Automated auditory brainstem response hearing screening in NICU graduates

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5.1 Abstract

Severe congenital hearing impairment is an important handicap affecting 0.1% of apparently healthy liveborn infants and 1-2% of graduates of neonatal intensive care units. The prognosis for intellectual, emotional, language and speech development in the hearing-impaired child is improved when the diagnosis is made early and intervention is begun before the age of 6 mo. Universal screening is preferable, since about 50% of infants with hearing loss are not discovered if neonatal hearing screening is restricted to high-risk groups. The automated auditory brainstem response (AABR) screener is a dedicated hearing screening device, which provides information not only about the outer/middle ear and cochlea but also about the auditory pathway up to the brainstem. AABR has an agreement with conventional auditory brainstem up to 98%. It uses a 35 dB near hearing level click. No operator interpretation is needed and it can be used on the ward and during oxygen therapy without disturbance from ambient noise. Reported referral rates in a hospital-based screening programme at the time of discharge vary, with an average of 4%. AABR has also been used in a home-based setting, with the same results. The time necessary for screening varies with the setting, but ranges from 4 to 15 min.

Initial costs range from $15 to $25 per test, which is similar to neonatal screening for metabolic diseases. In addition to individual healthcare savings, early diagnosis may lead to savings on costs of intensive speech-language intervention and educational facilities.

5.2 Introduction

Normal hearing during very early life is of utmost importance for laying the basis for speech and language development. Hearing impairment leads to sensory deprivation with failure to develop communication skills, which in turn leads to learning problems and subsequently to problems with social and emotional development. Congenital hearing loss is apparent in 0.1% of the normal population and is much higher in a defined at-risk population (1-2%). Infant hearing has a high priority in preventive healthcare schemes in many European countries. Money and resources are made available for screening programmes, for example, in The Netherlands. Hearing screening is carried out in approximately 90% of infants at the age of 9 mo using distraction methods. However, the mean age of detection of bilateral congenital hearing loss is at least 18 mo in many countries. This is later than the age of 6 mo in which habilitation of hearing loss is significantly associated with better speech and language development, well documented in a recently published prospective study.
Table 6  
*Risk criteria for hearing loss in the neonatal period according to the American Joint Committee on Infant Hearing.*

- Family history of congenital sensorineural hearing impairment
- Congenital perinatal infections (Syphilis, TORCH)
- Craniofacial anomalies
- Birth weight < 1500 g
- Hyperbilirubinaemia exceeding a level needing exchange transfusion
- Use of ototoxic medication in potential toxic dose (e.g.: aminoglycosides, diuretics)
- Bacterial meningitis
- Severe birth asphyxia (Apgar ≤ 4 at 1 min or ≤ 6 at 5 min)
- Mechanical ventilation > 4 days
- Syndromes associated with sensorineural hearing loss

Neonatal hearing screening has become a topic of interest since objective measurement of hearing loss became possible in the newborn in the late 1980s. Following the 1994 Joint Committee on Infant Hearing (JCIH) consensus statement neonatal hearing screening is rapidly becoming "standard care" in the USA. In Europe, the European Consensus Development Conference in 1998 was a strong impulse for efforts to run universal or high-risk register screening programmes.\(^3,6\)

Both statements agree that the development of children with bilateral hearing impairment can be improved with early detection (<3 months) followed by habilitation started before the age of 6 months.

Two methods for neonatal hearing screening based on different physiological phenomena are available: otoacoustic emissions (OAE) and auditory brainstem response (ABR). This article presents the use of automated ABR (AABR) in term and preterm infants.

5.3 Methods

*Automated auditory brainstem response.*

For the neonatal period, conventional ABR is considered to be the most reliable method for assessment of the hearing level.\(^7,8\) The conventional ABR method is not widely used for screening because it is time consuming and it needs a well-qualified technician and audiologist to perform the test and evaluate the results. Now, a second-generation Automated ABR infant hearing screener (ALGO-1e and ALGO 2, Natus Medical, San Carlos, CA, USA) is available for screening purposes. The ALGO-AABR screener is extensively described by Jacobson et al.\(^8\) and Kileny.\(^9\) The AABR uses 35 dB near hearing level (nHL) click stimuli, presented mono-aurally at a rate of 37 pulses/second. The clicks have an acoustic spectrum, which is flat from 750 to 5000 Hz. After artefact reject-
on for ambient noise and myogenic activity, an internally programmed template-matching algorithm measures ongoing electroencephalographic activity for the presence or absence of the ABR. This sampling uses a statistical test, the likelihood ratio. After reaching a likelihood ratio of 160, the ALGO-1 Plus stops collecting data and displays a "pass" for the ear being tested. This indicates that the data collected were sufficient to discriminate between the presence of a "response plus noise" and the presence of pure noise, or a "no response" condition at better than the 99.80% confidence level. AABR hearing screening is safe, simple to operate, quick to administer to large populations and can be used by personnel who have no special audiological training. An AABR neonatal hearing screening programme could adequately fulfil the principles laid down by Wilson and Jungner in a WHO report (see Table 2). Several clinical trials have been carried out comparing the use of the first-generation ALGO1-Plus with the conventional ABR. In a multi-institutional clinical trial, Hall et al. found a sensitivity of 100% with a specificity of 96.7%. Kiliny found a sensitivity of 100% and a specificity of 96.15%. Using a controlled protocol Peters found a sensitivity of 100% and a specificity of 98.7% and Jacobson et al a sensitivity of 100% and a specificity of 96%. In a clinical trial in Colorado Mehl et al. found no evidence of a single false-negative test result. The sensitivity of newborn screening is therefore at or near 100%. A trend towards greater specificity is noted in second-generation AABR equipment, using improved technology. A more meaningful number is the positive predictive value of the test, namely the number of true positive results divided by the total number of positive screening tests. The positive predictive value for all newborn hearing screening in Colorado for the second-generation AABR equipment is as high as 19%. This compares favourably with positive predictive values for screening for haemoglobinopathy (1%), cystic fibrosis (4%) and hypothyroidism (3%) only exceeded by screening for phenylketonuria (PKU) (80%).

Automated auditory brainstem response as a universal screening device. The world literature on neonatal hearing screening projects comes from countries where practically all births take place in hospital, as in the USA. At present the AABR method is widely used in neonatal hearing screening programmes in the USA. A survey of practice (July 1998) in 17 states (accounting for 28% of total births in USA) participating in the Marion Downs National Center programme showed that more than half of the centres used screening ABR, 25% used OAE and the remainder used either conventional auditory brainstem response or a combination ABR-OAE protocol. Centres used a variety of personnel, including nurses, audiologists, technicians, and volunteers, to perform the screening. Most of the babies were screened within 36 hours of birth, which is before the hospital discharge in nearly all cases. The time necessary for screening is supposed to be 4-7 min for second-generation devices in an inpatient setting. The major challenge is to track referrals and transition of these infants and
families in a follow-up programme. For this reason a low referral rate at the first stage is of utmost importance. The performances of the AABR screening method (0.6-5.3%) in this context exceeds that of OAE screening (5-20.7%).¹⁷

The initial costs of AABR hearing screening have been reported to be in the a range of $17-33 per inpatient screening.¹³,¹⁸ By comparison, the true cost of a blood screening test, including laboratory, phlebotomy, and personnel costs, for any one of the newborn genetic screening diseases in Colorado is estimated to be $3 per infant. The cost of screening for congenital hearing loss, however, must be interpreted in the context of a disorder with a higher prevalence. The screening costs required to identify one new case of congenital hearing loss are calculated to be $9600-17750 and are similar to hypothyroidism ($10000), haemoglobinopathy ($23000), and PKU ($40000).¹³,¹⁸

In other countries, such as The Netherlands, a less "captive population" is available for neonatal hearing screening. In The Netherlands, approximately 35% of all births take place at home and of those neonates born in hospital, approximately 35% leave the hospital within a few hours of birth.¹⁹

A neonatal hearing screening programme in The Netherlands therefore needs to be flexible enough to allow screening outside the hospital and could be embedded in an outstanding network of well-baby clinics. In a feasibility study, Oudesluys-Murphy and Harlaar showed that the AABR method can also be used for neonatal hearing screening in the home setting.²⁰ Even with the first-generation AABR screener a referral rate was reached of 6.3% in a one-step screening procedure and 2.5% in a two-step procedure. At present, a neonatal AABR hearing screening programme outside the hospital setting is running successfully in Flanders (Belgium).²¹

Automated auditory brainstem response screening in the high risk group

Although universal neonatal hearing screening should be the goal, some hospitals or countries may prefer to start with a high-risk register hearing screening programme. Nearly all neonatal intensive care unit (NICU) babies belong to this high-risk register group. (Table 6) Besides, neonatal intensive care has been centralized in many countries. Hearing screening in the NICU is therefore very cost-effective. Because of the high risk of congenital hearing impairment, the National Institutes of Health (NIH) recommends hearing screening of all NICU graduates before discharge.³

Although transient evoked otoacoustic emissions (TEAOE) have been advocated as a first-stage technique for universal healthy newborn hearing screening, several problems have to be solved to make this technique suitable for at-risk neonates and for use in preterm newborns. In preterm babies Brienesse could detect a click-evoked OAE in 69% of the attempts, of which 40% had a reproducibility above 40% and 44% reached a signal-to-noise ratio >3dB.²² AABR is the method of choice for neonatal hearing screening in NICU settings. It is feasible for use on the ward and in the incubator, even during nasal CPAP oxygen therapy or artificial ventilation, without disturbance from either ambient noise or
technical equipment. Van Straaten et al. reported a 92% pass after the first screening and 98% pass after the second screening. All neonates referred after the second screening were proved to have congenital hearing loss. In this prospective study no false negatives were found at follow-up, which made the AABR neonatal hearing screening of great value in the total evaluation of the at-risk newborn, in which anticipation of probable disorders of neurological development is an important part of the follow-up. In a retrospective study of the Nottingham AABR screener used in a NICU population Mason et al. reported a sensitivity of 90% and a specificity of 93%, which compares favourably the results of OAE screening in an equivalent group, which had a sensitivity of 80% and a specificity of 92%.

The feasibility and excellent performance are not only the reason for using AABR in the at-risk population. Of even greater importance is the higher prevalence of neurological sequelae in this population. One of the main advantages of AABR screening is that it provides information not only on conductive hearing loss and cochlear pathology, as does OAE screening, but also on the more central auditory pathology up to the midbrain. Stein has drawn attention to the entity of auditory neuropathy. This is a form of hearing impairment characterized by normal or near-normal hearing sensitivity but reduced auditory perceptual or speech-processing skills and electrophysiologically and physiologically characterized by an absent or a grossly abnormal ABR but normal evoked OAE. The presence of auditory neuropathy in an undetermined number of NICU graduates raises questions about whether evoked OAE should be used as the initial screening tool for infants in a NICU.

The mean screening time in the NICU setting seems to be somewhat longer than in universal neonatal hearing screening. Since the introduction of the second-generation AABR screening devices the mean screening time has been reduced from 26 to 13 min, mainly as a result of a reduction in preparation time.

The click-evoked ABR typically appears during the 27th week of gestation, but may occur as early as the 25th week. Development of the infant ABR is usually complete by the second year of life. Use of AABR is officially recommended from 34 weeks gestational age onwards. Clinical practice in most countries with a centralized system for neonatal intensive care involves discharge of the high-risk newborn as soon as possible to the referring hospital. This is generally between 30 and 34 weeks corrected postmenstrual age for the very preterm babies. To ensure quality of care as well as cost-effectiveness AABR screening should be performed before discharge. In a prospective study it was shown that AABR screening is possible even in very preterm newborns. More than 80% passed the screening from 30 weeks onwards and 100% from 31 weeks corrected postmenstrual age in infants born between 26 and 30 weeks gestational age.

AABR neonatal hearing screening is being implemented in the NICU in The Netherlands. The screening is the first part of a nation-wide neonatal hearing screening programme. Central data registration ensures the quality of the screening programme. Data from
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NICU screening will be combined with data from universal hearing screening organized by the well-baby clinics. This will provide a national databank with information on the outcome of screening and ensure that diagnostic investigation and intervention pathways are followed. The screening process will be monitored continuously in this manner.

5.4 Conclusions

For the optimal development of a newborn child universal neonatal hearing screening should be the ultimate goal and all neonates with hearing impairment should be allowed to benefit from early diagnosis and intervention. AABR is a high-quality neonatal hearing screening method. It is easy to perform in a hospital setting as well as outside the hospital. It is suitable for NICU graduates before discharge to the referral hospital as well as for healthy newborns. The objective results should be registered for central monitoring of a nation-wide programme which includes tracking of referrals, further diagnostic audiolgic assessment, hearing-aid fitting for very young infants and intensive monitoring of the speech and language development, as well as socioemotional development of the young hearing-impaired child. Costs are a critical issue. Initial costs ranges from $15 to $25 per screening, resulting in a similar cost per case as for neonatal screening of metabolic diseases. Early diagnosis will primarily lead to savings on costs of intensive speech-language intervention and educational facilities, rather than a reduction of healthcare costs.
Chapter 5

5.5 References

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