Chapter 1

INTRODUCTION

Chapter one introduces the problem this study confronts; the rationale therefore, describes the terminology used, and presents an overview of the content and organization of the study.

1.1 ORIENTATION TO THE STUDY

The practice of pediatric audiology is based on the principle that the best outcomes for a child with hearing loss are achieved when his or her hearing status is determined as early as possible, followed by timely intervention (Cone-Wesson, 2003:253). Hearing loss in newborn infants can go undetected until as late as three years of age without specialized testing (Hayes & Northern, 1997:4). When hearing loss is detected in the newborn period, infants can benefit maximally from amplification (hearing aids) and intervention to facilitate speech and language development (Sininger, Doyle & Moore, 1999:11). Evidence regarding neural development strongly supports such early intervention for optimal outcomes of communication ability and hearing in infants (Sininger, Doyle & Moore, 1999:11). Pediatric audiologists, therefore, should facilitate early detection of hearing loss and intervention through screening programs and in-depth hearing assessments, determining which technologies and habilitation programs are best suited to the needs of both the infant and the family.

In order to capitalize on the positive aspects of early identification, the Year 2000 Position Statement on Infant Hearing Screening produced by the Joint Committee on Infant Hearing (JCIH), recommend that all infants' hearing should be screened using **objective, physiologic** measures in order to identify hearing loss (JCIH, 2000:10). Audiologic and medical evaluations should be conducted before three months of age. Infants with confirmed hearing loss should receive intervention before six months of age from health care and education professionals with expertise in hearing loss and deafness in infants and young children (JCIH, 2000:10).

Infants identified according to these recommendations are too young for the use of traditional audiometric procedures to determine an audiogram (Cone-Wesson, 2003: 254). Instead, electrophysiologic methods such as Auditory Evoked Potentials (AEP) must be used to estimate hearing thresholds. Ongoing evaluation of hearing function is furthermore needed to monitor the effects of early intervention and AEP tests may also form an important part in this process (Cone-Wesson, 2003:270).

According to Diefendorf and Weber (1994:56), four criteria need to be addressed before amplification of a hearing loss can occur. The degree, the configuration, the symmetry and the type of hearing loss must be determined. Amplification and habilitation strategies, such as choice of hearing aids, cochlear implants and choice of mode of communication, are based on the above criteria – usually revealed by the audiogram (Ross, 2001:3). Electrophysiological tests can assist in the process of characterizing hearing loss for infants, since these tests can measure auditory function objectively - giving frequency-specific information needed to fit appropriate amplification in this young population.

Apart from assisting in the process of diagnosing the hearing loss, it is important to verify that a specific hearing aid provides adequate amplification (Picton, Dimitrijevic, Van Roon, Sasha-John, Reed & Finkelstein, 2002:63). According to Picton et al. (2002:64), verification of a hearing aid fitting provides some indication of how well sounds are heard when the aid is used at its prescribed settings. Electrophysiological tests can assist in this process of verification in infants and young children. Studies have indicated that both the Auditory Brainstem Response (ABR) and the Auditory Steady State Response (ASSR) can be used to estimate threshold when the stimuli is transduced by a hearing aid (Garnham, Cope, Durst, McCormick, & Mason, 2000:268; Picton, Durieux-Smith, Champagne, Whittingham, Moran, Gigueve & Beauregard, 1998:315). Since the early identification of hearing loss through newborn hearing screening has resulted in a younger population being served by the audiologist, electrophysiological tests are becoming a more essential part of the required test-battery.

1.2 BACKGROUND

The audiologist serves as the professional primarily responsible for the assessment and non-medical diagnosis of auditory impairment. Assessment includes, but is not limited to, the administration and interpretation of behavioral, electroacoustic, and electrophysiologic measures of the status of **peripheral** and **central auditory nervous systems** (Stach, 1998:3). The main purpose of a hearing evaluation is to define the nature and extent of hearing impairment. A comprehensive description of hearing ability serves as a first step in the rehabilitation of a hearing handicap that result from an impairment. An individual with a hearing disorder must therefore rely on an assessment of auditory function as the foundation of the rehabilitation process (Swanepoel, 2001:1).

Pure tone audiometry represents the first and, arguably, the most fundamental measure of hearing acuity (Harrell, 2002: 71). The behavioral pure tone audiogram is a measure of auditory threshold as a function of frequency. The audiogram configuration provides fundamental baseline information for the selection of a suitable amplification system (Stach, 1998:68). It is understandable then that pure tone audiometry has remained the audiometric procedure of choice. It embodies the gold standard for frequency-specific threshold establishment against which all other audiometric measures are compared (Swanepoel, 2001:3).

It is clear that the standard clinical pure tone technique will not be effective for all clinical populations. The most obvious is the pediatric population. This is also true of the developmentally delayed and, in some cases, the severely physically impaired (Harrell, 2002:73). Conditioned audiometric test techniques (such as visual reinforcement audiometry), that can provide increasingly accurate information in older children (>6 months), are not suitable for quantifying hearing loss in very young infants or for those with visual or developmental disabilities (Rance & Briggs, 2002:237) as suprathreshold stimulation is required to elicit reflexive responses. These conditioned audiometric methods are limited to the detection of hearing loss greater than 50 dB HL (Diefendorf & Weber, 1994:57).

Therefore, as the age of identification is reduced the need for accurate, reliable, **objective** methods for determining hearing thresholds, is becoming more urgent. When an infant is identified as having a hearing loss, there is an immediate need to characterize the degree, the configuration, and the type of loss, since appropriate recommendations regarding the selection and fitting of devices such as hearing aids and cochlear implants, can only be made with an understanding of both the degree and configuration of a child's hearing loss (Rance & Briggs, 2002:237; Vander Werff, Brown, Gienapp, & Schmidt Clay, 2002:228).

Different techniques have therefore been developed over the years in order to address the problem of quantifying hearing loss in young infants. One such a measure, that was received positively for diagnostic purposes in the pediatric field, was otoacoustic emissions – discovered in 1978 by David Kemp. Otoacoustic emissions (OAE's) are sounds that originate in the cochlea and propagate through the middle ear and into the ear canal, where they can be measured using a sensitive microphone (Prieve & Fitzgerald, 2002:440). It has been known for the last 20 years that individuals with significant cochlear hearing loss have no measurable Evoked Otoacoustic Emissions (EOAE's). Hall (1997:265) has indicated that EOAE's are either not measurable from subjects with hearing loss or are substantially reduced in amplitude compared to normal hearing individuals. The minimum level required for measurable EOAE's ranges from 25 – 40 dB HL (Prieve & Fitzgerald, 2002:452).

Given this specific population and the need for specific information, with regard to degree, configuration, symmetry and type of hearing loss, work in the area of OAE's does not provide compelling evidence to indicate that EOAE's could be used to predict the nature of an infant's hearing loss (Prieve & Fitzgerald, 2002:456). This procedure is, however, a powerful tool for newborn hearing screening and should form an integral part of the differential diagnosis test battery in the diagnosis of hearing loss in infants (Hall, 2000:391).

The ABR is a far field evoked potential that is used frequently to estimate auditory sensitivity in children too young to be tested using standard behavioral methods. The ABR to click stimuli is the most commonly used clinical procedure in predicting hearing sensitivity (Vander Werff, Brown, Gienapp, & Schmidt Clay, 2002:228). Although the abrupt onset of the click stimulus is ideal for generating the ABR, the drawback is that the click is a broad-spectrum signal, containing energy across a wide range of frequencies. Owing to this frequency spread, clicks cannot be used to assess sensitivity in specific frequency regions, but rather to provide a gross estimate of hearing sensitivity (Arnold, 2000:454).

ABR to brief tones can be used to obtain more frequency-specific threshold information than available from the click ABR. Stapells (2000a:13) has shown across studies that tone-ABR thresholds have been found to be a reliable method in obtaining frequency specific information in this young population. Stapells (2004: conference presentation) maintains that the tone-evoked auditory brainstem response (tone-ABR) is currently the only measure that can reliably provide information with regard to severity, configuration and nature of hearing loss in infants. Some studies have however questioned the frequency-specificity and reliability of threshold estimation with low frequency tone-evoked ABR (Cone-Wesson et al., 2002:174). In addition, toneburst ABR waveforms, especially to low-frequency stimuli, tend to be less distinct and more difficult to identify than the click ABR (Arnold, 2000:459). Output limitations are also a concern with tone burst stimuli, particularly for low-frequency tone bursts for which thresholds are elevated relative to behavioral thresholds. These concerns may limit the implementation of toneburst ABR protocols (Vander Werff et al., 2002:229).

1.3 RATIONALE

In the past two decades, the Auditory Steady State Response (ASSR) has been developed as an alternative frequency specific Auditory Evoked Potential (AEP) approach to quantify hearing loss (Rance et al., 1998:499). Vander Werff et al. (2002:228) define the ASSR as "*an alternative evoked potential technique that uses continuous rather than transient stimuli to elicit a response from the auditory system".* Unlike the ABR, the ASSR use stimuli that is continuous. The stimuli used to evoke ASSR, are modulated tones, which are frequency specific due to the fact that spectral energy is contained only at the frequency of the carrier tone and the frequency of the modulation (Hood, 1998:117). Responses from the neural system that responds to the changes or modulations in the stimuli are recorded. The ASSR appears to be generated by the same neural anatomical regions from which the ABR evoked by clicks or tone-bursts is produced (Cone-Wesson, 2003:267).

The ASSR shows potential to address some of the limitations associated with ABR testing in the early diagnosis and amplification of infants. One of the limitations of ABR is the lack of frequency specificity. The nature of the ASSR stimuli offers advantages over other short duration stimuli (Rance et al, 1998:499). Because the threshold estimates obtained from ASSR testing are frequency specific, it allows for testing across the audiometric range and for the generation of evoked potential audiograms (Rance et al., 1998:499). This feature will address the problem of determining the configuration of hearing loss in infants.

Due to the limitations in maximum output with the ABR, the absence of wave V in ABR test results does not inevitably imply the absence of hearing (Arnold, 200:457). Rance et al. (1998:506) demonstrated the advantages of using ASSR's to determine residual hearing thresholds in infants and children for whom ABR's could not be evoked at 100 dBnHL. The continuously modulated tone used to elicit the ASSR can be presented at levels as high as 120 dB HL. Absence of a click- or tone-burst ABR does not inevitably indicate profound deafness, and ASSR tests may reveal enough residual hearing to consider the use of amplification or help determine the preferred ear for cochlear implantation (Rance et al., 1998:506). The advantages offered by the ASSR in this regard are most beneficial for infants and children as the severity of the hearing loss can be determined in a more accurate manner.

Cone-Wesson et al. (2002:270) mentions a third limitation of the ABR for audiologic application as the subjective nature of response detection. Although methods for "automatic detection" of ABR exist, these algorithms have been successfully applied for click-evoked ABR. There are no published data on the use of automatic detection criteria for detecting the response to tonal stimuli. In contrast, there has been extensive research on the efficacy for detecting a steady-state response automatically (Cone-Wesson et al., 2002:175; Rance et al., 1995:499). The objective nature of response detection in the ASSR measures may lead to more accurate diagnosis of hearing loss in infants.

According to Rance et al. (1998:506) a further advantage of the ASSR as opposed to the ABR, is the speed with which a response can be detected. Although Hall (2005:conference presentation) disputes the speed of the ASSR to be faster than the ABR, several researchers have concluded that ASSR offers the possibility of estimating frequency-specific hearing thresholds in babies in a more time-efficient way (Luts, Desloovere, Kumar, Vandermeersch & Wouters, 2004:995; Swanepoel, 2001:112; Rance et al., 1998:506). A constant unpredictable factor in testing infants is that they may awake at any moment during the procedure. The fast detection speed of the ASSR thus reduces the need to have the infant asleep or under sedation for long periods of time.

It is techniques such as the ASSR that underlie successful early amplification of hearing necessary to preclude or limit the auditory sensory deprivation effects (Ross, 1996:13). The amplification process begins directly after the diagnosis of a hearing loss has been made. Functional evaluation of a hearing aid seeks to determine whether the child benefits from such amplification. The functional evaluation of hearing aids is as essential as an electroacoustic evaluation thereof. The aided audiogram can evaluate whether the child is able to hear soft sounds within expectations, based on the electroacoustic fitting of the hearing aid. Kuk (2004:1) also maintains that using the levels obtained through functional gain must be a reassurance to the parent to ensure that the optimal opportunity is given to develop a child's potential. Although ABR measures have been used in the past to assist in the fitting of hearing aids in children, the clinical use of these procedures are technically challenging (Garnham et al., 2000:268; Mahoney, 1985:351).

ASSR's have been used to demonstrate the gain provided by amplification (Picton et al., 1998:315). ASSR's can be obtained in the sound-field condition – measuring an unaided response as well as the aided response. The difference in ASSR threshold obtained in the aided condition is then used to predict the functional gain of the hearing aid (Cone-Wesson, 2003:272). According to Glockner in Cone-Wesson (2003:272), hearing aids appear to transduce the modulated tones with good fidelity; the spectral characteristics of the modulated tones played through an analog hearing aid with no compression are well preserved. The fact that the stimuli are much more stable over time than brief transients means that they are more reliably transferred through the free field speakers and hearing aids – even when the hearing aids are non-linear (Picton et al., 2002:66). After being diagnosed with a hearing loss and fitted appropriately with hearing aids, the adequacy of the fitting needs to be validated. Validation is an ongoing process designed to ensure that the infant is receiving optimal speech input from others and that his or her own speech is adequately perceived (Pediatric Amplification Protocol, 2003:15).

1.4 PROBLEM STATEMENT

The dawn of an era of early identification of hearing loss in newborns and infants confronts audiologists with new challenges and opportunities. The advent of universal newborn hearing screening has made it all the more common for audiologists to see infants less than two to three months of age who have been identified as being at risk of having a hearing loss. The process of fitting a hearing aid or determining the candidacy for cochlear implantation requires detailed knowledge of these infants' residual hearing abilities (Vander Werff et al., 2002:228). For newborns and infants, evoked potential estimates of audiometric thresholds may be the only information about hearing status that is available at the time when these critical decisions need to be made.

The transformation of new discoveries into practical clinical procedures has been a frequent occurrence in audiological test development over the past three decades (Gorga, 1999:29). Several recent studies have therefore explored the relationship between ASSR electrophysiological thresholds and audiometric behavioral thresholds for normal-hearing and hearing impaired listeners (Dimitrijevic et al., 2002:205; Herdman & Stapells, 2001:41; Lins et al., 1996:81; Rance et al., 1995:499). These investigators have reported finding significant correlations between ASSR thresholds and behavioral audiometric thresholds for individuals with a range of hearing losses. Other studies have focused on the correlations between the ASSR and ABR as threshold prediction technique (Cone-Wesson et al., 2002:173, Vander Werff et al., 2002:227). Although these results appear promising, it is difficult to make definite conclusions about the application of ASSR to the infant population, as these studies were based on the responses of adults or older children rather than infants to evaluate the efficacy of the ASSR as a threshold estimation tool.

Stapells (2002a:14 & 2004: conference presentation) cautions that too few studies are available concerning the infant population to recommend the ASSR method for clinical use. However, the potential advantages of the ASSR that come from continuous rather than transient stimuli, including potentially better frequency-specificity and the ability to obtain higher output levels, warrant further investigation of the clinical application of the ASSR in the infant population.

In addition to the need of a tool for frequency specific estimations of hearing in infants the validation of amplification early on is also an essential component. Seewald (2001:70) emphasizes the need for improving the quality of pediatric hearing aid fitting, as the consequences of decisions made will be with a child forever. Yet, after fitting hearing aids on infants, validation of the fitting in most cases occurs through the use of subjective questionnaires and variables being evaluated such as auditory awareness, speech-production abilities, rate of language acquisition and social development (Scollie & Seewald, 2002:702) Aided thresholds are generally done only when the infant is mature enough to complete behavioral audiometry which may be several months after the initial fitting. In the age of early identification, this needs to be addressed. A limited number of studies on ASSR and functional gain have been performed (Glockner in Cone-Wesson, 2003:272; Picton et al., 2002:63; Picton et al., 1998:315). These studies focused on adults and older children and although the results are promising, the application possibilities of the ASSR in addressing the specific needs of the infant population need further investigation.

Bess (2000:250) and Gravel (2005:19) urge audiologist to collect, evaluate and integrate evidence about procedures in order to become evidence-based practitioners[1]. This implies that as new procedures become available, clinicians must be willing to continually evaluate and modify their clinical protocols. Therefore, with the advent of the ASSR in clinical practice and in light of the crucial importance of early identification of hearing loss and of the intervention process that follows, the question that arises is:

What is the clinical value of Auditory Steady State Response for early diagnosis and for evaluation of amplification in infants with hearing loss?

It was the purpose of this research endeavor to find answers to this particular question.

[1] Evidence Based Practice is an approach to clinical service delivery that has become increasingly advocated in the past decade. EBP is defined as the 'conscientious, explicit, and judicious use of current best evidence in making decisions about the care of patients (Oxford-Centre for Evidence Based Medicine, 2004: online).

1.5 DEFINITION OF TERMS

For the purposes of this study, the following terms will be defined and discussed in order to promote a mutual understanding of the basic and primary concepts dealt with in it. The terms provided are adapted from the work of Mendel, Danhauer & Singh (1999) in the *Illustrated Dictionary of Audiology,* unless otherwise stated.

Amplification of a hearing loss – amplification refers to an increase in the intensity of sounds. This is a collective term used for devices such as hearing aids. When referring to hearing aid assessment, the term functional gain is often used when validating a hearing aid fitting. Functional gain refers to the difference in performance between aided and unaided thresholds measures.

Auditory Evoked Potential – electrical activity evoked by sounds arising from auditory portions of the peripheral or central nervous system traveling from cranial nerve VII to the cortex, recorded with electrodes and also known as auditory evoked response. In this study the focus will be on the following evoked potentials:

- *Auditory Brainstem Response* (ABR) – an objective test that measures the electrical potential produced in response to sound stimuli by the synchronous discharge of the first- through sixth- order neurons in the auditory nerve and brainstem; also known as brainstem auditory evoked potential (BAEP) and brainstem auditory evoked response (BAER).
- *Auditory Steady State Response* (ASSR) – an auditory evoked potential in which the response wave-form approximates the rate

of stimulation; also referred to as steady-state evoked potential (SSEP).

Clinical value – Audiology is the health-care profession devoted to hearing. It is a clinical profession that has as its unique mission the evaluation of hearing ability and the amelioration of impairment that results from hearing disorders (Stach, 1998:2). Pediatric audiologists play a crucial role in early identification of hearing impairment in infants and evaluation of their hearing abilities. In addition, pediatric audiologists evaluate the need for hearing aid amplification in the pediatric population and monitor the success of these fittings. This study focuses on the potential value of the ASSR as an assessment tool that could aid the pediatric audiologist in fulfilling his/her clinical responsibilities.

The ASSR have been used in audiology research centers around the world. The results from the clinical studies have shown that ASSR thresholds can be used to predict pure-tone thresholds in sleeping infants and young children. It has also shown success in evaluating hearing aid fittings by determining functional gain. As with other discoveries in the field of audiology where transformation of new discoveries into clinical procedures has occurred, this study investigates the adoption of the ASSR into the clinical setting, comparing this promising technique with the traditional approaches used in the difficult-to-test populations.

Early diagnosis – The Healthy People 2000 initiative established the goal to reduce the average age at which children with significant hearing impairment are identified to no more than 12 months of age by the year 2000 (Diefendorf, 2002:469). With the implementation of universal hearing screening programs, the Joint Committee on Infant Hearing (1994) recommendations were that all infants with hearing loss should be identified by three months of age, and receive intervention by six months of age. In this study the time of identification varied between three months of age and 6 months of age. Intervention (amplification) was implemented immediately after diagnosis – trying to conform to the guidelines of the JCIH.

Infant - refers to a child during earliest period of life – before age 1 after the neonatal period (The concise Oxford dictionary, 1982:512).

1.6 DIVISION OF CHAPTERS

A research endeavor, consisting of both an empirical and theoretical component was conducted, in order to answer the research question stated above. The following section delineates the division of chapters and provides a short summary of the contents of each chapter.

Chapter one: ***Background and rationale***

This chapter provides an overview of the importance of the need of electrophysiological procedures in the diagnostic process of hearing loss in young infants and the difficult-to-test population. The ABR is contrasted with the ASSR technique with regard to its potential for estimating pure tone behavioral thresholds. The use of ASSR in estimating functional gain in the young population is discussed. The rationale for the study and the problem statement is provided. Definitions of the terms and concepts fundamental to this study are provided and clarified.

Chapter two: ***Clinical application of Auditory Evoked Potentials in infants: Comparing the auditory brainstem response and auditory steady state response***

The current procedures of choice for early intervention for infants are discussed – considering early identification and diagnosis of hearing loss and amplification for infants with hearing loss. Attention is given to the diagnostic process and hearing aid fitting in the young infant population - focusing on the problems and challenges. A critical discussion of AEP's in pediatric audiology will follow thereafter – comparing the ABR method with the ASSR method.

Chapter three: ***Research Methodology***

This chapter describes the operational framework implemented to conduct this study. The aims of this present study are outlined. The research design and method are discussed. The ethical issues related to this study are considered. The subjects, material and apparatus used in the study are described as well as the procedure that was followed to conduct this study.

Chapter four: ***Results and Discussion***

The results are presented according to the sub-aims stipulated in chapter three in order to address the main aim of the study. The results are presented – utilizing the results from each individual subject. Thereafter the collective results of the six subjects will be considered. Interpretation and discussion of the results are performed. The value and meaning of the research findings in relation to other studies and literature in this regard is discussed.

Chapter five: ***Conclusions and Implications***

The results from this study are summarized. This chapter provides an outline of the significant results and the way they contribute to current literature. Using *critical appraisal* methods, the research evidence are assessed – considering the value, validity, reliability and relevance thereof. Future research recommendations are provided and a conclusion regarding the study is formulated.

1.7 SUMMARY

This chapter aimed to provide relevant background information in order to focus on the research endeavor and to provide a broad perspective of the rationale underlying the study. Attention was drawn to the infant population as a difficult-to-test population and the special need for objective audiometric measures in this population at a time when critical decisions need to be made about intervention strategies.