

CHAPTER 1

INTRODUCTION AND ORIENTATION



CHAPTER AIM: To provide an outline of the problem against a backdrop of the conditions that lead to the research question, and to provide a rationale to justify the investigation of the research problem.

“If we truly desire to afford the best possible services to children and their families, we must be willing to continually modify our clinical protocols as new evidence emerges.”

~ Fred Bess (Bess, 2000:250)

1.1 INTRODUCTION

The provision of early intervention services to children with hearing loss begins with an accurate early diagnosis of the hearing loss and the fitting of appropriate amplification (Kuk & Marcoux, 2002:504-505). This forms the foundation on which a child with a hearing loss can start to develop auditory-oral communication skills in order to function optimally in a variety of environments and to materialise the main goal of auditory habilitation programs, namely, the development of these skills comparable to that of their normal-hearing peers (Mecklenburg et al., 1990, as cited in Blamey et al., 2001:265). Consistent audibility of all speech sounds in a number of listening environments is critical for the development of speech and oral language skills (Kuk & Marcoux, 2002:504; Palmer, 2005:10; Stelmachowicz, Hoover, Lewis, Kortekaas, & Pittman, 2000:209). Guidelines for best practice in the process of diagnosis and fitting of appropriate amplification have been compiled (Paediatric Amplification Guideline in Bentler et al., 2004) to ensure consistent audibility for the child with hearing loss. These guidelines provide a comprehensive overview of candidacy for amplification, diagnostic test batteries, selection of appropriate amplification, verification and validation of the hearing aid fittings, and suggest a structure for follow-up appointments and further referrals as deemed necessary.

Despite these guidelines, children with a moderate to severe sensorineural hearing loss (MSSHL) still do not seem to benefit from mere amplification of the sound signal (Miller-Hansen, Nelson, Widen, & Simon, 2003:106). Altering the sound signal by

applying frequency transposition was consequently considered a possible solution, but problems in sound quality remained. Recently, an alternative hearing aid processing strategy, namely, linear frequency transposition has been introduced. However, very little outcomes data of the possible effect that this type of frequency transposition might have on children's speech perception, is available to date.

Since word recognition is a fundamental aspect of speech perception and language development, the proposed study will focus on the possible effect that linear frequency transposition may have on word recognition abilities of young children with moderate-to-severe sensorineural hearing loss.

1.2 BACKGROUND AND RATIONALE

As the auditory system of children with normal hearing develops, improvements in the threshold where they start to respond in the presence or absence of noise are noted. Their temporal resolution, frequency discrimination and frequency resolution also improve, enabling them to detect very small differences between frequencies (Hall & Mueller, 1997:432). This auditory speech perception capacity is the ability of the auditory areas in the cortex and sub-cortex to conduct accurate representations of the sound patterns of speech to the higher centres of the brain (Boothroyd, 2004:129). Aslin and Smith (1988, as cited in Carney, 1996:30) also introduced the idea of a hierarchical development of speech perception on three levels. The sensory primitive level constitutes the most peripheral level of auditory development, and its primary focus is on detection of sound, whether it is present or absent. The next level is the perceptual representation level and it involves the categorising of sounds based on acoustic features, before recognition and comprehension of the signal takes place. Auditory processing occurs within the central auditory nervous system in response to auditory stimuli (Lucks Mendel, Danhauer, & Singh, 1999:22) and at the cognitive/linguistic representation level the acoustic features of the signal from the perceptual representation level are processed into meaningful words, rather than phonemes, and word recognition follows. The term "speech perception" thus refers to the ability to understand speech through listening (Lucks Mendel et al., 1999:242), and "word recognition" forms an integral part of speech perception as a

whole and refers to the listener's ability to perceive and correctly identify a set of words (Lucks Mendel et al., 1999:285).

Approximately 126,000 – 500,000 babies with significant hearing loss are born worldwide each year and 90% of these babies live in developing countries (Olusanya, Luxon, & Wirz, 2004:287) such as South Africa. Hearing loss in children may be caused by environmental factors, for instance prematurity, prenatal and postnatal infections (including congenital cytomegalovirus infection and rubella embryopathy), head trauma, subarachnoid haemorrhage and pharmacological ototoxicity. Up to 50-60% of hearing loss in children is also associated with syndromic and non-syndromic genetic causes (Morton & Nance, 2006:2151). It has been speculated that hearing loss in the high frequencies may be associated with “dead” areas in the cochlea (Moore, 2001:153). The inner hair cells on the basilar membrane of these areas may be either completely missing or non-functioning, preventing the transduction of vibrations along the basilar membrane (Moore & Alcantara, 2001:268). This may result in an inability to detect and process high frequency speech sounds (Miller-Hansen et al., 2003:106).

If a hearing loss is present it may lead to limited perception and resolution of the speech signal (Flynn, Davis, & Pogash, 2004:479), resulting in relaying a misrepresentation of the speech signal to the higher centres in the brain. It may finally result in a delayed or distorted cognitive/perceptual representation of oral language (Carney, 1996:32). This would result in the child missing some or all of the important acoustic cues in the speech signal, resulting in language development delays, articulation disorders and learning difficulties, depending on the degree of hearing loss. A child with a moderate hearing loss of 30-50 dB can hear vowels better than consonants, since word endings (such as –s and –ed) and short unstressed words (such as prepositions and relational words) carry less stress and may be very difficult to hear and are often inaudible. This would lead to numerous semantic as well as grammatical difficulties (Northern & Downs, 2002:22).

Speech sounds of the English language are usually described according to the way they are produced: whether they are voiced or voiceless, their place of articulation and the manner in which they are articulated (Bernthal & Bankson, 1998:16). These

properties will determine their frequency composition, relative intensities and duration (Northern & Downs, 2002:16) and sort the different speech sounds into the following sound classes: vowels (including /a/, /i/ and /o/), nasals (/n/, /m/, and /ŋ/), glides (including /w/ and /j/), liquids (including /l/ and /r/), fricatives (including /s/, /z/, /ʃ/, /f/, /v/, /ð/, /θ/ and /ʒ/), affricates (including /ts/, /tʃ/ and /dʒ/) and stops (including /p/ and /b/). Vowels, nasals, glides and liquids tend to be dominated by low spectral frequency energy, whereas strident fricatives and affricates have more energy centred in the high spectral frequencies (Bernthal & Bankson, 1998:46).

The so-called high frequency English speech sounds thus include the /s/, /z/, /ʃ/ and /ʒ/, and would be negatively affected by high frequency hearing loss (Bernthal & Bankson, 1998:45). Audibility of the /s/ and its voiced cognate /z/, play an integral role in the acquisition of speech and language skills (Kortekaas & Stelmachowicz, 2000:646). The word-final /s/ indicates plurality, possessiveness and verb tense, and is the third or fourth most common consonant in the English language (Rudmin, 1981:263). The spectral frequency region of peak energy for the /s/ varies – for the male talker it occurs at 5000 Hz and for the female and child talker at about 9000 Hz (Stelmachowicz, Pittman, Hoover, & Lewis, 2002:317). High frequency speech sounds also contain prosodic information in the auditory signal and contribute to the development of the suprasegmental aspects of speech and language (Grant & Walden, 1996:230). Decreased audibility for high frequency sounds thus puts a child at risk for phonological errors as a result of affected speech perception, and will adversely influence language development (Stelmachowicz, 2001:168; Rees & Velmans, 1993:53).

Using assistive devices that provide amplification of the speech signal may minimise the consequences and negative effects of hearing loss. This timely provision of amplification attempts to restore the original speech signal for the listener with hearing impairment, by processing and modifying the signal based on the configuration of the hearing loss. This is done on the assumption that the adjustments made to the speech signal will be adequate for the development of accurate perceptual and cognitive/linguistic representations necessary for word

recognition and discourse comprehension (Carney, 1996:32). These adjustments are, in part, dependent on the level of amplification technology that is used to process the incoming speech signal. The first generation digital conventional hearing aids utilise sequential processing, where a microchip analyses the incoming signal as a function of frequency and intensity. Each process that adjusts the signal is then performed in isolation and in succession according to the predicted needs of the listener, which could compromise the goal of amplification in a complex listening environment (Kroman, Troelsen, Fomsgaard, Suurballe, & Henningsen, 2006:3). This goal of amplification is to amplify low, moderate and high intensity sound to a level where it is audible but not uncomfortable, and to provide excellent sound quality in a variety of listening conditions (Bentler et al., 2004:46).

The second generation digital conventional hearing aids utilise parallel processing, where a microchip still analyses the incoming signal, but where like-processes can be managed simultaneously, allowing for more features and processing to happen in real-time (Kroman et al., 2006:3). However, true interactive communication between the processes is not possible, and the goal of amplification in a complex listening environment may again be compromised (Kroman et al., 2006:3). The third generation digital conventional hearing aids utilise integrated signal processing (ISP), where the central microchip allows for a two-way communication between each process of the signal, so that each process is adapted based on the “decisions” made by another process in order to tailor the output from the hearing aid. This ensures audibility of all speech sounds across a variety of listening environments (Kroman et al., 2006:4).

Paramount to successful amplification, in addition to the level of amplification technology utilised by the hearing aid, is the provision of appropriate gain. Gain refers to the difference in amplitude between the incoming and outgoing signals of the hearing aid (Dillon, 2000:7). The amount of gain (or adjustment made to the sensory primitive) that is needed to attain this goal of amplification depends on the prescription target that is used for the electroacoustical fitting of the hearing aid (Moodie, Scollie, Seewald, Bagatto, & Beaulac, 2007:1). The *Desired Sensation Level multistage input/output algorithm* (DSL m[i/o]) has been developed to address the issue of audibility in quiet as well as noisy conditions. It is aimed at normalising

loudness and prescribes amplification targets for children based on the avoidance of loudness discomfort, the audibility of important acoustic cues in conversational speech, the prescription of hearing aid compression that attempts to make soft, average and loud speech inputs available to the child, and accommodates different listening requirements within noisy and quiet listening environments (Moodie et al., 2007:9).

Another prescription algorithm is the *National Acoustic Laboratories' Non-linear Version 1* (NAL-NL1) algorithm. This formula does not attempt to restore equal loudness perception at each individual frequency, but aims at optimising speech intelligibility by taking into account the effect of sensorineural hearing loss (Dillon, 2000, p. 255). Although audibility of all speech sounds is crucial for the development and maintenance of auditory-oral communication, amplification of all speech sounds to equal loudness perception in the presence of a sensorineural hearing loss may not always seem desirable (Ching, Dillon, & Katsch, 2001:141). The benefit of high frequency amplification depends on the way residual hearing is used and how well the signal is perceived through the hearing aid (MacArdle et al., 2001:17). A study involving adults with hearing loss demonstrated that amplification of the high frequencies in some patients with dead areas may have a detrimental effect on word recognition (Hogan & Turner, 1998:440). Another study conducted by Vickers, Baer, and Moore (2001:1174) found that other patients demonstrate an increase in performance if the high frequencies 50-100% above the estimated dead area are amplified. Surprisingly, these authors also emphasised the fact that listeners with no evidence of dead areas **do** benefit from high frequency amplification. Research findings seem to indicate that it cannot be assumed that all patients with high frequency hearing loss will not benefit from high frequency amplification.

Kortekaas and Stelmachowicz (2000:657) suggested that children need more audibility in the form of greater stimulus levels, greater signal-to-noise ratios and the provision of a broader bandwidth in order to perform the same as adults with similar hearing loss. Children should be provided with adequate high frequency audibility as soon as possible after diagnosis in order to facilitate the development of auditory processing skills (Ching et al., 2001:149). Palmer and Grimes (2005:513) state that children with mild to moderate-severe hearing loss would benefit from amplification

that uses wide dynamic range compression with a low-compression threshold, moderate compression ratio, and fast attack time and which would provide increased compression to limit the maximum output of the hearing aid. Unfortunately, the fitting of digital conventional hearing aids may be of limited use to some children with MSSHL in providing high frequency amplification (Rees & Velmans, 1993:54). The amplified high frequency sounds are often still inaudible due to the severity and configuration of the hearing loss (Simpson, Hersbach, & McDermott, 2005:281). Conventional hearing aids are rarely able to provide gain above 6000 Hz and acoustic feedback may limit the high frequency gain output in the hearing aids of young children and infants despite feedback cancellation (Ricketts, Dittberner, & Johnson, 2008:160).

For the child with severe-to-profound hearing loss using only hearing aids, this goal of audible amplification may also be unattainable due to the severity of the hearing loss and the limitations of appropriate “well-fitted” hearing aids (Sininger, 2001:187; Johnson, Benson, & Seaton, 1997:91). In these circumstances a cochlear implant may be able to provide the child with more audibility so that the goals of amplification may be more realistically materialised (Stach, 1998:563). Assistive listening devices such as FM systems can also help all children with hearing impairment to detect important speech signals optimally in noisy situations by improving the signal-to-noise ratio (Northern & Downs, 2002:328).

The strive towards improving the audibility and usefulness of high frequency amplification has lead to the investigation of modifying conventional hearing aids and the effect that it would have on speech perception and word recognition. One such attempt has been to shift high frequency information (where residual hearing is poor) to lower frequencies where residual hearing is better, and therefore modifying the outgoing signal of the hearing aid (Ross, 2005). This type of processing has been recommended as an option prior to determining whether the child is a cochlear implant candidate or not, because it might provide the child with improved access to high frequency information (Launer & Kuhnelt, 2001:118). If adequate amplification is achieved with frequency lowering, it can be seen as an alternative option to a cochlear implant in some cases, as it is less costly than a cochlear implant and does not require surgery (Johnson et al., 1997:92). Therefore, amplification by means of

hearing aids may still remain the most appropriate solution for children with hearing impairment and even if a child uses a cochlear implant in one ear, fitting the non-implanted ear with a hearing aid (bimodal amplification) may preserve the residual hearing of that ear (Ching, Psarros, Incerti, & Hill, 2001:40).

There are three main types of frequency-lowering technology available at present, and these are summarised and depicted in Table 1:

Table 1: Frequency lowering circuitries available at present

CIRCUITRY	SIGNAL PROCESSING
Proportional frequency compression <i>(this term is sometimes used interchangeably with the broader term “frequency transposition”)</i>	Hearing aids that utilise proportional frequency compression shift the entire sound signal downwards by a constant factor, thus preserving the natural ratios between the frequency components of speech (Turner & Hurtig, 1999:884).
Non-linear frequency compression	Hearing aids with non-linear frequency compression compress only the high frequencies of the sound signal in increasing degrees (McDermott & Glista, 2007).
Linear frequency transposition	Linear frequency transposition technology only shifts the high frequencies of the sound signal downwards by a fixed amount and not the whole frequency spectrum (Kuk et al., 2006)

Results obtained from adult studies using proportional frequency compression have shown individual-dependent results. Mazor, Simon, Scheinberg and Levitt, (1977) found positive evidence for the practical application of this processing scheme (Mazor et al., 1977:1276). In a study conducted by Parent, Chemiel and Jerger (1997:360), an improvement in performance was noted for two of the four adult participants used in the study. Turner and Hurtig (1999:884) found significant improvements in speech recognition for many of their participants with hearing loss. McDermott and Dean (2000:359) however, found that this type of frequency lowering did not improve the speech perception of adults with steeply sloping high frequency hearing loss. Simpson et al. (2005:289) found that proportional frequency compression improves the recognition of monosyllabic words. However, in a later study by Simpson, Hersbach and McDermott (2006) using adults with steeply sloping hearing losses, no significant benefit was found when proportional frequency

compression was used (Simpson et al, 2006:629). Studies involving children and proportional frequency compression also reported varied results. MacArdle et al. (2001:27) noted that although this type of technology improved the performance of some children on speech perception tests, speech intelligibility and communication mode, this was not the case with all the children in their study. However, Miller-Hansen et al. (2003:112) reported that all the children in their study showed a significant improvement of approximately 12% on word recognition scores.

A recent development in non-linear frequency compression hearing aids has shown some promising results for adults and children with severe-to-profound hearing loss. Significant improvement in at least one speech recognition task was seen as well as an improvement in the production of high frequency speech sounds (Bagatto, Scollie, Glista, Parsa, & Seewald, 2008), and more studies are in progress to validate its efficacy.

In literature, linear frequency transposition has also produced mixed evidence. The use of a linear frequency transposition device, named a frequency recoding device (FRED), was explored in children and adults. Rees and Velmans (1993:58) found that children demonstrated a marked benefit from linear frequency transposition. However, Robinson, Baer and Moore (2007:305) found that although linear frequency transposition increased the recognition of affricates and fricatives of adults, in some cases it was at the expense of other speech sounds.

A more recent development in integrated signal processing introduced the option of linear frequency transposition that addresses the limitations of analogue signal processing and unnatural sound quality by lowering only the frequencies that are necessary and applying the correct amount of processing to the signal, as well as preserving the temporal structure of the original signal (Kuk et al., 2006). These hearing aids are sophisticated devices designed to provide optimum audibility of speech by using integrated signal processing in order to provide high definition sound analysis and sound processing. Multiple programs designed for specific listening situations can be stored in the hearing aid, including a program dedicated to linear frequency transposition. The hearing aid defaults to a master program as a start-up program and all the other programs are based on this program. The program

dedicated to linear frequency transposition can also be set as the start-up program, depending on the needs of the hearing aid user.

It has been shown that adult listeners using this particular device demonstrated improved recognition of high-frequency sounds (Kuk et al., 2006). Another study indicated that linear transposition resulted in better perception of consonants, that the improvement was seen without an acclimatisation period or prior experience with the hearing aid, and that more improvement was seen by using low-input stimuli (Kuk, Peeters, Keenan, & Lau, 2007:63).

Disadvantages of previous frequency lowering hearing aids include the higher cost compared to conventional hearing aids, the requirement of specialised knowledge of the fitting process, the inclusion of a habilitation program, wearing more apparatus and more hardware maintenance (Johnson et al., 1997:92). It would therefore seem that the most recently developed linear frequency transposition may be a suitable option for the child with a high frequency hearing loss, as it aims to improve the sound quality and addresses some of the limitations (raised by Johnson et al., 1997:92), namely:

- the hardware and apparatus used with this device is the same as for a conventional hearing aid of the same manufacturer, and is thus available in ear-level hearing aids, as opposed to the earlier body-worn versions.
- information on the fitting procedure is readily available from the manufacturer and involves little deviation from standard fitting procedures of conventional hearing aids. Therefore it does not require extensive and technical fitting procedures.
- a habilitation program does not seem to be necessary to experience **initial** benefit from the device as improvement in speech perception was seen without prior experience with the hearing aid (Kuk et al., 2007:62).

1.3 RESEARCH QUESTION

All of the above-mentioned studies have been conducted using adults as participants. Limited studies using linear frequency transposition have been

documented with children, but all have shown promising results. Rees and Velmans (1993:58) found that congenitally deaf children's ability to discriminate between auditory contrasts of high frequency consonants was improved without prior training, and that a hearing loss exceeding 70dB averaged over the frequencies 4000, 5000, 6000, 7000 and 8000 Hz in the better ear was a reliable indicator whether a child may benefit from a transposition hearing aid or not. Auriemmo, Kuk and Stenger (2008:54) also presented two case studies where better production of high frequency speech sounds, increased word recognition performance and awareness of environmental sounds were demonstrated in children with steeply-sloping hearing loss.

Due to the complex nature of high frequency hearing loss, high frequency amplification poses a challenge to paediatric audiologists (Moodie & Moodie, 2004:247). Studies providing evidence for the efficacy of using frequency transposition in children seem to be limited, but linear frequency transposition may be beneficial for children during the critical period for acquiring speech and language skills (Kuk et al., 2006) as it may theoretically provide the child with hearing impairment with more audible high frequency information that may lead to better speech perception. Stelmachowicz (2001:174) stresses that a distinction should be made between a **decrease in performance** and a **failure to observe** an improvement when working with hearing-impaired children and dealing with the issue of high-frequency amplification, as high frequency amplification may not provide much benefit in quiet environments, but may be helpful when listening in noise.

Therefore, the need exists for data on the performance of children using linear frequency transposition in quiet and in noise. Clear candidacy criteria and case studies reporting on the degree and configuration of the hearing loss and other concomitant factors are also needed (Gravel & Chute, 1996:269-270). Thus, in light of the above discussion, the following question arises:

Does linear frequency transposition affect the word recognition abilities of children with moderate-to-severe sensorineural hearing loss, and if so, in which way?

1.4 OUTLINE OF CHAPTERS

The chapters of this study are presented as follows:

Chapter 1: In this chapter the background and rationale of the research question is discussed and the outline of chapters is presented. Definitions of terms are provided as well as clarification of acronyms used throughout the study.

Chapter 2: The second chapter provides a context-specific discussion of the prevalence and aetiology of MSSHL in children. Communicative, educational, and socio-emotional outcomes of this population are presented against the backdrop of the variables that may affect these outcomes.

Chapter 3: In Chapter three an overview of the normal development of the auditory system is presented and the neurophysiology of the auditory system is linked with word recognition. Several theories of word recognition are discussed, and the effect of deprivation on the ability to recognise spoken words is considered. Assessment of word recognition in children is discussed briefly.

Chapter 4: Chapter four focuses on hearing aid technology and children. Conventional advanced digital signal processing schemes as well as frequency transposition technology are described, and placed within the context of evidence-based principles.

Chapter 5: In this chapter a description of the methodology is provided. The aims of the research are stated and the research design is explained. Selection of the participants is described, as well as the data collection and research procedures. A detailed account of the ethical considerations is also provided.

Chapter 6: Chapter six presents a discussion of the results obtained according to the aims set for this study. In addition, a discussion and interpretation of the results are provided.

Chapter 7: In the final chapter, the conclusions and clinical implications for the study are stated. The study is critically evaluated together with recommendations for future research.

1.5 DEFINITION OF TERMS

The following terms are defined in order to clarify their meaning for this study.

Cochlear dead areas/regions

An area in the cochlea where the inner hair cells are non-functioning, preventing transduction of the sound in that region (Moore, 2001:153).

Fast attack time

A diminished amount of time the compressor of the hearing aid needs to react to an increase in signal level (Dillon, 2000:161).

Frequency lowering

A general term that refers to signal processing that lowers high frequency sounds to lower frequencies (Ross, 2005).

Integrated digital signal processing

A central microchip allows for a two-way communication between each process of the signal, so that each process is adapted based on the “decisions” made by another process in order to tailor the output from the hearing aid (Kroman et al., 2006:4).

Linear frequency transposition

Frequency lowering by only shifting the high frequencies of the sound signal downwards by a fixed amount and not the whole frequency spectrum (Kuk et al., 2006).

Low-compression threshold

A low sound pressure level above which the hearing aid begins compressing in order to ensure the audibility of soft speech sounds (Dillon, 2000:165).

Maximum output of the hearing aid

The highest value of sound pressure level that the hearing aid can produce (Dillon, 2000:9).

Non-linear frequency compression

Frequency lowering by compressing only the high frequencies of the sound signal in increasing degrees (McDermott & Glista, 2007).

Parallel digital signal processing

A microchip still analyses the incoming signal, but like-processes can be managed simultaneously, allowing for more features and processing to happen in real-time, but without true interactive communication between the processes (Kroman et al., 2006:3).

Proportional frequency compression

Frequency lowering by shifting the entire sound signal downwards by a constant factor, thus preserving the natural ratios between the frequency components of speech (Turner & Hurtig, 1999:884).

Sequential digital signal processing

The first form of digital signal processing where a microchip analyses the incoming signal as a function of frequency and intensity and each process that adjusts the signal is then performed in isolation and in succession (Kroman et al., 2006:3).

Speech perception

Speech processing through sound detection, speech sound discrimination, word recognition and comprehension (Thibodeau, 2000:282).

Wide dynamic range compression

Digital signal processing that results in compression that is applied more gradually over a wide range of input levels (Dillon, 2000:161).

Word recognition

The ability to correctly recognise a word by comparing it to other possibilities stored in the auditory memory (Luce, Goldinger, Auer, & Vitevitch, 2000:615).

1.6 ACRONYMS

The following acronyms are used throughout the study and are clarified as follows:

AE:	Audibility Extender
AIDS:	Acquired immunodeficiency syndrome
CMV:	Cytomegalovirus
DSL m[i/o]:	Desired Sensation Level multistage input/output algorithm
DSP:	Digital signal processing
FRED:	Frequency recording device
HIV:	Human immunodeficiency virus
ISP:	Integrated signal processing
MSSHL:	Moderate to severe sensorineural hearing loss
NAL-NL 1:	National Acoustic Laboratories' Non-linear Version 1
PTA:	Pure-tone average
QoL:	Quality of life
SII:	Speech Intelligibility Index
SNR:	Signal-to-noise ratio
WIPI:	Word Intelligibility by Picture Identification

1.7 CONCLUSION

The development of oral speech and language skills depends primarily on the audibility of all speech sounds for the child with hearing loss. Conventional hearing aids are often unable to provide children with hearing loss with sufficient high frequency information in order to develop adequate oral language skills due to the risk of feedback and the frequency spectrum of conventional amplification. This may lead to numerous semantic as well as grammatical difficulties for the hearing-impaired child acquiring oral speech and language skills. A modification of the output of hearing aids in the form of linear frequency transposition may improve the word

recognition scores of some children with hearing loss. Linear frequency transposition technology attempts to provide the listener with better audibility of high frequency sounds, by shifting high frequency information (where residual hearing is poor) to lower frequencies where residual hearing is better, by modifying the output signal of the hearing aid. Linear frequency transposition may be beneficial for children during the critical period for acquiring speech and language skills (Kuk et al., 2006) as it may theoretically provide the child with hearing impairment with more audible high frequency information that may lead to better speech perception. This study will aim to determine whether linear frequency transposition affects the word recognition abilities of children with moderate to severe sensorineural hearing loss, and if so, in which way.

CHAPTER 2

CHILDREN WITH MODERATE TO SEVERE SENSORINEURAL HEARING LOSS



CHAPTER AIM: To provide an overview of literature reporting on the child with moderate to severe sensorineural hearing loss specifically, in order to provide a holistic description of this population.

“Not only those severe and profound losses so devastating to speech and language, but even the mildest losses with their sequelae in delayed expressive language must be identified early enough to allow interventions that will lessen their problems.”

~ Marion P Downs (Northern & Downs, 2002:5)

2.1. INTRODUCTION

A comprehensive review of available data on the child with moderate to severe sensorineural hearing loss (MSSHL) needs to consist of appropriately detailed analyses of the auditory and linguistic components and mechanisms related to the child with hearing impairment. It should also aim to provide a characterisation of the “whole child” in terms of socio-emotional development as well as educational outcomes, in addition to communicative outcomes (Donohue, 2007:713). The majority of outcomes research in the field of paediatric audiology has centred mainly on the child with severe and profound hearing loss, and less focus has been placed on the child with hearing loss of lesser degrees (Donohue, 2007:713), thus exposing a gap in the data-base available to practitioners from which clinical decisions can be made. This chapter will attempt to report holistically on the state of the world’s children with MSSHL in terms of prevalence and aetiology, as well as the communicative, socio-emotional, and educational outcomes.

2.2. PREVALENCE OF MODERATE TO SEVERE SENSORINEURAL HEARING LOSS IN CHILDREN

The audiological community has strived towards the early identification of hearing loss in children during the past 60 years (Northern & Downs, 2002:259). The techniques used to accomplish this, have also developed during this time, from the introduction of the Electric 4-C group speech test (McFarlane, 1927, as cited in Northern & Downs, 2002:259) to the current screening practices using otoacoustic emission (OAE) technology and automated auditory brainstem response (A-ABR) testing (Johnson et al., 2005:664). The possible age of diagnosis has also decreased with the development of objective physiologic tests, making it possible to identify and confirm the presence of hearing loss before 3 months of age, and starting early intervention services before 6 months of age, therefore minimising the negative consequences of hearing loss on the child's language, cognition and social-emotional development (Joint Committee on Infant Hearing, 2007:898). The benefits and importance of early identification and intervention has been well-documented (Yoshinaga-Itano, 2001:221; Yoshinaga-Itano, 2003a:266; Yoshinaga-Itano, 2003b:205; Watkin et al., 2007:e699; Yoshinaga-Itano, 2004:455; Flipsen, 2008:563; Verhaert, Willems, Van Kerschaver, & Desloovere, 2008:606), and this has lead to the implementation of universal newborn hearing screening (UNHS) as the *de facto* medical/legal standard of care in the USA (White, 2003:85), where currently >90% of all newborn babies are screened (Johnson et al., 2005:663). UNHS has also found its way to the UK and other European countries, as well as Australia (Parving, 2003:154; Davis, Yoshinaga-Itano, & Hind, 2001:6; Ching, Dillon, Day, & Crowe, 2008). These projects together with the focus-shift from research conducted on children with profound hearing loss, to children with lesser degrees of hearing loss has brought about reports on prevalence data of children in the developed world with moderate to severe hearing loss specifically, and these are summarised in Table 1:

Table 1: Prevalence data for moderate to severe hearing loss
(adapted from Fortnum, 2003:158)

REPORT	LOCATION	TYPE OF HEARING LOSS	PREVALENCE OF HEARING LOSS (% or prevalence/1000)		
			Overall	Moderate hearing loss 41 to 70 dB	Severe hearing loss 71 to 95 dB
Maki-Torkko, Lindholm, Varyrinen, Leisti and Sorri (1998:97)	Finland	All	1.3	55.3%	14.2%
		Congenital	1.1	—	—
Vartiainen, Kemppinen and Karjalainen (1997:177)	Finland	Sensorineural	1.12	42.3%	15.4%
Drews, Yeargin-Allsop, Murphy and Decoufle (1994:1165)	Atlanta, USA	Not defined	1.1	>70 dB = 0.6-0.9	
Baille et al. (1996, as cited in Fortnum, 2003:158)	France	Not defined	—	>70 dB = 0.66	
Hadjikakou and Bamford (2000, as cited in Fortnum, 2003:158)	Cyprus	All	1.59	0.43	0.59
		Congenital	1.19	—	—
Haddad (1994, as cited in Fortnum, 2003:158)	England	Congenital	0.63	50-69 dB = 0.23	70-89 dB = 0.16
Nekahm, Weichbold, and Welzl-Muller (1994:199)	Austria	All	1.32	0.7	0.32
		Congenital	1.27	0.67	0.3
		Sensorineural	1.15	—	—
Van Naarden, Decoufle and Caldwell (1999:571-572)	Atlanta, USA	All	0.78	40-64 dB = 0.2	65-84 dB = 0.13
		Congenital	0.71	—	—
		Sensorineural congenital	0.54	—	—
Davis and Parving (1993, as cited in Fortnum, 2003:158)	Denmark	Sensorineural/mixed	1.45	41%	21.8%
		Congenital	1.34	40.3%	20.8%
Davis and Parving (1993, as cited in Fortnum, 2003:158)	England	Sensorineural/mixed	1.21	47.5%	22.0%
		Congenital	1.1	51.1%	21.7%
Shiu et al. (1996, as cited in Fortnum, 2003:158)	England	All	0.86	0.35	0.23
		Congenital non-progressive	0.74	0.31	0.2
Pitt (1996, as cited in Fortnum, 2003:158)	Ireland	Sensorineural	1.23	0.76	0.16
		Sensorineural congenital	1.1	0.68	0.13
Fortnum and Davis (1997:439)	England	All	1.33	0.74	0.28
		All Congenital	1.12	0.64	0.23
		Sensorineural	1.27	0.68	0.28
		Sensorineural congenital	1.06	0.59	0.23
Uus and Davis (2000:193)	Estonia	All	1.72	0.74	0.52
		Congenital	1.52	0.69	0.45
Fortnum, Summerfield, Marshall, Davis and Bamford (2001:5)	United Kingdom	All	1.07	0.6	0.22
National Institute on Deafness and Other Communication Disorders (2005)	USA	All	1.11	30-45 dB = 1.66-2.37	45-75 dB = 0.2-0.51
Watkin, Hasan, Baldwin and Ahmed (2005:179)	England	All	1.27	0.73	0.3
Uus and Bamford (2006: e889)	England	All	1.0	38.8%	26.3%
Declau, Boudewyns, Van den Ende, Peeters and Van den Heyning (2008:1121)	Flanders	All	1-2	22%	13%
De Capua, Constantini, Martufi, Latini, Gentile and De Felice (2007:604)	Italy	Sensorineural congenital	1.78	30-50dB = 0.1	50-80 dB = 0.46

Of all the reports presented in Table 1, only four reports give an estimation of the prevalence of **sensorineural** moderate to severe hearing loss specifically (Vartiainen et al., 1997:177; Pitt, 1996, as cited in Fortnum, 2003:158; Fortnum & Davis, 1997:439; De Capua et al., 2007:604). Overall, the incidence of moderate hearing loss seems to be more prevalent than severe hearing loss. A lower prevalence for moderate to severe hearing loss was also seen for all types of hearing loss, compared to congenital hearing loss. This suggests that the inclusion of late-onset hearing loss (which is not routinely screened for) may reflect a higher, true prevalence of hearing loss in children. In the USA, an overall estimated prevalence (all types and degrees of hearing loss included) of 1.86 per 1000 births seems reasonable. This prevalence increases during childhood and reaches a rate of 2.7 per 1000 before the age of 5 years and 3.5 per 1000 during adolescence (Morton & Nance, 2006:2152). Prevalence data from the UK have shown that for every 10 infants born with hearing loss, similar late-onset hearing loss will manifest in 5 to 9 children before the age of nine years (Fortnum et al., 2001:5).

It is evident from the above-mentioned studies that the collection of prevalence data rests primarily on the availability of screening practices and manpower. This has been problematic in developing countries such as South Africa, where resource-poor countries have to deal with the challenge of infectious and deadly diseases, and non-life threatening conditions such as hearing loss, are often neglected (Olusanya, 2000:167-168). There are 146 countries in the developing world. These countries are divided into six regions, and the regions with the number of countries they represent, are depicted in Table 2 (UNICEF, 2008:148):

Table 2: The number of countries in the developing world (UNICEF, 2008)

REGION	NUMBER OF COUNTRIES
Sub-Saharan Africa (SSA)	46
Middle East & North Africa (MEN)	21
South Asia (SOA)	8
East Asia & Pacific (EAP)	29
Latin America & Caribbean (LAC)	33
Central/Eastern Europe & Baltic States	10

Although these countries are classified as developing countries, variations in per capita income, immunisation uptake and under-five mortality may vary considerably within regions and socio-economic contexts in some of the countries (Olusanya et al., 2004:289). Even though hearing impairment is seen as a non-life threatening disease, it contributes significantly towards the global burden of disease (Olusanya et al., 2007). Two-thirds of the world's population with hearing-impairment live in developing countries, and it is estimated that of the 120 million babies born each year in the developing countries, 718 000 will have a bilateral congenital hearing impairment (Olusanya & Newton, 2007:1315). Therefore, numerous small scale and pilot hearing screening studies have been initiated in some of the developing countries, and prevalence data could be derived from the results of these studies (Olusanya et al., 2004:291-293). It is estimated that a prevalence rate of 4-6/1000 seems reasonable, but may underestimate the true prevalence of hearing loss in children (Olusanya & Newton, 2007:1315). This is significantly higher than the 1.86/1000 prevalence rate of the developed world, and it may be due in part to deprivation, as the child from a poorer socio-economic background might have less access to healthcare (Kubba, MacAndie, Ritchie, & MacFarlane, 2004:123). This seems to be especially true of the South African context, where it has been reported that the audiology-profession is underrepresented in the public health sector, where the majority of South Africa's population receive their medical care (Swanepoel, 2006:266).

Prevalence data for MSSHL specifically is limited, but some of the small scale and pilot studies conducted in the developing countries distinguished between the different degrees of hearing loss, and these prevalent rates are presented in Table 3:

Table 3: Prevalence of MSSHL in developing countries

REPORT	LOCATION	POPULATION	PREVALENCE OF HEARING LOSS (% or prevalence/1000)		
			Overall	Moderate hearing loss 41 to 70 dB	Severe hearing loss 71 to 95 dB
Olusanya, Wirz, and Luxon (2008:997)	Nigeria	Infants	5.2	0.75	2.3
Olusanya, Okolo, and Ijaduola (2000:175)	Nigeria	School-aged	33.2	5.5	–
Abdullah et al. (2006:61)	Malaysia	Infants	4.2	1.0	–
Mukari, Tan, and Abdullah (2006)	Malaysia	Infants	1.6	0.6	
Yee-Arellano, Leal-Garza, and Pauli-Muller (2006:1866)	Mexico	Infants	1.63	1.6	–
Habib and Abdelgaffar (2005:840)	Western Saudi Arabia	Infants	0.18	0.3	0.5
Chapchap and Segre (2001:35)	Brazil	Infants	1.07	0.4	0.2

In addition, Westerberg et al., (2005:522) have found a prevalence of 3.4/1000 moderate to moderate-to-severe hearing loss in school-aged children in Zimbabwe. Unfortunately, this includes conductive and mixed losses as well, and may not reflect the true prevalence of sensorineural hearing loss for that population. Due to the lack of prevalence data, it has been *estimated* that in 1999 the following number of children 0 to 19 years of age presented with MSSHL (Table 4):

Table 4: Estimated number of children (1000s) 0 to 19 years of age with MSSHL

(compiled from Davis and Hind, 1999:S52)

REGION	TOTAL POPULATION OF CHILDREN	NUMBER OF CHILDREN WITH MSSHL
	AGED 0 to 19 years of age	
World	6 228 254	3 317
Developed countries	1 277 963	463
Developing countries	4 950 291	2854
Africa	856 154	622
Latin America	522 962	289
North America	305 881	115
Europe	523 749	174
Oceania	30 967	13
Asia	3 691 579	1975

It is well-known that prevalence may differ across ethnic, cultural and genetic backgrounds (Fortnum, 2003:162). If the overall prevalence rate of sensorineural hearing loss in children is higher in the developing world, it can reasonably be

assumed that the specific prevalence of MSSHL in children would also be higher in South Africa compared to the developed world.

2.3. AETIOLOGY OF MODERATE TO SEVERE SENSORINEURAL HEARING LOSS IN CHILDREN

The aetiology of MSSHL is defined in terms of time of onset of hearing loss: it may be either congenital/early-onset, or acquired (late-onset or progressive) (Smith, Bale, & White, 2005:881). MSSHL can be attributed to **genetic** and **non-genetic** causes of hearing loss (Pappas, 1998:53). **Genetic** hearing losses are associated with chromosomal abnormalities and can be classified according to the mode of inheritance: autosomal dominant, autosomal recessive and X-linked, and can either manifest as syndromic or non-syndromic hearing loss. The specific location where a gene responsible for hearing loss resides on the chromosome is called a specific locus name. A locus name consists of a pre-fix followed by a number. Autosomal dominant loci are represented by the pre-fix DFNA, autosomal recessive loci by the pre-fix DFNB and X-linked by a DFN (Mazzioli et al., 2008). Syndromic hearing loss usually has other distinctive clinical features associated with the hearing loss, of which the hearing impairment may be an inconstant or clinically significant feature, depending on the different mutations of the genes responsible for the hearing loss (Morton & Nance, 2006:2153). Nonsyndromic hearing loss has no other clinical features associated with the hearing loss, and accounts for up to 70 to 80% of genetic deafness (Nance, 2003:113).

Non-genetic forms of hearing loss are not associated with chromosomal abnormalities, and the hearing loss is usually caused by environmental factors. These factors can be divided into three categories: prenatal, perinatal and post-natal factors (Morzaria, Westerberg, & Kozak, 2004:1194). Prenatal factors cause hearing loss that are present at birth, but not necessarily detectable and perinatal factors may be those that lead to hearing loss, at birth or soon thereafter (Fortnum, 2003:159-160). Post-natal factors may be responsible for hearing loss that is acquired post-lingually (Roizen, 2003:124).

Although most forms of sensorineural hearing loss are caused by either genetic or non-genetic factors, the interaction between these two factors cannot be denied (Smith et al., 2005:881). Chromosomal abnormalities may lead to a heightened susceptibility for environmental factors like noise (Noben-Trauth, Zheng, & Johnson, 2003:21) or aminoglycosides (Morton & Nance, 2006:2160). A major difficulty in determining the aetiology of hearing loss is that not all permanent childhood hearing loss may be identifiable at birth, either due to late-onset, or due to the mild degree of the hearing loss at birth, and thus may be not detectable by modern hearing screening practices (Fortnum, 2003:155). A further problem with the determination of the aetiology of hearing loss is that for a large number (up to 44%) of diagnoses, the aetiology may remain unknown (Declau et al., 2008:1122). The known aetiology of MSSHL in children is outlined in Figure 1. It is clear from Figure 1 that causes of hearing loss are almost equally represented in genetic and non-genetic aetiology.

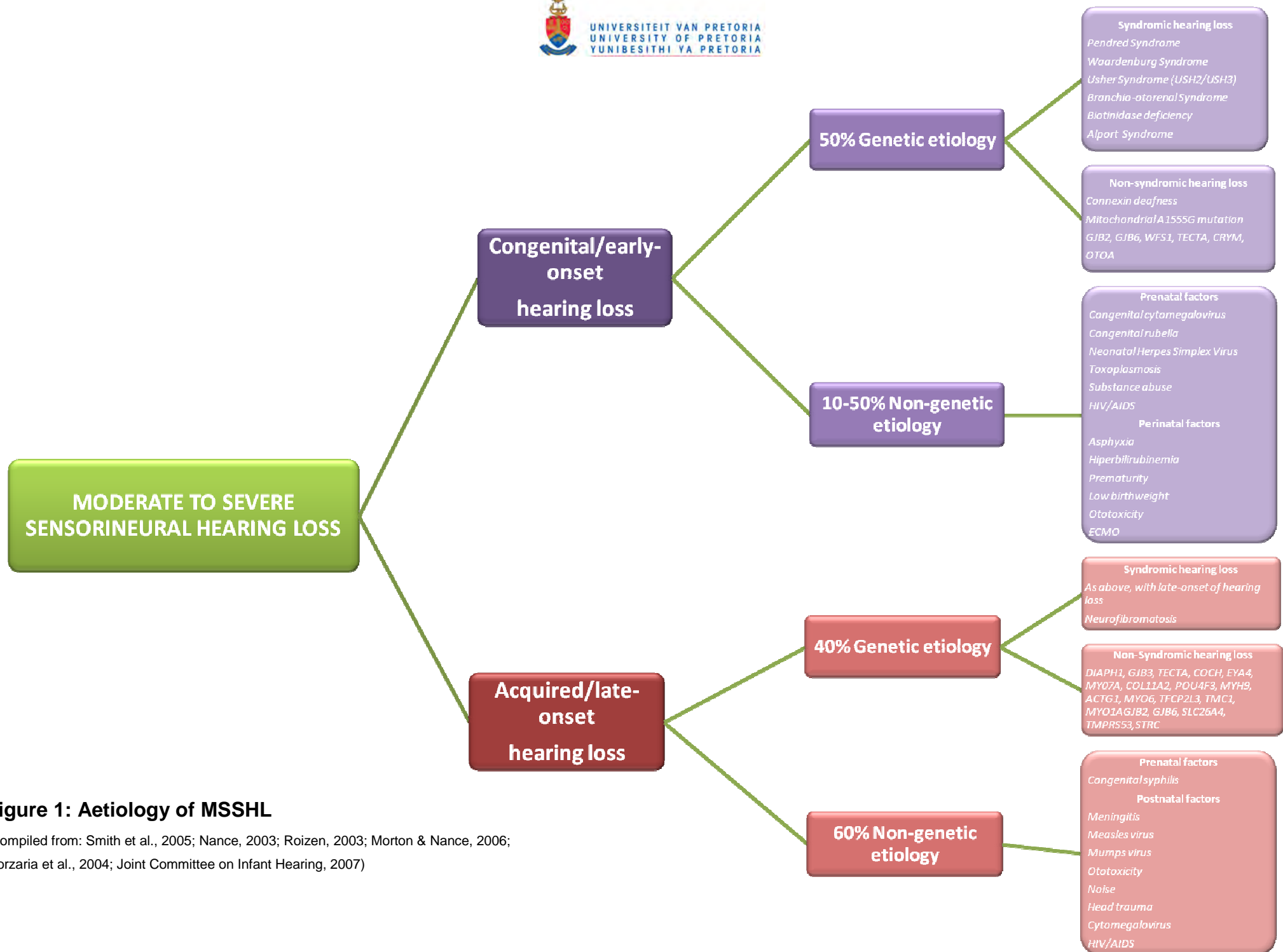


Figure 1: Aetiology of MSSHL

(Compiled from: Smith et al., 2005; Nance, 2003; Roizen, 2003; Morton & Nance, 2006; Morzaria et al., 2004; Joint Committee on Infant Hearing, 2007)

2.3.1 Genetic syndromic hearing loss

About 400 forms of syndromic hearing loss have been identified (Nance, 2003:110), of which Alport, Branchio-otorenal, Norrie, Pendred and Waardenburg syndromes are the most significant where the aetiology of MSSHL is concerned (Van Camp & Smith, 2008). Syndromic hearing loss accounted for 2.3% of the aetiologies of congenital hearing loss in a study conducted by Declau et al. (2008:1122). This corresponds with the percentage of syndromic hearing loss as stated by Morzaria et al. (2004:1195), where a prevalence of 3.15% was found. Of the 3.5% of syndromic hearing loss, 1.92% was accounted for by Waardenburg Syndrome and 0.32% for Pendred Syndrome. This is in contrast with literature stating that Pendred is the most common form of syndromic hearing loss (Morton & Nance, 2006:2154; Smith et al., 2005:882). Usher syndrome is also a common form of syndromic deafness, and is associated with retinitis pigmentosa (RP), a progressive disorder of degeneration of the retina (Nance, 2003:111). Three subtypes exist, namely Usher syndrome type I, II and III. Type II and III are usually associated with less severe sensorineural hearing loss than Type I, which is associated with profound sensorineural hearing loss (Rosenberg, Haim, Hauch, & Parving, 1997:317). Biotinidase deficiency is an autosomal recessive inherited trait and results from a deficiency of the enzyme responsible for encoding vitamin biotin. It is associated with skin rashes, seizures, hair loss, hypotonia, vomiting and acidosis (Nance, 2003:112). In a study by Wolf in 1985 (as cited in Straussberg, Saiag, Korman, & Amir, 2000:270), high frequency hearing loss was found in 40% of the 31 patients, and was reversed in some of the cases when treated with biotin. However, in most cases the hearing loss remained a constant feature (Straussberg et al., 2000:270). Neurofibromatosis Type 2 is associated with bilateral masses on the VIIIth nerve, and a unilateral acoustic neuroma, or bilateral neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity (Neary et al., 1993:6). Hearing loss may vary from mild to profound, and is usually late-onset (Neary et al., 1993:8-9).

In some forms of syndromic hearing loss, the hearing impairment can be the most salient feature, either detectable at birth or late-onset/progressive. Thus it is important that newly diagnosed infants with hearing loss be referred to other specialists (developmental paediatricians, neurologists, ophthalmologists,

nephrologists, and cardiologists) to rule out any other associated hidden clinical features (Joint Committee on Infant Hearing, 2007:908).

2.3.2 Genetic non-syndromic hearing loss

It is reported that 70 to 80% of genetic hearing loss is nonsyndromic (Nance, 2003:113). The identification of genes and their loci responsible for sensorineural hearing loss has developed rapidly over the last 5 years and the rapidly evolving knowledge regarding the genetics of hearing loss, has contributed considerably towards the understanding of the aetiology of sensorineural hearing loss (Smith et al., 2005:881). Table 5 depicts the number of loci of the causal genes identified by 2003 compared to those in 2008:

Table 5: The number of loci of causal genes (Van Camp & Smith, 2008; Nance, 2003)

LOCI OF CAUSAL GENE	2003	2008
Autosomal dominant	41	57
Autosomal recessive	33	77
X-linked	5	8

The genes and their loci depicted in Table 5 are responsible for all types of genetic non-syndromic hearing loss (degree and time of onset of hearing loss included). Genes and their loci have been identified which are responsible for MSSHL with prelingual and postlingual onset specifically, and these are depicted in Tables 6 and 7:

Table 6: The genes and their loci responsible for prelingual moderate to severe sensorineural hearing loss in children (compiled from Morton and Nance, 2006)

LOCUS	GENE	HEARING LOSS
DFNA3	<i>GJB2</i>	<ul style="list-style-type: none"> Moderate to profound K⁺ recycling defect May be some hearing at birth
	<i>GJB6</i>	<ul style="list-style-type: none"> Moderate to profound K⁺ recycling defect High frequency progressive
DFNA6/14	<i>WFS1</i>	<ul style="list-style-type: none"> Moderate to severe Low frequency with tinnitus
CRYM	<i>CRYM</i>	<ul style="list-style-type: none"> Moderate to profound Possible K⁺ recycling defect
DFNB1	<i>GJB2</i> & <i>GJB6</i>	<ul style="list-style-type: none"> Some moderate to profound Some pass UNHS
DFNB22	<i>OTOA</i>	<ul style="list-style-type: none"> Moderate Hair cell defect

Table 7: The genes and their loci responsible for postlingual moderate to severe sensorineural hearing loss in children (compiled from Morton and Nance, 2006)

LOCUS	GENE	HEARING LOSS
DFNA1	<i>DIAPH1</i>	<ul style="list-style-type: none"> Moderate to profound Low frequency progressive Hair cell defect Onset 1st – 4th decade
DFNA2	<i>GJB3</i>	<ul style="list-style-type: none"> Moderate to severe High frequency progressive Tinnitus K⁺ recycling defect Onset 4th – 6th decade
	<i>KCNQ4</i>	<ul style="list-style-type: none"> Moderate High frequency progressive Vertigo K⁺ recycling defect
DFNA4	<i>MYH14</i>	<ul style="list-style-type: none"> Moderate to profound Fluctuating, progressive Hair cell defect
DFNA5	<i>DFNAS</i>	<ul style="list-style-type: none"> Moderate to severe High frequency progressive
DFNA8/12	<i>TECTA</i>	<ul style="list-style-type: none"> Severe U-shaped/high frequency progressive Tectorial membrane defect
DFNA9	<i>COCH</i>	<ul style="list-style-type: none"> Moderate to profound High frequency progressive Tinnitus, vertigo, poor balance Endolymphatic hydrops Onset 2nd – 7th decade
DFNA10	<i>EYA4</i>	<ul style="list-style-type: none"> Moderate to severe U-shaped progressive Defective transcription factor 1st – 4th decade
DFNA11	<i>MYO7A</i>	<ul style="list-style-type: none"> Moderate to severe High frequency progressive Onset 1st – 6th decade Haircell defect
DFNA13	<i>COL11A2</i>	<ul style="list-style-type: none"> Moderate to severe U-shaped Tectorial membrane defect
DFNA15	<i>POU4F3</i>	<ul style="list-style-type: none"> Moderate to severe Progressive Onset by 5th decade Defective haircell transcription factor

(Table 7 continued)

DFNA17	<i>MYH9</i>	<ul style="list-style-type: none"> • Moderate to profound • High frequency progressive • Haircell defect
DFNA20/26	<i>ACTG1</i>	<ul style="list-style-type: none"> • Moderate • Progressive • Defect in intracellular cytoskeletal protein
DFNA22	<i>MYO6</i>	<ul style="list-style-type: none"> • Moderate to profound • Progressive • Onset by 5th decade • Haircell defect
DFA28	<i>TFCP2L3</i>	<ul style="list-style-type: none"> • Moderate to severe • Progressive • Onset by 5th decade • Defective transcription factor
DFNA36	<i>TMC1</i>	<ul style="list-style-type: none"> • Moderate to profound • Rapidly progressive by 3rd decade • Defective transmembrane protein in haircells
DFNA48	<i>MYO1A</i>	<ul style="list-style-type: none"> • Moderate to severe • Progressive • Probable haircell defect
DFNB4	<i>SLC26A4</i>	<ul style="list-style-type: none"> • Variable high frequency hearing loss • Enlarged vestibular aqueduct
DFNB10	<i>TMPRSS3</i>	<ul style="list-style-type: none"> • Moderate • Progressive
DFNB12	<i>CDH23</i>	<ul style="list-style-type: none"> • High frequency
DFNB16	<i>STRC</i>	<ul style="list-style-type: none"> • High frequency • Stable

About 75 to 80% of prelingual non-syndromic hearing loss is autosomal recessive inherited. Twenty to 25% is thought to be autosomal dominant, and 0.15 to 1.5% is X-linked (Declau et al., 2008:1124). Although the aetiology of a large section of hearing loss is unknown, prospective studies are predicting that 30 to 50% of hearing loss with unknown origin may in fact be a form of nonsyndromic hearing loss (Morzaria et al., 2004:1197).

Despite the heterogeneity of nonsyndromic hearing loss, mutations of one gene, GJB2, are responsible for up to 50% of autosomal recessive nonsyndromic hearing loss (Kenneson, van Naarden, & Boyle, 2002:262). GJB2 are responsible for encoding connexin 26, a protein that is found largely in the cochlea and that is important for the maintenance of K⁺ homeostasis during the transduction of auditory stimuli (Snoekx et al., 2005:946). Biallelic truncating and biallelic nontruncating mutations of GJB2 were found to be the most prevalent in children with MSSHL (Snoekx et al., 2005:949).

Mitochondrial defects also cause nonsyndromic hearing loss. Mutations of A1555G can cause a hypersensitivity to aminoglycosides, but hearing loss without

aminoglycoside exposure has also been documented (Hutchin & Cortopassi, 2000:1928).

Consanguinity is mentioned as a factor related to the higher prevalence of genetic deafness in children in the developing world (Attias et al., 2006:533). Family intermarriage is practised as a social custom, and it was found that 10 to 12% of children whose parents are related, have a genetic hearing loss (Zakzouk, 2002:813).

2.3.3 Non-genetic causes of MSSHL in children

Non-genetic causes of hearing loss have been remarkably reduced over the past 30 years due to the development of vaccines and antibiotic treatment. Furthermore, developments in technology and medicine have lead to an increased number of neonatal intensive care unit (NICU) survivors, who are presenting as a medically complex, developmentally at-risk population with a further risk of hearing loss (Roizen, 2003:120). The reduction in number of non-genetic causes of hearing loss are directly related to the availability of vaccines, medicine and technology, and in resource-poor countries, these may be unattainable, leading to an increased number of non-genetic causes of hearing loss in developing countries (Olusanya & Newton, 2007:1316). This implies that the child has poorer access to vaccinations against infections such as mumps, measles and rubella, and specific illnesses such as malaria and tuberculosis might be rudimentary treated with ototoxic drugs. Neonatal jaundice accounts for a higher prevalence of hearing loss in children in developing countries as well, and injuries sustained from armed conflicts and noise-related disasters could also have an effect on the prevalence-rate of childhood hearing loss in the developing world (Olusanya & Newton, 2007:1316).

The human immunodeficiency virus (HIV) causes the acquired immunodeficiency syndrome (AIDS). This syndrome causes a progressive immunological deficit, and creates vulnerability for infectious diseases (Matas, Leite, Magliaro, & Goncalves, 2006:264). HIV/AIDS is considered a global pandemic, with an estimated 2 million children living worldwide with the disease, and 230 000 – 320 000 children are estimated to live with HIV in South Africa (UNAIDS, 2008). Sensorineural hearing

loss associated with HIV/AIDS, may manifest as a direct result of the effects of the virus on the peripheral auditory nerve, or as a secondary effect to infections and the administration of ototoxic drugs (Matas et al., 2006:264).

Studies reporting on the proportion of prevalence of pre-, peri- and post-natal factors have produced slightly different results. This prevalence is depicted in Table 8:

Table 8: Prevalence of pre-, peri- and post-natal factors

Study	No of children	%Prenatal	%Perinatal	%Post-natal	%Unknown
Maki-Torkko et al., 1998	112	52.7	8.0	1.8	37.5
Vartiainen et al., 1997	52	53.8	9.6	11.5	25.0
Nekahm et al., 1994	165	37.0	22.4	4.2	36.4
Fortnum, Marshall and Summerfield, 2002	17160	33.8	8.0	6.9	49.4

It is important to note that these studies included genetic and non-genetic causes in their estimation of prenatal factors, but perinatal and postnatal factors all refer to non-genetic causes. Morzaria et al. (2004:1197) have found a prevalence of non-genetic prenatal factors of 12%, perinatal 9.6% and postnatal 8.2%. A large number of diagnoses' aetiology remains unknown, and the differences in prevalence between different studies may be due to the different environments and locations where these studies were conducted (Fortnum, 2003:161).

Prenatal factors

Prenatal non-genetic causes of MSSHL are usually defined as intra-uterine infections (congenital cytomegalovirus, rubella, toxoplasmosis, syphilis and herpes infections), and substance abuse during pregnancy. Since the introduction of universal rubella vaccine in 1982, cytomegalovirus (CMV) infections have replaced rubella as the most prevalent cause of non-genetic congenital hearing loss (Declau et al., 2008:1125; Morton & Nance, 2006:2158). The prevalence of CMV in children with late-onset moderate-to-severe hearing loss can be as high as 35% (Barbi et al., 2003:41), and is present in only 3.9% of all infants with hearing loss at birth (Morton & Nance, 2006:2158). Congenital CMV may be clinically asymptomatic at birth, but half of the 10% of infants who do present with clinical symptoms at birth, have sensorineural hearing loss of varying degrees (Smith et al., 2005:883). Thus it is

important to continually monitor children where CMV infection is a concern, due to the high incidence of delayed-onset of hearing loss.

In the developed world the rubella vaccination has successfully eliminated rubella as a cause for hearing loss (Vartiainen et al., 1997:183). Currently, rubella vaccine is available in 123 countries, and confirmed cases of congenital rubella syndrome was decreased by 98% from 1998 to 2006 (WHO, 2008). In countries where widespread vaccination is not implemented, congenital rubella is still the most prevalent non-genetic cause of hearing impairment (Banatvala & Brown, 2004:1130). Sixty-eight to 93% of children born with congenital rubella syndrome present with sensorineural hearing loss (Roizen, 2003:123).

Herpes simplex virus infection is also a non-genetic cause of hearing loss in children. Although intra-uterine infection is rare, most mother-to-child transmissions occur during delivery (Whitley et al., 1991). Hearing loss occurs when the transmission was intra-uterine (Westerberg, Atashband, & Kozak, 2008:935), and has been reported as moderate to severe in degree (Dahle & McCollister, 1988:257). Congenital syphilis is a condition that has increased in prevalence during the past 20 years, and hearing loss resulting from this condition is usually late-onset, high-frequency and progressive, and may develop with vertigo (Roizen, 2003:123). Hearing loss occurs in 3% of children with congenital syphilis (Valley, 2006:4). Congenital toxoplasmosis may lead to hearing loss in 10 to 15% of children with this infection, but may be preventable with timely treatment (McGee et al, 1992). Hearing loss can be mild-severe, and stable or progressive (Noorbakhsh, Memari, Farhadi, & Tabatabaei, 2008).

Substance abuse during pregnancy may also have a profound effect on the foetus, and this has also been reported to cause hearing loss in children (Morzaria et al., 2004:1195). Foetal alcohol syndrome may affect hearing as a sensorineural, conductive or central hearing disorder (Church & Abel, 1998). The ingestion of ototoxic drugs during pregnancy may also cause a high frequency progressive hearing loss in the unborn child (Roizen, 2003:123). These drugs may cause aplasia of the inner ear, damage to the inner and outer hair cells, absence of VIIth and VIIIth nerves, dysplasia of the organ of Corti, and a decreased number of ganglion cells.

Prenatal exposure to trimethadione and methyl mercury and an iodine deficiency have all been associated with congenital hearing loss occasionally (Jones, 1997, as cited in Roizen, 2003:123).

Perinatal factors

Perinatal factors are present in adverse neonatal events, and constitute prematurity, low birth weight, hyperbilirubinemia, and ototoxic drug therapy. The Joint Committee on Infant Hearing (2007) has identified the following perinatal risk-indicators associated with permanent hearing loss:

- NICU-stay for more than five days
- NICU-stay for any of the following, independent of length of stay: extracorporeal membrane oxygenation (ECMO), assisted ventilation, ototoxic drug therapy or exposure to loop diuretics (furosemide/Lasix) and hyperbilirubinemia that requires an exchange transfusion

Prematurity in infants require more intensive care in the perinatal period, leading to a higher prevalence of respiratory disorders, and to a higher exposure to ototoxic medications than term infants (Marlow, Hunt, & Marlow, 2000:141). This may be inevitable, due to the fact that the neonate's survival is at stake. ECMO is used when there is acute, reversible respiratory or cardiopulmonary failure in neonates, and is a form of prolonged cardiorespiratory bypass. This allows the lungs of critically ill neonates to rest and to avoid oxygen toxicity (Fligor, Neault, Mullen, Feldman, & Jones, 2005:1519). However, this kind of therapy has shown to be associated with a high incidence of neurodevelopmental disorders, such as hearing loss. The duration of the time that the child receives ECMO, is correlated with an increased risk of sensorineural hearing loss. If the duration was more than 160 hours (6 to 7 days), then the child is >7 times more likely to develop sensorineural hearing loss than a child who only received the therapy of <112 hours (Fligor et al., 2005:1526). The hearing loss associated with ECMO can range from mild to profound, and may be delayed in onset and progressive (Fligor et al., 2005:1523).

The use of diuretics also has an adverse effect on the neonate's hearing and the longer the duration of diuretic-use, the higher the prevalence of sensorineural

hearing loss (Robertson, Tyebkhan, Peliowski, Etches, & Cheung, 2006:221). Furosemide inhibits the Na-K-2CL transporter system in the stria vascularis, and this leads to oedema and a decreased endocochlear potential. Other ototoxic drugs such as aminoglycosides, vancomycin and neuromuscular blockers may also cause hearing loss that is usually high frequency, and may be progressive and late-onset (Robertson et al., 2006:219).

Bilirubin-induced pathology of the auditory system may usually lead to a bilateral symmetric high frequency hearing loss, and an incidence of moderate-to-severe hearing loss has also been reported, and the amount and duration of hyperbilirubinemia are both risk factors (Shapiro, 2003:413).

Postnatal factors

Postnatal factors are mostly associated with bacterial meningitis infections, and in rarer cases with head trauma, noise and ototoxic drugs. Bacterial meningitis is seen as the most prevalent cause of acquired hearing loss in childhood. The vaccination for Haemophilus Influenza Type B (Hib) has eliminated meningitis as a cause for hearing loss in most countries, but other strains of bacteria can cause hearing loss for which there is no current vaccination (Koomen et al., 2003:1049). Seven percent of children who survive non-Hib bacterial meningitis presents with sensorineural hearing, of which the degree can vary from severe to profound. Hearing loss may deteriorate, and ossification of the cochlea may compromise later cochlear implantation (Koomen et al., 2003:1051).

Measles is also a preventable highly infectious viral disease which is associated with sensorineural hearing loss in children. The illness presents with high fever, running nose, Koplik's spots on the buccal mucosa and a distinctive generalised maculopapular rash. In some cases the measles virus can be found in the cochlea, thus leading to sensorineural hearing loss. The triple mumps, measles and rubella (MMR) vaccine has been proved efficient in preventing measles (Olusanya, 2006:7). Mumps infection affects the salivary glands, and the incidence of mumps-related sensorineural hearing loss is estimated to be 5/100,000. Hearing loss is usually profound, but milder losses have also been reported. The introduction of the MMR

vaccine also resulted in a decline in the overall incidence of mumps (Olusanya, 2006:8).

Ototoxic drugs are mainly administered to young children to fight infections or cancer (Knight, Kraemer, & Neuwelt, 2005:8588). These drugs include cis-platinum (oncology drug), acetylsalicylic acid, aminoglycosides, chloramphenicol, chloroquine phosphate, dihydrostreptomycin, neomycin, nitrogen mustard, nortriptyline, pharmacetin, polymyxin B, quinine, ristocetin, streptomycin, thalidomide, vancomycin and viomycin (Roizen, 2003:123). Evidence has also been produced that noise is a growing factor in the aetiology of acquired hearing loss in children. Children are also exposed to hazardous levels of noise (fire crackers, toys, portable stereos, referee whistles, musical concerts) which may cause a noise-induced hearing loss (NIHL). Twelve percent of children from a study in the US have presented with NIHL in at least one ear. Of these children, 4.9% had a moderate to profound hearing loss, typical of noise-induced hearing loss with a notch in the high frequencies at 3000, 4000 and/or 6000 Hz (Niskar et al., 2001:41). In this study, NIHL was significantly more pronounced in boys than in girls, and more prevalent in older children. Head trauma has also been reported to cause moderate-to-severe hearing loss and the type and degree of the hearing loss depends on the site of lesion in the skull and brain, and most hearing loss resolves after a period of time (McGuirt & Stool, 1991, as cited in Roizen, 2003:124).

HIV/AIDS is also a prevalent viral perinatal cause of hearing loss in the developing world. The virus affects the CD4+ T cells of the immune system, and several causes of hearing loss are linked to HIV infection (Newton, 2006:11-12). Although most of these causes are also experienced by HIV negative children, they may present with more severe forms of hearing loss if they are HIV positive (Newton, 2006:12). The most common causes of sensorineural hearing loss due to HIV infection are acute/recurrent otitis media, otosyphilis, medications, HIV infection of the cochlea and VIIIth nerve, and opportunistic infections (Newton, 2006:11).

2.4. OUTCOMES OF CHILDREN WITH MSSHL

The experiences and outcomes of children with moderate to severe hearing loss and their families and the accessibility of audiological services depend strongly on the socio-economic context in which they reside (Swanepoel, Delpont, & Swart, 2007:3). However, the socio-economic context consists of a number of variables that may influence the outcomes of a child with hearing impairment (Ching et al., 2008), and this is depicted in Figure 2:

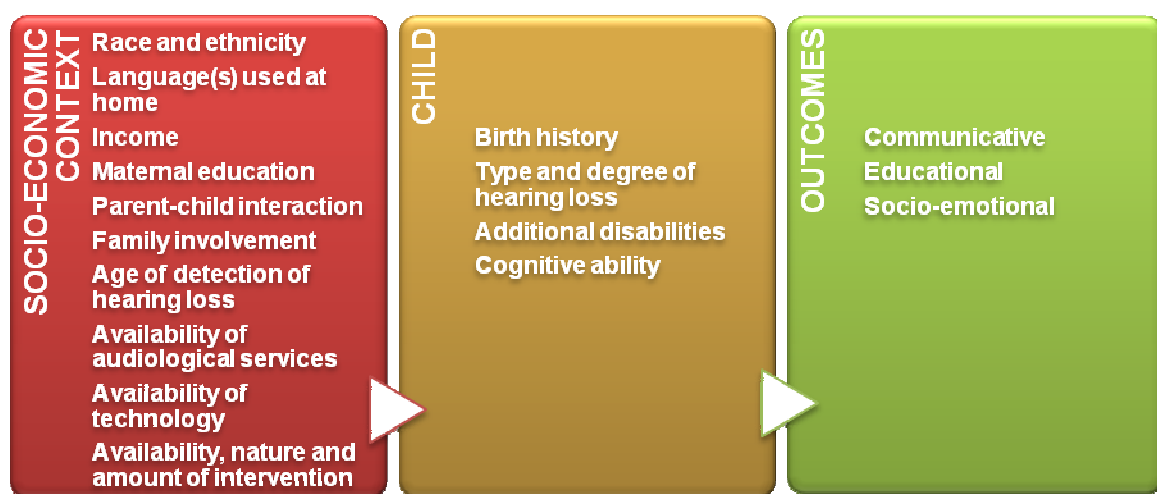


Figure 2: Variables related to the outcomes of children with MSSHL (adapted from Ching et al., 2008)

The socio-economic context and the characteristics of the child with hearing impairment inter-relate closely, and this may have a profound effect on the outcomes of the child. The effect of these variables on the outcomes of children with MSSHL will be discussed in the following sections.

Race, ethnicity, and languages used at home

According to the results obtained from the 2001 census, the South African population of 44.8 million can be divided into four races (Statistics South Africa, 2003). The distribution of these groups is depicted in Figure 3:

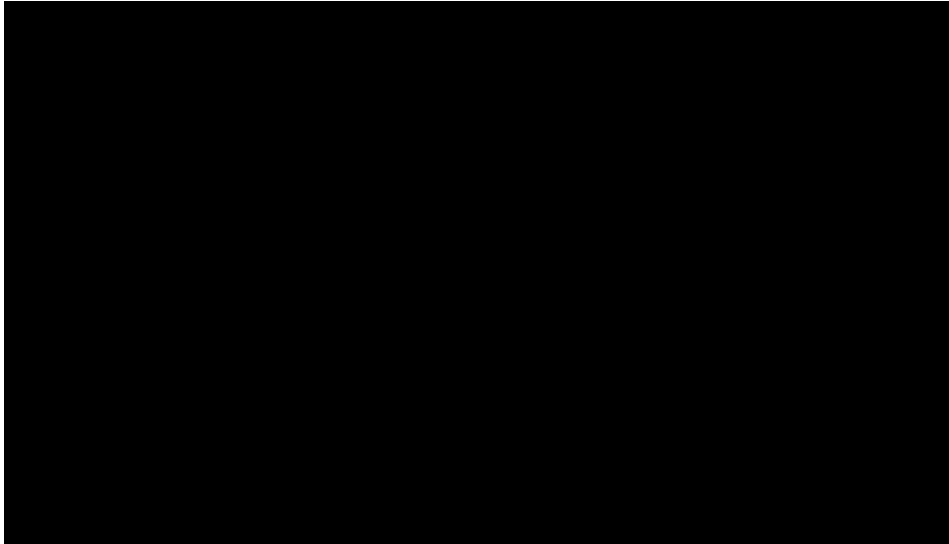


Figure 3: The distribution of race in the South African population (compiled from Statistics South Africa, 2003)

The Black African racial group is considered the majority (79%), with each of the other groups comprising 2 to 10% of the whole population (Statistics South Africa, 2003). Furthermore, within these races, a wealth of languages and cultures are found. South Africa has eleven official languages, and the percentage of people using these languages at home is depicted in Figure 4:

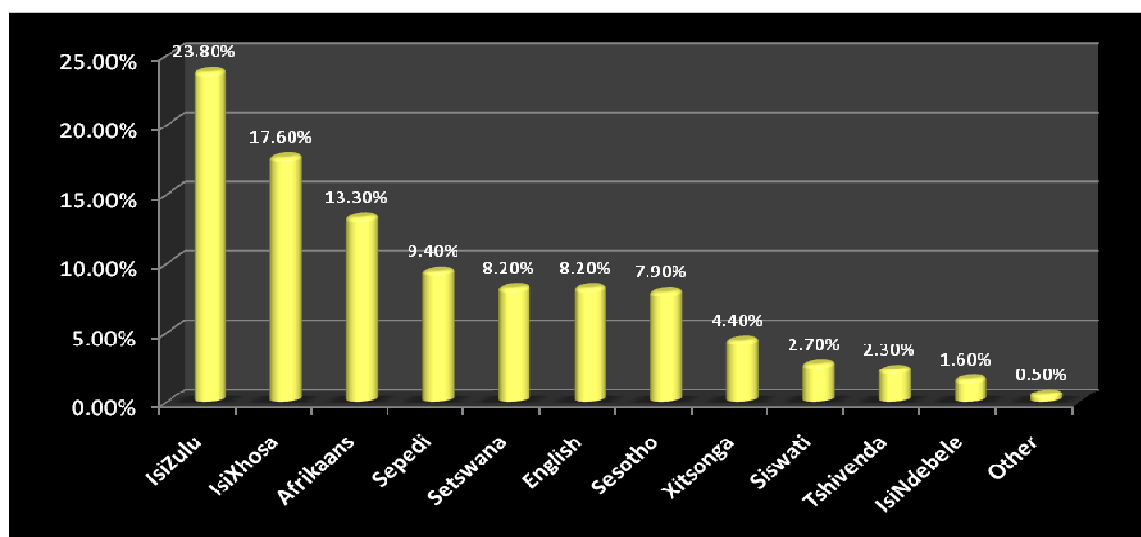


Figure 4: The percentage of people using each of the eleven official languages at home (compiled from Statistics South Africa, 2003)

It can be seen from these statistics that IsiZulu is spoken by 23.8% of the population, and IsiNdebele by 1.6%. This would affect the outcomes of the child with hearing impairment negatively if the intervention and educational services were not provided in the language spoken at home (Swanepoel, 2006:265).

Income

The gross national income per capita was estimated to be \$5390 for 2006 (UNICEF, 2008). However, 55% of South Africa's children are considered "ultra-poor" and live in households of a monthly income of ZAR800 or less, and 14.5% of South Africans live in make-shift shacks with no running water, toilets, electricity, and other basic services (UNICEF, 2007). This creates significant challenges for the implementation of intervention services for children with hearing impairment, and the costs of transport to programs offering intervention, costs of care and maintenance of amplification devices *et cetera*, may create a vast obstacle to the successful habilitation of hearing loss.

Maternal education, parent-child interaction, and family involvement

Parents can affect all three domains of a child's development, namely communicative, educational and socio-emotional outcomes, and these domains influence each other (Calderon, 2000:141). Earlier studies have indicated that maternal education was a significant predictor of parental involvement, which leads to better child outcomes (Stevenson & Baker, 1987:1356). A study by Calderon (2000) indicated that maternal education alone did not influence child outcomes significantly, and a shared communication mode was required. Maternal communication skills and interaction were found to be a prerequisite for parental involvement, and both were found to be predictors of the child's outcomes (Calderon, 2000:151). Late-identified children from families with low involvement are at risk for poor outcomes (Moeller, 2000:6). Maternal sensitivity was also found to be a predictor of language outcomes, with strong language gains made when mothers were more sensitive to the child's attempts to communicate (Pressman, Pipp-Siegel, Yoshinago-Itano, & Deas, 1999:302). Cultural differences in parental involvement and interaction may thus manifest in the acceptance of hearing loss, which might lead to the late identification of hearing loss and a fatalistic passive approach to intervention, which might affect outcomes negatively (Louw & Avenant, 2002).

Age of detection of hearing loss

The general consensus seems to be that earlier identification of hearing loss and subsequent intervention lead to improved outcomes for children with MSSHL (Yoshinaga-Itano, 2001:221; Yoshinaga-Itano, 2003a:266; Yoshinaga-Itano, 2003b:20; Watkin et al., 2007:e699; Yoshinaga-Itano, 2004:455; Flipsen, 2008:563; Verhaert et al., 2008:606). In the absence of universal neonatal hearing screening, the average age of diagnosis for this population is 24 to 30 months of age (Yoshinaga-Itano, 2003b:199). Children with MSSHL were generally later-identified than children with more severe degrees of hearing loss, and outcomes are negatively affected by the delay in identification and intervention (Watkin et al., 2007:e697). The majority of children with MSSHL in South Africa are still subjected to late identification (if identified), with ramifications in all outcomes of these children.

Availability of audiological services, intervention services, and technology

For most countries of the developed world, accessibility to audiological services seems to be well within reach for each child with hearing impairment. As mentioned before, newborn hearing screening programs are now widely implemented in these countries in order to facilitate early detection of hearing loss, followed by well-designed multi-disciplinary intervention programs (Joint Committee on Infant Hearing, 2007:908). Resources are also usually available to provide the child with amplification technology that meets the basic requirements set by the Paediatric Amplification Guideline (Bentler et al., 2004; Lutman, 2008; Bamford et al., 2001:214) and these requirements are depicted in Table 9:

Table 9: Basic requirements of circuitry-signal processing (Bentler et al., 2004:49)

BASIC REQUIREMENTS
<ul style="list-style-type: none"> • The system should be distortion-free • It should be possible to shape the frequency/output response of the system to meet the audibility targets of an appropriate prescriptive method • Tolerance issues should be avoided by frequency/output shaping based on a prescriptive method • Amplitude processing should be employed by the system to ensure audibility from a wide range of input-levels. Wide dynamic range compression may be necessary to allow for optimal audibility. • Output limiting should be possible independent of other sounds in the dynamic range • Sufficient electro-acoustic flexibility should be present in order to compensate for characteristics related to the growth of the child.

Although these requirements are only basic, it may provide the child with moderate to severe hearing loss with the first step towards an equal opportunity to develop spoken speech and language skills comparable to those of his/her normal hearing peers (Joint Committee on Infant Hearing, 2007:908).

Despite the well-documented positive effect that early hearing detection and intervention (EHDI) programs have on the communicative, educational, and socio-emotional outcomes of children with MSSHL, equal opportunities still remain largely out of reach for those children with hearing loss residing in developing countries with poor resources (Swanepoel, Hugo, & Louw, 2006:1241). However, some countries in the developing world such as Brazil, Oman and Chile, have implemented newborn hearing screening programs in multiple cities, thus making it clear that EHDI programs in the developing world are a feasible and viable possibility (Olusanya et al., 2007:13).

South Africa is also considered a developing country, although there is co-existent developed and developing contexts (Swanepoel, 2006:262). Eighty-five percent of the South African population is served by the public health sector, and only 15 to 20% of the population have the resources to afford private health care (National

Treasury Department, Republic of South Africa, 2007). Audiologists are employed in both the public and private health care sectors, with the majority of audiologists employed in the private sector. This creates an inverted relationship between the audiological manpower and the population they serve, with only a small number of adequately trained professionals available to serve the majority of the hearing-impaired population (Swanepoel, 2006:264).

Both the developed and developing contexts of South Africa have explored the feasibility of UNHS in the form of small-scale pilot studies (Swanepoel et al., 2006:1246; Swanepoel, Ebrahim, Joseph, & Friedland, 2007:884; Theunissen & Swanepoel, 2008:S28). Private hospitals and the subsequent intervention services usually comprise of world-class medical personnel and advanced equipment, while hospitals and services in the public health sector are still considered equal to those in the developing world (Saloojee & Pettifor, 2005). Therefore, only a small number of infants in South Africa are being identified with hearing loss before the age of 6 months (Theunissen & Swanepoel, 2008:S28). In some of the public hospitals in South Africa, children are given a priority and those with MSSHL are fitted with digital signal processing hearing aids (Coetzee, 2008, personal communication.) These hearing aids meet the basic requirements set by the Paediatric Amplification Guideline (2004) and would affect the outcomes of children positively. In the private sector, advanced signal processing hearing aids are available to those who can afford them, and this would increase the opportunity of children with MSSHL to develop successful oral spoken language skills. Unfortunately, amplification technology is only available once the child has been diagnosed, and due to the lack of screening programs, many children with MSSHL are not identified, and fail to develop sufficient spoken language skills (Theunissen & Swanepoel, 2008:S25; Yoshinago-Itano, Sedey, Coulter, & Mehl, 1998:1169).

Intervention services for children with MSSHL are rendered by clinicians, speech-language therapists, audiologists, special nursery schools for the deaf and hard of hearing children, and community-based programs (Friedland, Swanepoel, Storbeck, & Delport, 2008).

Birth history, type and degree of hearing loss, additional disabilities and cognitive ability

An analysis of the birth history would determine whether there are any prenatal or perinatal causes of hearing loss that would have an impact on the type and degree of hearing loss. This is important, as the type and degree of hearing loss would have an effect on the management of the hearing loss, which would affect the outcomes of children with hearing loss (Northern & Downs, 2002:19). Twenty-five to 40% of children with hearing loss have additional disabilities, which would affect outcomes as well (Tharpe, Fino-Szumski, & Bess, 2001:32). Cognitive ability would also play an unfavourable role in the outcomes of children, as language development is often delayed (Owens, 1999:29).

All of the discussed factors play an intricate role in the outcomes of children with MSSHL, and the communicative, educational, and socio-emotional outcomes will be discussed in the following section.

2.4.1 Communicative outcomes of children with MSSHL

Infants with normal hearing, who are developing typically, acquire significant motor and auditory/perceptual experiences as they go through the prelinguistic vocal stages, with a progressive move towards approximating speech with their vocalisations (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:606). Four prelinguistic stages of vocal development have been identified in literature (Oller & Eilers, 1988:441):

- **Phonation stage (0 to 2 months):** “comfort sounds” with normal phonation are mainly produced, which are precursors to vowel production. Syllables are rare during this stage.
- **Gooing stage (2 to 3 months):** phonetic sequences are produced that are a combination of the sounds of the previous stage, paired with sounds formed at the back of the vocal cavity. These sounds may be the precursors to consonant production, are usually not well-formed and mature.
- **Expansion stage (4 to 6 months):** a variety of new sounds are introduced in this stage, namely raspberries, squeals, growls, yells, whispers, isolated

vowel-like sounds and marginal babbling, which is a precursor to syllable production.

- **Canonical stage (7 to 10 months):** production of reduplicated sequences such as /mamama/, /bababa/, and /dadada/. True syllable production is apparent, which are the precursors to true words.

Interaction between infants and adults also develops in different stages. During the first 9 months of life, adult-child interactions are mainly visual and involve social transactions or physical manipulations of objects. The development of joint attention and pointing as a means of collaborating with adults follows, and is closely linked with the development of reference, which is necessary for the learning of new words (Zaidman-Zait & Dromi, 2007:1167). Deictic gestures (showing, giving, reaching, pointing) as well as referential gestures (the symbolic manual label of objects and actions) start to emerge around the same time as first words (Caselli & Volterra, 1990, as cited in Zaidman-Zait & Dromi, 2007:1167).

These prelinguistic stages are very important in the transition to words, as first words tend to contain syllables and consonants mastered during the prelinguistic stages (Ferguson & Farwell, 1975, as cited in Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007b:629). The stabilisation of vocal-motor control is also regarded as a prerequisite for the emergence of first words (McCune & Vihman, 2001, as cited in Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007b:629). The single-word stage is characterised by utterances that contain babble, jargon, unintelligible word attempts, as well as true words, which usually contain simple syllable structures, like CV, CVC and CVCV (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007b:629). Development of phonological skills continues during the second year of life, although intelligibility is usually limited (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007b:630). Typically developing 2 year olds learn about two to nine words per day (Golinkoff et al., 2000, as cited in Lederberg & Spencer, 2008:1), and between 24 and 30 months they acquire the ability to learn the meaning of new words in situations where the speakers give no direct cues for reference (Lederberg & Spencer, 2008:2). By 3 years of age, a typically developing child should be able to produce between 900 and 1000 different words in 3 to 4 word utterances that usually contain a subject and

a verb (Owens, 2008:454). By 4 years of age a child should use utterances that contain basic structure rules, like subject-noun-verb or noun-verb-object. Their utterances should contain at least 6 words, and they should begin to use auxiliary and modal verbs. By 5 years of age a child should have a vocabulary of at least 1500 words, and should speak clearly in nearly correct sentences (Owens, 2008:454).

It is imperative for a child to have hearing thresholds of 15 dB or better in order for all these skills to develop normally and on time (Northern & Downs, 2002:14). If a child with MSSHL is not fitted with appropriate amplification, most conversational speech sounds will be inaudible and language and speech may not develop spontaneously. Vowels may be heard better than consonants, and in some cases only when spoken at a close range. The endings of words and short unstressed words may be very difficult to hear or may even be inaudible (Northern & Downs, 2002:22). However, with appropriately fitted amplification, these children may respond well to language and educational activities, and with intervention may function very well (Northern & Downs, 2002:22).

At present, there seems to be a lack of consensus regarding the impact of degree of hearing loss on communication outcomes (Moeller, Tomblin, Yoshinaga-Itano, McDonald, & Jerger, 2007:740). Studies have shown that vocalisations of the precanonical stages of children with normal hearing and hearing impairment have some similarities (Oller & Eilers, 1988:448), but the onset of canonical babbling was found either to be within normal limits (Davis, Morrison, Von Hapsburg, & Warner Czyz, 2005:21), a little delayed with a few months (Nathani, Oller, & Neal, 2007:1426), or substantially delayed (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:612). In the latter study, children with MSSHL did not meet the criteria for canonical babbling until 14 – 20 months of age. Small sample sizes, age of amplification and the ability to control for all the variables influencing communication development may account for these differences. Auditory experience is a key factor in the development of canonical babbling (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:621), and this would be affected by earlier versus later provision of amplification and hearing aid retention once amplification has been provided. Early identified children with MSSHL may present with smaller

repertoires of consonants than their normal-hearing peers, on average between 5.5 and 9 consonants at 13 to 18 months of age, and between 8 and 12 consonants between 19 and 24 months of age (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:622). Children with hearing impairment may also be slower in their development of the production of fricatives and affricates (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:623). Possible explanations for this phenomenon may be two-fold, namely the complexity of this class of speech sounds, and the limited bandwidth of conventional hearing aids that would provide sufficient audibility of these speech sounds to develop (Moeller, Hoover, Putman, Arbataitis, Bohnenkamp et al., 2007a:623; Stelmachowicz, Pittman, Hoover, & Lewis, 2001; Stelmachowicz et al., 2002). Consonant blends are absent in the phonetic repertoires of children with MSSHL up until 31 to 42 months, and initial blends start to emerge at this age (Yoshinaga & Sedey, 1998). The development of vowels seems to be near age-matched norms during the first year of life for children with MSSHL, although this development is more marked in this population than in children with profound hearing loss (Nelson, Yoshinaga-Itano, Rothpletz, & Sedey, 2008:118). Speech production characteristics of a population of 5 to 14 year old children with moderate-to-severe hearing loss reflected that all these children had at least one deviant or borderline-deviant speech/voice behaviour, typically longer than usual voice-onset time or a higher fundamental frequency (Higgins, McCleary, Ide-Helvie, & Carney, 2005:553).

The development of receptive vocabulary in early identified children between 8 to 22 months of age is also delayed in comparison to normal-hearing peers, but this delay is expected to be less pronounced than in later-identified children (Mayne, Yoshinaga-Itano, Sedey, & Carney, 1998). In a study conducted by Mayne et al., (1998), it was found that the expressive vocabulary of early-identified children with MSSHL aged 32 to 37 months of age fell below the 25th percentile for children aged 30 months with normal hearing. For later identified children, it was found by Davis et al. (1986:57), that vocabulary development was delayed by one to three years for children with MSSHL older than 12 years of age. This seems to be apparent as well in younger later-identified children with MSSHL. A Swedish population of late-identified 4 to 6 year old children demonstrated a delay in vocabulary development of 1.5 to 2 years (Borg, Edquist, Reinholdson, Risberg, & McAllister, 2007:1076).

Studies involving expressive and receptive vocabulary are closely related to the ability to learn new words. Gilbertson and Kamhi (1995, as cited in Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:742), found that half of their population with mild to severe hearing loss demonstrated novel word learning skills comparable to their normal-hearing peers. The other half showed a significant difficulty in learning phonologically complex words and required more trials (or repetitions) of the novel words in order to learn them. Lederberg, Prezbindowski and Spencer (2000:1581) found that children with MSSHL were delayed in the development of rapid-word learning skills, but this development improved in explicit naming contexts. Children with MSSHL were also found to categorise words appropriately into semantic categories, and this skill deteriorated as the degree of hearing loss increased (Jerger et al., 2006). Syntactic development is also impaired by the presence of MSSHL. Elfenbein, Hardin-Jones and Davis (1994, as cited in Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:745), found that patterns of development were delayed in comparison with normal hearing children, and that complex syntax, verb structures, bound morphemes and pronouns are amongst those errors most frequently observed. Results from a study conducted by McGuckian and Henry (2007:27-28), showed that children with moderate hearing loss are not simply delayed in their acquisition of grammatical morphemes, and that the order of acquiring of these morphemes are similar to those children acquiring a second language. Interestingly, the morphemes that the children with hearing impairment had the most difficulty with, were the third singular –s, past –ed, and possessive –s. These are, according to Brown (1973, as cited in McGuckian & Henry, 2007:29), the least frequent occurring morphemes, and children do not have as much access to these morphemes compared to others. However, as mentioned earlier, limited bandwidth characteristics of conventional amplification may also decrease the amount of auditory input children with moderate hearing loss have for these morphemes (Stelmachowicz et al., 2001; Stelmachowicz et al., 2002).

Overall, a study by Yoshinaga-Itano, Coulter, and Thomson (2000:S133), showed that early-identified and fitted children with hearing impairment have an 80% chance of developing communication skills comparable to their normal-hearing peers by 5 years of age. However, due to the lack of screening practices in South Africa, the

majority of children with MSSHL are still identified late, and may be subjected to poor communication outcomes due to this late-identification.

2.4.2 Educational outcomes of children with MSSHL

Children should be linguistically prepared for the educational setting, otherwise significant delays in language skills may result in academic, socio-emotional and self-esteem challenges (Moeller, 2000:7). Five factors have been introduced which might predict the educational setting where a child with hearing loss will receive a formal education: hearing capacity, language competence, nonverbal intelligence, family support, and speech communication attitude (Northern & Downs, 2002:357). These five factors are all interrelated, but a degree of hearing loss and the resulting language skills seem to be the most important factor regarding choice of educational setting, as this may affect the child's ability to comprehend the curriculum, to follow directions and classroom rules, to conduct themselves with appropriate classroom behaviours, to follow discussions, all the while using appropriate language and intelligible speech (Matkin & Wilcox, 1999:149). Educational methodologies differ from the mode of communication used as the language of learning. These methodologies can be divided into three categories: auditory-verbal (auditory/oral) approach, which uses spoken language alone for communication and teaching, manual communication, which relies on signs and/or finger spelling, and total communication, which utilises the simultaneous use of speech and signs with finger spelling (Northern & Downs, 2002:358). Children with severe and less than severe sensorineural hearing loss tend to be included in programs where oral speech is the primary mode of communication (Karchmer & Mitchell, 2003:26). A study by Madden et al. (2005:1195) reported on the educational outcomes of 21 children infected with CMV. Of these 21 children, seven presented with MSSHL. These seven children made use of auditory-verbal or total communication and were either included in mainstream settings or special education. Those included in the special education settings had additional disabilities like cognitive impairment, and made use of total communication. The children who were included in mainstream schools had also been identified before 6 months of age, thus most probably providing them with the opportunity to spend more time in intervention and with amplification, thus raising

their chances to develop oral speech and language skills comparable to their normal-hearing peers (Moeller, 2000:7).

Further and formal education for children in South Africa with hearing impairment are provided in the form of 37 special schools for the Deaf, three special schools for the hard of hearing, and eight units for the Deaf and hard of hearing attached to schools primarily providing for other disabilities (National Institute of the Deaf, 2004). Inclusive education was introduced in 2001 by the South African Education White Paper no 6 (2001), which states that children with disabilities may be placed in a range of different educational environments, namely from ordinary schools to special schools or resource centres. It is envisioned that full service schools will be developed in order to provide support for the whole range of learning needs, but at present this has not been implemented, and there is still a reliance on pull-out programs for children with disabilities in the general education classroom (Yssel, Engelbrecht, Oswald, Eloff, & Swart, 2007:357). Although inclusive education has been introduced in 2001, children with disabilities have been integrated in the mainstream schools since 1994 (Yssel et al., 2007:357). These children may also have been early-identified children with MSSHL, fitted appropriately with amplification shortly after diagnosis, who attended early intervention services with the emphasis on development of spoken language on a regular basis, and whose family support have been adequate.

Academic achievement is rated with regards to grade-to-grade advancement, as well as the mastery of curricular units (Karchmer & Mitchell, 2003:27). Of particular interest is the development of **literacy** in children with MSSHL, which is defined as “a sociocultural activity of meaning construction using text” (Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:746). Literacy comprises both reading and writing skills, and forms an integral part of the education of a child. Poor oral language skills have been found to be the greatest indicator of reading difficulties, with specific reference to the development of phonological processing abilities and the development of lexical, sentence, and discourse processes (Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:746-747). Auditory experience may form the foundation on which good reading ability can be build, and children with hearing impairment may be especially at risk for reading difficulties

(Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:746-747). Older studies have reported that on average, the reading comprehension of children with MSSHL were one or two grade levels below those of the hearing peers (Stinson & Antia, 1999:168). Surprisingly, newer studies have reported no evidence for poorer reading skills compared to hearing peers (Briscoe, Bishop, & Norbury, 2001:338; Gibbs, 2004:24), but a very recent study by Most, Aram and Andorn (2006:19-25), found that children with MSSHL presented with poorer word recognition, phonological awareness, letter identification, and orthographic knowledge than age-matched hearing peers.

New technology and new practices may be significant in improving the educational outcomes of children with MSSHL (Moeller, Tomblin, Yoshinaga-Itano, McDonald Connor, & Jerger, 2007:749), but it is expected that children who are not exposed to this technology and these practices will continue to perform less than optimally in educational settings, due to poor language skills.

2.4.3 Socio-emotional outcomes of children with MSSHL

A child with MSSHL is at risk in the area of psychosocial development due to the increased risk of communicative delays, limited access to communicative exchanges and effects like noise, reverberations and distance (Moeller, 2007:729). Quality of life (QoL), the social-emotional context of early learning, self-concept and identity formation, and social access have been identified as areas of psychosocial development that may be influenced by MSSHL (Moeller, 2007:730).

Quality of life

QoL may be defined as an “overall mental and physical health and well-being” (Moeller, 2007:730). Hind and Davis (2000:200) identified nine categories of QoL that are applicable to families with children with hearing impairment. These categories are:

- Communication
- Health
- Independence

- Family activities
- Family functioning
- Relationships
- Roles
- Wealth
- Work

It has been shown that these areas that affect QoL may be subjected to the degree of hearing loss, as stated in Hind and Davis (2000:204). All of these categories may be affected in families with children with MSSHL, but it has been reported that these families report less impact on QoL as families with children with severe to profound sensorineural hearing loss. Also, the child's communication and the time spent with the child, had large effects on the families' QoL, and 50% of the families with children with moderate hearing loss reported that the family health is affected by the child's hearing impairment (Hind & Davis, 2000:205). Overall, QoL was found to be significantly lower for children with hearing loss than for hearing children (Keilmann, Limberger, & Mann, 2007:1750; Petrou et al., 2007:1050).

The social-emotional context of early learning

A two-way communication process develops between caregiver and infant from a very early stage, where both parties are aware of the other's emotions and respond accordingly (Louw & Louw, 2007:120). This interaction is crucial for the development of warm, consistent and predictable relationships between caregivers and their children. Maternal sensitivity and emotional availability have been shown to be indicators for a healthy psychosocial developmental context (Moeller, 2007:732), but the presence of a hearing loss may form a barrier to the normal development of parent-child interactions (Obrzut, Maddock, & Lee, 1999:240), thus creating a risk in the area of socio-emotional and language development of a child, and especially with the presence of MSSHL (Pressman et al., 1999:294).

Self-concept and identity

Self-concept refers to the "stable set of attitudes about the self including a description and an evaluation of one's attributes and behaviours" (Piers, 1984, as cited in Moeller, 2007:734), and is dependent on the socialisation process with family

and friends (Silvestre, Ramspott, & Pareto, 2007:40). From this definition, it is clear that hearing loss can affect the construction of self-concept and identity forming. It seems that degree of hearing loss in itself is irrelevant when it comes to the construction of self-concept (Silvestre, Ramspott, & Pareto, 2007:51), but the educational setting has more influence on self-concept construction due to the fact that peer-relationships are so important in this process (Obrzut et al., 1999:248). Self-concept findings were similar for children with hearing impairment in schools for the deaf and hard of hearing, and children with normal hearing in mainstream schools. Children with hearing loss' self-concept scores were significantly lower when placed in a mainstream school. This has been further explored in a study by Israelite et al. (2002:144), where it was found that high-school students with hearing impairment often felt marginalised when they were included in mainstream education, and that interaction with other hearing-impaired peers were of more importance in the development of meaningful peer relationships.

Social access

Social access refers to the socialisation process through which the self-concept and identity is constructed, and social relationships are affected directly by academic achievement (Moeller, 2007:735). It has been shown that a lack of peer acceptance impedes academic performance (Flook, Repetti, & Ullman, 2005:319). This is important for the professionals working with children with hearing impairment, as these children may be at risk for difficulties with peer acceptance (Moeller, 2007:735). A study by Cappelli, Daniels, Durieux-Smith, McGrath and Neuss (1995:205), revealed that children with MSSHL experience significant rejection by their peers than children with normal hearing. Davis et al., (1986:59-60) demonstrated that children with MSSHL scored significantly higher on aggression and psychosomatic complaints. They were also perceived by their parents as having greater difficulty at school, interacting with others, and establishing friendships. Externalised behaviour problems were also noted, such as impulsivity, immaturity, and resistance to discipline and structure. These children were also afraid of telling other children about their hearing aids, as they often get teased about it. Friendship formation increases academic gain, (Newcomb & Bagwell, 1995:306; Ladd, 1990, as cited in Moeller, 2007:734), and social access to normal-hearing learning groups may be limited for children with MSSHL (Power & Hyde, 2002).

There is a great need for research to report on the communicative, educational, and socio-emotional outcomes of children with MSSHL specifically, and to analyse the differences in development of early and late-identified children. These studies would yield a significant amount of insight into the difficulties experienced by these children, and would give valuable guidelines in order to provide better services to them.

2.5 CONCLUSION

All the major factors relating to children with MSSHL are closely connected with the socio-economic status of the society in which these children and their families reside. Data on the prevalence of MSSHL in children are lacking especially in the developing context, but it seems that the prevalence of MSSHL in children is much higher in the developing world than in more developed countries. Also, the aetiology of MSSHL is dependent on the availability of health resources, as immunisation and prenatal care play an integral part in the prevention of MSSHL. These variables also affect the outcomes of children with MSSHL, as the outcomes of children with MSSHL in the developed context are much more positive due to the availability of services and technology. Although by no means exhaustive, this chapter attempted to provide a deeper understanding of children with MSSHL in the developing and developed contexts.

CHAPTER 3

THE RECOGNITION OF SPOKEN WORDS: A DEVELOPMENTAL APPROACH



CHAPTER AIM: To describe the developmental processes that are necessary for the child with hearing loss to recognise spoken words.

“The hearing ear is always found close to the speaking tongue...”

~ Ralph W Emerson (1857:26)

3.1 INTRODUCTION

Speech can be described as complex sounds that are generated and shaped by the vocal organs and the structures surrounding them, such as the lungs, trachea, larynx, pharynx, nose and nasal cavities, and mouth. The movements of the structures above the larynx (called the vocal tract) generate sound sources, and these sound sources are filtered by the position of the structures in the vocal tract (Moore, 2004:301; Stevens, 1998:243). Thus, the acoustic patterns of speech are complex and constantly changing (Raphael, Borden, & Harris, 2007:214). The movements of the vocal organs are highly coordinated, and represent the audible manifestation of a specialised linguistic system, whose elements are stored in memory (Stevens, 1998:243). Words are considered to be the most familiar units of speech, and can be segmented into syllables. Syllables in turn are made up of speech sounds or phonemes (Moore, 2004:300), and these phonemes are distinct from each other based on their spectral information. Thus, the speech signal can be decomposed into a finite and well-defined set of acoustic features (Koch, McGee, Bradlow, & Kraus, 1999:305).

The speech signal can be described in terms of its temporal and spectral information. In the temporal domain, the speech sound wave consists of a series of valleys and peaks within an amplitude envelope. The differences in energy between sounds that are produced with an open vocal tract (such as vowels) and sounds that are produced with a constricted vocal tract (such as consonants), result in these

amplitude variations. Furthermore, filter characteristics of the vocal tract produce rapidly changing amplitude peaks and valleys across the frequency spectrum, resulting in the enhancement or attenuation of spectral energy in certain frequency regions (Koch et al., 1999:306). These features can be visualised by plotting the frequency and changes in frequency amplitude against time on a spectrogram (Baken & Orlikoff, 2000:243).

The manner in which speech sounds are articulated and their place of articulation, directly results in their unique spectrotemporal characteristics. Speech sounds can be categorised as sonorants, such as vowels (/i/, /e/, /a/, /u/, /o/ et cetera), glides (/j/ and /w/), liquids (/l/ and /r/) and nasals (/n/ and /ŋ/), or obstruents, such as fricatives (/v/, /f/, /ð/, /θ/, /z/, /s/, /ʒ/, /ʃ/ and /h/), stops (/b/, /d/, /p/, /t/, /k/, /g/ and /ʔ/) and affricates (/dʒ/ and /tʃ/) (Koch et al., 1999:306; Kent, 1998:10, 18). When producing sonorants, air passes relatively freely through the oral or nasal cavities. The glottis is the primary source of sound (Stevens, 1998:258) and as vowels are usually produced with voice, they are relatively high in intensity, with a rich harmonic structure with clear formants (Koch et al., 1999:306; Raphael et al., 2007:214). The frequency and the patterning of the formants are important for the listener to identify vowels (Raphael et al., 2007:214), and the formant frequency varies with tongue position when producing vowels (Koch et al., 1999:306). Changes in the formants thus lead to changes in bandwidth, and vowels have a relatively narrow bandwidth of approximately 54 to 73 Hz (Stevens, 1998:258-259). Listeners usually only require the first and second formant in order to correctly identify the vowel (Raphael et al., 2007:214). Glides typically are lower in intensity than vowels, with vowel-like formants. Liquids also have a clear formant structure, and is characterised by a sudden drop in intensity. Nasals are characterised by their strong low-frequency murmur, together with their clear formant structure (Koch et al., 1999:306), as there is a complete closure at some point in the vocal tract, but with an open velopharyngeal port (Stevens, 1998:487).

Stop consonants are characterised by a complete closure of the vocal tract, followed by a sudden burst of broadband energy, lasting no longer than 100ms. Thus, their acoustic properties consist of a period of low energy (during the closure), followed by

the broadband energy, usually in the high frequencies (Niyogi & Sondhi, 2002:1064). Fricatives are produced when a turbulent air stream passes through a constriction in the vocal tract, which produces a relatively extended period of noise caused by the friction. Fricatives can be divided into two categories, namely sibilants and nonsibilants. Sibilants (/s/, /z/, /ʃ/ and /ʒ/) have relatively steep, high frequency spectral peaks and are produced posterior compared to nonsibilants. Nonsibilants (/θ/, /ð/, /v/ and /f/) have flat spectra and are produced anterior to the sibilants (Raphael et al., 2007:226). Affricates are a combination of a stop consonant, followed by a fricative, and thus contain acoustic properties found in both classes.

Human infants typically enter the world pre-wired for the detection of the complex speech signal and its acoustic properties, with an emphasis on the learning of speech and language through the auditory modality (Werner, 2007:275). In-depth knowledge of the basic acoustic properties of the speech signal, the development of the auditory system (pre- and postnatal) and how the auditory system codes the acoustic properties of speech is of more than academic interest for the paediatric audiologist, as audition is considered the first step in speech perception (Raphael et al., 2007:207). The development of speech perception also influences the decisions regarding optimal amplification input over time, as infants and children may use different types of acoustic information at different stages, which is very useful in determining hearing aid characteristics and signal processing algorithms (Stelmachowicz et al., 2000:903).

3.2 NORMAL DEVELOPMENT OF THE AUDITORY SYSTEM

The auditory system develops and matures in the typically developing child in a fairly predictable manner. In addition, these developments are shaped by exposure to sounds and speech, and are of vital importance in order for word recognition to occur and the subsequent acquisition of language (Northern & Downs, 2002:127-128). The inner ear reaches adult full-size at 5 months gestation, and is the only sense organ that reaches full differentiation by foetal midterm (Northern & Downs, 2002:41). Thus, the foetus is exposed to fluid-conducted sound for 4 months before birth, and is physiologically ready to respond to sound by this time (Northern & Downs,

2002:128). Although babies are born with an adult-like peripheral auditory system, development and maturation of the central auditory system continues through-out the life-span, with the most rapid growth during the early years up until 3 years of age (Northern & Downs, 2002:128). These stages of development and maturation will be discussed in the following section.

3.2.1 Embryonic development and prenatal hearing

During the first three weeks after gestation, the embryo is organised in three layers as a cellular disk. These three layers are superimposed on each other, and consists of the ectoderm (responsible for the development of the skin, nervous system and senses), the mesoderm (associated with musculo-skeletal and circulatory systems, kidneys and reproductive system) and the endoderm (creates the digestive and respiratory systems). The disk is divided by the primitive streak, which continues to become the primitive groove and primitive fold. This groove deepens and becomes the ectodermal-lined neural pit. The neural folds close off to form the neural tube. The cephalic end of the neural tube is characterised by an enlargement, which is to become the head of the foetus (Northern & Downs, 2002:37). Five branchial grooves form laterally in the lower head and neck area with corresponding endodermal-lined pharyngeal pouches on the inside of the embryo. These are collectively known as the branchial arches (Northern & Downs, 2002:41). The inner ear develops from the ectoderm, the middle ear from the endoderm and the outer ear from all three layers (Northern & Downs, 2002:37). A discussion on embryonic development of the inner ear, middle ear, outer ear as well as the central auditory system follows separately.

Inner ear

Towards the cephalic end of the neural tube, two auditory placodes on each side of the tube arise from the ectoderm during the 3rd week after gestation. These placodes are the earliest beginnings of the inner ear and start to invaginate into the ectoderm on approximately the 23rd day after gestation, to become the auditory pits (Northern & Downs, 2002:38). The auditory pits close off to form the sphere-like auditory vesicles on approximately the 30th day after gestation (Martin, 1997:293). During the 5th week after gestation, the auditory vesicle divides into its vestibular and cochlear portions. At 6 weeks gestation age, the utricle and saccule is present in the

vestibular portion of the auditory vesicle, and the semi-circular canals start to form (Northern & Downs, 2002:38). One coil of the cochlea can be seen in the cochlear portion of the vesicle during the 7th week, and the sensory cells develop in the utricle and saccule. In the 8th week, the sensory cells in the semi-circular canals start to form, and by the 11th week the cochlear duct has formed 2.5 coils, and the VIIIth nerve fans its fibres across the whole length of the cochlear duct. By the 12th week, the sensory cells of the cochlea start to appear, the membranous labyrinth is complete and the otic capsule starts to ossify (Northern & Downs, 2002:40-41).

Middle ear

During the 3rd week after gestation the first pharyngeal pouch forms an elongation of the lateral-superior edge of the pouch, called the tubotympanic recess, from which the tympanic cavity and the auditory tube (later known as the Eustachian tube) originates. The tubotympanic recess approaches the embryo surface between the first and second pharyngeal branches during the 8th week (Northern & Downs, 2002:41). The tympanic cavity is present in the lower half of the recess, and the upper half is filled with mesenchyme, which is to become the ossicles (Martin, 1997:241). In the 9th week after gestation, the ectodermal groove deepens towards the tympanic cavity, until it meets the meatal plug, which consists of epithelia cells. Mesenchyme forms between the ectodermal lining of the groove and the endodermal lining of the tympanic cavity, to form the three layers of the tympanic membrane (Northern & Downs, 2002:41). At the 15th week, the cartilaginous stapes is present, and the malleus and incus start to ossify during the 16th week (Martin, 1997:241). During the 18th week ossification of the stapes begins. As the ossicles ossify, they become loose from the mesenchyme and the mesenchyme becomes less cellular and is absorbed by the membrane of the middle ear cavity. Each ossicle stays connected to the walls of the middle ear cavity with a mucous membrane, which becomes the ligaments supporting the ossicles. By the 21st week, the meatal plug disintegrates, exposing the tympanic membrane. The tympanum is pneumatized during the 30th week, and by the 32nd week ossification of the malleus and incus is complete. In the 34th week, the middle ear cavity forms outpouches which will become the mastoid cells. The antrum is pneumatized by the 35th week and the epitympanum is pneumatized by the 37th week (Northern & Downs, 2002:42).

External ear

The first evidence of the external ear appears during the 5th week after gestation, when the development of the primary auditory meatus commences from the first branchial groove (Martin, 1997:219). The ectodermal lining of the first branchial groove and the endodermal lining of the first pharyngeal pouch are in contact for a short period, during which mesodermal tissue forms between the two layers, separating the groove from the pouch (Northern & Downs, 2002:42). During the 6th week six hillocks (tissue thickenings) form on either sides of the first branchial groove. These are arranged three on a side facing each other, and become the auricle. The auricles start to move from the original ventromedial position to a more ventrodorsal position during the 7th week, as they are displaced by the development of the mandible and face. In the 8th week after gestation, the primary auditory meatus moves towards the middle ear cavity, and becomes the outer third of the external auditory meatus (Northern & Downs, 2002:44). By the 20th week, the auricle reaches adult-shape, but growth continues until the 9th year. The external auditory meatus is fully formed by the 30th week, and maturation continues until the 7th year.

Central auditory system

During the 4th week after gestation, a group of cells separate from the auditory vesicle to become the statoacoustic ganglion, which will ultimately form the VIIIth cranial nerve (Moore & Linthicum, 2007:461). The cochlear division of the VIIIth cranial nerve consists of ganglion cells, which winds around the modiolus of the cochlea, to form the spiral ganglion. Neurons from these cells extend axonal processes towards the cochlea, and towards the brainstem. The axonal processes that are developing towards the brainstem contact the brainstem neurons at 5 to 6 weeks gestational age (Cooper, 1948, as cited in Moore & Linthicum, 2007:461). The axonal processes developing towards the cochlea enter the base of the Organ of Corti in the 9th week after gestation. At 10 to 12 weeks gestational age, the axonal branches form synapses with the developing hair cells (Pujol & Lavigne-Rebillard, 1985, as cited in Moore & Linthicum, 2007:461).

Neurons of the central auditory system originate from the ventricular zone in the brain and after they turn post-mitotic, they migrate to the appropriate destination in the brain tissue (Illing, 2004:6). All auditory centres and pathways are identifiable by

7 to 8 weeks in the brainstem. Between 9 and 13 weeks, the structures increase in size, but remain in their basic configuration. On the edge of the brainstem, groups of neurons form synapses with the axons from the cochlear nerve, and is called the cochlear nuclei. From the cochlear nuclei, the axons of the trapezoid body cross the brainstem towards the superior olivary complex. Axons also ascend towards the lateral lemniscus from the cochlear nuclei towards the inferior colliculus. At the 8th week, the medial geniculate nucleus is visible on the posterior surface of the thalamus, and receives the axons from the inferior colliculus (Moore & Linthicum, 2007:461). The neurons develop visible cytoplasm, and axons that contain neurofilament proliferate in the auditory nerve, the trapezoid body, and the lateral lemniscus (Sininger, Doyle, & Moore, 1999:5). At 24 to 26 weeks, the axons start to branch at the terminal ends in their target nuclei and short dendrites are visible on the neurons of the cochlear, olivary and collicular nuclei. At 28 weeks gestation, the appearance of myelin in the auditory nerve and brain stem pathways signals the synchronised conduction of stimuli. During the last three months before term birth, the myelin increases in density and conduction velocity is rapidly increased in the auditory pathways (Sininger et al., 1999:5).

The auditory cortex matures much later than the auditory brainstem. At 4 weeks gestational age, the forebrain is divided into two cerebral hemispheres, which are balloon-like expansions containing fluid in a ventricular cavity. Cells are generated in the innermost lining of the ventricular cavity, from where they migrate towards the outer surface. At the 8th week, these cells near the surface form the cortical plate. By 22 weeks, the cortex appears thicker, and cells in the cortex express reelin, acetylcholinesterase and calcium-binding protein that may attract migrating neurons and guide them towards the correct placement on the cortical plate. At 27 weeks, the temporal lobe is distinctly visible, and axons from the auditory system increase in the marginal layer (Moore & Linthicum, 2007:466). At 4 months gestation age, the neurofilament-containing axons penetrate layer 4, 5 and 6 of the auditory cortex in parallel arrays (Sininger et al., 1999:5). At the end of the third trimester, a clear separation between the primary and secondary auditory cortex is formed (Moore & Linthicum, 2007:466). At birth the auditory cortex is only half of the adult thickness, with an indistinct cytoarchitectural laminar pattern (Moore, Guan, & Shi, 1996, as cited in Sininger et al., 1999:5).

As the central auditory system develops, neurons are formed in excess of the number that the brain actually needs, and systematic pathways and neuronal connections are established as the neurons conduct electrical activity from the sensory organs towards the brain. This electrical activity stimulates the neurons to form long axons with multiple branches so that synaptic connections with thousands of other neurons can be formed (Northern & Downs, 2002:128). The electrical activity can alter the location, number and strength of the synaptic connections between the neurons (Pallas, 2005:5). Both intrinsic and extrinsic activity is needed for the development of the central auditory system. Intrinsic activity is independent of sensory input, because the neurons in the cortical pathways are active and spontaneous neuronal activity occurs. Sense organs drive the extrinsic cortical activity and the brain will reflect patterns of stimulation within a critical period (Pallas, 2005:5-6). The neurons that are not stimulated during this critical period will be discarded (Northern & Downs, 2002:129).

The foetus is able to detect sound in utero from 20 weeks gestation age and it has been shown that bone conduction is the primary pathway through which soundwaves in the surrounding amniotic fluid is conducted to the inner ear (Sohmer, Perez, Sichel, Priner, & Freeman, 2001:109). Sounds found in the uterus contain external sounds of 60 dB or louder in the close vicinity of the mother, including her own vocalisations (Lecanuet, Granier-Deferre, & Busnel, 1995:240). The response of the foetus depends on the frequency, type, intensity and duration of the sound (Lecanuet et al., 1995:99), and is evident in the form of a motor response, a cardiac accelerative change, or a change in behavioural state (Morokuma et al., 2008:47-48). The foetus start to respond to low sounds first, as high sounds are attenuated through the mode of transduction of sound (Hepper & Shahidullah, 1994, as cited in Lecanuet et al., 1995:100). The foetus may be able to discriminate between contrasting consonants, such as /l/ and /f/ during the last trimester, and the near-term foetus may perceive differences in voice characteristics of two speakers (Lecanuet et al., 1995:240, 256). The foetus may also respond to music (Lecanuet et al., 1995:103).

Prenatal hearing is of great importance in the development of the auditory system, as intra-uterine sounds are already transduced as electrical activity from the inner

ear towards the auditory cortex, forming and strengthening synapses along the way. In order for stimulus recognition (such as word recognition) to occur, the synaptic circuitry requires experience to mature (Pallas, 2005:7). In other words, in order for the infant to develop word recognition skills, full access and exposure to spoken language is necessary, even in utero. Thus, typically developing newborns are able to process sounds and analyse their loudness and pitch accurately, as well as discriminate between speech sounds, due to the early development of the auditory system in utero and simultaneous exposure to sound (Sininger et al., 1999:6).

3.2.2 Postnatal maturation of the auditory system

As the infant is born with a relatively mature peripheral auditory system, postnatal maturation is focused primarily on the central auditory system. Up until 6 months of age, final maturation of the olivocochlear system occurs. The olivocochlear neurons increase in size and the dendritic branches of the efferent neurons acquire an adult-like morphology (Moore & Linthicum, 2007:470). Between 6 to 12 months of age, two important cortical changes occur, namely a marked reduction in the marginal layer of the auditory cortex, and maturation of thalamic input. The intrinsic axons in the marginal layer disappears, as the cortical neurons are now completely mature, and the potential arises for the auditory cortex to be stimulated by a continuous flow of input from the core of the brainstem pathway (Moore & Linthicum, 2007:470). Simultaneously, the infant's response to speech sounds changes accordingly. Until 6 months, infants display good discrimination of all speech sounds, irrespective of the language in which they occur. From 6 to 12 months, infants begin to attend differently to native and non-native languages. Discrimination of pairs of speech sounds may improve, deteriorate, or remain the same, depending on the characteristics of the native language. Infants between the ages of 6 to 9 months, listen longer to monosyllables with a high probability of occurrence in the ambient native language, and words that have the same stress patterns as those in the native language. During the second half of the first year of life, infants start to attend to speech sounds as bearers of meaning (Moore & Linthicum, 2007:470).

Between the ages of 2 to 5 years, the cortical neurons enlarge and extend. There is a continual axonal maturation in the deep layers of the auditory cortex, reaching an

adult density by 5 years. Myelin also increases in density until 6 years of age. Final maturation of the auditory system occurs during 6 to 12 years of age. Axonal maturation occurs in the superficial layers of the auditory cortex, and at 11 to 12 years the density equals that of an adult. The neurons in layers 2 and 3 are interconnected vertically and horizontally to neurons within the same column of the auditory cortex and neurons in adjacent areas. These neurons are also interconnected with neurons from the auditory cortex in the opposite hemisphere. Maturation of the neurons in the superficial layers broadens the scope of intracortical interaction (Moore & Linthicum, 2007:471). This is apparent in the gains the child makes in the ability to discriminate speech in difficult listening situations. Perception of speech in noise improves markedly across late childhood. Children also demonstrate an improvement in the ability to discriminate masked and degraded speech (Moore & Linthicum, 2007:472).

3.3 THE NEUROPHYSIOLOGY OF THE AUDITORY SYSTEM AND WORD RECOGNITION

The development of the auditory system (pre- and postnatal) culminates in the maturation of the structures so that sounds (such as sounds from the complex speech signal) can be coded by the system in order for further processing to occur. Sound waves enter the outer ear and are transduced by the tympanic membrane and ossicles from acoustic vibrations to mechanical vibrations (Kramer, 2008:80). These mechanical vibrations are conducted via the footplate of the stapes towards the oval window of the cochlea, where it set the fluids of the cochlea in motion (Martin, 1997:291). This disturbance in the fluids causes the basilar membrane to move up and down, producing a back and forth movement of the stereocilia of the outer hair cells, which are connected to the underside of the tectorial membrane (Kramer, 2008:86). When the movement by the stereocilia of the outer hair cells are sufficient, the stereocilia of the inner hair cells will also bend back and forth. This movement of the inner hair cells causes an in and out flow of K^+ -ions which increases and decreases the intracellular potential (Kramer, 2008:88).

Two general theories have been developed to explain how the cochlea codes the different frequencies of the sound wave, namely the place theory and the frequency theory (Kramer, 2008:96). The place theory of hearing proposes that the frequency information is coded at the place on the basilar membrane where the peak of the travelling sound wave occurs. The hair cells are orderly arranged along the basilar membrane, with the hair cells that respond to the low frequencies (starting at about 20 Hz) near the apical end of the cochlea, and the high frequencies from 2000 up to 20 000 Hz in the bottom half of the cochlea (Martin, 1997:293). This tonotopic arrangement is preserved throughout the auditory pathways, ranging from the cochlea to the auditory cortex (Kramer, 2008:96). The frequency theory assumes that the hair cells will transmit an impulse that is similar to its input, for example if the tone is a 100 Hz tone, then the neurons would fire 100 times (Martin, 1997:293). The place theory explains pitch discrimination well, but have some limitations in explaining why pitch discrimination becomes poor at the auditory threshold (Martin, 1997:290), whereas the frequency theory fails to describe frequency coding for mid and high frequencies, as the auditory nerve can only fire up to 400 times per second (Kramer, 2008:99; Martin, 1997:291). Thus, although there is no consensus at the moment regarding frequency coding, it is known that hair cells along the basilar membrane respond to specific frequencies, and these frequencies are coded and transduced by the auditory pathways to their specific area in the auditory cortex.

During the development and maturation of the auditory system, frequency-specific neural information is coded and conducted along the nerve fibres of the auditory nerve towards the auditory cortex. At the auditory cortex, the place-frequency tonotopic arrangement of the basilar membrane is preserved, and the frequency-specific neural impulses stimulate designated areas for that specific frequency region in the auditory cortex (Raphael et al., 2007:210-211). This phenomenon of the brain to re-organise itself based on the input it receives is called neuroplasticity. As the maturing auditory cortex is exposed to speech and all the different acoustic features, the frequency-specific electrical impulses carrying the neural information to the cortex causes systematic pathways to be established through coordinated routes that are used repeatedly (Northern & Downs, 2002:128). Once the neurons reach their target in the designated area of the cortex, connections between these neurons form in order to create physical “maps” of the acoustic features so that learning can

take place (Northern & Downs, 2002:129). This has been demonstrated in the auditory pathways of a rat (Illing, 2004:9), where it was found that after two hours of exposure to sound that the rat has not heard before, the neurons conducting these impulses to the brain started to respond to this new sound by changing their gene expression. This argues towards the notion that a sensitive period exists during which plasticity of the auditory system appears to be high. The period during which the human auditory system remains maximally plastic has been established to be up until 3.5 years of age (Sharma, Dorman, & Spahr, 2002:538), although the system may remain plastic until 7 years of age in some children. During this period, the brain is able to assimilate and master new information rapidly, accounting for the high-speed development of word recognition as part of speech perception during this time (Northern & Downs, 2002:131).

Research in the domain of speech perception commenced in the 1950s. The structure for studying the underlying active mechanisms involved with speech perception was influenced by issues that were under investigation in the field of language development. Taxonomic linguistics proposed that language is a hierarchy organised in a number of distinctive levels, and an accurate description of language requires a description of each level, independent of the higher levels. In order to describe language, an account of four levels of language should be presented:

- Phonetic level: how acoustic properties map into phonetic segments
- Phonemic level: how phonetic segments map into particular phonemes
- Morphemic level: how these phonemes are combined in order to form morphemes
- Syntactic level: how the morphemes are constructed in order to form a sentence

Thus, in order to provide a structural analysis of speech perception, research was focused on the apparent basic level of speech perception, namely, how the acoustic signal arriving at the ear is transformed into phonetic segments (Jusczyk & Luce, 2002:2). It was found that a great amount of variability exists in the acoustic signal for the same speech sound. It was found that a single set of acoustic features that

identifies a phonetic segment across all contexts does not exist, and that the acoustic features of a segment is greatly influenced by the surrounding speech sounds (Delattre, Liberman, & Cooper, 1955, as cited in Jusczyk & Luce, 2002:3). In addition to the variability of acoustic features of phonetic segments in the proximity of other phonetic segments, variability in the acoustic features also exists between different speakers, and within utterances produced by the same speaker. This extra variability depends on the speaker's gender, state of articulators, vocal folds and speaking rate (Jusczyk & Luce, 2002:3). Liberman et al. (1967, as cited in Jusczyk & Luce, 2002:4), found that, due to co-articulation, the beginning portions of the phonetic segment already carries information about the speech sound following the phonetic segment, and thus no consensus could be reached in the determination of the basic perceptual unit.

A structural analysis of speech perception failed to provide an explanation of how listeners perceive fluent speech, and attention was focused on spoken word recognition (Jusczyk & Luce, 2002:12). Four major models of the word recognition process have been developed, and these models all share the common assumption that the recognition of words involves two components, namely, activation and competition. This assumption underlines the phenomenon that there is a competition among multiple representations of words that are activated in the memory (Luce et al., 2000:615). These major models will be presented in the subsequent discussion:

3.3.1 The Cohort model

According to the Cohort model of spoken word recognition, when a word is heard, a set of possible word candidates are activated in memory that are similar in their initial sound sequence (the "word-initial" cohort) (Tyler, Marslen-Wilson, Rentoul, & Hanney, 1988:368). Once these words are activated, the possibilities are narrowed down with a simultaneous bottom-up (acoustic-phonetic) and top-down (syntactic-semantic) process, until a single candidate remains, and word recognition follows (Marslen-Wilson & Welsh, 1978, as cited in Jusczyk & Luce, 2002:13; Tyler et al., 1988:368). In order for the word to be recognised, a corresponding representation of the full acoustic-phonetic form of the word must be present in the mental lexicon. This is called the full listing hypothesis, and implies that access to the word-initial

cohort can only be gained if there is a full listing of the lexical forms of the word (Tyler et al., 1988:369). In contrast with the full-listing hypothesis, a decomposition hypothesis has also been proposed. According to this hypothesis, possible word candidates in the word cohort can also be activated on the basis of a shared sublexical unit, such as the word's stem. The speech input has to be broken down into the sublexical units before the word cohort can be activated (Tyler et al., 1988:369). The recognition system tracks the input closely, and any minimally discrepant features in the acoustic-phonetic information are sufficient to eliminate a word candidate as a match for the input word (Jusczyk & Luce, 2002:13). The level of activation of a possible word in the word cohort is not affected by the other words, and the effect that a competitor word has on the other words is derived merely from its presence in the cohort as a candidate for recognition (Jusczyk & Luce, 2002:13). A shortcoming of this model is that it preserves the notion that lexical competition occurs without lateral inhibition, and this was addressed in the development of the TRACE model (Jusczyk & Luce, 2002:13).

3.3.2 The TRACE model

The TRACE model has been developed by McClelland and Elman (1986). This model proposes that word recognition occurs in three levels, which correspond to the primitive processing units, namely features, phonemes and words (McClelland & Elman, 1986:8). At the feature level, a number of feature detectors are present. Similarly, in the phoneme and words level, detectors for the different phonemes and words are also present. Excitatory connections exist between levels, and inhibitory connections among levels. Features such as voiced/voiceless, manner of articulation *et cetera* that are present in the input-word are activated at the feature level, which in turn will cause all the phonemes containing those features in the phoneme level to be activated. These activated phonemes will cause all the words in the word level containing the activated phonemes to be activated. Those units that are only momentarily consistent with the input-word, will be inhibited by the lateral inhibition that exists among units within a level (Jusczyk & Luce, 2002:13), thus addressing the shortcoming of the Cohort model. However, the architecture of this model consists of nodes and connections that are probably psychologically implausible when dealing with the temporal aspects of spoken word recognition (Jusczyk & Luce, 2002:14).

3.3.3 The Shortlist model

The Shortlist model has been developed by Norris and colleagues (1994), and is similar to the TRACE model in that it is also a bottom-up model. The candidate words are programmed into a lexical competition network (similar to the word level of the TRACE model), but this model accounts for the speed with which word recognition occurs, by simplifying the process by which the candidate words are programmed into the lexical network, and by selecting the target word from a much smaller word pool (Norris, 1994:202). The model proposes that word recognition occurs in two stages. During the first stage, a shortlist of words is generated that consists of the same lexical items as the target word, based on bottom-up evidence. These words enter the lexical network during stage two, and overlapping words inhibit each other in proportion to the phonemes with which they overlap (Norris, 1994:202). Conventional programming techniques are used to wire the lexical network, and no more than 30 words are generated as possible candidates for recognition (Norris, 1994:202). This is a purely bottom-up approach, and does not account for the top-down lexical influences on word recognition, but remains an attractive alternative to the TRACE model (Jusczyk & Luce, 2002:14).

3.3.4 The Neighbourhood Activation Model (NAM), and Paradigmatic and Syntactic model (PARSYN)

The NAM constitutes an approach in which the stimulus input activates a set of similar sounding acoustic-phonetic patterns in memory (Luce & Pisoni, 1998). The activation level depends on the similarity of the pattern, and the activation level is higher for patterns with greater similarity. Word decision units which are tuned to specific patterns are responsible for deciding which pattern best matches the input. The probability of each pattern for matching the input is computed based on the frequency of the word to which the pattern corresponds, the activation level of the pattern, as well as the activation levels and frequencies of all the other words activated in the system (Luce & Pisoni, 1998). The acoustic-phonetic pattern with the highest probability of matching the input word is considered the target word. The neighbourhood density of the acoustic-phonetic patterns for the input word influences the speed with which processing can occur. Words that share fewer

acoustic-phonetic patterns with other words will be processed more quickly than words that share many acoustic-phonetic patterns (Luce & Pisoni, 1998).

The NAM was reviewed and adjusted to account more accurately for the spoken word recognition effects as a neighbourhood density function, and the PARSYN model was developed (Vitevich, Luce, Pisoni, & Auer 1999:310). This model also consists of three levels, namely an input level (the allophone input level), a pattern level (the allophone pattern level), and a word level. Connections exist between the units within a level, and these are mutually inhibitory, with one exception: the links among the allophone units in the pattern level are facilitative across the temporal positions. The connections between the levels are facilitative, also with one exception: once a single word has gained an advantage over the others, the word level sends inhibitory information back to the pattern level, thus quelling activation in the system (Luce et al., 2000:620). Another important feature of this model proposes that allophones that occur more frequently than others have a higher resting level of activation, and allophones that frequently occur together will excite one another through facilitative links (Jusczyk & Luce, 2002:15). Although PARSYN accounts for simultaneous bottom-up and top-down processing of word recognition, it fails to account for the processing of the rich information embedded in the speech signal (Jusczyk & Luce, 2002:16).

It is evident from the above discussion that the development of word recognition skills already starts in utero, and that prenatal hearing results in the establishment of neural pathways which are crucial for the processing of speech sounds. Thus, the presence of a hearing impairment may significantly affect the development of word recognition skills, and will be discussed in the following section.

3.4 THE EFFECT OF DEPRIVATION ON WORD RECOGNITION

As audition is the first step in the recognition of spoken words, an accurate representation of the words must be conducted to the auditory cortex for processing of the speech signal and organisation of the cortical neurons (Raphael et al., 2007:207). This is compromised considerably when the cochlea is unable to code the frequency information due to the presence of a sensorineural hearing loss, and

thus the subsequent neural information that is conducted to the auditory areas lacks important information, resulting in an altered organization of the auditory areas due to deprivation (Sininger et al., 1999:7).

“Auditory deprivation” refers to a deviation of auditory input to the auditory cortex that is different from the expected or needed input for optimal development of auditory function (Gravel & Ruben, 1996:86). Auditory deprivation can cause extensive degeneration in the auditory system (Sharma et al., 2002:532). The evidence of this degenerative effect can be found in the peripheral as well as central auditory structures. The spiral ganglion in the cochlea, anteroventral cochlear nucleus and ventral cochlear nucleus may suffer from loss of cell density, and alterations in neural projections between brainstem nuclei may be seen (Nordeen, Killackey, & Kitzes, 1983, as cited in Sharma et al., 2002:532). The generation of significant activity in the infragranular layers by the auditory cortex may be absent, activation of the supragranular layers may be temporally delayed, and the amplitude of synaptic currents may be significantly smaller (Kral, Hartmann, Tillein, Heid, & Klinke, 2000:723). In the absence of auditory stimulation of the auditory cortex, input from other sensory modalities (such as vision) may start to stimulate the auditory cortex, and may occupy those areas designated for auditory processing (Finney, Fine, & Dobkins, 2001:1173). Total absence of auditory stimuli (as it is the case in profound hearing loss), will result in the most severe form of auditory deprivation. The effect of auditory deprivation is also much worse if the deprivation occurred during the sensitive period for development, compared to deprivation at a later postlingual age (Sininger et al., 1999:6).

Children with congenital moderate to severe sensorineural hearing loss (MSSHL) may not experience the effect of total deprivation, as some of the speech sounds may still be audible even without amplification (Gravel & Ruben, 1996:101). Figure 1 shows the audibility of different speech sounds plotted as intensity and fundamental frequency at a normal conversation level in the presence of MSSHL (adapted from Northern & Downs, 2002:18; Harrell, 2002:82):

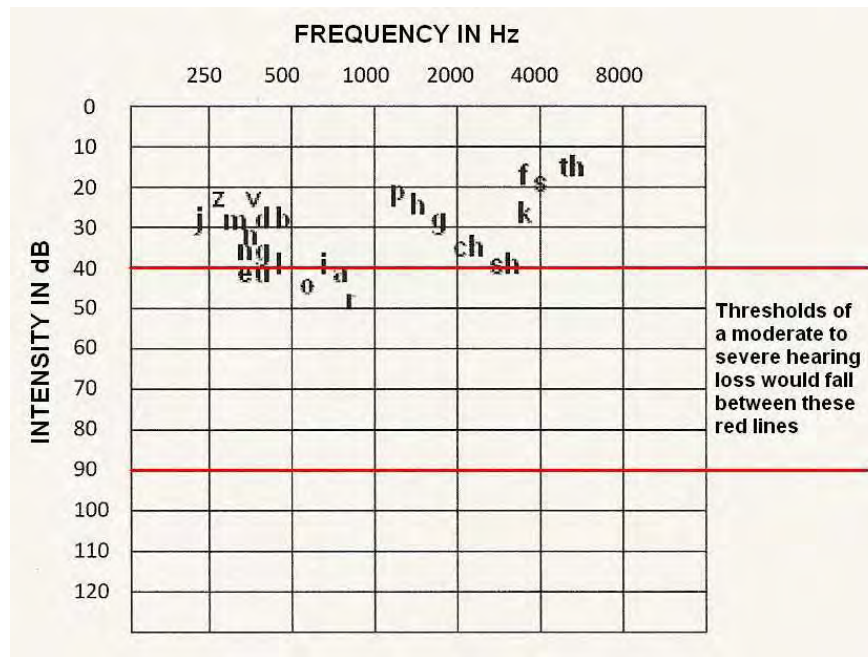


Figure 1: Audibility of the different speech sounds in the presence of MSSHL

(adapted from Northern & Downs, 2002:18; Harrell, 2002:82)

It can be seen from Figure 1 that some of the low frequency speech sounds may be audible if the child has thresholds at 250 to 1000 Hz of about 40 dB. Therefore, some of the acoustic information in the speech signal can be coded into frequency-specific neural information, and stimulate the corresponding areas in the auditory cortex to form connections between the neurons. This has been demonstrated by Harrison, Nagasawa, Smith, Stanton, & Mount (1991), where a high frequency hearing loss induced at birth in cats, resulted in altered sensory representation in the auditory cortex in the high frequencies only, and the low frequency areas of the auditory cortex remain similar to that of a normal hearing cat (Harrison et al., 1991:14). A study by Tibussek and colleagues (2002) demonstrated that children with MSSHL may present with some synchronisation in the firing of neurons up to the brainstem level, as these children demonstrated detectable waves I-III-V when the auditory brainstem response was measured, but the latencies were prolonged (Tibussek, Meiste, Walger, Foerst, & Von Wedel, 2002:128). These occurrences may account for the outcomes listed in Chapter 2 for children with MSSHL when left unaided during the sensitive period, but also substantiate the statement that, due to the amount of residual hearing, these children can be successfully integrated into mainstream settings provided that they receive appropriate intervention in a timely

manner. The routine assessment of the speech perception capacity of a child with hearing loss may provide valuable insight regarding the prognosis of the development of speech, language, reading and cognitive skills (Iler Kirk, Diefendorf, Pisoni, & Robbins, 1997:101). It may also aid in the delineation of habilitation choices, including amplification and education, and has been found to be especially helpful in the comparison of outcomes of different sensory aids and/or processing algorithms (Eisenberg, Johnson, & Martinez, 2005). The following section will describe issues related to the assessment of word recognition skills in children as part of a full battery of paediatric speech perception assessments.

3.5 ASSESSMENT OF WORD RECOGNITION SKILLS IN CHILDREN

Erber (1982, as cited in Thibodeau, 2000:282) proposed that speech perception assessments should reflect the following four elements, namely speech awareness/detection, speech discrimination, speech identification or recognition, and speech comprehension. Speech awareness tasks would require only an indication that a sound was heard, such as the establishment of a speech detection or speech reception threshold. Speech discrimination tasks would require the detection of a change in stimulus. Speech identification or recognition tests would require the young child to attach a label to the stimulus-word by pointing to a picture or repeating the word verbally. Speech comprehension would entail the child to attach a meaning to a stimulus-word by answering questions verbally, or by pointing to a picture that conveys the meaning of the word (Thibodeau, 2000:282). Currently, no true tests of speech discrimination or speech comprehension is used in day-to-day speech audiometry, but age appropriate test materials for the determination of speech awareness thresholds and speech identification/recognition scores have been well-developed (Thibodeau, 2000:282).

The sub-classification of speech identification/recognition assessments is dependent on the stimulus-type used for administration of the test (Brewer & Resnick, 1983:205). Monosyllabic words are usually used for the determination of a word recognition score, and sentences are likewise used for sentence recognition tests.

Three sets of variables should be considered when selecting test material for the determination of a word recognition score. Internal variables, including the size of the child's vocabulary and his/her language competency, chronological age, cognitive abilities, state of alertness, and attentiveness during the testing could influence the test outcomes (Eisenberg et al., 2005). External variables, such as the selection of an appropriate response task, the utilisation of reinforcement and the memory load that is required in order to perform the task, need to be considered. The consideration of methodological variables also is of great importance, and includes audio cassette versus monitored-live-voice presentations, open- versus closed-set tests, and unrestricted and restricted task domains in closed-set construction (Iler Kirk et al., 1997:103).

The mode of presentation of the stimuli during the determination of a word recognition score can be either via a pre-recorded version of the word lists, or by monitored-live-voice. A recording of the word lists can eliminate differences in consistency across one listener to the next and repeated assessment of the same listener, although it does not rule out differences in recordings of different speakers. Monitored-live-voice provides the clinician with more flexibility, especially when it comes to testing young children, and good test-retest reliability has been mentioned (Iler Kirk et al., 1997:103).

When an open-set word recognition test is used, the listener is not provided with a possible set of responses. The listener must process the word and compare it with other words that are stored in the auditory lexical memory. Therefore, the listener's response is dependent on the familiarity of the stimulus-word (Brewer & Resnick, 1983:205). These kinds of tests are not appropriate for all children, as some children's articulation may be so poor that the examiner may be unable to make a judgement on the correct recognition of the word (Iler Kirk et al., 1997:207). For this reason, several closed-set tests have been developed. Closed-set word recognition tests require that, upon hearing the stimulus word, the child must choose from a set of possible alternatives (usually pictures) that best match the stimulus-word. A drawback of this kind of test is that it elevates the "guessing floor," thus increasing the word recognition score that does not accurately reflect the child's true word recognition score (Brewer & Resnick, 1983:206). The guessing floor may be lowered

by introducing more possibilities from which to choose (Iler Kirk et al., 1997:104). The task domains for closed-set word recognition tests differ in their restriction of the target signals. An unrestricted task domain has the target signals embedded in range of items that represent phonemic confusions, such as the stimulus-word “socks,” embedded in a page alongside pictures of “box,” “fox,” and “blocks.” This prevents the child to guess the word in a process of elimination (Iler Kirk et al., 1997:104), and truly reflects the child’s sensory capabilities, as no top-down processing is required for responding to the word. A restricted task domain closed-set test specifies beforehand what the listener would expect to hear, and the target item would stand alone. These expectations trigger a top-down approach to processing, as the context of the sensory event may provide clues to the target word. This type of test may give an indication of the conceptual processing of the signal instead of a reflection on the sensory capabilities (Iler Kirk et al., 1997:105).

3.5.1 Paediatric open-set word recognition assessments

The earliest development of word recognition test material for children utilised the open-set paradigm. Haskins (1949, as cited in Brewer & Resnick, 1983:213), developed the first phonetically-balanced monosyllabic word lists for children, the PB kindergarten lists. This test (PBK-50) consists of three lists of 50 words each, and the words are taken from kindergarten vocabulary lists. The child is required to repeat the word after it is heard, and due to this response format and the fact that some of the words may not be familiar to children, it is not recommended for use when testing children 5½ years or younger (Brewer & Resnick, 1983:214). The generation of the Manchester Junior (MJ) lists followed ten years later by Watson, (1957, as cited in Jerger, 1984:71). The words from these lists are considered to be age-appropriate for children over 6 years of age, and are phonetically balanced (Iler Kirk et al., 1997:106). Boothroyd (1967, as cited in Brewer & Resnick, 1983), compiled 15 lists of ten monosyllabic words each, called the ABL test. This test required a shorter testing time, and is scored according to the number of phonemes correctly recognised, rather than the number of words (Brewer & Resnick, 1983:208).

As these tests rely on the verbal response of a child, a language disorder, vocabulary deficit, an articulation disorder or lack of motivation will render the interpretation of responses unreliable (Diefendorf, 1983:247). Therefore, alternative tests were developed that utilise a closed-set paradigm, with the aid of pictures.

3.5.2 Paediatric closed-set word recognition assessments

Siegenthaler (1966, as cited in Diefendorf, 1983:247), urged that the response formats and test items of the word recognition tests should fit a child's interest and ability (Jerger, 1984:71). The Discrimination by Identification of Pictures (DIP) word recognition test was consequently developed, and consists of 48 pairs of monosyllabic words that differ in distinctive features (Brewer & Resnick, 1983:214). These word pairs are depicted on picture cards, and only one word-pair is depicted on a card. The child is asked to point to the stimulus word. A major disadvantage of this test is that only two pictures are displayed per card, so the child has a 50% chance to guess the stimulus word, and thus the word recognition score may reflect inaccurate word recognition ability.

This disadvantage was addressed in the development of another closed-set word recognition test within an unrestricted task domain. The Word Intelligibility by Picture Identification (WIPI) test was developed by Ross and Lerman in 1970. This test consists of four test lists of 25 words each, and also includes picture plates with six pictures per plate. Several of the pictures depict words that rhyme and have similar acoustic/phonetic similarities, and the other pictures are added as foils. Upon hearing the stimulus word, the child is required to choose from six different pictures and point to the picture of the target word. Embedding the target word in six different pictures increases the difficulty of the test, as well as the validity of the test results, as it becomes more difficult to correctly guess the target word (Ross & Lerman, 1970:48-49). It is appropriate for hearing-impaired children aged 5 to 8 years, and was standardised on 61 hearing-impaired children. Ross and Lerman also concluded that the word recognition score obtained with the WIPI should exceed open-set word recognition score by 25%, and has good test-retest reliability (McLauchlin, 1980:269). Norms for this test have been established by Sanderson-Leepa and Rintelmann (1976, as cited in Thibodeau, 2000:303), and Papso and Blood

(1989:236). The test can be administered relatively fast, is not as limited by differences in speech and language abilities among children, and is noted to be an interesting task for children (McLauchlin, 1980:270). Due to the effect that cultural differences may have on the test results, some of the words and pictures of the test were adapted to use with the South African population (Muller, personal communication, 2008).

The Northwestern University-Children's Perception of Speech (NU-CHIPS) test was developed by Elliott and Katz (1980, as cited in Iler Kirk et al., 1997:108) as a word recognition test for use with children as young as 3 to 5 years of age (Thibodeau, 2000:302). This test is also in the unrestricted task domain, and is similar to the WIPI in the sense that it is also in a picture-pointing response format. The test consists of 50 monosyllabic words as test items, and four pictures appear on each page of the picture book. Foils that are phonetically similar also appear in the test. The test demonstrates construct validity, but older children tend to score better on the NU-CHIPS than on the WIPI, mainly because of the somewhat more difficult vocabulary of the WIPI (Iler Kirk et al., 1997:108).

In the restricted task-domain of closed-set word recognition tests, two tests were developed in 1980. The Auditory Numbers Test (ANT) was developed by Erber (1980, as cited by Iler Kirk et al., 1997:109), and is a spectral/pattern perception test for use with children who exhibit severe speech discrimination difficulties. Five coloured picture cards with groups of one to five ants are used with the corresponding numerals, and the child scores a point each time the correct number of ants is pointed out. Thus, children should be able to comprehend the words representing the numbers of 1 through to 5, and valuable information is gained regarding the use of minimal spectral cues and gross intensity patterns (Iler Kirk et al., 1997:110).

The Paediatric Speech Intelligibility (PSI) test was also developed in 1980, by Jerger, Lewis, Hawkins and Jerger (as cited in Diefendorf, 1983:248). The effect of receptive language on the test results was extensively researched in order to prevent the contamination of results (Diefendorf, 1983:248), and the test can be used for the evaluation of children from 3 to 6 years of age (Thibodeau, 2000:302). The test items

consist of both words and sentences. Twenty words and ten sentences were selected for this test, and are adjustable to account for differences in receptive language skills (Diefendorf, 1983:249). Four plates with five pictures each are used, and a picture-pointing response is requested from the child (Iler Kirk et al., 1997:110).

The determination of a word recognition score (regardless of the procedure) should contribute towards two objectives when assessing children, specifically the quantification of the effect that hearing loss has on the word recognition skill of a child, as well as the facilitation of hearing aid fittings (Thibodeau, 2000:304). Thus, a word recognition score that is obtained with hearing aids *in situ*, would yield valuable information regarding the amount of acoustic cues that is available to a child with hearing impairment, as well as areas of acoustic information that the child is missing. All aspects related to the selection of assessment material should be taken into account, so that the test is maximally dependent on the sensory capacity, and difference in performance should reflect changes in sensory capacity, and not linguistic or cognitive status (Hnath-Chisolm, Laipply, & Boothroyd, 1998:94).

3.6 CONCLUSION

When determining the outcomes of children with hearing impairment, an in-depth understanding of the development of the auditory system is imperative. The physiology of the auditory system and how this relates to the recognition of spoken words also provides valuable insight when interpreting word recognition scores of children with hearing impairment. The effect of deprivation on the auditory system is devastating and should be taken into account when designing early intervention programmes and when providing amplification. The assessment of word recognition skills in children forms an integral part in the validation of paediatric hearing aid fittings, and the unique needs of children should be taken into account when selecting appropriate assessment tasks.

CHAPTER 4

LINEAR FREQUENCY TRANSPOSITION TECHNOLOGY AND CHILDREN: AN EVIDENCE-BASED PERSPECTIVE



CHAPTER AIM: To provide a critical description of paediatric hearing aid technology issues and the development of appropriate signal processing for children.

“No matter how logically appealing a new treatment may seem to be, it cannot be assumed to perform as planned until there is specific effectiveness data that verifies this.”

~ Robyn Cox (Cox, 2005:421)

4.1 INTRODUCTION

The developing speech perception abilities of children require special amplification considerations (Dillon, 2000:412). Children have unique needs compared to adults, such as the need for higher sound pressure levels and audibility of speech sounds (regardless of the listening situation) in order to perform the same as adults on word recognition tasks (Stelmachowicz et al., 2000:911). Furthermore, children’s audiologic configurations may vary considerably across individuals and may display larger asymmetries than those of adults, and the physical size of the ear canal may be much smaller and may change more as the child grows up (Palmer, 2005:11). Thus, paediatric hearing aid fitting procedures should be designed to incorporate objective, valid, and reliable methods to measure audibility of speech across a variety of listening environments over time (Scollie & Seewald, 2002:689). These fitting procedures should follow a step-wise approach, which include assessment of ear- and frequency-specific hearing thresholds, selection of appropriate amplification devices, objective verification of the output of those devices, validation of aided auditory function, as well as informational counselling and follow-up (Paediatric Working Group, 1996, as cited in Scollie & Seewald, 2002:689).

Cost and electroacoustic flexibility are two major factors to consider when selecting appropriate amplification devices for children (Beauchaine, 2002:48). This is particularly relevant within the South African context, where hearing aids are not

funded by the government for children over six years of age, and parents are dependent on private medical aids which rarely cover the full amount of a hearing aid. For the paediatric audiologist working in an ever-changing technological environment, the need clearly exists for evidence of “current best practice” guidelines regarding the amplification choices for children with hearing loss. Evidence-based practice (EBP) guidelines are derived from clinical outcomes research that is both well-designed and client-centred (Cox, 2004:10), and is defined as “the conscientious, explicit, and judicious use of current best evidence in making decisions about the care of individual patients” (Sackett, Rosenberg, Gray, Haynes, & Richardson, 1996:71). Information on the normal developmental and functional aspects of the auditory system, as well as the effect of deprivation and subsequent intervention on the auditory system should be gathered from sources that produce excellent clinical evidence, so that informed clinical decisions can be made regarding amplification and habilitation strategies (Northern & Downs, 2002:33).

Outcomes research is guided by three concepts: efficacy, effectiveness and efficiency. Efficacy refers to the outcomes of a measured intervention as it presents under ideal circumstances (“Can it work?”). Effectiveness is the outcome of this measured intervention in usual daily circumstances (“Does it work in practice?”) and efficiency measures the effect of the intervention in comparison to the resources that it demands (“Is it worth it?”). Historically, research has contributed considerably towards the “efficacy” information-pool, and more studies are needed that evaluate the effectiveness and efficiency of treatment/intervention (Haynes, 1999:652).

The practitioner must be cautious in the use of evidence for clinical decision-making (Palmer & Grimes, 2005:506). The evidence obtained from clinical research can be graded into different levels of acceptance based on the research design, by acknowledging that certain research designs are more susceptible to error and bias (Cox, 2004:16). These research designs produce evidence ranging from most convincing (Level 1) evidence to least compelling evidence (Level 6) and are presented in Figure 1:

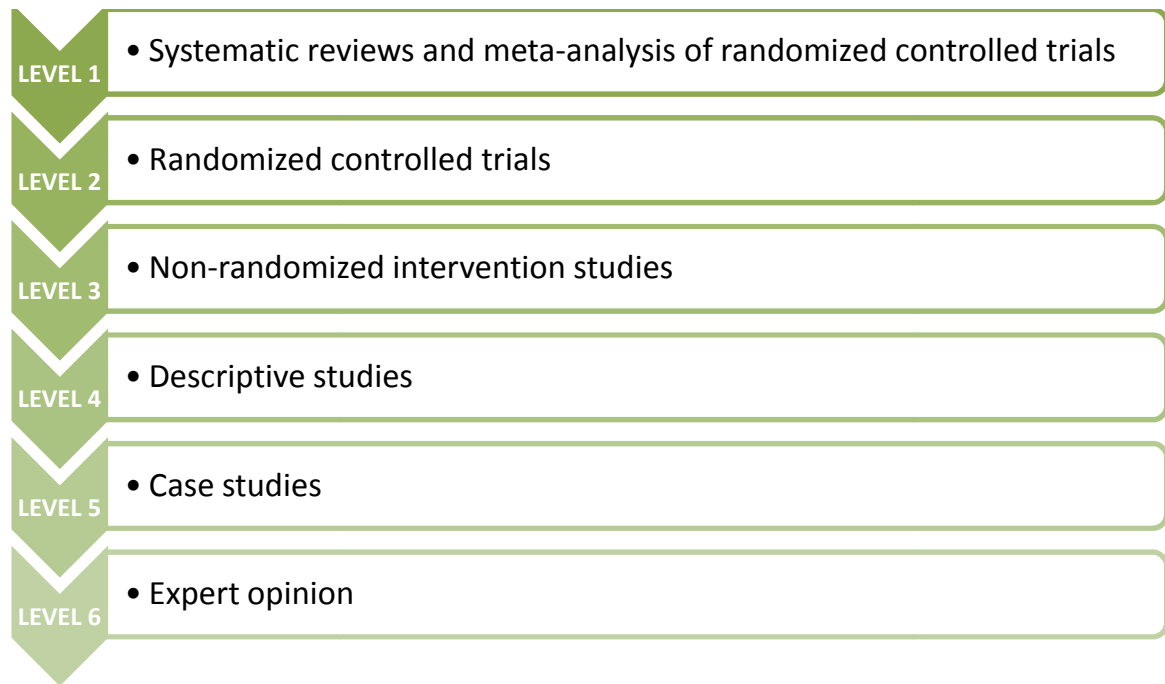


Figure 1: Levels of evidence produced by clinical research

The level of confidence that certain evidence instils in the decision-making process is awarded a grade of recommendation. These grades are presented in Figure 2:

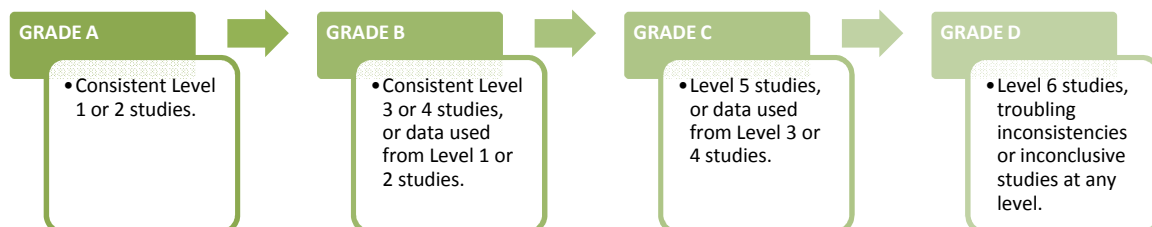


Figure 2: Grades of recommendation

These levels of evidence will be used to grade the relevant literature in this chapter. When assessing the appropriateness of amplification strategies for children, it is important not to focus on analogue versus digital signal processing alone, but to evaluate which processing scheme is appropriate by reviewing studies that have appropriately studied these schemes (Palmer & Grimes, 2005:506). However, due to the overwhelming evidence that digital signal processing provides the paediatric audiologist with the best electroacoustic flexibility, essentially all hearing aids employ this type of signal processing at present (Mueller & Johnson, 2008:291). Therefore, the paediatric audiologist is more than likely to choose from a range of digital hearing

aids when selecting amplification for the child with moderate to severe sensorineural hearing loss (MSSHL) than other types of circuitry.

4.2 CONVENTIONAL ADVANCED DIGITAL SIGNAL PROCESSING SCHEMES AND CHILDREN

The most basic function of amplification by means of hearing aids is to amplify sound signals to a level that is audible to the listener, so that the residual hearing can be used optimally (Staab, 2002:631). This is especially true when providing amplification to children, as the hearing aids should provide them with access to the full acoustic spectrum of speech in a variety of listening environments, so that the auditory cortex can receive maximal representation of the speech signal, and speech and language skills can develop age-appropriately (Hnath-Chisolm et al., 1998:94).

The development of hearing aid technology over the last 350 years incorporated the latest technology available at that specific time (Dillon, 2000:12), in order to amplify speech to optimal levels for the listener. The earliest amplification device was anything shaped like a trumpet, funnel or horn in the 1650s (Dillon, 2000:13). This was followed by the carbon hearing aid that emerged during the early-1900s (Staab, 2002:631), and subsequent development of the vacuum tube hearing aid in the early- to mid-1900s (Dillon, 2000:13). All these hearing aids were too big to be worn at ear-level, and had to be worn on the body. In the 1950s, the invention of the transistor by Bell Telephone Laboratories heralded the arrival of much smaller hearing aids that operated on a significant reduction in battery power (Staab, 2002:631). Analogue hearing aids were the first hearing aids to employ the transistor in the signal processing scheme, and this led to the development of flexible response shaping and multi-channel processing of sound, class D amplifiers to achieve less distortion in the output-signal, and the use of two microphones in one hearing aid, so that the listener could select directional or omni-directional sensitivity of the microphone (Dillon, 2000:16). However, digital electronics invaded hearing aid technology in 1980s, when the first wearable body-worn digital hearing aid was designed. The introduction of digital control circuits increased the flexibility and accuracy of adjustments made to the output-signal, but due to the style of the hearing aid, it was not a commercial success (Dillon, 2000:16). In 1996, the first

generation digital hearing aids were developed in behind-the-ear (BTE), in-the-ear (ITE) and in-the-canal (ITC) styles, making them much more cosmetically acceptable (Dillon, 2000:17).

The digital hearing aid technology evolved during the subsequent years into second and third generation digital hearing aids (as described in Chapter 1), and digital hearing aids currently consist of the following basic components (Mueller & Johnson, 2008:291):

- ✧ **Microphone:** to convert the acoustical signal into an electrically manipulated signal.
- ✧ **Amplifier:** enlarges the small electrical signal into a large electrical signal.
- ✧ **Digital conversion:** the electrical signal is converted to binary digits (bits), to manipulate the sound signal and apply different processing algorithms to the signal. In turn, the digital signal is reconverted to an electrical signal, and sent to the receiver.
- ✧ **Receiver:** converts the amplified and modified electrical signal into an acoustic output signal
- ✧ **Battery:** a power source to the hearing aid
- ✧ **Volume control:** to manually adjust the output signal of the hearing aid, although the volume control is de-activated where possible in paediatric hearing aid fittings.
- ✧ **Telecoil:** to convert electromagnetic information and deliver it to the amplifier, as it is used in some cases to improve the signal-to-noise ratio).

As discussed in Chapter 2, the guidelines provided by the Paediatric Amplification Guideline (Bentler et al., 2004:48-49), stipulate that the above-mentioned components should be able to provide audibility of the speech signal at low, mid and high intensity levels, without distortion, and without damaging the auditory system even further. In order to approximate the amplified speech signal as close as possible to the original signal across different listening environments, advanced features were added to the digital hearing aids, such as directional and adaptive directional microphones, automatic digital noise reduction, spectral speech enhancement, and an increased bandwidth of the frequency response of the hearing

aid (Bentler et al., 2004:49-50). Controversy surrounds the application of these advanced digital signal processing schemes in amplification choices for children, due to the lack of evidence that is available regarding efficacy, effectiveness and efficiency of hearing aids utilising these schemes for the paediatric population (Palmer & Grimes, 2005:506).

4.2.1 Directional microphone technology

The overall goal of directional microphone technology is to reduce the output of the hearing aid for sounds coming from the back and sides, without affecting the output for sounds that are coming from the front (Mueller & Johnson, 2008:293). Directional microphone technology has been developed in order to improve the signal-to-noise ratio (SNR) in a noisy environment. Fixed directional microphone arrays do not vary from moment to moment, and automatic/adaptive microphone arrays adapt in an environment so that noise coming from different directions is minimised (Dillon, 2000:188). A hearing aid with automatic directionality will automatically switch between omni- and directional microphone arrays, and a hearing aid with adaptive directionality will change the polar plot of sensitivity based on the noise-input's intensity, spectrum, and location (Mueller & Johnson, 2008:293). A study producing Level 5 evidence, reported that older children with MSSHL using a directional microphone system in their hearing aids presented with better closed-set speech recognition scores in noise than when they used an omni-directional system (Gravel, Fausel, Liskow, & Chobot, 1999). This may not reflect functional outcomes in open-set recognition or more reverberant environments. Functional outcomes of directional microphone technology suggest that this type of technology may benefit the child with MSSHL in some situations, provided that the child's head angle is optimally positioned for maximum benefit of the directionality (Ricketts & Galster, 2008:522). However, this may not be relevant for young children as they are not always positioned so that they can look at the speaker directly (Dillon, 2000:408). As with adults, directional technology may not be desirable in all situations, and may result in some control to be exerted by the older child or parent over the type of microphone array to use in a specific situation (Scollie & Seewald, 2002:696).

4.2.2 Digital noise reduction

Automatic digital noise reduction is also a strategy aimed at decreasing the effect of background noise on the speech signal. This can be accomplished by Wiener filtering or spectral subtraction. The gain in a Wiener filter is dependent on the SNR in each frequency band or channel, and will thus decrease the gain of that particular frequency if the SNR is poor. A spectral subtraction system subtracts the amplitude of the noise spectrum from the amplitude of the speech plus noise system, so that only the speech spectrum remains (Dillon, 2000:196). This subtraction-system and frequency-specific gain reduction may result in a decreased audibility of speech sounds in children using this type of technology. However, as was mentioned before, children require a better SNR for speech perception, and any attempt to improve the SNR may result in better speech perception in noise (Marcoux, Yathiraj, Cote, & Logan, 2006:711). This was demonstrated in a study by Gravel, Hanin, Lafargue, Chobot-Rodd, & Bat-Chava (2003), where they found that children with MSSHL may benefit from digital noise reduction, and this type of technology used with directional microphones provide a strong signal processing platform to increase speech perception in noise (Gravel et al., 2003:38). This study did not include a control group, and produced Level 5 evidence with a Grade C level of confidence.

4.2.3 Spectral speech enhancement

Spectral speech enhancement depends on the idea that the acoustic features of speech can be detected and exaggerated, in order to make the speech sound more recognisable. The spectral shape can be enhanced, as well as the duration of sounds, especially vowels (Dillon, 2000:206). The consonant/vowel intensity ratio (CVR) can also be increased, and this has led to increased consonant recognition in children with MSSHL, stops and fricatives alike (Smith & Levitt, 1999:418). However, this was only demonstrated in consonant recognition in syllables, not in words or continuous speech, and may thus change when the formant characteristics of the surrounding phonemic neighbourhood context change. This study also used case studies as a research design, and is classified as Level 5 evidence with a Grade C level of confidence.

4.2.4 Extended high frequency amplification

As mentioned in Chapter 1 and 3, the audibility of all speech sounds is crucial in the development of age-appropriate speech and language skills. The audibility of high frequency speech sounds seems particularly important, as these sounds are in addition to normal word recognition also important for the grammatical development of plurality, possessiveness and verb tense (Rudmin, 1981:263). The development of grammatical morphemes is usually delayed in children with moderate hearing loss, especially the development of the word-final /s/ (McGuckian & Henry, 2007:32). These authors suggest that the delay is due to an interaction between the fact that the word-final /s/ indicating possessiveness and plurality occurs less frequent than other grammatical morphemes, and that audibility is decreased for these high frequency speech sounds even though the child is appropriately fitted with hearing aids (McGuckian & Henry, 2007:31). This may be due to the reality that the peak energy of /s/ spoken by female and child talkers is approximately in the 6.3 – 8.8 kHz range (Stelmachowicz, Lewis, Choi, & Hoover, 2007:483), and that most modern hearing aids only provide amplification up to approximately 6000 Hz (Ricketts et al., 2008:160). Figure 3 depicts roughly the amount of speech energy that is “lost” when most modern hearing aids amplify speech:

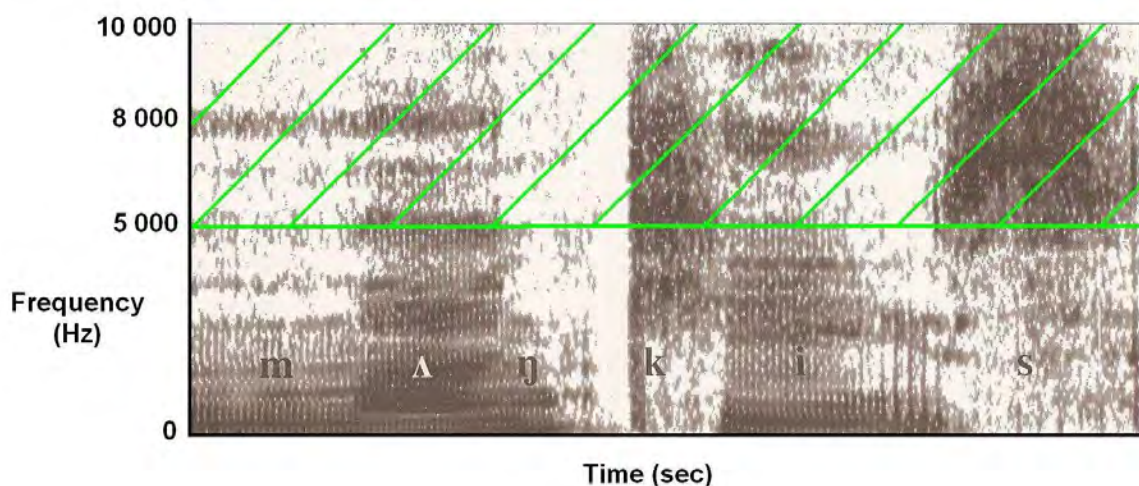


Figure 3: A spectrogram of the word “monkeys” as spoken by a female talker
 (adapted from Bernthal & Bankson, 1998:400)

The green shaded area in Figure 3 depicts the amount of speech energy that is lost when conventional hearing aids amplify speech. Thus, audibility and word recognition may be reduced.

The importance of high frequency audibility for children has been debated, but Kortekaas and Stelmachowicz (2000:657) conducted a study that produced Level 4 evidence and found that children with hearing impairment require a broader bandwidth to perform the same as adults with similar hearing loss on speech perception tasks. Ching and colleagues cautioned against the downwards spread of masking due to too much high frequency amplification as this may have a detrimental effect on perception of low frequency speech sounds, and that the amount of residual hearing should be taken into account when prescribing high frequency amplification gain for children (Ching et al., 2001:150). Recent studies producing high levels of evidence however, have all demonstrated the value of providing as much high frequency information as possible for children with MSSHL (Stelmachowicz et al., 2007:493; Pittman, 2008), although this should be done without the risk of feedback, which may limit the practical application of hearing aids that utilise an increased bandwidth of frequency response (Horwitz, Ahlstrom, & Dubno, 2008:799).

4.3 FREQUENCY LOWERING TECHNOLOGY

The limitations of conventional advanced digital processing schemes in providing adequate high frequency information to the listener, has lead to the development of alternative strategies in order to provide the listener with hearing loss with high frequency speech cues to facilitate better word recognition (Simpson et al., 2005:281). Three current frequency lowering signal processing strategies have been mentioned in Chapter 1, namely proportional frequency compression, non-linear frequency compression and linear frequency transposition. However, investigations so far produced mixed results regarding its efficacy, mainly due to a heterogeneous participant population, different processing schemes and test material through which efficacy could be established (Madell, 1998:116).

4.3.1 Terminology issues

Despite the publication of excellent reviews of frequency lowering technologies, some of the terminology for the different processing schemes is often used interchangeably. A review by Erber (1971) used the term “frequency-shifting amplification systems” as the generic term for all devices that utilise frequency lowering (Erber, 1971:530). As can be seen in Table 1, Braida, Durlach, Lippman, Hicks, Rabinowitz and Reed (1978) referred to “frequency lowering” as the generic term for all these type of strategies, and “frequency transposition” and “frequency shifting” as subdivisions of frequency lowering schemes (Braida et al., 1978:102). Gravel and Chute (1996:253), used “frequency transposition” as the generic term for all types of frequency lowering technologies. Furthermore, proportional frequency compression incorporates both transposition and compression strategies, and are referred to by some authors as a frequency transposition device, and by some as a frequency compression device (Ross, 2005).

In order to provide some guidance, all these frequency lowering techniques can be roughly classified by asking three questions (McDermott & Glista, 2007):

1. Does the lowered frequencies overlap with unaltered lower frequencies (*“transposition”*) or is the bandwidth of the frequency spectrum reduced (*“compression”*)?
2. Is the whole frequency spectrum lowered (*“linear”* or *“proportional”*) or only a portion of the spectrum (*“non-linear”* or *“non-proportional”*)?
3. Is the frequency lowering active the whole time (*unconditional lowering*) or only in selected conditions (*conditional lowering*)?

For the purpose of this study, the following terminology will be used according to these questions: **proportional frequency compression** refers to the reduction of the frequency bandwidth by lowering all the spectral components of the signal by a constant factor. **Non-linear frequency compression** reduces the bandwidth by lowering only the high frequencies in increasing degrees, and **linear frequency transposition** to the lowering of only the high frequency spectrum.

4.3.2 Early frequency lowering strategies and their implementation in hearing aids

A review by Braida et al. (1978) provides an overview of early frequency lowering techniques. These are summarised in Table 1:

Table 1: Early frequency lowering circuitries (compiled from Braida et al., 1978)

CIRCUITRY	SIGNAL PROCESSING
Transposition	Only high frequency components of the frequency spectrum are shifted downwards by a fixed displacement.
Zero-crossing-rate division	Speech is filtered into four passbands, and the resulting signal is processed to achieve a frequency spectrum band reduction by a factor of two.
Vocoding	Bandwidth reduction is achieved by filtering speech through a low-pass filter. The vocoder transmits a signal that is descriptive of the vocal sound source and the fundamental frequency of voiced sounds.
Slow-playback	Pre-recorded sounds are replayed at a slower rate than the original signal, and each spectral component is lowered in frequency by a multiplicative factor equal to the slowdown factor.
Time-compressed slow-playback	Essentially the same as slow-playback, but speech is compressed in time deleting segments periodically, deleting successive pitch periods of voiced sounds, and deleting segments according to phonological rules.
Frequency shifting	All spectral components of the entire frequency spectrum are shifted downwards by a fixed displacement.

These lowering schemes have been incorporated in various frequency lowering hearing aids. The first wearable frequency transposition hearing aid was developed in the 1960's by Bertil Johansson (Erber, 1971:530). This hearing aid, the Oticon TP 72, aimed to shift only high frequency phonemes to lower frequencies without affecting the main vowel formants. This was achieved by filtering energy above 4000 Hz, modulating it with a 5000 Hz signal, and presenting the difference components mixed with the original speech sound below 1500 Hz (Erber, 1971:530). Early studies reporting on the efficacy of this device produced low levels of evidence in the form of case studies. Johansson (1966:367) investigated the use of the Oticon TP 72

on children with profound hearing loss, and found that three of the five participants demonstrated 100% discrimination of /sa/ and /ja/ after two training sessions, and in a related experiment three of five participants also increased their discrimination scores of 25% to 75% with the discrimination of /sa/ - /ja/ - /ka/ - /tja/ after several training sessions. However, an unpublished study by Hirsh, Greenwald, and Erber (as cited in Erber, 1971:530) produced a higher level of evidence, because of the non-randomised intervention study design (Level 3). Three children used the Oticon TP 72, and three children received conventional amplification. Both groups received the same amount of training on speech perception and speech production for four weeks. Comparison of word recognition scores after this period revealed no significant differences (Erber, 1971:530). Ling (1968, as cited in Erber, 1971:531) also investigated the use of the Oticon TP72 in eight children with profound hearing loss by means of case studies (producing a Level 5 of evidence), and found no significant differences for performance with and without frequency transposition.

The next development in frequency transposition hearing aids occurred in 1967, when Ling and Druz developed a hearing aid that produced pure-tone analogue signals in the 750-1000 Hz range whenever speech energy existed in the 2000-3000 Hz range (Erber, 1971:531). A study that produced Level 3 evidence was conducted by Ling and Druz (1967, as cited in Erber, 1971:531) which evaluated the efficacy of their device in four children with profound hearing loss, paired to four children who were trained similarly with their own hearing aids. All the children in both groups demonstrated significant progress with the instruments with which they had been trained, thus dismissing the notion that the device itself was responsible for the progress (Erber, 1971:531).

In 1968, Guttman and Nelson designed a frequency-dividing speech coder for children. This coder isolated high frequency speech sounds by means of selective filtering, and then produced a low frequency pulse for a proportion of original wave-zero-crossings. This low frequency pulse is presented together with the original speech signal (Erber, 1971:532). Guttman, Levitt, and Bellefleur (1970) tested this device in a Level 3 study, where six children with hearing loss received amplification through the coder and two children in a control group were fitted with conventional

amplification. It was found that after an extensive training session, articulation of the /s/ and /j/ by the children using the coder, improved slightly more than the control group (Guttman et al., 1970:19).

Another variation of the speech coder was developed in 1969, by Ling and Doehring. This device divided speech energy in the 1000-4000 Hz range into ten logarithmic intervals, and transposed it to the 100-1000Hz range in ten 100 Hz-wide analogue channels (Erber, 1971:532). An extensive Level 3 study produced evidence that children did not perform better with the device than with conventional amplification (Erber, 1971:532). Another Level 3 study also did not demonstrate a difference in performance with the coder (Ling & Maretic, 1971:37).

A frequency recording device (FRED) was developed in the 1970s by Velmans. This device subtracted 4000 Hz from every sound in the 4000-8000 Hz area and superimposed it on the unaltered low frequency signal (Robinson et al., 2007:294). A Level 5 evidence study with a Grade C level of confidence indicated that seven of the eight participants with severe to profound hearing loss using FRED demonstrated better performance in quiet conditions without training (Rees & Velmans, 1993:59). This was particularly noted for discriminating contrasts involving the absence or presence of /s/ and /z/ (Rees & Velmans, 1993:58).

The development of power ear-level amplification has lead to a diminished interest in frequency lowering during the 1970s and 1980s. However, by 1996 two frequency lowering devices were commercially available, namely the Emily™ device, and the TranSonic™ FT 40 MKII. The Emily was described as a signal processor rather than a transposition aid, as it created additional harmonics by the multiplication and division of a tone representing the peak energy in the second formant of the phoneme, and imposed these harmonics onto the conventional amplified signal (Gravel & Chute, 1996:257). Although this device was reported by its manufacturers to be of some use to children with even moderate degrees of hearing loss, studies reporting this are lacking (Gravel & Chute, 1996:259).

The TranSonic™ hearing aid operates on the basis of “slow-play” frequency transposition or proportional frequency compression. Consonants and vowels are detected separately and if energy is detected above 2500 Hz, the spectral information is divided by the specific transposition coefficient for that class of phonemes, and proportionally shifted to a lower frequency region (Gravel & Chute, 1996:260). The use of the body-worn model, the TranSonic FT 40 MK II, with children, was investigated by MacArdle and colleagues in 2001. In their study, 11 of the 36 participants demonstrated better performance on auditory discrimination tasks and two participants presented with better speech intelligibility with this device (MacArdle et al., 2001:26-27). However, due to the case study design, only Level 5 evidence was produced by this study, with a Grade C confidence level. The behind-the-ear version of the FT 40, the ImpaCT DSR, became commercially available in 1999, and was tested in a study by Miller-Hansen et al., where 78 children participated. This study compared the aided performance of children using this device to the results obtained from a small subgroup using conventional hearing aids. Hearing loss ranged from mild to profound with flat, sloping or precipitous configurations. Results indicated that the participants showed significant better performance with the ImpaCT than with their previous conventional hearing aids (Miller-Hansen et al., 2003:112). As this study included a control group, the evidence produced by this study can be graded as Level 3, with a Grade B level of confidence.

4.3.3 Linear frequency transposition

The most recent development in frequency transposition is the introduction of the Audibility Extender (AE) in the Widex Inteo hearing aid in 2006. This type of technology utilises linear frequency transposition as its means for lowering high frequencies. Linear frequency transposition does not alter the harmonic relationships between the original and transposed sounds, and preserves the naturalness of the sounds. Only one or two octaves are usually transposed, and the amount of transposition as well as the gain of the transposed sounds may be adjusted manually. The AE picks the frequency with the highest peak energy within a source octave and locks it for transposition. This frequency region will be linearly transposed down an octave, and placed on the slope of the hearing loss where the hearing loss is aidable (Andersen, 2007:21). Auriemma et al. (2008) report on the outcomes of

two case studies using the AE. Two unpublished case studies from the University of Melbourne in Australia also investigated the use of the AE in one adolescent and a nine-year old girl. All the studies evaluated speech perception, speech production as well as listening behaviour and perceived benefit. Although the case studies only produced Level 5 evidence, the inclusion of a wide range of testing materials (speech as well as non-speech sounds) increased the reliability of the results and they are summarised in Table 2:

Table 2: Case studies related to the use of the AE in children and adolescents

AUTHOR(S)	AGE OF CHILD	HEARING LOSS	OUTCOMES
Smith, Dann, and Brown (2007a)	14 yrs 4 months	Extreme ski-slope sensorineural hearing loss	<ul style="list-style-type: none"> • Positive at first, but then requested for the AE to be removed • Accepted the AE after fine-tuning • Significant and rapid improvement in speech perception • Fewer errors in producing final consonants • Self-correction of fricatives based on what was heard • Improvement in classroom listening • High perceived benefit
Smith, Dann, and Brown (2007b)	9 yrs 6 months	Severe sensorineural hearing loss	<ul style="list-style-type: none"> • Significant improvement in speech perception • 14.8% improvement in speech production, especially consonant clusters • Significant positive changes in listening behaviour
Auriemmo et al. (2008)	13 yrs 0 months	Ski-slope sensorineural hearing loss	<ul style="list-style-type: none"> • Improvement in consonant recognition • Significant improvement in consonant production • Improved awareness of environmental sounds
Auriemmo et al. (2008)	8 yrs 0 months	Extreme ski-slope sensorineural hearing loss	<ul style="list-style-type: none"> • Consonant and vowel recognition improved dramatically, especially at low-input levels • Significant improvement in accurate production • Significant improvement in hearing environmental sounds

These studies all seem to favour the application of linear frequency transposition in a population with steeply-sloping high frequency sensorineural hearing loss. However, linear frequency transposition may provide more high frequency speech cues to children with MSSHL than their conventional hearing aids, as these hearing aids rarely provide gain over 6000 Hz. This advantage is demonstrated in Figure 4:

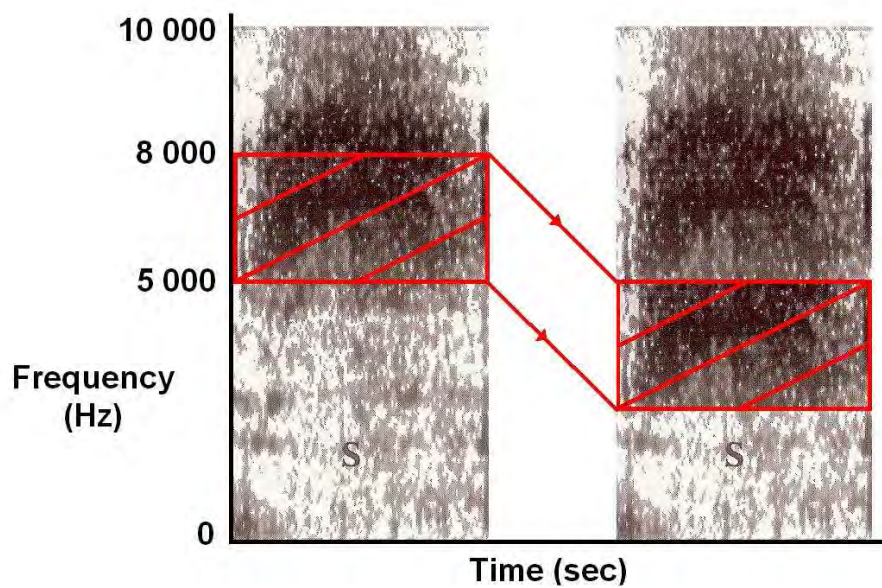


Figure 4: The extra speech cues provided by linear frequency transposition for the speech sound /s/ (adapted from Bernthal & Bankson, 1998:400)

The speech energy in the shaded red block of the spectrogram on the left depicts the amount of speech cues that might be lost with conventional amplification, leaving very little energy with which recognition can occur. These speech cues might become available again if they are transposed to lower frequencies, regardless of the degree and configuration of the hearing loss (Stelmachowicz, personal communication, 2008).

In general, there seems to be consensus that all these advanced digital signal processing strategies aim to ensure consistent audibility across a number of different listening environments (Kuk & Marcoux, 2002:517). Hearing aids employing advanced signal processing schemes are being recommended for infants and young children although empirical evidence regarding the evidence of the superiority of these systems over more basic amplification is limited (Gravel et al., 2003:34). Thus, the need clearly exists for research on these signal processing schemes that would produce high levels of evidence regarding its efficacy, effectiveness and efficiency.

4.4 CONCLUSION

Evidence-based practice guidelines indicate that amplification choices for infants and young children should be made from a knowledge-base that provides evidence regarding the efficacy, effectiveness and efficiency of the particular signal processing feature in question. Conventional advanced digital signal processing schemes have been developed that strive to provide better audibility across a wide variety of listening environments. The efficacy, effectiveness, and efficiency of using some conventional advanced digital signal processing schemes in children have been proved. The importance of high-frequency audibility in children has led to the development of frequency lowering hearing aids. Linear frequency transposition has evolved since the 1960s, and its use in children is currently investigated in local as well as international studies. Some of these studies lack a larger sample size and control of variables, and there is a need for studies that address these weaknesses.

CHAPTER 5

METHOD



CHAPTER AIM: To present the methodology of the procedures followed in this study.

“It is common sense to take a method and try it. If it fails, admit it frankly and try another. But above all, try something.”

~ Franklin D Roosevelt (1932, as cited in Kennedy, 1999:104)

5.1 INTRODUCTION

The introduction of advanced digital signal processing strategies (such as linear frequency transposition) in the development of hearing aid technology has possibly had the biggest influence on creating better opportunities for children with moderate to severe sensorineural hearing loss (MSSHL) to develop oral speech and language skills comparable to those of their normal-hearing peers. These signal processing strategies and their use in children are currently under investigation in numerous international studies. However, research is inextricably connected to the social and historical issues of the present time and place (Struwig & Stead, 2004:21). Current research topics in South Africa seem to be closely linked to fields of study in the developed world, and therefore researchers are encouraged to produce research findings that are relevant to the unique South African context (Stead & Wilson, 1999, as cited in Struwig & Stead, 2004:22). Studies investigating issues (such as paediatric amplification), should be carefully integrated into the relevant cultural context, without ignoring international developments in the related disciplines (Struwig & Stead, 2004:22). Research methods used in the developed world should be examined and adapted for the South African context. These considerations need to be taken into account when planning and executing a research project.

5.2 AIMS OF RESEARCH

The following aims have been formulated for this study:

5.2.1 Main aim

To determine whether linear frequency transposition has an effect on the word recognition abilities of children with a moderate-to-severe sensorineural hearing loss, and if so, what the nature and extent of such an effect would be.

5.2.2 Sub aims

To determine:

- word recognition scores of children using previous generation digital signal processing hearing aids in quiet and noisy conditions respectively.
- word recognition scores of children using integrated signal processing (ISP)-based hearing aids, without linear frequency transposition, in quiet and noisy conditions respectively.
- word recognition scores of children using ISP-based hearing aids, with linear frequency transposition activated, in quiet and noisy conditions respectively.
- to compare word recognition scores of each child as obtained with and without linear frequency transposition in both quiet and noisy conditions.

5.3 RESEARCH DESIGN

The purpose of social research may be three-fold, namely, that of **exploration**, **description** and **explanation** (Babbie, 2002:79). Due to the empirical nature of the study, research was conducted within a **quantitative** paradigm and was distinguished from a qualitative approach due to its purpose, process, data collection procedures, data analysis and reported findings (Leedy & Ormrod, 2005:102-103). The purpose of this study was to **explore** the topic of the effect of linear frequency transposition on the word recognition abilities of young children in an attempt to provide a good basic understanding of it, as the need exists for exploration in this

field due to the dearth of studies reporting on linear frequency transposition and children. This was accomplished by **describing** or determining the word recognition abilities in a number of case studies and by attempting to **explain** the causality between frequency transposition and word recognition abilities. Quantitative studies are conducted within carefully structured guidelines set by the researcher in order to exert some control over dependent and independent variables (Neuman, 2006:253). This is achieved by defining the concepts, variables, and methods beforehand (Leedy & Ormrod, 2005:102). Data collection according to the quantitative approach is specifically related to these variables, and is collected from a sample of a specific population.

A **quasi-experimental single subject time-series** research design was selected to form the structure of the methodology of this study (De Vos, 2002:145), but the inclusion of several subjects enhanced the validity of the single subject study. Advantages of experimentation include the establishment of causality, the exertion of control over the experiment and the opportunity to observe change over time (Babbie, 2002:219). A true experiment starts with a hypothesis, modifies a situation, and then compares the outcomes with or without the modification (Neuman, 2006:247). Random assignment of subjects is also needed to create similar groups in order to facilitate comparison (Neuman, 2006:249). Some variations from this classical experimental design were made in order to materialise the aims of research due to the characteristics of this study. A quasi-experimental design still allowed for testing of causal relationships in a variety of situations (Neuman, 2006:256), but accounted for the lack of randomness in the selection of subject group members in this study, as only a small number of children fitted the selection criteria (Leedy & Ormrod, 2005:237-238).

A unique concept in the research design of this study is the addition of a **time-series design**. The time-series design allows for making observations of a dependent variable over time, before as well as after introducing intervention or treatment. If a substantial change has occurred after the intervention, then it can be reasonably assumed that the intervention brought about the change in the system (Leedy & Ormrod, 2005:238). A weakness in this design is the possibility that an unknown event may occur at the same time as the experimental intervention, and that this

event brings about the measured change. If this is the case, then deducting that the *intervention* caused the change may be erroneous. An unknown event did not occur to the knowledge of the researcher in this study and in order to control for this weakness, a standardised word recognition test was used in this study. This test already provided the norm of the word recognition abilities of South African children with normal hearing and hearing impairment in this age-group.

5.4 SUBJECTS

Due to the quasi-experimental design of this study where random assignment of subjects is not possible, true quantitative sampling techniques were not used. Nonprobability sampling techniques are usually associated with qualitative research designs, as the cases are selected with the characteristics of the case determining whether it is selected or not (Neuman, 2006:220). Purposive sampling as a type of nonprobabilistic sampling was used to identify subjects for this study. This kind of sampling technique was appropriate because particular types of cases for in-depth investigation was identified and these cases needed to be unique and especially informative in order to obtain the necessary information regarding the purpose of the study (Neuman, 2006:222).

5.4.1 Selection criteria

The following criteria have been established in order to select appropriate subjects for this study from a selected centre for hearing-impaired children:

Table 1: Subject group selection criteria

CRITERIA	JUSTIFICATION
<p>Configuration and degree of hearing loss</p> <p>All subjects had to have a bilateral sloping moderate-to-severe sensorineural hearing loss, which must not have progressed more than 10 dB at two consecutive frequencies or 15 dB at one frequency during the last year (Skarzynski, Lorens, Piotrowska, & Anderson, 2006:935).</p>	<p>This type of hearing loss was found to be a good indicator for a child to benefit from frequency transposition (Rees & Velmans, 1993:58).</p>
<p>Middle ear functioning</p> <p>All subjects were required to present with normal middle ear functioning, established by normal otoscopy results and a type A tympanogram.</p>	<p>Middle ear pathology will result in adding a conductive component to the hearing loss (Rappaport & Provencal, 2002:19). Children with conductive hearing loss experience different amplification needs than children with sensorineural hearing loss (Dillon, 2000:256).</p>
<p>Age</p> <p>Subjects between the ages of 5 years 0 months to 7 years 11 months at the time of fitting of the advanced hearing aid were selected.</p>	<p>The children in this age-group are developmentally mature enough to understand what is expected of them during tasks and to cooperate well and consistently during the extended assessments (Louw, Van Ede, & Louw, 1998:335).</p>
<p>Language</p> <p>Subjects had to use English as the primary language.</p>	<p>The language used in international studies was English (MacArdle et al., 2001) and comparison of results between studies would be more accurate. High-frequency speech sounds carry a high informational load in English (Rudmin, 1981:263).</p>
<p>Current hearing aids</p> <ul style="list-style-type: none"> - Subjects had to have at least two years' experience with binaural conventional hearing aids utilizing serial or parallel processing set according to the amplification targets prescribed by the DSL m[i/o] (Scollie, 2006:10). - Subjects should have had regular daily use of the hearing aids for at least 10 hours/day - The hearing aids should not have had feedback during normal use (Schum, 1998). - Subjects should have been followed-up by the same audiologist for at least 2 years prior to the fitting of the advanced hearing aid. 	<p>The child's current hearing aids must be optimised to reflect the current best practice (Flynn et al., 2004:480) so that accurate comparisons can be made between different technologies.</p>
<p>Educational environment</p> <p>All subjects should have attended the selected centre for hearing-impaired children in for at least 2 years prior to the fitting of the advanced hearing aid.</p>	<p>A pre-primary school gives an opportunity for sensorimotor, language and socio-emotional growth (Owens, 1999:363), and all the subjects should be subjected to the same educational environment where uniform opportunities for growth in these areas are created.</p>
<p>Speech therapy</p> <p>All subjects should have received at least 1 year of weekly speech- language and hearing therapy.</p>	<p>Intervention aimed at auditory perceptual development should be provided for the child with hearing loss as part of a comprehensive service delivery model in order to maximize the use of residual hearing through amplification (Moeller & Carney, 1993:126).</p>

5.2.4 Subject selection procedures

The selection of subjects as multiple single case studies was determined by their characteristics according to nonprobabilistic purposive sampling (as described above). Children in the English Grade R and Grade 1 classes were identified as possible subjects based on observation and experience working with these children in the past two years. This could be done accurately as the researcher had been working at this centre for hearing-impaired children for two and a half years before the commencement of this study. Permission from the institutions involved was requested. Clearance from the hospital with which the centre is affiliated as well as the centre itself was obtained. Letters requesting informed consent for participating in the study were given to the primary caregivers/legal guardians of the identified subjects (see Appendix C). After informed consent was granted, their school files were obtained and the personal and audiological information in the file was used to verify whether the child fitted the selection criteria. Informed assent was also obtained from the subjects (see Appendix D). The size of the subject group depended on the number of appropriate subjects who gave consent/assent to take part in the study. Letters requesting participation were also given to the principal and class teachers of all the subjects (see Appendix F).

5.4.2 Sample size

The sample size of this study consisted of seven subjects. These subjects were all the children in the centre who matched the selection criteria. The other centres in the Western Cape are all far removed from this particular centre, and thus it would have been very cumbersome to include subjects from other institutions in order to increase the sample size due to the time it would take to travel to the centre for all the assessments. A larger sample size may increase the likelihood of precision and reliability (Struwig & Stead, 2001:119), but a smaller sample size for the purpose of this study allows for cost-efficient in-depth monitoring of possible changes in the word recognition skills of children with moderate-to-severe sensorineural hearing loss.

5.5 DATA COLLECTION

Data was collected using the following apparatus and materials:

5.5.1 Data collection apparatus

Otoscopy: A Heine mini 2000 otoscope used with specula was used to perform otoscopy in order to determine any abnormalities of the outer ear and tympanic membrane before assessments commenced.

Tympanometry: Tympanometry was performed with a GSI 38 Autotymp and probes, calibrated on 19/06/2007, to detect any abnormalities of the middle ear system as this may have influenced the accuracy of the assessments.

Audiometry: An Interacoustics Clinical Audiometer AC40, calibrated on 19/06/2007, was used for pure tone and speech audiometry in order to assess hearing thresholds. Stimuli were presented through Eartone 3A insert earphones and a Radio Ear B-17 bone conductor to determine unaided thresholds. Stimuli were also presented through speakers at 90° azimuth for assessing aided thresholds. All these assessments took place in a 2m x 2m sound treated booth.

ISP-based hearing aids with the option of linear frequency transposition: The Widex Inteo 9 and 19 VC hearing aids were selected to use as amplification devices in this study. The models (Widex IN 9 VC or IN 19 VC) selected for the subjects in this study depended on the severity of the hearing loss, and were behind-the-ear hearing aids connected by standard #13 tubing to a full shell acrylic earmould with appropriate venting.

Programming of the hearing aids: A Mecer Celeron personal computer and a GN Otometrics NOAHlink system were used to program the hearing aids together with the cables and programming shoes supplied by the hearing aid company. Initial amplification values were calculated using the Compass v4.2 software provided by the company.

Verification: The Audioscan Verifit was used to verify the output from the hearing aids according to the amplification targets prescribed by the DSL m[i/o] and to check whether the distortion levels of the hearing aids were within acceptable limits. Listening checks were also performed using a stethoclip. In addition to the verification performed by the Audioscan Verifit, audibility of the transposed sounds when the frequency transposition was activated were verified using the SoundTracker provided in the Compass software, as this would have given a more accurate verification (F. Kuk, personal communication, 2007).

Sterilization fluid: Alcohol swabs were used to disinfect the specula and Milton sterilization fluid was used to disinfect the probes for proper hygiene control.

5.5.2 Data collection materials

Results of the otoscopy, tympanometry and pure-tone audiometry were recorded on an audiogram (see Appendix A). Word recognition scores were measured with the Word Intelligibility by Picture Identification (WIPI) test (Ross & Lerman, 1971). This test has been standardised for the South African context. Results of the speech audiometry measurements were recorded on separate forms (see Appendix B).

5.6 RESEARCH PROCEDURE

Research was conducted using the following procedures for data collection, data recording and analysis:

5.6.1 Data collection procedures

The subject group was divided into two groups in order to make the data collection procedure more manageable in terms of the number of children assessed per day and to keep the interruptions of the classroom activities to the minimum. The two subject groups alternated weekly and the schedule for these assessments is depicted in Table 2. The data collection process was divided into six research phases for each subject group. An overview of the total 7-week research process for this study is depicted in Figure 1:

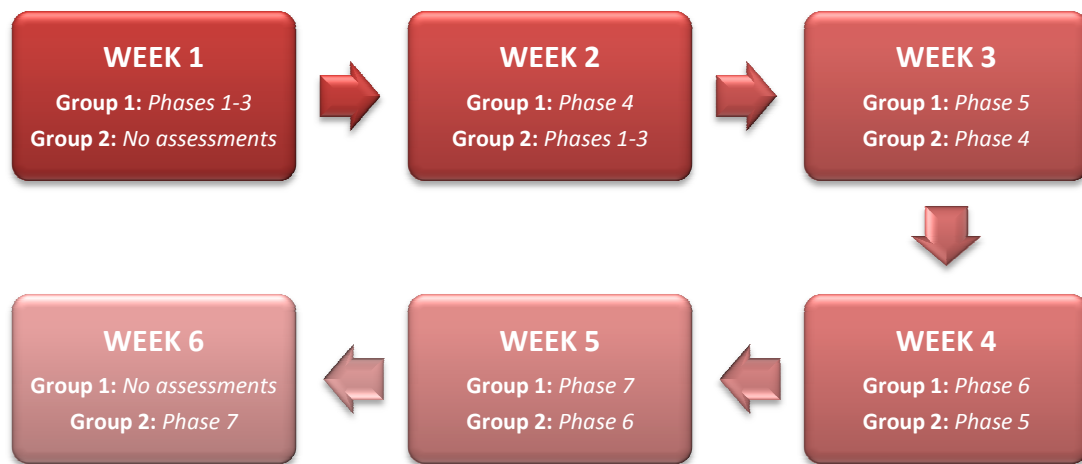


Figure 1: An overview of the research phases

Table 2: Assessment schedule for the subject groups

WEEK 1				WEEK 2		
Group 1	Phase 1: First assessment with digital signal processing (DSP) hearing aids Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI	Phase 2: Second assessment with DSP hearing aids Hearing aid check Otoscopy Tympanometry Pure-tone audiometry WIPI	Phase 3: Third assessment with DSP hearing aids Hearing aid check Otoscopy Tympanometry Pure-tone audiometry WIPI <i>Fitting of ISP-based hearing aid without linear frequency transposition</i>	Phase 4: Acclimatization period		
	No assessments			Phase 1: First assessment with DSP hearing aids Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI	Phase 2: Second assessment with DSP hearing aids Hearing aid check Otoscopy Tympanometry Pure-tone audiometry WIPI	Phase 3: Third assessment with DSP hearing aids Hearing aid check Otoscopy Tympanometry Pure-tone audiometry WIPI <i>Fitting of ISP-based hearing aid without linear frequency transposition</i>
WEEK 3				WEEK 4		
Group 1	Phase 5: Assessment with ISP-based hearing aids without linear frequency transposition Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI <i>Activation of frequency transposition</i>			Phase 6: Acclimatization period		
	Phase 4: Acclimatization period			Phase 5: Assessment with ISP-based hearing aid without linear frequency transposition Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI <i>Activation of linear frequency transposition</i>		
WEEK 5				WEEK 6		
Group 1	Phase 7: Assessment with ISP-based hearing aid with linear frequency transposition Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI			No assessments		
	Phase 6: Acclimatization period			Phase 7: Assessment with ISP-based hearing aid with linear frequency transposition Hearing aid check and verification Otoscopy Tympanometry Pure-tone audiometry WIPI		

The assessments conducted in each phase were completed within 60 minutes and the test battery will be discussed below:

5.6.1.1 Phases 1 and 2: Assessments with previous generation digital signal processing (DSP) hearing aids

The assessments from Phases 1 and 2 are discussed below:

Otoscopy

Otoscopy was performed prior to the assessment to detect any excessive cerumen or abnormalities of the tympanic membrane and external auditory meatus, by inspecting the ear canal with an otoscope. If any abnormalities were found, the subject would have been appropriately referred and excluded from the study. No subjects presented with abnormalities of the outer ear and tympanic membrane, and therefore no referrals were indicated.

Hearing aid check and DSL m[i/o] verification

All subjects' hearing aids were checked to see whether the hearing aids were in an excellent working condition with no distortion or intermittence. The subject's current hearing aid was removed from his/her ear and connected to the 2cc-coupler in the test chamber of the Audioscan Verifit and a distortion test was run to detect any harmonic distortion at the frequencies 250 Hz to 4000 Hz. Total harmonic distortion of less than 5 dB at all the frequencies were accepted (Dillon, 2000:87). A listening check with a stethoclip was performed. Real-ear measurements were then performed to objectively verify the hearing aid's output to comply with the amplification targets set by the DSL m[i/o] (Scollie, 2006:10). A real-ear-to-coupler-difference (RECD) transducer was connected to the 2-cc coupler of the Audioscan Verifit and the coupler response to a given signal was measured. The RECD transducer was taken off from the 2-cc coupler and connected to a foam tip, and both the foam tip and the probe tube were inserted into the child's ear canal so that the tip of the probe tube is +/- 2 to 5 mm from the ear drum. A real-ear response was measured and Audioscan Verifit calculated the difference between the real-ear and the coupler response as the RECD. Adjustments to the gain output of the hearing aid can be made, if necessary, in order to match the amplification targets set by the

DSL m[i/o]. It was found that matching the output of the hearing aid to the targets of amplified speech at 55 dB SPL and 70 dB SPL provided the most assurance that fitting goals are being met (Moodie et al., 2007). This was done separately for each ear.

Tympanometry

Tympanometry using a 226 Hz-probe tone was conducted (Fowler & Knowles, 2002:176). A probe-tip was placed into the child's ear while measurement of middle ear functioning takes place. If any abnormalities were detected, the subjects were appropriately referred and excluded from the study. No subjects presented with any abnormal tympanograms, therefore no referrals were done.

Pure-tone audiometry

Pure-tone air-conduction audiometry was performed using insert-earphones in a sound treated room in order to establish unaided air-conduction thresholds (Diefendorf, 2002:479). These thresholds were established using the Modified Hughson-Westlake procedure (Harrell, 2002:72). The child was seated between two free field speakers, approximately 1.5m away from the speakers. The child was instructed to press the response button every time a sound was heard. This was practiced a few times to make sure that the child understood the procedure. Insert-earphones were placed in the ear canal, and testing began by presenting 2000 Hz pulsed warble-tones 40 to 50 dB above the estimated threshold. If the child responded to the sound, descends in stimulus intensity commenced in 10 dB steps until the child did not respond anymore. The stimuli were increased in 5 dB steps until the child responded again. Threshold was established as soon as the child responded twice on the ascending presentation (Northern & Downs, 2002:186). This procedure was repeated for the frequencies 250, 500, 1000, 4000 and 8000 Hz in both ears.

After these thresholds were established, the insert-earphones were taken out and a bone-conductor was placed on the mastoid process behind the ear. The same procedure was followed as for the unaided air-conduction thresholds for the frequencies 500, 1000, 2000 and 4000 Hz. Bone conduction audiometry was performed as this forms part of a standard audiometric test battery.

Aided air-conduction thresholds of the subject group were determined as well using the Hughson-Westlake method (as described in the above section) with narrowband noise as stimulus type. This procedure uses an ascending method of establishing threshold (Harrell, 2002:72). The hearing aids were placed in the child's ear and only one hearing aid at a time was switched on in order to determine ear-specific aided information as far as possible. The hearing aid to be switched on first was the hearing aid that was fitted to the better ear. The narrowband noise stimulus was presented via the free field speaker (on the side of the hearing aid to be tested) at 10 dB below the estimated threshold and increased in 5 dB steps until the child responded and decreased again in 5 dB steps until the child did not respond. Threshold was established again as soon as the child responded twice to the same level on the ascending presentation.

Speech audiometry

The WIPI was administered to the child with both hearing aids in-situ and switched on at the same time in a sound treated room with speakers at 0° and 180° azimuth approximately 1.5m from the child's head. A female talker who was unfamiliar to the child presented 25 pre-recorded monosyllabic words from the WIPI in quiet at 35 dB HL and the child was asked to point to the named picture from a set of nine pictures. The test procedure was repeated at 55 dB HL, and at 55 dB HL with signal-to-noise ratio of +5 dB. This was performed by presenting the female talker through the free field speaker at 0° azimuth and presenting speech noise through the speaker set at 180°. This corresponded with the signal-to-noise ratios of typical classrooms (Flexer, 2006).

5.6.1.2 Phase 3: Third assessment with previous generation DSP hearing aids

Assessments from Phase 3 and the fitting of the ISP-based hearing aids without linear frequency transposition were performed as follows:

Otoscopy, tympanometry, pure-tone audiometry and speech audiometry

These assessments were performed as described in Phases 1 to 2 in Section 5.6.1.1.

Fitting of ISP-based hearing aids without linear frequency transposition

The new hearing aids were connected to the computer and fitted using the manufacturer's software and cables and set to the master program with no frequency transposition. The output of the hearing aids were also verified using probe-microphone measurements described in Section 5.6.1.1 and fine-tuned to match the DSL m[i/o] targets as close as possible.

5.6.1.3 Phase 4: Acclimatisation period

A period of 12 days was allowed for the subjects to acclimatise to their new hearing aids, as it was found that 1 week seemed to be sufficient time (Marriage, Moore, Stone, & Baer, 2005:45).

5.6.1.4 Phase 5: Assessment with ISP-based hearing aid without linear frequency transposition

The assessments from Phase 5 and the activation of the linear frequency transposition are discussed below:

Otoscopy, tympanometry, pure-tone audiometry and speech audiometry

This assessment was identical to the procedure followed in Phases 1 to 2 in Section 5.6.1.1.

Activation of the linear frequency transposition

The frequency transposition program was activated as the start-up program after the assessment with the ISP-based amplification. The software calculated a default start frequency that represented the region where transposition occurred based on the listener's hearing loss (Kuk et al., 2007:60). When the frequency transposition was activated and set as the start-up program, audibility of /s/ and /ʃ/ was checked with the SoundTracker software (Kuk, 2007). The hearing aids were connected to the Audioscan Verifit and computer with the NOAHlink and set to the frequency transposition program. The SoundTracker in the Compass software was used to display an objective and visual display of the audibility of the transposed sounds in

addition to the Audioscan Verifit (Kuk, personal communication, 2007). The input stimuli from the Verifit were used to verify the audibility of the transposed sounds, and were presented at soft (55 dB SPL) and average levels (70 dB SPL).

5.6.1.5 Phase 6: Acclimatisation period

A period of 12 days was allowed again for the subjects to acclimatise to the new modified input signal (Marriage et al., 2005:45).

5.6.1.6 Phase 7: Assessment with ISP-based hearing aid with linear frequency transposition

Assessments with the ISP-based hearing aid with linear frequency transposition are discussed below:

Otoscopy, tympanometry, pure-tone audiometry and speech audiometry

This assessment was identical to the procedure followed in Phases 1, 2, and 5, in Section 5.6.1.1.

5.6.2 Procedures for data recording and analysis

Data was recorded and analysed using the following procedures:

5.6.2.1 Recording of data

The results from all the tests (except the speech audiometry scores) were depicted on an audiogram (see Appendix A) and the word recognition scores were recorded on a separate form (see Appendix B). The researcher recorded all the data during the assessments.

5.6.2.2 Procedures for analysis of data

The data was analysed with the help of a qualified statistician using statistical procedures. The Pearson correlation coefficient was used because the strength of

two variables can be measured and expressed as the strength and direction of a bivariate relationship (De Vos, 2002:244). Inferential statistics were used in order to interpret the word recognition scores from norms provided by Papso and Blood (1989).

5.7 ETHICAL CONSIDERATIONS

Ethical considerations form an integral part of quasi-experimental research due to its “intrusive” nature. Quasi-experimental research requires placing a subject in a setting where his/her behaviour may be manipulated by introducing an independent variable and measuring the change in behaviour as the dependent variable (Neuman, 2006:269). Ethics provide the norm and standards about the most correct conduct towards subjects, sponsors and other researchers (De Vos, 2002:63). Researchers want to obtain knowledge, perform problem solving and design new methods of treating diseases and disorders. All of this needs to be done in an honest, responsible, open and ethically justifiable manner (Hegde, 1987:414). The underlying foundation of ethical research is to preserve and protect the human dignity and rights of all the subjects taking part in a research study (Jenkins, Price, & Starker, 2003:46).

The ethical principles of autonomy, beneficence and justice were incorporated in this study (Hyde, 2004:297), and are discussed below:

5.7.1 Autonomy

According to the Oxford Advanced Learner’s Dictionary (Crowther, 1995:68), the term autonomy refers to independence and control over one’s own affairs. In research, autonomy means strictly voluntary participation (Leedy & Ormrod, 2005:107). Informed consent, withdrawal of subjects, privacy, confidentiality, and anonymity, disclosure of information, debriefing of respondents and ethical clearance are discussed in Table 3:

Table 3: The components of autonomy relevant to this study

COMPONENT	DESCRIPTION
Informed consent	The subject involved in the study should have the legal capacity to give consent (Jenkins et al., 2003:47), by making an informed decision whether or not to take part in the study. As the subjects in this study are all children, the parent or legal guardian acted in the best interest of the child as a minor. Written informed consent was obtained by presenting the parent or legal guardian of the child with a letter explaining the purpose of the study, the procedures to be followed during data collection by the researcher, the child as well as the caregiver, and the possible advantages and disadvantages of taking part in the study (De Vos, 2002:65). It stated that information will be kept strictly confidential (see Appendix C). Informed assent also obtained from the child in order to preserve their autonomy. The child was provided with an age-appropriate brochure explaining the procedures to be followed, the issue of confidentiality and was given the opportunity to refuse to take part in the study (see Appendix D).
Subject withdrawal	The norm for social research is that all participation in social research should be voluntary (Babbie, 2002:521). The child and the caregiver therefore reserved the right to withdraw at any time from the study, without being penalized or sacrificing any tangible benefits they might receive for participating in the study.
Privacy, confidentiality and anonymity	The subjects' right to privacy was protected by handling all information as confidential and anonymous (De Vos, 2002:67). Permission was also obtained from the centre for hearing-impaired children to use information in the children's school files for selection procedures.
Disclosure of information	Subjects were informed that the information gained from the study would be used for academic purposes – either as an article or presentation. This was done in an objective manner, keeping the principle of confidentiality and the language accurate, objective and unambiguous. When writing the report after the results have been obtained, all forms of bias and plagiarism will be avoided. Errors and limitations of the study were admitted and recommendations were made for future research (De Vos, 2002:72).
Debriefing of respondents	Debriefing of respondents is a way of minimizing harm (De Vos, 2002:73). The results from the study was summarized in a letter and sent to the subjects. The dissertation will also be available in an academic library upon request. Interviews with the primary caregivers/legal guardians were also conducted after the participation of the study in order to rectify any misperceptions that may exist and to provide an overview of their child's performance with the advanced technology, so that an informed decision could be made which hearing aid is best for their child.
Ethical clearance	Ethical clearance was obtained from the Research Proposal and Ethics Committee from the University of Pretoria (see Appendix E) and the Committee for Human Research from the University of Stellenbosch (see Appendix G). Permission was also obtained from the centre for hearing-impaired children (see Appendix F).

5.7.2 Beneficence

Beneficence refers to showing active kindness (Crowther, 1995:100). It also refers to the conferral of benefits (Hyde, 2004:297). Subjects were not exposed to undue physical or psychological harm (Leedy & Ormrod, 2005:107; Babbie, 2002:522). This was accomplished by including the following components discussed in Table 4:

Table 4: Beneficence as a relevant ethical principle for this study

COMPONENT	DESCRIPTION
Competency	The researcher is qualified to conduct research due to her qualification and experience in the field of Audiology. Two research supervisors from the University of Pretoria supervised the study, and valuable input was gained from leaders in the local and international field of paediatric audiology. The researcher (STA 0024554) and the tutors are registered with the Health Professions Council of South Africa.
Relevance	As all clinicians are urged to display evidence-based practice especially in the field of paediatric audiology, this study is highly relevant and may yield valuable information regarding the prescription of technology to meet the needs of the hearing-impaired paediatric population.
Risks	Taking part in a quasi-experimental research study may involve the disruption of regular, daily activities (Babbie, 2002:521). However, the risk of participating in this study did not unreasonably exceed the normal risk of day-to-day living. No medical risks were involved in the study. Standard procedures for hearing aid fittings were followed, but protocols were conducted more frequent than for ordinary fittings in the 6-week period following the commencement of assessments of the subject group. The subjects were thus removed from the classroom for one hour each day for six days in total. It was arranged with the teachers and the parents to create opportunities to make sure that the subject did not fall behind the rest of the class due to these absences.
Discrimination	Subjects were not discriminated against due to their gender, race or economic status.

5.7.3 Justice

Justice refers to the honesty with professional colleagues (Leedy & Ormrod, 2005:108). The researcher has a responsibility towards other colleagues in the scientific community as well as sponsors or contributors towards the study (Babbie, 2002:526). All co-workers were therefore acknowledged. As this study was dependent on a sponsorship from a hearing aid manufacturer, namely Widex Denmark, it was clarified beforehand that the sponsor would not be prescriptive towards the study, nor that the identity of the sponsor would remain undisclosed. The researcher did not modify or conceal the real findings of the study in order to meet the expectation of the sponsor, or concealed the real goal of the experiment (De Vos, 2002:71). Justice also refers to the fairness in the distribution of benefits among members of the society (Hyde, 2004:297). Thus the subjects were given a choice to keep the hearing aids donated by Widex Denmark for this study.

5.8 RELIABILITY AND VALIDITY

Reliability and validity are two issues that cannot be ignored when conducting research within the unique South African context. This is true especially in the field of hearing loss and subsequent communication disorders, where language and culture play a major role in the development of language and treatment of communication disorders. **Reliability** is a reflection of the accuracy, consistency and stability of the

word recognition scores obtained (Struwig & Stead, 2001:130), and **validity** refers to the soundness of the quasi-experimental research design employed in this study (Struwig & Stead, 2001:136), whether or not the causal effect of the independent variable on the dependent variable can be measured accurately (Bailey, 1994:239).

The main subject of concern in quasi-experimentation refers to the reliability of the instrumentation used to measure the possible causal effect (Bailey, 1994:239). Three types of reliability are found in quantitative research methods, namely stability reliability, representative reliability, and equivalence reliability (Neuman, 2006:189). These terms and their definitions are provided in Table 5:

Table 5: The three types of reliability in quantitative research methods (compiled from Neuman, 2006:189)

TERM	DEFINITION
Stability reliability	Refers to the reliability of the measurement over time, verified by using a test-retest method.
Representative reliability	Refers to the consistency of test results for different social and cultural groups
Equivalence reliability	Refers to the reliability of results when different specific indicators are used

In this study the Word Intelligibility by Picture Identification (WIPI) was used in order to measure word recognition scores of the subjects. According to Ross and Lerman (1970:50) the WIPI provides excellent stability reliability with reliability coefficients of 4.7 to 7.7. It also provides good equivalence reliability, with a learning effect that is clinically insignificant. Representative reliability was achieved by using a South African version of the WIPI that has been adapted to allow for cultural differences in vocabulary (Müller, personal communication, 2008).

Furthermore, the reliability of this study was also increased by clearly conceptualising all the constructs beforehand. The level of measurement was increased so that the precise measurement of word recognition could be achieved, and subjective assessments of word recognition scores were corroborated by objective verification of the hearing aid fittings (Neuman, 2006:191). The clinical

instrumentation for administering the WIPI was also calibrated prior to the study, in order to ensure accurate presentation levels.

In order to increase the validity of quasi-experimental research methods, the level of external and internal validity should be increased (Struwig & Stead, 2001:136). Although external validity refers to the generalisation of the results to other populations (which would be difficult in this study, as the sample size is very small), care was taken in the sampling procedures, time, place and conditions in which the research was conducted in order to increase the external validity (Struwig & Stead, 2001:136). The internal validity was increased by controlling for extraneous variables, as presented in Table 6:

Table 6: The controlling of extraneous variables in this study (compiled from Struwig & Stead, 2001:239)

VARIABLE	DEFINITION
Maturation	In order to control for the maturation effect, the assessments were schedules within a 5-week period, as this was reported by Ross and Lerman (1970:50) to yield test scores independent of the maturation effect.
History	Changes in word recognition scores due to unrelated factors were controlled for by monitoring the subjects closely for any middle ear pathology, as well as checking their hearing aids every time before assessments took place.
Testing	In order to control for the influence of previous testing, all the word lists were produced in a random sequence.
Instrumentation	The measuring instrumentation was not altered during the assessments, and the same clinical instrumentation and test environment were used for all the assessments.
Selection	The subjects were selected and divided into two groups that alternated weekly. This also provided a control for selection and other issues, such as instrumentation and testing that could have confounded the results.
Attrition	All the subjects participated in the study until it was completed.
Diffusion of treatment	As children participated as subjects in this study and were unaware of the settings in their hearing aids, they could not communicate information about the research project to the other group of participants.

As with all experimentation, it may be difficult to measure the reliability and validity of the measures that were used to conduct the research project, but the greater the degree of control over all the variables, the more accurate the measures of causality (Bailey, 1994:239).

5.9 CONCLUSION

Research should be conducted in order to investigate the applicability of international studies and recommendations within the South African framework. In order to accomplish this, research methods should be examined and adapted for the unique South African context. This empirical research was planned in order to produce context-specific results regarding word recognition and linear frequency transposition in children with moderate-to-severe sensorineural hearing loss. The ethical principles of autonomy, beneficence, and justice were considered in this study due to the intrusive nature of quasi-experimental research design, in order to protect and preserve the human dignity and the rights of all the subjects (Jenkins et al., 2003: 297). Measures were taken to ensure increased validity and reliability of this investigation.