Hearing Screening of High Risk Register (HRR) in Neonatal Intensive Care Unit [N.I.C.U]

Thesis submitted for partial fulfillment of master degree in Audiology

Presented by
Marwa Mohamed Saleh EL-Shabrawy
M.B.BCh

Supervised by

Prof.Dr. Ahmed Sameh Farid
Professor of Otolaryngology & Dean of Faculty of Medicine-Cairo University

Prof.Dr. Iman Abdel Salam Seoud
Professor of Pediatrics & Head of Neonatology Department- Faculty of Medicine- Cairo University

Prof.Dr. Zeinab Mohamed Mounir
Professor of Pediatrics in Child Health Department National Research Center

Faculty of Medicine – Cairo University
2007

List of Tables

(14)	Overall result in the second stage screening.	82
(15)	Outcome of second screening test using TEOAE.	84
(16)	Duration in seconds per ear for AABR screening.	85

List of Figures

Contents

INTRODUCTION

Hearing loss is one of the most common congenital anomalies, occurring in approximately 2-4 infants per 1000. Screening only those infants who meet the high-risk register (HRR) criteria is not enough because as many as 50% of infants born with hearing loss have no known risk factors. Reliable screening tests that minimize referral rates and maximize sensitivity and specificity are available (De Michele et al., 2005).

Hearing screening programs are intended to identify individuals who may have, or who are likely to have, hearing problems, thus separating the tested individuals into two groups; those who have normal results (pass) and those who have abnormal results, who need further testing(re-screening or referral) (Johnson, 2002).

Mass screening of hearing in children is based on the concept of secondary prevention. In recognition of congenital or early acquired (i.e. neonatal) hearing disability, numerous hearing screening programmes have been introduced in the world (Parving, 2001).

In contrast to the recommendations of the Joint Committee on Infant Hearing, neonatal hearing screening programs are still not universally available, and many countries implement elective screening in high-risk newborns (Korres et al., 2005).

It was noted that detection of hearing loss at earlier ages will improve the success of rehabilitation programs (Turan and Apaydin, 2002). Moreover, the recent studies supported by National Institute of Health (NIH) have concluded that children whose hearing loss is identified and who receive appropriate intervention prior to 6 months of age, develop significantly better language ability than whose who are identified later

(Still, 1999). The same observations were stated by Hall (2000), regardless the degree of hearing loss. On the other hand, delayed identification and management of severe to profound hearing impairment may impede the child ability to adapt to life in a hearing world or in the deaf community (Dort et al., 2001).

The frequency of various causes of hearing loss in children has changed over the past thirty years and will continue to change as newborn hearing screening becomes available and as more ways develop to prevent hearing loss. Hereditary causes and neonatal intensive care unit (NICU) graduates are contributing now to a major part of the causes of hearing loss in children. The category of NICU graduates easily identifies a group of children who are at risk for hearing loss since they are exposed to a unique health experience (Roizen, 1999).

Automated Auditory Brain Stem Response (AABR) has been used for a targeted screening of neonates at risk of having a sensorineural hearing loss (SNHL) (Homer et al., 2000). Transient Evoked Otoacoustic Emissions (TEOAEs) on the other hand, have been used to screen newborns for hearing loss for several years. Since TEOAEs are present in over 95% of population, they could appear to be an appropriate parameter to measure in a screening program (Stevens et al., 1991 and Maxon et al., 1995). The existence of OAEs excludes with high probability a peripheral hearing loss, which can influence the development of speech, yet it can not detect the hearing threshold and can not exclude retrocochlear pathology (Schorn, 1993).

In particular, emissions in children are stronger and easier to measure than in adults (Kimberely and Nelson, 1989). So it is a fast and

non invasive technique to screen auditory dysfunction in high risk newborns (Plinkert et al., 1990).

When combining auditory brain stem response to otoacoustic emissions test, in evaluation of hearing impairment gives very highly sensitive and specific results (Mason and Herrmann, 1998).

DISCUSSION

Infant hearing impairment stands as the most common congenital sensory disorder, it is considered to be 20 times more prevalent in neonates than other disorders that are routinely screened for, including phenlyketonuria, sickle cell anemia, and hypothyroidism (Oghalai et al., 2002). Moreover the negative impact of late detection of congenital or prelingual hearing loss on spoken language and cognitive skills development has long been confirmed by several studies (Yoshinaga-Itano et al., 1998; O'Donoghue, 2000).

Objective physiological measures must be used to detect newborns and very young infants with hearing loss. Current physiologic measures include otoacoustic emissions (OAE), transient-evoked (TEOAE) or distortion-product (DPOAE), and/or auditory brainstem response (ABR). Both OAE and ABR technologies have been successfully implemented for Universal Newborn Hearing Screening (UNHS) programs (Finitzo et al., 1998).The goal of these neonatal hearing screening programs is to identify hearing loss as early in life as possible allowing the initiation of appropriate treatment and/or rehabilitation thereby preventing the demonstrated educational, social, communicative consequences of hearing loss (Kerschner, 2004).

The Joint Committee on Infant Hearing (JCIH) recommends a "1-3-6" plan: all infants should be screened prior to **1** month of age, preferably before discharge; diagnostics should be completed by **3** months of age for those referred after re-screening; and, for those with confirmed hearing loss, intervention should be initiated by **6** months of age (JCIH, 2000).

Prior to the implementation of the UNHS programs the average age of diagnosis of hearing loss was reported to be between 14 months to 2.5 years (American Academy of Pediatrics (AAP) Task Force on newborn and Infant Hearing, 1999). This led to an average delay in the onset of intervention and many cases started to learn spoken language by the age of 3 years. Such delays in diagnosis and intervention have been linked to irreversible delays and/or deficits in speech and language development (Hall, 2000).

The present study was carried out on 130 newborn, hearing screening was done using TEOAE. Out of 130 neonates, 30 had no risk factors of hearing loss (60 ears); they were screened in well-baby nursery in the maternal hospital, faculty of medicine Cairo university using TEOAE, their age ranged from 1-4 days (Table 2),15 male and 15 female (Table 5). This screening revealed that 26 (86.7%) neonate had passed the test and 4 (13.3%) had a refer result (Table 3). This prevalence of failure among well-baby nursery was higher than cited by many authors (White, 1996; Bonfils et al., 1998; Bener et al., 2005) they found prevalence rates of hearing loss ranging from 0≈5 %. However, all the previous authors had screened their cases after the second day of life.

The results were completely different when TEOAE was recorded in the first 48 hours of life as reported by Levi et al., (1997) they found a failure rate of 22.5% when screening neonate aged 10-48 hours. Another study by Kok et al., (1993) revealed failure rate of 22% when recording TEOAE within the first 30 hours of life. When the test was repeated after elapsing 108 hours from life this failure rate dropped to about 1%. This could be explained by the presence of vernix caseosa in

the external canal or effusion in the middle ear, which could be a residual amniotic fluid. As result of this the majority of authors agreed to do screening for hearing loss after the first 48 hours of life and to be associated with otoscopic examination and external ear canal cleaning or suction when necessary (Norton et al., 2000).

The other one hundred neonates were screened in the Neonatal Intensive Care Unit (NICU) in El-Monuria Children Hospital, Cairo University. They had at least one of the risk factors or indicators for hearing loss listed by the JCIH (1994). The screening was done to those HRR neonates between 3-23 days of life, a time by which the vernix and amniotic fluid have cleared spontaneously from the ear, and inspection was done before screening to check the external ear canal, otoscopic examination was planed but was not possible in all cases, beside we intended testing the baby in a state of natural sleep, tendency to perform otoscopic examination would wake them up and accordingly the test could not be completed. However, despite all these measures, still some fallacies were encountered during conducting the survey in the NICU. These included the noise present in the field of testing, internal noise of the newborn resulting from breathing sound and heart beats and the activity state of the newborn. To overcome the problem of noise, the test was done in a quiet room whenever possible. The test was carried out while the newborn was in state of quiet sleep or at least alert and quiet, no sedatives were used in this study.

Many authors stated that despite all these measures, still some fallacies could not be avoided. The most important was the presence of otitis media or middle ear effusion especially among the preterm population which had an incidence in the literature ranging from 6.5 %

according to Smurziniski et al., (1993), 13.8% as reported by Veen et al., (1993) and as high as 21.1% according to Stevens et al., (1991).

In the presence study, 29 % of infants got Refer result in the initial screening (Table 8) and the final percentage of failure (after adjustment for dropouts) was 15.4%. Among the Egyptian NICU neonates El-Danasoury et al., (2003) reported the presence of hearing impairment in 19.1 % of HHRs. Makky and Rashid (2004) found 24 % hearing affection in the HHRs. Also El-Gamal et al., (2001) reported hearing loss in 20% of NICU neonates. Gupta et al., (1991) demonstrated a 30.88% hearing loss among NICU graduates in India. On the other hand, a lower prevalence of hearing loss was reported; it varied between 1.5 to 7 % in the HHRs (Veen et al., 1993; Korres et al., 2005). The difference of prevalence of hearing loss among different studies could be attributed to the variants of each group, regarding the risk factors present, their number and associated general condition.

Although risk factors for hearing loss have been used for screening in many countries world wide, these factors are constantly refined by the JCIH. However, we should not consider these factors as the "gold standard" with same relative importance because the situation in different countries or time periods may vary considerably. For example, a developing country may use more ototoxic agents, without assessing drug blood levels because of the lack of resources; whereas another developed country may not use such agents. In contrast, neonates who could have died as a result of severe prematurity and birth complications in a remote center with limited funding and equipment may very well have survived in a large state-of-the-art neonatal intensive care unit. Therefore, continuous investigation of the relative

importance of high-risk factors of hearing loss is essential to assess, refine these factors and modify accordingly to the current clinical practice. Moreover, such studies are very useful when funding is limited, and screening should be targeted to the most vulnerable neonates (Korres et al., 2005).

In this work, the high risk newborns were categorized according to the number of risk factors in every high risk neonate into: Groups with two risk factors and groups with multiple risk factors. The prevalence of hearing loss in those with 2 risk factors was 11.1% while those with multiple risk factors had a referral rate about 44.45% (Table 15). So it is clear that infants who suffer many clinical adverse effects during their neonatal course, are more liable to develop hearing impairment. This was supported by several studies; El-Gamal et al., (2001) studied 90 cases selected from NICU. They reported a failure rate of 54% in group with multiple risk factors and 20% in group with single risk factor. A lower rate was found by El-Danasoury et al., (2003) which were 27.7% in multiple risk group and 14.3 in single risk group, such discrepancy was due to lack of sharp demarcation between the groups in which was due to defective data of the newborn.

However, this observation of multiple risk factors and prevalence of hearing loss should be taken cautiously as the number of infants in the multiple risk groups ranged from 2-3 cases only in this study and the other mentioned studies. So this combination of factors should be studied on a larger scale in order to get more accurate percentages (Korres et al., 2005).

The most frequent risk factor encountered in the NICU was ototoxicity (100%), followed by hyperbilirubinaemia (55%) and then low birth weight (14.5%) and mechanical ventilation for more than 5 days (11.5%) and lastly craniofacial anomalies (1%) (Table 10). Vohr et al., (2000) found that the four most frequent risk factors in the NICU were ototoxic drugs, low birth weight, mechanical ventilation for more than 5 days and low Apgar score. Korres et al., (2005) found that toxic levels of ototoxic drugs, mechanical ventilation for more than 24 hours, prematurity and LBW were the four frequent risk factors. However, there a slight difference between the two studies, yet ototoxic drugs, mechanical ventilation and LBW are still the three most frequent risk factors in both studies. This fact becomes more important if we consider that the study by Vohr et al., assessed children born in the United States between 1994 and 1996, whereas the study done by Korres et al., assessed children born seven years later, in south Europe (Greece), highlighting the constant and universal frequency of these factors in the developed world.

On the other hand, hyperbilirubinaemia was the most frequent factor encountered in our study and other studies carried out in Egypt (Makky and Rashid, 2004; El-Danasoury et al., 2003; El-Gamal et al., 2001). This is a point which needs further research in coordination with pediatricians to assess its magnitude and effect, as neonatal jaundice are more likely to develop central rather then peripheral hearing loss. This necessitates the combination of TEOAE and ABR in cases with neonatal jaundice (Hazaa and El-Danasoury, 1995; Jakobikova et al., 2003).