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NEWBORN HEARING SCREENING IN THE SPECIAL CARE NURSERY OF HOSPITAL UNIVERSITI SAINS MALAYSIA KOTA BHARU

By

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DEDICATION

I would like to dedicate this work to those whose contribution never ceased in their presence and in their absence:

The dear memory of my father (late), my mother, my wife and my daughters Sarah and Shaima'a.

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LIST OF ABBREVIATIONS

ABR	Audiometry brainstem response
dB	Decibel
HL	Hearing Loss
HUSM	Hospital Universiti Sains Malaysia
Hz	Hertz
IDT	Infant Distraction Test
ILO	Institute of Laryngology and Otology
JCIH	Joint Committee on Infant Hearing
NIDCD	National Institute of Deafness and Communication Disorder
OAEs	Otoacoustic Emissions
OHCs	Outer Hair Cells
ORL	Otorhinolaryngology
SCN	Special Care Nursery
TEOAEs	Transient Evoked Otoacoustic Emissions
WHO	World Health Organization
SNHL	Sensorineural hearing loss
NICU	Neonatal intensive care unit

ABSTRACT

Neonatal hearing loss is a common health problem. Early detection, identification and management prevent the development of hearing loss and its consequences such as delay in speech and language development. The prevalence of sensorineural hearing loss among all newborns has been reported to be 1 - 2 per 1000, whereas it rises to 1-2 per 100 newborns in with high-risk criteria. Recent research provides evidence for the value of transient-evoked otoacoustic emissions (TEOAEs) in neonatal hearing screening. This study is an experimental hearing screening using TEOAEs, carried out at the special care nursery, Hospital Universiti Sains Malaysia aimed at detecting hearing loss in newborns, to determine the prevalence and size of the problem. A total number of 530 neonates from the special care nursery were screened with TEOAEs using a two-stage process. The first test was performed prior to discharge. Those who failed the first test were re-screened after 4 weeks. Those who did not pass the second-stage TEOAE screening were referred for diagnostic audiological evaluation for confirmation of hearing loss. All newborns detected with hearing loss were found to be among high-risk group, in which 2 had neonatal jaundice, 1 with hydrocephalus, 1 with sepsis and 1 with premature birth only failed on the left ear. The prevalence of hearing loss obtained in this study was 5 (0.94%) out of 530 newborns.

ABSTRAK

Pengesanan awal dan pengurusan terhadap masalah pendengaran terhadap bayi baharu lahir adalah mustahak. Memandangkan implikasi masalah pendengaran di kalangan bayi baru lahir ini boleh mengakibatkan masalah dalam perkembangan berbahasa dan pertuturan. Kadar prevelan masalah pendengaran saraf di kalangan bayi baru lahir ialah 1 hingga 2 bagi setiap 1000 kelahiran, di mana ia meningkat di kalangan kumpulan yang berisiko tinggi hingga 1-2 bagi setiap 100 bayi baru lahir. Kajian ini di jalankan terhadap bayi baru lahir yang terdapat di dalam SCN HUSM sebelum mereka discaj. Ujian saringan pendengaran menggunakan transient-evoked otoacoustic emissions (TEOAEs) yang terbahagi kepada dua proses. Proses pertama ialah seramai 530 bayi dari SCN di uji dengan saringan pertama kali. Bagi bayi yang telah dikenal pasti gagal dalam ujian pendengaran perlu mengikuti pengujian semula selepas 4 minggu dan seterusnya dirujuk untuk penilaian audiologikal diagnostik. Semua bayi yang mempunyai masalah pendengaran adalah dikalangan risiko tinggi, 2 orang mempunyai masalah jaundis neonatal, seorang hydrocephalus, seorang sepsis, dan seorang lahri kurang matang yang gagal di sebelah telinga. Keputusan kajian dengan kadar prevelan 5(0.49%) dari 530orang bayi.

CHAPTER I

INTRODUCTION

Hearing is one of the very important five senses. Helen Keller called deafness "A worse misfortune than being blind because when you lose your vision you lose contact with things, when you lose your hearing you lose contact with people". Hearing loss is invisible, and usually occurs gradually. It can mimic forgetfulness, inattentiveness or mental dullness.

Normal speech and language development depend upon a child's ability to hear spoken language. Early infancy is the most appropriate time for a child to acquire the foundation of language and communication. The most important period for language and speech development is generally regarded as the first 3 years of life. Therefore, early detection and early identification of hearing loss is very important. This should be followed by a timely and effective therapeutic intervention and rehabilitation programs to minimize the negative effects of hearing loss on the development of cognitive, psychosocial and verbal communication skills and social interactions. (National Institute of Hearing, 1993).

Significant hearing impairment is one of the most common major abnormalities present at birth and, if undetected, will impede speech, language, and cognitive development. Universal detection of infant hearing loss requires universal screening of all infants. Shannon et al., 1984, Watkin et al., 1991, Watson et al., 1996, Hess et al., 1998, Meyer et al., 1999) studies indicate that screening by high-risk registry alone (e.g., family history of deafness) can only identify about 50 % of newborns with significant congenital hearing loss. Reliance on physician observation and/or parental recognition has not been successful in the past in detecting significant hearing loss in the first year of life (AAP and JCIH1994). The impact of hearing loss on early language development

has been well documented. Although published studies on efficacy of early intervention are more limited, the majority demonstrates that children with hearing loss who received early intervention had higher score of improvement on acquired language than those not connected to service early (NIDCD 1997).

A study by (Yoshinagaitano et al., 1998 and Gopal et al., 2000) indicated that early identification followed by proper intervention as early as 6 months of age results in essentially normal language acquisition later on and minimize the negative effects of hearing loss. In contrast, a delay in detection of up to 2 to 4 years may result in abnormal language acquisition. Hearing loss in newborns and infants are not readily detectable by routine clinical procedures (behavioral observation), although parents often report the suspicion of hearing loss, inattention, or erratic response to sound before hearing loss is confirmed (Arehart et al., 1998). Hearing impairment may be conductive or sensorineural or a combination of the two (mixed) hearing loss. Hearing impairment affects one or both ears, varies from mild to profound in degree. It may be congenital, acquired, transient, fluctuating, recurrent, progressive or permanent (Kinney et al., 2000).

1.1 Prevalence of Hearing Impairment

According to the World Health Organization (WHO) deafness and hearing impairment are common health problems throughout the world. Referring to WHO's most recent estimate (2001), 250 million people in the world have disabling hearing impairment of moderate or worse nature. Two-thirds of these people live in developing countries.

WHO estimates that around seven million children in developing countries have disabling hearing difficulties Many more have mild hearing difficulties (WHO, PDHI. 2001).

In the USA, it has been estimated that approximately one per thousand children suffers from hearing impairment. Many more are born with less severe degrees of hearing impairment while others develop hearing impairment during their life (NIH statement1993). In France, a prevalence of 1.4 / 1,000 would represent a total of 1000 deaf infants every year, (with reference to about 700,000 births / year). It suggests that universal screening programs would substantially increase the rate of early-identified infants with significant hearing impairment (Aidan et al., 1999).

Watson et al., (1996) noted a prevalence of 1-2 per thousand live births having significant permanent hearing loss averaging 50 dB in the speech frequencies. Significant bilateral hearing loss is present in about 1 to 3 per 1000 newborn infants in the well baby nursery population, and in about 2 to 4 per 100 infants in the intensive care unit population (Erenberg, et al., 1999). However, the most disturbing fact is that over 50 % of hearing loss may be prevented. The majority of people with hearing problems live in the developing world where there are limited resources and facilities for diagnosis and management of hearing problems. What is even worse is that there are no reliable data available to make a precise assessment of the problem (WHO, 1995). In the Asian Pacific Congress on deafness held in Beijing in August 1998 it was reported that in China, 24 million out of 1.2 billion people suffer from hearing problems. There are 1.5 million children in school for the deaf, half of them are congenital hearing loss (Prasansuk et al., 2000).

Robinette et al., (1997) cited a number of prevalence studies of newborn hearing loss that ranges from 1 per thousand live births to 6 per thousand live births. The prevalence estimated for hearing impairment varied depending on age and criteria used in the diagnosis. Severe congenitally and/or prelingually acquired losses range from 1–3 per thousand live births. .

1.2 Delay of Detection of Hearing Impairment

Hearing loss is often not suspected by the parents or the pediatrician until language development is significantly delayed. Currently, in USA the average age of detection of significant hearing loss is about 14 months. A study by Yoshinaga-Itano (1998) demonstrated that any intervention after the age of 6 months will most likely yield less than optimal speech and language development. Generally, the period between the first 6 months of life and 18 months has been widely postulated as the critical phase for this development. The American Academy of Pediatrics supports the statement of the Joint Committee on Infant Hearing (1994), which endorses the goal of universal detection of hearing loss in infants before 3 months of age, with appropriate intervention no later than 6 months of age.

In a recent article by Daniels, (2001) found that approximately 840 children are born each year in the United Kingdom with profound hearing loss in both ears. Around 400 children with hearing loss are not detected until they are over 1 year old and a further 200 remain undetected until the age of over 3 years. This often leads to lower educational achievements and a poorer quality of life. In a recent study in the United

States of America, it has been shown that more than 90% of children with hearing loss identified by neonatal hearing screening developed a normal range of vocabulary ability in the first 3 years of life. Only 40 % of parents of children with a hearing loss of at least 55 dB noticed behavioral indicators before the age of 3 months. Another study found that less than 10% of parents of infant with hearing loss between 40 and 80 dB had concerns about their child's hearing at the time the hearing loss was diagnosed (Garganta et al., 2000).

1.3 Prevalence of Hearing Impairment In Malaysia

In Malaysia, two previous studies regarding hearing impairments have been performed. The first study was carried out in 1984 by (Said and Abdullah 1984) utilizing 38 school children attending classes for the hearing impaired. It was found out that the majority (75%) of cases were identified as having hearing impairment after the age of one year and the remaining children were detected after 3 year's of age.

The second study was carried out by (Maisarah et al., 1992) and involved children with sensorineural hearing loss attending ENT clinic of National University of Malaysia during the period extending from January to December 1990. In the majority of cases the diagnoses were confirmed at the ages of 3 to 5 years. In the remaining cases, hearing impairment was confirmed after 7 years of age. Only about 25% of cases had been confirmed before the age of 2 years.

These studies showed that confirmation of hearing loss and its rehabilitation in Malaysia are somewhat delayed. Such delay does not comply with the recommendations of the Joint Committee of Infant Hearing. The studies also emphasize that all infants with significant hearing loss should be identified by the age of 3 months and receive intervention by the age of 6 month before they reach a critical period in their development.

A pilot project for the national early hearing screening program has been decided after a meeting held in Kuala Lumpur in early this year (2001). This project will be conducted in a few selected districts in Selangor and Kelantan. The screening will be configured around distraction test perform by the nurses to the children age 6-12 months.

1.4 Recommendations For Hearing Screening

The Joint Committee on Infant Hearing has recommended that infants with significant hearing loss should be identified by 3 months of age, and received intervention by 6 month of age. The European consensus statement of hearing screening program (1998) confirmed that neonatal hearing screening should be considered to be the first part of a program of rehabilitation of children with hearing impairment. This includes the availability of facilities for diagnosis and assessment. In considering the implementation of hearing screening program we should also look into the efficiency of the different screening methods including the sensitivity and specificity, the cost price, need for training, time consumed, and resources needed. Screening without having a rehabilitation program can also be considered as a west of resources.

1.5 Screening Methods For Hearing Impairment

During the past, infant hearing screening has been attempted using a number of different test methods classified as behavioral audiometry and measurement of acoustic reflexes. For the past 15 years, electrophysiological methods are most commonly used which include auditory brain stem response (ABR.). More recently, attention turned to measurement of otoacoustic emission (OAE), which seem to be a promising method since it is fast, inexpensive, and a noninvasive test of the cochlear function. That is why we have chosen OAE as the method of choice in our study.

Infant hearing screening was started in the USA more than 30 year's ago by (Downs and Sterritt, 1964) using behavioral audiometric 'arousal' technique. High rates of false positives and false negatives were detected, according to the Joint Committee On Infant Hearing, and recommended the alternative use of audiometry tests for infants with high-risk criteria. Low sensitivity and specificity in conventional screening procedures such as the arousal technique apparatus render the technique suitable for screening only and not for diagnostic procedures. In 1988 Screening for hearing impairment in infancy in most districts in the United Kingdom was done with infant distraction test (IDT) at 7 to 8 months of age, a targeted high risk babies Johnson et al., (1990) reported the distraction test was sensitive (91%) but non-specific (82%) in the high-risk population. The effectiveness of the screening program was limited. Recently, the use of TEOAEs together with ABR was shown to be reliable and high sensitivity and specificity in universal hearing screening programs. The two techniques (TEOAEs and ABR) showing maximal promise as universal screening tools for the newborn, each has its unique advantages and disadvantages (Geert De Ceulaer et al., 1999).

1.6 Definition of OAE

The normal cochlea does not just receive sound. It also produces low-intensity sounds called OAEs. Otoacoustic emission is the sound emitted by the cochlea generated by motion of outer hair cell that can be recorded within the external canal although they occur spontaneously in 50% to 60% of ears (Parving, 1999). Otoacoustic emissions, though their name suggests a unity, cannot be considered to be a single phenomenon. Different types of emissions can be distinguished on the basis of the type of stimulus and of the latency onset with respect to the stimulus onset.

1.7 Classification of OAE Types

The phenomena of acoustic emission can be observed by various methods, this it can be classified into;

1.7.1 Transient Evoked Otoacoustic Emissions (TEOAE)

Kemp (1978) used a transient excitation to measure the OAEs. He found that after 5 ms post stimulus the original excitation had decayed to a negligible level, but a slowly decaying response component was present between 5 and 20 ms post stimulus. This OAEs has been termed the transient evoked OAEs, or delayed OAE, and is commonly referred to as the cochlear echo. Clicks are the most commonly used stimuli (tone-burst stimuli may be used). Most commonly, 80- to 85-decibel (dB) SPL stimuli are used

clinically. Stimulation rate is less than 60 stimuli per second. TOAEs generally occur at frequencies between 500-4000 Hz.

1.7.2 Distortion Product Otoacoustic Emissions (DPOAE)

Sounds emitted in response to 2 stimulations tones of different frequencies. That is the emissions have components at a frequency, which is not present in the stimulation. The lower tone is usually the F1 and the higher tone the F2. The relative merits of TEOAEs and DPOAEs are widely discussed. Essentially, DPOAEs allow greater frequency specificity and can be used to record at higher frequencies than TEOAEs. DPOAEs has been introduced recently in hearing screening though most screening OAE machines use the transient evoked OAEs.

1.7.3 Stimulus Frequency Otoacoustic Emissions (SFOAE)

Emissions can be evoked at the stimulus frequency by continuous tone. In this method of observation the detailed amplitude and phase variations of the sound in the ear canal are monitored in relation to frequency stimulus. This is caused by the emission interacting with the stimulus, producing cancellation and addition with the stimulus tone. (Wilson, 1980) used a lock-in analyzer to measure the stimulus frequency OAEs from several subjects, and concluded, from measurement of the emission delay, that it must be a function of the cochlea.

1.7.4 Spontaneous Otoacoustic Emissions (Soaes)

Gold (1948) hypothesized that the same active mechanism in the ear, which overcame the damping of the membrane resonance, could result in a spontaneous emission if the positive feed back was too high. Such emission has been found to exist. Several investigators have shown the presence of spontaneous emissions in 30-40% of normal ears.

Only the first 2 types are currently used clinically. Transient evoked otoacoustic emissions are a major subclass of evoked OAEs, because these responses are commonly elicited by the use of brief acoustic stimuli. Commonly used transient stimuli are clicks, single sinusoids, or tone bursts. A major condition to register these emissions, elicited by different stimuli, in the outer ear canal is the reverse conductance of the vibratory energy from the cochlea, through the middle ear (ossicular chain, tympanic membrane) and the outer ear canal. In the outer ear canal, this vibratory energy is transformed to acoustic energy by using the tympanic membrane as a kind of loudspeaker. In our study we used the TEOAEs (Echocheck) using click Stimulus tone bursts.

1.8 Limitation of OAE

Spector, et al., (1991) reported that TEOAEs limitations is due to their inability to provide good frequency specific information, because the click is a wide-band signal which stimulates the entire cochlea. Another limitation of TEOAEs is that it cannot quantify the degree of hearing loss of the subjects. It is well documented that OAE testing has a high false positive rate (up to 15.6%) in the first 24 hours of life, falling to

about 4 % by 72 hours. Some of this is related to middle ear effusion and debris in the external ear canal, and it may also be related to neurological immaturity. According to the American Academy of Pediatrics, the recommended median age of testing is 48 hours, thereby eliminating any early neonatal problems (Kei et al., 1997).

1.9 Prevalence of OAE

From the first report of OAEs it was found that they were present in normal ears but were absent in cases of deafness. For otoacoustic emissions to be an effective indicator of normal physiology. Kemp, (1978), & Johnsen and Elberling (1982) found that emissions occurred for the entire subject they tested with normal ear 100%. It is apparent that OAE has a high prevalence, but not all the researchers were able to measure emissions in all normal subjects tested. Dijk et al., (1987) found emissions present in 85% of the 210 normal subjects tested. Although all these studies used transient stimuli to evoke the emissions.

1.10 Clinical Applications of OAE

The clinical applications of otoacoustic emissions are mainly focused on the identification of sensorineural losses in the auditory periphery. Despite the fact that the otoacoustic emissions signals are affected by alterations in the sound transmission chain (outer ear to middle ear and middle ear to outer ear) there are no current applications based on the transmission loss concept. The presence of OAEs provides direct evidence of the existence of an active mechanism in the cochlea. Otoacoustic emissions have

potential for the study of the detailed mechanical function of the cochlea in a noninvasive and objective manner otoacoustic emissions have potential clinical importance and will function in the near future as a supplement to other standard clinical methods. Therefore measurement of otoacoustic emissions in neonates and young infant is rapidly becoming widespread.

1.11 Role of OAE In Neonatal Hearing Screening

Rutten, (1980) concluded that physiology vulnerability of the OAEs seems important for early detection of progressive hearing loss. Kemp suggest that the potential application of OAEs is the registration of the detailed otoacoustic parameters of the patient for future use indicating early changes in the ear. The OAE test has possible application such as;

- The patients with handicaps children in special school.
- Neonatal hearing screening (targeted or universal)
- Children hearing screening
- Monitoring of the course of a potentially ototoxic medications
- Noise induces hearing loss monitoring in industrial, and or military environment.
- Differential diagnoses (between OAE present and ABR altered).

1.12 Effect of The Ear Pathology of The Presence of OAE

Many studies have been performed on the occurrence of OAE in abnormal ears. Kemp, (1978) found no emissions in subjects who had best threshold of greater than 30 dB HL. (Rutten 1980) found that if an OAE was present at a given frequency, then the audiogram threshold at this same frequency was better than 15 dBHL.

Bray and Kemp, (1987) found that subjects with conductive losses, due to diseased middle ears had no measurable. Even though the cochlea may well have been functioning normally, the poor transmission of the emission, from the cochlear to the eardrum, resulted in the emission being immeasurably small. In addition, the stimulus is also attenuated as it is propagated from the ear canal to the cochlear, and as a result the cochlear receives less stimulation.

Anderson & Kemp (1979) and Johnsen & Elberling, (1982), have both investigated the effect of ototoxic drugs on the OAE. Johnsen & Elberling induced a flat sensorineural hearing loss of 25-30 dB HL using serum salicyate. They found that the emission virtually disappeared. However, after 2 days complete recovery of the emission occurred. Anderson & Kemp used injection of both furosemide and ethacryic acid in laboratory primates to study the effect on the emission caused by these drugs. They found that administration of each drug caused a substantial reduction of the emission intensity, within minutes followed by some degree of recovery (within hours).

1.13 Anatomy and Physiology Underlyng of The OAE

Because OAEs may be new to some clinicians, a brief review of the relevant anatomy and physiology is provided. When sound is used to elicit an emission, it is transmitted through the outer ear, where the auditory stimulus is converted from an acoustic signal to a mechanical signal at the tympanic membrane and is transmitted through the middle ear ossicles; the stapes footplate moves at the oval window causing a traveling wave in the fluid filled cochlea. The cochlear fluid's traveling wave moves the basilar membrane; each portion of the basilar membrane is maximally sensitive to only a limited frequency range. The arrangement is a tonotopic gradient. Regions closest to the oval window are more sensitive to high-frequency stimuli. Those regions further away are most sensitive to lower-frequency stimuli for OAEs, therefore, the first responses returned and recorded by the probe microphone emanate from the highest-frequency cochlear regions, because the travel distance is shorter. Responses from the lower-frequency regions, closer to the cochlear apex, arrive later. When the basilar membrane moves, the hair cells are set into motion and an electromechanical response is elicited, while an afferent signal is transmitted and an efferent signal is emitted. The efferent signal is transmitted back through the auditory pathway, and the signal is measured in the outer ear canal. As described above, the responses from the high-frequency region arrive first, progressively followed by responses from lower-frequency regions. Outer hair cells are located in the Organ of Corti on the basilar membrane. These hair cells are motile; an electrochemical response elicits a motoric response. The 3 rows of outer hair cells have stereocilia arranged in a 'W' formation. The stereocilia are linked to each other and, therefore, move as a unit. These are the outer hair cells believed to underlie OAEs generation. The ear canal supports (resonates or enhances) sound vibrations best

at the frequencies, which the human ears hear most sharply. This resonance amplifies the variations of air pressure that make up sound waves, placing a peak pressure directly at the eardrum. For frequencies between approximately 2 KHz and 5.5 KHz, the sound pressure level at the eardrum is approximately 10 times the pressure of the sound at the auricle. There are two types of nerves at the base of the hair cells: "afferent nerve fibers" carry sensory information away from the cells to the brain while "efferent nerve fibers" bring information from the brain to the hair cells. These afferent neural pulses are then collected and sent out the internal acoustic meatus via the auditory nerve thus translating mechanical information into neural information. Once the auditory nerve has received the neural impulses, it continues the signal through various pathways in the brainstem. From the auditory nerve, signal information sent to the cochlear nucleus, then proceeds to the superior olivary complex, to the lateral lemniscus, to the inferior colliculus, and to the medial geniculate body, until reaches its final resting place in the brain, the auditory cortex. The auditory cortex then interprets the signal into sound where, from previous experience, we are able to understand what that sound represents.

1.14 Signal Morphology of TEOAE

A typical TEOAEs signal consists of acoustic "burst spindles", the main frequencies, which decrease with increasing time distance from the stimulus. This phenomenon is caused by the tonotopic organization of the cochlea. High frequency components of the acoustic input signals stimulate the more basal parts of the cochlea and cause the earliest responses as a result of the traveling wave whereas the lower frequency components stimulate the apical hair cells.

1.15 Hearing Loss

The Aetiology

Hearing loss can be broadly defined as the decreased ability to receive or process acoustic stimuli. There are many causes of hearing loss in newborns. Some may be temporary and easily corrected for example, a blockage in the ear canal, or fluid in the middle ear may cause a hearing loss. Some hearing loss is permanent and may only be corrected by hearing aids or other listening devices. Some infections that mothers may have during pregnancy, such as Rubella, herpes, may cause an infant's hearing loss at birth. Hearing loss may also be passed on in families. Sometimes there is no known cause for hearing loss in newborns.

1.16 Types of Hearing Loss

There are three basis types of hearing loss;

A - Conductive hearing loss

B - Sensorineural, hearing loss

C - Mixed hearing loss.

1.16.1 Conductive Hearing Loss

Conductive hearing loss occurs when sound is not conducted efficiently through the outer and middle ears, including the ear canal, eardrum, and the tiny bones, or ossicles,

of the middle ear. Conductive hearing loss usually involves a reduction in sound level, or the ability to hear faint sounds. This type of hearing loss can often be corrected through medicine or surgery. Absence or malformation of the pinna, ear canal, or ossicles can cause a conductive hearing loss. Presence of a foreign body; impacted ear wax (cerumen) fluid in the ear associated with colds, allergies, ear infections (otitis media) or a poorly functioning eustachian tube are all examples of conditions that may cause a conductive hearing loss.

1.16.2 Sensorineural Hearing Loss

Sensorineural hearing loss occurs when there is damage to the inner ear (cochlea) or to the nerve pathways from the inner ear (retrocochlear pathway of the acoustic nerve) to the brain. Sensorineural hearing loss not only involves a reduction in sound level or ability to hear faint sounds, but also affects speech understanding or ability to hear clearly. Sensorieneural hearing loss can be caused by diseases, birth injury, drugs that are toxic to the auditory system, and genetic disorder with or with out syndromes. Sensorineural hearing loss may also occur as a result of noise exposure, viruses, head trauma, aging, and tumors. Sensorineural hearing loss cannot be corrected medically or surgically, it is a permanent loss.

1.16.3 Mixed Hearing Loss

Sensorineural hearing loss occurs in combination with a conductive hearing loss. In other words there may be damage in the outer or middle ear and the cochlea or auditory nerve. When this occurs, the hearing loss is referred to as a mixed hearing loss.

1.17 Degree of Hearing Loss

Degree of hearing loss refers to the severity of the loss. There are 5 categories that are typically used. The numerical values are based on the average of the hearing loss at three frequencies 500 Hz, 1000 Hz, and 2000 Hz in the better ear without amplification.

Degree of hearing loss accordingly to the (WHO) classification;

1- Normal no impairment = 0 - 25 dB (better ear).

2- Mild impairment = 26- 40 dB (better ear).

3- Moderate impairment = 41- 60 dB (better ear).

4- Severe impairment = 61- 80 dB (better ear).

5- Profound impairment = 81 dB or greater (better ear).

1.18 High Risk Criteria

According to Joint Committee on Infant Hearing and American Academy of Pediatrics, followings are the high-risk criteria;

1. Family history of hereditary childhood sensorinural hearing loss
2. In utero infections such as toxoplasmosis, cytomegaly, rubella, herpes simplex and syphilis.
3. Craniofacial anomalies including those with morphologic abnormalities of the pinna and ear canal
4. Birth weight less than 1500 g.

5. Hyperbilirubinemia at serum level requiring exchange transfusion

6. Ototoxic medication

7. Bacterial meningitis

8. Postnatal asphyxia (Apgar < = 5 at 1 minute or < = 6 at 5 minutes).

9. Mechanical ventilation lasting 5 day's or longer Stigmata or other findings associated with syndrome known to include a sensorineural and or conductive hearing loss. (NIH, statement 1993).

10- Stigmata or other findings associated with syndrome known to include a sensorineural and or conductive hearing loss. (NIH, statement 1993).

CHAPTER 2

AIMS AND OBJECTIVES

Significant hearing loss is one of the most common health problems present at birth and, if undetected, will impede speech, language, and cognitive development. Early detection, intervention, treatment and rehabilitation prevent the consequences of neonatal hearing. The statement of the Joint Committee on Infant Hearing (1994), supported by the American Academy of Pediatrics (AAP), endorses the goal of universal detection of hearing loss in infants before 3 months of age, with appropriate intervention no later than 6 months of age. Infant distraction test has the disadvantage that it cannot be performed until 6 months of age. By doing such research, we hope that a protocol on neonatal hearing screening can be developed in Hospital Universiti Sains Malaysia.

2.1 Objectives

1- To determine the prevalence of hearing loss using the screening tool Echocheck among newborn of the special care nursery in HUSM during the study period.

2- To determine distribution of common risk factors in newborns with hearing loss.

2.2 Research Questions / Hypothesis

1- What is the prevalence of hearing impairment in Special Care Nursery, HUSM?

2- What are the common risk factors in newborns with hearing loss?

CHAPTER 3

MATERIALS AND METHODS

Study design cross sectional study, was carried out in the special care nursery, at Hospital of University Science Malaysia (HUSM), Kota Bharu, Kelantan, using otoacoustic emissions method, The Special Care Nursery (SCN) unit is a relatively quiet unit situated on the first floor of the main Hospital's building. The admission in the unit ranging from 80 to 120 newborns per month. It receives almost all the Neonatal Intensive Care Unit (NICU) graduates who are stable enough to be transferred out while waiting to be discharged. Apart from the NICU graduates, it also receives the problematic newborns, which do not need NICU treatment but have to be admitted such as mild to moderate neonatal jaundice.

3.1 Materials

The subjects of this study were composed of 530 neonates admitted to the Special Care Nursery at the Hospital of University Science Malaysia (HUSM). The testing started in February 1999 and lasted until July 2001. The neonates were screened twice. Echocheck first after delivery and second, a month later (only those who failed the firs screening test using the same screening tool. The mean duration of hospitalization was 3 days. Recordings were made systematically for all neonates. Testing was carried out after 24 hours aged avoiding TEOAEs false positive results, which is more likely occurred due to the accumulation of debris in the ear canal after delivery. TEOAEs test was carried out every day except holidays. The newborns were admitted to SCN for different causes, the majority were neonatal jaundice, premature, sepsis, low birth weight, and other clinical aspect.