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REVIEW PAPER

Evaluation of hearing screening in newborns in Poland

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ABSTRACT

The Universal Newborn Hearing Screening Program (UNHSP), which has been in operation in Poland since 2002, enables the use of modern, impartial techniques in detecting congenital hearing impairment. It is free and generally available, and so it gains high attendance. The examination uses globally accepted methods of hearing screening: Otoacoustic Emission and Auditory Brainstem Response. The first examination takes place on the obstetric or neonatal ward and allows the examiner to discover any hearing impairment at an early stage. It is important in the context of psychosomatic development of the children. Special attention is paid to newborns with inherited risk factors of hearing impairment. It turns out that < 10% of examined newborns are referred for a check-up in a higher reference centre, and only < 1% of examined children have actual hearing impairment. This review article presents an evaluation of the UNHSP in Poland.

KEY WORDS:

hearing impairment, newborns, hearing screening.

INTRODUCTION

The first attempts of neonatal hearing screening took place in the 1960s and used mostly behavioural techniques [1]. Then, also for the first time, hearing impairment was linked to particular risk factors. Sadly, the early behavioural techniques had their limitations – they enabled the diagnosis of children only older than 8–10 months. Therefore, only infants with known risk factors were tested, which caused undervaluation of the number of children with hearing impairment [1, 2]. In the mid-1970s, attempts to use more modern techniques began. One of those was breathing electroimpedantography, which is the observation of changes in the pattern of breathing in response to acoustic stimuli. It was followed by other methods examining children's movement influenced by acoustic stimuli [1]. A breakthrough was made by the invention of otoacoustic emission (OAE), auditory brainstem response (ABR), and the further downsizing and development of the devices. Nowadays, those 2 methods are the most commonly used worldwide in hearing screening programs due to their simplicity of use, low cost, availability of devices, and easiness of interpretation of results [1, 2].

Otoacoustic emission is widely used in screening due to its simplicity, short time of examination, impartial results, and lack of necessity of the patient's cooperation. This method utilises measurement of silent sounds generated by the outer hair cells located in the inner ear. Those signals are carried further by the auditory ossicles to the tympanic membrane, which strengthens it. The measurement is made by a very sensitive probe placed in the outer ear. Those sounds can be generated spontaneously (SOAE); however, they are present among only 30–45% of people, who can hear properly. Hence, for diagnostic purposes, transiently evoked otoacoustic emission is used [3, 4]. This method uses a tone burst that generates a response in up to 98% of well hearing people [3]. The ABR method uses a single high-level click, which generates the activation of nerve fibres in the auditory nerve and brainstem. This can be observed as a sequence of normally 5 (up to 7) waves, labelled numerically. Each wave

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TABLE 1. Risk factors for congenital hearing loss

Hearing loss risk factors
Using an ototoxic drug in a child for over 7 days or using 2 or more ototoxic drugs at the same time
Family hearing loss
TORCH infections
Craniofacial anomalies
Congenital defect complexes
Prematurity (< 33 HbA)
Low birth weight (< 1500 g)
Apgar < 4 in the 1st minute
Apgar < 6 in the 5th minute
Hyperbilirubinemia requiring blood transfusion
ICU stay > 7 days
Mechanical ventilation > 5 days
Meningitis

ICU – intensive care unit

is generated by another part of the auditory pathway with a precisely defined latency. The evoked potentials are collected by an electrode placed on a newborn's scalp [5, 6].

In Poland the hearing screening program has been running since 2002 thanks to the founding of the Universal Neonatal Hearing Screening Program. This was possible, due to money collected by a foundation called The Great Orchestra of Christmas Charity, during its ninth annual finale in 2001 [1, 7]. Among the Program there are 409 facilities of the first referential level, in which every newborn undergoes an OAE in the second day of life. If the result is incorrect, then a second examination is done on the day of discharge from the hospital. At the same time the risk factors are assessed [7–9]. Every newborn with an incorrect test result or stated risk factor is referred to one of 73 facilities of the second referential level, where an additional ABR is conducted [7]. The basis of the program is to diagnose infants with hearing impairment no later than at 3 months of life, which is essential for accurate speech development. All diagnosed children are referred to one of 25 of the third referential level facilities, where therapeutic, rehabilitative, or observational actions are undertaken. Help should be initiated by the age of 6 months [7–9].

RISK GROUPS, WITH PARTICULAR REFERENCE TO PREMATURE BABIES

Both genetic and environmental factors are responsible for congenital hearing loss. They can coexist or be isolated causes [10]. Currently, it is estimated that 1–3 per 1000 full-term infants have congenital hearing loss [10]. This rate increases 10 fold in premature babies [4].

The most important risk factors for hearing loss are ototoxic drugs, a positive family history of hearing loss, the presence of congenital TORCH infections, and genetically determined syndromes with hearing loss, such as Down's syndrome [10]. It is estimated that together they are responsible for over 50% of the causes of in-depth diagnostics in patients suspected of hearing impairment [10].

Other risk factors include low birth weight, significant hyperbilirubinemia, and intensive care unit treatment for more than 7 days, which are much more common in newborns born below 34 HbA [4]. All risk factors for hearing loss are presented in Table 1.

A retrospective study conducted in 2013 by Wróbel *et al*. shows that among newborns included in the Universal Newborn Hearing Screening Program (UNHSP) in Poland, on average, 71% have an isolated risk factor for hearing loss, 13.5% have 2 factors, and the remaining 15.5% have at least 3 factors [10]. However, studies show that the most common risk factors are not the most common causes of hearing loss. The reasons for this phenomenon are sought in the continuation of the diagnosis of hearing loss in patients with correct results of screening tests due to the existing risk factors. For example, the actual rate of diagnosed hearing loss in infants with craniofacial abnormalities is much higher than, for example, due to ototoxic medications taken by a pregnant mother [10].

The study of Ratyńska *et al*. from 2001 showed that in neonates with genetic factors, TORCH infections and administered ototoxic drugs, TEOAE signal-to-noise ratio (SNR) values were significantly reduced compared to neonates without these factors. On the other hand, the records of the TEOAE study in neonates with hyperbilirubinemia showed an unexpectedly higher SNR compared to the records of newborns without this risk factor [11].

We should also pay attention to the correlation between the number of risk factors and the occurrence of hearing loss – the greater the number of risk factors, the greater the likelihood of hearing loss.

Current data show that genetic factors cause the development of various forms of hearing loss in about 50%, of which 30% are so-called isolated hearing loss. The most important genes responsible for congenital hearing loss are the genes *GJB2*, *GJB*6, *SLC26A4*, *MYO15A*, and *STRC* in an autosomal recessive manner. On the other hand, the *TMC1* and *KCNQ4* genes, which are the rarer causes of hearing loss, are inherited in an autosomal dominant manner. The remaining 20% of inherited hearing loss falls within the spectrum of symptoms of genetic syndromes such as Pendred syndrome, Usher syndrome, Lange-Nielsen syndrome, Wolfram syndrome, and Charcot-Marie-Tooth disease (X-linked inheritance). Any of these syndromes in a child can predispose them to progressive hearing loss, sometimes with a delayed onset [12, 13].

Premature babies are particularly vulnerable to hearing loss. Statistically, congenital hearing loss is 10 times more common in this group. It is a consequence of the coexistence of multiple risk factors resulting from prematurity. The most important are as follows: very low birth

weight, low Apgar score, mechanical ventilation, and craniofacial defects. Greater immaturity of a premature baby leads to a greater number of risk factors; therefore, an inverse relationship between foetal age and the occurrence of hearing loss in premature babies is observed [4].

ATTENDANCE AT FIRST AND SECOND SCREENING

Universal Newborn Hearing Screening is free of charge and covers all children born in Poland. At the end of November 2021 a total of 6,809,197 newborns were examined, counting from the beginning of this program. The average number of children examined annually between 2010 and 2019 is 364,009. The formula of the screening program determines that the greatest number of children undergo examinations in facilities of the first referential level [7]. Both studies, one from 2016 [6] and one covering the years 2003–2013 [14], show that the attendance on first-level screening is 96% on average among live newborns. The same studies show also that about 8.5% of patients are referred for check-up in a facility of the second referential level. Surprisingly, the attendance there according to official data is low. Data covering the years 2003–2016 show that only 55.8% of reported children attend the check-up [14], whereas data from the years 2006–2015 state that the average attendance is 47.9% [8]. A study was conducted on that matter [8]. A telephone survey was conducted among 3239 parents of children referred to the check-up in the facility of the second referential level between 01.06.2014 and 30.11.2014. The study showed that the percentage of children appearing on the check-up was actually much higher, at 83.6%. After analysing the data, the researchers pointed out the reason for this variance. It turned out that it was a reporting error. Newborns with incorrect results of the first examination or those who had not been examined were reported in the system, whereas the information received from the second examination in the neonatal ward were often not reported. This leads to an error, where newborns with correct results are still guided by the system to the check-up. However, some parents, despite incorrect screening results, decide not to attend the check-up with their children. The authors of this study point out that this is due to a lack of social consciousness and insufficient parental education on the topic of children's hearing disorders. Another reason for abandoning the check-up are long queues at the doctor's office, which comes from the fact that children are often referred to the same doctors as adult patients [8].

OUTCOMES OF STUDIES

Przewoźny *et al*. analysed the results of the UNHSP at the Department of Otolaryngology of the Medical University of Gdańsk in 2011 [15]. At that time, 715 children were examined, of whom 453 newborns were referred for further audiological diagnostics (second-level reference centre), which constituted 60.2%. Hearing impairment risk factors were detected in 360 children. Incorrect test results were obtained in 133 children (28.7%).

Ultimately, hearing loss was confirmed in only 33 children (7.3%), of whom 24 (72.7%) had risk factors. The most common were head and neck congenital anomalies, a family history of hearing impairment, and congenital defect syndrome associated with hearing loss.

The study also noted a high (22.1%) rate of false-positive results [15].

Greczka *et a*l. summarized the activity of the UNHSP in Poland in the years 2003–2013. Over a 10-year period, the program included a population of 4,345,326 children. 415 first-degree centres, 70 second-degree centres, and 23 third-degree centres participated in the program. Annually in the above-mentioned years, an average of 96% of the population of live born children in Poland was examined. Children requiring further diagnostics accounted for 8.5% annually. On average 55.8% of them were referred annually to the next stage of the program.

In the analysed population, any hearing impairment was eventually found in 0.3% of the children. After detailed diagnostics, 58.2% of children were referred to prosthetic hearing aids, 34% to surgical treatment, and 7.8% to further rehabilitation [14].

In the work of Greczka *et al*. the results of the UNHSP in Poland in 2006–2015 were analysed. It was estimated that 373,477 children were examined annually. 24,486 of them (6.6%) required further diagnosis. According to the analysis of data from the central database (CDB), only 55.8% of them actually engaged in in-depth research at a second -level reference centre. Therefore, the authors of the study created a questionnaire for parents of unexamined children, which showed that the actual level of further diagnosis was 83.6%, and that the underestimated CDB data resulted from reporting errors. To estimate the above-mentioned errors, data from the program implemented between June and November 2014 were used. The results of the survey also showed that in 2014 as many as 96% of newborns were included in the Universal Screening Program, and 91.78% of them obtained a positive result [8].

In the work of Zych *et al*., the structure and results of UNHSP in Poland in 2016 were analysed. Screening tests were carried out in 496 centres, of which 401 were in the first stage of reference, 69 in the second, and 26 in the third. Up to 22 August 2017, 5,458,114 newborns took part in the program, which is 96.3% of the population born during that period. 91% of them obtained a correct test result and were not referred for further diagnostics. At the second stage, 96.7% of the children obtained a correct test result, and only 3.3% of them were referred to a third-level reference centre. As many as 55% of them required only observation, 25% received hearing aids, 13% were referred for surgical treatment, and 7% underwent rehabilitation [9].

The work of Greczka *et al*. analysed changes in the UNHSP in Poland over a period of 15 years (2012–2017). According to data from the CDB, 5.5 million newborns have already participated in the program, and the percentage of examined children increased from 97.8% in 2004 to 98.4% in 2017. The percentage of children participating in hearing tests at level 2 also increased from 41.4% in 2009 to 83% in 2016. Reasons for the above-mentioned increases are sought to increase the awareness of parents and improve the IT and technical side of the program [16].

COMPARISON WITH DATA FROM ANOTHER COUNTRIES

The Polish Newborn Hearing Screening Program is similar in its grounding to similar programs in other European countries.

Data from the Tuscany region in Italy show many similarities to the Polish program – the OAE is used as the screening method, and the ABR is used in further diagnostics of children with incorrect screening results or those with correct results but from high-risk groups. The percentage of examined children is similar as well – data from 2011 show that the attendance is about 98%. The major difference is that in Tuscan hospitals children with incorrect screening results have mandatory urine tests for the presence of the cytomegalovirus genome [17].

In Germany the Neonatal Hearing Screening has been in operation since 2009. The methods used as well as the basis of the program are the same as those in Poland. The percentage of children in need of further diagnostics after screening in a neonatal ward is rather constant and reaches 4% [18].

Such programs are developed outside the Europeans Union as well. In Israel the Neonatal Hearing Screening Program was established in 2010. During 2014–2016 it allowed 98% of live born newborns to be examined. The basis is the same as those in previously mentioned European countries. Data from Israel show that the percentage of children with incorrect screening results for 2014, 2015, and 2016 are, respectively, 1.8%, 2.5%, and 2.6% [19].

TREATMENT, REHABILITATION, AND FURTHER SCREENING OPPORTUNITIES OF CHILDREN WITH HEARING IMPAIRMENT IN POLAND

Child hearing impairment negatively influences speech, language, developmental, cognitive, and educational outcomes, as well as quality of life [2, 12]. Hence, children with confirmed hearing impairment are under constant medical care. Depending on the test results from the higher referential level facility treatment, rehabilitation or surveillance is offered [9]. The most important thing to focus on is correct psychosomatic development; therefore, speech rehabilitation and psychological consultations are essential [7]. Possible hearing rehabilitation management in children includes hearing aids and cochlear implants [12]. The decision of which method should be proposed to a particular patient is made on the basis of the kind and profundity of hearing loss. It is worth mentioning that the guidelines for cochlear implant have been expanded over time due to studies showing that this method has advantages over use of a hearing aid also in less severe hearing loss [9]. The current indication for cochlear implantation in children is bilateral sensorineural hearing loss > 80 dB HL after rehabilitation with the use of hearing aids that lasts approximately 6 months. Bilateral implantation is currently not a common practice, although it is widely discussed in the world [20].

Even though the Universal Hearing Screening Program covers a very large number of children, some preschool and school children still suffer from hearing impairment. A study performed in 2008 among 95,411 preschool and first-grade children, i.e. children 6 and 7 years old, showed that as much as 13.9% of first-graders had positive hearing screening results. Further examination showed that 3.5% of them had severe hearing loss and 22% suffered from moderate hearing loss. The study also enabled the authors to observe that as much as 58.8% of parents and carers were unaware of their children's problems. Based on the outcomes, the authors suggest that hearing screening in schools should be performed on a regular basis [21].

CONCLUSIONS

The percentage of newborns taking part in the Hearing Screening Program is growing every year. The number of facilities conducting neonatal hearing screening has also grown.

The Neonatal Hearing Screening Program allows faster diagnostics and treatment of hearing impairment, which decreases the risk of psychosomatic development retardation in children.

The Polish Neonatal Hearing Screening Program uses the same techniques and basis as similar programs in other developed countries.

DISCLOSURE

The authors declare no conflict of interest.

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