Hearing loss in neonates and infants. Audiological neonatal screening: an international group experience

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Abstract:

Objective: The aim of the study has been the development of an universal screening for congenital hearing loss in the neonatal period in Europe.

Methods: 1021 infants (both healthy and sick new-borns) were screened for congenital hearing loss between September 2000 and January 2008. All newborns were screened primarily with otoacoustic emission and otoacoustic products of distortion as first stage screening; with automated auditory brainstem response audiometry and auditory steady state responses as a second stage screening for those who failed the otoacoustic emission test.

Results: Hearing loss was confirmed in 11patients (5 unilateral and 6 bilateral). The incidence of congenital hearing loss was 0.84%in presumed healthy infants and 2.10% in infants admitted to the intensive care nursery.

Conclusions: Several protocols are available for universal newborn hearing screening programs: although otoacoustic emission is quicker and easier to perform than automated auditory brainstem response audiometry, otoacoustic emission is affected by external ear wax or fluid. To avoid misinterpretation, we consider important combined measurements of otoacoustic emissions and otoacoustic products of distortion with automated auditory brainstem response audiometry and auditory steady state responses. Screening for congenital hearing loss can be carried out with a very low rate of referrals and a low rate of false positive tests, particularly if there is access to otoacoustic emission as well as automated auditory brainstem response testing and auditory steady state responses.

Key words: ABR, auditory brainstem response, audiological neonatal screening; OAE, otoacoustic emissions.

Introduction:

In children, undiagnosed hearing loss often leads to permanent developmental delays: congenital or acquired hearing loss in infants and children has been linked with lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties [1,2]. Identification of hearing loss through neonatal hearing screening as well as objective hearing screening of all infants and children can prevent or reduce many of these

adverse consequences [1,2]. Significant hearing loss is present in 1 to 6 per 1000 newborns [2-4].

Although some congenital hearing loss may not become evident until later in childhood, most children with congenital hearing loss have hearing impairment at birth and are potentially identifiable by newborn and infant hearing screening [5].

Universal detection of infant hearing loss requires universal screening of all infants: since September 2000, we have performed an audiological screening of all newborns of our hospital.

Materials:

1021 children were included in this neonatal audiological screening and tested between September 2000 and January 2008: the screening protocol included otoacoustic emission (OAE), otoacoustic products of distortion (DPOAE), automated auditory brainstem response audiometry (ABR) and auditory steady state responses (AUDIX).

This initial screen was designated stage 1: in this stage, all newborns were screened primarily with OAE and DPOAE (OAE and DPOAE were recorded using the Otodynamics ILO92 Version 5.6).

The infants who failed the otoacoustic emission test were examined with ABR (carried out using MK 72- ABR screener with natus-ALGO2e) and AUDIX, this was designated as stage 2.

AUDIX thresholds were recorded after ABR testing. ASSRs were measured from the forehead to mastoid/earlobe electrodes (impedance maintained at <3 KΩ) using the ERA run on a personal computer Pentium processor. Stimulation of a repetitive sinusoidal amplitude/frequency (AM/FM)-modulated tone was delivered using ER3A 10Ω insert earphones. Test frequencies were 500, 1000, 2000, and 4000 Hz. If time permitted, 250, 1500, and 8000 Hz were also tested. Total testing time with all frequencies completed was approximately 1.5 hours with 30 minutes reserved for anesthesia recovery time.

Results:

During the study, 831 healthy term newborns (81%) were born at San Martino Hospital (Genoa), of which 831 (100%) had stage 1 hearing screen performed. All the infants (n=29) who failed the stage 1 screen were referred for stage 2 follow-up screening (3%). All of these children completed stage 2 screen, and of these 7 (0.84%) failed: four had severe bilateral hearing loss and 3 had mild to moderate hearing loss (three of the 4 children with severe bilateral hearing loss met high-risk registry criteria). Three infants who failed stage 2 screening were later found to have normal hearing (stage 2 false-positives). The overall stage 1 false-positive rate was 2% (n=17).

In the same period (from February 2000 and January 2008), we analyzed 190 infants (19%) admitted to the intensive care nursery of San Martino Hospital (Genoa), all (n=190) had stage 1 hearing screen performed. All the infants (n=11) who failed the stage 1 screen were referred for stage 2 follow-up screening (5.7%). All of these children completed stage 2 screen, and of these 4 (2.1%) failed: one had severe bilateral hearing loss and 3 had mild to moderate hearing loss (the child with severe bilateral hearing loss didn't present high-risk criteria). One infant who failed stage 2 screening was later found to have normal hearing (stage 2 false-positives). The overall stage 1 false-positive rate was 2,1% (n=4).

Because we did not collect data on false-negative tests, sensitivity and negative predictive value of the screening process could not be calculated.

Discussion:

Objective screenings for hearing impairment should be performed on all infants and children: the technology used for hearing screening should be age appropriate and the child also should be comfortable with the testing situation; young children may need preparation. Screenings should be conducted in a quiet area where visual and auditory distractions are minimal.

Hearing disorders affect the perception of complex sounds in a variety of ways, depending on the sites of lesions: early-onset hearing impairment can seriously impede language development. Language cannot develop normally without adequate speech stimulation and deafness is more prevalent than any other handicapping condition for which mandated neonatal screening programs exist. Sensitive and inexpensive techniques are available for performing neonatal hearing screening, and early intervention has a documented positive effect on development of language skills in hearing-impaired children [6,7].

Children who have high-risk indicators for hearing loss should be referred promptly for audiologic evaluation: electrophysiologic techniques such as ABR and OAE are universally used for the identification of significant hearing loss in newborn infants [7,8]; [Table 1.]

Family history of SNHL
In utero infection associated with SNHL
Ear and other craniofacial anomalies
Hyperbilirubinemia at levels requiring exchange transfusion
Birth weight less than 1500 g
Bacterial meningitis
Low Apgar scores: 0–3 at 5 min; 0–6 at 10 min
Respiratory distress
Prolonged mechanical ventilation for more than 10 days
Ototoxic medication administered for more than 5 days or used in combination with loop diuretics

Table 1: High risk factors for hearing loss in children from birth to 24 momth of age (SNHL: sensorineural hearing loss).

To justify universal screening, at least five criteria must be met: an easy-to-use test that possesses a high degree of sensitivity and specificity to minimize referral for additional assessment is available; the condition being screened for is otherwise not detectable by clinical parameters; interventions are available to correct the conditions detected by screening; early screening, detection and intervention result in improved outcome; the screening program is documented to be in an acceptable cost-effective range [3,9,10,11].

Although additional studies are necessary, review of both published and unpublished data indicates that all five of hese

criteria currently are achievable by effective universal newborn hearing screening programs [9-11].

ABR, OAE and AUDIX are objective tests of peripheral function that can be administered to individuals of all ages and are acceptable methodologies for physiologic audiological screening.

The OAE test is an effective screening measure for inner and middle ear abnormalities, because at hearing thresholds of 30 dB or higher, there is no OAE response. The OAE test does not further quantify hearing loss or hearing threshold level and does not assess the integrity of the neural transmission of sound from the eighth nerve to the brainstem and, therefore, will miss auditory neuropathy and other neuronal abnormalities. Infants with such abnormalities will have normal OAE test results but abnormal ABR test results [12,13].

ABR can test each ear individually and can be performed on children of any age. Motion artifact interferes with test results. For this reason, the test is performed best in infants and young children while they are sleeping or, if necessary, sedated [13].

Testing for profound hearing impairment is limited, however, by the upper limit of ABR testing at 90 to 100 dB nHL, depending on the equipment used. Thus, the identification of exact hearing levels for individuals with profound hearing impairment is not possible.

In our department (University of Genoa), we began to consider auditory steady-state evoked responses (AUDIX) as a new method for objective hearing testing, since 2000.

Our investigation has shown AUDIX testing to be superior to ABR testing for the evaluation of hearing level in newborns. Compared with ABR, AUDIX offers frequency-specific information that allows for an estimated audiogram based on algorithms. AUDIX is an objective test that can extend to the severe to profound levels of hearing loss: AUDIX testing provides threshold information in the 90 to 120 dB range of hearing loss where amplification is not beneficial. On the other hand, ABR is a test of exclusion for these children to demonstrate that hearing is worse than 90 to 100 dB nHL without giving the exact level of hearing [14,15].

ABR and OAE are tests of auditory structural integrity, but are not true tests of hearing. Even in the presence of a normal ABR and normal OAE, there is no way to guarantee that a child "hears" until he or she is mature enough to indicate so behaviorally: thus, follow-up evaluations should be scheduled until a reliable audiogram can be obtained. In addition to rescreening infants before discharge, the use of AUDIX as a screening tool is an important factor in minimizing false-positive results: it has been shown to consistently produce lower false-positive rates than the otoaccoustic emissions test, when other screening tests are used screening [16].

The false-positive rates previously reported for universal newborn hearing one-stage screening programs range from 2.5 to 8%; therefore, minimizing false-positive results is critical in developing a more reliable newborn hearing screening program: our two-stage screening test was effective in obtaining a lower false-positive rate [17].

Undiagnosed hearing loss often leads to permanent developmental delays: the goal of early diagnosis and intervention for a congenital hearing loss is to enable the child to develop language and communication skills that correspond to his chronological age and

innate cognitive abilities. The routine evaluation of hearing should include the identification of parental concerns regarding infant hearing as well as the assessment and diagnosis of infants with potential hearing impairment: identification of hearing loss should be followed by early interventions to prevent developmental delays. Therefore, these observations and the data of our study endorses the implementation of universal newborn hearing screening.

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