

Results of Applying a Universal Protocol for Early Detection of Hypoacusia in Newborn Infants for 42 Months

José I. Benito Orejas,^a Beatriz Ramírez Cano,^a Darío Morais Pérez,^a José L. Fernández-Calvo,^b and Ana Almaraz Gómez^c

^aServicio de Otorrinolaringología, Hospital Clínico Universitario de Valladolid, Valladolid, Spain

^bServicio de Pediatría, Hospital Clínico Universitario de Valladolid, Valladolid, Spain

^cUnidad de Apoyo a la Investigación, Hospital Clínico Universitario de Valladolid, Valladolid, Spain

Introduction and objectives: We present the results from applying over a period of 3.5 years a universal newborn hearing screening programme aimed at the early detection of hearing loss. Our goal has been to diagnose and treat all unilateral or bilateral permanent hearing losses ≥ 40 dB nHL.

Methods: The detection strategy consists of 2 screening phases with transient evoked otoacoustic emissions (TEOAE) and 1 diagnostic phase with auditory brainstem responses (ABR). ABR tests were performed on newborns "not passing" the TEOAE screening, as well as those with risk factors for hearing loss.

Results: Of 4568 live newborns (NB), we evaluated 97.6%. Of these, 3.7% presented risk factors for hearing loss. The first TEOAE was "passed" in both ears by 91.7% of the newborns; after the second TEOAE, only 1.3% of cases were referred to the diagnostic phase. We detected 2.7/1000 cases of severe/profound bilateral hearing loss.

Conclusions: This programme was designed for early application of appropriate treatment. Although it is possible to reach the objectives of screening and diagnosis of newborn hearing loss, the intervention stage is much more complicated. This is because intervention depends on different disciplines that must act in co-ordination, as well as multiple environmental and family factors that are difficult to control. Therapeutic effectiveness still remains our challenge and a motive for discussion in fully justifying early congenital hearing loss screening programmes.

Key words: Hearing loss. Neonatal screening. Otoacoustic emissions.

Resultados de aplicar durante 42 meses un protocolo universal de detección e intervención precoz de la hipoacusia en neonatos

Introducción y objetivos: Presentamos los resultados de aplicar durante 42 meses un programa de detección precoz universal de la hipoacusia neonatal. Nuestro objetivo ha consistido en diagnosticar y tratar todas las hipoacusias permanentes unilaterales o bilaterales ≥ 40 dB nHL.

Métodos: La estrategia de detección consta de 2 fases de cribado con otoemisiones acústicas transitorias (OEAT) y una fase de diagnóstico con potenciales evocados auditivos del tronco cerebral (PEATC). Los PEATC se realizaron tanto a los que "no pasaban" el cribado con OEAT como a los neonatos con factores de riesgo de hipoacusia.

Resultados: De los 4.568 recién nacidos (RN) vivos, hemos evaluado al 97,6%. Un 3,7% de los nacidos presentaba factores de riesgo de hipoacusia. El 91,7% de neonatos "pasó" la primera OEAT en ambos oídos, y tras la segunda sólo se derivó a la fase de diagnóstico al 1,3%. Hemos detectado 2,7 hipoacusias severas-profundas bilaterales/1.000.

Conclusiones: La finalidad de este programa reside en la aplicación precoz del tratamiento adecuado. Aunque resulta posible alcanzar los objetivos de cribado y diagnóstico de la hipoacusia neonatal, es mucho más complicada la fase de intervención, pues depende de diferentes disciplinas que deben actuar coordinadamente y múltiples factores ambientales y familiares difíciles de controlar. La efectividad terapéutica sigue siendo nuestro reto y el motivo de discusión para justificar plenamente los programas de detección precoz de la hipoacusia congénita.

Palabras clave: Hipoacusia. Cribado neonatal. Otoemisiones acústicas.

The authors have not indicated any conflict of interest.

Correspondence: Dr. J.I. Benito Orejas.
Avda. Salamanca, 16, 1.ª. 47014 Valladolid. España.
E-mail: jibenito@ono.com

Received September 25, 2007.

Accepted for publication January 6, 2008.

INTRODUCTION

Although hearing loss is the most common congenital disease,^{1,2} for years we have only had brainstem auditory evoked potentials (BAEP) as an objective early detection procedure.³ The time needed to carry out this test kept us from evaluating all newborns (NB),⁴ so a small group had to be selecting in which the prevalence of hearing loss was

greatest. Starting in 1972, the Joint Committee on Infant Hearing (JCIH) of the United States has published⁵⁻⁹ a set of "risk factors for hearing loss" that reduce the study population to 8%-10% of the general population.¹⁰ However, BAEP screening in NB with risk factors for hearing loss identified less than half of all cases of congenital, severe-profound bilateral hearing loss.^{1,11} In the light of the inefficiency of this "high risk" hearing screening in our setting,^{11,12} the introduction of otoacoustic emissions¹³ and the positive experience of the universal programmes in clinical practice that were launched in Spain and abroad,¹⁴⁻²³ we began to draft a protocol for universal detection of congenital hearing loss that we implemented in our hospital starting in March 2001.²⁴

The purpose of universal hearing screening consists of diagnosing all neonatal hearing losses, whether permanent, sensorineural or conductive (due to anatomic alterations of the outer and/or middle ear), unilateral or bilateral, ≥ 40 dB HL.^{25,26} The total incidence, which varies depending on the author,²⁷ is 14/1000. The results obtained by Mehl et al²⁵ reveal that most (68%) are bilateral sensorineural hearing loss; however it may also be unilateral sensorineural (20%), unilateral conductive (7%), and bilateral conductive hearing loss (5%). Just over 1% (1.2%) of the cases of congenital hearing loss would present only a permanent alteration of the middle ear.²⁸

According to JCIH (2000) guidelines⁹ endorsed by other groups,^{26,29-33} an early neonatal hearing loss detection and care programme should contemplate hearing screening of all NB during the first month of life, obtain an audiological diagnosis at 3 months of age and initiate treatment at around 6 months. A neonatal screening programme would be of no use and, in fact, would be unethical^{10,26} if it were not followed by early and effective treatment of the cases detected. However, just as the screening and diagnostic phases are conducted in the hospital setting, the treatment phase—including amplification (hearing aids, FM systems, and cochlear implants), speech rehabilitation (in its different modes of communication), and family support and intervention²⁶—has the tremendous inconvenience that it is the result of actions carried out by several different teams (multidisciplinary intervention)^{10,34} working in different settings, with different knowledge yet they must, nevertheless, maintain close collaboration (interdisciplinary intervention) to achieve the greatest efficacy. The complexity of this multidisciplinary and interdisciplinary intervention process, together with other additional factors highlighted below, make this treatment phase the most vulnerable of the entire programme.

The results obtained after conducting a universal screening programme with TEOAE over a period of 42 months (from March 2001 to August 2004) are presented. The impediments encountered during the early care of neonatal hearing loss are commented on from today's perspective.

MATERIAL AND METHOD

Our universal hearing screening protocol consists of 2 successive tests using TEOAE. A BAEP test is administered

to those who "do not pass" both TEOAE tests in 1 or both ears and to NB with risk factors for hearing loss (according to JCIH "high risk indicators for hearing loss" [1994]⁸) prior to the age of 3 months, corrected for prematurity.²⁴ For some 1300 NB per year in our centre, there are 2 nurses assigned to the Otorhinolaryngology Department with enough training to conduct the audiological screening and diagnostic tests; they work Monday to Friday in the morning. One of them is in charge of doing the auditory screening everyday and the other one performs BAEP in children and adults for later interpretation by the ENT specialist co-ordinating the paediatric hearing loss programme. The first screening test is carried out prior to hospital discharge, except for neonates who are discharged over the weekend. The second screening test is repeated on an appointment-only basis, at our department, during the first month after birth. At 6 months of age (corrected for prematurity), those NB who "did not pass" the 2 auditory screening phases and who have V-wave thresholds >30 dB nHL on the BAEP testing, confirmed with a second test, begin the treatment phase. Basically, this process comprises: auditory amplification (hearing aid centres with the means and ability to fit infant hearing aids are recommended); "early care" and language rehabilitation (performed at the Centro Base de Atención al Discapacitado [Base Centre for Care of the Disabled], dependent on the Social Services), and support and family intervention at specific centres Amigos y Padres del Sordo [Friends and Relatives of the Deaf], Agrupaciones de Sordos [Associations for the Deaf], etc). This entire multidisciplinary intervention programme, which varies depending on the child and family's demographic and socio-cultural conditions and appropriate to the patient's age and development, is co-ordinated by the social worker at our hospital in close collaboration with the ENT specialist in charge of the programme.

Finally, we complete the audiological diagnosis with auditory behaviour tests, performed by the same nursing staff at our department and by the infant hearing aid centres, starting at 8 months of age.

Depending on the V-wave thresholds obtained in the BAEP, we classify hearing loss as mild (40-50 dB nHL), severe (60-90 dB nHL), and profound (≥ 100 dB nHL). Children with mild bilateral hearing loss are referred to the Base Centre, where their capacities are stimulated and the advisability of a hearing aid is considered. Neonates with severe bilateral hearing loss are referred to the Base Centre and also to the infant hearing aid centres. Finally, we will also consider the advisability of a cochlear implant in children born with profound bilateral hearing loss.

The TEOAE tests were performed using ILO 92 clinical equipment (Otodynamics Ltd.) and the BAEP with Centor-C equipment from Racia-Alvar.

The validation criteria used for the TEOAE were:

- Validation of the test: background noise <45 dB SPL, at least 50 stimuli, stimulus intensity <86 dB SPL, stimulus stability $\geq 75\%$
- Validation of response: general reproducibility $\geq 70\%$, general response ≥ 10 dB SPL, response to at least

3 frequencies (1500 Hz, $\geq 50\%$, or ≥ 3 dB; 2200-3000 Hz; and 3700 Hz, $\geq 70\%$, or ≥ 6 dB)

The BAEP validation criteria are the presence of V-wave at 30 dB nHL.

The results of the different phases of the programme were entered into a database, previously designed in Microsoft Access.

RESULTS

Table presents the data obtained after applying our universal hearing screening protocol with TEOAE for 3.5 years (March 2001 to August 2004).

Results of Conducting a Universal Neonatal Hearing Screening Protocol With TEOAE Over a Period of 42 Months^a

Study period: March 2001 to August 2004
Live newborns: 4568
Newborns with risk factors: 3.7%
Newborns evaluated: 97.6%
Results of the first TEOAE (phase 1)
Failure to pass: 8.3% (3% in both ears)
Failure to attend: 1.1%
Ratio of false positives: 95.4% (children who fail to pass and do not have the illness)
Specificity: 91.7%
Coverage of the first phase: 97.6%
Results of the second TEOAE (phase 2)
Failure to pass: 18% (1.2% of the total)
Failure to attend: 4%
Ratio of false positives: 87% (total false positives: 0.94%)
PPV: 13% of the newborns without RF, who do not pass this second phase, have some kind of hearing loss (8.7% have severe-profound bilateral hearing loss)
Specificity: 98.9%
Coverage of the second phase: 96%
Continuity index from phase 1 to phase 2: 96%
Results of phase 3 (diagnosis)
Referral: 5% (3.7% for RF and 1.3% for "failure to pass")
Bilateral severe-profound hearing loss: 2.7/1000
In the general population: 1.4/1000
In newborns with risk factors: 6.6%
In the Gypsy population: 1.9%
Continuity index from phase 2 to phase 3: 95.5%

^aPPV indicates positive predictive value; RF, risk factors; TEOAE, transitory evoked otoacoustic emissions.

We detected 17 patients with hearing loss (3.8/1000): 9 profound bilateral, 3 severe bilateral, 2 severe unilateral, and 3 mild bilateral. From another point of view, 11 (65%) of the cases of hearing loss were bilateral sensorineural; 4 (23%) were bilateral transmission, and 2 (12%) unilateral transmission. Of the 12 patients (2.7/1000) bilateral severe-profound hearing loss, 8 (67%) were associated with risk factors. Hearing loss was present in 6.6% of the neonates with risk factors and 1.4/1000 healthy births. More than 7% (7.8%) of the population studied was of Gypsy ethnic origin, in which there is a 2% incidence of hearing loss (vs 3.8/1000 in the rest), and 10% of them failed to attend the diagnostic phase (vs 1% of the rest).

The rapid increase of other races in our population is particularly striking, as revealed by the fact that this category grew from 1.8% in the first semester of our study to accounting for 5.7% in the last semester, with a predominance of individuals from South American countries (51%) and from Eastern Europe (30%).

The results achieved during the treatment intervention phase were as follows:

- The diagnostic phase with BAEP was completed in all the patients in the first 3 months of life and rehabilitation was initiated at that age (stimulation and/or speech therapy, depending on the case at hand)

- Of the 9 children with profound bilateral hearing loss, 3 (belonging to the same Gypsy family tree) deceased between 5 and 7 months due to progressive familial encephalopathy; the remaining 6 were fitted with bilateral hearing aids around the age of 6 months; 3 of them received a cochlear implant around the age of 2 years and another 2 refused them (one of Gypsy origin and the other, the son of parents with profound deafness) and continued with their hearing aids. Finally, another patient displayed significant psychomotor delay as a result of candidiasis of the brain during birth. The responses to sound attained in one of the ears led to the hearing loss being considered as severe instead of profound and the patient was not admitted into the cochlear implant programme for the time being

- There were 3 children with severe bilateral hearing loss. In one case, the parents, who lived in another province, refused to have their child outfitted with a hearing assistive device and did not come for the check-up appointment. Another patient with Treacher-Collins syndrome exhibited bilateral auricular atresia. At 4 months of age, he was fitted with a bone conduction hearing aid that he continues to use at the age of 5 years (the parents, content with the results, have not opted in favour of an osteointegrated prosthetic device). Finally, another child, with V-wave thresholds of 60-65 dB nHL on the BAEP, presented low-tone levels of 40 dB, with good evolution with speech therapy during the first year. The family later moved to another city and the parents have reported that at 4 years of age, he was fitted with bilateral hearing aids because of a certain degree of delay in language development

- Three children with mild bilateral hearing loss were detected. All received speech therapy from the time of diagnosis. These children suffer frequent bouts of otitis

media serosa (OMS) that continually modify the audiometric thresholds. Given their evolution, 2 of them were fitted with bilateral hearing aids, 1 at 2.5 years of age, and the other at 4 years of age. All display age-appropriate language development

– There were 2 children with congenital auricular atresia and severe hearing loss on that side. One of them, as a result of the common episodes of OMS in the healthy ear, required follow-up and speech therapy during the first years of life

In summary, of the 9 cases of severe-profound bilateral hearing loss we could treat (3 of these children died before treatment could be started), 3 patients rejected our treatment proposal, 1 moved, preventing follow-up and another child presented severe psychomotor and language delay, making it difficult to determine the relationship between the core deficit and the peripheral sensory deficit. In short, in 4 of 9 patients we were unable to achieve the treatment objectives we had initially proposed.

DISCUSSION

Although convinced, in the light of our prior experience,^{11,12,24} that the best auditory screening is universal screening, we are aware of the tremendous difficulties this entails.

First of all, the physiological tests used in the screening (TEOAE) and diagnosis (BAEP) of hearing loss do not have 100% sensitivity and specificity,³⁵ and false positives and false negatives may emerge. Even though we can decrease the false positives by repeating the tests (2 phases of TEOAE and 2 of BAEP),³⁶⁻³⁸ certain secondary effects are inevitable, such as the emotional trauma for the family, the label of the disease, iatrogenic effects of repeated unnecessary testing, and increased costs.¹ Given that TEOAE are incapable of assessing the integrity of sound transmission from the internal ciliary cells to the brain, we performed BAEP in all neonates with risk factors (even in those who “passed” in the TEOAE). Nevertheless, in the 6 years that have elapsed since the beginning of this protocol, we have not detected a single case of auditory neuropathy or a single false negative.

In contrast, the prevalence of permanent sensorineural hearing loss continues to rise during childhood.^{39,40} Although the children with risk factors for progressive hearing loss or for late onset hearing loss undergo follow-up every 6 months for the first 3 years of life,^{9,41} we believe it is very important for primary care paediatricians to be involved, since with their surveillance and control, they participate in early detection of the postnatal hearing loss.⁴²

However, if the screening and diagnostic phases of childhood hearing loss entail certain difficulties and even, as we have seen, inaccuracies, we believe it is possible to achieve the widely accepted objectives.^{9,29-31,33} During the course of the 42-month period of this study, we have screened the hearing of more than 95% (97.6%) of all NB, completing this phase during the first month of life. The rate of false positives (“failure to pass” both screening phases and no hearing loss) was 0.94% and, hence, lower than the

recommended 3%. Just over one percent (1.3%) of the children who “failed to pass” were referred for diagnostic study (accepting up to 4%) and we have completed the audiological diagnosis with BAEP in the first 3 months of life of the neonate (although the degree and configuration of the hearing loss were obtained later).

Therefore, in our opinion, the fundamental hurdle in an early hearing loss detection programme lies in the treatment intervention phase. In 1994, the audiologist Fred Bess and the paediatrician Jack Paradise⁴³ published an interesting article in which, while acknowledging the prevalence and importance of identifying severe congenital hearing loss early, they state their reservations with respect to treatment, specifying that it must be “efficacious, available, easily performed, and have reliable tests that demonstrate that early treatment is more effective than late treatment.” In 1998, Christine Yoshinago-Itano and her team in Colorado demonstrated⁴⁴ that children with congenital severe-profound bilateral hearing loss identified and treated before 6 months of age displayed better language acquisition than if treatment had been given later. Nevertheless, in 2001, the US Preventive Services Task Force (USPSTF)⁴⁵ stated again that there were not enough prospective or controlled studies analyzing whether neonatal auditory screening and early intervention in hearing loss result in improved language and educational development. We agree with the reply from Yoshinago-Itano’s group⁴⁶ and although it is true that there are not enough randomized clinical trials, the many retrospective works that do exist provide sufficient evidence of the benefits early intervention offers.²⁷ Professionals with experience in the education of deaf children are convinced that early identification of hearing loss and appropriate, immediate intervention can potentially improve the deaf child’s language, speech and social and emotional development, as well as that of his/her family.^{8,14,29,47,48} However, applying treatment that is “efficacious, available, and easily performed” is a complicated affair.⁴⁹ In our setting, despite having sufficient resources to provide the hearing impaired child with all the care he/she requires and having a well-trained and efficient multidisciplinary team working in a co-ordinated fashion, we have found that there are other factors that come to bear on the process of intervention. Population movements, cultural, and socio-economic differences, a rural setting, communication difficulties, concomitant illnesses, and respect for the parents’ or caregivers’ decisions, amongst others, hinder the obtention of the desired results and make the holistic treatment of hearing impaired children a permanent challenge. In 4 of the 9 children whose severe-profound bilateral hearing loss we sought to treat, we did not achieve the proposed purposes. As in the screening and diagnostic phases, we believe that it is essential for the primary care paediatricians to be committed and involved, and for them to be familiar with the treatment intervention protocol, and permanently redirect the child and the family toward the priorities at each given point in time.⁵⁰ Finally, although mild hearing loss is a priori deleterious for the child’s language acquisition and psychological and intellectual development, its frequent interrelation with OMS,⁵¹ sometimes due to the persistence

of amniotic liquid or mesenchyme in the middle ear,⁵² fosters its continuity and represents a challenge and considerable expense, in terms of follow-up until a definitive diagnosis can be made.

We have improved our results with respect to those obtained after the first year of screening with TEOAE.²⁴ The risk factors with the highest index of pathogenicity continue to be craniofacial malformations and family history of hearing loss, closely followed by infection and low birth weight. As we had anticipated,²⁴ the total number of cases of severe-profound bilateral hearing loss has fallen from 5.8 to 2.7/1000, which would be comparable to outcomes reported by other authors.²⁷ According to Mehl et al,²⁵ 65% of cases of hearing loss were bilateral sensorineural hearing loss. The high percentage of hearing loss detected among Gypsies is remarkable (accounting for 8% of our study population), with an incidence that is 5 times that of the rest of the population (2/100 vs 3.8/1000) and with loss of patients in the diagnostic stage (failure to attend appointments) 10 times greater (10% vs 1%). In cases where social marginalization also hinders follow-up (we are beginning to see this problem in the immigrant population, which is growing slowly), we believe that it is particularly important, at the very least, to establish a diagnosis of "normality" prior to discharge, that is, that the screening tests performed should have the highest specificity.

In September 2004, our Regional Government in Castilla-León launched the widespread Early Detection and Comprehensive Attention Programme for Childhood Hearing Loss,⁵³ agreed upon by all the public hospitals. This consensus led to the choice of the automatic brainstem auditory evoked potentials (BAEP-A) as the screening procedure, considered more effective than TEOAE. We have finished an initial stage of universal hearing screening with TEOAE in our hospital, that we have maintained over the last 3 and a half years and are beginning another new one using BAEP-A, the results of which over time and its comparison with the one presented here will prove whether the decision to change from TEOAE in favour of BAEP-A as the screening procedure is a good one.

CONCLUSIONS

We feel that the universal detection strategy is the best early diagnostic procedure for congenital hearing loss.

The main disadvantages of the use of TEOAE in the auditory screening phase are: the high percentage of false positives and the inability of the test to detect retrocochlear involvement. Both factors increase the number of children who are referred and make it easier for patients to be lost in the process. If this patient drop-out is significant, the specificity of the neonatal auditory screening procedure should be increased.

In our setting, bilateral OMS constitutes the most common difficulty impeding audiological diagnosis, above all in the case of mild hearing loss.

In a early detection and intervention of childhood hearing loss programme, we find the treatment phase to be the most

difficult and vulnerable of the entire process, since it is a multidisciplinary and interdisciplinary task. Its proper functioning not only depends on the availability of means and resources, but also on social, cultural, economic, and medical factors, among others, that should be taken into account.

Someone must co-ordinate continuity between the hospital setting (where the hearing loss screening and diagnostic phases take place) and the extrahospital setting (where treatment intervention is carried out). In our case, the ideal person is the hospital social worker.

Finally, in order for the programme to run properly, the primary care paediatrician must necessarily be involved, through the Well Child Programme, collaborate in controlling screening, diagnosis and intervention in the hearing-impaired child, correcting any deviations that might arise.

Acknowledgements

The good results of the screening and diagnostic phases are due to the ENT nursing staff, Esther, and Mamen. The interdisciplinary co-ordination of the treatment phase is led very competently by Begoña, our social worker. We are grateful to the paediatrics and obstetrics departments at our hospital and to the paediatricians, midwives, and obstetricians in primary health care for their attitude toward our work and for the help provided. We would also like to thank all those who were there with us.

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