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Original article

Universal newborn hearing screening: the experience of the University Hospital of Parma

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Summary. *Background and aim:* Early diagnosis of congenital deafness is fundamental to minimize the negative consequences on a child's educational and psychosocial development. To lower the age of hearing-impaired children at the time of diagnosis, universal neonatal hearing screening (UNHS) is considered essential. The aim of this study was to review tha data of the first 4 years of implementation of UNHS in the University Hospital of Parma. *Methods:* 11624 infants born between February 2010 and December 2013 were included into the study. Transient evoked otoacoustic emissions were used as screening test. When the newborn had failed at the initial test, he was re-tested within 3 weeks from birth. If the same result was obtained at the second step, the newborns were referred for clinical auditory brainstem response. We calculated quality indicators and compared them with international guidelines. *Results:* 11592 infants (99.7%) were screened during the birth admission. 10359 (88.5%) were well-babies, while 1233 (11.5%) had audiological risk factors. Among 11592 newborns screened, 42 (3.59‰) had a final diagnosis of sensorineural hearing loss. The incidence of deafness was 1.64‰ in well-babies, and 2.02% in neonates with audiological risk factors. Only 71 infants (0.6%) did not complete the screening program. False-positive rate was 1.7%. *Conclusions:* The analysis of benchmarks and outcomes of UNHS demonstrated the good quality of our hearing screening program. Introduction of automated auditory brainstem response as well as enhanced enrollment of patients who do not complete the screening could further improve the quality program. (www.actabiomedica.it)

Key words: congenital deafness, universal neonatal hearing screening (UNHS)

Introduction

Hearing loss (HL) during the first 3 years of life can hinder speech and language acquisition with significant negative consequences on a child's educational and psychosocial development (1). HL can be classified as conductive or sensorineural in nature; conductive hearing loss (CHL) is related to a disorder of the external and/or middle ear, which impacts on sound transmission toward the inner ear. Depending on the etiology of the CHL, the rehabilitation includes drug therapy, external and/or middle ear surgery, and hearing aids (2-5).

Sensorineural hearing loss (SNHL) affects the cochlea, which transforms sound vibration into a neural signal or, the cochlear nerve, which transmits this signal to the auditory brain. Depending on its severity, SNHL can be rehabilitated by means of prostethic devices, such as traditional or semi-implantable hearing aids, cochlear implants, and auditory brainstem implants (6-8).

Regardless of the rehabilitative method, early hearing deficit detection and precocius intervention are fundamental to maximize linguistic competence and literacy development in deaf children (9). For this reason, universal newborn hearing screening (UNHS) programs have been implemented worldwide with the aim to identify hearing deficits > 40 dB present at birth.

With the Regional Decree n.694 of May 23st 2011, the local regional government of Emilia Romagna approved the guidelines for the implementation of the neonatal audiological screening program in all the birth centers of the region. In our hospital, the Neonatal Unit in collaboration with Otorhinolaryngologic Unit, have already started a UNHS program in 2010. To evaluate UHNS programs, benchmarks and quality indicators have been proposed by the Joint Committee on Infant Hearing (JCIH) (10-11). The aims of this study were to present the results of the UNHS in the University Hospital of Parma and to analyze its evolution during the first 4 years of implementation.

Methods

Study design was based on a retrospective analysis of 4 annual databases (2010-2013) from the UNHS program in the University Hospital of Parma. The UNHS protocol recommends different tests depending on the presence or the absence of risk factors for SNHL. In well babies, transient evoked otoacoustic emissions (TEOAE) are executed with the device Accuscreen Pro-Gn Otometrics in a two-step screening program using a default "PASS-REFER" algorithm. Audiologists or paediatric nurses perform the first step 24-48 hours after birth in silent rooms while the neonates are asleep. In presence of TEOAE (*PASS*) on both ears the screening is considered successfully completed and the neonate is discharged from the program. If TEOAE are absent (*REFER*) in one or both ears the test should be repeated within 3 weeks from birth. If this second step still produce a refer result, infants undergo auditory brainstem response (ABR) test. Neonates with a normal hearing threshold on ABR are discharged from the screening program, while patients with a SNHL undergo audiogical and otorhinolaryngologic evaluation and start investigation on the causes of the hearing impairment. Neonates with risk factors for SNHL (based on the list from the JCIH Position Statement 2007) (11) are screened by means of both TEOAE and ABR. Hearing impairment severity was classified according to the BIAP (Bureau International d'AudioPhonologie) criteria (mild, between 21 and 40 dB; medium, between 41 and 70 dB; severe, between 71 and 90 dB; profound over 91 dB. We calculated quality indicators and compared them with JCIH guidelines. Outcomes measured were: percentage of infants screened before 1 month of corrected age, percentage of infants referred for diagnostic testing, percentage of false positive, percentege of infants lost to the follow-up, and prevalence of SNHL. Due to the retrospective nature of the study and the maintenance of anonymity of all subjects, the consensus of the ethical committee was unnecessary. The study was performed in accordance with the Declaration of Helsinki, under the terms of relevant local legislation.

Results

From February 2010 to December 2013, 11624 neonates were born in the University Hospital of Parma. Among these infants, 11592 (99.7%) have undergone UNHS. Thirty-two newborns have not been tested because parents refused the screening (4 cases), because life-threatening conditions and subsequent transfer in other hospitals (18 cases), because they died (10 cases). There were 10359 (88.5%) well-babies, while 1233 (11.5%) had audiological risk factors, based on JCIH Position Statement (10). Overall, 344 well-babies (3.3%) did not pass the birth admission screen and were admitted to the second step. At the re-testing, 253 (73.5%) resulted PASS, 27 (7.8%) were missed, and 64 (18.7%) were still REFER and underwent audiological evaluation. Overall, the percentage of well-babies referred for audiological and medical evaluation was 0.6%; this percentage was 0.93% in the first 2 years and lowered to 0.28% in the 2 subsequent years. The infants lost at the second step (n=27) were 13 in 2010, 10 in 2011, and 2 in 2012 and 2013. Overall, the percentage of well-babies screened before one month of age was 99.7%. Among the 64 infants

admitted for audiological evaluation, 45 had normal hearing acuity bilaterally and 17 had a final diagnosis of hearing deficit; two subjects did not execute audiological evaluation because parents did not agree to further tests. Overall, 17 well-babies (1.64‰) presented SNHL. Five infants (0.48‰) had unilateral SNHL (2 medium, and 3 severe-to-profound); twelve patients (1.15‰) had bilateral SNHL (2 mild, 3 medium, 4 severe, and 3 profound). Summary of the screening program results in well-babies is reported in Tables 1 and 2.

Among the 1233 neonates with audiological risk factors, 30 (2.4%) did not complete the screening program, 1026 (83.3%) resulted PASS, and 177 (14.3%) resulted REFER and underwent ABR. At the second step, 152 out of 179 had normal hearing acuity bilaterally, and 25 (2.07%) had SNHL (Table 3). Seven patients (0.56%) had unilateral SNHL (1 medium, 2 severe, and 4 profound); eighteen infants (1.45%) had bilateral SNHL (2 mild, 4 medium, 7 severe, and 5 profound). Overall, among 11592 newborns screened, 42 (3.6‰) had a final diagnosis of hearing impairment. Bilateral SNHL was found in 30 patients (2.56‰); among these, 18 (60%) presented audiological risk factors.

As for the etiology of SNHL, 13 patients were prematurely born or had suffered from respiratory problems, 8 had connexine 26 gene mutations, 7 presented syndromes related to SNHL, 5 had a malformation of the inner ear/cochlear nerve, 4 had idiopathic SNHL, 3 had family history of infantile deafness, and 2 had congenital Citomegalovirus infections. Table 4 shows the analysis of the quality indicators of UNHS in the University Hospital of Parma.

Table 1. Results of the first step of well-babies hearing screening in the four years analyzed

Year	Screened infants (n)	Pass (n) (%)	Refer (n) (%)
2010	2601	2470 (94.97%)	131 (5.03%)
2011	2771	2664 (96.14%)	107 (3.86%)
2012	2563	2512 (98.02%)	51 (1.98%)
2013	2424	2369 (97.74%)	55 (2.26%)
	Total: 10359	Total: 10015 (96.67%)	Total: 344 (3.33%)

Table 2. Summary of the results of the second step of hearing screening and audiological evaluation in well-babies.

Number of infants	Second step of screening			Audiological evaluation		
	Pass (n)	Refer (n)	Lost (n)	Normal hearing (n)	Hearing loss (n)	Lost (n)
344	253	64	27	45	17	2

Table 3. Summary of the hearing screening results in newborns with audiological risk factors

Infants (n)	Pass (n)	Lost (n)	Referred for audiological evaluation (n)	Normal hearing (n)	Hearing loss (n)
1233	1026	30	177	152	25

Table 4. Analysis of quality indicators of Parma UNHS and comparison with JCIH guidelines

Quality Indicator	JCIH guidelines	Results of Parma UNHS (overall)	Results of Parma UNHS (well-babies)	Results of Parma UNHS (babies with ARF)
Screening	> 95%	99.5%	99.7%	97.5%
Referral	< 4%	2.09%	0.61%	14.3%
False-positive rate	< 3%	1.7%	0.45%	12.3%
Lost to follow-up	< 5%	5.9%	8.4%	1.1%

Legenda. JCIH: Joint Committee on Infant Hearing; UHNS: universal newborn hearing screening; ARF: audiological risk factors

Discussion

SNHL is the commonest neonatal sensorial deficit with an estimated prevalence of 1 to 4 cases for 1000 newborns; this incidence mounts up to 4-5% in neonates with risk factors for SNHL (12). Unidentified bilateral SNHL > 40 dB at birth can adversely affect speech and language development as well as academic achievement and social-emotional development. It has been demonstrated that early identification and rehabilitation of SNHL (no later than 6 months of age) allows hard of hearing infants to perform as much as 20 to 40 percentile points higher on school-related measures (9). The aim of the universal screening is hence to identify as early as possible the highest number of infants with permanent bilateral hearing impairments. Medium-to-severe SNHL can be successfully treated by means of traditional hearing aids; cochlear implantation has long been proven to provide hearing restoration to children with severe-to-profound SNHL. The indications for cochlear implantation have expanded dramatically over time; currently, deaf chidren with inner ear malformations, additional handicaps and cochlear obliteration can be successfully rehabilitated by means of cochlear implants (13-19). Early intervention and a short duration of deafness before implantation are associated with the best language acquisition results, taking advantage of sensitive periods of auditory development.

The implementation of a UNHS program is complex and related to several factors, including economic resources, social nad cultural context, and available health professionals. In addition, different quality indicators codified by JCIH (10-11) have to be respected and monitored to ascertain whether a program is achieving expected benchmarks and outcomes. According to JCIH guidelines (10), the percentage of infants screened before 1 month of corrected age must be > 95%. This goal is easily achievable testing newborns before hospital discharge and immediately scheduling the test for the infants discharged without having carried out the screening. Following this approach we were able to test 99.5% of the patients. The percentage of infants who do not pass the birth admission screening and, thus, admitted to medical and audiological evaluation, is an important quality indicator; a value ≤ 4% is considered acceptable. Overall, in our program, 2.09% of patients were referred for diagnostic testing; over the 4-years period analyzed, this percentange lowered from 5.04% (2010) to 1.98% (2012) and 2.26% (2013). The growing experience of the professionals dedicated to the screening allowed us to achieve this result. However, while analyzing separately infants with audiological risk factors, the percentage of these patients who underwent audiological evaluation was 14.3%. Although there was a trend towards a decrease over the years, this value is not acceptable in a screening program of quality. As reported in the literature (20), the percentage of infants who do not pass the birth admission screen can be further lowered with a two-stage screening, based on otoacoustic emissions and automated auditory brainstem response (A-ABR). Furthermore, utilizing A-ABR offers two additional advantages. First, A-ABR is fundamental to identify infants with auditory neuropathy, a recently identified clinical/audiological entity characterized by normal TOAE and altered auditoy brainstem response (21). Secondly, A-ABR can reduce false-positive rate as well as audiological referral also in well-babies; in effect, unlike TOAE, A-ABR results are not adversely affected by middle ear fluid or ear canal debris (22). As for the percentage of false positives, a value <3% is considered adequate. In our program, an overall percentage of 1.7% was achieved. Yet again, a too high value (12.4%) was found in infants with audiological risk factors. Obviously, this was a direct consequence of high referral rate. Although values up to 30% are considered acceptable, the percentage of infants lost to follow-up should be <5% (23). Overall, we were able to achieve a percentage equal to 5.9% with a trend towards a constant and progressive decrease over the years. The percentage of patients lost to follow-up was considerably higher in well-babies (8.4%) than in infants with audiological risk factors (1.1%). Probably, we were not effective enough in explaining to parents the role of the universal screening program. Finally, the incidence of SNHL was found to be within the ranges reported in the international literature (24-25) both in well-babies (1.6‰) and in newborns with audiological risk factors (2.02%).

In conclusion, the analysis of the results of this study demonstrated a good quality of UNHS implemented in the University Hospital of Parma. Feedback

of our evaluation and quality indicators identified the necessity to improve referral and false-positive rate, especially in newborns with audiological risk factors. Introducing a two-step screening, based on TOAE and otoacoustic A-ABR could allow the achievement of the above-mentioned goals. A further improvement has to be realized in the communication process with families regarding the importance of completing the screening program in infants without audiological risk factors.

It is important to keep in mind that a normal hearing screening does not exclude a subsequent diagnosis of an acquired SNHL, nor does it rule out conductive or progressive hearing deficit with onset later in childhood. Thus, clinicians and family pediatricians are required to have a vigilant approach to assessment of hearing acuity and language development, even when UNHS has been resulted normal.

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