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Original Research Article

Second stage of Universal Neonatal Hearing Screening – A way for diagnosis and beginning of proper treatment for infants with hearing loss

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ABSTRACT

Purpose: To analyze retrospectively the results of hearing testing in infants at the second stage of the Polish Universal Neonatal Hearing Screening Program carried out in the Department of Otolaryngology at the Medical University of Warsaw.

Material/methods: A total of 351 infants referred to our Department for the second stage of UNHS were included in the study. There were 39.60% infants referred due to positive result of hearing screening at the first stage of the Program performed in neonatal units, 55.27% with negative screening but risk factors present, and 5.13% without any tests due to equipment failure in the maternity unit.

Results: Risk factors were identified in 86.61% of the infants. The most frequent ones were hyperbilirubinemia (71.51%), premature birth (63.25%), and ototoxic medication (62.11%). Otoacoustic emission test showed fail results in 17.66% of the infants, and auditory brainstem responses confirmed hearing loss in 16.81%. Correlation between risk factors and confirmed hearing loss was found for hyperbilirubinemia, low birth weight, intensive therapy for at least 7 days, low Apgar scores, and craniofacial abnormalities.

Conclusions: The early identification of infants with hearing loss is essential for early intervention. Not only infants who fail the initial screening but also the ones with risk factors of hearing impairment should be referred to the centers that are capable of providing the necessary diagnostic services required for the second stage of the UNHSP. Those two steps are needed to both minimize the risk of overlooking a child with hearing loss and properly diagnose hearing impairment.

1. Introduction

High incidence of hearing loss among newborns draws our attention. Current statistics indicate an overall hearing impairment rate of 1–3 in 1000 live births in healthy newborns nursery and 2–4 in 100 in neonatal intensive care units. Hearing impairment in children, undiagnosed or diagnosed with delay, may negatively result in child's language, cognitive, social, emotional, and academic development. The hearing loss itself and its mentioned negative results may significantly affect the child's quality of life. When hearing impairment is diagnosed early and proper intervention is undertaken, children with hearing loss are likely to make great progress, and be more successful in life. Recent advances in technology made the Universal Newborns Hearing Screening (UNHS) feasible and enabled early intervention of congenital hearing impairment. It is one of the greatest advances in medicine of the last century [1–5].

The Joint Committee on Infant Hearing 2000 Position Statement [6] with later update in 2007 [7] and supplement in 2013 [8] includes guidelines for the development of Early Hearing Detection and Intervention programs. These guidelines specify recommendations for the audiologic tests in infants with negative results of hearing screening. They also recommend accurate further interventions depending on the final hearing diagnosis. The audiologic tests include auditory brainstem response (ABR), otoacoustic emission (OAE), impedance audiometry including tympanometry with high frequency probe stimuli, and acoustic reflexes.

Polish Universal Neonatal Hearing Screening Program (UNHSP) started back in 2002. It was initiated by a group of Polish otorhinolaryngologists and neonatologists with a great support

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from The Great Orchestra of Christmas Charity Foundation. From the very beginning, it covered all neonatal units in Poland using transient evoked otoacoustic emission (TEOAE) as a tool for hearing screening. TEOAE is performed in all newborns in their second or third day of life. With years, almost all newborns and infants (96–98.5%) are screened for hearing impairment [9,10]. The UNHS Program was created to meet three main purposes, that is, to detect, diagnose and treat. The first purpose is to detect hearing loss in newborns before they leave the neonatal units and identify neonates with normal hearing but with hearing loss risk factors in their perinatal history. The newborns who do not meet the TEOAE pass criteria and/or have hearing loss risk factors are referred to the second stage of the Program – the diagnostic process. It is at the same time the second purpose of the Program, i.e., to verify positive results from the first stage, establish final diagnosis of hearing loss, and refer patients for treatment. Not only neonates with positive TEOAE results are referred to the second stage, but also those with hearing loss risk factors and those that skip TEOAE first screening regardless of reasons. According to the Program, no child is missed from screening at this point and the possibility of overlooking subjects with hearing loss due to the risk factors in their early life is limited. The Polish UNHSP is carried out very carefully. It prevents the exclusion of children with various degrees of hearing impairment. The third purpose of the Program (third stage) is to provide the child with hearing aids, and/or introduce proper treatment, including surgical treatment with cochlear implants and rehabilitation [9,10].

Before the Polish UNHSP was introduced, there had been an earlier successful attempt to create a coordinated care system for children with hearing impairment [11], but it had its limitations. The problems mainly resulted from the care system of that time, limited availability of diagnostic equipment and insufficient technology. Hearing tests in infants and children were limited mostly to observing the behavioral response to sound, such as a ringing bell, and to conducting behavioral audiometry for hearing thresholds detection in children who were old enough to participate in such tests. The ABR or OAE was not available at that time, but since then a lot has changed in the Polish medical care system and in medicine and technology worldwide. Currently, even the most modern diagnostic equipment is available in Poland. The use of objective auditory measurements including auditory threshold detection in infants and young children enabled early intervention programs such as The Polish UNHSP.

The Polish UNHSP has been proven to achieve its goals and prevent the adverse consequences of delayed diagnosis of hearing loss not only on speech and language, but also on cognitive development [9].

The purpose of this study was to analyze retrospectively the results of hearing testing in infants at the second stage of the Polish UNHSP carried out in the Department of Otolaryngology at the Medical University of Warsaw by means of hearing loss confirming diagnosis and hearing loss risk factors.

2. Material and methods

In this retrospective study, we analyzed data of 351 infants who were referred to our Department for the second stage of the Polish UNHS Program in the years 2007–2011. Our Department is one of those in Poland that meets the criteria of the second stage of the Program – diagnosis of hearing loss in infants referred from the first stage. At the same time, it also covers the third stage goal – treatment with cochlear implantation in children and their further speech and language rehabilitation.

We analyzed the following data: presence of hearing loss risk factors, results of hearing testing at the second stage (confirmation of hearing loss diagnosis), false-positive results from the first stage of the program, and correlation between confirmed hearing loss at the second stage and risk factors.

Data of each subject were collected using a questionnaire, on the presence of hearing loss risk factors, answered by the parents of an infant. Infants were considered to be at risk for hearing loss based on the recommendations of Joint Committee on Infant Hearing (JCIH), 1994 Position Statement [12]. The JCIH in its 1994 statement listed 10 risk factors that identify infants who are at the greatest risk for hearing impairment, in an attempt to optimize infant hearing screening. The risk factors include the following:

1. Family history of hearing loss
2. Craniofacial abnormalities
3. Low birth weight (less than 1500 g)
4. Postnatal asphyxia (low Apgar scores of 0–4 at the 1st minute or 0–6 at the 5th minute)
5. Hyperbilirubinemia more than 15 mg/l (including serum level requiring exchange transfusion)
6. Congenital perinatal TORCH infection
7. Bacterial meningitis or bacteriology-proven sepsis
8. Mechanical ventilation lasting 5 days or longer
9. Ototoxic medication
10. Stigma or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss (syndrome associated with congenital hearing loss).

Additionally, we considered premature birth (<33 Hbd) and neonate intensive therapy for at least 7 days as perinatal risk factors and included them in the questionnaire.

Hearing testing was performed with transient evoked otoacoustic emission and auditory brainstem response. Otoacoustic emissions are routinely conducted in our Department as part of the evaluation of infants failing the first stage of the newborn hearing screening. For this purpose we use Distortion Product Otoacoustic Emission (DPOAE). DPOAE are evoked by two pure tones of slightly different frequencies (F1 and F2) presented to the ear. Five different pairs of tones were used to test different regions of the cochlea covering frequency range from 1500 Hz to 6000 Hz. The pass criterion was set to be at least 6 dB sound-to-noise ratio (SNR).

We routinely use ABR procedure as a part of the second stage hearing evaluation, not only when appropriate responses to DPOAE testing are not obtained but in all cases. The acoustic stimuli used for ABR consisted of broadband clicks with 100 μs duration and with alternating polarity. The clicks were delivered monaurally by a handheld TDH-49 headphone, at a rate of 29.4/s. The analysis time was 20 ms. The electrode impedance was ensured to be below 5 kΩ. The ABR protocol consisted of testing each ear at 70, 60, 50, 40, 30, and 20 dBnHL with 5 dB steps at threshold. Thresholds, the lowest intensity level that evokes an electrophysiologic response, were established to judge the presence or absence of hearing loss. If a response was not observed at 70 dBnHL, testing was performed at 80 and 90 dBnHL. An infant was considered to have passed the ABR test if a replicable wave V response was present at least at 35 dBnHL in both ears. Our facility performs ABR frequency-specific tests, but we do not use it as a part of the diagnostic evaluation of infants failing the newborn screening. We use those tests as immediate follow-up in infants who fail the screening at the second stage in order to establish hearing thresholds for frequencies of 500 Hz, 1000 Hz and clicks (a click is considered to cover the frequency range of 2000–4000 Hz) and start proper treatment. All subjects were tested while they were in natural sleep. Sedation was not used.

Statistical analysis was performed using Statistica software (StatSoft, Inc., 2011, data analysis software system, Version 10, Oklahoma, USA). This is a retrospective study and no free informed consent form for this study was needed; subject's identity was not divulged.