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Short Communication

Deafness: The Neglected and Hidden Disability

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Hearing loss has become the fourth leading cause of disability globally. According to a range of studies and surveys conducted in different countries, around 0.5 to 5 in every 1000 neonates and infants have congenital or early childhood onset sensorineural deafness or severe-to-profound hearing impairment [1]. Hearing loss is a hidden problem because children, especially infants and toddlers cannot tell us that they are not hearing normally. Nearly all their parents have normal hearing with little or no knowledge about hearing loss in children. Deaf and hearing-impaired children often experience delayed development of speech, language and cognitive skills, which may result in slow learning and difficulty progressing in school. Congenital and early childhood onset deafness or severe-to-profound hearing impairment may affect the auditory neuropathway of children at a later developmental stage if appropriate and optimal interventions are not provided within the critical period of central auditory pathway development. The critical language learning period of a child is from birth to about three and half years of age. If the hearing deficit is corrected before the child is of six months age, the development of speech and language is more or less normal and equal to that of a normal hearing child. Therefore, early detection is a vitally important element in providing appropriate support for deaf and hearing-impaired babies that will help them enjoy equal opportunities in society alongside all other children.

According to the World Health Organization (WHO), a hearing loss is considered to be disabling for adults (15 years or older) if it is greater than 40 dB in the better hearing ear. For children (0 to 14 years), the WHO has defined a hearing loss exceeding 30 dB in the better hearing ear as disabling. However, hearing loss at lower thresholds also has a negative impact. For example, children whose hearing loss exceeds 26 dB have trouble understanding soft speech from a distance or in background [1,2].

Handicapping effects of hearing loss in children

Fortunately many of the adverse implications of deafness in babies can be prevented substantially. Many research studies had demonstrated that early intervention in hearing-impaired children results in improved language development, increased academic success and increased lifetime earnings. If it is to be effective, early intervention with deaf children should begin before the Childs first birth. To do a much better job of early identification and early intervention, and to reduce the unnecessary suffering, poor educational performance and lack of productivity that so often accompany deafness, three groups of people must work together:

- Parents are in the best position to identify their child's hearing difficulties. We as health care providers need to do a better job of making parents aware of normal developmental milestones in speech and language, the danger signals suggesting the presence of hearing loss in their child, and of the sources of help that are available to them.
- Physicians need to become more responsive to parents concerns about their child's hearing. Too often, those concerns are brushed aside or ignored completely. Yet, a recent study found that parents of hearing-impaired children knew about their baby's hearing loss an average of 7 months before it was diagnosed and that almost all of them were given poor advice, such as " don't worry about it" or " wait until the child starts school," when they told their doctors about their concerns.

Average hearing level (500-2KHz)	Description	Possible condition	What can be heard without amplification	Handicapping effects (if not treated in 1st year of life)	Probable needs
0-15 dB	Normal range	Conductive hearing losses.	All speech sounds	None	None
15-25 dB	Slight hearing loss	Conductive hearing losses, some sensorineural hearing losses	Vowel sounds heard clearly; may miss unvoiced consonant sounds	Mild auditory dysfunction in language learning	Hearing aid, speech reading, auditory training, speech therapy, preferential seating
25-30 dB	Mild hearing loss	Conductive or sensorineural hearing loss	Only some speech sounds, the louder voiced sounds	Auditory learning dysfunction, mild language retardation, mild speech problems, inattention	Hearing aid, speech reading, auditory training, speech therapy
30-50 dB	Moderate hearing loss	Conductive hearing losses, chronic middle ear disorders; sensorineural hearing loss	Almost no speech sounds at normal conversational level	Speech problems, language retardation, learning dysfunction, in attention.	All of the above plus consideration of special class room situation
50-70 dB	Severe hearing loss	Sensorineural or mixed losses	No speech sounds at normal conversational level	Severe speech problems, language retardation, learning dysfunction, inattention	All of the above plus probable assignment to special classes
70+ dB	Profound hearing loss	Sensorineural or mixed losses	No speech or other sounds	Severe speech problems, language problems retardation, learning dysfunction,	All of the above, plus possible cochlear implant and long-term educational support

Table 1

- State Agencies are helping by sponsoring initiating and conducting newborn hearing screening and early intervention programs. Studies suggest that such programs can identify up to 95% of infants who are born with various degrees of hearing impairments.

During the formative years, the focus in shifted on infant hearing screening and on the risk factors for deafness. Presently two types of neonatal hearing screening are practiced:

- "Universal Neonatal Hearing Screening" program. In this method, all live births are screened for hearing impairment within 48 hours of birth. This has 100% specificity and no case of congenital hearing loss is missed.
- "High Risk Neonatal Hearing Screening" program. In this process only babies born out of high risk pregnancies are screened for deafness.

Fortunately, the implementation of newborn hearing screening has resulted in the fact that 97% of newborn in European countries receive simple hearing screening procedures to help identify the presence of hearing loss prior to discharge from their birthing hospital. In developing countries like India the challenge of a very large population and a high annual birth rate approaching 25 babies per 1000 makes neonatal screening a difficult task. Moreover, 75% of the population lives in rural areas and over 50% of births occur at home and are frequently attended by a trained birth attendant. So more focus is on identifying high risk groups [2].

The joint committee on infant hearing (JCIH, 2019) [3] recommended that all infants at risk for deafness should be identified. Downs and silver 1972 turned the five risk factors into a widely used and easy- to- remember acronym known as the "ABCD'S of the high risk register for deafness".

- Affected family (excessive intake of alcohol or ototoxic drugs by the mother during pregnancy/Family history of permanent marked sensorineural hearing loss).
- Bilirubin level (Any illness requiring hospitalization for 48 hours or more in the first 4 weeks of birth).
- Congenital rubella syndrome(History of in utero infections such as rubella, CMV, herpes, toxoplasmosis, syphilis).
- Defects of the ear, nose, and throat(Any recognizable syndrome at birth where hearing loss is a known component of the syndrome like down's syndrome, treacher-collin's syndrome etc.).
- Small birth weight (Birth weight of a baby below 1500 grams).

Protocol for neonatal hearing screening

In 2006, India launched the National Programme for Prevention and Control of Deafness. This programme is currently running in over 60 districts of the country and its aim is to identify babies with bilateral severe-profound hearing losses by 6 months of age and initiate rehabilitation by 9 months of age. Under this programme, the following two-part protocol for infant hearing screening is being implemented:

- Institution-based screening - to screen every baby born in a hospital or admitted there soon after birth using OAE. Those who fail the test are re-tested after 1 month. Those who fail the second screening are referred for ABR testing at the tertiary-level centers.
- Community-based screening - to screen babies who are not born in hospitals. Such screening is carried out using a brief questionnaire and behavioral testing. The screening is performed when the baby attends for immunization at 6 weeks of age and onwards. A trained health care worker at the sub centre administers immunization and conducts the hearing screening. The protocol is repeated at every immunization. Any baby failing the Screening is referred for formal OAE screening to the district hospital, and if they fail OAE they are then sent for ABR testing. Once an individual is identified as hearing impaired, they are referred for hearing aid fitting and for suitable therapy.

Evoked otoacoustic emission test (EOAE) should be performed within 48 hours of birth. If the child passes the EOAE test, the child is considered to have normal hearing at birth, however, even if the child have passed the EOAE test, a behavioral observation audiometry like free-field/visual response audiometry is suggested at 8, 12, 24 and 36 months in high risk cases [2,3].

If the child fails the EOAE test, the child is retested 7 to 10 days later. However if the baby fails in the second test, then a more precise and more accurate evaluation of the hearing threshold level is done by "BERA" (Brainstem evoked response audiometry). If BERA test shows normal or near normal hearing, then middle ear pathology is suspected, and in these cases.

Figure 1: Otoacoustic Emission Test (OAE) and Brainstem Evoked Response Audiometry Test.

Tympanometry test must be done to rule out any middle ear pathology like collection of fluid in the middle ear space, congenital ossicular fixation. If any middle ear pathology is found then the condition is treated by surgical/medical means as early as possible. If surgical correction will not be possible at that age, then a hearing aid is prescribed immediately. If those cases where, the EOAE is absent and the BERA shows evidence of deafness, but tympanometry test show normal findings, hearing aid should be provided immediately before the child is 6 months of age. Such a child should be followed up at monthly intervals by speech therapist. If in spite of using hearing aids for at least 6 months child is not hearing at all i.e. not responding to sounds, then the child should be considered for cochlear implantation preferably between the ages of 1-2 years, the earlier the better. After cochlear implantation, auditory verbal training and speech therapy will have to be pursued.

The multidisciplinary team

The ongoing evaluation and assessment of an infant/neonate identified with hearing loss should be performed by a team of

professionals working in conjunction with the parent/caregiver. A full team including the otolaryngologist, audiologist, neonatologist, pediatrician, speech language pathologist, and a wide array of nurses involved in infant care The multidisciplinary team must include the parent.

All the health care providers from primary health care centre to tertiary centre should manage and generate appropriate referrals. They should closely coordinate with hearing care team members.

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