International Journal of Pediatric Otorhinolaryngology 73 (2009) 607–612

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Newborn hearing screening and strategy for early detection of hearing loss in infants

Janka Jakubíková [a,*], Zuzana Kabátová [b], Gabriela Pavlovčinová [a], Milan Profant [b]

[a] Pediatric Otorhinolaryngology Department of Medical Faculty of Commenius University and Children's University Hospital, Limbova 1, 833 40 Bratislava, Slovak Republic
[b] I. Otorhinolaryngology Department of Medical Faculty of Comenius University and University Hospital, Slovak Cochlear Implant Center, Antolska 12, Bratislava, Slovak Republic

ARTICLE INFO

Article history:
Received 10 November 2008
Received in revised form 12 December 2008
Accepted 12 December 2008
Available online 31 January 2009

Keywords:
UNHS
TEOAE
Tympanometry
ASSR
Etiology of hearing loss

SUMMARY

Objective: More than 80% of permanent hearing losses (HL) in children are congenital. Newborn hearing screening (NHS) is the best method for early detection of suspected hearing loss. If the NHS is not universal more than 30% permanent hearing losses are not identified. There are various methods of NHS: otoacoustic emissions (TEOAE, DPOAE) and automatic auditory brainstem response (AABR). After hearing screening, and when hearing loss is suspected, tympanometry and audiological methods then used for determination of hearing threshold; these include ABR, ASSR or/and behavioral methods. The goal of this study is to evaluate the influence of UNHS on the early detection of hearing loss in children before and after the implementation of obligatory universal newborn hearing screening in Slovakia, and also on the etiologic evaluation of hearing impaired infants identified by screening.

Method: In Slovakia NHS started in 1998 and was provided in ENT departments. From May 1, 2006 UNHS has been mandatory in Slovakia, using two stages TEOAE in all newborn departments in Slovakia (64 newborn departments). In year 2005 — 42% of newborns in Slovakia were screened, in 2006 — 66% newborns and in 2007 — 94, 99% (three small newborn departments do not yet have equipment for OAE screening). For determination of hearing thresholds ASSR are used in two ENT departments and ABR in the other four ENT departments.

Results: Comparing the number of identified cases with bilateral severe permanent HL or deafness before and after UNHS, 22.8% more cases of PHL were identified in the first year of UNHS. Also the average age of diagnosis of PHL was lower. In the year 2007, 94% of newborns were screened. We found 0.947/1000 newborns with bilateral severe PHL (35.9%) more than before UNHS). After audiologic and etiologic assessment of the 76 infants who failed screening, 5 (6.58%) were found to have normal hearing, 16 (22.54%) had unilateral and 55 (77.46%) had bilateral SNHL. A non-syndromic genetic cause was present in 25.45% of cases, syndromic in 9%, perinatal cause (31%), congenital CMV infection in 7.27%, bilateral cochlear anomalies without other abnormality in 1.83% and unknown etiology in 25.45%.

1. Introduction

Early detection of hearing loss (HL) in newborns and infants by newborn hearing screening is necessary for early treatment. Undetected bilateral hearing loss in a child causes delay in speech and cognitive development; normal speech and language, may not develop at all.

More than 80% of hearing losses in children are congenital or acquired in the neonatal period [1,2]. Permanent hearing loss (PHL) can be conductive, due to congenital anomalies of the external and/or middle ear, or sensorineural: sensory (cochlear 80%) and neural which may include auditory neuropathy [3,4].

The first step for early detection of HL in newborns in any country is universal newborn hearing screening program (UNHS). If the newborn hearing screening (NHS) is not universal, more than 30% of cases of permanent hearing loss remain undiscovered [2,5]. Two methods are used in newborn hearing screening: otoacoustic emissions (OAE) and automated auditory brainstem response (AABR) [2,6,7]. In most European countries UNHS uses a two-stage OAE program. In some countries (Czech Republic, Hungary) UNHS is not yet obligatory. In the literature there are some reports about use of two-stage ABR screening, or about using OAE as a first step, followed by AABR [2,6].

The TEOAE test is fast and inexpensive and is appropriate for universal newborn hearing screening; moreover once the hearing loss has been detected, appropriate treatment can take place [1]. This method has a high rate of false positive results due to middle ear pathology; there is also a possibility of false negative results,

* Corresponding author.
E-mail address: jakubikova@dfnsp.sk (J. Jakubíková).

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doi:10.1016/j.ijporl.2008.12.006

such as auditory neuropathy/auditory dys-synchrony (AN/AD) [3,4,8]. AABR screening in newborns can also give false positive results due to an immature CNS [5]. Psarommatis et al. described transient auditory neuropathy in high risk neonates and they recommend repeating ABR at the age of 6 months [9].

If there is a suspicion of hearing loss after the hearing screening, the next step is tympanometry, followed by audiological methods for determination of the hearing threshold: ABR, auditory steady state response (ASSR) or/and behavioral methods [10]. The frequency-specific ASSR technique has a number of advantages over click-evoked ABR. There is very strong relation between behavioral tests (BHTs) and ASSR thresholds (10 dB deviations), but the time delay is approximately 5 months with BHTs [11,12].

In infants the audiologic determination of type of hearing loss, appropriate treatment, and etiologic assessment are very important. The etiology is important not only for determining the risk of deafness within the family, but also knowledge of the etiology allows the anticipation of potential health problems and offers appropriate referral for any therapeutic option.

The goal of this study is to evaluate the influence of UNHS in early detection of hearing loss in infants in Slovakia and on the etiologic evaluation of hearing impaired infants.

2. Newborn hearing screening in the Slovak Republic and incidence of permanent hearing loss

2.1. Material and methods

Newborn hearing screening using TEOAE started in Slovakia in 1998. Initially, NHS was provided in ENT departments using ILO 92. From year 2000 NHS started in newborn departments and in year 2002 all newborns in Bratislava, the capital of Slovakia, were screened. In January 2005 the Ministry of Health provided screening TEOAE equipment to all eight special care baby units in Slovakia. From May 1, 2006 UNHS has been mandatory in Slovakia, using two-stage TEOAE in all 64 newborn departments. The first step of hearing screening is on the third day of life in healthy newborns, using portable screening TEOAE equipment. In high risk newborns hearing screening is performed before discharge from hospital. The second step of TEOAE screening is done 1 month after the first screening. In year 2005 – 42% newborns, in 2006 – 66% newborns and in 2007 – 94% were screened in Slovakia (only three small newborn departments do not yet have equipment for OAE screening).

If there is a suspicion of hearing loss after the TEOAE hearing screening, it is followed by tympanometry as the next step, in order to clarify the status of middle ear. If the middle ear effusion (MEE) persists for more than 3 months, the insertion of ventilation tubes (VTs) is indicated before ASSR/ABR examination.

To estimate the hearing threshold, ASSR is used in two ENT departments and ABR in the other four ENT departments in Slovakia. Infants with bilateral severe PHL and deafness are forwarded to the Slovak Cochlear Implant Center in Bratislava (I. ORL Department). After the hearing loss is confirmed, children are scheduled for early intervention (hearing aids and rehabilitation), followed by behavioral audiologic tests and/or repeat ASSR (Fig. 1).

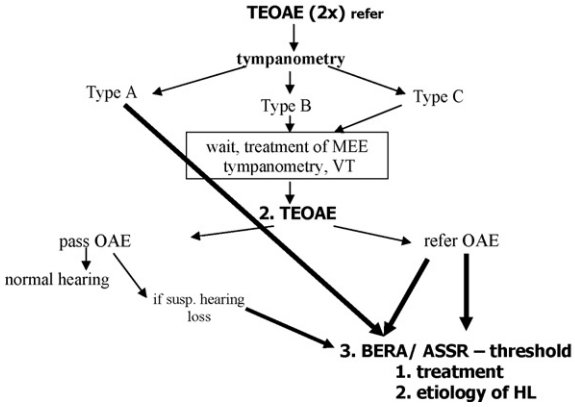

Fig. 1. Steps in diagnosis of hearing loss in newborn.

2.2. Results

The data were collected from the National Statistics Office, from all newborn departments in Slovakia and from six audiologic centers within Otorhinolaryngology Departments.

Comparing the number of cases of bilateral severe permanent HL or deafness before and after UNHS, there was an increase of 22.8 % in identified cases of PHL in the first year of UNHS. Also the average age of diagnosis of PHL was lower. In the year 2007, 94% of newborns were screened. We found less than 1 child of 1000 newborns (0.947/1000) with bilateral profound PHL (35.9% more than before UNHS), however not all infants have a complete audiologic assessment (Table 1).

3. Etiology of permanent hearing loss in infants after UNHS

3.1. Material and methods

A prospective analysis was done of 76 children (born in the years 2006 and 2007), who were referred to Pediatric ENT department and Slovak Cochlear Center after two failures of TEOAE testing. The pattern of tertiary center referral is based on geographical areas according to the children's residence. Most children come from Bratislava and the western region of Slovakia.

In all infants the detailed analysis of congenital hearing loss, medical history and all risk factors were evaluated. Medical evaluation was performed by ENT specialist, with examination of the ear, nose and throat and the clinical examination of the head and face in search of ear anomalies and syndromic features.

The standard methods for audiologic evaluations were TEOAE, tympanometry and auditory steady state response (Audera equipment). In all infants with permanent hearing loss of more than 60 dB, and in those with external ear canal atresia or

Table 1
Identification of bilateral permanent severe hearing loss (PHL) or deafness in newborns, born in the year 2005, 2006 and 2007 (before and after UNHS in Slovakia).

Year of birth	2005	2006 (22.8% more)	2007 (35.9% more)
Birth number in Slovakia	54,430	53,904	54,424
Number of newborns who pass NHS	22,330 (42%)	35,574 (66%)	51,702 (94.99%)
Number of infants with PHL	32 (0.588/1000)	44 (0.816/1000)	49 (0.947/1000)
Average age of diagnosis	12 months	8.8 months	
Minimal age	3 months	2 months	
Maximal age	27 months	15 months	
Number of infants with PHL diagnosed before 6 months of life	4 (12.5%)	21 (47.7%)	29 (59.2%)

Universal Newborn Hearing Screening is obligatory since May 1, 2006.