CHAPTER 2

LITERATURE REVIEW

2. 0. INTRODUCTION

This chapter gives an overview on the prevalence of hearing loss and the various screening tests used for early identification of hearing loss with emphasis on otocoustic emissions and the various related studies on their potential use as either a screening or diagnostic tool. Some findings from previous research on middle ear conditions affecting the OAE screening tests and the test used to detect this problem are also discussed.

2. 1. PREVALENCE OF HEARING LOSS

One to 2 of 1000 newborns suffer from congenitally or perinatally acquired hearing disorders. The prevalence of neonatal hearing disorders has been reported to be increased 10 to 50 fold in infants at risk (Mason, Herman 1998). Moderate to severe bilateral hearing loss, greater than 40 dB distorts the developing child's perception of his or her attempt at speech production. If this type of hearing disorder remains undetected through the critical period of language acquisition within the first year of life, a profound impairment of receptive and expressive speech and language development will result. This in turn will lead to a decreased acquisition of expected milestones in speech and language development. For acquisition of hearing, a sensitive period up to 6 months of age has been demonstrated (Peck, 1995). Accordingly introduction of hearing aids within this period will improve subsequent hearing development among hearing impaired children. To ensure timely therapy a reasonable goal is to establish the diagnosis of severe neonatal hearing impairment before the age of 6 months.

2. 2. Language of early and later identified children with hearing loss

Hearing loss that is bilateral and permanent is estimated to be present in 1.2 to 5.7 per 1000 live births (Mauk et al, 1993). The typical consequences of this condition include significant delays in language development and academic achievement. These delays are apparent for both children with mild and moderate hearing loss, as well as for those whose losses fall in the severe and profound ranges.

Despite advances in hearing aid technology, improved educational techniques and intensive intervention services, there has been virtually no change in the academic statistics of this population since the systematic collection of national data more than 30 years ago (Wrightstone & Furth,1966). These data indicate that the average deaf student graduates from high school with language and academic achievement levels below that of the average fourth grade hearing student (Allen, 1986) Similarly, for hard of hearing children achievement is also below that of their hearing peers with average reading scores for high school graduates at the fifty grade level.

These limitations in reading have a pervasive negative impact on overall academic achievement (Quigley, 1979).

Many professional in both health care and special education have supported early identification of hearing loss and subsequent intervention as a means to improving the language and academic outcomes of deaf and hard of hearing individuals(Ross,1990). In 1994 the joint committee on infant hearing released a position statement endorsing the goal of universal detection of infants with hearing loss as early as possible, preferably by 3 months of age. This position statement was endorsed by American Academy of Pediatrics. All of this position statement support the need to identify all infants with hearing loss.

In a study done by Yoshinaga- Itano (1997), to compare the language abilities of earlier and later identified deaf and hard of hearing children, receptive and expressive language abilities of 72 deaf or hard of hearing children whose hearing loss were identified by 6 months of age was compared with 78 children whose hearing losses were identified after the age of 6 months.

Results indicated that children identified by 6 months of age and with early intervention demonstrated significantly better language scores than children identified after 6 months of age. This language advantage was evident across age, gender, socioeconomic status ethnicity, cognitive status, degree of hearing loss, mode of communication and presence /absence of other disabilities. In this study there was no significant difference between the earlier and later identified children on a wide variety of demographic variables based on cognitive ability, age at testing, communication mode, minority status, gender, degree of hearing loss, and socioeconomic status, frequently associated with language ability.

Retrospective epidemiological studies on the prevalence of bilateral congenital permanent hearing impairment in Europe have indicated estimates of 1.2 to 2.07 per 1000 live births (Fortnum & Davis,1997). In these cases auditory deprivation has a serious effect on the speech and language development and thus on the social, emotional and cognitive development of the child. Several authors have demonstrated the benefit of an early identification and intervention with (hearing aids) on the later outcome of language skills in hearing -impaired children (Robinshaw,1995; Eilers et al, 1994; Kuhl, 1992).

Over the last decade two major new technologies have emerged which make it possible to take objective measures to estimate the likelihood of adequate hearing function in newborn babies. The *Evoked Otoacoustic emissions* (EOAEs) and the *Auditory Brainstem response* (ABR) testing. Furthermore when compared to the classical infant distraction test which is only possible from around the age of nine months, these new neonatal screening programs have produced better figures of sensitivity and specificity.

2. 3. AUDITORY FUNCTION DEVELOPMENT

An understanding of the development of auditory behavior is requisite to the testing of hearing in infants and children. A review of the literature from many discipline - specific journals begins naturally at the pre-natal stage.

2. 3. 1. Pre-natal hearing

Elliot and Elliot, (1964) confirmed physiologically that the human cochlea has normal adult function after the 20th week of gestation. Johansson and his associates (1964) were among the first to report testing the fetal hearing, using high frequency pure tones presented by means of a microphone placed on the mother's abdomen, fetal heart rate increases response to the tones was recorded after the 20th week of gestation.

The demonstration of fetal hearing has value in contradicting the theory that the child is born a tabula rasa insofar as hearing is concerned. The newborn infant has actually been hearing sounds for at least 4 months fluid-borne sounds, to be sure but nonetheless, true auditory signals.

It remained for a zealous experimenter, Bench (1968) to measure the acoustic qualities of the sounds that reach the fetal ear through the specific gravity of the amniotic fluid. With a specially prepared crystal microphone inserted at the cervix of a woman in the 38th week of gestation, Bench measured the noise level of sounds presented through a loudspeaker placed on the abdomen, over the fetal head. The internal background noise level was first measured representing the mother's pulse (80 beats per minute) at a sound level of 72 dB (C- scale).

What is interesting to us in this study is the revelation that the fetus is surrounded by a noise level that many people complain of as being too loud in the environment. Yet, the fetus has listened to this noise level for 4 months. Other investigators have measured Electro-encephalographic evoked auditory response of fetuses during the first stages of labour (Ostwald & Peltzman, 1974). A miniature earphone was introduced through the cervix and directed to the fetal ear. Electrodes were then clipped to the scalp and bursts of tone at a moderately high sound level were presented. Only one of six fetuses showed an evoked response to the sound – the only one whose mother had not been given drugs to stimulate contractions.

2. 3. 2. Newborn hearing – stimulus properties

It has been hypothesized that because the fetus is accustomed to listening to the heartbeat of the mother, he will be quieted effectively by the same sound in his early days in the outer world. Brackbill et al (1966) found that a tape recorded heartbeat is no more effective than any other continuous, low frequency stimulus in lowering the arousal level of the infant. Lowered arousal level means a specific reduction of overt behavior and physiological patterns. The infant's heart and respiration rates becomes lower, they cry less and move about less. Kagan (1972) suggests that stimulus change is another parameter that should be explored

In another study Bench and Bosack (1970) applied ***signal detection theory*** to infants responses to three different stimulus levels of 300HZ (55, 75, and 95 dB). The infants pre-stimulus states were rated as deep sleep, light sleep, and limb activity. They found that the signal detection is affected by both the sound pressure level of the stimulus and the state of the baby before stimulation. Taylor and Mencher (1972) also identified infant state as a significant variable in neonatal testing, reporting that light sleep is the optimal state for evaluating a response to auditory stimulus.

Downs (1967) has catalogued a variety of neonatal behavioral responses to sound and has pointed out ways of observing these responses.

- Eye blink or eyelid activity.
- Moro reflex or startle reaction.
- Cessation of activity.
- Limb movements.
- Head turn away from sound or head turn toward sound. These are commonly seen either toward the sound or away from it. Both responses are valid.
- Grimacing.
- Sucking
- Arousal : when the baby is sleeping quietly or is awake but quiet
- Widening of the eyes: when the baby is sleeping, his eyes may open wider than they were before even momentarily. These responses consist

of those observed to a 90dBSPL signal, when presented in a nursery environment

2. 3. 2. 1. *The older infant*

The maturation of auditory processing proceeds after birth in ways that have been demonstrated in research designs. Already at 4 weeks the infant can distinguish phonemic contrasts in sound signals as measured by heart rate changes. By age 3 months according to Turnure (1969), babies attend better to mothers voices on a tape recording than to stranger's voices even when mother's are modified by filtering. Turnure reported differences in the way the infants attended to the voices as a function of age level. The 9 month olds tended to be quieter to the natural mother's voice and progressively less attentive to the distortions.

Perception of rhythm in the 2month old infant was demonstrated by Demany et al (1977) utilizing varied sequences of time bursts in a habituation paradigm relating duration of fixation on a visual figure. The infants were able to perceive intervals of time as subjective links between sounds, showing that they apprehend a succession of several sounds as a psychological unit. This ability presents a necessary prelinguistic skill, since language comprehension requires the same kind of process in grasping a semantic unit as a whole despite its sequential character.

Linguistic stress is also discriminated by infants 1 to 4 months old. Spring and Dale (1977) demonstrated with a high amplitude sucking paradigm that young infants could discriminate the acoustic correlates of stress, location, fundamental frequency, intensity and duration. This study completes the number of suprasegmental aspects of speech that have been shown to produce discriminatory responses : intonation, rhythm and now stress.

Babies show a great range of discriminative listening to a wide variety of natural, disguised and synthetic language as well as to other auditory stimuli as the following examples indicate:

(a) a 12 month old infant preferred to listen to a strangers voice with bright intonation than to his mothers voice speaking in a flat monotone.

(b) a 14 month old baby could not decide for several days whether to select the strangers voice or the mother's distorted voice. After several days he made an enormous burst of listening to the mother's and after that he ignored completely the switch that turned on the stranger's voice. This choice is regarded as an " Aha" effect with the child ultimately recognizing the mother's voice despite its distortion.

Its seems inescapable to conclude that these young babies are using some cognitive process to make their listening selections. They must formulate internal linguistic models against which to compare the recurrent inputs. Such intellectual activity challenges Piaget's idea that cognitive development in the infant begins only after the sensorimotor period ends at 18 months. Exploring the responsiveness of the 0 to 6 month old infants to various stimuli, Bench et al (1977) found that both younger and 6 month olds were notably unresponsive to tonal stimuli and even to band widths of 300Hertz. But broad –spectrum noise gave better responses in the younger infants (1week and 6 weeks).

For the 6 month old, voice stimuli were most responded to. Moderate intensity signals were not effective for awake 6 month olds. The younger infants were mostly in sleep states when studied, so stimuli of 90dBSPL were necessary for responses. Kagans work (1972) suggests a varied assortment of response parameters for investigations. One of them is the duration of the infants sustained attention, which Kagan believes is a rough index of how easy or how difficult it is for the infant to understand a new experience.

2. 4. HEARING SCREENING TESTS – AN OVERVIEW

A consensus is growing that neonatal hearing screening is important. Several programs are being implemented throughout the western world. In many countries people are still looking for a feasible program that fits with the national health care system. By necessity most programs will still be based on existing ones, with slight modifications to cope with the local circumstances.

Early detection and intervention are extremely important to alleviate the disabling and handicapping effects of significant congenital sensorineural hearing impairment. It is generally accepted that delay in provision of appropriate amplification and rehabilitation measures will have marked effects on speech, intellectual and social development.

In recent years much attention has been focused on the possibility of screening neonates for hearing impairment. A variety of screening devices have been marketed and evaluated, for example the ***' Crib- O – Gram'*** in the USA (Marcellino,1981, Hosford-Dunn et al,1987) and the ***'Auditory Response Cradle'*** in the UK (Bennett and Wade,1980,Mccormick et al, 1984, Bhattacharya et al , 1984) both of which monitor behavioral response to sound and give a pass or fail decision.

2. 4. 1. Behavioral Auditory tests.

Behavioral Auditory assessment particularly during the early months of this age range is primarily limited to behavioral response, involving elicitation of reflexive responses to relatively intensive auditory signals. The more useful responses are the eye blink, startle reflex and arousal the startle response is seen as a small "jump" of the infants body immediately following a stimulus.

2. 4. 1. 1. Neonatal period.

Various procedures have been developed for the testing of hearing in newborn infants. Such methods are generally applied within the neonatal unit itself as screening procedures for the testing of at risk babies, usually pre-term infant neonates.

2. 4. 1. 2. Reactometry Test (3 – 5^{th} day)

The stimuli consists of a modulated pure tone, white noise or narrow band noise centered around 3- 4 kHz. This frequency range is chosen because in the neonate any significant damage to the hearing system will almost always include a high frequency hearing loss. Sound is presented in a sound field conditions at levels of 60 – 90 dB or 500 ms at a distance of 20 cm from the test ear, and is generated by a sound generator unit called ***Reactometer.*** Typical responses include the moro reflex, blinking, sucking, startle reflex, and crying. The procedure can be repeated at least twice / day until the neonate leaves the unit.

2. 4. 1. 3 Auditory Response Cradle Test (ARC) (first 5 days)

This automatic screening procedure, introduced by Dr. Mike Bennet in 1979, involves the electronic monitoring of motor and respiratory response in the neonate. It can therefore be considered an objective procedure since it does not depend on the subjective evaluation of neonates reaction to sound on the part of the examiner, which inevitably causes much variance. Motor responses include head turn, body activity and backward head movements and are sensed by a micro- processor unit.

Respiratory changes are sensed by a transducer filled inside a cloth belt which is strapped around the abdomen of the infant when he is transferred from his cot to the cradle. The stimulus used is a broad band noise (resonance's range from (125 - 4000Hz) presented at 87dBASPL for 2 seconds. Responses in neonates differ accordingly to the state of arousal where a positive response is taken to be a change in motor or respiratory activity during the test. The test has drawbacks, that is such a loud stimulus may easily pass a mild - moderate hearing loss or record a false positive response in the presence of sensorineural loss.

For the past 40 years, infant hearing screening has been attempted with a number of different psychoacoustic methods. For the last 15 years, ABR testing has been considered the most objective screening technique. The development of auditory evoked potentials in the assessment of hearing has led to the ABR being widely investigated as a neonatal screening technique (Cox, 1984). The majority of studies have used ABR testing to screen neonates admitted to the special care baby unit (SCBU) just prior to discharge.

The reported prevalence rates for significant bilateral sensorineural hearing loss in this SCBU population are indeed higher than for the general population. The more recently developed Cochlear emissions or Otoacoustic emission techniques are also currently being investigated for suitability for neonatal screening (Stevens et al 1987, Johnson et al 1988).

TEOAEs, represent a recently developed method that could be applied to all newborns prior to hospital discharge. The joint committee on infant screening proposed that screening begin with a TEOAE test.

2. 5. OTOACOUSTIC EMISSIONS : AN OVERVIEW

The physical mechanism of the middle ear and cochlear serve to collect and concentrate sound energy on to the sensory hair cells, but just as you can turn an old horn type hearing instrument around and blow like a trumpet so the ear can operate in reverse, vibrations generated inside the cochlear are magnified by the middle ear and transmitted into the air as sound. By sealing a receiver microphone probe into the ear canal, sounds made by the cochlear can be evoked by external sound and recorded from almost any ear with normal mid frequency hearing. With the latest techniques EOAE can be registered so quickly and efficiently that they offer a very effective new objective screening method especially in infants (Stevens et al, 1987, Bonfils et al, 1988).

2. 5. 1. New concepts

An old idea put forward by gold in 1948 has been revived to explain Evoked Otoacoustic Emissions and other discoveries over the last 20 years. It is, that the organ of corti acts as a kind of biomechanical hearing instrument overcoming the naturally high energy wastage in the cochlear. Contrary to Bekesy's observations on dead ears, the healthy cochlear can focus sound energy on to the sensory hair cells much more efficiently and sharply than is physically credible.

Vibration amplification, powered by the metabolism of the cochlear, is the only viable explanation for the cochlear's superb sensitivity and frequency selectivity. The outer hair cells with their newly discovered motility and ability to generate vibration are the favored site of the amplifier. This radical new thinking about the cochlear is changing the way we investigate and interpret hearing loss.

As otoacoustic emissions are now thought to reflect the activity of active biological mechanisms within the cochlea responsible for the exquisite sensitivity, sharp frequency selectivity, and wide dynamic range of the normal auditory system, there is significant indirect evidence that these mechanism are the outer hair cells (OHCs) at least in the mammalian cochlea (Kiang, Moxon and Levine, 1970, Khanna and Leonard, 1986, Liberman and Dodds, 1984).

It is well documented, that when OHCs are absent or damaged, auditory sensitivity is reduced by 40 - 60 dB, and the" tips" of tuning curves are elevated or absent. Responses to auditory stimuli as a function of stimulus level grow abnormally. The absence of outer hair cells is associated with a lack of otoacoustic emissions, supporting the hypothesis that OHCs are responsible for OAE generation.

Numerous observations support a cochlear origin for Otoacoustic emissions. (1) OAEs are independant of synaptic transmission and are pre-neural. That is if the eighth nerve activity is blocked either chemically (Arts, Norton and Rubel, 1990) or physically by severing it (Siegel and Kim, 1982; Martin, Lonsbury-Martin, Probst and Coats, 1987). OAEs can be measured while neural responses to sound are absent. (2) OAEs are unaffected by stimulus rate, unlike neural responses.(3) Evoked otoacoustic emissions are frequency dispersive (i.e., the higher the emission frequency, the shorter its latency) and their amplitudes grow nonlinearly with stimulus level.

(4) OAEs are vulnerable to noxious agents, such as ototoxic drugs, intense noise and hypoxia, which are known to affect the cochlea. (5) Finally, they are absent in frequency regions with cochlear hearing losses greater than 40 - 50 dB, and present where hearing sensitivity is normal.

Although there are many unanswered questions concerning Cochlear function in general and OAEs in particular, there is growing interest in OAEs as an objective, noninvasive measure of cochlear status in clinical populations.

There are several types of otoacoustic emissions including Spontaneous (SOAEs), Stimulus frequency, Transiently evoked (TEOAEs) and Distortion product (DPOAEs) otoacoustic emissions.

Spontaneous otoacoustic emissions are present in approximately 40% to 75 % of normal hearing ears and occur without any external stimulation. Current clinical research is focusing on TEOAEs and DPOAEs. The TEOAEs are elicited by a click stimulus and are present in nearly all healthy ears. Hearing loss>30dB hearing level (HL) has been found to obliterate these emissions. The TEOAEs have clinical use in hearing screening, in differentiating cochlea from neural hearing loss and in monitoring cochlear function.

2. 5. 2. Transient evoked otoacoustic emission

Several studies indicate that TEOAEs are able to detect hearing loss of greater or equal to 30dBHL, whereas others found that hearing loss > 20dBHL can be detected by TEOAEs. In addition, the literature on the effect of different middle ear conditions on TEOAEs is sparse, unclear and at times contradictory. Therefore the effects of tympanic membrane (TM) retraction, middle ear fluid, pressure-equalizing (PE) tubes and TM perforation on TEOAEs are not clear.

In a prospective observational study in a tertiary care children's hospital, 89 patients (169 ears) with age range from 8 months to 18.1 years were studied to determine the association of middle ear status (assessed clinically and by tympanometry) with the outcome of TEOAE recording, to determine the level of hearing loss that can be identified by TEOAEs and to determine whether middle ear dysfunction or hearing loss has a greater impact on the presence of TEOAEs.

Statistical analyses showed that of the 8 middle ear and TM conditions evaluated, only the presence of middle ear effusion and normal examination findings had a significant effect on the results.

The study concluded that Type B and Type C tympanogram and the presence of middle ear effusion, which reflect abnormal middle ear status have an adverse effect on TEOAEs. The presence of hearing loss is the most significant predictor of TEOAE results. The above study recommended that TEOAE testing be completed in conjunction with tympanometry.

Transient evoked otoacoustic emissions (TEOAEs),also referred to as click evoked OAEs are frequency dispersive responses following a brief acoustic stimulus such as a click or tone burst. Because this was the first emission type reported in the literature by Kemp (1978), the term evoked OAEs is often applied specifically to Transient Evoked emissions. They are also known as Kemp echoes and delayed evoked otoacoustic emissions. TEOAEs are obtained using synchronous time - domain averaging techniques similar to those used to measure auditory evoked potentials.

TEOAEs are measurable in essentially all normal hearing persons with normal middle ear and normal cochleas (Kemp1978; Johnsen and Elberling et al,1982). In response to tone bursts, the emissions are quite frequency specific. If there are spontaneous emissions one or more SOAEs can be "synchronized" by a transient evoking stimulus and strong components will appear at the SOAE frequencies throughout the evoked emission averaged waveform.

In general however the frequency content of a transient evoked emission is determined by the spectrum of the evoking stimulus. This is true for both the level and bandwidth of the stimulus. There is some confusion about the "frequency specificity" of Transient evoked emissions. This arises at least in part because of the properties of transient stimuli. The shorter the stimulus duration the broader the spectrum and the less energy at any one frequency. Because emissions can be evoked at most, if not all locations in the normal cochlear, the broader the stimulus spectrum the broader the emission spectrum. That is if there is more than one frequency in the stimulus, emissions will be evoked at more than one frequency.

Each emission however is frequency specific in that the frequency of a given emission is specific to the frequency of the evoking stimulus. If clicks and tone bursts are equated for band levels, the emissions within the bandwidth of the tone burst are nearly identical (Stover and Norton 1992)

Although recent research provides some evidence for the value of (TEOAEs) in neonate hearing screening, data are needed from larger scale clinical evaluation about the value of using TEOAEs for screening not only high risk but also healthy neonates. In a study (Aidan, et al,1999) on a cohort of 1421 neonates (2842) from the well baby- nursery was screened with TEOAEs in a 2 stage process. The purpose of this study were to evaluate whether TEOAE recording could be used accurately as a universal hearing screening tool for every newborn and to estimate the prevalence of neonatal sensorineural loss.

Neonates were referred from the first test prior to being discharged from the hospital. Those who failed were re-screened before the end of the first month. Those who did not pass the second stage TEOAE screening, were referred for diagnostic audiological evaluation for confirmation of hearing loss. Neonates transferred to a neonatal intensive care unit were not included in this study. Two neonates with bilateral sensorineural hearing loss of more than 40 dB hearing level were identified from this cohort.

This study demonstrates the feasibility and the limitations of using TEOAEs as a universal hearing screening tool for all neonates. It confirms that the prevalence of hearing impairment in neonates has to be taken into account even in a group of children without high-risk criteria. In France a prevalence of 1.4 per 1000 would represent 1000 deaf children born every year with reference to about 700,000 births per year.

This study suggests that such universal screening programs would substantially increase the rate of early identified infants with significant hearing impairment.

Analyses of results in this study showed that of all tested neonates, 1183 (83.25%) passes the first test. Chang et al studied a group of 41 full *term newborns, without perinatal disease with TEOAE recording. The authors* compared the results of the test before and after cleaning the external canal. The pre-otoscopic examination TEOAE pass rate of 82 ears was 76%, while the pass rate improved to 91% after external canal debris removal.

The first stage failure rate in this study could have been influenced by probe fitting problems. Obstructing debris in the external duct can attenuate TEOAE responses. White et al reported 37 neonates identified with a conductive hearing loss in a group of infants referred for diagnostic audiological evaluation.

Middle ear dysfunction can indeed confound the interpretation of TEOAEs. Middle ear evaluation by tympanometry prior to screening was not performed in this study, and even though middle ear disorders are not frequent in neonates, this factor could have influenced the results of the screening (Lonsbury-Martin1994). Another important characteristic of evoked otoacoustic emissions is that their amplitude grows non- linearly as a function of stimulus level.

Much of the clinical work related to OAEs has focused on click evoked emissions. One such neonatal screening program using Transient otoacoustic emissions (TEOAEs) was started in St.Augustinus hospital, Antwerp in 1993. The cost of the test was not covered by the public health insurance, so the parents had to pay for the full cost for screening their child. Since 1993 the program strategies have been changed on several occasions to improve the quality and efficacy.

A retrospective analyses was performed on the test pass rate, the coverage and the number of children who become " lost to follow up" after failing the initial test. The data shows a steady learning curve with a time course of several years. They also demonstrate that it is worthwhile and feasible to run a high quality screening program in a private establishment. The initial screening protocol in 1993 was as follows: The neonates were tested in a sound proof cabin on a quiet room at the audiological centre of the University ENT department. Typically the neonates were brought to the test location by their mother, about 1.5 hours after feeding. Testing was done as soon as possible after the parents registered for the test. The pass - fail criterion was a test of qualitative visual scoring . In the case of bilateral failure of the first test, the parents were immediately and verbally invited for a re-screen 3 weeks later, if after six weeks no re-screen had been done, a letter was sent once or twice to the parents to urge them to make an appointment, if a child failed the TEOAE test twice it was referred for a diagnostic auditory brainstem test (ABR) at the age of 3 months.

The global evaluation of the screening program for the whole period from 1993 to 1997 showed that 16 of the 3751 screened neonates were referred for diagnostic ABR testing. Half failed this test and were identified as being bilaterally hearing impaired. All were referred for early rehabilitation and intervention.

The wide spread application of TEOAEs is also due to the availability of commercial hardware and software. In the presence of hearing loss, Transient evoked OAEs have been shown to decrease in incidence as hearing thresholds increase (Kemp, 1978, Kemp et al 1986). Generally if the hearing loss exceeds 40-50 dB, an emission cannot be evoked to a transient stimulus. Kemp et al (1986) reported that the upper limit is 30dBHL

The relationship between audiometric threshold and OAEs in response to suprathreshold stimuli is not yet well defined. There is good correlation between pschophysical threshold and Transient EOAE for the same stimuli (Stover and Norton, 1992). In general responses to suprathreshold stimuli decrease as sensitivity increases. If one is interested in sensitivity ,one may need to measure emissions at several stimuli levels and determine emission threshold. If interested only in cochlear reserve or integrity, one may be able to use a single high level stimulus.

TEOAEs also decrease in magnitude with age in normal hearing persons. They are extremely robust in normal hearing, full term newborn babies. Current results from large clinical trials (Maxon, Norton, White and Brehens,1991) indicate that Transient OAEs can be a rapid sensitive tool for detecting hearing loss in both normal and at risk newborns. The primary difference in terms of testing time is the additional preparation needed for ABR.

Another advantage of OAEs for screening is that they examine a broader frequency range than ABR, however particularly with neurologically at risk population, it must be remembered that EOAEs are pre-neural. EOAEs therefore could be normal if the cochlear were normal in cases of central auditory system dysfunction. For this population EOAEs and ABR can be used together to help identify the site of lesion.

2. 5.3. Middle Ear conditions/ status and Otoacoustic emissions:

It is also important to note that in testing cochlear function, OAE also assess the conductive status of the middle ear. Thus their measure are sensitive to both sensorineural hearing loss and otitis media with effusion, the two most common cause of hearing loss in newborn. Regardless of the potential complexities introduced by the sensitivity of emissions to middle ear dysfunction, they potentially provide an excellent test for both screening situations, because evoked OAEs can be evaluated in the absence of a sound treated room.

Targeted neonatal screening of babies on the special care baby unit (SCBU) plus those with certain other high risk factors is now widely used for the early detection of congenital sensorineural loss. Such targeted hearing programs may detect up to 64% of deaf babies by testing only 7 % of births (Davis, 1993). Click evoked OAE are commonly used for the screening often in conjunction with auditory brainstem response testing (Stevens et al,1990).

Despite the positive OAE properties listed above, their use may be nevertheless, subject to some impediment . It is well known that the structural properties of the external and middle ears affect sound transfer to the cochlea (Pickles, 1988) and therefore, will affect OAEs. OAEs may be affected by the presence of middle ear fluid / effusion debris in the external canal or negative pressure on the ear drum (Kemp et al 1990).

As one apparent weakness of OAE testing is the failure rate of roughly 18% obtained in neonates during the first 3 days of life. This is higher than the 10% of neonates who are reported to fail an auditory brain-stem response screening test in comparable environments. Both OAE and auditory brain stem response fail rates are higher than the reported incidence of hearing impairment, indicating a high false-positive error rate requiring follow up. This apparent limited specificity diminishes the cost effectiveness of OAEs as a mass screening device.

One of the hypothetical explanations for the increased failure rate in the first days of life is the presence of middle ear fluid that attenuates the OAE response. Middle ear negative pressure or fluid have previously been shown to decrease the OAE below 2000 HZ. In screening of newborns in the neonatal intensive care unit , 20% fail to have recordable OAEs, corresponding with the higher incidence of middle ear abnormalities in this population. However, middle ear problems in normal full-term infants have been reported to be rare.

One area that has not been explored in the literature is the effect of debris in the external ear canal on the recorded OAE. Balkany et al, reported that examination of the external auditory canal of 50 normal term infants revealed that in all infants younger than 24 hours old, the canal was partly filled with vernix caseosa. Vernix caseosa drains out in the first 24 to 48 hours after birth, and the ability to successfully record OAE improves.

In a study to determine the relationship between the external auditory canal and middle ear status and the characteristics of the click - evoked OAE, 41 full term neonates were tested initially on the OAE, followed by an otoscopic examination, removal of debris, and a second post examination OAE was completed.

Results of the study indicated that the pre - otoscopic examination pass rate of 82 ears was 76%. The OAE pass rate improved to 91% after debris removal. Results indicate that an examination of the ear canal is warranted whenever an ear fails to pass the OAE test, especially when it falls into the fail study category. Since abnormal OAEs may reflect sensorineural loss, canal blockage, or middle ear dysfunction, it appears that routine screening with this method may result in an increased false - positive rate and referral for further evaluation. In conclusion an examination and cleaning of the external ear canal are important components of a neonatal screening process.

The conditions of the middle and external ear can be assessed by performing an ear examination or Otoscopy and Tympanometry.

2. 5. 4. Tympanometry: An Overview

Tympanometry is currently enjoying extensive popularity worldwide in the evaluation of the middle ear system and particularly (Meyer 1997). The initial description of the clinical application of this technique was made by Metz (1946). It was only since the late 1960s that it's use with infants and children was investigated and validated by several researchers (Robertson et al, 1968, Brooks 1971, Bluestone et al, 1973, Jerger et al, 1974, Keith 1975, Groothuis et al 1979, and Weaver 1979). The Impedance battery, as currently employed, typically consists of three measures: Tympanometry, Static compliance, and Acoustic reflex thresholds.

Tympanometry is a measurement of the relative change in the compliance of the middle ear system as air pressure is varied in the external canal. Tympanograms, the graphic display of the tympanometric measurements, are generally classified in terms of the depth, shape, and point of the middle ear pressure (Feldman, 1976).

The most common cause of hearing loss in children is middle ear disease and by far the most common form of middle ear disease is Otitis media and its associated complications (Goin,1975). In fact, 90% of all ear problems in children are the result of some form of otitis media (McCandless,1979).This disorder refers to the accumulation of fluid in the middle ear as a result of infection

In addition, the prevalence of this disorder has raised concerns about the effect of the concomitant conductive hearing loss upon speech and language development (Holm & Kunze,1969, Kaplan, Fleshman, Bender, Baum, & Clark,1973). Further, Brooks (1980) has recently suggested that recurrent or chronic middle ear disease in childhood may interact with the expected age - related hearing loss in the elderly, producing the need for amplification at a significantly earlier age. Brooks (1968) was among the first to demonstrate the efficiency of using tympanometry testing in the detection of middle ear disease in children. Brooks observed that the primary effects of serous otitis media were a reduction in the depth of the tympanogram, and associated decrease in static compliance.

The relationship between degree of Otitis media and tympanogram type was examined by Orchik, Dunn, and McNutt (1978). Tympanograms were classified using a system similar to that devised by Jerger (1970). A total of 142 ears of 75 patients were examined, and the results suggested that when a type B tympanogram was found, there was approximately a 90% probability of significant middle ear effusion. Other tympanogram types were not efficient predictors of serous otitis media.

When tympanometric testing indicates the presence of middle ear disease, the results will typically be corroborated through otoscopic or ear examination by a physician. An important consideration is the degree of agreement one can expect between the results of tympanometry and otoscopy. An early investigation by McCandless and Thomas (1974) indicated excellent agreement between the two measures. Tympanometric data and otoscopy were compared in 730 children and the results of the two procedures agreed in 93% of the cases. McCandless and Thomas recommended criteria for medical referral as negative pressure in excess of -100mmH2O.

2. 5. 5. Middle ear effusion /fluid and neonatal screening

Some authors have suggested that middle ear effusion (MEE) is prevalent among babies on the Special care baby unit (SCBU) (Berman et a, 1978). Hence because MEE is likely to reduce or abolish OAEs (Kemp et al 1990), it may be a major reason for the failure of neonatal hearing screening using OAE in these high risk babies, and account for much of the high false positive rate typically 20 % (Stevens et al, 1990)

The relationship between OAEs and middle ear effusion in these high risk neonates is therefore of great interest, but there have been no systematic studies on the topic. The only reports (Chang et al, 1993,Vohr et al,1993, Thornton et al,1993) have been on full term babies tested within the first 3 days of life among whom midldle ear effusion is rare and where there are also confoundly factors including vernix caseosa in the ears and variable maturation of OAE responses (Kok et al, 1992; Thornton et al, 1993).

As part of a larger prospective study into the prevalence of middle ear effusion and its effect on hearing on 100 special care and 100 full term babies in the first year of life, OAE and Otoadmittance testing using 678HZ probe on both ears of 84 babies in special care were performed.

Results of the study showed that abnormal tympanometry is strongly associated with OAE failure. Otoscopy showed little vernix or wax in the canals of these babies, and none of the abnormal tympanometric or OAE results could be ascribed to this cause.

In cases of middle ear pathology OAEs may be measurable because they are not effectively transmitted by the middle ear. Generally if the air bone gap for pure tones thresholds exceeds 30 -35dB, OAEs cannot be measured. EOAEs can be measured in ears with patent pressure equalization (PE) tubes, if the air bone gap is small and middle ear cavity healthy. In testing newborns, debris including wax and vernix in the external ear canal can reduce and block EOAEs (Chang, Vohr, Norton and Lekas1992). Collapsed canals can also interfere with emission measurements.

Middle ear effusion or abnormal middle ear pressure (MEP) would be expected to affect OAEs both by reducing the transmission from the cochlear through the middle ear and by attenuating the stimulus reaching the cochlear (Kemp et al 1990). It has been shown that Evoked OAEs are absent in ears with middle ear effusion in older children (Owens et al 1992). Erwy et al, 1991 found that OAEs are observable in only 12% of children with flat tympanogram and that they were always absent when the conductive loss exceeded 20dB.

There are also some reported data on the effect of middle ear pressure on OAEs. Owen's et al, (1992) reported that even mildly abnormal middle ear pressure could effect OAEs, although not necessarily abolishing them. Positive middle ear pressure (above about 100dapa) has been found to abolish evoked OAEs (Owen's et al 1992).

There are very few other relevant data relating OAEs to middle ear effusion in neonates. Studies in full- term babies tested in the first 3 days of life (Vohr et al,1993; Chang et al,1993; Thornton et al,1993) do not cast light on the question because middle ear effusion is rare in this group (Balkany et al, 1978; Pestalozza et al,1988). Failure to record OAEs in these babies is more likely due to significant vernix or debris in their ears or the immaturity of the cochlear response in the first few days after birth (Kok et al,1992, Thornton et al, 1993).

The statement in Vohr et al, (1993) that their findings suggested that for full term infants in the normal nursery, middle ear status has little impact on OAE response clearly does not mean that fluid in the middle ear has no effect on OAEs but only that middle ear effusion has little effect on an OAE screening programme in full term babies because it is rare in this population. This conclusion would not hold true for OAE screening targeted on SCBU babies.

There are 2 other studies that have included a small amount of data relating middle ear effusion to OAEs in special care babies. Kennedy et al in 1991 reported on neonatal tympanometric results on 10 babies with middle ear effusion in their first year, although they did not give OAE results on all the ears with abnormal tympanometry neonatally, 4 out of the 5 ears that were reported with flat tympanograms failed the OAE test. Smurzynski et al, 1993 found that 8 out 10 ears with middle ear effusion failed the evoked OAE test. All reports are therefore quite consistent with the conclusion that OAEs are sensitive to middle ear effusion, and usually abolished by it.

Middle ear effusion appears to be uncommon in normal full term babies, but much common in special care baby unit babies (Balkany et al,1978, Berman et al, 1978, Pestalozza et al, 1988). For this study cohort the prevalence of abnormal tympanometry was 20 % of ears, or 29 % of babies (95% confidence interval 19 - 41 %) with 18 % unilateral and 11 % bilateral. This result is in line with the higher values reported in the literature for middle ear effusion in special care infants (30% of babies) in (Berman et al 1978).

2.6. AUDITORY BRAINSTEM RESPONSE TEST (ABR)

In 1967 an important contribution was made to the physiological measurement of auditory response using click stimului. Two Israeli physicians, Sohmer and Feinmesser, observed an evoked response recorded from the vertex of a human subject. This evoked response consisted of five positive direction waves occuring within the initial 12.5 millisecond post stimulus. In

1970 Jewett recorded from the vertex of humans within the first 8 milliseconds following the acoustic stimulation a series of 5-7 potentials probably evoked by cochlear process and auditory relays within the brainstem.

Jewett et al, (1970) commented on the remarkable stability of this series of evoked wave forms which have shorter latencies and more consistent potentials than other "early" evoked responses (between 20 and 50 millisecond post stimulus) of late evoked response patterns longer than 50 millisecond. Jewett and Williston (1971) systematically recorded early component evoked responses of varying stimulus and recording parameters. They labelled the seven peaks from1 to VII (Fig.15a)

The extensive literature on the origins of ABR in laboratory animals and in man has been thoroughly reviewed by Buchwald (1983).The early interpretations of the likely origins of ABR have been gained from experimental lesions in animals and from human brain structures during operations and in pathological conditions. The ABR morphology and responsiveness to various experimental manipulations in experimental animals are similar to those in man (Stockard et al,1978).

Numerous studies have now confirmed the clinical application of ABR measurement in various population from premature infants to adults. Helcox and galambos (1974) demonstrated the feasibility of detecting hearing loss in full term neonates with ABR. Subsequently, Schulman-galambos and galambos(1975) obtained ABR data from premature infants. Other investigators have demonstrated the practicability of ABR as a diagnostic tool with children, including information about the presence of conductive disorders (Mendelson et al, 1979).

ABR in Paediatric audiology

ABR has been used systematically to examine suspected hearing loss in small children (Hecox and galambos1978,Schulman-Galambos & galambos 1975). A study done on children from material compiled in 1977to1981 consists of 93 children, 74 of whom were tested under sedation. Only 19 older children during the first year of study were tested without sedation. With the exception of 1977 the first year of study the annual mean age of children was 2.1 to 3.7 years. The youngest patients were 4 months. Results of the study showed that that ABR threshold is acceptably accurate for rehabilitation purposes.

225 infants referred from intensive care nursery over a 2year period were studied using Auditory Brainstem response test. Approximately 64% of the infants were considered to have normal peripheral auditory sensitivity in each ear and no evidence of neuropathology. An additional 15% appeared to have normal peripheral sensitivity in at least one ear, while 21% showed abnormal results in both ears.

Of the 16% suspected of having a bilateral hearing loss, initial evaluation indicated that 12% were conductive, 2% Cochlear and 2% mixed hearing loss. If these 2 groups were combined, then 5% of the infants showed Cochlear involvement. These results are consistent with those reported by Schulman - galambos and galambos (1979), Marshall et al, (1980) and Cox et al (1981). A total of 25% were suspected of having a conductive hearing loss. This percentage is consistent with reported incidence of middle ear effusion in infants in ICNS (Balkany, Berman, Simmons, Schaffer & Angell 1977)

Otoacoustic emissions (OAEs) versus Auditory brainstem response (ABR)

Auditory thresholds using ABR were determined in 30 ears of normally hearing neonates and infants and in 70 ears of infants with Sensorineural hearing losses. In the same population, OAEs were recorded in response to a click of 30dBHL. The mean time necessary to measure ABR thresholds in this series was about 40 minutes for the 2 ears whereas the time necessary to record EOAEs was less than 5 minutes. EOAEs could be recorded in all normally hearing ears.

EOAEs were present in ears with either a pure Conductive or a sensorineural loss and a wave V ABR threshold equal to or lower than 25dBHL. When the ABR threshold was greater than 30dBHL, EOAEs were no longer present.

In a study to evaluate the effect of external and middle ear abnormalities on the specificity of OAE measurements compared to those of the Auditory brainstem response (ABR),20 neonates from Special care baby unit (SCBU) were tested using procedures for obtaining auditory brainstem responses (ABR) and Transient evoked otoacoustic emissions (TEOAE)

All 40 ears passed the initial ABR screen, while the pass rate for the TEOAE screen was only 52.5% ears with no external or middle ear abnormalities (group A) had a significantly higher TEOAE pass rate (94.7%) than those with at least one abnormality (group B) as detected by otoscopic examination and tympanometry (14.3%). Other variables, such as age at test, gestational age at birth and birth weight, did not differ significantly between groups A and groups B. We conclude that external/middle ear abnormalities in this group of neonates had no effect on the ABR screening results, but had a significant effect on the TEOAE screening results.