

Universal newborn hearing screening: preliminary experience at the University Hospital of Cagliari

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Abstract

Bilateral congenital or acquired sensorineural hearing loss is a pathological condition affecting 1-2 children per 1,000 live births; it represents a major issue in public health because its late identification can negatively affect speech and language development. The aim of hearing screening is to obtain diagnosis and management of hearing loss as soon as possible; in fact early diagnosis and treatment allow children with congenital hearing impairment to acquire adequate linguistic competence. The present study reports our preliminary experience in newborn hearing screening at Neonatology services of University of Cagliari (Italy). During the first semester of surveillance, between January 2012 and June 2012, hearing screening was performed on a total of 901 babies using two different methods, TEOAEs in healthy neonates and automated ABR in high-risk babies. All infants were screened prior to hospital discharge; in some cases, especially for preterm infants of Neonatal Intensive Care Unit and Puericulture Institute, the screening was performed after discharge, to achieve a possible better global and acoustic maturation; 5 cases of hearing impairment were found.

In the present study the Authors confirmed that it is possible to start a universal hearing screening in a relatively short time reaching the percentages suggested by Joint Committee on Infant Hearing.

Keywords

Hearing screening, automated Auditory Brainstem Response, Transient Evoked Otoacoustic Emissions, Neonatal Intensive Care Unit.

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Introduction

Bilateral congenital or acquired sensorineural hearing loss is a pathological condition affecting 1-2 children per 1,000 live births [1-2]. It is defined as a hearing impairment of at least 40 dB hearing level or higher [3-5].

Infant deafness represents a major issue in public health and it has been long recognized that its late identification can negatively affect speech and language development in children and, therefore, it can have adverse effects in school performance and social integration that results in adulthood difficulties [4, 6].

Several studies [4, 6] have demonstrated that early diagnosis and treatment, before 6 months of age, allow children with congenital hearing impairment to acquire adequate linguistic competence.

Unfortunately, the mean age for identification of hearing loss has been reported to be 18 months of age or older, if hearing screening is not routinely performed. According to international guidelines, such as those suggested by Joint Committee on Infant Hearing (JCIH), the hearing of all infants should be screened at no later than 1 month of age [4, 6].

Usually, hearing loss is early detected only in newborns admitted in Neonatal Intensive Care Unit (NICU) and in high-risk children, but studies have shown that only half of neonatal hearing loss is identified in these groups [6].

The evaluation of the hearing status in all newborns is now possible thanks to the modern research in auditory technology, which allows to obtain feasible, cost effective, rapid and convenient instruments for hearing screening.

From April 2009, at the University Hospital of Cagliari (Italy), the hearing screening was started preliminarily by automated Auditory Brainstem Response (aABR) only in the high-risk newborns and NICU children. Since January 2012, it has been extended to all newborns, with the introduction of Transient Evoked Otoacoustic Emissions (TEOAEs). The aim of this study is to report our preliminary experience in newborn hearing screening, during the first semester of surveillance, between January 2012 and June 2012, and to compare our results with those recommended by international guidelines.

Materials and methods

Between January 2012 and June 2012 we performed the universal hearing screening programme on all newborns admitted to Neonatology services

at the University Hospital of Cagliari (Italy). The Neonatology services care includes the NICU, the Puericulture Institute (PI) and the Neonatal Section (NS). The NS is the post-natal ward that accepts healthy infants for at least 48 hours after birth; NICU and PI receive newborns with health problems occurred after or during birth, both from our hospital and other hospitals in Cagliari, suburban area and different parts of Sardinia.

The project employed two main approaches: in NS, we used only TEOAEs whereas in NICU and PI, the aABR was used for newborns with risk factors of hearing loss while TEOAEs was used in all the other cases. The newborns with high risk of deafness were recognized following criteria of Joint Committee on Infant Hearing (**Tab. 1**) [4].

Both TEOAEs and aABR were successfully used for universal hearing screening. These technologies allow non-invasive recordings of auditory system and they are easily performed in neonates and infants [4].

The TEOAEs routinely used is Otoport Lite (Otdynamics®, Hatfield, UK). Measurements are obtained by a probe inserted in external auditory canal that records cochlear responses (from cochlear outer hair cells) to acoustic stimuli, reflecting the status of the peripheral auditory system.

The aABR routinely used is Algo 3i (Natus Medical Inc.®, San Carlo, CA, USA). In this case, measurements are obtained from surface electrodes that record neural activity generated in response to acoustic stimuli delivered via an earphone. These measurements reflect the status of the peripheral

Table 1. Risk factors associated with neonatal sensorineural hearing loss.

1	Family history of permanent childhood hearing loss
2	Neonatal intensive care of more than 5 days, assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide) and hyperbilirubinemia that requires exchange transfusion
3	In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis or postnatal infections associated with sensorineural hearing loss (including bacterial and viral meningitis)
4	Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
5	Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
6	Syndromes associated with hearing loss (neurofibromatosis, osteopetrosis, etc.)

auditory system, the acoustic nerve, and the brainstem auditory pathway.

Both TEOAEs and aABR give two possible responses: *pass* or *refer*. *Pass* means that the result of the screening is good and the baby is not deaf, while *refer* is indicative of a probable hearing impairment that needs further investigations.

The tests were performed in a quiet room by a team including the otorhinolaryngologist, the audiologist and the paediatrician.

All infants were screened prior to hospital discharge; in some cases, especially for preterm infants of NICU and PI, the screening was performed after discharge, in order to achieve a possible better global and acoustic maturation.

Those who didn't pass the first screening (*refer*) were re-evaluated to ENT outpatient clinic for both ears, even if only one ear failed at the initial screening, no later than 1 month of age, with possibility of age correction for preterm.

Those who failed even the second step of the screening were admitted to a comprehensive audiological evaluation, including repeated TEOAEs, tympanometry, stapedial reflexes, Auditory Brainstem Response (ABR). The behavioural audiometry, has been recently introduced in our department and it is now part of the protocol. This is an audiological test based on observing a change in behaviour, such as startling to loud noise and stirring from sleep, in response to sound; an assortment of noisemaker are used and they are classified as low, mid or high frequency sounds.

If hearing loss is confirmed, the child is submitted to further clinical assessments, including computed tomography (CT) scan and/or magnetic resonance (MRI) and genetic consultation. At the same time, an auditory rehabilitation service was offered, including selection and fitting of hearing aid device and speech therapy.

Results

Between January 2012 and June 2012, a total of 647 children were born at the University Hospital of Cagliari (Italy); 511 of them were admitted to NS and 136 were admitted to PI and NICU because of some health problem occurred after or during birth.

In the NS unit, hearing screening was performed using TEOAEs on 498 (97.45%) on a total of 511 children. At first examination 397 *pass* and 101 *refer* (52 bilateral and 49 unilateral) were found; 75 on 101 *refer* passed the second screening (36 of bilateral *refer* and 39 of unilateral); 25 on these 101 did not attend (DNA) to the follow-up (16 of bilateral *refer*

and 9 of the unilateral); 1 on these 101 confirmed the unilateral *refer* after 4 retests (**Tab. 2**).

In the PI and NICU, in the period under evaluation, a total of 403 children (109 from NICU and 294 from PI), including inborn and patients admitted from outside the University Hospital, were evaluated for hearing. In the NICU, counting out deaths, hearing screening was performed using aABR on all 109 children of whom 1 bilateral *refer* was confirmed at retest (**Tab. 3**).

In the PI 294 babies were evaluated for hearing. In this group of patients, aABR was performed on 98 high-risk children; 95 passed the test at first examination; 1 passed the test at the second examination and 2 children have bilateral *refer*, confirmed at retest. TEOAEs was performed on the others 195 children without risk factors for hearing loss; 168 of them passed test at first examination, 20 passed at second examination and 7 DNA.

One case (1/294) of Treacher-Collins Syndrome, with bilateral microtia and atresia of external conduct, was recently diagnosed. Because of malformation it hasn't been possible to perform usual hearing screening and the child will be submitted to bone conduction ABR during the surgical removal of the appendixes (**Tab. 4**).

Table 2. Results of hearing screening in NS.

Screened	498
Pass 1 st test	397
Pass 2 nd test	75
DNA	25
Hearing impaired	1 (unilat.)

Table 3. Results of hearing screening in NICU.

Screened	109
Pass 1 st test	108
Pass 2 nd test	0
DNA	0
Hearing impaired	1 (bilat.)

Table 4. Results of hearing screening in PI.

Screened	98 aABR	195 TEOAEs	1 no screening
Pass 1 st test	95	168	/
Pass 2 nd test	1	20	/
DNA	0	7	/
Hearing impaired	2 bilat.	0	1 Treacher-Collins

Discussion

According to World Health Organization, a health screening is a medical procedure that allows the identification of pathological conditions using an easily available, inexpensive, rapid, reliable test, not invasive or involving pain for the neonate. Usually, the pathology identified is an important health problem that benefits from an early intervention.

Neonatal permanent hearing loss is a relatively more frequent disease than other congenital diseases that are routinely tested in newborns, such as metabolic disorders like PKU or hypothyroidism; therefore, for a long time, it has been recognized the importance of a programme for early detection and intervention in hearing loss [7, 8].

Despite concrete evidence of the benefits of newborn hearing screening, both for families and public health, in Italy, unlike other countries, it is not required but just suggested. For this reason, the implementation of hearing screening programme is an initiative of every single Region and, inside the Region of each Hospital and, consequently, its diffusion and continuity over time, throughout the country, can be considered still inadequate in certain regions.

In our screening programme we used two different instruments: TEOAEs and aABR. In neonatal permanent hearing loss, the sensorineural damage may be located in several locations of the auditory system, including the cochlea, the 8th cranial nerve, the brainstem or the auditory cortex. Because central disorders typically affect children who require care in NICU, it is necessary to screen this group with instruments able to detect auditory neuropathy, like aABR measurement. In the remaining cases, the instrument employed should be TEOAEs.

In the present study, we performed hearing screening in 498 healthy babies born at the University Hospital of Cagliari and admitted in the NS, reporting a covering percentage of 97.45%. Recommended benchmark by international guidelines is 95.5% [4], therefore our results can be considered satisfactory by International Guidelines.

We experienced relatively high rate of *refer* results at the first examination with TEOAEs (101/498, 20.2%). The main reason may be the presence in the external auditory canal of vernix and debris, wax or because of poor probe fit. However, *refer*, checked with retest two or three weeks from birth in the outpatient clinic, was not confirmed

and only one confirmed *refer* (1/498, 0.2%) was observed.

In NICU and PI, the percentage of *refer* at first examination, using TEOAEs and aABR is lower (30/403, 7.4% excluding the patient affected by Treacher-Collins syndrome who was not assessable with these methods because of malformation of the external auditory canal). There are two possible reasons for the aforementioned result: aABR is a test with higher sensibility than TEOAEs, and since the children are usually hospitalized in these wards for a longer time and the test performed after a while, not immediately after birth, there is the possibility that certain maturation of the auditory system occurs and, in addition, the ENT usually perform suction of the external auditory canal.

In those cases with confirmed *refer* test, the child is submitted to a comprehensive audiological and multidisciplinary evaluation in order to confirm hearing impairment and recognize its origin and possible related pathology.

Fundamental steps for the neonatal diagnosis are the ABR, tympanometry and acoustic reflex thresholds. These are audiological objective tests that offer results independently from collaboration of the patient, but they should be performed when the child is sleeping and in a quiet environment. Therefore, it's often necessary to perform the procedure under general anaesthesia or sedation. Ideally, all the assessments, also including CT or MRI performed with the aim to detect anatomical abnormalities, should be achieved during the same general anaesthesia to reduce costs, and to avoid a repeated anaesthesia and delay in diagnosis.

Infant sensorineural hearing loss has two main origins: non-genetic origin, or environmental, and genetic.

The most common environmental factor is congenital Cytomegalovirus (CMV) infection; in all infants in whom hearing impairment was confirmed, CMV infection should be investigated by CMV-DNA urine research [9].

Moreover, a fundamental evaluation for diagnosis is genetic consultation. In fact, epidemiological studies showed that infant hearing loss, in approximately 50% of cases, is due to genetic causes. In one-third it is an aspect of a syndromic disease, such as Usher, Pendred or Alport syndromes, in two-thirds the hearing loss is non syndromic [3, 10]. The genes implicated in non-syndromic hearing loss are 46 with over 100 different mutations. In the majority of cases

the cause is an autosomal recessive mutation on gene of Connexin (GJB2 locus); rarely mutation is autosomal dominant, X-linked or mitochondrial [1, 3, 11]. Therefore, excluding syndromic hearing loss, genetic consultation should include routinely DNA sequencing of GJB2.

In addition, thin-cut CT or MRI of the temporal bones is required to rule out the presence of an inner ear abnormality and to prepare to an eventual cochlear implantation [3].

In our experience 5 suspected hearing impaired children were found, genetic and imaging assessments are still in progress.

When all the investigations are completed and hearing impairment is diagnosed, the child should receive as soon as possible the appropriate hearing aids. In 3/5 patients of the present study the electroacoustic prosthesis have been already fitted. Cochlear implantation should be suggested for any child with severe to profound hearing loss that seems to receive limited benefit from a few months trial (usually three months) with appropriately fitted hearing amplification device associated to appropriate speech therapy. Benefits of hearing aids should be assessed evaluating if speech, language, communication and listening skills are appropriate for the age [12, 13].

When a cochlear implant becomes necessary, the treatment should start between the age of 12 months and 18 months [9, 12, 13].

Regardless of previous hearing-screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development, in order to identify other types of children hearing deafness, such as hearing loss with late onset. This global evaluation, including global development, middle-ear status, hearing and language skills, should be performed periodically or at any time if the professional health care or family have concerns [4].

Patients with the risk factors listed below should undergo audiological evaluation every 6-12 months until 3 years old and then every 12 months until children have 6 years old [4, 9]:

- congenital CMV infection;
- family history of permanent childhood hearing loss;
- syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson;

- neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome;
- culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella), meningitis;
- head trauma, especially basal skull/temporal bone fracture that requires hospitalization;
- chemotherapy or ototoxic drugs.

Conclusions

Late identification of hearing loss is responsible of an important delay in language, cognitive, social and emotional development, that results in a significant public health issue.

In line with guidelines of Joint Committee on Infant Hearing, the first step of the screening should be performed at no later than 1 month of age, both in healthy newborns and high-risk infants. The aim of the hearing screening is to obtain diagnosis and management of hearing loss as soon as possible, at no later of 6 months of age, in order to avoid negative effects in children development.

The protocol that has been implemented in our Department is encouraging since the percentage of screening can be considered above the standard suggested by Joint Committee on Infant Hearing. The DNA patients we experienced can be easily avoided with a better information of the relatives about the importance of further hearing evaluation if the first refer has been found.

Considering the importance of the congenital deafness, we firmly believe that it is necessary to extend the hearing screening program in all the neonatal health facilities in Sardinia, in order to obtain a universal hearing screening, as suggested by international guidelines. The audiological surveillance program should also be performed by primary care paediatricians in order to identify hearing loss with late onset or delayed language development.

Declaration of interest

Authors declare no conflict of interest.

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