July 2008, United State preventive service and task force made following recommendations (B recommendation) [14].

1. Screening for all newborns within one month with two step screening process, first with OAE and followed by ABR who fails the first test.
2. Newborn delivered at home and hospitals without hearing screening facilities, should have some mechanism for referral
3. Early intervention should be designed to meet the individual needs of the infant and the family
4. Babies with risk factors should undergo periodic screening evaluation till three years.

How should we screen the infants?

Evoked OAE and ABR are the two screening tests recommended for hearing assessment [3, 13-15]. Otoacoustic emissions are acoustic energy originating in the normal inner ear, which can be detected by acoustic probe connected in the external ear canal. Otoacoustic emission was first described by Kemp. These are produced by the action of outer hair cells. There are two types of OAE. One is spontaneous, present in the absence of external stimulation and the second is evoked OAE. Evoked can be further divided into transiently evoked OAE (TEOAE) which are evoked by acoustic transient such as click or tone burst, stimulus frequency OAE (SFOAE), elicited by single continuous tone, and distortion product OAE (DPOAE), which are generated by two continuous pure tone separated by specific frequency. Emissions are not measurable if conductive or sensorineural hearing loss exceeds 30dB to 35dB HL at 0.5 kHz, 2 kHz and 4 kHz and 40dB at 1 kHz. Background noise in nursery does not interfere with the procedure if the probe is well fixed, but internally generated noise, such as sucking or swallowing may interfere in estimating low frequency emissions [16-19].

Auditory brainstem response is the second screening test used if babies fail in the EOAE. The auditory stimuli produce electrical activity in the brain stem that can be detected by surface scalp electrodes. ABR is used for estimating thresholds, absolute latencies, and inter-wave latencies. There are seven positive waves, the presumed origins of which are as follows:

Wave I – VIIIth nerve
Wave II – cochlear nucleus
Wave III – superior olivary complex
Wave IV – lateral lemniscus
Wave V – inferior colliculus
Wave VI – medial geniculate body
Wave VII – auditory cortex

The procedure is not affected by sleep or sedation. Advantages of this test include objective measurement of auditory system, provide ear specific information and are independent of subject state [16-19].

Other forms of hearing evaluation include electrocochleography, visual reinforcement audiometry (VRA), play audiometry and sound field audiometry. However only EOAE and ABR are suitable for neonatal screening [17,18].

Management of hearing impaired children:

Early intervention includes evaluation for amplification or sensory devices, surgical and medical evaluation and communication assessment therapy. Cochlear implants are considered in case of severe to profound hearing loss only after inadequate response to hearing aids [3,18].

The purpose of treatment is to obtain as much hearing as possible and to achieve adequate communication for speech, language and social development. Hearing aids, cochlear implants and speech therapy are the cornerstones of the rehabilitation [13,14].

Selection and fitting hearing aid is a challenging process. Obtaining accurate, frequency-specific threshold measurement is difficult for infants. Also selecting hearing aid type, setting hearing aid output is difficult in infants. Since ear pinnas in these children are small it is difficult to hold the hearing aid. The ear mould must be remade according to child's growth. [18,20]. Prior to fitting hearing aids, diagnostic tests should be done including OAE, ABR, acoustic reflexes, VRA and audiometry depending upon the age. If the ABR or VRA is suggestive of sensori-neural hearing loss and the OAE are absent, hearing aid is advised. If OAE is normal with absent ABR further investigation for auditory neuropathy are warranted. The treatment option include traditional amplification, frequency modulated (FM) systems, vibrotactile stimulation, cued speech and cochlear implantation

Hearing aids are essential and should be fitted early. They are useful even in profound hearing losses. They are electronic instruments which amplify the sound and help the patient in using the remaining hearing for communication. Hearing aids can be behind-the-ear (BTE), completely inthe-canal (CIC), in the ear (ITE) and over body parts. Implantable hearing devices are hearing aids which are implanted inside the body. They are currently used for external or middle ear impairment with good bone conduction Bone conduction hearing devices transmit amplified signals by skull vibration into the inner ear. Over body aids are usually not used in children because they do not have sound processing and programming flexibility. Completely in the canal can only be used after seven to eight ears of age. Since concha is smaller in size ITE also cannot be used in infancy. Because of all these reasons BTE is the choice in infancy. Initially during early infancy directional microphone system is used as it enhances the sound coming from the front. Once child starts crawling this can be changed to omni-directional. Tamper proof battery are recommended [18,20].

Cochlear implants are electronic devices which detect environmental sounds and transform them to electrical impulses that are transmitted to the acoustic nerve. The basic components are a microphone, a processor and an electrode [3,19,21]. They are used in case of profound bilateral sensorineural hearing loss, which cannot be helped by hearing aids. Cochlear implantation in children requires a team approach. Children typically undergo a trial period of six months. Child's skills are re-evaluated. Child who shows little or no improvement with hearing aid is the usual candidate. All children who have earphone threshold of 90 dB hearing level or more are considered potential candidates for cochlear implant. Radiological evaluation of the temporal bone with high resolution computed tomography is required before surgery. Surgery entails the placement of electrode into the scala tympani of the cochlea and placement of the receiver stimulator in the skull. Post-operative programming and training is required in these children [21,22].

Counseling of parents is another important issue. Parents should be counseled about the disease and treatment options. They should be given option when to start treatment and what hearing aid to select. Unnecessary delay in hearing aid fitting because of parental confusion should be avoided [23,13].

The screening tests and treatment are not without concern. Though EOAE and ABR both are sensitive and specific, both can have false positive and false negative results False positive test is associated with long term parental concerns. These concerns persist in about 10% of parents even after repeat test shows negative result [25]. There is also risk of meningitis in children with cochlear implants. Facial nerve injuries, cerebrospinal fluid leakage, damage to cochlear nerve are other complications

Conclusion

Screening for hearing impairment fulfills all the criteria for compulsory testing of all babies. Early intervention for positive screen improves the language development and communication. However screening should be done with minimal false positivity. Delay in diagnosis and treatment results in irreversible damage to the child's development.

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