

Early audiological diagnostic in newborn and infant

Abstract

Congenital hearing loss is a frequently met congenital condition, invisible, with extremely severe consequences upon child and his family. Late detection of this sensorial handicap, deafness, and absence of an appropriate treatment leads to double handicap - children become deaf and mute. Newborn hearing screening can be performed since day 2 or 3 of life, by automatic methods which do not imply audiology trained personnel. Otoacoustic emission and brainstem auditory evoked response are fast, non-invasive tests which can be performed in baby's natural sleep. The utility of newborn hearing screening is obvious and demonstrated by medical reports. This led to international available specific testing protocols and also an ideal scheme of testing in order to assure hearing aid provider and speech therapy from 6 month of age. In Romania, the National Program of newborn hearing screening started in 2006, at the Institute of Phono-Audiology and Functional Surgery's initiative, but not until no full national cover, law-driven screening and rigorous follow-up of REFER babies was obtained.

Keywords: congenital hearing loss, hearing screening, auditory abilitation, deaf and mute

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Introduction

Permanent congenital bilateral hearing loss has a high incidence in newborns (1,3-3,1/1000)^(1,2). In the European Union the majority of states respects the 2000 Joint Committee on Hearing line, with neonatal hearing screening in the first month, audiologic diagnostic until the age of three month and auditory rehabilitation with hearing aids by the age of six month. Unfortunately, in Romania neonatal hearing screening is just a pilot National Program, which covers just a small part of the country, due to few equipment units and lack of compulsoriness for performing the test in the maternity wards.

Auditory handicap induced by bilateral profound congenital hearing loss is invisible at birth and deaf newborns look and act as normal hearing newborns. For this reason, active detection of the congenital deafness is the only method useful for early detection of congenital hearing impairment. If we rely on family's observation of the infant's auditory behaviour, detection occurs around age of 24-36 month, which is too late in order to have a normal speech development of the child.

Clinical studies in this field already demonstrated the validity of the "auditory deprivation" concept and its irreversible effects on auditory and verbal outcome in congenital deaf children. The length of the period without normal hearing influence speech and language acquisition, even though if the child receives hearing aids. Late detection and intervention in congenital deaf children has severe consequences upon speech, language but also cognitive development⁽³⁾ and long-term social and emotional difficulties.

Besides its negative impact on verbal communication, profound bilateral hearing loss (HL) affects also education, mental health, self-esteem and job opportunities.

Speech and language acquisition is significantly better in children diagnosed with profound bilateral HL by the age of 6 months who received immediately appropriate hearing abilitation (i.e. hearing aids or cochlear implant) compared with infants with delayed detection of hearing impairment^(4,5). About 90% of 5 years old hearing impaired children has permanent hearing loss since neonatal period⁽⁶⁾.

Nowadays we have the technological means (i.e. otoacoustic emissions (OAE), auditory brainstem response (ABR)) for effective, automatic hearing screening in newborns during natural sleep. These audiological methods evaluate different aspects of auditory system functionality: OAE evaluates inner ear function (i.e. the most widely used test for hearing screening) and ABR evaluates auditory pathways from inner ear to brainstem.

Neonatal hearing screening program was introduced in Romania in 2006 as a National Pilot Program for Universal Newborn Hearing Screening (UNHS) and it was designed to have two stages: first one in the maternity wards from Bucharest (two maternity wards), and Timișoara (one maternity ward) and the second one in the audiological centers (very few, unfortunately - Bucharest/Institute of Phono-Audiology and Ear, Nose and Throat (ENT) Functional Surgery and Timișoara), where audiological diagnostic was performed for infants referred from the screening stage.

First stage of UNHS - maternity ward

All newborns has to be screened for hearing loss, regardless presence or absence of hearing loss risk factors since 50% of congenital deaf children has no risk factor for HL.

A targeted hearing screening is not efficient since it would lose 50% of the deaf infants. The algorithm for

this first stage of UNHS is different is risk factors for HL are present or not:

■ **newborns without risk factors for HL** - hearing screening is performed during sleeping in day 2 or 3 of life with OAE test.

- ✓ If OAE are present in both ears (PASS result) parents receive recommendation of monitoring normal language acquisition (200 correctly words and simple sentences at the age of 2 years) and hearing testing before kindergarten and school
- ✓ If OAE are absent at least in one ear (REFER result), infant has to be rescreened one month later either in the maternity ward or in an audiological centre. If he has one REFER result at retesting, in the same session ABR should be performed in both ears and if at least one REFER for ABR is obtained, the infant is referred to an audiological centre for diagnostic.

■ **Newborns with risk factors for HL** are screened in the maternity ward with both methods, OAE and ABR one day before discharge from the hospital. If at least one REFER result is obtained in ABR screening, infant has to be rescreened one month later either in the maternity ward or in an audiological centre with both hearing screening methods. If at least one REFER for ABR is obtained, the infant is referred to an audiological centre for diagnostic.

Second stage of UNHS - audiological centres

All new-borns who have a REFER result from maternity ward hearing screening must be referred to a specialised audiological centre⁽⁶⁾.

The second stage of UNHS National Program includes a battery of objective clinical audiological test (1000Hz tympanometry, acoustic stapedius reflex, OAE, ABR and auditory steady state response (ASSR)) which should be performed either in natural sleep or profound sedation.

No single test is sufficient for hearing loss diagnostic, since false negative results can be present due to specific conditions (ex. middle ear effusion, auditory neuropathy spectrum disorder).

The cross-check principle is the best option in order to establish with high accuracy the presence of hearing loss, type (conductive, sensorineural or mixed) and degree (mild, moderate, severe or profound) of hearing loss, in one or both ears (unilateral or bilateral)⁽⁷⁻¹⁰⁾.

These hearing screening methods are used since 1990^(7,8).

UNHS program based on both automated methods has a very high sensitivity (almost 100%) in detecting hearing impaired new-borns and a high specificity (over 90% - percentage of normal hearing new-borns with PASS results)^(8,11).

Modern equipment have a low rate of false positive results (2%)^(11,12).

For the university centres in Romania where UNHS National program was implemented, mean age of HL detection is between 3 to 6 months, similar with literature results^(8,9).

For targeted hearing screening programs (i.e. dedicated only to hearing impaired new-borns) mean age of hearing loss diagnostic is 24 months⁽⁹⁾.

Following aspects are considered risk factors for hearing impairment⁽¹³⁻¹⁷⁾:

- Gestational age less than 32 weeks and birth weight below 1500g;
- NICU admission for more than 5 days;
- Mechanical assisted ventilation for more than 5 days;
- Pulmonary hypertension and mechanical assisted ventilation for more than 5 days;
- Aminoglycoside antibiotics (Gentamycin, Tobramycin, Vancomycin, Amikacyn) for more than 5 days, without monitoring the serum levels;
- Association of aminoglycoside antibiotics with loop diuretics;
- Severe hyperbilirubinemia with prolonged phototherapy;
- Perinatal severe asphyxia (APGAR score less than 4 at 1 minute and less than 6 at 5 minutes);
- Postnatal infections with viral or bacterial meningitis associated;
- Family history of congenital or first childhood acquired hearing loss;
- Genetic syndromes which includes hearing loss;
- Craniofacial anomalies;
- Intrauterine infections (especially with Toxoplasma, rubella virus, cytomegalic virus, herpetic virus, or treponema pallidum); and
- Alcohol or drug abuse during pregnancy.

Hearing screening and diagnostic has different aims⁽¹⁸⁾

Hearing screening intends to select a small number of infants at highest risk for hearing loss among general population (REFER results mean suspicion of hearing impairment)

Hearing loss diagnostic confirms the hearing loss and quantifies it.

It is very well established now that UNHS is feasible, justified and brings certain benefits as well for hearing impairment children as for their families and society as well. Some things should be mentioned:

- Ethical and equity principles require a UNHS to cover all Romania as soon as possible;
- Early detection of hearing loss is a must and early intervention (hearing aids or cochlear implants and specialised speech therapy) has to be part of the process of management of the hearing impaired newborns;
- In order to be a success program, UNHS should fulfil some conditions:
 - ✓ to be universal, not a targeted hearing screening program^(9,12);
 - ✓ to be comprehensive in approach - it must include training and supervision of the personnel involved, to ensure a high quality and accuracy of results, to follow-up hearing impaired infants, to include report and monitoring IT systems and to ensure psychological support for parents⁽⁴⁾;

■ UNHS National Program should be integrated in maternity-wards all over Romania and hearing screening should be part of the base-package medical services offered during hospitalisation. By this way a large number of new-borns will be screened at birth. Of course follow-up is mandatory^(6,16);

■ UNHS cannot replace parent's and family doctor's vigilance in continuous monitoring of the auditory behaviour and speech and language development in order to detect as early as possible hearing loss acquired during first childhood. The incidence of hearing impairment is 10 times larger in children age 5 than in new-borns due to acquired hearing loss.

Regarding organising a UNHS National Program, dedicated coordinator and governmental support is mandatory.

Conclusions

A success UNHS program allows hearing screening in the first month of life, early audiological diagnostic of bilateral permanent hearing loss (i.e, by 3 months age) and early hearing rehabilitation by the age of 6 months.

These are the appropriate landmarks of normal auditory and verbal development of a congenital hearing impaired infant. ■

References

1. Bamford J, Davis A. Neonatal hearing screening: a step towards better services for children and families. *British Society of Audiology* 1998, 32, 1-6.
2. Barsky-Firkser L, Sun S. Universal newborn hearing screening: a three-year experience. *Pediatrics* 1997, 99, 6.
3. American Academy of Pediatrics Taskforce on Newborn and Infant Hearing (1999). Newborn and infant hearing loss: Detection and intervention. *Pediatrics*, 103, 2, 527-30.
4. Yoshinaga-Itano C, Sedey A, Coulter D, Mehl A. Language of early-and-later identified children with hearing loss. *Pediatrics* 1998, 102(5), 1161-71.
5. Moller M. Early intervention and language development in children who are deaf and hard of hearing. *Pediatrics* 2000, 106(3), e43.
6. Kuhl P, Williams K. Linguistic experience alters phonetic perception in infants by six months of age. *Science* 1992, 225(5044), 606-8.
7. Davis A, Bamford J, Wilson I, Ramkalawan T, Forshaw M, Wright S. A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment. *Health Technology Assessment* 1997, 1(10), 1-176.
8. Arehart K, Yoshinaga-Itano C, Thompson V, Gabbard S, Stredler-Brown A. State of the States: The status of universal newborn hearing identification and intervention systems in 16 states. *American Journal of Audiology* 1998, 7(2), 101-14.
9. White K, Maxon A. Universal screening for infant hearing impairment: simple, beneficial, and presently justified. *International Journal of Pediatric Otorhinolaryngology* 1995, 32, 201-11.
10. Coplan J. Deafness: Ever heard of it? Delayed recognition of permanent hearing loss. *Pediatrics* 1987, 79, 206-14.
11. Birtles G, Modson F, Lovegrove R, Mutton P, Rosen J, Starte D, Williams K. Early identification of hearing impairment in children in NSW. Sydney: Parent Council For deaf education, 1998.
12. Mehl A, Thompson V. Newborn hearing screening: The great omission. *Pediatrics* 1998, 101, (1), E4.
13. White K, Vohr B, Behrens, T. Universal newborn hearing screening using transient evoked otoacoustic emissions: Results of Rhode Island Hearing Assessment Project. *Seminars in hearing*, 1993, 14, 18-29.
14. Wilson JM, Junger YG. Principles and practice of mass screening for disease [Spanish] *Boletín de la Oficina Sanitaria Panamericana* 1968, 65(4), 281-393.
15. National Institute of Health. Consensus statement: Early identification of hearing impairment in infants and young children. National institute of health Consensus Development Conference Proceedings. Bethesda, MD: National Institute of Health, 1993.
16. Grandori F, Lutman ME. European consensus statement on neonatal hearing screening finalised at the European Consensus Development Conference, 15-16th May, 1998. *International Journal of Pediatric Otorhinolaryngology* 1998, 44(3), 309-10.
17. Joint Committee on Hearing. Year 2000 position statement: principles and guidelines for early hearing detection and intervention programs. *American Journal of Audiology* 2000, 9, 9-29.
18. Pascu A, Georgescu. Early detection of hipoacoustic at newborn and immediate intervention. *Revista Română de Pediatrie* 2004, LIII(4), 411-4.