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Introduction

1.1 Introduction

"It seems that even a short exposure to language, a brief moment during which the curtain has been lifted and oral communication established, is sufficient to give a child some foundation on which much later language may be based". This assumption of Lenneberg in 1967 marks the beginning of attempts to detect a hearing impaired child as early as possible.[1] Normal hearing during very early life is of utmost importance for laying the basis for speech and language development. Hearing impaired children identified late are at risk of substantial delay in their acquisition of language and communication skills, with consequent longer-term risk to educational achievement, mental health and quality of life.[2-4]

This notion may explain the lasting tradition of behavioural hearing screening in young children in many western countries. In the Netherlands the universal EWING behavioural hearing screening was introduced in 1965. Recently a modification of this test has been developed which uses recorded sounds produced via loud speakers. This is called the Compact Amsterdam Paediatric Audiometric Screener (CAPAS). The EWING/CAPAS hearing screening programme resulted in detection of impaired hearing at a median age of 1.5 year.[5] Children with multiple handicaps, proportionally more widely distributed in neonatal intensive care graduates, are not eligible for the EWING/CAPAS screening. This contributes perhaps to the later age at detection of hearing loss in neonatal intensive care (NICU) graduates. In the Dutch follow up study of preterm and small for gestational age infants (POPS) more than 50% of the children with sensorineural hearing loss were detected later than 2 years of corrected age.[6]

From recent studies it has become clear that a detection age of 1.5 year is too late to achieve optimal development of a hearing impaired child. Present opinion favours the view that hearing loss should be detected by 3 months of age and habilitation should have been started before 6 months of age.[7-9]

The prevalence of congenital hearing loss (CHL) in the normal population is 1:1000. There is a 10 to 20 fold increase of CHL in NICU graduates who fulfil the at risk criteria of the Joint Committee of Infant Hearing (1:100).[7] Despite this high risk, routine determination of the hearing status as part of a more general neurologic evaluation has been a major omission in neonatal intensive care graduates.
Technical developments for hearing screening in the newborn have been warmly welcomed in view of the high prevalence of CHL in NICU graduates and the late age of detection of CHL when the EWING/CAPAS screening method is used. This thesis reflects a period of introduction, investigation, exploration and implementation of neonatal hearing screening in neonatal intensive care, in the Netherlands.

Introduction of the AABR hearing screening method

The aim of neonatal hearing screening is the detection of congenital hearing loss. This sounds easy but there is more to it because the definition of hearing loss is not clear-cut. The risk-benefit ratio of neonatal hearing screening depends critically on this definition. Most commonly, the aim of neonatal hearing screening is to detect what is called "significant hearing loss". This is hearing loss that interferes with sufficient development of spoken language.[10] A bilateral hearing loss of $\geq$ 40 dB is regarded as fulfilling this criterion.[11]

In the late eighties two different methods were developed for neonatal hearing screening: the Automated Auditory Brainstem Response (AABR) hearing screener and a screener based on detection of Oto-Acoustic-Emissions (OAE).

The OAE technology is based on a physiologic phenomenon of the inner ear. OAE's appear to be preneural in origin but the exact origin is still subject to investigations. One of the theories is that OAE's may be a by-product of active movements of the outer hair cells in the healthy inner ear passage which enhance the vibration caused by a sound stimulus. The resulting vibrational energy partially leaks out of the cochlea through the middle ear and can be recorded in the outer ear canal. Using the proper stimulus, OAE's can be detected in 98 % of normal hearing humans. They are absent when there is a hearing impairment of more than 20-40 dB.[12] Measurement of OAE's is not limited by age, but detection of OAE in premature newborns is rather unsuccessful due to the small external ear canal and unfavourable signal-noise ratio.[13]

The Automated Auditory Brainstem Response hearing screener was developed as a screening device. Using a bipolar EEG recording, it detects an auditory brainstem response following a 35 dB HL click stimulus. The built-in algorithm technology, based on ABR recording of normal hearing newborns, produces an objective and reliable interpretation of the response and results in a "pass" or "refer" indication. The AABR hearing screener has a noise and myogenic artefact rejection system.

Theoretically AABR hearing screening is the superior technique because it not only detects hearing loss of preneural origin but also hearing loss due to auditory neuropathy. This may be an advantage in a population at risk for neurologic sequelae. The incidence of central auditory neuropathy is not known.

Investigation

Initially, the introduction of AABR hearing screening on the neonatal ward was met with scepticism. Hearing screening resulting in reliable results in an environment with ambient noise and possible disturbance from technical equipment was considered to be almost

impossible. Therefore, a feasibility study of the use of the ALGO 1 Plus AABR hearing screener device (Natus Medical Inc, California, USA) was started in 1992 in the Academic Medical Centre in Amsterdam. To establish the applicability of this screening method a follow-up study was performed.

Exploration
AABR hearing screening is based on the ABR responses of normal term newborns. Use of this device is advised by the manufacturer from 34 weeks gestational age onwards. Hearing screening in high-risk newborns, mostly preterm infants, should therefore preferably be carried out the end of their stay at the neonatal ward, when their auditory system would be mature enough to ensure reliable results. However, due to limited availability of NICU facilities in the Netherlands, many preterm newborns before 34 weeks postmenstrual age are transferred back to the local hospitals that lack screening facilities.
After exploration of the theoretical background of the developing ABR and the electronic properties of the AABR screener, there seemed to be a place for AABR hearing screening even in very preterm newborns. This resulted in an efficacy study of the use of AABR hearing screening in very preterm newborns.

The next problem to be solved was whether it would be possible to set up AABR hearing screening as part of regular health care for NICU graduates and, finally, to set up a nation-wide AABR hearing screening programme in the NICU's in addition to a future nation-wide neonatal hearing screening programme.

Implementation
General implementation of AABR hearing screening in NICU's should meet screening programme criteria. This involves not only recruitment of structural financial support for training personnel and carrying out the AABR hearing screening in the NICU's, but also setting up a registration system that provides tracking of referred newborns and of monitoring the overall quality of the screening programme.
Our implementation study focused on key outcomes of this programme: capture rate, first stage success rate, pass/refer rates, rescreen compliance, diagnostic referral rates, age at first diagnostic evaluation and prevalence of congenital hearing loss.

This thesis describes the process of evaluating the feasibility and applicability of the AABR neonatal hearing screening method in the NICU and ends with the nation-wide implementation of this method in NICU graduates.

1.2 References

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