Original Article

Newborn Hearing Screening: Present Scenario

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ABSTRACT

In 2009 many countries of the world met to discuss newborn and infant hearing screening current issues and guiding principles for action under World Health Organization (WHO) banner, though most of the countries who had begun this work as universal program or high risk screen do not have exact data and protocol. The developing countries also decided to become part of it and common guideline was proposed. India being part of it included hearing screening as one of the 30 diseases to be screened under Rashtriya Bal Swasthya Karyakram (RBSK). This article discusses all these issues of newborn hearing screening in the world and India.

Keywords: Hearing screening, neonatal hearing screening, Rastriya bal swasthya karyakram 2013, TEOAE

Current Approaches to Newborn and Infant Hearing Screening in the World

As per World Health Organization (WHO) in its report of new born and infant hearing screening current issues and guiding principles for action November 2009, various countries have started newborn screening for hearing in one way or other.[1] Though the exact data of the fact is not available with most of them. In some countries, newborn and infant hearing screening has become a widespread tool for the early detection of hearing impairment, whereas in other countries such screening is considered to be too costly and its value is questioned. Even when it is available, there is no consistent approach to newborn and infant hearing screening, and there is often great variation within individual countries. The reasons for this are not always financial—some wealthy countries have fragmented and ineffective programs while a number of less-wealthy countries have very successful programs. In countries where newborn hearing screening is conducted it is assumed that the vast percentage of babies born deaf can be helped and their futures immeasurably improved. However, issues such as quality control, screening methods, follow-up, and cost effectiveness need to be thoroughly discussed and reviewed. Quality assurance issues in particular, are vital to successful newborn and infant hearing screening and related interventions. In some settings it is estimated that the poor training and performance of screeners renders up to 80% of screening useless. As in each country, separate set of people are carrying out these programs.

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Not only the exact data is unavailable in most of countries, the minimal level of hearing loss being taken in account and method of testing is different. Members of the International Association of Logopedics (IALP) Audiology Committee recently [1]reported on the activities of their newborn hearing screening programs during 2008.[1] The data presented was either for the whole country (Australia; Brazil; China; Germany; Philippines; Serbia; and Sweden) or for specific regions (India: Maharashtra, and Mumbai; the United States: Colorado, and Washington DC). The detection threshold targeted by newborn hearing screening ranges from 20 dB HL (Brazil) to 40 dB HL (India) and is performed bilaterally in all replying countries. The screening methods used in all replying countries are transient evoked otoacoustic emissions (TEOAE) testing and automated auditory brainstem response (AABR), with distortion product otoacoustic emissions (DPOAE) testing also used in some countries. Most countries use AABR in neonatal intensive care units (NICUs) or for babies at risk of early infant hearing loss. The protocols used in the first stage

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are TEOAE alone (Brazil, India, and Serbia); TEOAE/DPOAE (China); TEOAE/AABR (some regions in Germany, Sweden, and the United States); or AABR alone (some regions in Germany). In the second stage, TEOAE alone is used in India and Serbia; TEOAE/AABR in Sweden and the United States; and AABR alone in Germany. In India, AABR is also used for a third stage. The first and second stages of screening are performed mostly in hospitals (China, Germany, India, Serbia, and the United States), while the third stage screenings are performed in hospitals (India), in Hearing Health Care Services (Brazil), or in pediatric-audiological or ENT departments and practices (Germany).

In some countries, screening is performed on a national basis, and is either non-compulsory (China and the United States) or mandatory (Germany and the Philippines; pending in Australia). In other countries, screening is performed at the district or other sub-national level (Brazil, India, and Serbia). Screening is performed by nurses (China, Germany, Serbia, and the United States); by audiologists/technicians (Brazil, China, India, and the United States); by midwives (Germany); and by physicians (Germany and Serbia). Financing for the newborn hearing screening program comes from parents (China, and partially in Brazil and the United States); health insurance (Germany, and partially in Brazil and the United States); the government (Brazil for public hospitals, India and Serbia); or hospitals (partially in the United States).

The reported prevalence of permanent hearing loss identified by newborn hearing screening programs were: ~ 1/1000 (Brazil, bilateral and Sweden); 1-3/1000 (China, bilateral) and ~ 5/1000 (China, unilateral); 1.6/1000 (Germany, bilateral) and 0.7/1000 (Germany, unilateral); 1.61/1000 of at-risk infants (India, bilateral); 1/1000 (Serbia, bilateral) and 0.3/1000 (Serbia, unilateral); 1.05/1000 (United States, Colorado, bilateral) and 0.45/1000 (United States, Colorado, unilateral); 1.83/1000 (United States[1] Washington DC); and 3/1000 (Philippines).[1]

All countries reported that physiological hearing screening methods were preferred over screening based on questionnaires or behavioral methods. The reported prevalence from all member countries justify universal newborn hearing screening, and even developing countries are highly interested in such programs. Earliest screening by 1month, complete diagnostic evaluation by 3months of age and interventions by 6months of age.

Approximately 50% of all cases of congenital hearing loss are attributable to environmental factors such as congenital hyper-bilirubinemia, ototoxic medication exposure, neonatal hypoxia, viral infections, and meningitis. The other 50% of cases are thought to be inherited, that is of genetic causes. Of these hereditary cases, approximately 30% are classified as syndromic. About 400 named syndromes are associated with hearing loss, the associated auditory features being quite variable—sensorineural or conductive, unilateral or bilateral, and progressive and stable. This small subset 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. The other 70% of hereditary cases are classified as nonsyndromic. This group is the otherwise perfectly normal child with the exception of hearing loss.

The inheritance patterns of nonsyndromic genetic deafness are autosomal recessive in 75%, autosomal dominant in 22% and X-linked in 3% of cases. The associated 'DeaFNess' genes are designated as DFN A (for autosomal dominate gene), DFN B (for autosomal recessive gene) and DFN (for X-linked gene). To date, more than 50 deafness gene has been identified and genetically sequenced, out of which more than half are identified from syndromic form of hereditary deafness. It is likely that hundreds of genes still awaits discovery. As a general rule cases with autosomal recessive inheritance are typically born with bilateral, profound deafness to normal hearing parents. Those with autosomal dominant inheritance have a variable pattern of severity and progression and more often have hearing impaired parents. Interestingly, most genetic acquired hearing losses are caused by single gene defects and no traceable family history is apparent. The most common cause of nonsyndromic deafness is *Connexin 26,* and in India the cause still needs to be worked out. This *Connexin 26* protein, a gap junction protein, is present throughout the inner ear and is important in K^+ concentration regulation.[2,3] The absence of K^+ circulation is responsible for the hair cell's inability to generate action potential in response to sound.

Indian Context

In our country an estimated 5.82 persons have congenital hearing losses per lakh of population at one point of time;[4] two deaf babies are born per hour which amounts to 1/2000 to 1/0000 live births,[4] 18000 deaf babies are added to our population every year, 5% of India population have speech and hearing problem due to congenital sensorineural hearing (SNHL) with delayed development of speech and language (DOSL). The incidence of the later is 0.9/1000 in ENT OPD cases.[4] Thus, at least 10,000 of genetically deaf children are added to our population.

M. V. V. Reddy *et al*. 2004, conducted Interview based prospective study in children below 14 years of age with

hearing loss which showed the results on the type of the hearing impairment are presented in their study; Out of 743 children with hearing loss 18.57% were found with syndromic hearing impairment and 81.73% constituted for only isolated (nonsyndromic) deafness. The results on etiology of hearing loss in children with deafness shows that in 15.22% of children, deafness was inherited, in 13.77% it was acquired, and in 71.01% the etiology was unknown.[5]

Rajiv Dhawan *et al*. 2006 conducted comparative study to evaluate TEOAE as screening modality for hearing impairment in neonates. Brainstem Evoked Response Audiometry (BERA) was used as gold standard diagnostic tool in this study. The factors affecting the specificity of TEOAE were also studied. They concluded that TEOAE is a simple and rapid test with relatively higher acceptability. But, the low sensitivity and specificity are the main shortcomings that take away from TEOAE, the status of independent screening modality for hearing impairment in neonates. TEOAE cannot completely replace BERA as screening modality for hearing impairment in neonates, however can complement it.[6]

Sharing his 10years experience of deaf mute children Dr. Mangal Singh *et al*. has mentioned non genetic causes as 33% of his total patients as the etiological agents, genetic causes responsible for 15.8% and remaining as idiopathic.[7]

Etiological factors for deafness

- Non-genetic causes 33.3%
  1. Embryopathies
     a. Infection
     b. Toxaemia of pregnancy
     c. First trimester bleeding
     d. Ototoxic drugs
     e. Jaundice
     f. Rh incompatibility
  2. Perinatal causes (10.8%)
     a. Low Apgar score
     b. Low birth weight (<2.5 kg) or prematurity
     c. Breech presentation
     d. Post-term
  3. Post-natal causes (12.5%)
     a. Eruptive fever
     b. Meningitis
     c. Hyperbilirubinemia
     d. Traumatic
     e. Cerebral palsy
     f. Delayed mi1estones
- Genetic causes (15.8%)
  1. Family history (10.8%)
     a. Paternal
     b. Maternal
     c. Siblings
  2. Congenital syndromes (5.4%)
- Idiopathic (50.6%)

The NSS 58th round also enquired about probable causes of hearing loss in India. Nearly 1% of hearing disabled persons were reported to have German measles or Rubella as the cause of their hearing disability.

Rashtriya Bal Swasthya Karyakram (RBSK)

This very year in February 2013 Rashtriya Bal Swasthya Karyakram (RBSK)[8] is a new initiative aimed at screening over 27 crore children from 0-18 years for 4 Ds: Defects at birth, Diseases, Deficiencies and Development Delays including Disabilities, has been launched by ministry of health and family welfare. Children diagnosed with illnesses shall receive follow up including surgeries at tertiary level, free of cost under National Rural Health Mission (NRHM).

This programme Child Health Screening and Early Intervention Services under NRHM plans to cover 30 identified health conditions for early detection and free treatment and management.

Identified Health Conditions for Child Health Screening and Early Intervention Services

Defects at birth

1. Neural tube defect
2. Down's syndrome
3. Cleft lip and palate/Cleft palate alone
4. Talipes (club foot)
5. Developmental dysplasia of the hip
6. Congenital cataract
7. Congenital deafness
8. Congenital heart diseases
9. Retinopathy of prematurity

Deficiencies

10. Anaemia especially severe anaemia
11. Vitamin A deficiency (Bitot spot)
12. Vitamin D deficiency (Rickets)
13. Severe acute malnutrition
14. Goiter

Childhood diseases

15. Skin conditions (scabies, fungal infection and, eczema)
16. Otitis media
17. Rheumatic heart disease
18. Reactive airway disease
19. Dental caries
20. Convulsive disorders

Developmental delays and disabilities

21. Vision impairment
22. Hearing impairment
23. Neuro-motor impairment
24. Motor delay
25. *Cognitive delay*
26. *Language delay*
27. *Behaviour disorder (Autism)*
28. Learning disorder
29. Attention deficit hyperactivity disorder
30. Congenital hypothyroidism, sickle cell anaemia, β-thalassemia (Optional).

Though this program includes a lot of causes which may be responsible for SNHL in children but it still misses out genetic causes of hearing loss. This program has strategy of screening by dividing into target groups of which heel prick sample are to be collected for screening of various disorders and study of otoacoustic Emission for hearing screening, but misses out in coordinating for screening of genetic defect in cases of children who are screened to have hearing loss supposing it to be of genetic origin.

Discussion

Most of countries of the world including India has just began this work in one form or other without having proper data for the same in some form or other, realizing hearing loss to be most common sensory deficit. Still the RBSK has not been allocated any budget, surgical treatment has been included in the program, having very high cost. Indian cochlear implant still to be developed completely, which promises low cost remedy. India which will be most populous country of the world by 2030 will have the highest load of these patients, atleast 23000 being added to the population each year as congenitally deaf. What outcomes do we expect?

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