REVIEW

Newborn hearing screening: Opportunities and challenges

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Abstract  Congenital hearing impairment in infants and children has been linked with lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties. Great emphasis is placed on the importance of early detection, reliable diagnosis, and timely intervention with better chances of hearing impaired infants developing skills equivalent to their peers. This article describes the long journey that the newborn hearing screening process has passed through in the developed countries. It also discusses the requirements, equipment, programs and benefits. It emphasizes promoting newborn hearing screening as a national program with government involvement and elaborates the challenges facing newborn screening in developing countries. The main goal was encourage implementing national newborn screening in developing countries with a discussion of Egypt’s experience.

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1. Historical background

Remarkably, rehabilitation services to deaf subjects are the earliest hearing health services developed, it dates back to the 16th century. It was born as a systematic formal teaching in Spain, then progressed slowly to other European countries & the USA.1,2

In the mid 20th century after the 2nd world war, the diagnostic audiology was introduced.1,2 Since then, hearing services have progressed fast with in depth diagnostic abilities to measure and accurately identify hearing impairment. These measures, in addition to the technological advances of the amplification devices including hearing aids, Assistive Listening Devices (ALDs), Cochlear Implants (CIs) and other implantable devices have great influence on the evolution of rehabilitation services with good outcome in language acquisition and psychosocial development for hearing impaired children.

The overall success of the rehabilitation process however, has been shown to be dependent on the provision of timely and effective diagnostic and intervention services.3,4 Consequently, the value of providing early detection services and the importance of hearing screening has been recently realized.

2. Hearing screening

Screening can be defined as a medical service that aimed at the early detection of a particular condition in a population of those likely to have it. Screening results divide the population into a group with the condition or its antecedents and those without it. Hearing impairment is a good application of the value of screening services.

Congenital hearing impairment has been recognized for decades as a serious disability for affected children. Studies showed that newborns that have significant hearing impairment are estimated to range from 1 to 3 per 1000 live births and are considered as the number one congenital birth defects among neonates5, and it is more than twice as prevalent as other conditions that are screened at birth, such as sickle cell disease, hypothyroidism, phenylketonuria, and galactosaemia.6

In the study by Yoshinago-Itano et al.3, the age of identification was the only significant variable identified to affect the development of language skill. In comparison with hearing peers, children with congenital hearing loss also have low educational attainment with poorer academic outcomes, particularly reading attainment.7 Congenital hearing loss has also been associated with increased behavioral problems, decreased psychosocial well-being, and poor adaptive skills.8 A delay in the diagnosis of 2 years or more being the rule rather than the exception9 with an irreversible delay of speech and language development. In this essence, hearing screening is viewed as an effective means for early identification of hearing impairment.4

Screening services went through great steps. The initiation of screening for hearing defects in USA can largely be attributed to the career-long efforts of the audiologist Marion Downs as long ago as 1964.9 Her efforts paved the way to develop national screening programs in many countries around the world.

At the beginning, screening had been performed in some countries as part of the comprehensive observation scheme by using the behavioral distraction test when the infant was developmentally old enough (6–9 months).10 Many of the hearing impaired children are however, either not detected or if detected, not soon enough. Also, the definition of early identification and intervention has been evolved and now been defined as diagnosis as early as the age of 3 months, with intervention by the age of 6 months rather than 18 months in the recent past.11 This was based on studies that have shown that children identified with hearing impairment prior to 6 months of age have a better chance of developing skills equivalent to their peers by the time they enter kindergarten. Children not identified until later, may ultimately suffer from irreversible and permanent impairments in speech, language, and cognitive abilities when compared to their peers.3,4,7,8,11

Moreover, the discovery of Otoacoustic emissions OAEs has a peculiar impact on hearing screening in newborns.

3. Newborn hearing screen

Newborn hearing screening was initially targeted toward those newborn “at risk” for hearing loss called High-Risk Register [HRR].12 This group included infants who had asphyxia, meningitis, congenital or peri natal infections, anatomic defects or stigmata, hyper bilirubinemia, family history of hearing loss, low birth weight, ototoxic medications, and neonatal illnesses requiring mechanical ventilation.

HRR screening resulted in around 50% of congenital HL being undetected.13 It was soon realized that a more logical approach is to implement universal newborn hearing screening aiming at early identification of most, if not all children with congenital hearing loss.

4. Universal newborn hearing screen

The year 2000 Joint Committee on Infant Hearing [JCIH] position statement has endorsed the early detection of, and intervention for infants with hearing loss through integrated, interdisciplinary state and national systems of universal newborn hearing screening, evaluation, and family centered intervention.14 Universal newborn hearing screening has become the standard of care to provide early detection and intervention for infants with permanent hearing loss.

The expansion of newborn hearing screening in the past decade has helped reduce the average age of identification of infants with permanent childhood hearing loss, to record a mean age of diagnosis was 3–4 months, with a mean age of intervention of 6 months.4,11,15

A recent systematic review by the US Preventative Services Task Force (USPSTF) revealed good-quality evidence of
5. Universal hearing screening programs

Although the need for successful universal newborn hearing screening programs cannot be denied, the legislative support, technology, and expertise needed to implement such programs on a national level has only recently been realized. Screening programs need to be part of the government health service provision in the country concerned to ensure their contribution. There are ongoing needs for staff training provision, maintenance of equipment and establishing links with education, social and other support agencies. Many countries have adopted national programs aimed at universal newborn hearing screening e.g., the United Kingdom, the United States, Canada, Australia and a growing number of other countries. This has been shown to be feasible in developing countries as well as in developed countries.

The aim is identifying all infants born with permanent hearing loss within 4–5 weeks of birth and ensuring the provision of safe, age-appropriate assessments and support for these infants and their families. The core goals are described as “1–3–6” goals.

1. Infants to be screened at 1 month of age.
2. Audiology assessment completed by 3 months of age.
3. Initiation of appropriate medical and audiological, and early Interventional services at 6 months.

Improving technology and experience-based training protocols implemented during the past decade had its greatest impact on the success of these programs in different counties. It is also apparent that implementing a national universal newborn hearing screening mandates the feasibility and efficiency of referral hospitals or centers for appropriate diagnostic services and optimal interventional services.

6. Screening tests and equipment

One important development that has greatly facilitated the implementation of universal newborn hearing screening was the application of objective noninvasive physiological tests that could be administered by nonprofessional personnel.

The ideal screening test would have a high sensitivity and a high specificity. A high sensitivity is particularly important to enable catching up of all infants with significant hearing loss without a delay in the diagnosis of hearing impairment. A high specificity is required, as a false positive result could lead to much workload on the diagnostic services (over referrals) and undue parental anxiety.

Two objective methods were used in most universal hearing-screening programs. They are automated otoacoustic emissions (OAEs) and automated ABR (AABR). They are available as handheld portable equipment with a pass/fail criterion.

Behavioral audiometry, as mentioned before has been shown to be not sufficiently sensitive nor specific for use in a screening program.

6.1. Otoacoustic emissions

OAEs are used to assess cochlear integrity and are physiologic measurements of the response of the outer hair cells to acoustic stimuli. They serve as a fast objective screening test for normal preneural cochlear function through the use of probe in the ear canal.

Currently, two types of evoked OAE measurements are used for newborn hearing screening: transient evoked otoacoustic emissions (TEOAEs) and distortion product otoacoustic emissions (DPOAEs). Provided that the patient’s middle ear function is normal, these measurements can be used to assess cochlear function in the 500-6000 Hz frequency range. The presence of evoked OAE responses indicates hearing sensitivity in the normal to near-normal range.

OAEs are fast, efficient, and frequency-specific measures of peripheral auditory sensitivity. OAE screening has been shown to be a highly cost-effective tool. However, the effectiveness of the test is reduced by contamination with low-frequency ambient noise in a busy nursery, vernix in the ear canal, or any middle ear pathology. Moreover, OAEs are not a sufficient screening tool in infants who are at risk for neural hearing loss (e.g., auditory neuropathy/dyssynchrony) where cochlear function, and therefore OAE measurements, are usually normal.

6.2. Automated auditory brainstem response

AABR is an electro physiologic measurement that is used to assess auditory function from the eighth nerve through the auditory brainstem. AABR measurements are generally obtained by placing disposable surface electrodes high on the forehead.

Most AABR systems compare an infant’s waveform with that of a template developed from normative ABR infant data. A pass or fail response is determined from this comparison. Most commercially available systems can be used as an effective screening tool in infants younger than 6 months. However, AABR needs efficient training and extra costs of disposable supplies.

The new Joint Committee of Infant Hearing [JCIH] 2007 Position Statement recommended separate protocols for neonatal intensive care units (NICUs) and well-baby nurseries. Auditory brainstem response screenings are recommended for all NICU babies, as well as babies admitted for greater than 5 days, so that neural hearing loss (auditory neuropathy/dyssynchrony) will not be missed.

Some centers recommend the use of 2-staged screening OAEs & AABR before hospital discharge. Both AABR and OAE technologies are accepted as reliable measures for newborn hearing screening.

7. Newborn hearing screening in developing countries

The success of newborn screening after its fairly long journey in developed countries has had enormous personal, social, and economic benefits. The consensus is, any infant born with a hearing loss has the chance to experience normal speech and language development as a result of early intervention.

On the other hand, the challenges facing hearing screening in newborns in developing countries are great. Finding the resources to implement solutions for the detection and treatment of newborns is a major problem. Most developing countries
have a high birth rate with heavily dense populations.\textsuperscript{1} Hearing impairment prevalence rate in the newborns is estimated to be higher in developing countries considering the relatively higher rate of exposure to risk factors.\textsuperscript{26–28} The burden of hearing impairment falls disproportionately on the poor, because they are unable to afford the preventive and routine care necessary to avoid hearing loss, or to afford hearing aids to make the condition manageable. Standardizing screening and intervention programs remains an important goal to establish national newborn hearing screening programs in developing countries. Also, it needs to consider the local culture and be acquainted with local resource limitations and strengths.

8. Newborn hearing screening in Egypt

Still, there is no national program for early detection and intervention of hearing impairment but some steps have been taken to promote a national hearing screening program. A field study on screening for hearing impairment in 1500 neonates was conducted\textsuperscript{29} taking the advantage of the implemented metabolic newborn screening for hypothyroidism and the feasibility of providing hearing aids by health insurance to all hearing impaired infants and children. Results have shown great opportunity to implement newborn hearing screening. There was a significant high rate of hearing impairment (9 for 1000 newborns). Also, the Primary Health care Centers [PHCs], with some modifications, can be efficient to provide newborn hearing screening. However, the great challenge was the very limited resources available to provide diagnostic hearing services within the ministry of health hospitals. Since then, implementing and expanding hearing screening services are going in phases and hand in hand with the establishment of audiological services for each sector and governorate together with strengthening of the link to health insurance services to provide the amplification devices. Covering all governorates and implementing comprehensive national newborn hearing screening would be the ultimate target and is expected to become an attainable realistic goal aiming at providing rehabilitation services at optimal time.

National newborn hearing screening remained of high priority to achieve equality in the quality of health and life.

References