Automated auditory brainstem response hearing screening in NICU graduates
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Citation for published version (APA):

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Evaluation of an Automated Auditory Brainstem Response Infant Hearing Screening Method in at Risk Neonates

Based on the article:

*Evaluation of an Automated Auditory Brainstem Response Infant Hearing Screening Method in at Risk Neonates.*
H.L.M. van Straaten, M.E. Groote, A.M. Oudesluys-Murphy
4.1 Abstract

An automated auditory brainstem response (ABR) method-the ALGO-1 Plus-has been developed for hearing screening in healthy neonates. The aim of this study was to test the validity of this automated ABR screening method in at-risk neonates in a neonatal intensive care unit. Two hundred and fifty at-risk neonates were selected for screening according to the criteria of the American Joint Committee on Infant Hearing. All 250 neonates were screened with the ALGO-1 Plus for bilateral hearing loss. When two consecutive screenings pointed to bilateral hearing loss ("refer") further audiological investigations were performed and where necessary therapeutic measures were taken. All children who "passed" the screening unilateral or bilateral enrolled in a nationwide behavioural screening programme at the age of 9 months as well as in a 6-monthly follow up programme documenting speech and language development.

A total of 245 (98%) neonates passed the ALGO-1 screening, 230 (92%) at the first attempt and 15 (6%) at the second attempt. Five (2%) were referred with bilateral hearing loss. One of these died of congenital rubella shortly after screening and bilateral congenital hearing loss of >35 dB was confirmed in the other 4. None of the infants who passed the screening were discovered to have moderate to severe bilateral hearing loss (>40 dB) with behavioural screening (n=183/233) or at follow up (n=233/233). In this study, all at-risk neonates with bilateral congenital hearing loss were detected with ALGO-1 Plus screening. No false-negatives were discovered.

Conclusion: The ALGO-1 Plus infant hearing screener can be used as a valid automated ABR-screener to detect hearing loss in at-risk neonates in a neonatal intensive care setting.

4.2 Introduction

The development of children with bilateral hearing impairment can be improved with early detection (<3 months) so that treatment can start before the age of 6 months. When this is taken into account, behavioural screening for hearing loss at the end of the 1st year (as carried out in many European countries) is rather late, but this was the best available screening method up to now. For the neonatal period, conventional auditory brainstem response (ABR) is considered to be the most reliable method for assessment of the hearing level. Despite the significant incidence (1%-5%) of mild to severe sensorineural hearing impairment in at-risk neonates, the conventional ABR method is not widely used for screening because it needs attendance and it is time-consuming. An automated ABR infant hearing screener (ALGO-1 Plus, Natus Medical Inc, San Carlos, California, USA) is available now, for screening purposes. The sensitivity of this method is
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reportedly 100%, with specificity at 97.6%, in term neonates, compared with the conventional ABR method.\textsuperscript{6,9,13}

The aim of this study was to test the use and validity of this automated ABR screener, in detecting hearing loss in at-risk neonates in a neonatal intensive care setting. Detection of bilateral hearing loss was the goal of this study, because normal speech and language development is still possible in cases of unilateral hearing loss. Follow up data from behavioural screening methods, as well as from speech and language development, are presented to confirm the validity of the automated ABR screening method in this group.

4.3 Subjects and Methods

Newborns admitted to the NICU of the Academic Medical Centre in Amsterdam between July 1992 and December 1993 were eligible for entry into this study, provided they were at risk for hearing loss according to the criteria of the American Joint Committee on Infant Hearing Screening (see Table 4).\textsuperscript{7}

The Academic Medical Centre serves a multi-cultural and multi-lingual population. Neonates are often transferred from other hospitals, either post-natally to the NICU, or pre-natally to the obstetric ward. When intensive care is no longer necessary, the neonates are transferred back to the referring hospital for high care and medium care where they often stay for several weeks before being discharged home.

Informed parental consent was obtained for 250 of the 252 neonates eligible for entry into this study. The median gestational age of the 250 neonates was 30.0 weeks (range 24-42 weeks). The median birth weight was 1350 g (range 570-4395 g). Most of these premature and/or dysmature neonates had signs of respiratory distress syndrome requiring assisted ventilation, circulatory problems, hyperbilirubinaemia, neonatal infection, asphyxia etc.

Table 4  

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 dosis (e.g.: aminoglycosides, diurectis)
- 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
Automated ABR screening (ALGO-1 Plus) was performed in 250 neonates before transfer back to the referring hospital. The screening was performed by three medical students without special audiological background. The portable ALGO-1 Plus screener uses 35 dB near Hearing Level click stimuli, presented mono-aurally at a rate of 37 pulses/s. The clicks have an acoustic spectrum, which is flat from 750-5000 Hz. After artefact rejection for ambient noise and myogenic activity, an internally programmed template-matching algorithm measures ongoing EEG activity for the presence or absence of the ABR. This sampling provides the use of a statistical test, the likelihood ratio. After reaching a likelihood ratio $>160$, the ALGO-1 Plus stops collecting data and displays a “pass” for the ear being tested.\textsuperscript{9,13} This indicates that the data collected were sufficient to discriminate between the presence of a "response + noise", versus the presence of pure noise, or a "no response" condition at better than the 99.80% level of confidence. To avoid disturbance from myogenic activity screening took place whenever possible with the neonate extubated, after feeding and during sleep. Noted were screening results, postnatal age, duration of procedure, and situation during screening (nasal continuous positive airway pressure (CPAP) therapy, position in incubator or cot, disturbance from ambient noise, or from routine technical equipment). Neonates who did not pass the test for both ears were rescreened after 4 weeks or at term. Those who did not pass the second automated ABR screening at least unilaterally were referred for conventional ABR and audiological evaluation. If hearing loss was confirmed, early habilitation was started (see Fig 7).

Those who passed for screening unilateral or bilateral were enrolled in:
1. the 6-monthly follow up programme of the NICU, where reaction to sounds as well as speech and language development were screened according to Egan and Illingworth\textsuperscript{1} and,
2. a nationwide behavioural hearing screening programme (Ewing, or CAPAS) at the age of 9 months\textsuperscript{4,10} in the well baby clinics. Infants who failed repeated behavioural hearing screening were referred to an otorhinolaryngologist for further diagnostic investigations.

4.4 Results

\textit{Neonatal screening with the automated ABR method}
Screening was possible in the incubator in the NICU, even during nasal CPAP oxygen therapy, without disturbance from ambient noise or from technical equipment. One child needed to be tested during artificial ventilation. Mean duration of the procedure was 25 min (SD 13.9), including preparation of the skin, the screening itself and cleaning the material. The three students who performed the screening were also being trained in
The first screening took place when the neonate had been extubated and in a stable circulatory condition, at a median postnatal age of 21 days (range 1-119 days). Of 250 neonates, 230 (92%) passed the screening and 20 had the result "refer" at first screening. Two of these 20 neonates died, and 1 moved abroad, before a second ALGO-1 Plus screening could be performed. In 17 (mean gestational age 27.9 weeks, mean birth weight 1070 g), a second screening resulted in 15 "passes" and 2 "refers". A total of 5 neonates (2%) did not pass either first or second screening. One of these five died from encephalomyopathy due to a respiratory chain disorder (cytochrome C oxidase dysfunction). In this case conventional ABR confirmed a congenital hearing loss >35 dB. Another child died from congenital rubella in the 3rd week of life. In this case conventional ABR could not be performed, but congenital hearing loss was highly probable. One other suffered from a severe form of bronchopulmonary dysplasia and laryngotracheomalacia. It was not possible to detect the hearing level with conventional ABR testing in this infant. When behavioural distraction testing failed at the end of the 1st year of life, further audiological evaluation demonstrated a permanent mild hearing loss of 40 dB. Conventional ABR testing confirmed bilateral hearing loss of >35 dB in the remaining two infants of this group of five. These infants were enrolled in a therapeutic programme, and fitted with hearing aids because of sensorineural hearing loss detected with the ALGO-1 in the neonatal period. Fig 8 shows the results of the ALGO-1 screening.

Fig 7: Flow chart ALGO-1 Plus neonatal hearing screening
Distraction screening in the well baby clinics

Of the 250 children who entered the study, 9 died before the age of 1 year and 5 were lost to follow up. Three of the 250 had already been referred for audiological evaluation because of the results of the ALGO-1 neonatal screening. Although available to all, behavioural distraction screening was performed in only 183 of the remaining 233 children (78.5%) of whom 161 (88%) passed. Twenty-two of 183 (12%) infants failed the behavioural screening after repeated measurements. Sixteen of these 22 infants ultimately turned out to have transient conductive hearing loss. The parents of 6 of these 22 infants refused further diagnostic investigations.

Table 5  
Sensitivity, specificity and predictive values after the first and second ALGO-screening

<table>
<thead>
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<th>First ALGO-1 screening (%)</th>
<th>Second ALGO-1 screening (%)</th>
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<tr>
<td>Negative predictive value</td>
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Follow up in the NICU programme

All 233 infants were enrolled in the follow-up programme of the NICU for at-risk neonates. There were no cases where moderate, severe, or profound (>40 dB) bilateral hearing loss was suspected. Speech and language development were adequate in all infants, according to the method of Egan and Illingworth.¹

When compared with the combined results of the behavioural screening method in the well baby clinics and the normal speech and language development in the NICU follow up programme, the ALGO-1 Plus screening has a sensitivity for congenital sensorineural hearing loss of 100% (n=5). The specificity after the first screening was 94%, after a repeat screening at term 100% (n=233). The positive predictive value was 25% after the first attempt, and in this study 100% after the second screening. The negative predictive value was 100% (Table 5).

In this at-risk group, 4 neonates had moderate to severe bilateral hearing loss. In one other child congenital hearing loss was very probable. Therefore, the incidence of bilateral congenital sensorineural hearing loss was 2%. All were detected with ALGO-1 Plus screening. Three of the five children with bilateral congenital hearing loss were still alive at the age of 1 year.

4.5 Discussion

Neonatal hearing screening is very desirable for at risk infants (Table 4) in whom the 1%-5% incidence of sensorineural hearing loss is tenfold the incidence in the general population.²,⁶,⁷ For logistical reasons it may be necessary to perform screening before the neonate is transferred from the NICU to the referring hospital. At transfer most of the at risk neonates are preterm, and may still be in an incubator with or without nasal CPAP oxygen therapy. This study shows that a major advantage of the ALGO-1 Plus hearing screener is the possibility of testing under NICU conditions, without disturbance by ambient noise.

The reliability of the automated ALGO-1 Plus hearing screener compared with conventional ABR detection in the neonatal period, has been proven in several studies.⁶,⁹,¹³ In this study, follow up data on at-risk neonates, selected according to the AJCH criteria confirmed the validity of the ALGO-1 Plus hearing screener. Although transient evoked otoacoustic emissions have been advocated recently as a first stage technique for universal healthy newborn hearing screening,⁸ several problems have to be solved to make this technique suitable for at risk neonates and for use under NICU conditions.⁵ Our study confirmed the suboptimal cover of the behavioural screening programme (78.8%).¹⁰,¹⁵ Even after being repeatedly referred for behavioural screening, the parents
of 6 of the 22 referrals refused further diagnostic investigations. In contrast, the neonatal hearing screening was accepted by most parents.

When compared with the results of behavioural hearing screening at the age of 1 year and with the results on hearing, speech and language development at further follow up, the specificity of the ALGO-1 Plus test was 94% after the first attempt and 100% after a repeat screening. Maturation phenomena of the central nervous system may be responsible for this difference. 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 2nd year of life. The small group of children who failed at the first screening had a median gestational age of 27 weeks, and the first screening was performed within 3 weeks of birth. Probably, the ABR of the very preterm neonate cannot be compared with the algorithm of the standard ABR, based on data of 30 normally hearing term neonates. To avoid disappointing attempts when screening preterm neonates, it is advised to wait until the neonate is in a stable condition and has reached the post-conceptional age of 30 weeks.

A sensitivity of 100% means that none of the children who passed the ALGO-1 Plus screening has a moderate to severe sensorineural hearing loss at follow up. For screening purposes this high sensitivity is important, especially when combined with a high specificity. All children with a bilateral sensorineural hearing loss were detected in the neonatal period with the ALGO-1 Plus screener, but further investigations are necessary to confirm the high sensitivity with confidence.

Early detection of hearing loss and early habilitation is important for the development of the child with bilateral hearing impairment. The ALGO-1 Plus infant hearing screener can be used in the neonatal intensive care unit as an automated ABR screener to detect hearing loss in at-risk neonates. Our follow up data confirm the validity of this safe and simple screening method.
4.6 References
