CHAPTER 2: OVERVIEW AND DISCUSSION OF THE LITERATURE AND ISSUES SURROUNDING SENSORINEURAL HEARING LOSS IN INFANTS/CHILDREN RELEVANT FOR PEDIATRICIANS IN THE SOUTH AFRICAN CONTEXT.

2.1 INTRODUCTION

The following quote applies to a child with a serious to profound, congenital hearing loss: "*he cannot ask, for he has no words. He has no words in which to think; no words to understand explanations. Never having heard any, he does not even know such things as words exist*" (Greinwald & Hartnick, 2002).

To give infants and children the best possible health care, especially those with a severe-to-profound sensorineural hearing loss, professionals must integrate their knowledge to form a meaningful whole. Therefore one needs a positive attitude surrounding teamwork and learning from other professional team members' field of specialty. Such a team is willing to learn from one another and is collaborative, not competitive (Morsink, 1991).

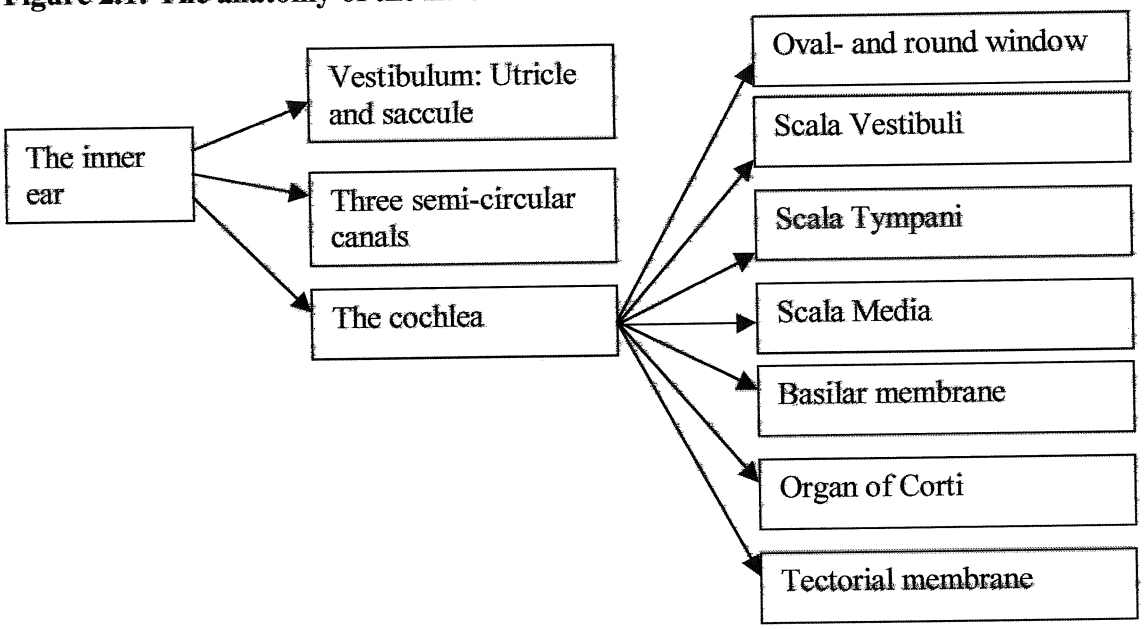
The factors (neurological and physiological) surrounding sensorineural hearing loss are presented in this chapter and a discussion on early hearing screening and early intervention is provided. The need for teamwork is discussed quoting previous research by emphasizing the role of both the audiologist and the pediatrician. A need exists to provide pediatricians with the newest research and technical development for diagnosis and intervention of babies and children with sensorineural hearing loss.

2.2 THE ANATOMY AND THE STRUCTURE OF THE (INNER) EAR

The hearing mechanism is an amazingly intricate system. The hearing organ is divided into three different parts. The external ear which receives and transfers the sound waves to the middle ear. The middle ear which transfers the vibration of the tympanic membrane to the fluid in the cochlea, amplifying the sound energy in this process. In the inner ear are the receptors for hearing which are found in a highly specialized membrane structure, the cochlea (Sherwood, 2001).

The anatomy of the inner ear is presented schematically in figure 2.1.

Figure 2.1: The anatomy of the inner ear.

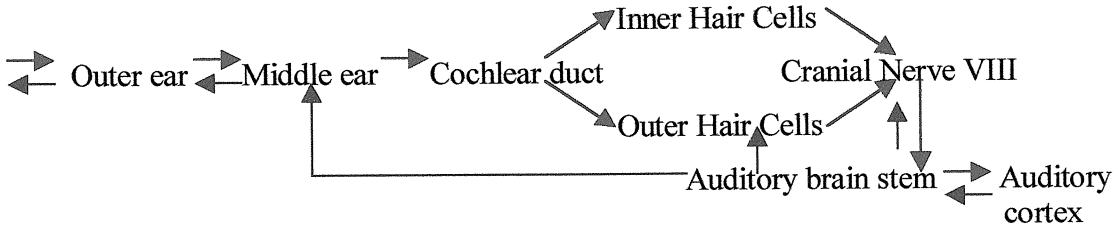


Sources: Sherwood (2001) and Vander, Sherman & Luciano (2001)

The cochlea houses the sensory system for hearing. It is a fluid-filled space interfaced between the middle ear and the auditory nerve. The cochlea acts as a device for converting sound into electro-chemical energy that transmits information to the brain concerning the frequency, decibels and the phase of the sound waves (Vander, Sherman

& Luciano, 2001). In figure 2.2 a schematic representation of the structures and functions of the auditory system is given, illustrating both the afferent- and efferent pathways.

Figure 2.2: A representation of the structures and function of the auditory system.
(* afferent; * efferent)



Sources: Sherwood (2001) and Vander, Sherman & Luciano (2001)

Information concerning the functions of the structures involved in the cochlear mechanism could help towards understanding the causes and effects of sensorineural hearing loss during cochlear development (Lenoir, Puel & Pujol, 1987).

2.3 OVERVIEW ON SENSORINEURAL HEARING LOSS

The aim of this paragraph is to show the importance of being knowledgeable about sensorineural hearing loss. The discussion on this type of hearing loss is to aid the understanding of this complicated problem and the interwoven issues that lead to barriers in early diagnosis and intervention. The knowledge important for understanding hearing loss motivates the rationale underlying this study.

A pathology of the inner ear is a lesion or obstruction that occurs between the inner ear and the brain, resulting in the prevention of sound being perceived (Hall, 2000). Sensorineural hearing loss is the type of hearing loss that occurs when there is an interference with the action of the hair cells in the cochlea or the acoustic nerve (Cranial Nerve VIII) (Shipley & McAfee, 1998). Hair cell action may be mechanically impeded or the hair cells may be damaged or destroyed. About 1/600 infant has a congenital hearing loss (occurs at birth) and many more acquire a hearing loss (occurs after birth) owing to conditions encountered during the neonatal period (Guralnick, 1997). Children requiring

intensive medical treatment during the neonatal period are 1/25-30 (Guralnick, 1997). This puts them in a high-risk category for hearing loss. 2/1000 infants are born with a severe-to-profound hearing loss and 5-6/1000 have a mild-to-moderate hearing loss (Guralnick, 1997). More than 90% of congenital hearing losses are of cochlear origin (Jakubikova, Kabatova & Zavodna, 2003).

The term sensorineural hearing loss indicates uncertainty whether the loss of hearing is due to a lesion in the inner ear or the neural pathways (Berkow et al, 1992). It is important therefore to differentiate between sensory- (cochlear) and neural pathologies (N VIII – vestibulocochlearis) since the results can be life threatening if left untreated (for example a tumor). In table 2.1 the differences between sensory and neural hearing loss is listed.

Table 2.1: Differences between sensory hearing loss and neural hearing loss

Test applied	Sensory hearing loss	Neural hearing loss
Discrimination of speech	Moderate decrement	Severe decrement
Discrimination with increasing intensity	Improves	Deteriorates
Recruitment	Present	Absent
Acoustic reflex decay	Absent	Present
Sensitivity to small recruitments in intensity	Good	Poor
Tone decay	Mild	Marked
Waves in auditory brainstem response (ABR)	Well formed with normal latencies	Absent or with abnormal long latencies

Source: Berkow et al, 1992

Sensorineural hearing loss has three fundamental effects:

- Reduction in cochlear sensitivity;
- Reduction in frequency resolution;
- Reduction in the dynamic range of the hearing mechanism (Berkow et al, 1992).

(It is important to note the differences between the different losses for adequate diagnosis and intervention but for the purpose of this study, the term sensorineural hearing loss will be used throughout the text.)

2.4 THE POSSIBLE CAUSES OF SENSORINEURAL HEARING LOSS

The detection of an infant/child with a hearing impairment does not only rely on the results of the audiometry tests, but on the understanding of the relevant high-risk conditions as well as the behaviours and responses that might suggest a hearing impairment (Calderon, Bargones & Sidman, 1998).

In the following paragraphs a discussion on the possible causes of sensorineural hearing loss is given. The reason being to emphasize why pediatricians must have sufficient knowledge of the given risk factors in order to identify a possible loss of hearing.

A) Prenatal causes

In newborn babies structural defects, known as congenital abnormalities, can be due to embryological malformations or structural changes secondary to infections (Berkow et al, 1992).

Congenital abnormalities:

- **Syndromes and /or inherited disorders**

This is one of the most common causes of congenital hearing loss. Some babies are born with hearing loss that is either an autosomal recessive or an autosomal dominant genetic condition (Martin, 1997). The autosomal recessive inheritance accounts for about 80% of genetic hearing losses while autosomal dominance accounts for about 50% of inherited hearing losses (Martin & Clark, 2000).

A syndrome is the combination of symptoms. Hearing loss might occur as only a part of other medical and physical disorders that occur together. A group of symptoms present can be considered diagnosing a particular disorder. Examples of syndromes that usually present hearing loss are Waardenburg; Klippel-Feil; Treacher-Collins etc. (Martin & Clark, 2000)

Hearing loss that occurs together with a syndrome or an inherited condition might be partial or complete.

- **Virus infections**

A virus infection results from teratogenic effects of an infection of the mother during the embryologic development of a fetus (during the first three months of pregnancy) (Berkow et al, 1992). Hearing losses presented are usually sensorineural with a greater loss at the higher frequencies (from 2000Hz upwards). The degree of the hearing loss can range between 20dB and a 100dB and can occur uni- or bilaterally (Berkow et al, 1992).

(a) Cytomegalovirus (CMV)

This type of herpes virus when contracted by the mother can be transmitted into the uterus (Berkow et al, 1992). This condition can be a result of the human immunodeficiency virus (HIV). 31% of infants infected have a serious hearing impairment, together with complications like microcephaly and mental retardation (Ladewig, London & Olds, 1998). Hearing impairment is a particular concern and

close monitoring beyond the neonatal period is thus of great importance. Another reason is that hearing loss usually has a delayed onset. The characteristics of this type of hearing loss are sensorineural, asymmetric and progressive (Berkow et al, 1992).

(b) Rubella (German Measles)

During the first three months (trimester) of pregnancy the cells of the ear and central nervous system differentiate rapidly. Congenital Rubella results from a primary maternal infection (Berkow et al, 1992). This viral infection can cross the placental barrier and may invade the developing inner ear (viral endolymphatic labyrinthitis), resulting in much destruction and a profound sensorineural hearing loss (mostly unilateral) (Berkow et al, 1992). Effects on the fetus may vary, but close observation is required. (See Table 2.2) Other common signs to look out for by the pediatrician are cardiac defects, pneumonia, otitis media, brain damage, blindness and mental retardation (Ladewig et al, 1998; Enkin, Keirse, Neilsen, Growther, Duley, Hadnett & Hofmeyr, 2000).

Table 2.2: Possibility for defects at different stages of the pregnancy

Duration of pregnancy	Percentage chance of defect
First gestational month	30% to 100% chance of defect
Second gestational month	20% to 50% chance of defect
Third gestational month	13% to 40% chance of defect

(Berkow et al, 1992)

(c) Syphilis

Syphilis is a venereal (contagious systemic) disease (Farber & Ballard, 2000). It can affect any tissue or vascular organ of the body and can be transmitted from an infected mother to the fetus via the placenta. There is a 60% to 80% chance of the fetus being infected (dependant on the stage of the mother's infection and the stage of pregnancy) (Berkow et al, 1992). Most babies are asymptomatic at birth. A progressive loss of hearing (uni-or bilateral) may appear at any stage from birth

onwards. Other related symptoms may be skin lesions, failure to thrive, meningitis, convulsions, optic atrophy, fissured skin lesions, etc. (Enkin et al, 2000).

(d) Human immunodeficiency virus (HIV)

This infection caused by a cytopathic human retrovirus, results in a continuously changing and progressive spectrum of immune deterioration and associated clinical conditions of which the end stage is acquired immune deficiency syndrome (AIDS) (Farber & Ballard, 2000). In short, it affects the immune system. During the course of the virus, it is characterized by a wide range of complications, and conditions that progress as the immune systems deteriorates. Mothers with HIV have a 50% chance of delivering a baby with the disease (Berkow et al, 1992). Congenital or perinatal infections like fever, meningitis and otitis media usually manifest during the first and second year of life (Enkin et al, 2000). Hearing is at risk mostly from opportunistic infections such as meningitis that are secondary to the disease or from the ototoxic drugs used as treatment (Berkow et al, 1992). With the increase of individuals who test positive for HIV, hearing loss, related to bacterial and viral infections may increase dramatically.

- **Incompatibility of the mother's and the baby's blood types**

A protein molecule, called the Rh-factor is found in the fetus blood, but is absent in the mother (Berkow et al, 1992; Martin & Clark, 2000). The mother's body produces antibodies for protection against the harmful effects of the Rh-factor. The count of antibodies increases with each succeeding pregnancy (Berkow et al, 1992; Martin & Clark, 2000). If a sufficient number of antibodies are produced the developing red blood cells of the fetus are damaged, to the extent that they cannot properly carry oxygen to the essential body parts, that includes the cochlea. The cochlea then becomes damaged, and a sensorineural hearing loss is present (Berkow et al, 1992).

- **Metabolic disturbances**

The most common example here is diabetes mellitus. Cardiac effects, congenital malformations of major organs including the ear, have been associated with the use of

oral hypoglymic agents in the first trimester. Because of the elevated glycosylated hemoglobin (HbAlc) concentrations at conception and during embryogenesis the baby's inner ear degenerates and leads to hearing loss at birth (Berkow et al, 1992).

- **Ototoxic drugs or agents**

This indicates agents or medication that is poisonous (Martin & Clark, 2000) to the ear. The ear especially is very sensitive to certain medication and chemical substances, for example neomycin, dihydrostreptomycin, nicotine and alcohol (Ladewig et al, 1998; Enkin et al, 2000). These medication or substances generally damage the outer hair cells of the cochlea at its basal end. The hearing nerve, more than any other, is highly susceptible to chemicals and medication, since it leads to the degeneration of the hair cells (Berkow et al, 1992). The hearing loss that may result is usually permanent, bilateral and symmetric (Martin & Clark, 2000).

B) Acquired hearing loss

(I) Perinatal causes (occur during the process of birth)

- **Anoxia**

A difficult, long or early birth can cut off the oxygen supply, which leads to oxygen deprivation to important cells. The ears are extremely susceptible to oxygen deprivation, especially the hair cell and the stria vascularis (Martin, 1997). The structures of the ear can degenerate and lead to sensorineural hearing loss (Berkow et al, 1992). Anoxia is a common cause of damage to the cochlea and the central nervous system.

- **Prematurity**

This classification refers to newborns who are born before 37 week gestation and weigh less than 1 500 grammes (Berkow et al, 1992). Prematurity is a matter of concern, since the level of organ system maturation is determined primarily by the infant's gestational age (Martin, 1997).

- **Trauma to the fetal head**

Fractures of the temporal bone may be due to the uterine contractions which can lead to damage to the brain and can even cause bleeding in the inner ear. A profound sensorineural hearing loss is present in 35% of the cases (Berkow et al, 1992; Martin, 1997).

(II) Postnatal causes (occur after birth)

- **Kernicterus**

This cause is usually evoked by maternal and fetal blood group incompatibility. After birth, bilirubin builds up in the newborn's circulation, and high levels can be deposited in the basal ganglia of the brain, resulting in Kernicterus. Clinical symptoms are poor feeding, flaccidity, seizures, apnea, sensorineural hearing loss and neonatal death (Berkow et al, 1992).

- **Purulent Labyrinthitis**

The invasion of the inner ear by a bacterium leads to the inflammation of the membranous labyrinth of the cochlea (Martin, 1997). This might be secondary to serous otitis media or purulent meningitis. During the case of serous otitis media, bacterial toxins may gain access to the inner ear through the membranes of the oval or round windows (Berkow et al, 1992). A sensorineural hearing loss may occur since the pressure of the fluid damages the inner ear structures (Jókay, Papp, Soós, Sziklai,& Dezsö 2001). If the inner ear itself is damaged or infected, a complete hearing loss may occur (Martin, 1997).

- **Ototoxins and medication**

Already discussed. (See A)

- **Infection of the baby/child**

Infections can lead to severe and complete hearing losses and have been identified as the causative factors in cochlear hearing loss. There is usually a sudden onset and the hearing loss can be uni-or bilateral (Martin, 1997).

(a) Measles

Measles is a highly contagious, acute disease and may cause a sudden, unilateral hearing loss. Other complications are fever, pneumonia, otitis media and encephalitis (Martin &Clark, 2000).

(b) **Mumps**

This acute, viral disease, causes painful enlargement of the salivary glands (Berkow et al, 1992). The highest incident occurs between the ages of 5 and 15 years. Mumps may occur along with a sudden, permanent, profound, unilateral sensorineural hearing loss (Martin & Clark, 2000).

(c) **Meningitis**

Inflammation of the meninges is due to a bacterial invasion, and usually occurs during the first four weeks of life (Berkow et al, 1992). This process includes the filling of the labyrinth with pus, and as the healing takes place, the membranes and other loosely attached structures of the labyrinth are replaced by bone. This happens to 2 out of 10 000 full-term babies and 2 out of 1000 low-birth-weight babies (Berkow et al, 1992). Due to the damage to the labyrinth a profound-complete, bilateral hearing loss can be found.

- **Fever**

During excessive fever, cells, including those of the cochlea, may become damaged (Martin, 1997).

- **Otitis media**

A factor that enjoys a lot of attention in terms of research and the fact that it may be related to the possible development of sensorineural hearing loss is otitis media (Papp, Rezes, Jókay & Sziklai, 2003; Mutlu, Osabasi, Metin, Basak & Erpek, 1998). Otitis media is the most common childhood disease of which the highest prevalence is between 75% and 95%, amongst neonates and children between the age of 6 and 12 months (Arick, 1995; Papp et al, 2003).

Several hypotheses have been made to confirm these associations. One of the reasons being that otitis media occurs along a continuum, and thus may result in a secondary pathology other than middle ear infection (Paparella, Schachern & Cureoglu, 2002). It is believed that the inner ear is vulnerable to chronic supportive otitis media (Papp, Rezes, Jókay & Sziklai, 2003). The proximity of the sensory cells to the potential source of harm (inflamed middle ear) may lead to more exposure, as reflected by the fact that sensory cells processing higher frequencies may be more seriously damaged (Papp, Rezes, Jókay & Sziklai, 2003).

Another theory on the relation between otitis media and a possible sensorineural hearing loss is that the toxins from bacteria in the middle ear may enter the inner ear by way of the round or oval window, or pus may enter the labyrinth from the middle ear or the meninges, the protective covers of the brain and the spinal cord (Papp, Rezes, Jókay & Sziklai, 2003).

According to the study being done by Cherow et al (1999) a large percentage of pediatricians and physicians are of the opinion that a hearing test by an audiologist is not necessary in the case of otitis media. Another issue that arises is that some pediatricians view the presence of otitis media primarily as a medical problem and not as a resulting hearing loss with possible devastating effects and consequences (White, 2002).

- **Idiopathic causes**

Other causes are usually of unknown (idiopathic) origin, and cause a sudden, unilateral hearing loss.

As seen in these paragraphs, this type of hearing loss can be caused by various factors, either congenital (occur by birth) or acquired (occur after birth as the result of pre- or postnatal factors) (Greinwald & Hartnick, 2002; Martin, 1997). Knowledge of patophysiology forms the basis for intelligent and successful prevention and intervention of a hearing impairment. Signs of these serious otologic (ear) diseases indicate the need for an evaluation by a specialist. "*Physicians should provide recommended hearing screening, not only during infancy but also through early childhood for those children at risk for a hearing loss, and for those demonstrating clinical signs of a possible loss.*" (American Academy of Pediatrics, 1999, p.1)

2.5 ASSESSMENT OF SENSORINEURAL HEARING LOSS

Modern technology has enabled audiologists to test the hearing of newborns and children with great accuracy. These tests use reflexive, behavioural and physiologic responses to auditory stimuli of controlled intensity. Infants with normal hearing show reaction to sound only a few hours after birth (Martin & Clark, 2000). Identification audiometry, a component of a hearing conservation program involves screening or testing individuals at risk for hearing impairment (Yasuhara, 1996).

A screening procedure is part of the preventative health care to detect disorders before clear symptoms of an impairment exists. The goal of screening is to identify an infant with a possible hearing impairment and to initiate treatment by six months of age (Yoshinaga-Itano, 2001). The purpose of identification is to differentiate between individuals with a hearing loss and those with normal hearing (Joint Committee on Infant Hearing, 2000).

Hearing impairment is difficult to detect, thus extraordinary efforts must be made to assess hearing during infancy and early childhood in order to prevent irreversible impairments (Joint Committee on Infant Hearing, 2000). During the past 25 years newborn hearing screening programmes were compiled to improve early identification of profound hearing loss and to ensure habilitation as soon as possible (Diefendorf, 1997). As a result of advances in understanding the auditory anatomy, physiology and advances in test equipment and procedures, audiologists are now able to differentiate between sensory and neural sites of lesions. The goal for all children with hearing loss should be early detection and the correct diagnosis, followed immediately by appropriate intervention (Guralnick, 2000). The underlying rationale is to decrease or even eliminate the effect that hearing loss has on speech, language, literacy, academic skills and social development (McGrath, 1998).

Successful prevention, identification, the correct intervention and possible medical treatment of mentioned causes are dependant on early screening programmes as well as the knowledge of professionals regarding hearing loss (Hugo & Pottas, 1997). In short, the essentials for an effective hearing-screening program are:

- Initial screening;
- Identification;
- Evaluation;
- Tracking and follow-up; and
- Intervention.

Universal hearing screening is necessary. The high-risk registry, for instance, is a procedure that can detect almost half of newborns with hearing loss (Lutherman & Kurtzer-White, 1999). Unfortunately if only the infants and children who fall in a high-risk category are being screened, 30-50% of infants and children with hearing loss will be missed (Oudehuys-Murphy, Van Straaten, Bholasingh & Van Zanten, 1997). Therefore, it is important for infants to undergo ongoing attempts at being diagnosed within the first three months of life. A combination of tests must be used as a cross-check principle in order to make the correct identification and diagnosis of neonatal hearing impairment.

Screening of infants' hearing before they leave the hospital or clinic, is part of the law in most states of the USA (Hall, 2000). Impressive gains in the development of instrumentation and test protocols have been made to identify hearing loss in infancy, yet many communities still do not have any hearing screening programmes or they are unaware of these tests (Hall, 2000). As soon as hearing loss is suspected, audiologic assessment should be undertaken. Electrophysiologic testing includes auditory brainstem response (ABR), immittance measurements and oto-acoustic emissions (OAE). The use of these objective techniques does not require the patient's behavioral responses and is ideal for infants (Joint Committee on Infant Hearing, 2000).

The two electrophysiological techniques which are mostly used during screening of hearing, is the auditory brainstem response (ABR) and oto-acoustic emissions (OAE) (Hall, 2000). Both tests are recordings of the physiological activity that underlies normal auditory function, and are easily applied to infants and children and both correlate with the degree of peripheral hearing sensitivity (Joint Committee on Infant Hearing, 2000). These tests are also capable of detecting unilateral or bilateral hearing losses of approximately 30dB and above (Diefendorf, 1997).

A screening procedure should use objective criteria to define the method for a technically correct screening test and to set a guideline for a "pass versus refer" outcome. Thereby the need and driving force for automated testing in screening technology is - (a) control and (b) monitoring of testing. In short the number of babies who require testing far exceeds highly skilled professionals available to perform the conventional tests. Automated technology for ABR and OAE is at present highly recommended and favored for using. The reasons being:

- Less skilled personnel can now do the above screening.
- It test signals and objectively analyses data, thus being efficient and cost-effective.
- Maintain accurate, consistent and effective hearing outcomes (Katz, 2002).

This automated system has proven to be a valid and reliable way to screen babies and children successfully (Katz, 2002).

2.5.1 Automated brainstem response (ABR)

ABR testing is the cornerstone of objective audiometry in infants and children (Tomaski & Grundfast, 1999). ABR reflects the activity of the cochlea, auditory nerve and auditory brainstem pathways. Using ABR, frequencies between 1 and 4 kHz can be obtained from neonates, young children, or uncooperative children (Tomaski & Grundfast, 1999). ABR is well suited for estimating hearing levels in infants, and is unaltered by sleep or sedation (Greinwald & Hartnick, 2002). When appropriately used, premature infants as young as 30 weeks of gestational age can be tested at low stimulus intensity levels. ABR is highly accurate in detecting sensorineural hearing losses in infants in excess of 30dB HL (Martin & Clark, 2000).

2.5.2 Oto-acoustic emissions (OAE)

OAE screening is an objective and quick procedure and tests a wide frequency range simultaneously (Diez-Delgado Rubio, Espin Galvez, Lendinez Molinos, Ortega Montes, Arcos Martinez & Lopez Munoz, 2002).. Hearing screening programmes based on transient-evoked otoacoustic emissions (TEOAE) can expect to identify hearing impairment in approximately 80% of infants screened (Jakubikova, Kabatova & Zavodna, 2003). It is particularly useful in screening for peripheral auditory dysfunction, especially in infants, since OAE's tend to be immeasurable in ears with greater than 25-30 dB of peripheral hearing loss (Martin & Clark, 2000). OAE's are sensitive to outer hair cell dysfunction, detecting cochlear (inner ear) hearing loss. The disadvantage of OAE is that it is unable to detect neural (eight cranial nerve or auditory brainstem pathway) dysfunction, and cannot be used when the middle ears are abnormal (Martin & Clark, 2000).

2.5.3 Auditory steady-state evoked potentials (ASSEP)

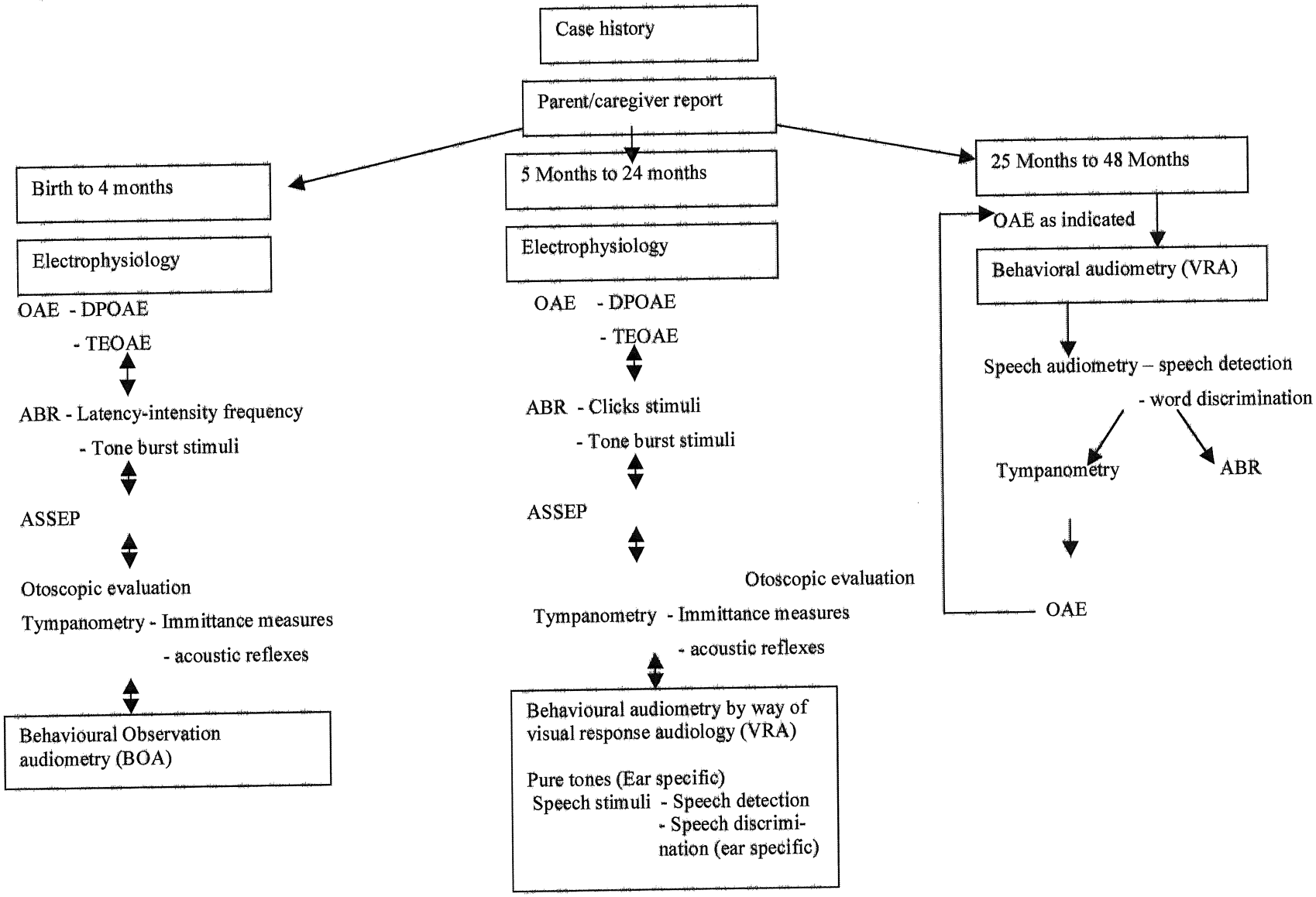
Another test that has been proved reliable during neonatal hearing screening is the auditory steady-state evoked potential (ASSEP) (Rance & Briggs, 2002). ASSEPs are continuous scalp-recorded potentials that arise in response to periodically time-varying

stimuli such as amplitude- and frequency-modulated tones. Rates around 70 to 100Hz have been most effective for infant testing. This procedure provides an objective, frequency specific assessment of hearing that can be used to confirm results obtained behaviourally, particularly in cases of significant sensorineural hearing loss (Sininger & Cone-Wesson, 2002). Studies proved that the ASSEP-thresholds obtained can be used as the basis for hearing-aid fittings for infants and children with sensorineural hearing loss and that all babies tested through this process, were successfully amplified within one month after evaluation (mean age is 4 months) (Rance & Briggs, 2002).

The basic hearing assessment/evaluation through a test battery must not be underestimated (Hall, 2000). The test battery is the foundation that leads to responsible and effective auditory assessment. The results of all given tests provide information on the auditory functions; normal and abnormal. This information is critical during the management planning after the diagnostic phase is completed.

The test battery approach is based on the cross-check principle. This implies that the results of a single test are never accepted and interpreted in isolation as conclusive proof of the nature or the site of the hearing impairment without support from various objective assessments (e.g. ABR) (Katz, 2002). The reason for applying the cross-check principle during hearing assessment is because of the fact that the results of the assessments will have a direct impact on the infant's/child's life. In figure 2.3 a flowchart outlines the various assessments used with infants and young children.

Figure 2.3: Pediatric audiometric flowchart



2.6 INTERVENTION METHODS MOST SUCCESSFUL FOR SENSORINEURAL HEARING LOSS

Intervention services are designed to help individuals overcome the challenges posed by their disability. These services are designed to provide an individual with the most appropriate technological support that will help maximize the use of residual auditory abilities and ensure the best possible hearing for the development of oral language and speech (Boothroyd, 1998). Early intervention is secondary prevention. The goal is to prevent, evaluate and provide relevant treatment in order to minimize potential negative outcomes (Fair & Louw, 1999).

The majority of hearing losses are caused by damage to the inner-ear structures. When the hair cells in the cochlea are damaged, the transferring of sound to the rest of the structures is obstructed. The problem occurring with a sensorineural hearing loss is that the damage to the inner ear is mostly not medically treatable (Martin, 1997). The most effective way of intervention is usually the use of hearing aids or sensory aids (Pediatric Working Group of the Conference on Amplification for Children with Auditory Deficits, 1996). Prompt initial probatory treatment with a conventional hearing aid is necessary to take advantage of the sensitive phase of development and maturation of the auditory pathways (Martin, 1997).

An increasing number of very young children with marked developmental delays, especially in speech and language, are being referred to the audiologist. This trend reflects a growing awareness within the professional community and amongst parents about the importance of early identification and habilitation of infants/children with a hearing impairment. Several reasons exist for providing amplification as early as possible:

- To avoid possible effects of sensory deprivation, which refers to both the physiological and psychological aspects of development.
- The enhancement of language development during critical periods of readiness.

The fitting of the hearing aid is the first step in the intervention process. Hearing aids provide the hearing-impaired infant/child with optimal use of the residual hearing so that speech and language milestones can be achieved at appropriate age levels.

Hearing aids increase the intensity of environmental sounds and may modify the sound spectrum and tailor/amplify it to the patient's particular pattern of hearing loss at higher or lower frequencies. A hearing aid's secondary function is to increase the quality of life of a hearing-impaired person. It cannot provide normal hearing but increases the ability to communicate; makes a person an active participant in life because the person is able to experience sounds of every day life. Every hearing aid is selected and adapted to the individual's needs. Unfortunately it is true that not every person with a hearing loss receives the same benefit from a hearing aid because it depends on the cause, nature and degree of the hearing loss. The fitting of an appropriate hearing aid is essential to ensure an optimally aided performance. Assessment of the hearing aid fitting is critical, as it provides valuable information regarding the patient's ability to make use of auditory cues in speech, and therefore aids in the prediction of potential cochlear implant performance.
Current results in young infants and children with a hearing impairment have exceeded the expectations of professionals. Technology is continually improving in terms of better speech processing paradigms (Northern & Downs, 2002).

Hearing aid optimization is an essential part of the pre-operative assessment for cochlear implant patients. The patient's hearing aid fitting must be rigorously evaluated before an informed decision van be made as to whether a cochlear implant would be of more benefit compared to a standard hearing aid fitting. A cochlear implant is a useful procedure for those with severe and profound sensorineural hearing loss but a trial period with hearing aids must first be followed (Al-Shaikh, Zakzouk, Metwalli, Dasugi & Metwalli, 2002).

A device that has developed in the last 20 years to an aid of high standard and safety for even the pediatric population with a profound to severe sensorineural hearing loss is the cochlear implant (Arts, Garber & Zwolan, 2002; Gysin, Papsin, Daya & Nedzelski, 2000). A cochlear implant is an electronic device that, through the strategic placement of electrodes in the cochlea, provides direct stimulation of the

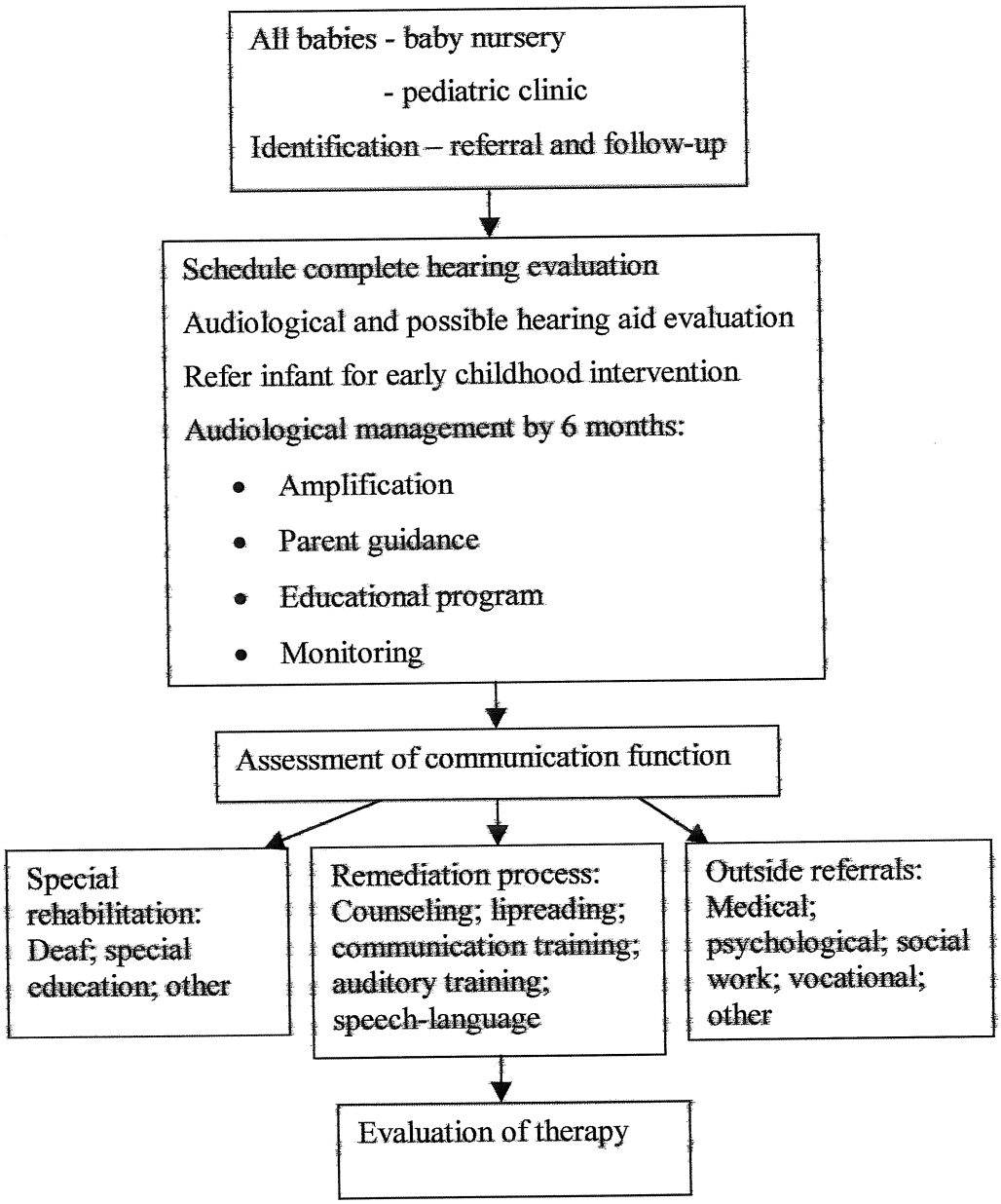
auditory nerve (Martin, 1997). It consists of a surgically implanted receiver and electrode array connected to an external transmitter coil, microphone and speech processor. A cochlear implant provides meaningful auditory information to the person that was implanted, which increases the functional perception of speech and communicative skills (Franz, 2002). Cochlear implants are now firmly established as effective options in the habilitation and rehabilitation of individuals with a severe and profound hearing impairment who do not benefit from traditional personal hearing aids when maximum stimulation of the auditory system is desired.

Children with cochlear implants show continuous improvement in their speech perception and use of oral language. The younger the child was when he/she received an implant, the greater the improvement and the better their oral language (Family Practice News, 2002). The importance of early referral to a cochlear implant clinic cannot be underestimated. An early referral allows time to plan the approach to be followed for intervention. An additional important task at the time of referral is to obtain and collate existing medical and audiological records of the child. The positive benefits of implantation for both pre- and postlingually profoundly deaf children are beyond doubt. Such benefits have been shown to include increased capacity for spoken language acquisition. The improved auditory information following implantation further improves the ability of the individual to learn, which has rendered mainstreaming a more attainable social and academic goal.

The goal of amplification (both hearing aids and cochlear implants) is to give infants and children maximum accessibility to sound and speech. The period since the confirmation of the hearing loss until the fitting of the amplification system must be minimal (Arehart, Yoshinaga-Itano, Thomson, Gabbart & Stredler Brown, 1998). Because early intervention is a prerequisite for successful speech- and language development, infants and children must be referred to an Intervention Programme, like the Cochlear Implant Team, for an assessment as soon as his/her hearing loss is diagnosed (Sirimanna, 2001; Nucleus Cochlear Implant, 2000). With professionals becoming more confident in implanting younger children and realizing the benefits of early implantation, especially for speech and language development, there is a growing demand for referring infants as early as possible to the implant programmes. Providing amplification is only the beginning of the process of habilitation.

Technology growth will continue to result in more possible options for children. Unfortunately, without appropriate audiological services and technological support, these children will not have the opportunity to access information and participate in learning, as well as they deserve. Without intervention children with a bilateral sensorineural hearing loss of approximately 50 dB may have an academic delay of as much as 3 years (Lewis, 2000). An interdisciplinary team approach is extremely important in the process of rehabilitation (Sirimanna, 2001). (Figure 2.4 provides a summary of the rehabilitation process from screening to intervention.)

Figure 2.4: Flow chart of the rehabilitative audiology process



Source: Hall (2000); Finitzo & Crumley (1999)

2.7 THE IMPORTANCE OF EARLY DIAGNOSIS AND REHABILITATION

The most crucial period for the development of phonology (speech) is during the first twelve months, while language development must be developed during the first three years of life. Hearing is a key component in the infant's development of speech, language and cognition, therefore early detection of an infant's hearing loss is critically important (Boothroyd, 1998).

The importance of early diagnosis of hearing loss is that the type and degree of the hearing impairment can be measured, hearing aids can be fitted and parents are put in touch with the correct and relevant professional personnel (Boothroyd, 1998). The primary objective of early diagnosis of hearing impairment in babies/children should be followed by intervention (Purdy, 2000). Intervention means a goal-orientated and time-limited process aimed at enabling the baby or child to reach an optimum functioning level. It can involve measures intended to compensate for a loss of function or a functional limitation or to prevent and/or reduce the impact of impairment on their development (Boothroyd, 1998).

The emotional and psychological development, as well a communication development of a baby/child is greatly dependent on normal hearing. A baby/child with a hearing loss foregoes all of this. His/her needs are similar to the baby/child with normal hearing, as is the need to make his/her needs known. The link between infant hearing loss and speech and language deficits is well established. The baby/child is excluded from normal everyday conversations and therefore has difficulty in understanding language, in expressing him-/herself, in forming words and in all the other areas of language (Boothroyd, 1998). These difficulties are due to the fact that a part of the ear or nervous system is not functioning and his/her brain is deprived of information provided by listening under normal conditions (Purdy, 2000). This has a profound impact on speech and language development, since the child is not able to hear adequately during the most critical period of brain growth and speech- and language development. These inabilities lead to frustration, emotional disturbances, school and learning problems as well as possible behavioural problems.

Appropriate pediatric care may help other professional members to identify and diagnose a hearing loss at an earlier age, thereby preventing some of the influences of a hearing impairment. The astounding effects of early diagnosis and intervention confirm this statement. Studies have shown that hearing-impaired children who are amplified and who receive educational support within first 6 months of their lifes have significantly greater potential for speech and language development than do children who receive intervention after six months (Rance & Briggs, 2002; Yoshinaga-Itano, 2001).

Once a child has been diagnosed by a pediatrician, audiologist or ear-nose and throat specialist as having a sensorineural hearing loss, immediate audiological and educational management should be instituted. Children with a newly identified sensorineural hearing loss should be assessed by an audiologist, preferably every 3 months during the first year, every 6 months during their preschool years and at least once a year while in school (Tomaski & Grundfast, 1999). Early audiological and educational treatment is crucial to children's later performance in the formal educational arena (Tomaski & Grundfast, 1999).

Pediatricians' ability to diagnose and treat hearing loss is related to suspicion, physical examination, evaluation of the audiogram, knowledge of hearing loss and common disorders and armamentarium of where and how to make a timely referral for further workup and expeditious treatment (Tomaski & Grundfast, 1999).

A useful eponym for pediatricians to approach children with a suspected hearing loss is as follows:

ALOUD:

A: Ask about family history.

L: Look for physical findings known to be present in some types of hearing loss.

O: Obtain appropriate assessments to include an audiogram, temporal bone CT scan and other studies that may be indicative of a hearing loss.

U: Use consultants to include otolaringologists and an audiologist experienced in dealing with hearing loss.

D: Determine an appropriate plan of action for children newly diagnosed with a hearing loss. (Tomaski & Grundfast, 1999, p.42).

The above-mentioned is relevant and well functioning in developed countries but unfortunately these standards are still not met in South Africa (a developing country). The reasons have been discussed in chapter one as well as chapter 2, paragraph 2.8.

Infants and children that have been identified with hearing problems through the use of these hearing tests must be referred as soon as possible (before 3 months of age) for a complete audiological evaluation (Joint Committee on Infant Hearing, 2000). The relationship between hearing, oral language development, cognitive development and psychosocial development defines the need for early diagnosis and intervention and teamwork (Purdy, 2000). The importance of early diagnosis and intervention cannot be overemphasized in order to make it a reality. During this evaluation the presence of a hearing loss is confirmed, as well as the nature, degree and possible cause of the hearing loss is discussed (Shonkoff & Meisels, 2000). The diagnosis of a hearing loss is only the beginning of a complex intervention process that varies on a continuum of medical intervention to speech and language intervention (Purdy, 2000; Katz, 2002).

In conclusion it can be said that the core of problems surrounding a hearing loss is not only the defective functioning of the hearing mechanisms, but it includes the whole person. This statement is confirmed by the words of Helen Keller: " *The problems of deafness are deeper and more complex, if not more important, than those of blindness. Deafness is a much worse misfortune. For it means the loss of the most vital stimulus – the sound of the voice that brings language, sets thoughts astir and keep us in the intellectual company of man*" (Whetnall, 1946).

2.8 THE IMPORTANCE OF TEAMWORK

Because of the complex nature of most abnormalities which requires high standards and different levels of specialization, the success of early hearing detection and intervention is dependant on the professional people who work together as a coordinated team (Sraka & Bricker, 1996; Guralnick, 1997). Team approaches during the intervention of infants/children with special needs are being emphasized throughout the literature, since it constitutes a critical component of the larger system of services and support for patients and their families, (Bailey, 1996; Ryan-Vincek,

Tuesday-Heathfield & Lamorey, 1995). The approach that is generally advocated in infants and children with hearing impairment is the interdisciplinary approach (Rossetti, 1996). This approach is described as the conscious attempt of different disciplines to exchange and combine information, skills and knowledge in order for a practical approach to the problems that exist with a hearing impairment (Bailey, 1996 and Rossetti, 1996). (See figure 2.5)

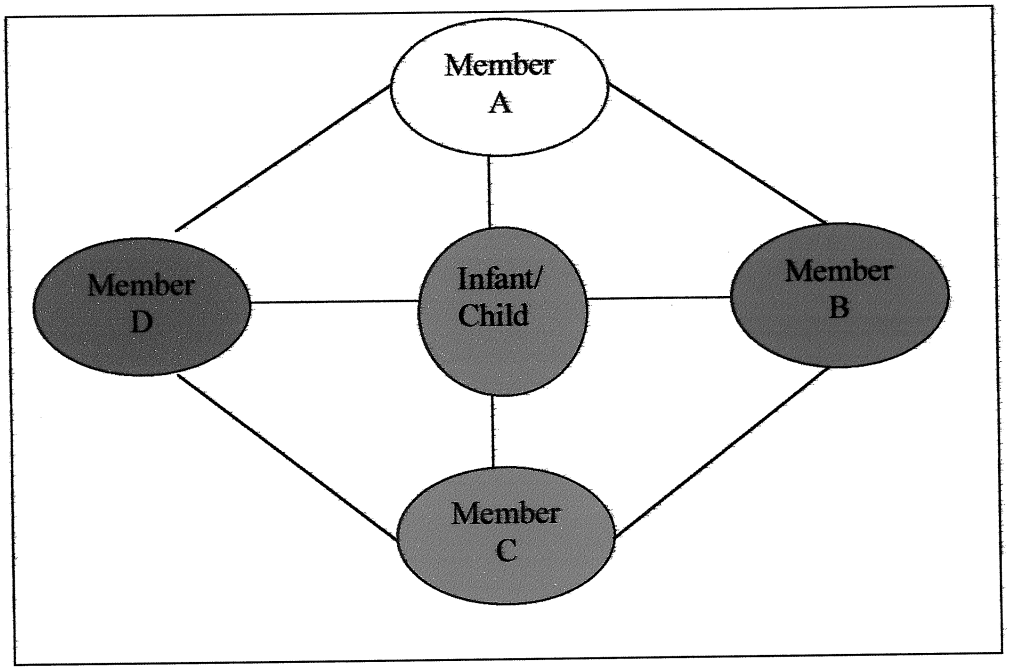
Members of the different professions work together in order to reach a joint goal and to share the responsibility regarding intervention and results (Halper, 1993). This is because the intervention for a patient with a sensorineural hearing loss, involves the combined efforts of audiology, medicine, education and speech-language pathology. No professional has all the knowledge and skills which is required for every patient's problem. The importance of gaining insights from many disciplines and exchanging one's knowledge with respect to a child's development is crucial for success in the health sector (Klein & Moses, 1994). Teamwork leads to a more effective intervention programme as well as a more positive attitude and greater participation amongst members (Krueger, 1990).

A need exists for expanded in-service educational opportunities. The explosion of information created a need for continuing education. Professionals from other disciplines need in-service education, both to establish a better understanding of hearing impairment and its implications and to enable them to assist with the identification of undetected hearing losses (White, 2002). Physicians are increasingly dependent upon one another for the highest quality care for their patients (White, 2002). With every discipline there are some tasks that cannot easily be modified. Yet many activities are not only amenable to broader participation but are greatly enhanced by the involvement of other professionals (Hall, 2002).

To develop an effective working relationship with other professionals, one must know what their roles are in relation to the services that one provides (Rossetti, 1996). Teamwork between the audiologist and the pediatrician is important not only for documenting and monitoring hearing losses, but also for determining the implications of hearing losses and the effects of various management strategies (De Conde, Benson & Seaton, 1997). In the above-mentioned paragraph the importance of coordinated

teamwork is emphasized. Therefore it is important to provide an overview on the audiologist and the pediatrician's expertise and the importance of coordinating their skills when an infant/child with a sensorineural hearing loss is involved.

Figure 2.5: The interdisciplinary model



Source: Rossetti (1996) & Roodt (1994)

2.8.1 The role of the pediatrician

Most pediatricians are the primary care practitioners who direct their services in keeping babies and children well (Finitzo & Crumley, 1999). This is done through illness prevention, early detection, treatment and provision of guidance and support to the parents (White, 2002). Pediatricians are especially concerned with any disturbances of the health or the orderly growth and development of babies and children (Finitzo & Crumley, 1999). Thus it is important for pediatricians to be sufficiently familiar with the normal patterns and milestones of babies and children in order to recognize deviation from the normal ranges that might prevent them from reaching their full potential.

The pediatrician is the front line and has an important role to fulfill in terms of the diagnosis, referral and treatment of infants/children with a hearing impairment.

(Tomaski & Grundfast, 1999). The pediatrician should review the infant's history for the presence of risk indicators that require monitoring for delayed onset and/or progressive hearing loss and should ensure periodic audiological evaluation for children at risk (Tomaski & Grundfast, 1999). The pediatrician's ability to communicate effectively to parents is particularly important in order for satisfactory care of babies and children in the first five years of their life. Pediatricians examine infants at month 1, 3, 6, 9, 18, and at 3 years of age in order to detect a hearing loss as early as possible (Yasuhara, 1996). Another reason for the frequent examinations is that 30% to 40% of children with confirmed hearing loss will demonstrate developmental delays or other disabilities (Family Practice News, 2000).

Pediatricians are asked to evaluate and treat an ever-increasing number of both routine and complex medical problems. It is important that the pediatrician be knowledgeable in a broad spectrum of routine medical problems, yet also recognize when the consultation of another specialist is required (Cherow et al, 1999). Through the necessary concerns, knowledge of the family history, a physical examination, evaluating the audiogram and efficient knowledge of the causes and nature of a hearing loss, the pediatrician can make a timely diagnosis and appropriate referrals (Tomaski & Grundfast, 1999). The assessment by the Cochlear Implant Team as part of early intervention for infants and children with a confirmed hearing loss usually occurs after a diagnostic assessment and the correct referral of professionals (Bagnato & Hofkosh, 1990 and Oski, DeAngelis, Feigin & Warshaw, 1990). Since pediatricians are usually in early contact with most infants and children with a high- or confirmed risk factor, they are also the first professionals to evaluate and treat this population. This puts the pediatrician in the ideal position to identify, counsel and make the necessary referrals of infants and children who may be considered candidates for amplification devices like hearing aids or cochlear implants (Wachtel & Compart, 1996).

Pediatricians must be familiar with many types of diseases that might be of concern regarding speech, language and auditory development. This serves as reference for the decision-making of whether the patient is going to be treated in the primary care environment and whether he/she should be referred (Cherow et al, 1999). Referrals to other medical specialists are important for additional help in the diagnosis and

treatment of a child with a hearing impairment. This avoids delay in detection of significant hearing impairment and the associated lack of essential skills in speech, language and social interaction (Tomaski & Grundfast, 1999). No child is too young for a hearing assessment.

2.8.2 Role of the audiologist

As experts in identification, evaluation, and auditory habilitation of infants who are hard- of-hearing and deaf, audiologists are involved in every component of the EHDI process (Knightly, 1994). For the hearing-screening component, audiologists provide program development, management, quality assessment, service coordination, and effective transition to evaluation, habilitative and intervention services (Knightly, 1994). For the follow up component, audiologists provide comprehensive audiological assessment to confirm the existence of the hearing loss, evaluate the infant for candidacy for amplification and other sensory devices and assistive technology, and ensure prompt referral to early intervention programs. Specialized audiologists also participate in the assessment of candidacy for cochlear implants (Family Practice News, 2000).

Audiologists specialize in the prevention, identification and non-medical management of hearing loss (Knightly, 1994). The goal of pediatric audiology is to lessen the impact of hearing loss on language acquisition and consequently on the social, emotional, educational and ultimately vocational status of the child (Knightly, 1994). This goal can only be achieved through early identification and prompt intervention. The audiologist possesses the skills and technology necessary to identify hearing loss and initiate management as early as the newborn status (Hall, 2002). Efficiency of the technology used by the audiologist is parallel to their skills and attitudes. Technology does not function in isolation, especially not in the healthcare system (Masterson, Wynne, Kuster, & Stierwalt, 1999). Table 2.3 provides an overview on the role of the audiologist.

Table 2.3: Functions of the audiologist

Function/role	Definition/description
• Identification	Planning, initiation, implementation, management and monitoring of any identification programmes for hearing problems.
• Assessment	All activities which are focused on the description (non-medical diagnosis) of people with a hearing loss as well as the decision-making regarding intervention. The goal: • To determine the presence, nature and degree of the impairment. • To describe the impairment. • To identify steps relevant to intervention. • To formulate a prognosis for progress. • To convey the implications of the impairment to the parents.
• Intervention	Activities that focus on the modification of the hearing impairment through the following strategies: • To provide hearing aids, cochlear implant as part of an aural rehabilitation programme. • To provide strategies for auditory stimulation for language acquisition. • To obtain compensation for irreversible hearing impairment. • Change the attitudes regarding hearing loss in order to improve

	hearing and communication. • To educate the parents to help them communicating effectively with their child.
• Counseling	Services being provided to the individual with the hearing loss and the family in order to decrease the problems that are associated with, or is the result of the hearing loss.
• Consultation	Activities that focus on service delivery to other professionals who are involved with the individual with a hearing impairment.
• Education	Planning, implementation and assessment of educational programs regarding aspects such as normal hearing and hearing disorders, professional roles in terms of knowledge, skills and attitudes.

(Hugo & Pottas, 1997; Knightly, 1994)

Referral to a skilled audiologist for an ongoing management of a child with a hearing loss is essential. By developing a positive relationship with pediatricians, the audiologist can best address the client's needs as well as to secure a stable referral base (White, 2002). Pediatricians, together with other professional personnel, play an important role in the linking of families to the relevant healthcare facilities (Cherow et al., 1999).

Early detection and intervention are best done through an interdisciplinary team approach (Tomaski & Grundfast, 1999). The audiologist can assist the pediatrician in making the diagnosis of a hearing loss. It is important to document whether the hearing loss is stable or progressive, and ongoing audiological follow-up is imperative. The pediatrician needs the diagnostic, treatment and support services provided by audiologists (White, 2002). It is important that the audiologist has a proactive role in seeking opportunities to inform pediatricians of the value and importance of hearing assessments (White, 2002).

2.8.3 The role of the parents or caretakers

The roles of the professionals have shifted to acquiring information from the parents about their needs, strengths, resources and intervention priorities (Kricos, 1993). By making parents partners in the effort to facilitate the child's development the parents are empowered to master the problem areas in their lives. The family-centered perspective, enables each unique family to capitalize on its strengths in meeting the challenge of raising a hearing-impaired child (Kricos, 1993).

Parental involvement during the habilitation process of their hearing-impaired children is crucial to the success with amplification. The child's needs are best met by meeting family needs. It is the parents, not the professionals, who must make the decisions regarding the child's habilitation because they must accept and take the ultimate responsibility (Northern & Downs, 2002).

Parents should be considered as expert informants about the child's communicative competence. Caregivers have opportunities to observe and interact with the child far more frequently and in a more familiar and emotionally secure situation than professionals (Watson, Crais & Layton, 2000).

The family plays a significant role in providing the context in which the hearing-impaired child will grow (Kricos, 1993). The hearing-impaired child's chances for successful communication, academic achievement and life satisfaction are enhanced when the parents can accept the child and are dedicated to maximizing his/her potential (Kricos, 1993).

With new technology at hand, a greater sensitivity and understanding of family systems, culture beliefs and diversification of children and their families, audiologists and pediatricians can make a critical difference in the lives of children with hearing loss (Boswell, 2001). Joint assertation of quality indicators among all EHDI stakeholders avoids future misunderstandings.

2.9 CONTRIBUTING FACTORS LEADING TO THE LATE DIAGNOSIS AND INTERVENTION OF HEARING LOSS IN SOUTH AFRICA

In the less developed countries, like South Africa, child development and health is dominated by problems of nutritional deficiency, infectious diseases, diversity and multi-cultural characteristics of the population (Kane-Berman, Henderson & De Souza, 2001; Morley, 1985). Culture includes the total of customs, beliefs, attitudes, values, goals, laws, tradition and moral codes of the people (Morley, 1985). This might include, for instance, the different group views on medical support, hearing loss, hearing aids and cochlear implants as well as their ignorance on technological progress. According to the Medical Research Council (MRC), between 75% and 80% of the South African population rather consult traditional healers than educated medical personnel (Kane-Berman, Henderson & De Souza, 2001). The high incidence of illiteracy, poverty and unemployment contribute to the fact that a great deal of the South African population is deprived of general necessities namely water, electricity, sanitation, housing and transport. All these aspects contribute to unsatisfactory attendance of health services (Kane-Berman, Henderson & De Souza, 2001).

Negative attitudes towards children with hearing loss and the reasons for the perception of such attitudes within a community can be divided into three categories:

- Socio-economic conditions: puts additional strain on the family, making the deaf family member an undesirable burden;
- Lack of understanding concerning the nature of disability; and
- Beliefs about the etiology of the disability (Stephens et al., 2000).

The above reasons have a tendency to lead to the concealment of the condition of the child. The overriding attitude toward these children is one of pity, sympathy and even negative attitudes (Stephens et al., 2000). Some parents will initially either ignore the impairment or take their children to traditional healers before accepting rehabilitative intervention from professionals. This would appear to stem in part from traditional attitudes and in part from the relatively high cost of hearing aids in the country relative to outcomes (Stephens et al., 2000).

Parents of infants/children diagnosed with hearing loss may experience such intense grief, losing the hopes, dreams and aspirations they had for their child (Kricos, 1993). This might result in the non-acceptance or denial of their child's condition. Hearing impairment is relatively easy to deny since it is invisible. Parents might deny the hearing impairment in a number of ways. One being that they might reject the diagnosis or they will start 'shopping' around (Kricos, 1993). This meaning that they will take their child to a number of different professionals and clinics in the hope of a more positive outcome. Many parents of infants and children with a hearing impairment have thus not sought professional help or followed advice given to them at the time being (Shimon, 1992). This leads to a time consuming process where important time goes by for the hearing impaired baby/child.

Another concern is the unavailability of health services. According to the director of labour relations, Department of Health, professor Rachel Gumbi, only one doctor is available for every 800 people in the city areas, and only one doctor for 26 000 people in the country (rural) areas (Kane-Berman et al., 2001). There appears to be a shortage of medical personnel in South Africa. A shortage of pediatricians, especially in the country (rural) areas exists. The realization of early diagnosis of infants and children in South Africa with hearing loss causes concern because of the fact that South Africa is a developing country with limited economical growth (Kane-Berman et al., 2001).

As discussed in chapter 1, hearing loss is a 'disguised' impairment (Joint Committee on Infant Hearing, 2000). A baby/child with hearing loss does not look any different and one cannot detect a hearing loss by looking into the patient's ear (Hampton, 1999). To identify a possible loss of hearing within 48 hours after birth is ideal. However, a lack of infrastructure at hospitals and clinics as well as a lack of knowledge has deprived many babies and children of the opportunity of being diagnosed early and of receiving adequate rehabilitation (Bhengu, 2002). It is important that professionals, who do not deal with hearing impairment on a daily basis keep well informed of the current changes and developments in the assessment and intervention of hearing impairment (Hall, 2000). The technology is evolving at a rapid pace and professionals must keep up to date with current developments. A deficiency in knowledge can lead to problems during the initial phases of referral to the relevant professional instance.

Tremendous technological advances regarding amplification in the hearing aid as well as cochlear implants have been made over the past decades. Technology regarding amplification such as cochlear implants is a new and highly specialized field. Improvements in technology lead to the rapid development and broadening of the selection criteria for candidates who qualify for cochlear implants as well as other forms of amplification (Lenarz, 1998). (See Table 2.4 and 2.5 for the changes in the selection criteria for infants and children for cochlear implants).

The most important intervention to help infants/children with hearing impairments is amplification – hearing aids. The ideal is to fit hearing aids on the pediatric patients as soon as hearing loss is confirmed. The process of selecting the correct amplification is a complex one though and several problems may arise, especially in a developing country like South Africa. The selection of children's hearing aids requires special consideration. The hearing aids must have maximum flexibility, so that significant ranges of adjustments and modifications can be made without the need to purchase new hearing aids as the child's needs change. Unfortunately, higher technology in hearing aids costs more, for example the costs of batteries, new ear molds that need to be made every six months while the child is growing. It also requires accurate individual threshold measurements and may depend on a remote control unit to operate the hearing aid. A child's hearing aid must also be durable and an insurance protection plan for extended warranty or loss or damage is required. Since poverty is a problem in South Africa, there are a lot of parents, clinics and hospitals that will not be able to endure the high costs.

The reason for the focus being on hearing aids, is because cochlear implants are very expensive and few parents will be able to carry the cost for the operation. It is therefore assumed that the majority of hearing-impaired children in South Africa will receive hearing aids prior to cochlear implants. Another reason being cochlear implantation can only be considered after the infant/child was fitted with appropriately fitted hearing aids and little or no benefit from this amplification system was indicated.

A major problem encountered in the initial stages of referral for cochlear implants is the amount of inaccurate information about implants in the community. This arises

from a number of different sources including groups who are apposed to implants in children, media information which is exaggerated or incomplete, opinions based on subjective information, or even professionals with little knowledge of this field (Stephens et al., 2000).

In terms of early identification, diagnosis, intervention and referral especially for cochlear implants, the following factors may be of importance:

- Rapid advance of medical technology,
- Involvement of other relevant professional areas (ie. Ear-Nose-and-Throat Specialist, Speech-Language Therapist, Audiologist, etc.).
- The devices that are currently used: In the UK once a referral is made, the patient falls under the care of the World of Cochlear Implants and the referring agent no longer needs to be concerned. These professionals do not have the opportunity to see the progress being made (Novy, 2003).

Table 2.4 refers to the history of the selection criteria for infants and children in order to receive a cochlear implant, while table 2.5 gives the current selection criteria.

Table 2.4: An overview of the selection criteria for candidates from 1985 – 2000 (Nucleus Coclear Implant System, 2000)

	Infants and children
1985	• The FDA (Food and Drug Association) does not recommend Cochlear Implants for infants/children.
1990	• The FDA approves implantation candidates that are 2 years and older in the following scenarios: ➢ Severe to profound bilateral sensorineural hearing loss. ➢ Little or no benefits received by a hearing aid. ➢ The candidate as well as the family must be motivated and must have realistic expectations.
1995	• The criteria of 1990 stay unchanged.
1998	• 18 Months or older. • 3-6 months hearing aid trial period. • Less than 20% on the MLNT or the LNT tests.
2000	• 12 months and older (FDA approval). • 3-6 months hearing aid trial period. • Lack of improvement of the auditory skills or less than 30% on the MLNT or the LNT tests.

Table 2.5: The current selection criteria according to the Cochlear Corporation (Nucleus Cochlear Implant System: General Selection Criteria; 2000)

Pediatrics and children candidacy population
❖ Bilateral, severe to profound sensorineural hearing loss. ❖ No minimal age for referral is necessary (Hall & Meuller; 1998). ❖ Receives no to little useful benefit from the appropriate hearing aids: • Younger than 5 years: does not develop the basic auditory skills e.g. localization. • Older than 5 years: 50% or less on the open set word recognition. • Amplified thresholds are outside the speech spectrum at 2kHz +. ❖ No medical contra-indications are apparent e.g. cochlear ossification, hearing loss because of a retro-cochlear or a central auditory scarring, chronic otitis media, absence of cochlear development. ❖ The family and the candidate must be motivated and need to show realistic expectations.

2.10 SUMMARY

The most important information in this chapter is that the hearing system is a complex one and that there are many aspects surrounding hearing loss to be taken into account, especially sensorineural hearing loss. These factors are interwoven with different professionals' field of expertise and therefore the early diagnosis and intervention of an infant/child with sensorineural hearing loss cannot be realised if professionals work in isolation.

A role description of both the audiologist and the pediatrician are provided. It is evident that both members contribute a great deal within an interactive team towards the effectiveness of early diagnosis and intervention. Cooperation between the mentioned team members lead to a positive attitude and working relationship. Buchholz and Roth (1987: 43) states: "*...in a work unit where shared responsibility is encouraged, you can also expect to find employees who show a sense of excitement, who express a feeling of camaraderie, and who feel they are a part of something bigger than what they themselves are responsible for singly*".

Technology is improving at such a fast and rapid rate that it is becoming increasingly difficult to keep up-to-date with current issues. It is thus important to determine how one can combine the skills, knowledge and attitudes of the above-mentioned professionals to the benefit of all hearing-impaired infants/children and their families.

A well-coordinated team leads to early identification and intervention. All practitioners are therefore required to update their knowledge and clinical skills to meet the challenge of universal neonatal hearing screening (Hall, 2000).

The earlier in life a baby or a child with a hearing impairment is identified and intervention is implemented the greater the chance they have to develop normally and be successful (Martin, 2000). Therefore, testing an infant's hearing immediately after birth is none too soon (Martin, 2000). Unfortunately, there is a delay between suspected hearing loss and diagnosis, which means the goal to intervene with all

infants before or by 6 months is not met. In this study an investigation will be made in order to determine the possible cause(s) of late diagnosis and intervention.

In viewing an infant/child with sensorineural hearing loss holistically, and taking into account the need for interdisciplinary teamwork of the literature, the importance of this study can be detected as well as the positive implications it holds for the long term.