Acta Scientific Otolaryngology

Volume 3 Issue 6 June 2021

Research Article

New Born Hearing Screening-An Essential Tool in Early Identification of Deafness in Children

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Received: May 12, 2021

Published: May 21, 2021

Abstract

Background: Hearing impairment is one of the most critical sensory impairments. Universal Newborn Hearing Screening (UNHS) is a standard practice in most developed countries. However, in developing countries like INDIA many obstacles like unawareness, inaccessibility, lack of infrastructure reduce the sustainable rate of screening for early identification. This study aims to review the benefits and results of universal newborn hearing screening, to propose a standard protocol for screening of all newborns irrespective of high-risk infants, hospital/home deliveries.

Materials and Methods: This cross-sectional study conducted by Department of ENT, GMERS Medical College, Gandhinagar, Gujarat along with TARA foundation screened 31460 neonates, infants and children, between 2017 - 2018. All infants were initially screened with OAE, and those who failed were subjected to ABR/BERA.

Results: All children were screened with OAE as 1st stage screening procedure, out of which 4557 (14.48%) showed positive results. At 2nd stage OAE refer children were subjected to ABR, in which 1322 (29.01%) children were found having deafness which was confirmed by audiologist and otorhinolaryngologist. Out of the 1322 diagnosed with deafness, 746 (56.42%) patients had history of NICU admission, 268 (20.27%) patients had history of consanguineous marriage, while 308 (23.31%) patients had no risk factors. Thus highlighting the risk factors in congenital deafness detection, and the need for universal screening over selective high risk screening.

Discussion: Current recommendations are to screen all infants for hearing loss early in life for timely intervention. A two-step screening with OAE and ABR has to be implemented which can reduce the no of missing cases due to lost follow up. A new initiative 'DHWANI on wheels' has been implemented with OAE and ABR carried out by trained audiologists in a mobile van, to tackle the issue of inaccessibility and unawareness in remote areas.

Conclusion: As normal hearing is critical for speech and language development, UNHS is a strategy that enables to identify congenital hearing loss early and provide appropriate intervention as early as possible for optimal benefits.

Keywords: UNHS; OAE; ABR; Neonatal Hearing Screening; Congenital Hearing Loss

Abbreviations

UNHS: Universal Newborn Hearing Screening; OAE: Oto Acoustic Emissions; ABR/BERA: Auditory Brain Response/Brain Evoked Response Audiometry; ASHA: Accredited Social Health Activist; DEIC: District Early Intervention Center; RBSK: Rastriya Bal Swasthya Karyakram

Introduction

Worldwide reporting of hearing loss finds that the prevalence of moderate and severe bilateral hearing deficit (> 40 dB) is 1-3 per 1,000 live births in well baby nursery population [1,2] and 2 - 4 per 100 infants in an high risk/NICU babies [3-7]. These numbers signify that hearing impairment is one of the most common potentially disabling conditions in infancy and most frequent congenital anomalies which usually goes undiagnosed [8-10]. Current recommendation by WHO is to screen all infants for hearing loss preferably at birth and hearing augmentation by 6 months of age [11,12]. The definition of hearing loss and grading of hearing deficit varies in different classification systems. Hearing loss is usually categorized as mild (21 - 40 dB HL), moderate (41 - 70 dB HL), severe (71 - 95 dB HL), and profound (> 95 dB HL). Deafness is the term reserved for profound hearing loss [13]. Hearing loss can be classified as: Conductive, Cochlear (Sensory: defect in the cochlea and Neural: defect in the 8th cranial/auditory nerve), Retro-cochlear (defect at the level of auditory nerve, brainstem auditory pathway or both) and Central (defect in the auditory area in cerebral cortex). Sensorineural hearing loss is most relevant and more commonly due to cochlear causes.

Universal newborn hearing screening (UNHS) is a strategy that enables to identify congenital hearing loss at a very early age. The major objective of UNHS is to identify children with all kinds and degrees of hearing impairment, both unilateral and bilateral hearing loss, and to lower the age at the time of diagnosis for early hearing amplification, to maximize their linguistic competence and literacy development [14,15]. Typically, the UNHS programs are a two-stage approach. First stage: New born babies are screened for hearing loss before discharge from the hospital within first few days of life. This involves one or two-step OAE testing or OAE and AABR in high risk infants. Second stage: Children who fail in-hospital screening test are referred for a repeat testing between 2 and 8 weeks after discharge and are examined by means of OAE followed by AABR. Positive second stage results should be validated by otolaryngologist and audiological consultation, diagnostic ABR testing and other electrophysiological testing performed by the third month of age. Once diagnosed, all infants identified with hearing loss should receive appropriate early intervention by 6 months of age.

In India there are central level national health programs that propose the guidelines and the protocol for screening newborn babies. Screening should ideally be 'universal' i.e. everybody is screened and at a minimum, screening should be 'targeted' i.e. 'high risk' babies are screened, a centralized facility catering to all hospitals in a city is a practical option.

Aim of the Study

The aim of our study is to review the benefits and results of universal newborn hearing screening, to propose a protocol for universal screening all newborns irrespective of high risk infants, hospital or at home deliveries. Selective screening based on high risk criteria fails to detect half of all infants with congenital hearing loss, thus calling for a need to screen all neonates. In our study, we are also writing on how we were able to cover the gap in screening all infants of even out-reach areas with skilled personnel.

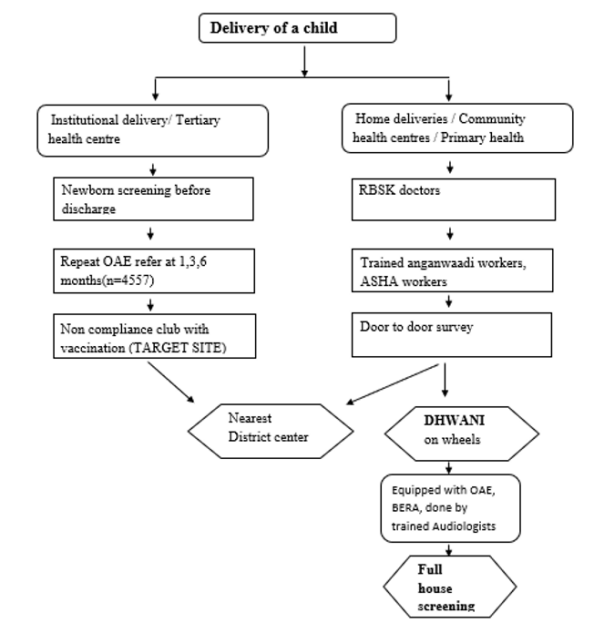
Materials and Methods

A cross sectional study and an observational study conducted by Department of ENT, GMERS Medical College, Gandhinagar, Gujarat, along with the TARA foundation, Ahmedabad. The study covered 14 districts of Gujarat, which screened 49519 infants and children, between 2017 - 2018 (1 year), which included both institutional and at home deliveries, late onset childhood hearing loss.

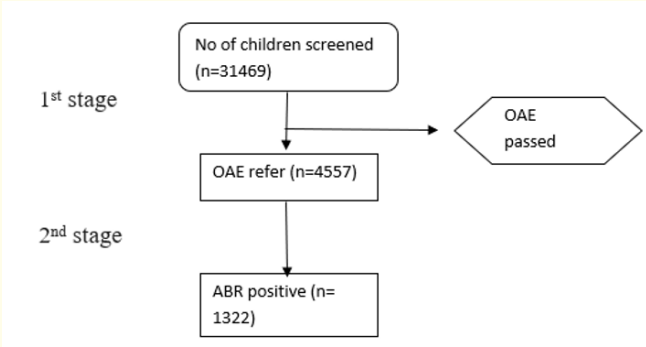
All infants were initially screened with OAE, and those who failed the screening test were subjected to ABR, which was performed by trained audiologist (Flowchart 1). Hearing loss was confirmed by the audiologist and otorhinolaryngologist.

Results

This is an observational study, which covered 14 districts of Gujarat, which screened 31460 infants and children, between 2017 - 2018 (1 year), which included both institutional and at home deliveries, late onset childhood hearing loss. All infants were initially screened with OAE, and those who failed the screening test were subjected to AABR, which was performed by trained audiologist. Demographic details: Out of 31460 infants and children, 17891



Flowchart 1: Protocol of screening.



Flowchart 2: Demographic distribution- results.

(56.86%) were males and 13569 females (43.13%). 18562 children were aged between 0 - 1 year and 12898 children aged between 1 - 2 years.

All children were screened with OAE as 1st stage screening procedure, out of which 4557 (14.48%) showed positive results. As 2nd stage OAE refer children were subjected to AABR, in which 1322 (29.01%) children were found having deafness which was confirmed by audiologist and otorhinolaryngologist (Flowchart 2). Out of the 1322 diagnosed with deafness, 746 (56.42%) patients had history of NICU admission, 268 (20.27%) patients had history of consanguineous marriage, while 308 (23.31%) patients had no risk factors. Thus, highlighting the risk factors in congenital deafness detection, and the need for universal screening over selective high risk screening.

Discussion

Hearing impairment is one of the most critical sensory impairments with significant social and psychological consequences. Failure to detect congenital or acquired hearing loss at an early age may result in lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, emotional and behavioral problems [16]. Neonatal hearing screening is the main mode for early detection of hearing loss. The procedure should be fast and simple and select those most likely to have an alteration in auditory function. Based on the World Health Organization (WHO) screening guidelines [17], successful screening would include the following components: availability of accurate, reliable screening tools; demonstration of earlier diagnosis; consideration for adverse effects of screening; evaluation of the availability and effectiveness of earlier intervention following diagnosis; consideration of the adverse effects of earlier intervention; and evaluation of the longer-term outcomes from earlier diagnosis and intervention.

Under the UNHS, first screening to be conducted before the neonate's discharge from the hospital -- if it 'fails', then it should be repeated after four weeks, or at first immunization visit. If it 'fails' again, then Auditory Brainstem Response (ABR) Audiometry should be conducted. All babies admitted to intensive care unit should be screened via ABR. All babies with abnormal ABR should undergo detailed evaluation, hearing aid fitting and auditory rehabilitation, before six months of age. The goal is to screen newborn babies before one month of age, diagnose hearing loss before three months of age and start intervention before six months of age.

A two-step screening procedure has been implemented in most UNHS programs as a cost-effective and accurate approach. UNHS includes a faster and less expensive OAE as the first screening test

in newborns with no risk factors, followed by ABR in those who do not pass the OAE. ABR is also recommended particularly in infants requiring neonatal intensive unit care as this population is at an increased risk of auditory neuropathy. There is strong evidence indicating that two-step screening is highly effective in identifying infants with hearing loss [18,19]. OAEs can be recorded in 99% of normally hearing ears. The response is generally absent in ears with a hearing loss of 30 dB or greater [20]. Otoacoustic emissions (OAEs) are quicker methods (as compared to electrophysiologic methods like ABR) for assessing hearing in newborns via a simple set-up. ABR test records brainstem electrical activity in response to sounds presented to the infant via earphones. In contrast to the OAE test, ABR evaluates the auditory pathway from the external ear to the level of the brainstem, enabling diagnosis of auditory neuropathy, which is a less common cause of hearing impairment [21].

According to the American academy of Pediatricians 1-3-6 goal is followed in neonatal hearing screening and rehabilitation. Where every neonate is screened at birth or before 1 month of age, confirmed diagnosis by 3 months of age, intervention in the form of cochlear implant or hearing aids done by the age of 6 months at the latest to assist better neural development and speech and language skills [22-24].

In India, National programme for prevention and control of deafness was implemented in 2006 by the ministry of Health and Family welfare. Under which institutional and community based hearing screening was implemented in > 200 districts initially and covers nationwide at present. According to the family and health welfare survey 2015-16, 88.7% are institutional deliveries while the rest are home deliveries. Congenital deafness rate is approximately 5.6-10/1000 live births in Gujarat. Birth rate in Gujarat is 20.1/1000 population according to 2011 census [25]. This is one of the states where a child who are born profoundly deaf can have completely free evaluation, diagnosis, early intervention and then a subsequent rehabilitation. This has been a far sighted, cost intensive and the most cost-effective scheme in the state. However, there is a unique challenge in this country because a lot of children are born at home. Hence, we are now trying to have a screening program in maternity hospitals but also combine it with the vaccination program with oral polio vaccination scheme wherein children who missed their hearing screening can be gathered and evaluated for the same with the help of RBSK, ASHA and Anganwadi workers. Initially an awareness program is conducted for these workers to sensitize them to the issue. They later help in distribution of pamphlets in remote and inaccessible areas as to when to get the child for screening.

The 'Child Health Screening and Early Intervention Services' Program (Rashtriya Bal Swasthya Karyakram) under National Rural Health Mission initiated by the Ministry of Health and Family Welfare of Government of India in 2013, implemented Universal hearing screening of all infants irrespective of high risk/hospital delivery or home deliveries. Congenital deafness was included as one of the conditions for early identification and remediation. It involves screening of infants and children under 18 years by a mobile team and provision of appropriate treatment at District Early Intervention Centres (DEICs). This ambitious scheme is likely to streamline the management of hearing disabilities. It consists of around 1700 RBSK doctors. Under each team there are approximately 42 Anganwadi workers who in-turn train ASHA workers who carry out door to door vaccination and surveys. These surveys include at risk infants like: Family history of hereditary childhood sensorineural hearing impairment, Intrauterine infection (TORCH), Craniofacial anomalies, Birth weight less than 1500 gram, Hyperbilirubinemia at a serum level requiring exchange transfusion, Ototoxic medications used in multiple courses, or in combination with loop diuretics, Bacterial meningitis, APGAR scores 0 - 4 at 1 minute or 0 - 6 at 5 minute, Mechanical ventilation for 5 days or longer, Stigmata of other findings associated with a syndrome known to include sensorineural and/or conductive hearing loss. Over a decade of its inception, institutional screening targeted at risk babies and all newborns at birth and follow up with rescreening, community based screening was carried out by trained ASHA workers or immunization workforce during polio vaccination drive. Though many states like Gujarat, Kerala, Tamil Nadu have taken concrete steps in implementing universal hearing screening, nationwide implementation lacks due to issues like inadequate human resources, lack of infrastructure, low priority, shortage of centers with diagnostic testing facilities and audiologists at district level hospitals, lack of public awareness [26-29].

In our tertiary care center, A new initiative DHWANI on wheels has been implemented (Figure 1), which houses facilities like OAE and BERA carried out by trained audiologists (Figure 2) in a mobile van running from one target site to another for new born hearing

screening purposes to tackle the issue of inaccessibility and unawareness amongst the public in remote areas.

Figure 1: DHWANI on wheels.

Figure 2: OAE being performed by the audiologist.

Universal neonatal screening and not targeted 'high risk' screening is ideal since about 50% of infants with hearing loss have no known risk factors for hearing loss and are discharged from well-baby nursery [30-32]. Delayed onset hearing loss should be considered and followed up. High birth rate associated with the high prevalence of hearing loss combined with frequent exposure to risk factors increases the difficulty of universal screening. Poor health systems, lack of facilities, high rate of home deliveries, lack of awareness, accessibility issues, lack of funding add to the reduced coverage of UNHS.

Factors like lack of awareness among parents/community/physicians, non-compliance by the family for evaluation, stigma attached to hearing aids, accessibility issues, financial constraints are the main hurdles we faced during our screening. Hence, we designed a work protocol to cater to all newborns who missed hospital screening (Flowchart 1), those who developed hearing loss at later days of life, those who cannot reach the tertiary centers for screening, which also included bringing awareness among health center workers and parents about the importance of early screening.

Conclusion

It's not about hearing alone- it's about brain development. As normal hearing is critical for speech and language development, it is recommended that during first 6 months of life, clinicians identify infants with hearing loss, preferably before 3 months of age. Universal Newborn Hearing Screening (UNHS) has become a standard practice in most developed countries. However, in developing countries like India there are many obstacles which we have to overcome in order to achieve a sustainable rate of screening newborns for early identification and required intervention for the betterment of the child. Newborn hearing screening will help to identify hearing loss at an earlier age and alleviate the double tragedy of inability to hear and speak. Early intervention is thus mandatory for best prognostic outcomes. One needs to act like an emergency when hearing loss is suspected - delay means poor outcomes.

Conflicts of Interest

The authors have no conflicts of interests to declare.

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