Early diagnosis and intervention on neonatal hearing loss

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Abstract

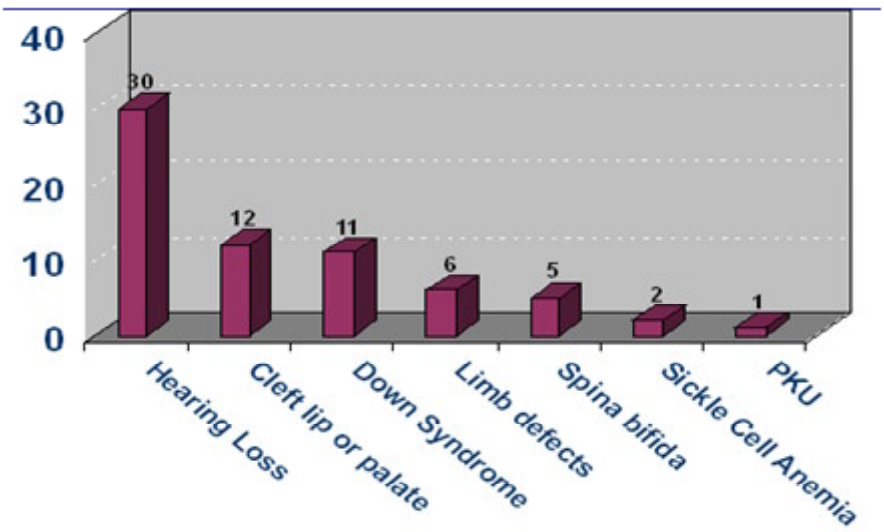
Aim: Congenital hearing loss is the most frequent disorder detected immediately after birth. The implementation of Newborn Hearing Screening (NHS) Programme is well-known in different countries. The purpose of this study was to identify and report experiences of different countries with the NHS programmes.
Methods: Data collection and analysis of information gathered by searching important professional websites, different journals, and research papers related to NHS.
Results: We found that programmes related to NHS are successfully implemented in different countries. We also found that there are some pilot studies carried out in Albania.
Conclusion: NHS is an individual human right widely recommended by the World Health Organization. In most developed countries this procedure is mandatory. It enables early detection, treatment and rehabilitation, preventing the consequences of linguistic, educational and psycho-social deafness, guarantying less disability and healthier children.

Keywords: deafness, hearing loss, newborn hearing screening.

Introduction

The ability to listen is essential to understanding the world around us and interacting with people. Congenital hearing loss is considered one of the most prevalent congenital abnormalities in newborns and is more than twice as prevalent as other conditions that are screened for at birth (1).

Figure 1. Incidence of congenital anomalies per 10,000



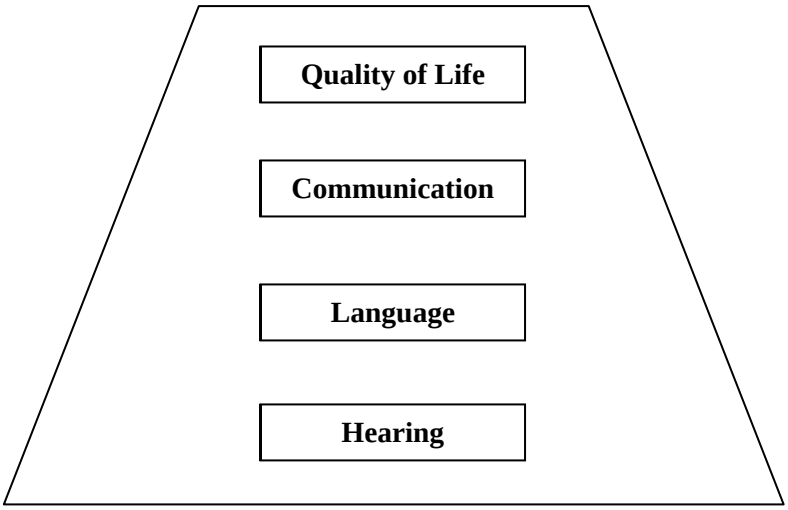
Hearing loss is defined based on the degree of loss, measured in logarithmic decibels. Hearing loss is further categorized based on aetiology (sensor-neural, conductive or mixed), and may be fixed or progressive (2). On average, 1 out of 3 children are born completely deaf, while many others are born with different grades of hearing impairment. Others present late onset of hearing loss during the first years of childhood (3).

Risk factors for hearing loss include: genetic causes (mutations in connexin-26 genes; family history of permanent hearing loss, etc); intrauterine TORCH infections; inappropriate use of antibiotics and other medications with ototoxic side effects; prematurity and low birth weight (less than 1500g); high-level hyperbilirubinemia (Rh incompatibility); 5 days or more spent in a neonatal intensive care unit (NICU) or had complications while in the NICU; craniofacial abnormalities (aural atresia); head trauma; neurological disorder associated with hearing loss; bacterial meningitis, and the like (4). Yoshinaga-Itano states that there is a critical period for early language development within the first six months of life. This sensitive period derives from behavioural and neurological factors that can affect timelines for aspects of development, such as socio-emotional, auditory, speech, and language. Permanent childhood hearing impairment in children affects the development of auditory speech perception, speech production, and language acquisition. Moreover, this disorder affects not only the individual, but also his family's quality of life, as well as society in general because of the high financial costs involved and a possible economic loss for the country.

The Joint Committee for Infant Hearing (JCIH) and WHO recommend that the earlier in life hearing loss is detected and treated, the higher the likelihood of the affected children to conduct a normal life and prevent the impairment of speech, language, and social development.

Early detection is actually defined as diagnosis in the first 3 months of age, as well as treatment and rehabilitation in the first six months of age (5).

Figure 2. Developmental stages



Hearing loss disorders are very difficult to be identified clinically or by means of communication during infant age. Many countries have already implemented screening, early hearing detection and intervention programmes known as EHDI. These programmes are based on several simple screening tests and aim at decreasing age of diagnosis and early treatment, in order to reduce the chance of permanent hearing loss.

According to WHO, around 5% of world population or approximately 350 million people suffer from hearing loss. Based on studies performed in many countries, from 0.5 to 5 newborns per 1000 (‰) suffer from congenital hearing loss or early sensor-neural hearing loss (6).

Screening programmes for neonatal hearing loss

Screening programmes for neonatal hearing loss can be universal or selective. One of the most recommended screening protocols is the Universal Neonatal Hearing Screening (UNHS). Under this protocol, all newborns are tested and neonatal disorders of moderate or severe grade may be diagnosed (bilateral hearing loss, equal to or more than 40 dB HTL and between 0.5 to 4 kHz). A great number of studies show that screening efforts focusing only at high risk infants could be misleading, because approximately half of newborns with hearing loss do not present any risk factor (7). In their statements, the JCIH endorsed integrated, interdisciplinary state and national systems of UNHS, evaluation and family-centered intervention. They recommend that all infants should have access to UNHS and be screened before age of 1 month. Infants not passing the screening test should undergo hearing assessment and medical evaluations before age of 3 months. In other words, hearing screening is a quick and cost-effective way to separate people into two groups: a pass group and a fail group. Those who pass hearing screenings are presumed to have no hearing loss. Those who fail are in need of more detailed hearing evaluation by a qualified audiologist. Infants with confirmed hearing loss should receive appropriate intervention before age of 6 months. In addition, all infants with risk indicators should undergo periodic monitoring for 3 consecutive years.

Additionally, the JCIH recommends that all infants with specific risk factors for speech or language impairment (progressive or late onset hearing loss) and auditory neural conduction disorders (auditory neuropathy spectrum disorders (ANSD), should be

followed by surveillance programs for long-term communication development (JCIH, 2007).

UNHS is an effective and successful program in more than 55 countries where it is already part of health care service (6-8). The UNHS is very successful in countries like USA, Brazil, India, Australia, China, Canada, Philippines, Singapore, etc., as well as in 15 countries of European Union and other countries in Europe like United Kingdom, Austria, Belgium, Denmark, Netherlands, Luxembourg, Croatia, Poland, Serbia, in which more than 90% of newborns are covered. There are also many countries with partial coverage: Germany (from 2009 is a mandatory procedure), Italy, Spain, France, Sweden, Switzerland, Ireland, Greece, Turkey, Hungary, Lithuania, Malta, Czech Republic, Cyprus, Romania, Russia, Slovenia, etc. (6-8).

In the USA, there are on average 4 million newborns, of which 98% in hospitals where the NHS (neonatal hearing screening) is a recommended procedure before discharge and actually covers 95% of the newborns. The model of the implemented programme is the EHDI (Early Hearing Detection and Intervention) with all the respective standards and guidelines, according to which all newborns should be screened for hearing loss within the first month of age. The CDC (Centres for Disease Control and Prevention) has established a data reporting system according to which on average 1.8% of newborns in the US fail at the screening test. In states where the EHDI programme has a higher coverage, 2-3 newborns per 1000 are diagnosed with permanent hearing loss (6-8).

In China, 2 million babies are born every year, of which 60,000 with congenital hearing loss. The Ministry of Health has strongly recommended screening for all babies, but has not been able to cover the expenses from the insurance fund. Actually, families pay 7$ and 14$ for the tests of OAE and ABR, respectively (6-8).

In developing countries like Nigeria, this service is offered in health centres where the BCG vaccine is provided, and it is encouraged and supported by non-profit organizations. In a study among schoolchildren in 1995, the prevalence of hearing loss was quite high (14%) (8).

In the UK, the NHS started in 2006 and its coverage is 99.8%. Among 3.5 million newborns since 2008, 5200 were diagnosed with permanent hearing loss. Guidelines are established as in the US and the screening tests used are OAE and ABR (6-8).

In the Balkan region, NHS programs are not mandatory. In these countries there are many hospitals offering NHS Programs, mostly universal NHS, using different protocols and methods, but many others (especially public ones) do not offer such screening procedures for newborns. The reported prevalence of permanent hearing loss identified in Serbia is: 1/1000 (bilateral) and 0.3/1000 (unilateral) (6-8).

In Germany, the NHS started since the 1st of January 2009. The model is similar to that of the US and UK, and data is recorded in the newborn's health card to enable follow-up through the whole health care system. The hospitals must send annual reports and this information is gathered by the national programme (6,9).

Methods

Data collection and analysis of information gathered by searching important professional websites, different journals, and research papers on programs related to NHS (MEDLINE, PubMed, Google Scholar, American Academy of Pediatrics (AAP) site, Joint Committee on Infant Hearing and WHO position statements, research in scientific events, and the websites on experiences of Universal Newborn Hearing Screening).

Screening methods

Screening programs in general, are made possible in the face of appropriate procedures and resources available. Literature demonstrates that

Universal Newborn Hearing Screening is implemented in most developed countries and has dramatically improved outcomes for infants with hearing loss at birth. Early detection with universal infant hearing screening programs has made early intervention a priority, allowing health care professionals to immediately address the child's communication developmental needs. The most common screening tests are: Otoacoustic emission testing (OAE), the auditory brainstem response (ABR), and the automated ABR. Behavioural audiometry is not sensitive or specific to be used in screening programmes (10). All screening tests are simple and painless. The OAE test is fast, efficient, and specific in measuring the peripheral auditory sensitivity. It was developed in 1978 by David Kemp, becoming a simple technological alternative for routine use in maternity hospitals. Two types of OAE are used in neonatal hearing loss screening depending on the type of stimulus used: transient evoked otoacoustic emissions (TEOAEs) and distortion product otoacoustic emissions (DPOAEs). The registration of the transient evoked otoacoustic emissions (TEOAEs) is performed by a microphone that sends the sound and a probe that registers the response (10).

The auditory brainstem response (ABR) evaluates the auditory evoked potential of the cerebral trunk. There are screening programmes which combine different tests in their protocols due to the fact that their screening mechanisms are different. While the OAE test registers sound in the outer ear, the ABR test registers the neurophysiological response coming from the brain as a reaction to the sound (11). This combination of the screening tests (OAE and ABR), leads to the advantage of early detection as well as the cause of it. Combining OAEs and ABR in screening will assure that we are using state-of-the-art strategies, standard of care, and following clinical guidelines for hearing screening (12).

Screening is safe, reliable, and possible

Data regarding the safety of the screening procedure shows that it is totally safe and painless for the newborn. Literature shows that the sensitivity of a screening programme is around 90%, while specificity varies from 95-97%. This means that for every 100 healthy newborns performing the screening, 3-5 newborns will result in negative response (false positive). Also, among deaf newborns, about 10% may initially result healthy (false negative). Later, with increasing in age, the mother or paediatrician may notice a hearing anomaly and refer to a specialist, who confirms the diagnosis with the confirmatory tests (12).

Affordability of screening cost

Referring to the worldwide experience, the cost of neonatal screening tests is significantly lower than the cost of late tests, and especially the cost of linguistic rehabilitation and psychomotor development, without mentioning the cost of a permanent disability (12,13).

Children with hearing loss in Albania

Hearing loss in Albania is generally detected at around three years of age, which is considered a late age. This delay negatively affects the efficiency of logopedic therapy. Usually, school aged children with hearing loss are enrolled in the Institute of Deaf Children in Tirana or in other special education centres. This has a negative impact in the progress of the rehabilitation and social integration.

Anyway, there are pilot studies to evaluate the prevalence of hearing disorders in Albania. The first study was conducted by the ENT department at UHC "Mother Teresa" in cooperation with the University of Padua, Italy. This study included

15,163 school age children of the first elementary in Tirana schools, examined for a period of three consecutive years (2008-2011). All children diagnosed with a hearing loss of 35dB or more were sent for further audiologic examinations to ENT department. It was found that the prevalence of children with severe-profound hypoacusis was 4.4% (14).

During the period 2005-06, ENT specialists from UHC "Mother Teresa" assisted by ENT specialists from the University of Ferrara, Italy, carried out a selective hearing screening amongst 1561 newborns with high risk factors recovered at the intensive care unit in the Maternity hospitals in Tirana (15).

From 2008 to the end of 2011, it was implemented a new project for "Early detection and social integration of children with hearing impairment" (AID 8670). This was the first universal hearing screening programme implemented in three regions with the highest number of births: Tirana, Fieri and Shkodra, which account for about 17,000 births of a total of 35,000 births per year. In total, 47,341 newborns were screened for hearing loss through the OAE test, which comprised 86% of all newborns in these maternity hospitals. At the end of the project, after further examinations and follow-up, 2.3 newborns per 1000 were diagnosed with permanent congenital hearing loss.

If this new approach (UNHS) will be implemented in all maternity centres of the country, based on hearing loss prevalence founded and the fertility rate in Albania (35,000 children per year), will be detected at least 80 deaf children per year. This figure would increase if children with progressive hearing loss (which consist of 30% of the cases diagnosed with moderate or severe hearing loss) are included.

Conclusions

Literature provides evidence that children using hearing devices within 6 months of life may achieve total recovery of hearing and normal development. Use of technological devices (hearing devices and cochlear implants for the severe cases) help to minimize this handicap thus preventing consequences such as: lack of speech, language, understanding skills or mental retardation, resulting in children equal to their peers. Better results are recorded when these children are integrated in normal schools and social life (16,17). Universal Newborn Hearing Screening (UNHS) is an individual right widely recommended by WHO and in most of developed countries is compulsory. It enables early rehabilitation treatment by preventing speech, educational and psychosocial consequences of deafness, which means less disabilities and healthier children.

By raising awareness about these problems, it is easily understandable the specific contribution of early detection and intervention of this disorder and its importance in preventing social isolation in this category of children. It is up to the competent institutions to take into account the needs of these children and enable early detection, treatment and their integration in society.

Multidisciplinary teams of professionals, including audiologists, physicians and nursing staff, are needed to establish the UNHS component of EHDI programs. Hospitals and agencies should also designate a physician to oversee the medical aspects of the latest.

Hospital-based programs should consider screening technology (i.e., OAE or automated ABR testing); validity of the specific screening device; screening protocols, including the timing of screening relative to nursery discharge; availability of qualified screening personnel; suitability of the acoustical and electrical environments; follow-up referral criteria; referral pathways for follow-up; information management; and quality control and improvement. Reporting and communication protocols must be well defined and include the content of reports to physicians and parents,

documentation of results in medical charts and methods for reporting to state registries and national data sets.

In conclusion, early detection of hearing loss, use of hearing aids and early initiation of speech therapy and special education are key to maximising hearing and speaking in children with hearing loss disorders. Determination of the exact cause of hearing loss, establishment of constructive relationship in family, productive cooperation between professionals and involvement of a specialized logopedist in hearing rehabilitation are vital for a promising future for children with hearing loss.

Conflicts of interest: None declared.

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