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Piloting Universal Neonatal Hearing Screening in Albania

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ABSTRACT

Aim: *Universal newborn hearing screening (UNHS), being routinely implemented in developed countries, could detect hearing impairment early in life enabling its treatment and preventing complication. UNHS is not implemented in Albania. In this context our aim was to present the data form aUNHS pilot project conducted for the first time in Albania during 2009-2011.*

Methods: *During 2009-2011 a collaboration between Albanian experts and foreign experts enabled the conduction of a UNHS pilot study in three main cities of Albania. UNHS was realized in two steps: first, Otto-acoustic Emissions (OAE) test screening and those failing were subjected to the Auditory Brainstem Response (ABR) test.*

Results: *A total of 47341 newborns were screened during 2009-2011 study period (86% enrollment rate). The prevalence of bilateral hearing impairment was 2.3 cases per 1000 births per year. No significant relationships between hearing impairment and sociodemographic and economic factors were detected. Genetic, acquired and idiopathic factors accounted for 38%, 30% and 32% of hearing impairment cases, respectively.*

Conclusions: *An early hearing detection and intervention program in Albania is necessary in order to optimize the language, social, and literacy development for children with hearing impairment.*

KEYWORDS : hearing loss, newborns, early intervention, hearing screening,

Introduction

Hearing loss is one of the most common developmental disorders present at birth (1). The prevalence of significant hearing loss varies from 1 to 4 cases per 1000 newborns (2, 3) or even higher (4), depending on the study population.

Hearing loss in young children is difficult to be reliably detected just by observation of their everyday life activities (5). The selective screening misses almost half of the children with significant hearing impairment (6).

Early hearing loss can affect a child's ability to develop communication, language, and social skills especially during early development of the child (7, 8). Evidence shows 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 (7-9).

Several conditions could increase the risk of hearing impairment in newborns, including family history, various infections of the female reproductive tract, bacterial meningitis, trauma of the head, several congenital malformations, recurrent otitis media with effusion, neurodegenerative disorders, etc. (10). Evidence show that screening efforts focusing only at high risk infants could be misleading since about half of newborns with hearing loss are not present any risk factor (11). Therefore, universal newborn screening was introduced in order to enable the early detection of hearing impairment among this group of newborns without any known risk factor as well.

In the world, universal newborn hearing screening (NHS) is becoming a standard of care in an increasing number of hospitals in more than 50 countries (12).In Europe, NHS is being implemented in virtually all countries of the continent (12) whereas in developing countries the NHS started first in India in 1986, followed by Oman and Iran, etc. (13).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) (14).

Early Hearing Detection and Intervention (EHDI) programs are mandatory in about half of European countries and in the remaining countries such programs are run on voluntary basis or as pilot interventions (14).

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 by NHS programmes are: 1/1000 (Serbia, bilateral) and 0.3/1000 (Serbia, unilateral); approximately 1/1000 (Brazil, bilateral; and Sweden); 1–3/1000 (China, bilateral) and approximately 5/1000 (China, unilateral); 1.6/1000 (Germany, bilateral) and 0.7/1000 (Germany, unilateral); 1.61/1000 of at-risk infants (India, bilateral); 1.05/1000 (United States, Colorado, bilateral) and 0.45/1000 (United States, Colorado, unilateral); 1.83/1000 (United States, Washington DC); and 3/1000 (Philippines) (14).

The Joint Committee on Infant Hearing (JCIH) and the World Health Organization (WHO) recommend the starting of EHDI programs (14). Despite this recommendation, in Albania no efforts have been accomplished in this regard because of lack of health budget and appropriate health policies, lack of specialized diagnostic equipment and trained professionals. In our routine pediatric practice we often detect mild and moderate hearing loss in children aged 2 to 3 years old, when they already present with difficulties to develop speech, spoken language and achieve intelligibility skills and when it is too late to be referred to ear, nose and throat (ENT) specialists and audiologists. In the best of cases, these children are equipped with hearing aids to amplify their hearing abilities and followed up in private clinics and some of them were referred to the Educational Institute for Deaf Children. The others, due to stigmatization, have been isolated at home and get no intervention at all. This is why we started to look for collaboration with colleagues in Albania and abroad in order to improve the situation and start early detection of hearing impairment among newborns.

In the framework of the pilot project "Early diagnosis and social integration of deaf children" (AID 8670) supported by the Italian Cooperation (2009 – 2011), the Albanian pediatricians and audiologists collaborated with experts from the Audiology Service of the University of Padua, Modena, and Reggio Emilia, and decided to intervene in different aspects including:

- early diagnose: using for the first time the UNHS approach;
- early interventions offering hearing aids, speech therapy and fol-

low up; and,
- social integration of deaf children from special educational institutions to public schools for normal audio logically children supported by dedicated tutors, and involvement in social and cultural activities.

This paper presents the data of thisUNHS pilot project conducted for the first time in Albania during 2009-2011 with the support of the Italian Cooperation.

Methods

A pilot project of UNHS was supported financially and logistically in Albania during 2009-2011. In this project the Albanian experts collaborated with experts from the Audiology Service of the University of Padua, Modena, and Reggio Emilia, which offered medical guidelines and protocols. It was decided to carry out for the first time a large-scale Universal Newborn Hearing Screening effort to cover the entire population of newborns in selected cities of Albania, because selective screening misses almost half of the babies born with hearing impairment and half of the children born with any degree of hearing impairment don't fall into any risk group (7,15,16).

In Albania there are about 30000 – 35000 births per year. This pilot project covered three main cities of Albania: Tirana, Shkodra and Fier, with a total of around 17000 births or 50% of total births at national level. Following the Guidelines of the JCHI we decided to apply a two-step screening model (Figure 1):

STEP 1: All newborns (whose parents agreed to be part of screening procedures) were screened before leaving the maternity centers with Oto-acoustic Emissions (OAE) test, which checks the bilateral inner ear response to sound. The OAE test is easy and painless.Because these tests don't rely on a person's response behavior, the person being tested can be sound asleep during the test. The test takes a very short time, usually only a few (4-5) minutes. It was applied by well trained nurses at neonatal units. The test was considered PASS when the response was present at least at one ear and considered FAIL/REFER when was missing in both ears. The pediatricians at neonatal services communicated the results and advised parents of infants who showed "no clear response" (FAIL/REFER)on the initial test. A second test of OAE was repeated within the first month after birth.

STEP 2: All children withFAIL/REFER OAE test at both ears and those with risk factors were invited to repeat the OAE test and take the Auditory Brainstem Response (ABR) test within the first month of life. The ABR test that checks the brain's response to sound was applied by anENT pediatrician evaluating the presence of waves from I-V in 30 dB nHL. We decided to use the combination of OAE's and a-ABR, because we know that it provides a significantly reduced referral rate. Both instruments were fully automated to display a "PASS" or "REFER" test outcome. Another group of newborns which were submitted to ABR test was the group of infants recovered in the neonatal intensive care unit (NICU)for more than 5 days, because of the great risk for retro cochlear pathologies. The results of second step will not be part of this study.

After that, the parents of children in whom OAE and ABR could not be detected in both ears, were requested to allow a complete exploration of the auditory status of the children at the Department of Pediatric Audiology,as soon as possible.

Results

After three years of carrying out the program (between March2009 and December 2011) a total number of 47341newborns were screened for hearing impairment following the two-step screening model explained earlier. The enrollment rate in hearing screening was 86% among all newborns. By the end of the study the prevalence was 2.3cases with bilateral hearing impairment per 1000 births/year (Table 1).

We also studied the records of all the screened children. We didn't find significant relationships between mothers' social-demographicfactors such as age, gender, place of origin, level of education, economic situation, religion, etc.), and life style factors (smoking, alcoholconsumption, physical activity, nutrition, etc.).

However, we noted that the most frequent risk factors were as follows: genetic in 38% of cases (family history, syndromes, and craniofacial malformations), acquired in 30% of cases (prematurity, jaundice, hypoxia, congenital infections) and idiopathic in the remaining 32% of cases.

Discussion

This was the first large-scale effort to estimate the prevalence of any type of hearing loss in newborns in Albania, which included all the newborns during 2009-2011 in three major cities of Albania. We found that the prevalence of any hearing loss was 0.30%, 0.23% and 0.21% in 2009, 2010 and 2011, respectively, thus showing a declining trend over time.

Based on prevalence of hearing loss in countries with similar health system level, we expected the prevalence in Albania to be higher than that observed. However, we think that several reasons might be responsible for our findings:

First, about 14% of newborns didn't participate in the screening effort because their parents didn't accept to screen their hearing, mainly because of the low cultural and/or intellectual level, but also because sometimes they left the hospital (by their decision) within 24 hours after admission;

Secondly, some parents didn't return for the re-examination efforts. This hold true not only for the first or second level tests, but also for follow-up and rehabilitation procedures. The reason is the lack of audiology services in all the country (this service is offered only in Tirana), making it difficult for families from other cities to travel to the capital on periodic basis;

Thirdly, we believe that the exceptionally good performance of our national immunization program and the follow-up done in the mother and child healthcenters might haveimproved the overall health status of pregnant mother and their children's;

Moreover, we have found children with HL returning later in time, that is after the pilot study period of time, only when parents noticed their difficulty in communication. Unfortunatelythis is a waste of time in terms of prevention of the irreversible consequences by late stimulation of the auditory system within a sensitive period.

As mentioned earlier, the most frequent risk factors identified were: genetic in 38% of cases, acquired in 30% of cases and idiopathic in 32% of cases. However, it is still difficult to verify the genetic causes of deafness, because of the poor development of this sector in Albania.

Additionally,if the screening effort would cover all the country, we believe that we could have detected higher prevalence of hearing loss, based also on the fact that in the remote areas of Albania the socio-economic and health care level is low and the risk of hearing impairmentcould be higher, as suggested by the social gradient in health, involving an inverse relationship between health and socioeconomic status(17).

Based on the new recommendations of JCIH in 2013 and the guidelines for early hearing detection and intervention (EHDI) programs, it is very important to establish strong early intervention (EI) systems with appropriate expertise to meet the needs of children who are deaf or hard of hearing (D/HH). Based on these guidelines, screening and confirmation that a child is D/HH are largely meaningless without appropriate, individualized, targeted and high-quality intervention. The delivery of EI services is complex and requires individualization to meet the identified needs of the child and family (18).

Despite the difficulties to find the resources needed to implement universal newborn hearing impairment screening, especially in developing countries, evidence shows that its implementation results in considerable personal, social and financial benefits and, therefore, all countries should aim to achieve such a standard of care (19).

Conclusions

In summary, we can conclude that:

- Prevention is always the best policy, even for congenital hearing loss. UNHS, is a right of the individual and is widely recommend-

ed by WHO and is mandatory in most developed countries;

- Neonatal hearing screening using Otto-acoustic emissions is a reliable and easy-to-perform test and this is exactly the right way to fight the effects of deafness. The combination of TEOAE's and a-ABR provides a significantly reduced referral rate;
- As recommended by guidelines of JCIH, all children need to be screened for hearing loss by 1 month, diagnosed by 3 months, and appropriately fitted by 6 months (18);
- UNHS' and EHDI programs allow to start early detection and rehabilitation, preventing the linguistic, educational and psycho-social consequences of hearing loss, therefore preventing disability and promoting healthy children;
- In regions where hospitals are not appropriately equipped to newborn hearing screening, community-based screening can be considered (20). That's why we propose to use our good experience in the immunization of children during the first year of life, where mothers routinely bring their babies to immunization clinics and such centres may provide an opportunity for an effective infant hearing screening with a wide population coverage;
- It's time that authorities start to support the development of an EHDI Program in Albania. The ultimate goal of EHDI is to optimize language, social, and literacy development for children with hearing impairment. A network of multidisciplinary pediatric experts including audiologists, pediatricians, speech-language specialists, and other professionals, in primary and secondary health care level, should be supported with trainings, protocols, referral procedures, and diagnostic and therapeutic equipment to implement EHDI. Also a dedicated secretariat system should be implemented to follow-up each "failed" newborn and remind parents about their follow-up appointments.

Table 1. Results of the UNHS screening in Albania, 2009-2011

Year	Number of newborns tested	Proportion with normal hearing level (%)	Proportion with Fail/ Refer test result (%)	Prevalence of any type of hearing loss (%)
2009	13570	96,2	3,8	0,30
2010	16896	97,8	2,2	0,23
2011	13875	97,0	3,0	0,21

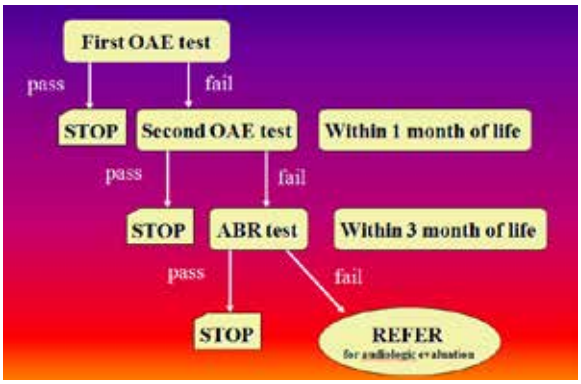

Figure 1. Diagram of UNHS carried out in Albania:

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