

**CLIENTS OF THE PRETORIA COCHLEAR  
IMPLANT PROGRAMME:  
CHARACTERISTICS AND PERCEIVED  
OUTCOMES OF CHILDREN AND THEIR  
FAMILIES.**

**MARGUERITE ANNE JESSOP**

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## Dedication

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## LIST OF ABBREVIATIONS

<b>ACE</b>	<b>Advanced Combination Encoders</b>
<b>ADD/ADHD</b>	<b>Attention Deficit Disorder/Attention Deficit and Hyperactivity Disorder</b>
<b>BPCIP</b>	<b>Birmingham Paediatric Cochlear Implant Programme</b>
<b>CHRIB</b>	<b>Clinic for High Risk Babies</b>
<b>CI</b>	<b>Cochlear Implant</b>
<b>CMV</b>	<b>Cytomegalovirus</b>
<b>ECI</b>	<b>Early Communication Intervention</b>
<b>ECMO</b>	<b>Extracorporeal Membrane Oxygenation</b>
<b>FDA</b>	<b>American Food and Drug Administration</b>
<b>HL</b>	<b>Hearing Loss</b>
<b>IE</b>	<b>Inclusive Education</b>
<b>JCIH</b>	<b>Joint Committee on Infant Hearing</b>
<b>NICU</b>	<b>Neonatal Intensive Care Unit</b>
<b>NPCIP</b>	<b>Nottingham Paediatric Cochlear Implant Programme</b>
<b>PET</b>	<b>Positron-Emission Tomography</b>
<b>SE</b>	<b>Specialised Education</b>
<b>SIR</b>	<b>Speech Intelligibility Rating Scale</b>
<b>PCIP</b>	<b>Pretoria Cochlear Implant Programme</b>
<b>WS</b>	<b>Waardenburg Syndrome</b>

**ABSTRACT**

**TITLE:** CLIENTS OF THE PRETORIA COCHLEAR  
IMPLANT PROGRAMME: CHARACTERISTICS AND  
PERCEIVED OUTCOMES OF CHILDREN AND  
THEIR FAMILIES

**NAME:** MARGUERITE ANNE JESSOP

**SUPERVISOR:** Dr A KRITZINGER

**CO-SUPERVISOR:** MRS P VENTER

**DEPARTMENT:** COMMUNICATION PATHOLOGY, UNIVERSITY OF  
PRETORIA

**DEGREE:** M.COMMUNICATION PATHOLOGY

In order to provide evidence demonstrating the efficacy of cochlear implantation in the children enrolled in the Pretoria Cochlear Implant Programme (PCIP), an in-depth analysis of the numerous variables involved in determining each individual child's success with his/her cochlear implant needs to be undertaken. As the PCIP has been operating for over a decade, a standardised database that includes the variables identified by similar cochlear implant programmes world-wide as being related to outcomes of children with cochlear implants and their families, has to be assembled. As the PCIP functions partly as a paediatric cochlear implant programme, an Early Communication Intervention (ECI) approach with the emphasis on the family unit is of critical importance.

To address this need, a comprehensive questionnaire was used in a cross sectional study combining qualitative, and predominantly quantitative methods. The aim was to determine the perceptions of parents/caregivers of children with cochlear implants in the PCIP of the children's outcomes. The questionnaire was

further used to gather relevant data pertaining to children and their families' biographical, medical, environmental, audiological, linguistic and educational histories and current functioning. A total of 45 participants, all mothers of children with cochlear implants, returned questionnaires.

Results indicated that several key factors played a role in determining a positive outcome in the children's audiological, linguistic, social and educational functioning leading to placement in an inclusive educational setting. These included an early age at diagnosis and prompt fitting of hearing aids and subsequent cochlear implantation, the absence of prenatal and perinatal complications including feeding difficulties, a higher level of maternal education, achievement of developmental milestones within normal age norms, the use of an FM system in the primary school phase, access to ECI, the presence of an older sibling to act as a language model, the absence of birth trauma and congenital rubella syndrome as cause of hearing loss, and later (acquired) onset of hearing loss. Children whose cause of hearing loss was non-syndromic and hereditary or unknown, were more likely to have positive outcomes.

The clinical implications for the PCIP were synthesised and presented as a developmental systems model, providing guidelines for the entire process from referral to the cochlear implant programme to exiting of the system. The urgency of the need for a universal newborn or infant hearing screening programme as well as reliable systems of early referral to cochlear implant programmes, emerged strongly in the recommendations of the study.

*Keywords: Pretoria Cochlear Implant Programme, outcomes, inclusive educational setting, paediatric cochlear implant programmes, early implantation, mode of communication, developmental systems model, variables affecting success, child with a cochlear implant, families.*

## **OPSOMMING**

<b>TITEL:</b>	KLIËNTE VAN DIE PRETORIA KOGLEËRE INPLANTINGSPROGRAM: KENMERKE EN WAARGENOME UITKOMSTE VAN KINDERS EN HULLE GESINNE.
<b>NAAM:</b>	MARGUERITE ANNE JESSOP
<b>STUDIELEIER:</b>	DR A KRITZINGER
<b>MEDE-STUDIELEIER:</b>	MEV P VENTER
<b>DEPARTEMENT:</b>	KOMMUNIKASIEPATOLOGIE, UNIVERSITEIT VAN PRETORIA
<b>GRAAD:</b>	M.KOMMUNIKASIEPATOLOGIE

Ten einde bewyse te lewer van die effektiwiteit van kogleëre inplantings in die kinders wat deel is van die Pretoria Kogleëre Inplantingsprogram (PKIP), asook om die verskeidenheid van uitkomste beter te verstaan, moet 'n in-diepte analise van elke individuele kind se sukses met sy/haar kogleëre inplanting uitgevoer word. Aangesien die PKIP vir meer as 'n dekade funksioneer, moet 'n gestandaardiseerde databasis, wat inligting aangaande veranderlikes deur soortgelyke kogleëre inplantingsprogramme wêreldwyd geïdentifiseer as verbandhoudend met uitkomste van kinders met kogleëre inplantings en hulle gesinne, saamgestel word. Aangesien die PKIP gedeeltelik as 'n pediatriese kogleëre inplantingsprogram funksioneer, moet 'n vroeë kommunikasie-intervensie (VKI) benadering gevolg word, waarin die klem op die gesinseenheid van kritiese belang is.

Om hierdie behoefte aan te spreek is 'n omvattende vraelys gebruik in 'n deursnit studie met 'n kombinasie van kwalitatiewe, en hoofsaaklik kwantitatiewe metodes. Die doel was om die persepsies van die ouers/sorggewers van hulle kinders met kogleëre inplantings in die PKIP se uitkomste vas te stel. Die vraelys is verder gebruik om relevante inligting te versamel oor kinders en hulle

gesinne se biografiese, mediese, omgewings-, oudiologiese, linguistiese en opvoedkundige geskiedenis en huidige funksionering. A totaal van 45 deelnemers, almal moeders met kinders met kogleêre inplantings, het vraelyste teruggestuur.

Resultate toon dat verskeie sleutelfaktore 'n rol gespeel het in die positiewe uitkomst van kinders se oudiologiese, linguistiese, sosiale en opvoedkundige funksionering wat gelei het tot plasing van sekere kinders in 'n insluitende opvoedkundige omgewing. Hierdie faktore sluit in 'n vroeë ouderdom van diagnose en spoedige passing met gehoorapparate gevolg deur kogleêre inplanting, die afwesigheid van prenatale en perinatale komplikasies insluitend voedingsprobleme, 'n hoër vlak van die moeders se opvoedkundige peil, die gebruik van 'n FM sisteem tydens die laerskool jare, toegang tot VKI, die teenwoordigheid van 'n ouer kind in die gesin om as taalmodel te dien, die afwesigheid van trauma gedurende geboorte en kongenitale rubella sindroom, en latere ontstaan van gehoorverlies (verworwe gehoorverlies). Kinders by wie die oorsaak van die gehoorverlies nie-sindromiese, oorerflike gehoorverlies of 'n onbekende oorsaak was, het in meer gevalle positiewe uitkomst getoon.

Die kliniese implikasies vir die PKIP is gesintetiseer en voorgestel as 'n ontwikkelings-sisteem model, wat riglyne bied vir die hele proses vanaf verwysing na die kogleêre inplantingsprogram, totdat die sisteem verlaat word. The dringendheid van die behoefte aan 'n universele neonatale/baba gehoorsiftingsprogram sowel as betroubare sisteme vir vroeë verwysings na kogleêre inplantingsprogramme het sterk in die studie se aanbevelings na vore gekom.

*Sleutelwoorde: Pretoria Kogleêre Inplantingsprogram, uitkomst, insluitende opvoedkundige konteks, pediatriese kogleêre inplantingsprogramme, vroeë inplanting, kommunikasiewyse, ontwikkelings-sisteem model, veranderlikes wat sukses beïnvloed, kind met 'n kogleêre inplanting, gesinne.*

## CHAPTER ONE

### INTRODUCTION

#### 1.1 Orientation

Cochlear implantation in children began in the mid 1980's, with the first multichannel cochlear implant device being implanted in a ten-year old boy in 1985 (Clark, Cowan & Dowell, 1997). Since then remarkable progress has been made in the technology of the device and its speech processor, in surgical techniques and in intervention programmes (Clark, *et al.*, 1997). Infants are now being implanted at increasingly younger ages (Wright, Purcell & Reed, 2002). This places a paediatric cochlear implant programme firmly within the realm of *early communication intervention (ECI) services*. Thus, in such a programme as with all ECI programmes, families are of the utmost importance as many *biological and environmental factors* play a role in the child's progress (Merenstein & Gardner, 1998; Rossetti, 1996). The family centred approach to intervention is one of the major principles of ECI. A major goal of ECI is early identification of risk factors for communication delay, including hearing impairment.

There is a worldwide trend toward *early identification of hearing loss* in infants and children (Wright, *et al.*, 2002). Families and their children are therefore entering cochlear implant programmes earlier. To best utilise this *time advantage* (Dennis, 2000; Mayne, Yoshinaga-Itano & Sedey, 2000), it is necessary that cochlear assessment protocols be fast, efficient and family-centred and that motivation for funding be handled quickly and effectively to decrease the time delay between the family's initial contact



with the cochlear team and approval for a cochlear implant, and between approval and implantation. The best possible early rehabilitation services must then follow, based on the specific needs of the client and family using an *individualised service approach*. In the words of Rossetti (1996, p.143) “...it becomes imperative that the early interventionist adopt the view that risk for communication delay and the corollary risk for school failure begins early and that communication-based intervention must, likewise, begin as early as possible.”

The outcomes of paediatric cochlear implantation are largely dependent on the *family context* in which the child finds him/herself (Rossetti, 1996). The selection criteria for implantation of children take into consideration the child’s support network (Katz, Burkard & Medwetsky, 2002), thus ruling out many children who would not have derived benefit from the implant due to a lack of follow-up support. Once enrolled in the cochlear implant program, however, it is imperative that the family be kept involved throughout the whole process.

Rossetti (1996) and Ostfeld & Gibbs (1990) support the idea of utilising parental report and of viewing the parents as accurate sources of information. This is in accordance with the notion of *family centred assessment*, which serves as a tool to include the family as team members, rally their co-operation, empower them as informed decision makers and so ensure their continued involvement in the programme. Increased levels of parental involvement also serve to enable parents to better fulfil their role as advocates for their children (Moog, 2002). Most and Zaidman-Zait (2003) studied the needs of parents of

children with cochlear implants, and found that parents expressed a need to access extensive information on a wide range of topics, as early as possible when enrolling in a cochlear implant programme.

Parents felt that a professional led multidisciplinary team was essential, each playing a dominant role at different times in the process. The need for emotional support from professionals and from other parents was rated highly. Parents further expressed the need for both group support systems and individual counselling. Such systems have been put into practice at the Pretoria Cochlear Implant Programme (PCIP), where ear-, nose- and throat surgeons, psychologists, audiologists, speech-language therapists and other therapists collaborate with teachers, parents and parent support groups. These services are, however, highly centralised and families living in areas far from Pretoria often miss out on opportunities for support and information sharing.

The PCIP has been operating for over a decade, and thus the gathering of descriptive data regarding this rapidly growing and changing population of patients is imperative for ongoing research, information sharing and efficacy of service delivery to families. There is increased pressure for outcome studies, or programme audits, to justify the cost of cochlear implantation and to demonstrate accountability for this cost in terms of efficacy (Summerfield & Marshall, 1999). If there is to be state subsidising of cochlear implants in South Africa for those children whose parents do not have the means to fund the procedure privately, strong motivation needs to come from sound research on the efficacy and cost benefits of cochlear implantation for children who are profoundly deaf. In order to describe the outcomes of the programme, a programme audit is required. Since

existing data is incomplete, the problem statement for the present study arises directly from this need. While both measured or current outcomes and perceived outcomes should be described for a complete database, the present study is undertaken with the focus on a family centred approach and so outcomes as perceived by parents are described. A critical review of research regarding the outcomes of other cochlear implant programmes is presented in the literature overview, in order to provide guidelines for the present study.

The Nottingham Paediatric Cochlear Implant Programme presented an outcomes report in 1997 on the first 100 children to be implanted at their centre (Nottingham Paediatric Cochlear Implant Programme, 1997). This represents the largest body of data on children from one centre world-wide.

Another large UK institution, the Birmingham Paediatric Cochlear Implant Programme, released an outcomes report (Birmingham Paediatric Cochlear Implant Programme, 2001) covering the ten-year time span 1990-2000. In this time 141 patients were implanted. The results were encouraging for the success of paediatric cochlear implantation, and are summarised in Table 1.

**Table 1: Outcomes of the Birmingham and Nottingham Paediatric Cochlear Implant Programmes.**

	<b>Birmingham Paediatric Cochlear Implant Programme (Outcomes Report 1999-2000).</b>	<b>Nottingham Paediatric Cochlear Implant Programme (Outcomes Report 1997)</b>
<b>Number of children used in study.</b>	The report covers the ten-year time span 1990-2000. In this time 141 children were implanted (20 per year).	The report is based on the first 100 children to be implanted at the centre.
<b>Number of children still using device</b>	Not reported	87.5% of children were wearing their device at all times three years post-implant All children implanted continued to wear the implant system.
<b>Percentage of children using speech as the main mode of communication</b>	Of all the children, 54% used total communication as their primary communication mode, and 28% were oral only. For 94% of children speech with or without sign was the primary communication mode.	A total of 87% of children used functional language as their primary means of communication three years after implantation.
<b>Aetiology of deafness</b>	50% had congenital hearing loss due to unknown causes; 7% due to perinatal complications, 35% due to genetic factors, and 8% due to prenatal infection. The majority with acquired hearing losses had lost their hearing through meningitis (65%), and a significant number had co-existing medical problems, either syndromal (18 children) or non-syndromal (11 children).	Not reported
<b>Ability to use and understand conversational speech three years post implant</b>	A total of 66% of children could understand common phrases without lip-reading and 64% were able to hold a simple conversation three years post-implantation.	57% of children were using speech one year post implant, with 69% being rated as intelligible after three years of implant use. A total of 87% of children were successfully able to carry out the McCormick Test of word discrimination three years after implantation.
<b>Percentage of children able to use a telephone</b>	Not reported	57% of children implanted before the age of five could use the telephone with a known speaker
<b>Percentage in inclusive educational settings</b>	The majority of children were in inclusive educational settings with a hearing impaired unit, and 17% in inclusive settings without support.	57% of the programme's preschool children with cochlear implants were placed in an inclusive educational setting two years post-implant.
<b>Audiometric outcomes</b>	The average audiometric configuration was a flat audiogram with thresholds around 40dB for all frequencies.	Typical high frequency response to pure tone audiometric testing was 35 dB.

As seen in Table 1, no major surgical complications have occurred in either of the programmes. It is interesting to note from the Birmingham study that clients' age at

implant has seen a change, from a mean of 9yrs in 1994, compared to 4yrs in 2000. The results reported by these two programmes, however, were attained in a developed country and caution should be used when comparing them directly to results attained in the South African context. The outcomes of the Birmingham and Nottingham programmes will now be presented and discussed.

Figures for educational placements of older children were not reported in the Nottingham report (see Table 1), and it can be argued that placements are likely to change somewhat as educational demands on the child increase following preschool. The present study will describe educational placements of children from infancy to age 16. According to Table 1 both programmes reported similar audiometric thresholds for children with cochlear implants, indicating good frequency-specific results of cochlear implantation, especially in the high frequencies. Although audiometric results based on pure tone testing provide valuable information on the perception of a wide range of frequencies through the cochlear implant, audiograms do not always relate directly to meaningful integration of sound and speech perception. Information on everyday functioning is thus required to supplement these results, and the perceptions of parents can render valuable information on how the child is using sound to function in his/her environment.

The two outcome reports further indicate that the majority of children are speaking intelligibly and conversing verbally after three years of cochlear implant use. However, information is lacking on the outcomes of the smaller percentage of children for whom this goal is not realised, and the variables surrounding their less successful outcome. A

rich description of all the variables involved in the child's life can serve to highlight contributing factors in these cases.

Although these studies by major centres deliver valuable information, neither of the studies provided an indepth description of biological, medical, biographical and environmental data nor any information about the children's families. Families and the context in which a child finds him/herself is a major focus of the present study. For this reason this study presents outcomes as perceived by the parents, rather than measured outcomes as in the two studies tabulated above.

In order to conduct a study involving outcomes of children with cochlear implants, descriptive data on the child as well as his/her context is needed. A large study undertaken by Fortnum, Marshall and Summerfield (2002) documented the *epidemiology* of the UK population of children with hearing impairment, with and without cochlear implants. Interesting results attained were that almost 30% of children with hearing impairment had another disability in addition to hearing impairment, and that children with cochlear implants were more likely to have a postnatal aetiology of deafness, and less likely to have additional disabilities involving learning or cognition. Possible reasons for this could be that selection criteria have, in the past, excluded children thought to have a below-normal IQ and limited potential for learning language (Lenarz, 1998). Children with cochlear implants were also more likely to come from more affluent families. Fortnum *et al.* (2002) found that the number of syndromal aetiologies as well as perinatal aetiologies had increased in children with cochlear implants. The

increases in reported numbers of children with these aetiologies of deafness could be due to increased awareness of syndromes and associations, and an increased survival rate of preterm infants respectively. The number of pre- and postnatal aetiologies of deafness in children had decreased, possibly due to vaccinations preventing many incidents of rubella and meningitis.

According to Fortnum, *et al.* (2002) there was a trend toward children with lesser degrees of hearing impairment receiving cochlear implants, suggesting a broadening of selection criteria regarding residual hearing of candidates as in the past only children with profound degrees of sensorineural hearing loss were considered for implantation. Similar data concerning the South African population of children with cochlear implants are needed, to give an indication of the trends surrounding selection criteria in local cochlear implant programmes. Since selection criteria ultimately dictate the profile of a programme's clients, it follows that careful inspection of both a programme's outcomes and selection criteria are required to build an accurate picture of the clients in that programme (Chester-Brown, 2005). In addition, knowledge of the variables surrounding a child with a cochlear implant and his/her family which either contribute to or hinder success can provide guidelines for the adapting of selection criteria to the population potentially being served by a programme.

As can be seen in the outcomes of the programmes in the UK described in Table 1 above, a revision and a constant shifting of the borders of *selection criteria* for paediatric cochlear implantation have accompanied the developments in technology and the

decreasing age of implantation (Eisenberg, Schaefer Martinez, Sennaroglu & Osberger, 2000; Katz, *et al.*, 2002; Lenarz, 1998). Since the South African context is unique, local research within its cochlear implant programmes is necessary to revise its own selection criteria. It is hoped that the information rendered by this study will provide guidelines in the important task of establishing the current outcomes of the PCIP, and in involving the parents of the programme as valuable team members to a greater extent by using them as sources of information.

Since information is the key to making informed decisions, it follows that a paediatric cochlear implant programme, which makes life-changing decisions regarding a child's future, requires detailed information about its clients.

According to Hebbeler (1993: p1). “ A data system is a way to produce information”. Burchill, Roos, Fergusson, Jebamani, Turner, and Dueck, (2000) emphasise that a standard system for data-collection and storage is essential to the accountability, efficiency, and scope for research and evaluation of any programme. An accurate data system thus promotes standardization and increases efficiency. The worldwide move toward justifying the cost of cochlear implantation and of rehabilitative therapy with outcome studies necessitates such a data system (Summerfield & Marshall, 1999).

The PCIP has, in the past, made use of various methods of data recording. Due to the lack of a clearly defined protocol for gathering specific client data, team members have recorded this information in differing ways, each using a different data recording style



and system and emphasising different areas of information. The information in past paper based files, though valuable, is therefore not complete. It follows that a standard system needs to be developed for the Programme, which can address the need for retrospective data collection to complete case histories of children implanted for the past ten years. In addition, the system should be capable of updating and verifying biographical records on previous patients. One system must be designed to store prospective data on clients and their families from enrollment onward, as well as to record retrospective data on existing clients. Reliability of retrospective data may, however, be compromised as the current records are incomplete. The main aim of a standardised system of data collection for the PCIP would therefore be to collect and store longitudinal data on all children and families enrolled in the programme.

A protocol firmly grounded in current research will fulfil the Programme's information needs as trends move toward earlier implantation, new surgical techniques, more sophisticated technology, changing selection criteria and the pressure for regular outcome studies (Wright, *et al.*, 2002). Analysis of the data may reveal trends that will assist the Programme's team in planning more efficiently for the changing profile of patients. To enable updating of current paper files and move toward greater efficiency, a paper-based data system which can be easily translated into a computerised data storage system is preferable.

Such a data storage system would enable longitudinal trends across clients to be tracked, supply individual client data and render information to be used to justify programme

funding (Hebbeler, 1993). In South Africa, state funding is scarce and many primary health care concerns are prioritised above cochlear implantation, which is still perceived as an elective treatment method (Summerfield & Marshall, 1999). Convincing data demonstrating the efficacy of cochlear implantation as a preferred treatment option for children with sensorineural deafness and their families, and possibly the decreased burden on state resources by a child implanted at an early age, is needed if state funding is to be applied for realistically.

The data system would allow background information as well as developmental variables to be easily stored, allowing for research into longitudinal relations among various variables (Bezruczko & Zell, 1993). In this way outcome studies can be facilitated, giving rise to the potential for greater exchange of information and collaboration with other similar programmes, programme evaluation and evidence-based revision of the programme and its protocols (Merenstein & Gardner, 1998).

The importance of ECI and early referral of possible candidates for cochlear implantation can be better substantiated by statistical data rendered by the data collection system. Such information, when disseminated to medical professionals, may serve to spur on early referrals of children and their families. With concrete evidence demonstrating the efficacy of early cochlear implantation, medical aid schemes may more readily fund the procedure. Results gleaned from the data demonstrating the necessity of rehabilitative therapy post-implant may also be used to motivate medical aid funding for ongoing speech-language therapy and long term support of children and their families.

Lastly, the standardised paper-based system can be used as the model for the development of a computerised database in the future. The aim of the current study is thus the establishment of a uniform data collection system, which may be used as a stepping stone toward the design of a computerised database system for the PCIP.

According to Mouton, (1996: p.107). “A well-defined research problem is a precondition for any study. The development of a research design thus follows logically from the research problem”. The research problem in this study arises directly from the needs of the PCIP explained in the rationale above, and can be expressed as the following question.

“Based on data collected from parents of children enrolled in the PCIP which can be stored in a specially designed, standardised data collection system, what are the defining characteristics and perceived outcomes of children and their families enrolled at this programme?”

## **1.2 Definitions of terminology used in the study**

The following list of terminology used in the study will be defined, for the purposes of this study, as such:

- **Early implantation:** this term is used in this specific study to imply that the child received a cochlear implant prior to the age of 3,5 years, as is reported by Sharma, Dorman and Spahr (2002) to be the critical early period for maximal plasticity in the auditory cortex.
- **Early Communication Intervention (ECI):** Intervention services focusing on facilitating the development of communication skills, directed at children from birth to 3 years who are at risk for developmental delays or who have a disability, and the families of these children (Kritzinger, 2000).
- **Child:** a child will refer to an individual up to and including the age of 16 years in this study.
- **Paediatric cochlear implant programme:** a programme involving children up to and including the age of 16 years, as opposed to an adult programme.
- **Communication mode:** the main manner in which the child communicates, being either by means of speech, total communication involving both speech and sign, or manual communication involving various forms of signing or a sign language (Birmingham Paediatric Cochlear Implant Programme, 2000).
- **Educational approach:** refers to either an auditory-oral, auditory-verbal, cued speech, total communication, or signing approach to the education and language stimulation of children (Clark, *et al.*, 1997).
- **Inclusion** in this context refers to the full participation of a child in a non-specialised school setting and among normal-hearing peers.
- **Perceived outcomes** will (in this study) refer to the outcomes of a child as reported by his or her parents or primary caregivers.

- **Current outcomes** will refer to the outcomes of the cochlear implant programme at one point in time, over a cross section of its client population and includes information such as the percentage of children who continue to use their cochlear implant devices and the number of children in inclusive educational settings.
- **Measured outcomes** will not be reported for this study, but will be referred to in discussions of the literature and implies results gained from clinical testing, such as percentage word discrimination and hearing thresholds.
- **Environmental factors** will be defined here to encompass all the factors external to the child which have a direct or indirect impact on him/her, such as socio-economic status and educational level of parents; geographical access to services; the family's support network; as well as the culture's views on disability and on child-rearing.
- **Biological and medical factors** will be used to refer to all the factors internal to the child such as genetics; illness and developmental progress; pre-and post-natal complications; additional disabilities and ability to process and use the sound received through the cochlear implant in a meaningful way.
- **Cochlear implant device variables** will include the type of internal and external cochlear implant devices used.
- **Programme variables** will refer to the specific approach to surgery, rehabilitation, counseling and management of clients followed by a cochlear implant programme. It includes issues such as funding, selection criteria, communication with other programmes and ongoing research and protocol revision.

### 1.3 Division of chapters

<p>CHAPTER ONE: Introduction</p>	<p>In this chapter the reader is given a brief orientation to the context of the study. The background to the study as well as its rationale are explained, and the research question posed. Terminology used in the text is then defined.</p>
<p>CHAPTER TWO: A literature review of factors that influence the success of a child with a cochlear implant.</p>	<p>Chapter two provides a comprehensive overview of the literature surrounding cochlear implantation in the paediatric population. The factors affecting success of cochlear implantation including biological and environmental factors, selection criteria, outcomes of cochlear implantation, educational approaches and the concept of inclusion are addressed. Special situations in which cochlear implantation is more complicated, such as in children with multiple disabilities; syndromes; auditory neuropathy and various etiologies of hearing loss and associated problems are discussed. Technological advances in implant technology, as well as binaural/bimodal amplification and bilateral cochlear implantation are discussed before the unique South African context is brought into this international perspective.</p>
<p>CHAPTER THREE: Research methodology.</p>	<p>The reader is taken through the methodological steps in the compilation of the questionnaire and execution of the study. The aims and objectives and procedures of the study are described.</p>
<p>CHAPTER FOUR: Results and discussion.</p>	<p>The data received from the questionnaires is assimilated into meaningful categories, analysed and presented as results. A profile of the children with cochlear implants and their families in the PCIP is compiled and important characteristics are highlighted and discussed, in order to establish a uniform data collection system.</p>
<p>CHAPTER FIVE: Conclusions</p>	<p>The relevant information from the study is used to draw conclusions about the population under study, and implications for future research are discussed.</p>

#### **1.4 Conclusion to Chapter One**

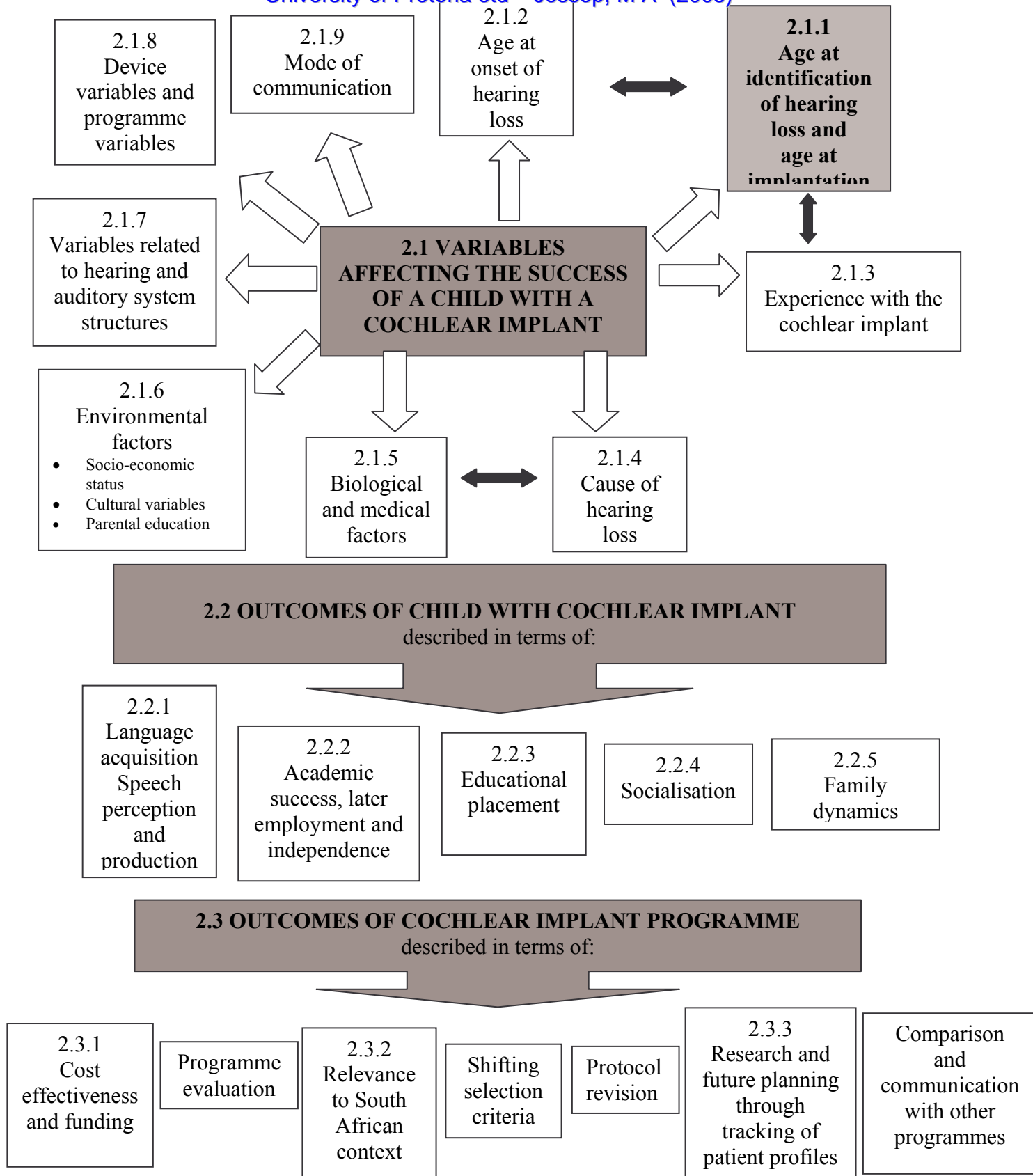
It can thus be concluded that the outcomes of cochlear implantation, now an accepted treatment option for individuals with profound and severe-profound sensorineural deafness of cochlear origin, are best viewed by analysing the population of children and families who make use of the technology with varying degrees of success.

## CHAPTER TWO

### **A LITERATURE REVIEW OF FACTORS THAT INFLUENCE THE SUCCESS OF A CHILD WITH A COCHLEAR IMPLANT.**

Throughout the literature on paediatric cochlear implantation, it is evident that the variables affecting the success of a child's use of this technology are numerous and complex, and of a highly individual nature in each case. As greater volumes of data become available and early paediatric implantees grow up and can be tracked, the profile of clients shifts and variables are better understood. This chapter will highlight some of the factors determining successful implantation of children as they have been explored by other researchers. Figure 1 summarises and illustrates some of the factors, which will subsequently be discussed.





**Figure 1: Summary of factors affecting client and programme outcomes.**

Conceptualised from: Clark, *et al.* (1997); Archbold, Nikolopoulos, Tait, O'Donoghue, Lutman & Gregory (2000); Summerfield & Marshall (1999) and Merenstein & Gardner (1998).

## **2.1 Variables affecting the success of a child with a cochlear implant**

### *2.1.1) Age at identification of hearing loss and age at cochlear implantation*

As illustrated in Figure 1 by the shaded block (2.1.1), the primary factors affecting the level of success a child achieves in language proficiency with a cochlear implant appear to be age at identification of hearing loss and age at implantation, as stated by Mayne, *et al.*, 2000. Closely related and also crucial is age at onset of hearing impairment (see number 2.1.2 in Figure 1), which determines whether or not a child has had any exposure to sound and to language prior to the onset of hearing loss. Age at onset of hearing impairment will be discussed in greater detail further on. It appears that earlier identification of hearing loss, and cochlear implantation as soon as possible after candidacy is established, may be the most important variables leading to an increased performance in the development of receptive and expressive language, speech perception and production, and in the neural development of the auditory cortex and central auditory processing pathways. Fryauf-Bertschy, Tyler, Kelsay, Gantz, & Woodworth, (1997) found a significant difference in open set speech recognition scores 24 months after implantation for children implanted before age five as opposed to children implanted after age five, when daily use of the device was controlled for. Snik, Vermeulen, Geelen, Brokx and van der Broek (1997) did not find age to be as significant a factor when implantation occurred between the ages of four and ten, although no children implanted prior to age four were utilised in the study. Children implanted after age ten scored significantly lower open set speech recognition scores. This may indicate that age four marks a boundary in expected performance, since children implanted prior to age four demonstrated better outcomes

in related studies (Lesinski, Battmer, Bertram & Lenarz, 1997; Kirk, Miyamoto, Lento, Ying, O'Neill & Fears, 2002).

Further research has indicated a sensitive period of approximately 3.5 years during which the central auditory system remains maximally plastic (Sharma, *et al.*, 2002). This plasticity remains in some children until age seven, highlighting the need for early cochlear implantation. Sharma, *et al.* (2002) have further shown, by use of the P1 cortical evoked potential, that auditory deprivation caused widespread degeneration in the central auditory system although functional neural connections were established initially in the absence of sound. After an extended period of auditory deprivation, the auditory cortical area was infiltrated and recruited by visual and somatosensory function. This could be expected to greatly reduce the efficacy of cochlear implantation. However, the rate of decrease of P1 latencies, indicating maturation of the auditory cortex, was higher for early-implanted children and reached age-equivalent levels within 8 months of switching on of the device (Sharma, *et al.*, 2002). Related studies investigated development of the auditory cortex in the absence of sound, thus prior to cochlear implantation.

A study by Ponton, Don, Eggermont, Waring and Masuda (1996) found, by using auditory evoked responses, that during the period of deafness in children with profound hearing loss, maturation of the cortical auditory function does not progress. However, upon stimulation by a cochlear implant, this maturation resumes at a normal rate. This appears to lend support to the cause for early implantation, since a shorter period of deafness seems to be correlated with better performance and a more normal pattern of auditory cortical maturation. Thus, the early critical period when this maturation is essential to language acquisition can be better utilised.

Szagun (1997), in a study on language development in children with cochlear implants, warned of possible changes in cerebral organisation of language as a result of auditory deprivation, which deprives the higher order cognitive processes of sufficient information on which to build during the language acquisition stage. O'Donoghue, Nikolopoulos, Archbold and Tait (1999) found that for children implanted before the age of five, intelligible speech generally developed within two years after the implant, and that these children showed greater benefit in speech production skills when compared with older candidates. Yoshinaga-Itano, Sedey, Coulter and Mehl (1998) demonstrated that children whose hearing losses were identified by six months of age and who received appropriate treatment (appropriate amplification or cochlear implantation and intervention), presented with expressive and receptive language abilities within the normal range regardless of all other variables except for normal cognition. Thus, the earlier a child's hearing loss is identified, the earlier cochlear implantation can be undertaken and experience with sound and language via the cochlear implant can commence.

#### *2.1.2) Age at onset of hearing loss*

A critically important factor in determining the outcome of cochlear implantation, as indicated in Figure 1, is *age at onset of hearing loss*. Infants with congenital deafness or those with severe to profound sensorineural hearing impairment due to illness in the perinatal period are at a disadvantage to those who lost their hearing later, after some exposure to sound and language. Not only is their auditory system confronted with a foreign sensory code upon cochlear implantation, but important neurological maturation has also failed to occur (Szagun, 1997). Children who acquired hearing

impairment postlingually often show immediate gains in speech perception, and remarkable progress in speech perception and production within 18 months (Katz, *et al.*, 2002). Children with severe hearing impairment and thus some residual hearing, or those who lost their hearing progressively, demonstrate faster progress than children with profound sensorineural hearing impairment. This rapid progress could be due to their greater experience with auditory input (Katz *et al.*, 2002) and due to the fact that they received linguistic input at the early stages when their auditory systems were ‘uniquely receptive’ to it (Ruben, 1995).

### *2.1.3) Experience with the cochlear implant*

Related to the age at which children with hearing losses are identified and implanted, is the concept of *hearing age and experience* with the cochlear implant, as shown in Figure 1. The earlier the implantation, the closer hearing age will be to chronological age and the longer the experience with the implant will be compared with that of later implanted children (Wright, *et al.*, 2002). Of crucial importance in determining the positive effect of a young age at implantation and corresponding experience with a cochlear implant is the amount of daily use of the device (Tomblin, Spencer, Flock, Tyler & Gantz, 1999). Geers (2002: p.173) maintains that “constant use of auditory input to monitor speech production and to comprehend spoken language provides the concentrated practice needed for optimum benefit from a cochlear implant”. Her review further found duration of device use to be the overriding factor in explaining variability in performance between children with cochlear implants. The U.S. Food and Drug Administration (FDA) has recently approved cochlear implantation in children as young as 12 months of age (Katz, *et al.*, 2002). Early-implanted children who wear their implant devices regularly are thus at a distinct advantage. Pisoni,

Cleary, Geers and Tobey (1999) found that age at implantation (closely related to age at identification of hearing impairment) and length of deafness to affect all outcome measures. Various causes or aetiologies of severe/profound sensorineural hearing impairment may imply different ages at onset, such as in the case of a congenital hearing impairment due to maternal rubella during pregnancy, or the progressive loss of hearing associated with some kidney disorders.

#### *2.1.4) Cause of hearing loss*

The *cause of hearing loss*, as seen in Figure 1, plays a significant role in determining the success of cochlear implantation in children. One in 1000 live births presents with congenital deafness in the child. 50% of these cases are hereditary and 80% of those are non-syndromic (Matsushiro, Doi, Fuse, Nagai, Yamamoto, Iwaki, Kawashima, Sawada, Hibino & Kubo, 2002). The developments in the field of genetics have helped to identify the GJB2 gene as the basic cause of autosomal-recessive non-syndromic congenital deafness. Since it is known that 60% of children with GJB2-mutations present with bilateral profound hearing losses at birth (Angeli, Utrera, Dib, Chiossone, Naranjo, Henriquez & Porta, 2000), there is a move toward early genetic screening of families and immediate referral to a cochlear implant team for these children. Genetic testing for deafness holds its own set of ethical issues which need to be contemplated before embarking on large-scale screening projects, and a family centred approach is critical.

Firstly, it is essential that families understand the uses and limitations of these tests: genetic testing can not be used to predict the degree of hearing loss, and a negative test for a specific gene does not mean the hearing loss is not genetic. Secondly, it is

essential that the professional carrying out the genetic testing and subsequent counseling to families be aware of the psychosocial impact the information may have on the particular family. Genetic testing is different from other medical tests in that it reveals facts about the entire family. Members of the extended family may need to be contacted to supply information. Parents may feel responsible for carrying over genetic defects to their children, and confidentiality issues become paramount (Smith & Robin, 2002). Therefore, geneticists have an obligation to contact other relatives should their health be implicated in the results of testing. A further problem is the possible loss of health insurance coverage due to a positive genetic test.

Furthermore, the issue of termination of a pregnancy when the genetic status of the foetus is known is a highly emotional, personal and individual one. Counseling for members of the Deaf Community holds further challenges: just as a hearing couple may prefer to have a hearing child, a Deaf couple may prefer to have a Deaf child (Smith & Robin, 2002). This tests the ability of the genetic counselor to respect individual families' viewpoints and to give an unprejudiced service, since "... the goal of genetic evaluation is to provide information and to assist families in making choices that are appropriate for them." (Arnos, 2003: p.326). The benefits of genetic counseling include emotional benefits for parents associated with knowledge concerning the cause of hearing loss, and better preparation and planning once the natural progression of the etiology of deafness in their child is known. Much medical expense on unnecessary and invasive medical procedures can be saved once a genetic disorder is diagnosed. As with early identification of hearing loss, early identification of the cause of hearing loss allows parents to go through the process of grieving early on and then focus energy and time on appropriate stimulation and treatment

procedures for their child with hearing impairment (Arnos, 2003). Knowledge of later developing associated conditions, such as visual impairment in children with Usher syndrome, may affect decision making concerning early cochlear implantation and educational options. Families will also be informed of the recurrence risks of the disorder and can thus make informed decisions regarding the planning of further pregnancies (Rehm, 2003). Clearly, it is essential to the success of cochlear implantation in a child that the correct diagnosis be arrived at, involving the whole family through-out the process of diagnosis, before cochlear implantation is undertaken. Diagnosis may be a long journey involving many tests to isolate the aetiology of hearing impairment and possible additional disabilities in the child.

Smith & Robin (2002: p.368) provide the following definitions for aetiologies of deafness. “Hereditary deafness can be distinguished from acquired/non-genetic deafness by otologic, audiological and physical examination complemented by family history and ancillary tests like temporal bone computed tomography, urinalysis, thyroid function studies, ophthalmoscopy and electrocardiography. Syndromic deafness implies the co-inheritance of abnormalities of other organ systems while with non-syndromic deafness, hearing loss segregates as the only abnormality”. While syndromic deafness is generally identified in an infant at birth due to the physical features of the syndrome, the majority of cases of congenital hearing loss in children is non-syndromic and is often overlooked until later.

Since the diagnosis of autosomal recessive non-syndromic deafness is a diagnosis of exclusion, it is incorrect in approximately one third of cases, where the correct diagnosis is congenital acquired deafness (Smith & Robin, 2002). Misdiagnosis can



cause families considerable stress, and the decision to choose cochlear implantation as an appropriate treatment for the child may be compromised or delayed until families are assured of a correct diagnosis. Syndromes involving hearing impairment are numerous, each holding different expectations for the development of an individual child with a cochlear implant.

About 400 genetic syndromes implicate hearing loss as part of their phenotype. Within the same syndrome the phenotypic expression of mutations of the affected gene may differ, leading to uncertainty in predicting outcomes of all children with cochlear implants that are affected by the same syndrome. According to Friedman, Schultz, Ben-Yosef, Pryor, Lagziel, Fisher, Wilcox, Riazuddin, Ahmed, Belyantseva and Griffith (2003) approximately 30% of individuals with hereditary hearing loss have abnormalities of other organ systems and thus are considered to have a syndrome involving deafness. A few examples of genetic syndromes, such as Waardenburg Syndrome, will be discussed here as they relate to cochlear implantation.

- *Waardenburg Syndrome.*

Waardenburg Syndrome (WS) occurs in two defined types, with three additional sub-types identified as the syndrome in addition to other clinical findings involving the upper limbs or Hirschsprung disease. All but one sub-type of WS have hearing loss as a possible phenotype. The frequency of sensorineural hearing loss in Waardenburg Type I is 20% and in Type II 50%. The syndrome expression is usually autosomal dominant with penetrance close to 100% (Keats, 2002). Visual problems requiring refractive lenses are a common concurrent finding in children with this syndrome, with vestibular system hypofunction and cleft lip and palate occurring in some cases.

A multidisciplinary team is thus required for the early and effective management of children and their families. The cochlea is often hypoplastic, necessitating careful surgical planning for cochlear implantation (Clark, *et al.*, 1997). More research is needed into the functioning of the central auditory system in children with Waardenburg syndrome and its impact on the processing of auditory stimuli through the cochlear implant. According to Shprintzen (2001) it is estimated that 2-5% of people with congenital sensorineural hearing loss have a type of Waardenburg syndrome. This syndrome thus warrants greater attention and research by professionals involved in paediatric cochlear implantation. Another syndrome implicating hearing loss, and possible cochlear implantation, is Stickler syndrome.

- *Stickler syndrome*

Stickler syndrome involves sensorineural hearing loss which is often of a progressive nature, in addition to severe deficits of vision, cleft palate, joint abnormalities and heart defects (Keats, 2002). Rehabilitation of children with Stickler syndrome who have received cochlear implants is thus more complex, and counseling must foster realistic expectations in parents and families regarding the amount and intensity of stimulation required, the time taken to reach language and communication goals and the educational options available to the child. Family involvement is crucial to the success of these children as consistent, high levels of input are needed and support systems for families are essential. Other syndromes, too, affect more than one system in the child in addition to hearing.

- *Pendred syndrome*

One of the most common types of autosomal recessive syndromic deafness is Pendred Syndrome, characterised by sensorineural deafness and thyroid dysfunction, as well as variable vestibular function. Special considerations for cochlear implantation include the associated temporal bone abnormalities such as enlarged vestibular aqueduct and Mondini dysplasia (Clark, *et al.*, 1997; Keats, 2002; Shprintzen, 2001), as well as the fact that the hearing loss may be fluctuating or progressive in nature (Friedman, *et al.*, 2003). Rehabilitationalists would need to take note of the associated symptoms of thyroid dysfunction, and sensory integration issues relating to abnormal functioning of the vestibular system. An occupational therapist would thus be an integral part of the rehabilitation team. A multidisciplinary approach including an occupational therapist is also warranted for children with syndromes affecting vision in addition to hearing.

- *Usher syndrome*

According to Shprintzen (2001), over half of all people who are both deaf and blind are affected by one of the subtypes of Usher syndrome. This syndrome is of autosomal recessive inheritance, and affects the auditory, visual, vestibular and central nervous system of an individual. The defective gene has been mapped. In Type I of the syndrome, profound bilateral sensorineural hearing loss is present at birth in the infant, as well as impaired vestibular function affecting motor and sensory development with clumsiness and ataxia as well as sensory integration problems occurring in the child. Retinitis pigmentosa develops near puberty, starting with night blindness. Occasionally, the syndrome is accompanied by cognitive impairment and psychiatric illness (Shprintzen, 2001). Multidisciplinary team involvement is required for children with Usher syndrome, and timely diagnosis may affect the decision to

undergo cochlear implantation early. Early implantation implies that, prior to the development of serious visual impairment, vision can be used to aid speech perception, lip-reading can be utilised, and environmental awareness can be maximised during the period of adjustment to the cochlear implant. The decisions regarding educational placements and mode of communication used by children will also be directly affected by knowledge of the presence of the syndrome. Oral communication may then be favoured over manual/visual communication modes, and Braille may be learnt before the onset of visual impairment. The concomitant cognitive and psychiatric involvement seen in some cases may have an effect on the efficacy of cochlear implantation, and families should receive counseling early on (Shrintzen, 2001).

- *Renotubular Fanconi syndrome*

Another syndrome in which children are affected by hearing as well as visual impairment is renotubular Fanconi syndrome. Müller, Wagenfeld & van Buuren (1996) reported on three cases of renotubular Fanconi syndrome, also called de Toni-Debre-Fanconi syndrome, in which they concluded that cochlear implantation was a successful and effective treatment option for individuals with this syndrome. Implantation in one case was prior to the onset of blindness. This syndrome is inherited or acquired, involving the urinary tract, pseudo-retinitis pigmentosa and sensorineural hearing loss. Rönnerberg, Samuelsson and Borg (2002) reported on a study in which the perceived world of the deaf-blind individual was described. Cochlear implantation was found to be "...one of the most worthwhile projects in the habilitation process." (Rönnerberg, *et al.*, 2002: p.137) since it provided sound detection and localisation, enabling the individual to attain a sense of security and

control through monitoring of the environment. The study involved persons with Usher syndrome, CHARGE association and Wolfram syndrome. It would appear that cochlear implantation can be considered a worth-while option for families of children who suffer from multiple sensory impairment in addition to hearing impairment, and that children's quality of life may be increased by using cochlear implantation as part of a holistic management plan.

- *Craniofacial syndromes*

Children with craniofacial syndromes and hearing impairment present special challenges for cochlear implant teams, in the counseling, surgical and rehabilitative stages of the process. MacArdle, Bailey, Phelps, Bradley, Brown and Wheeler (2002) stressed the importance of effective pre-operative interdisciplinary discussion, thorough radiological examination and realistic counseling of parents. The minimal requirements for these, and other children with multiple disabilities, to attain candidacy for cochlear implantation was stated by these researchers to be normal inner ear anatomy and the ability to condition to follow a rehabilitation programme. Thus, as confirmed by Clark, *et al.*, (1997) mild cognitive impairment in a child should not pose a sufficient obstacle to warrant rejection as a cochlear implant candidate. Additional factors to consider are the multiple hospitalisations and medical procedures which children with craniofacial syndromes must undergo, since these procedures will interfere with intensive auditory rehabilitation programmes and with general stimulation and development (MacArdle, *et al.*, 2002). A child's speech intelligibility will necessarily be affected by a cleft lip/palate, and the risk of middle ear infections due to incomplete muscle attachments around the Eustachian tube is higher. Due to abnormal nerve pathways associated with craniofacial syndromes in

children, facial nerve stimulation could be problematic during programming of the cochlear implant. Linguistic and auditory skills may be gained at a slower rate by children with an additional disability, as found by Waltzman, *et al.* (2000) in MacArdle, *et al.* (2002) in a study involving 31 children with multiple disabilities who had been implanted. Interestingly, no increase in surgical complications was noted for these children. The greatest variance in speech perception outcomes seemed to stem from involvement of the central nervous system, possibly indicating that neurological involvement should be investigated more closely in deciding on the candidacy of a child for cochlear implantation.

One of the craniofacial syndromes which may complicate cochlear implantation at many points along the process from selection to rehabilitation of a child after surgery is the *CHARGE association*. The CHARGE association is characterised by deficits in the central nervous system of the child as well as in the ocular, auditory, skeletal, cardiac and genital systems with craniofacial abnormalities and abnormal growth and development of children. The degree of cognitive impairment and central nervous system involvement will determine the child's language acquisition to a large degree in this disorder, thus the child should be seen and treated as a whole and realistic expectations set. Children with the CHARGE association are often very ill, with severe failure to thrive seen early in development (Shprintzen, 2001). Cochlear implantation should thus be seen as possibly contributing to overall quality of life in the child with multiple severe disabling conditions.

MacArdle, *et al.*, (2002) further report two cases of children with Goldenhar's syndrome and brachio-oculo-facial syndrome. The child with Goldenhar's syndrome

derived no benefit from the device, and upon re-examination was found to have eighth nerve hypoplasia and significant learning disabilities. The child with brachio-oculo-facial syndrome, despite requiring higher than usual stimulation levels during MAPping which prevented all the channels being used, as well as facial nerve stimulation, derived excellent benefit from the device and later entered a unit in an inclusive educational setting. MacArdle, *et al.*, (2002) warn that the time spent on assessment, processor programming and rehabilitation was approximately double that spent on less complicated cases. These researchers state children with visual impairments and cerebral palsy to be the children with multiple disabling conditions most commonly seen by cochlear implant centres, however they advocate the use of cochlear implants as part of a comprehensive treatment plan for children with other disabling conditions as well.

- *Mitochondrial deafness*

Figure 1 includes the cause of the hearing impairment (see 2.1.5), closely linked to the biological and medical factors unique to each child, as one of the major factors influencing the success of a child with a cochlear implant. Many etiological factors have been discussed in relation to the cause of hearing impairment. A less common hereditary cause of hearing impairment is mitochondrial deafness. Mitochondria are organelles in each cell which are responsible for the production of ATP (adenosine triphosphate, the chemical compound that is the source of intra-cellular energy) to fulfill the energy requirements of cells (Marieb, 2000). Mitochondria have their own DNA, are independent of the nucleus and are maternally inherited (Keats, 2002). Mutation of the DNA of the mitochondria affects those organs with the highest energy demands, thus symptoms would involve the muscles, nervous system, eyes, ears, and

cortex resulting in dementia, several neuromuscular syndromes and hearing/visual impairment in the individual. However, Rehm (2003) and Fischel-Ghodsian (2003) cited inherited mitochondrial mutations to be responsible for non-syndromic deafness with no involvement of other bodily systems, as well as a predisposition to aminoglycoside-induced hearing loss, such as in the treatment of severe neonatal infections in the neonatal intensive care unit (NICU). Deafness may be accompanied by generalised neuronal dysfunction in the central nervous system of the child (Fischel-Ghodsian, 2003) which may have implications for ECI, later rehabilitation and outcomes of cochlear implantation.

- *Multiple disabilities*

The subset of children who are congenitally deaf as a result of syndromes may present with multiple disabilities, which could in turn affect their progress with a cochlear implant. Katz, *et al.* (2002) stated cognitive disabilities to be more likely to interfere with performance with a cochlear implant than other disabilities such as blindness or cerebral palsy. Teagle and Moore (2002) found that although, in some cases, the presence of additional disabilities in a child contra-indicates implantation, the valuable sensory information which it could provide to such a child should be weighed up in each individual case. Issues which may complicate the course of therapy in specific cases are muscle weakness, high muscle tone or involuntary movements in cerebral palsy which may hamper the child's use of speech or signing; or the decreased potential for integrating auditory and visual information for speechreading in a child with visual deficits. Teagle and Moore (2002) emphasised that the presence of these factors should not preclude the child from consideration for



a cochlear implant. Educator and parental expectations should rather be adjusted accordingly.

- *Cochlear gene therapy*

Many causes of hearing impairment as well as other disabling conditions, are of genetic origin. As a result, much research has gone into deciphering the genetic code. An exciting development in the field of genetics is cochlear gene therapy, which holds the promise of “...arresting, reversing or curing deafness of a genetic and non-genetic origin” through gene transfer (Lalwani & Mhatre, 2003: p.342). This is achieved using a carrier virus or liposomes as gene transfer vectors into the cochlea via micro-injection through the round window, installation with a gelatin sponge or choleostomy infusion with a pump. These techniques are still being refined and tested to address safety concerns (Lalwani & Mhatre, 2003).

Despite the technological advances in genetics and the possibilities they offer, it remains questionable whether standardized genetic screening is a sustainable option at present in the South African health care context where the budget is already taxed by more life-threatening issues such as HIV/AIDS as well as the effects of poverty and inadequate living conditions on the health of a large percentage of the population.

- *Meningitis*

Medical factors such as accidents or illnesses are often the cause of hearing impairment, as illustrated in Figure 1. When discussing paediatric cochlear implantation and the factors which may affect a child’s progress, an important subset of children to consider is those who have contracted meningitis, since they are at

additional risk for communication delay. Meningitis as the cause of deafness is an important consideration for early cochlear implantation due to the fact that the cochleas of children who have suffered from meningitis are known to start ossification soon after illness, leaving a small window of opportunity for cochlear implantation in which all electrodes can be inserted. The need for early referral for cochlear implantation and a short period between identification of hearing impairment, approval as a candidate and implantation is thus significant in these children. *New technology* is, however, allowing improved insertion of electrodes and better results for these children with the use of split electrodes allowing electrodes to be dispersed in half the distance inside the cochlea (Bredberg, Lindstrom, Löppönen, Skarzynski, Hyodo & Sato, 1997). Studies have also shown that children who have recovered from meningitis are at risk for a variety of cognitive, motor, sensory and behavioural deficits (Clark, *et al.*, 1997). Nikolopoulos, O'Donoghue, Robinson, Gibbin, Archbold and Mason (1997) found children who were deafened by meningitis to achieve significantly lower scores on scales of auditory development at one year post-implantation than children with congenital hearing impairment, despite the former group's previous history of hearing. It is clear that a child who has acquired a hearing loss through meningitis requires a swift and thorough approach to management before and after cochlear implantation.

- *Auditory neuropathy*

An emerging problem specifically related to early cochlear implantation is that of auditory neuropathy. Auditory neuropathy is in some cases a contra-indication to implantation, yet it is difficult to identify in a child under six months of age due to the need for behavioural audiometric testing to confirm the diagnosis (Yoshinaga-Itano,

2000). According to Yoshinaga-Itano (2000), hyperbilirubinaemia and genetic causes are two possible causes of auditory neuropathy. She also found that most infants with auditory neuropathy spent time in NICU. Rance, Cone-Wesson, Wunderlich and Dowell (2002) mentioned oxygen deprivation as a further risk factor for auditory neuropathy. The diagnosis is characterised by sensorineural hearing loss in which no auditory brainstem response can be evoked in the presence of intact oto-acoustic emissions (Miyamoto, Iler Kirk, Renshaw, Hussain & Seghal, 2000). This indicates a more central locus for the hearing impairment than the outer hair cells of the cochlea. Rance, *et al.* (2002) described the condition as one in which a neural transmission or synchrony disorder is indicated by the absence of evoked neural activity at auditory nerve or brainstem level. For this reason Stredler-Brown (2002) used the term auditory dis-synchrony to describe this disorder, which is characterised by an unpredictable course including instances of worsening, fluctuating, and even complete resolving of the condition. Speech perception abilities in these children are disproportionately poor in relation to pure tone audiometric results. The problem which arises is the inability to predict the effects of the hearing impairment on speech perception and subsequent language development in a young child with auditory neuropathy. Hearing in noise also seems to present greater problems than in quiet (Stredler-Brown, 2002). Although tests indicate retrocochlear pathology, the exact sites of lesion are not yet determined.

There have, however, been several cases of successful implantation of children with diagnosed auditory neuropathy. Miyamoto, *et al.* (2000) described gains in closed set speech recognition for two children with auditory neuropathy and believed that results, although less than optimal, may exceed that which would have been achieved

with acoustic stimulation. Conventional amplification using hearing aids has been less successful, and remains a controversial issue (Rance, *et al.*, 2002). Often children with this disorder who have been fitted with hearing aids enjoy hearing more sound, however this does little to improve their speech perception since the sounds become louder with amplification but the integrity of the signal does not improve (Stredler-Brown, 2002). Children with auditory neuropathy whose pure tone thresholds fall within the severe-profound range are viewed by Rance, *et al.* (2002) to be candidates for cochlear implantation. Although pure tone levels cannot be related to speech perception in this population, there have been no instances in which speech perception was better than pure tone hearing thresholds. The goal of any treatment option for a child with auditory neuropathy is the development of language skills (Stredler-Brown, 2002). A variety of methods can be used to attain this goal, one of these being cochlear implantation. Visual communication methods have been used with success, with the children's dependence on this mode generally being related to their ability to benefit from auditory information. Stredler-Brown (2002) cited auditory-verbal therapy to be less effective for children with auditory neuropathy than for children with sensorineural hearing loss. This researcher recommended that language development should parallel cognitive development, and that whichever educational approach achieved this was most effective for the individual child.

- *Central auditory processing*

Figure 1 includes biological and medical factors specific to each child to be important variables influencing success of the child with a cochlear implant. An emerging area of research into these factors investigates central auditory processing as a primary factor in the large variation seen in cochlear implant recipients. In the very young

child, the development of perceptual, cognitive and linguistic abilities emerge after implantation and performance is thus difficult to predict. Pisoni, *et al.* (1999) thus suggested a new generation of outcomes research, focussing on the process rather than the outcome of success or failure of a child with a cochlear implant. In the words of these researchers: “Something else is happening at more central levels of processing in the nervous system beyond the auditory nerve...” (Pisoni, *et al.*, 1999: p.113). Higher level perception, attention, learning, memory, the interactions between sensory input and stored knowledge, and the integration of sensory input may well provide the key to understanding individual differences in performance.

#### *2.1.5) Biological and medical variables*

Biological and medical factors that can have an influence on a child’s success with a cochlear implant include prenatal, perinatal and postnatal complications which have been known to signal higher risk for sensorineural hearing loss in neonates and infants (see Figure 1). These can be delineated by high risk registers. Although high risk registers have limitations in terms of the early identification of hearing impairment in infants, they offer valuable guidelines to cochlear implant teams when examining the case history of a cochlear implant candidate pre- or post-implantation. The Joint Committee on Infant Hearing (JCIH) released a position statement in 1990 suggesting the following ten risk criteria for sensorineural hearing loss in neonates (0-28 days) (Mauk, Barringer & Mauk, 1995). These factors are tabulated in Table 2. In 2000 a revision of these criteria was released, which clustered the perinatal acute medical factors to ‘an illness or condition requiring admission of 48 hours or greater to an NICU’ (JCIH, 2000). An additional factor, listed in the updated risk criteria for infants (29 days to 2 years of age), is a history of conditions in the neonatal condition

requiring the use of extracorporeal membrane oxygenation (ECMO). For the purposes of this study it was decided to use the more descriptive 1990 set of criteria, which stipulate each condition in the neonatal period. The 2000 position statement further includes a previously lacking emphasis on conductive hearing loss and syndromes associated with conductive hearing loss, persistent otitis media with effusion, and Eustachian tube dysfunction.

**Table 2: JCIH risk factors for sensorineural hearing loss in the neonatal period (0-28 days).** (Mauk, *et al.*, 1995)

1) Family history of childhood hearing impairment.
2) Congenital perinatal infection (cytomegalovirus (CMV), rubella, herpes, toxoplasmosis, syphilis)
3) Anatomical malformations involving the head or neck (syndromal and non-syndromal abnormalities)
4) Birthweight less than 1500 grams.
5) Hyperbilirubinaemia at a level exceeding indications for exchange transfusion
6) Bacterial meningitis, especially haemophilus influenzae
7) Severe asphyxia (Apgar scores between 0 and 3 / infants who fail to institute spontaneous respiration by 10 minutes / hypotonia persisting to 2 hours of age)
8) Ototoxic medications used more than 5 days and loop diuretics used in combination with aminoglycosides
9) Stigmata associated with syndromes known to include sensorineural hearing loss
10) Prolonged mechanical ventilation for ten days or longer

Table 3 continues with the JCIH's list of risk factors pertaining to infants (29 days to 2 years). Mauk, *et al.* (1995) stated that 50% of children with severe to profound bilateral sensorineural hearing loss manifested with at least one of the high-risk criteria.

**Table 3: JCIH list of risk factors for sensorineural hearing loss in the infant (29 days to 2 years).** (Mauk, *et al.*, 1995)

1) Parental/caregiver concern regarding hearing; speech; language and/or
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developmental delay
2) Bacterial meningitis
3) Neonatal risk factors that may be associated with progressive sensorineural hearing loss e.g. CMV
4) Head trauma
5) Stigmata associated with syndromes known to include sensorineural hearing loss
6) Ototoxic medications used for more than 5 days and loop diuretics used in combination with aminoglycosides
7) Children with neurodegenerative disorders (e.g. neurofibromatosis; Tay Sach's disease)
8) Childhood infectious diseases known to be associated with sensorineural hearing loss (e.g. mumps; measles).

As seen in Tables 2 and 3, children who were deafened early due to illness or trauma pre-or peri-natally often have additional conditions. These children are also often graduates of the NICU, which presents its own set of complications (Merenstein & Gardner, 1998). Studies have shown that the *NICU environment* is negatively correlated with early parent-child attachment, an area which is already at risk in the dyad of hearing parent and child with hearing impairment (Moehn & Rossetti, 1996). Infants in the NICU may have been *preterm*, have had *low birth weight*, presented with *failure to thrive*, or have suffered any of a list of related *medical factors* and sequelae (Merenstein & Gardner, 1998). These factors are represented in Figure 1 as biological and medical factors and link directly the cause of hearing impairment, as represented in the figure by a double arrow.

*Preterm infants* demonstrate an increased risk for short term memory and specific language deficits in later development (Briscoe, Gathercole & Marlow, 1998) as well as attention, behaviour and learning problems, difficulty with social skills, hyperactivity, fine and gross motor deficits and some cognitive deficits (Robison & Stewart Gonzalez, 1999). Children who presented with *preterm birth* and were *small for gestation age* are at an even greater risk for developing speech/language delays

than infants who were only preterm (Gonzales, Montgomery, Fucci, Randolph & Mata-Pistokache, 1997). Infants who were preterm as well as being small for gestation age are at a greater risk for complications, such as congenital sepsis which is treated with drugs such as ototoxic aminoglycosides; respiratory distress or disease processes requiring mechanical ventilation or extracorporeal membrane oxygenation therapy and high frequency oscillatory therapy (Lasky, Wiorek & Becker, 1998) for extended periods. These treatments present a risk for sensorineural hearing loss, with ECMO being a recent addition to the list of risk indicators in the 2000 position statement (JCIH, 2000). These same infants are at risk for developing neurosensory diseases such as retinopathy, as well as intra-ventricular hemorrhage and seizures (Lasky, *et al.*, 1998). According to Rossetti (1996) the possible sequelae of low birth weight and preterm birth include respiratory distress syndrome; necrotising enterocolitis; bronchopulmonary dysplasia; patent ductus arteriosus; apnoea; bradychardia and inter-ventricular hemorrhage. Hyperbilirubinaemia in infancy has been linked to auditory neuropathy (Rance, *et al.*, 2002). An additional condition which can complicate and compound the effects of a hearing impairment on an infant is *failure to thrive*, either organic or non-organic, which is related to a deficiency of nutrients to the developing brain of an infant, leading to developmental sequelae (Wooster, 1999). Such conditions present a risk for hearing loss and hold the possibility of later developmental delays in all areas, including speech and language development.

Two further criteria in Table 2 are syndromes associated with hearing loss, and maternal infections. Many children with *syndromes* related to hearing loss spent time in the NICU due to the feeding difficulties associated with the syndromes, for



example Pierre Robin sequence, craniofacial abnormalities, Goldenhar syndrome and CHARGE syndrome (Cone-Wesson, Vohr, Sininger, Widen, Folsom, Gorga & Norton, 2000). *Maternal infection* during pregnancy can also result in acquired hearing loss and time spent in the NICU. The most common infections during pregnancy causing deafness are syphilis, toxoplasmosis, rubella, cytomegalovirus and herpes simplex. These infections also bear later consequences for children, such as motor delays, cognitive delays/deficits, impaired vision, epilepsy and balance problems (Clark, *et al.*, 1997). Of particular and growing concern in South Africa is the population of infants maternally infected with HIV/AIDS. These children carry a high risk for sensorineural hearing loss, although due to their chronic illness it is expected that they will pose an ethical dilemma to cochlear implant programmes in the future (Matkin, Diefendorf and Erehberg, 1998). It is evident that many children who receive cochlear implants carry with them an array of complex risk factors over and above their hearing loss, which could influence their progress negatively.

#### 2.1.6) *Environmental variables*

Aside from the biological factors discussed thus far, several environmental factors play a role in early success of implantation, as indicated in Figure 1. Closer analysis reveals some of these environmental variables to be the level of *maternal education*, *socio-economic* status of the family; the family's *support network*; the family's *cultural views* on disability and the level of *emotional availability* of both child and mother (Pressman, Pipp-Siegel, Yoshinaga-Itano, Kubicek & Emde, 2000). Parental education and income are related to the variables in the child's environment, like the quality of the language input he or she hears, the material resources available, the parents' knowledge about child development and their overt behaviour (Dollaghan,

Campbell, Paradise, Feldman, Janosky, Pitcairn & Kurs-Lasky, 1999). Pressman, *et al.* (2000) found a direct link between the quality of mother-child interaction and child vocabulary gain in young children with hearing impairment, and found the dyad to be at risk for these children and mothers. Mothers of a lower socio-economic status and thus with a lower educational status seem to be more likely to have preterm, low birth weight infants. This clearly presents a double risk for development and for parent-child interaction (Blair, Ramey & Hardin, 1995). The family's cultural views on disability will affect their views on child-rearing, and on participation in intervention efforts (for example ECI after cochlear implantation) and are important to bear in mind if true family-centred services are to be offered (Zhang & Bennett, 2001).

#### *2.1.7) Variables relating to hearing and hearing mechanism/structures*

All the variables mentioned thus far play a role in the careful selection of infants or children, and their families, who are candidates for cochlear implantation. Selection criteria, as illustrated in Figure 1, are thus an important variable affecting the outcome of a cochlear implant programme. As experience, technology, descriptive research and confidence in the reliability of the procedure of cochlear implantation in children progress, an inevitable shifting of the borders of *selection criteria* for cochlear implantation has occurred. According to research done at the Medical University of Hanover, Germany by Lenarz (1998), the shift has been in four domains: the areas of age, residual hearing, additional disabilities and complicated cases with specific etiologies. Children are now being implanted at *younger ages*, under the age of two. Children with severe hearing impairments, and thus some *residual hearing*, are receiving cochlear implants. Children with *additional disabilities* are being implanted, with the view that cochlear implantation will add to the general

development of the child and enable additional disabilities to be better managed through the improvement in communication skills. Visual, motor and many intellectual deficits in children are now being seen as irrelevant to the outcome of implantation if properly managed. Lastly, children with *specific malformations of the cochlea* due to syndromes, specific disease processes (e.g. meningitis) or head trauma are being successfully implanted with new electrode array designs and surgical techniques. Lenarz (1998) further added that with borderline cases it is generally the responsibility of the cochlear implant team to decide whether or not to approve cochlear implantation. The decision should be based on each individual case, and with information from parents and educators working with the child. In a different study at the same Centre, children who were rejected based on the early strict candidacy requirements were re-evaluated, and many were accepted for cochlear implantation (Hartrampf, Lesinski, Allum, Dahm & Lenarz, 1995). Absolute contra-indications to selection of a patient for cochlear implantation still include children with autistic spectrum disorders; complete obliteration of the eighth nerve; or substantial progress with the use of hearing aids.

Eisenberg, *et al.* (2000) presented evidence at the Seventh Symposium on Cochlear Implants in Children indicating that *candidacy criteria* can be shifted to include children with moderately severe hearing impairment, and with considerable open-set speech recognition with hearing aids. They found children with cochlear implants to perform as well or better than children with moderately severe hearing impairment and optimally fitted hearing aids on speech discrimination tests when phonetically balanced word lists were used.

An Australian study by Cowan, DelDot, Barker, Sarant, Pegg, Dettman, Galvin, Rance, Hollow, Dowell, Pyman, Gibson and Clark (1997) indicated better performance with cochlear implants of children with more residual hearing (in the severe range) as opposed to profoundly deaf children. Deguine, Fraysse, Uziel, Cochard and Cormary (1997) of the University of Toulouse in France recommended that 'borderline' cases be implanted earlier rather than later, due to the available evidence of the success of these candidates. Kiefer, von Ilberg, Reimer, Knecht, Gall, Diller, Stürzbecher, Pfennigdorff & Spelsberg (1998) found residual hearing to remain statistically unchanged in children implanted, and hearing in the contralateral ear to improve slightly, possibly due to cortical maturation effects of auditory stimulation. These authors warn, however, that in each individual case the potential hazards related to implantation in borderline cases, including the loss of residual hearing and the possibility that the same level of performance could have been achieved with hearing aids, must still be considered.

#### 2.1.8) *Device and programme variables*

Figure 1 further lists device variables, pertaining to the specifications of the child's cochlear implant or bilateral cochlear implants, and the hearing aid worn by some children on the non-implanted ear. Additional listening devices such as FM systems also fall under the heading of device variables. With the shifting and widening of selection criteria, the likelihood of a child being implanted who has useable *residual hearing* in the contra-lateral ear is increased. Thus there is renewed interest in the use of a hearing aid on the contra-lateral ear. Ching, Psarros, Hill, Dillon and Incerti (2001) explain that historically, hearing aid use was disregarded in the non-implanted ear since it was assumed that this stimulation was at best unnecessary, and at worst

confusing and a source of interference to the electrical stimulation provided by the cochlear implant. However, with regular and consistent use of both devices, optimal hearing aid setting and loudness balancing as well as frequent hearing aid checks, regular testing for changes in the hearing thresholds of the non-implanted ear, and preferably conversion to the ACE processing strategy, Ching, *et al.* (2001) have demonstrated definite binaural advantages to *binaural/bimodal amplification*. These include better frontal horizontal localisation; better speech perception in the presence of noise, and improved functional listening in everyday life. In addition, development and maintenance of the contralateral auditory pathways is a strong motivation for wearing an optimised hearing aid on the non-implanted ear. In a similar study Tyler, Parkinson, Wilson, Witt, Preece and Noble (2002) reported binaural advantages and definite binaural integration of the acoustic and electric signals in all children except one, who had the poorest performance out of the test group using the hearing aid alone. It can thus be concluded that the absolute as well as the relative levels of performance with the cochlear implant and hearing aid respectively will influence the potential of an implantee for binaural integration (Tyler, *et al.*, 2002).

Ching, Psarros, Hill, Dillon, Incerti and Hill (2002) found significant benefits in speech perception, localisation and aural/oral functions in everyday life when an adjusted hearing aid was worn on the non-implanted ear. Similarly, Armstrong, Pegg, James and Blamely (1997) demonstrated improved speech perception in noise in the binaural condition. These factors may be of benefit to children in noisy school situations. Some cochlear implant recipients, however, remain aware of a confusing interaural delay. Due to the relatively short period that cochlear implants have been in existence, there is no research documenting the effect on the auditory system or on the

device when a cochlear implant is used for more than 20 years. The rapid advances in technology also bear the possibility that a more sophisticated, fully implantable device could be available in the future (Katz, *et al.*, 2002; Lenarz, 1997). A *long term view* then must take cognizance of the question of wearing a hearing aid on the other ear in order to preserve it for possible later implantation.

Similar issues are addressed by *bilateral cochlear implantation*. The motivation for bilateral implantation, according to Müller, Schön and Helms (2002), is that although unilateral cochlear implant recipients show high levels of speech understanding in noise, they are faced with a monaural disadvantage in noise or reverberant environments. These researchers comment that bilateral implantation was only seen in this light since 1995, and that it has been shown that adult bilateral cochlear implant users have, to differing degrees, access to interaural time and level differences which are the basis for the binaural advantage to having two ears. The term *bilateral advantage* is used by Müller, *et al.* (2002: p.201) to indicate “the difference in speech understanding between listening with two cochlear implants compared with one” and includes the head shadow effect, the squelch effect and the binaural summation effect. The participative benefits of bilateral cochlear implantation are reported to be described as a richer and more natural sound, and ease in understanding speech at softer levels. Van Hoesel, Ramsden & O’ Driscoll (2002) found the squelch effect to be less available than the other two effects in adults fitted bilaterally with cochlear implants, since advantages associated with interaural level differences were more robust than benefits related to interaural time delays. These advantages could well make the task of developing speech and language easier for infants and children with bilateral cochlear implants.

A further development in hearing implant technology concerns those clients who, because their sensorineural deafness is of a *retrocochlear pathology* and not of a cochlear origin, are not suitable candidates for a cochlear implant. The *auditory brainstem implant* is an appropriate treatment for clients in whom both eighth nerves are dysfunctional. The internal portion of the device stimulates the cochlear nucleus within the lateral recess of the fourth ventricle of the brainstem, thus restoring a degree of auditory sensation. Nevison, Laszig, Sollman, Lenarz, Sterkers, Ramsden, Fraysse, Manrique, Rask-Anderson, Garcia-Ibanez, Colletti and von Wallenberg (2002) found it to be an effective treatment for patients who had had bilateral acoustic tumours removed due to *neurofibromatosis, Type II*. These researchers reported that users had access to environmental sounds and suprasegmental components of speech such as rhythm and stress which aid in lip-reading; and that some recipients had enough speech perception to follow a conversation without visual cues. Although the surgery is more complex and risky than that for cochlear implantation, and the likelihood of some non-auditory stimulation is relatively high, Nevison, *et al.* (2002) stated a favourable risk to benefit ratio. Positron emission tomography (PET) images used by Miyamoto and Wong (2001) indicated that the cortical areas classically associated with processing of speech were activated during listening to speech, but not to broadband noise. Improved processing of speech was found in a successful user of an auditory brainstem implant as well as users of cochlear implants. Interestingly, the non-successful adult cochlear implant user showed activation in the frontal regions of the brain during listening, showing that other processing strategies were being engaged.

As with all technological devices, cochlear implants are susceptible to damage and technical or *device failures*, and may need to be replaced or re-implanted. This could be a question in the minds of parents who consider the possible availability of more sophisticated technology in their child's future. Hamzavi, Baumgartner and Pok (2002) investigated the effects of *re-implantation* of a multi-channel cochlear implant in seven patients. All patients achieved the same or nearly the same level of speech recognition with the re-implanted device as with their original multi-channel cochlear implant, demonstrating no negative effects of re-implantation should surgery be successful and without complications.

#### 2.1.9) *Communication mode*

The final variable, namely *communication mode* (see 2.1.9 in Figure 1) will be discussed extensively in the following section on outcomes of children with cochlear implants.

All the variables affecting the success of a child with a cochlear implant discussed so far have a collective impact on the cochlear implant programme's outcomes, as illustrated in Figure 1. Careful consideration of all possible variables, and wise application of selection criteria and management of clients and their families will eventually result in a positive outcome when an audit of a cochlear implant programme is carried out.



## 2.2 Outcomes of a Child with a Cochlear Implant

The *outcomes of paediatric cochlear implantation programmes*, though variable, have overall been very positive. There is no longer a question of whether cochlear implantation is a viable option for children with early profound deafness, or whether the procedure is safe. “Outcomes in audition, speech and language achieved by children with implants are similar to those achieved by more mildly impaired children who are managed with hearing aids” (Summerfield & Marshall, 1999: p142). O’Donoghue, Nikolopolous and Archbold (2000) stated in August 2000 that over 10 000 children had received cochlear implants worldwide.

### 2.2.1) *Language acquisition, speech perception and speech production.*

The rate of language progress for children who were implanted early is reported by Rhoades and Chisolm (2000) to become normalised, implying one year of progress for every 12 months, however, overall language delays are still evident. In contrast, Schery and Love Peters (2003) reported the rate of language learning for children with similar hearing losses wearing hearing aids to be half this rate. The most difficult aspect of language for the child with a cochlear implant appears to be syntax. Speech outcomes of children with cochlear implants outweigh those with hearing aids, since cochlear implants appear to have a normalising influence on suprasegmental features of voice and articulation, including fundamental frequency, nasal-oral balance, breathiness, sibilants, intonation and stress (Schery & Love Peters, 2003).

Figure 1 lists outcome measures relating to the child with a cochlear implant. With numbers of cochlear implantees growing, clearly defined outcome measures are needed. The *most significant outcome measure* of the success of a cochlear implant is

*language*. The functional use of language necessarily implies adequate speech perception for this task (O'Donoghue, *et al.*, 2000). Moog (2002) reported more than 65% of the children with cochlear implants at the Moog Center for Deaf Education in St Louis scoring in the average range on standard tests of vocabulary and language ability. Benefits have been shown in the areas of *reading, academic progress and socialisation*. Spencer, Barker and Tomblin (2003) studied the link between cochlear implant users' gains in oral language comprehension and use, and their subsequent literacy development. The low levels of literacy attained by students with hearing impairment wearing hearing aids, as well as the low level of literacy affecting employment rates of adults with hearing impairment, have been much documented. The study by Spencer *et al.*, (2003) thus compared groups of hearing children to cochlear implant users as they made the transition from learning to read, to "reading to learn" (p.237). Findings were that the children's proficiency in oral language was directly related to both reading level and writing level at the early stages of writing development. Children with cochlear implants (implanted before school age) achieved equivalent reading levels to the normal hearing children, and only slightly lower reading comprehension scores. However, at the more complex level of writing the normal hearing children appeared to have dissociated their writing style and complexity from their oral productions, where-as the two were still closely linked in the cochlear implant group. Children with cochlear implants also had more trouble with the formulation of written sentences, grammatical structures, and complexity and length of sentences. It thus appears that expressive language, oral or written, is more difficult for children with cochlear implants than receptive skills (Spencer, *et al.*, 2003).

Factors internal to the child, such as temperament, cannot be ignored as they play a role in the success of paediatric cochlear implantation. The level of *autonomy in communication* shown by the child prior to cochlear implantation has been demonstrated by Tait, Lutman & Robinson (2000) to be a prognostic factor for speech production post-implantation, irrespective of whether the communication attempts are non-verbal or verbal.

#### *2.2.2) Academic success, later employment and independence*

Children with cochlear implants have been shown to reach a higher level of reading proficiency than deaf/hard of hearing children have in the past. Spencer, Tomblin and Gantz (1999) found a quarter of their children with cochlear implants to be reading at or above the appropriate age level. Moog (2002) demonstrated even better results, with over 70% of the children in her study reading within the average range for their age. Moog (2002) suggested that a new standard be established for the achievement of deaf children with cochlear implants. It is expected that the discrepancy between the skills of normal hearing children and children with cochlear implants will diminish as age of implantation becomes earlier.

#### *2.2.3) Educational placement and mode of communication*

A variable which appears in Figure 1 number 2.2, namely outcomes of the child with a cochlear implant, is educational approach and placement. This factor is closely linked to the mode of communication used by the child, listed as a variable (2.1.8) affecting the success of a child with a cochlear implant. The *educational approach* followed and the primary *mode of communication* (see 2.1.9) certainly seem to have an influence on the eventual performance of a children with cochlear implants.

Children in auditory/oral educational settings generally achieve higher levels of speech perception, production and intelligibility than do children in signing and even in total communication settings. Archbold, *et al.* (2000) found this to be true in a UK-based study of children at three, four and again five years after cochlear implantation. In part, this could be due to the amount of emphasis placed on listening and on the wearing and use of/dependence on the cochlear implant. In the same study, Archbold, *et al.* (2000) reported that after implantation, many children switched to an oral approach with good results despite pre-implant signing. This seems to indicate that pre-implant signing is not a disadvantage to developing later oral skills. In support of these findings, Tait, *et al.* (2000) have reported that the quality of pre-implant communication in children, regardless of mode, is a predictor of later success with a cochlear implant.

According to Pisoni (2000) the poorer performance of children using total communication could also be attributed to the fact that there is an interference and inhibition effect when two modalities are used during the learning of oral language through use of a cochlear implant (see 2.1.9 in Figure 1). It appears that knowledge or use of manual language competes with the processing of speech via the auditory modality. This causes an increase in the processing load on working memory and a slowing of the auditory language learning process in the child.

*Full inclusion* in an inclusive educational setting is a very realistic option for many children who were implanted before the age of five years (Summerfield, Marshall and Archbold, 1997). The capability of the speech processor to be fitted to an *FM system* for improved classroom listening, and the *ongoing support* of educational

audiologists, teachers, parents and speech language therapists as academic demands on the child change and grow, are vital ingredients in successful inclusion. According to Nevins & Chute (1996), children with cochlear implants in the inclusive educational setting achieve higher levels of academic achievement (see number 2.2.2 in Figure 1) as well as better speech intelligibility than do their peers in special school settings. Duncan (1999) found that there were no significant differences in most of the conversational skills of children with cochlear implants who had been integrated into a kindergarten setting with normal hearing peers. Differences related to topic changes, topic maintenance and initiations were difficulties related to extraneous factors inherent in the hearing impairment rather than the child's *communicative competence*. These children had been integrated as soon as a hearing loss was identified, demonstrating support for inclusion to develop appropriate social skills (see 2.2.4 in Figure 1).

An analysis by Nittrouer and Thuente Burton (2003) of the children with cochlear implants who were successfully placed in inclusive settings revealed that they had all received intensive language stimulation prior to formal schooling, and that their performance on tasks of phonetic awareness, working memory and sentence comprehension was equal to that of their normal hearing peers. The need for early and appropriate language intervention is thus highlighted if successful entry into the inclusive school situation is the goal.

There is a worldwide trend toward inclusion of children with cochlear implants as well as children with a wide range of other disabilities (see Figure 1, number 2.2.3). Archbold, Nikolopoulos, Lutman and O'Donoghue, (2002) examined the educational

settings of 42 children with profound hearing impairment who had received cochlear implants, three years post-implantation and found that 38% were attending inclusive schools. This percentage was equivalent to the number of children with severe hearing losses wearing hearing aids in the inclusive setting. They thus concluded that children with cochlear implants and profound hearing impairment were in similar educational settings to children with less severe hearing impairments. This is expected to hold implications for both special schools for the hearing impaired, who will receive fewer children with profoundly hearing impairment as numbers of children receiving cochlear implants increase; and for non-specialised schools. Future educational planning will thus be affected as a whole, with special schools serving as specialist centres providing information and support, as well as a bridge from special to mainstream settings (Archbold *et al.*, 2002). Children in this study who were implanted prior to educational placement were more likely to attend non-specialised schools. Children who were implanted prior to the decision being made about their educational placement were also more likely to be found in inclusive educational settings.

Number 2.2.2 in Figure 1 lists academic success as an outcome variable. Ertmer (2002) found a positive correlation between children's implant experience and progress toward educational independence, defined as functioning in the inclusive setting without support services. However, it was also found that inclusion initially involved an increased need for support services for children in order to adjust to the new and higher levels of linguistic and learning demands. Over time decreased reliance on support services occurred, implying cost savings in the long term, as verified by Archbold *et al.* (2002). Proficiency in oral language was stated by Ertmer

(2002) to be the determining factor in the choice of educational placements. In turn, the child placed in the inclusive setting is immersed in natural peer language which supports further language development, and leads to higher educational attainment due to the language-literacy link. However, since the lives of children with cochlear implants are not static, constant monitoring of the appropriateness of educational placement is required, and changes should be made on the basis of “facts rather than philosophy or expectation” (Archbold *et al.*, 2002: p.160). The goal should never be to dogmatically aim to place deaf children in regular classrooms, but to choose the most beneficial environment for the child at that time, in order for the child to reach his/her individual potential. Children with cochlear implants are faced with acoustic, academic, attention, linguistic, social and adjustment challenges in the mainstream school environment (Chute & Nevins, 2003) and the impact of these on the psychosocial wellbeing of the individual should not be overlooked.

#### *2.2.4) Socialisation*

As seen in the preceding section, social skills thus seem to be encouraged by early inclusion (Duncan, 1999) as well as by good speech perception and production skills (O’ Donoghue, Nikolopoulos, Archbold and Tait, 2000; Moog, 2002). These factors are also linked to early cochlear implantation.

#### *2.2.5) Family dynamics: parental stress*

Lowered levels of *parental stress*, listed in Figure 1 number 2.2.1 as family dynamics, seem to be positively affected by cochlear implantation. Parents of children with cochlear implants were found by Horsch, Weber, Bertram and Detrois (1997) to experience levels of stress equivalent to that experienced by parents of normal hearing

children, and substantially less than hearing parents of deaf children. Children with cochlear implants have a better chance of entering mainstream education systems and of being successfully employed later in life, if implanted early, than do children following the non-oral, signing approach of the Deaf Community or those with hearing aids and the same degree of hearing impairment. Children with cochlear implants thus have a greater chance of attaining independence and employment later in life (see number 2.3.7 in Figure 1).

### **2.3 Outcomes of the Cochlear Implant Programme**

As the focus of the study is the variables and outcomes of children with cochlear implants and their families, and not of the cochlear implant programme, only three variables in section 2.3 of Figure 1 will be discussed. The reader is referred to the relevant section of Figure 1 for a list of the possible programme outcome variables.

#### *2.3.1) Cost effectiveness and funding issues*

Outcomes must be measured not only for the child but also for the cochlear implant programme, as shown in Figure 1. This raises question of cost effectiveness and programme efficacy. Ruben (1995) stated that individuals with hearing impairment who had not received appropriate intervention were at a substantial economic and social disadvantage in the current communication and information based society. Mauk *et al.* (1995) added that hearing impairment and deficits in communication hold morbidity not only for the individual economically and socially, but for society in its productivity and socialisation. A cost analysis study in the Netherlands by Severens, Brokx and van den Broek (1997) divided the costs of cochlear implantation into selection, implantation, rehabilitation, long-term care and non-medical (e.g. travel)



costs. They found the ratio between cost and effect to be highly sensitive to the price of the implant hardware, and that predictions of later savings in educational and employment costs are difficult to predict.

In the UK, where cochlear implants are state funded, cost effectiveness studies have shown that children implanted early will be able to be integrated into inclusive schools (with support) and contribute positively to the economy of the country in later years. Summerfield, Marshall and Archbold (1997) found that paediatric cochlear implantation could be acceptably cost effective within the range of the British health care system. It is hypothesised that paediatric cochlear implantation leads to short term benefits (audition, speech and language) within 1-3 years; medium term benefits (educational placement and achievement, social and quality of life issues) in the next 5 years; and long term benefits (employment and adult quality of life) in adulthood. However, “implantation remains an elective treatment of last resort for a chronic condition”, which “reduces disability without restoring function” (Summerfield & Marshall, 1999: p.143). Thus it is ranked lower in priority than treatments which save lives in a health care budget, and must be able to justify its cost. This justification in the form of a later saving to society is most likely to be realised for those children who are implanted very young.

### *2.3.2) Relevance to the South African context*

In South Africa, no state subsidising of the procedure is offered as yet. The result is that many children who would have benefited from the procedure are unable to, due to financial reasons. With the *HIV/AIDS pandemic* taxing the health budget, and increasing numbers of children and pregnant mothers being infected daily (Matkin,

Diedendorf & Erenberg, 1998), it is of importance to generate positive cost benefit data if state subsidising is to be justified, and considered by policy makers.

### *2.3.3) Research and future planning*

Considering the array of variables discussed which may affect the success of cochlear implantation in children as well as the new developments in the profiles of patients, surgical techniques and technology, the paramount need for any cochlear implant programme to remain current with ongoing research in this dynamic field becomes clear.

The other outcome variables will be discussed in chapters four and five, as they relate to the PCIP specifically.

## **2.4 Conclusion to Chapter Two**

The critical review of current literature on the variables affecting the success, as well as the outcomes, of both a child with a cochlear implant and the programme serves as a framework for the methods employed in the empirical study undertaken. The study uses this framework to guide investigation of the characteristics of the PCIP, as well as the interpretation of the research results.

## CHAPTER THREE

### RESEARCH DESIGN AND METHOD

#### 3.1 Aims of the study

The **main aim** of this study is the description of defining characteristics and outcomes pertaining to the children with cochlear implants (16 years of age and younger) in the PCIP and their families, as perceived by their parents/caregivers. This will be done in order to gain insight into some of the variables affecting the success of cochlear implantation in children in this particular programme and provide a foundation for the development of a research database for the particular cochlear implant programme.

The four **sub-aims** involve:

- A description of the children's pre- and perinatal, medical, developmental, audiological and educational history;
- The compilation of a descriptive profile of the families of children with cochlear implants;
- A description of the children's current outcomes as perceived by their parents in terms of educational, audiological and communicative functioning with the cochlear implant;
- The identification of variables affecting the success of the child with a cochlear implant in the PCIP.

These sub-aims relate to the proposed content of the questionnaire, and results and implications are systematically discussed in chapters 4 and 5.

### **3.2 Research design**

The study is a descriptive or observational survey, since the aim of the survey method is to describe and produce information on the population being studied (Fink, 1995a). Thus, no control group is used since the study does not aim to compare different groups. The study is cross-sectional as it provides descriptive data at one fixed point in time, however much of the information will be attained retrospectively (Fink, 1995a; Leedy & Ormrod, 2001). Since the study involves mainly discrete data, from a fixed set of questions, it is a predominantly quantitative study (Bailey, 1994). The study is non-experimental, and no variables are manipulated, thus no dependent and independent variables exist (Leedy & Ormrod, 2001) and no causal relationships are explored.

It is also beyond the scope of the study to demonstrate relationships between the different sets of data gathered, although this may be a later application of the rich description rendered by the present type of study (Leedy & Ormrod, 2001) requiring a deeper level of analysis of the same data (Kritzinger, 2000). As in the study by Kritzinger (2000: p162), this descriptive, quantitative survey aims “to conduct a structured and in-depth inquiry into the characteristics of the clients enrolled in a particular programme”. The study will use only one uniform source of data gathering namely the questionnaire. The study utilises an approach grounded in the principles of ECI, therefore parental perceptions are sourced in order to emphasis the parents’/caregivers’ views regarding the PCIP.

### **3.3 Participants**

The participant selection criteria and selection procedures, as well as a description of the participants are presented in this section.

#### *3.3.1) Participant selection criteria*

These criteria separate those who are eligible for the study from those who are not. In this case the target group is the children enrolled in the PCIP and their families. No random sampling procedures were used to eliminate the problem of bias in participant selection (Leedy & Ormrod, 2001), instead non-probability convenience sampling was used. Therefore the results of the survey can only be generalised to the target group (Fink, 1995c) and not to the wider population of children enrolled in Cochlear Implant Programmes in South Africa since this would lead to distorted results. Since the study will attempt to collect data on as many of the children enrolled in the programme as possible the participant selection criteria are not restrictive in order to ensure larger volumes of data, as is desirable for the building of a data-base (Kritzinger, 2000). The selection criteria are presented in the following table, Table 4.

**Table 4: Participant Selection Criteria**

<b>Participant selection Criteria</b>	<b>Description of criteria</b>
<ul style="list-style-type: none"> <li>• <b>Age</b></li> </ul>	Participants must have children with cochlear implants, up to a maximum age of 16.
<ul style="list-style-type: none"> <li>• <b>Parent/guardian's language and literacy level</b></li> </ul>	The questionnaire is written in English and in Afrikaans. Adequate English/Afrikaans proficiency to complete the questionnaire is thus required of participants (parents/caregivers) completing the questionnaire.
<ul style="list-style-type: none"> <li>• <b>Geographic location</b></li> </ul>	Participants must be residing in an area in South Africa that renders them accessible by post and telephone to participate in the study. This is a practical criteria to conserve resources (Fink, 1995b).
<ul style="list-style-type: none"> <li>• <b>Cochlear Implantation</b></li> </ul>	Children must have received a cochlear implant, implanted by the Pretoria Cochlear Implant Team.
<ul style="list-style-type: none"> <li>• <b>Restriction to PCIP</b></li> </ul>	Participants' children must currently be enrolled in the PCIP.
<ul style="list-style-type: none"> <li>• <b>Willingness to participate</b></li> </ul>	Participants must be willing to participate in the study, and must have signed a written informed consent form.
<ul style="list-style-type: none"> <li>• <b>Knowledge of participant</b></li> </ul>	The caregiver completing the questionnaire should have had adequate knowledge of the child's history in all areas of the questionnaire, including prenatal and birth history, or have been able to make contact with the child's biological mother to gain this information. This pertained especially to children who may have been in a foster care situation. Section B of the questionnaire could only be filled in by, or with information from, the child's biological mother since it covers the history of pregnancy and birth.

### 3.3.2) Participant selection procedures

The research proposal and proposed questionnaire were submitted to the Research and Ethics Committee of the Faculty of Humanities, University of Pretoria, as well as to the Head of the PCIP. Permission was thus sought from the ear, nose and throat specialist in charge of the programme, to collect and use the participants' data for research purposes. His conditions as well as those of the Research and Ethics Committee were taken into account before undertaking the study (see letter of approval in Appendix B). The register containing the names and telephone numbers of the children enrolled in the Programme was requested, and parents were then

telephonically contacted to determine willingness to participate in the study and suitability as participants, according to the participant selection criteria listed in Table 4.

### *3.3.3) Description of participants*

Participants included the parents of all children with cochlear implants, up to 16 years of age, on the records of the PCIP as well as their families, provided the parent/s or caregivers could be contacted and were willing and able to complete a questionnaire. The respondents to the questionnaire were the parent/s or primary caregivers of the children included in the study, and thus the participants.

Only 71 of the 73 parents and children enrolled in the programme and their families qualified as participants in the study, since families two resided in remote geographic locations and were inaccessible by post. In order to draw accurate inferences about the population of children in the programme, it was desirable that all or most of this group partake in the study. However, it must be considered that the return rate of questionnaires is often low (Leedy & Ormrod, 2001) and that concerted efforts to motivate questionnaire returns should be undertaken with caution as voluntary participation must also be ensured for ethical reasons.

Of the 71 questionnaires which were sent out, 45 were returned. A description of these 45 participants and their children is given in Table 5. In general, as seen in Table 5, most of the children in the PCIP are white, Afrikaans speaking children of either gender, living with both parents and at least one sibling. The children were more often the youngest child, and spent the majority of their time with their mothers.

A maximum of 2 languages was spoken in the homes (with the exception of one home in which three languages were spoken).

Children were between 2 and 16 years of age, and were as likely to fall in the categories 2-6 years, 6-12 years or 12-16 years. A wide distribution of ages was thus seen in the participants. No children were under the age of two at the time of data collection. Regarding the families, the majority of parents were employed, with fathers/paternal caregivers employed in all families except one. Most of the families lived in Gauteng, and were thus relatively close to the PCIP for access to services.

The participants of the questionnaire were the child's biological mother in all cases except one, a mother of two children with cochlear implants in the PCIP, where the child's grandmother was the participant in consultation with the mother.



**Table 5: Description of participant, child and family characteristics (n=45)**

<b>Characteristic</b>	<b>Value, description or percentage</b>
Participant who completed the questionnaire	98% biological mother 2% grandmother assisted by mother
Residence: province	Gauteng province: n=39 (86%) Eastern Cape: n=1 North West province: n=3 Northern Cape: n=1 Mpumalanga: n=1
Languages spoken in home	English, Afrikaans, Shona, South African Sign Language, Zulu, Tswana, Sotho Maximum number of language spoken in a home was 2, except one home where 3 were spoken.
Main language	20% English 73% Afrikaans 2% Shona 5% South African Sign Language
Population group of participants and their families	White: n=41 Black: n=2 Indian: n=1 Coloured: n=0
Ages of children	0-2 years: n= 0 2-6 years: n=15 6-10 years: n=11 10-12 years: n=5 12-16 years: n=14
Gender of children	Female: n=22 (49%) Male: n=23 (51%)
Parents' marital status	76% married 24% divorced, separated, or one parent deceased (one family)
People other than nuclear family members living in the house	20% grandparents 7% another child (not a sibling)
Person with whom majority of time is spent	65% with mother 27% school: residence or aftercare centre 8% grandparents or siblings
Maternal and paternal parent/caregiver employment	75% maternal parent/caregiver employed 98% paternal parent/caregiver employed
Birth order of child with cochlear implant compared to siblings	64% youngest child 2% only child 34% eldest child
Number of siblings living with family	73% lived with at least one sibling Maximum number of siblings: 4

### **3.4 Apparatus and materials**

#### *3.4.1) Material for data collection*

Data was collected using a parent questionnaire compiled by the researcher, with input from other members of the Pretoria Cochlear Implant Team. The questionnaire consisted of discrete items and was divided into five sections over twelve pages covering the areas of biographical information, prenatal and birth history, medical history, developmental history, family history, audiological history and current functioning of the child post-implantation. The questionnaire was available in both English and Afrikaans, depending on the language preference of the participant. A copy of the questionnaires is provided in Appendix C. With the assistance of a statistician, a coding column was provided on the right hand side of each page for coding of answer categories. Since the questionnaire is extensive, question types were chosen for ease and speed of answering.

Along with the questionnaire parents received a covering letter outlining the aims of the study and the possible value thereof. The covering letter informed parents of the ethical guidelines followed in the study, including the confidentiality of the information they supply and their right to withhold information and withdraw from the study at any time. The letter informed parents/guardians of their role and level of involvement in the study and requested their participation. A consent form, to be signed by the parent/s or caregiver completing the questionnaire and returned with the completed questionnaire, was also included and is attached as Appendix E.

The questionnaire serves to operationalise the aims of the study, or link the key concepts in the problem statement to the actual phenomena being studied (Mouton,

1996). The questionnaire serves both as a data collection tool and a measuring instrument (Leedy & Ormrod, 2002).

#### *3.4.2) Apparatus for data recording and analysis*

Data was organised using manual coding strategies for each answer to questionnaire items, after which a Microsoft Excel spreadsheet was used for ease of processing. A computerised system for the analysis of descriptive statistics, namely the *SAS*® (Statistical Analysis System, 1999) was used to provide frequency and percentage information as in the study of Evens (2002).

#### *3.4.3) Justification for use of a questionnaire*

The aims of the study as well as the time and financial constraints in the particular situation point to the need for a data collection instrument that can collect large volumes of both quantitative and qualitative data from a large number of participants who are spread over a wide geographic area, in the shortest time span and the most standardised way possible (Leedy & Ormrod, 2001). The particular population under study was unique in that they are generally a highly motivated group of people, who are familiar with the institution conducting the research project and who make regular contact with the institution for maintenance of their cochlear implant devices. The nature of the information required was detailed, sometimes requiring that respondents consult their own previous records, and was in some instances of a sensitive nature.

With these specific criteria in mind, consultation of literature and previous studies of a similar nature indicated a questionnaire to be the data collection tool of choice.

A questionnaire allows for large volumes of data to be collected from many different sources simultaneously, at a single point in time (Bailey, 1994). It allows for easy standardization, since each respondent receives the same typed questions without the influence of the researcher, who may ask questions in a leading manner, express bias or prompt the respondent, so influencing the answers to questions (Leedy & Ormrod, 2001). A questionnaire offers both quantitative and qualitative data, since there is a fixed set of questions which are systematically coded (Bailey, 1994). It also allows for ease and speed of processing of a wide variety of data in one survey (Evens, 2002).

Since the nature of the information required in some instances is sensitive, such as questions regarding pregnancy (Section B) and family history (Section E), it was reasoned that questionnaires posted to respondents were the best option since such questions are often answered more truthfully and accurately without the anxiety-producing effect of the presence of the researcher (Leedy & Ormrod, 2001).

Due to the unique nature of the participant population, which is relatively well known to the researcher's colleagues, incomplete, ambiguous or unlikely answers could be verified using patient records or direct contact with patients during consultations for mapping sessions. This could help to cancel out the nullifying effect of incomplete answering of questionnaires on the data (Neuman, 2000). Respondents' contact details were also available to the researcher. For the same reason, ambiguities in the questionnaire could be cancelled out largely by the use of pre-testing via a pilot study (Leedy, 1993) with a respondent whose information was already well known to the researcher who had been working with them for some time. In this way, the degree to

which answers reflect understanding of the questions can lead to alterations in questionnaire items in terms of consistency, relevancy, wording, and format; and the time and degree of effort taken to complete the questionnaire could be assessed (Leedy, 1993).

Another criteria that the chosen data collection tool needed to fulfill was the remote geographic location of some of the respondents. An advantage of postal questionnaires is that it eliminates the distance-factor, allowing data to be collected from “people thousands of miles away whom the researcher may never see” (Leedy, 1993: p.187).

Financially, the study would not have been viable within the limited budget available if the survey was conducted telephonically. The questionnaire is long and there were 71 possible respondents. Travel costs would also have been high if the survey was conducted personally due to the large number of respondents and their wide geographic distribution.

Time, too, would have been a factor since many of the questions require that the adult observe the child’s listening behaviour in specific situations before answering, or locate records of information which may have been forgotten regarding birth (Section B) and developmental history (Section D). Some questions also required that other family members be consulted (Section E regarding family history), and the logistics of personally interviewing each member would thus have been difficult.

The low response rate generally cited for questionnaires (Leedy & Ormrod, 2001) could be addressed in this study in a variety of ways. Firstly, familiarity of the respondents with the institution where the research is being conducted and sometimes with the researcher, may have increased motivation to return questionnaires. Secondly, the nature of the population was a positive factor since it may be reasoned that inherent in their choice of a relatively new technology (cochlear implants) could be an attitude conducive to a higher return rate if it is made known to them that ongoing improvement and refinement through research is the goal.

As Leedy (1993: p.190) states: “The covering letter is all-important”. A covering letter stating the rationale for the study, as well as a return-addressed envelope and follow-up phone-calls or reminders during consultations with the Cochlear Implant Team were expected to influence the return rate favourably. The nature of the questionnaire is also a factor, therefore the majority of responses were yes/no answers to precise questions, which are easy and quick to answer. It should be noted that respondents were encouraged to participate but not pressurized, as voluntary participation is essential to ethical research (De Vos, 2002).

According to Leedy (1993: p.187): “Data can sometimes be buried deep within the minds or within the attitudes, feelings or reactions of men and women. As with oil beneath the sea, the first problem is to devise a tool to probe beneath the surface.” In this case and in accordance with the specific criteria set, the obvious choice of a data collection tool was the questionnaire.

3.4.4) *Content and compilation of the questionnaire*

Fink (1995b) warns that surveys should only contain questions that are pertinent to the survey's objectives. The questionnaire's content was compiled according to each objective (sub-aim) using various sources from various fields. The tool drew questions from the CHRIB case history form (Louw & Kritzinger, 1995) used by the Centre for Early Intervention in Communication Pathology at the University of Pretoria, as well as other questionnaires used at the Department of Communication Pathology, University of Pretoria. Further, the questions and justification for questions were influenced by various sources in the fields of early communication intervention (Rossetti, 1996), neonatal intensive care (Merenstein & Gardner, 1998), paediatric cochlear implants (Clark, *et al.*, 1997), assessment in childhood communication disorders including hearing impairment (Katz, *et al.*, 2002), and family assessment (Kritzinger, 2000). The format of the database being compiled by the Tygerberg Cochlear Implant Programme was consulted for comparative and evaluative purposes, and the questionnaire adapted to include information in this tool. The Tygerberg programme has been involved in paediatric cochlear implantation for a substantial time period and thus has experience in the field. The PCIP may be partly basing their computerised database on the client database programme compiled by the Tygerberg Cochlear Implant Programme, thus it follows that areas relevant for the database should be covered in the questionnaire if it is to be a useful tool for the future. The questionnaire's structure was compiled using various sources in the field of research, survey research and social research (Fink, 1995b; Leedy & Ormrod, 2001; Mouton, 1996). Other questionnaires on a masters level in the field of Communication Pathology were consulted to aid the development of an appropriate format (Evens, 2002; Gopal, 1999).

Since the questionnaire was reasonably lengthy, attempts were made to pose questions in the simplest/fastest response format as far as possible, with the emphasis on closed ended questions (Leedy & Ormrod, 2001). A logical flow and order of questions was used, with efficient spacing, unambiguous language and brief, clear instructions as well clarification of terminology (Bailey, 1994; Leedy & Ormrod, 2001; Neuman, 2000). After revision of the questionnaire following the pilot study, it was possible to give an estimation of the time taken to complete the questionnaire. Only questions that were answerable by the parents were asked.

#### 3.4.5) *Structure of the questionnaire*

The first section of the questionnaire involves *biographical information* pertaining to the child's family circumstance. Questions were chosen to render information about the socio-economic status of the family and the level of education of the people providing stimulation to the child (Dollaghan, *et al.*, 1999), as well as the family structure. The geographic area in which the family resides, as well as the number of people residing in the home, the number of siblings a child has and the employment status of the child's parents gives a further indication of socio-economic circumstances and the level of geographic and financial accessibility to support services. These questions, as well as those in the next four sections, were used to fulfill the need for information regarding the factors influencing the success of a child with a cochlear implant, visually depicted as the upper section in Figure 1. Identifying information, such as names of parents, addresses and telephone numbers serve to verify the patient records of the Programme. The language spoken as well as the number of languages spoken in the home were items included to inquire about ethnic origin of the family (Zhang & Bennett, 2001), and about the possibility of



many languages creating language confusion in the child. Where the nature of the information is sensitive, it was attempted to use subtle wording. This section necessitated the use of open questions as well as the preferred closed-ended questions, which can be answered by ticking appropriate blocks.

The next section, covering the *history of pregnancy and birth history*, was categorised into sections allowing parents to whom this was uneventful to move on quickly to the next section, and parents for whom this information is sensitive to provide as much or as little information as they wish. This section was included since many causes of hearing impairment have their origin in the prenatal period or with birth trauma (Merenstein & Gardner, 1998). A question inquiring about maternal concern during pregnancy is included to allow mothers to provide information about use of drugs or alcohol at their discretion (Rossetti, 1996). More specific, closed ended questions regarding the child's condition after birth were included in table format to cover all the possible risk factors for hearing loss and developmental delay in this time period (Kritzinger & Louw, 2000). Specific questions were asked here since open questions would not prompt parents to provide the amount of detail required.

The *medical history* of the child, including a filter question regarding hospitalisation as well as a table listing conditions associated with sensorineural hearing loss, additional disabilities and developmental delay influencing success with a cochlear implant, comprises the next section (Blair, *et al.*, 1995; Briscoe *et al.*, 1998; Moehn & Rossetti, 1996; Robison & Stewart Gonzalez, 1999; Rossetti, 1996). Only the specific information deemed relevant was included in this section.

Next, the *developmental history* of the child is covered in a table detailing developmental milestones, a filter question regarding the development of feeding and a closed set question judging parent's perceptions of their infants' development prior to onset of hearing loss as normal or abnormal. This question was included to attempt to control for developmental progress in the child post-implantation.

The *family history* surrounding the child is documented next, with emphasis on the presence of a hereditary component to the child's hearing loss (Angeli, *et al.*, 2000; Matsushiro, *et al.*, 2002), as well as to other disabilities which may affect progress with a cochlear implant such as learning disability or genetic syndromes (Pisoni, 2000). A question regarding parental membership to the Deaf Community was included to assess the possible influence of signing in the home on the child's mode of communication and acquisition of verbal language (Archbold, *et al.*, 2000).

The child's *audiological history* prior to and after cochlear implantation is then questioned by closed ended questions regarding suspected cause of hearing loss, diagnosis, nature of the hearing loss and period and nature of hearing aid fitting. The section moves on to include questions regarding funding (Severens, *et al.*, 1997), age at implantation (Snik, *et al.*, 1997), the type of professional by whom the referral was made and the ear chosen for implantation. The use of a hearing aid on the non-implanted ear (Ching, *et al.*, 2001) and use of FM systems is explored using yes/no type questions. These questions were chosen to gain information regarding the

variables influencing the success of the child with a cochlear implant, as illustrated in Figure 1.

The last section of the questionnaire uses categorical type multiple choice questions arranged in tables of options to describe the child's *current educational setting, use of his or her cochlear implant, level of communication, audiological development and the level of speech intelligibility attained by the child*. This section relates to the perceived outcomes of the child with a cochlear implant depicted in Figure 1. The scales used in the questionnaire for rating audiological development and the child's attained level of speech intelligibility were respectively adapted and included the Lip Profile (Listening Progress Profile) of Environmental Sounds (Archbold, 1993) and the SIRS Speech Intelligibility Rating Scale (Allen, Nikoloploulos & O'Donoghue, 1998). Further questioning involves the support services and rehabilitative therapy available to the child. The use of a telephone and the ability to enjoy music are the last questions of the questionnaire. The parents are then thanked for their co-operation.

The great majority of questions are of a categorical type, where the response is 'yes/no' (Fink, 1995b). Where more information is required where applicable to a certain child, the filter question type and subsequent response option of 'if yes, please specify' is included. In this way questions are kept closed ended as far as possible, while the option of providing more information is given (Fink, 1995b). Other closed-ended categorical type questions are presented in tables of options, where the respondent is instructed to 'tick one' or 'tick all applicable' (Fink, 1995b). The

‘other’ option is given in these cases to provide for the possibility of an option not accounted for by the researcher, thus making the options inclusive and exhaustive (Fink, 1995b). Scales used are of a nominal or interval type, with discrete values or categories to minimise ambiguity. For the same reason no attitude scales were included (Leedy & Ormrod, 2001).

The English and Afrikaans questionnaires are included in Appendix C.

### **3.5 Procedures**

The procedures of the pilot study and main study are presented in the following section.

#### *3.5.1) Ethical considerations concerning the research*

Researchers have an obligation to their research participants to maintain the highest possible levels of confidentiality regarding their personal information (Fink, 1995b). Since names were attached to the questionnaires, it was important that the questionnaires be read only by the researcher and her study leaders, and that this information not be shared with people other than those named. Permission was also granted by each participant to have access to their information, as well as from the head of the PCIP. In agreement with the recommendations of Leedy & Ormrod (2001), a letter of informed consent was sent to each parent involved in the study to confirm voluntary participation and to ensure that parents understand the nature, purpose and methods of the study as well as their role and level of involvement (see Appendix D). Parents were required to sign and return a consent form accompanying

the letter and questionnaire (see Appendix E). Parents will also be able to contact the researcher for access to the results of the study. The right of parents to withhold information or to withdraw from the study without consequences at any stage was made known to them. Parents were informed that their withdrawal from the study would in no way influence the quality of the service provided to them by the PCIP.

During statistical analysis of the data numbers were allocated to each questionnaire, and were no longer related to names. Dissemination of the results of the analyses did not contain any names. Should the information be fed into a data-base, it will out of necessity be attached to names and contact details. Access to the database, however, will be limited to members of the PCIP, and the person appointed to enter the data. Access will be via a password for each staff member.

The approval of the Research Ethics Committee of the Faculty of Humanities, University of Pretoria, was obtained prior to commencement of the study and their criteria for ethical conduct of the study after inspection of the research proposal and questionnaire were followed. Participants were not misled in any way, and data was recorded, analysed and reported in an honest and transparent manner (De Vos, 2002).

### *3.5.2) Reliability and validity*

According to Fink (1995a: p.4), “a reliable instrument is consistent; a valid one is accurate”. A questionnaire is reliable if data obtained from it does not vary as a result of the measuring instrument itself (Neuman, 2000). Fink (1995a) gives a checklist of the following factors. To help ensure **reliability** and a uniform set of responses, a **time limit** should be set, in this case in the form of only children currently enrolled in

the Programme. Reliability is a pre-requisite for measurement validity (Neuman, 2000). To help ensure *internal validity* selection criteria took note of the factor of *history*, thus only children who were a part of the PCIP since implantation were chosen. This enabled conclusions to be drawn about the population and aided *external validity*.

The issue of *instrumentation* was addressed by using the same questionnaire with all respondents, and not involving the researcher in the actual completion of the survey. The problem of *attrition*, or loss of data due to incomplete answering of survey items, is often found in this type of research design (Neuman, 2000). Via the use of simple response choices, answerable questions, user-friendly format, a terminology list and evaluation by a pilot study, attrition was minimised. It was hoped that all members of the target population would partake in the study. Since the study aimed to describe only this population, external validity would then be implied and no generalisations to a larger population could be made.

Reliability and validity of the research were also improved by the use of a pilot study and subsequent revision of the survey. The nature of a postal survey may be a threat to internal validity, since the generally low response rate may create distortion of results due to bias as the respondents who answer may not be representative of the population (Bailey, 1994; Leedy & Ormrod, 2001). Employing strategies to increase the response rate of participants can combat this threat. Another threat to internal validity would be a questionnaire which was not sufficiently grounded in the appropriate literature so as to be suited to the aims of the study. To address this issue,

a wide literature base was used and other relevant studies were examined (Evens, 2002; Gopal, 1999; Kritzinger, 2000).

*Face validity* of the questionnaire was enhanced by using a covering explanatory letter, structuring the format of the questionnaire in a logical way, and using appropriate language and terminology lists. All of these strategies were then tested by a pilot study. *Content validity* was addressed by using a wide and thorough literature base to justify and compose relevant, comprehensive questions. *Criterion validity* was addressed by comparing the instrument to the questionnaires used by other cochlear implant programmes. *Construct validity* required that the questionnaire contain only questions pertinent to the study's objectives and no vague, leading or biased questions (Leedy & Ormrod, 2001; Mouton, 1996; Neuman, 2000).

### 3.5.3) *Pilot study*

- *Aims of the pilot study*

The aims of the pilot study were refinement and adjustment of the structure and content of the questionnaire based on pre-testing. The time and effort taken to complete the questionnaire, as well as the ease of understanding and use, clarity, consistency and relevancy of the questions were assessed (Leedy & Ormrod, 2001). Ambiguities and unfamiliar terminology will be identified. This aided in enhancing the reliability and validity of the questionnaire as a data collection instrument.

- *Participants of the pilot study*

The participants included the biological mother of one child with a cochlear implant, who was slightly above the maximum age defined for participant selection in this study. The researcher is familiar with the participant and the child. The rationale behind the selection of this particular participant was the researcher's prior access to their information, enabling the researcher to compare the participant's answers to existing data gathered over the course of the researcher's contact with the participant. Answers could thus be better judged as reflecting understanding or misunderstanding of the intended meaning of the question items, fulfilling the aim of the pilot study. In addition, due to the age of the child, the problem often encountered in retrospective studies of loss of data to memory could then be assessed (Leedy & Ormrod, 2001). The questionnaire will take longer to be completed by the parents of older children in whose cases more information is available to be filled in, thus the time aspect of the questionnaire can be judged. The participant was given the choice to participate in the pilot study of own free will and complied with all participant selection criteria set for the main study, besides the age of the child (see Appendix A). A child who was too old to be included in the main study was chosen, since participation in the pilot study would have excluded a respondent from participation in the main study.

- *Materials of the pilot study*

The full questionnaire was used, along with an evaluation form in order to guide assessment of each section of the questionnaire. The areas of evaluation included content of questions, format of questionnaire, wording and user-friendliness. Relevance, consistency, brevity, clarity and appropriateness of questions, as well as the degree to which they can be answered based on parent knowledge were also evaluated. A personal interview with the participant then rendered additional



criticisms, suggestions and comments as well as reactions to questions of a personal/sensitive nature.

- *Procedures of pilot study*

The participant was contacted telephonically to request participation in the study. A questionnaire with a covering letter stating the purpose of the pilot study as well as the main study was then be given to the participant, along with an informed consent form. Attached was an evaluation form. The participant was contacted telephonically after one week to arrange a feedback session, and the questionnaire was collected from her prior to this session and analysed by the researcher. During the feedback session, relevant questions were asked regarding the answering of the questionnaire, and additional feedback was requested of the participant. The participant of the pilot study was not included in the main study.

- *Results of pilot study*

Valuable comments were made by the participant, which aided refinement of the questionnaire for the optimal collection of accurate information for the study. Alterations were made to the questionnaire based on the commentary supplied by participants of the pilot study. A statistician was then consulted again to aid in the changes in format.

Changes included re-organisation of the format of some of the questions, where too much information was requested to be filled into a table. In these cases the questions were divided into two or more questions, with separate tables for entry of information by parents. In addition, clarification of terminology was included in two questions

and a rating scale was included rather than a yes/no response for a question involving the manner in which the child communicates. Salient aspects of three questions were underlined, since these were overlooked by the respondent. Three questions were edited in favour of simpler language, with the use of examples in brackets. The time taken to complete the questionnaire was not found by the respondent to be excessive (20-30 minutes) and she reported that she did not find any questions offensive or of a sensitive nature. The respondent did not feel that any information requested was unnecessary, or that an area had been overlooked. She reported feeling positive about the questionnaire and the study. The respondent mentioned that she had not been able to recall exact ages at which developmental milestones were achieved by her child, but that she had entered approximate ages. This can be expected where participants are older. All questions were answered. The revised questionnaire was used for the main study.

### **3.6. Data collection procedures**

Data was collected via a comprehensive questionnaire filled in by the parents/care-takers of the children in the PCIP. Questionnaires were posted to all families as well as postage-paid, return-addressed envelopes and informed consent letters. Preceding posting of questionnaires, the families were contacted telephonically. This initial phone call served as a means to explain the nature of the study and its ethical issues, request permission to use data and determine willingness to partake in the study. Follow up phone calls three weeks after posting the questionnaires served as reminders to those respondents who had not yet returned their questionnaires. This time period allowed one week for postage time in either direction, as well as one week for completion of the questionnaire.

### **3.7. Data recording/preparation and analysis procedures**

A coding column was included in each questionnaire. Each questionnaire also had a number for ease of processing. Responses to questionnaire items were coded manually by the researcher, with the guidance of a statistician. A codebook was created, as advised by Fink (1995c), containing the number (code) for each variable, its location in the survey, a name for the variable and a brief description of its meaning. This will facilitate later use of the questionnaire for further outcome studies by different researchers. Coded data was entered into a Microsoft® Excel spreadsheet.

Analysing data involved two steps: firstly reducing the wealth of data into manageable proportions, and then identifying patterns or themes in the data (Mouton, 1996). Open questions were analysed and categorised according to themes (De Vos, 2002). Computer coded data which had been entered into an Excel spreadsheet for scanning of trends, organisation of data and ease of analysis was then quantitatively analysed using the SAS® (1999), a system of descriptive statistics in order to render quantitative information regarding the areas named as sub-aims of the study. Both dichotomous and categorical nominal (e.g. in biographical information), as well as ordinal and numerical measurement scales were used due to the variety of information being gathered. For the same reason, both measures of central tendency and of dispersion were used to provide the rich description required of the data. Frequency counts were widely utilised (Fink, 1995c). These will serve as a useful means of disseminating outcomes of the PCIP as described by the findings of the study, in the form of graphs and charts as demonstrated in the outcome studies of other

programmes (e.g. Birmingham Paediatric Cochlear Implant Programme Outcomes report 1990-2000; Nottingham Paediatric Cochlear Implant Programme Progress Report 1997).

## CHAPTER FOUR

### RESULTS AND DISCUSSION

#### **4.1 Introduction**

In this chapter the findings of the study will be presented and discussed according to the sub-aims. Where appropriate, the sample will be divided into two groups, namely those participants attending specialised schools and those in inclusive educational settings, and a comparison will be made between the groups. This will be used to aid the search for variables which could contribute to the success of a child with a cochlear implant in terms of the auditory, linguistic, everyday functioning and academic skills considered by parents and professionals when deciding on the choice of educational placement. As described in the first chapter and summarized in Figure 1, a number of audiological, biological and environmental factors affect the outcomes of child with a cochlear implant, and the family unit is the key to understanding the development of the child. For this reason the variables relating to the families of participants will be discussed in detail, in addition to variables directly related to the children in the study. The outcomes of children, as measured by parental report, will be presented and expanded on by comparison to outcomes of other programmes, and by linking these outcomes to various possible contributing factors in the children's histories.

The discussion begins with an overview of the number of children referred for cochlear implants (who were successful candidates and received cochlear implants) over the years 1992-2003, presented to guide the reader in the historical spread of data collection. Although it was a cross sectional study and data was collected at the same

time from all participants, a large chronological difference exists between data provided by various participants.

In accordance with the first sub-aim, namely a description of the children's prenatal; perinatal; medical; developmental; audiological and educational history, the background surrounding the participants' hearing impairment including the role of healthcare professionals at that stage is discussed. Factors related to age at diagnosis, fitting of amplification and cochlear implantation as well as funding issues are highlighted. This is followed by a breakdown of the age at implantation of the participants in the study. A discussion of biological and medical factors continues onto the cause and nature of hearing loss. The discussion concerning sub-aim one continues with various audiological variables, including side of cochlear implantation; the use of hearing aids on the non-implanted ear and bilateral cochlear implantation.

Following this, results relating to family histories of disorders; level of parental education; parental employment and the family's socio-economic status are explored in order to ascertain the role of environmental factors. Parental and familial variables are discussed in accordance with the second sub-aim, namely the compilation of a descriptive profile of the families of children with cochlear implants.

To fulfill the study's third sub-aim, namely a description of the children's current perceived outcomes in terms of educational, audiological and communicative functioning, a comparison is then made between numerous variables and outcomes pertaining to participants in inclusive education and those in specialised education. This discussion is taken further with a comparison of the medical histories of these

two groups, in order to bring to the surface any biological variables and co-existing conditions which could explain the difference in outcome of the two groups. Next, the auditory and linguistic outcomes of the participants are explored under the headings of use of the device; auditory performance; musical appreciation and mode of communication including language level to complete the third sub-aim of the study.

The fourth and final sub-aim entails the identification of variables affecting the success of the child with a cochlear implant in the PCIP, and serves as a summary of the various factors explored. A short list is provided of the variables found to be related to educational placement in an inclusive setting, which necessarily implies a high level of speech perception, production and language development.

Finally, the chapter ends with a critical discussion of the use of high risk registers for identifying risk factors in neonates as they relate to hearing impairment; family history data and the group of children with acquired hearing impairment. This is seen as a logical product of the study, and one which holds implications for the promotion of neonatal hearing screening in South Africa as opposed to the use of high risk registers.

Of the 45 questionnaires returned most included full data sets, however, where the number of respondents is not 45 in the results, questions were omitted. This was most frequently the case where parents could not remember the information asked, such as specific dates, ages or scores.

## 4.2 Presentation and discussion of results.

In the following section the results of the study will be presented, graphically represented and discussed. To give a perspective of the time frame of data collection, the number of participants of the study who received their cochlear implants in each respective year from 1992 to 2003 is given in Table 6.

**Table 6: Implant rate over years: time of referral to CI programme (n=37)**

<b>Year</b>	1992	1993	1994	1995	<b>1996</b>	<b>1997</b>	1998	1999	2000	<b>2001</b>	<b>2002</b>	2003
<b>No. of participants</b>	2	2	1	3	<b>4</b>	<b>4</b>	1	3	3	<b>4</b>	<b>9</b>	1

According to Table 6, In the 11 year period represented by these figures, 2002 stands out as the year in which most participants to this study were successfully referred to the PCIP, i.e. were referred and received cochlear implants. Only 14% were referred prior to 1995, with the figure rising to 30% for the next three year period (1995-1997), decreasing slightly to 11% for the third period (1998-2000) and increasing again to 38% for the fourth 3-year period from 2001-2003. Discussion with a member of the PCIP revealed that changes in medical aid schemes and funding issues had an effect on implant rate over the time span. Another possible explanation is that the researcher is familiar with a large number of the families whose children received cochlear implants in 2001 and 2002, possibly providing a favourable influence on the return rate of questionnaires from these families. For full analysis of these trends the entire population of paediatric cochlear implantees in this time span should be used as the present study can only report on data relating to respondents.



In the following section, the four sub-aims stipulated in the methodology are repeated with the results pertaining to each sub-aim put forward and discussed under the relevant subheadings.

*4.2.1) Results and discussion relating to Sub-aim One*

<p><b>SUB-AIM 1: A DESCRIPTION OF PARTICIPANTS' CHILDREN'S PRENATAL, PERINATAL, MEDICAL, DEVELOPMENTAL, AUDIOLOGICAL AND EDUCATIONAL HISTORY</b></p>
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In order to fulfill this sub-aim, the following factors were taken into account. They are described under the sub-headings named below.

- 4.2.1.1) Cause of hearing loss
- 4.2.1.2) Nature of hearing loss
- 4.2.1.3) Biological and medical variables
- 4.2.1.4) History of diagnosis of hearing loss and intervention
- 4.2.1.5) Age at cochlear implantation
- 4.2.1.6) Side of implant
- 4.2.1.7) Hearing aid use on the non-implanted ear
- 4.2.1.8) Bilateral cochlear implants

## 4.2.1.1) Cause of hearing loss

The causes of hearing impairment operational in the sample under study are illustrated by Table 7. The causes are listed in order from most to least frequent. Maternal cytomegalovirus infection is abbreviated to CMV.

**Table 7: Cause of hearing loss in children (n=45)**

<b>Cause of hearing loss</b>	<b>Percentage of children (%)</b>
Unknown	17 (38%)
Meningitis	4 (9%)
Genetic syndrome associated with hearing impairment	4 (9%)
Prenatal complications (maternal infections or medical conditions of mother or foetus during pregnancy)	3 (7%)
Birth trauma (foetal distress, asphyxia)	3 (7%)
Hereditary hearing impairment	3 (7%)
Congenital Rubella syndrome	3 (7%)
Maternal CMV infection	2 (4%)
Viral infection	1 (2%)
Kidney disorder	1 (2%)
Chronic otitis media	1 (2%)
Ototoxic medication	1 (2%)
Medication ingested by mother whilst pregnant	1 (2%)
Myelin sheath disorder	1 (2%)
<b>TOTAL</b>	<b>45 (100%)</b>

The largest group, namely those in which the cause is unknown to the parents, is of particular interest since this may in part represent the non-syndromal genetic mutations (BJB2 gene) as described in the literature as being frequent in the Caucasian population (Matsushiro, *et al.*, 2002). Angeli, *et al.* (2000) report that there are at least 20 different mutations of this gene, caused by hereditary or disease processes; and that the numbers of these children in cochlear implant programmes warrant standard screening for this aetiology. The Joint Committee on Infant Hearing (JCIH) (2000) state that, in the near future, advances in knowledge regarding

recessive genes responsible for non-syndromic hearing impairment could dramatically reduce the number of children whose aetiology of hearing impairment is unknown.

Of the children with genetic syndromes (see Table 7), two had Waardenburg Syndrome and one had renal Fanconi syndrome which further affected the child's kidneys leading to a progressive hearing impairment. The fourth child had a syndrome which was not specified by the mother, who completed the questionnaire, but which included cleft lip and palate, profound sensorineural hearing loss and Mondini abnormality of the cochlea.

Closer inspection of the data of children who lost their hearing prenatally due to maternal rubella reveals that other aspects of their general development were also affected, and this group was largely educated in specialised settings. The group who acquired hearing impairments due to meningitis formed part of those in inclusive education most often, indicating that despite the possible additional effects of meningitis (Clark, *et al.*, 1997), the children in this study were mostly able to function in regular schools.

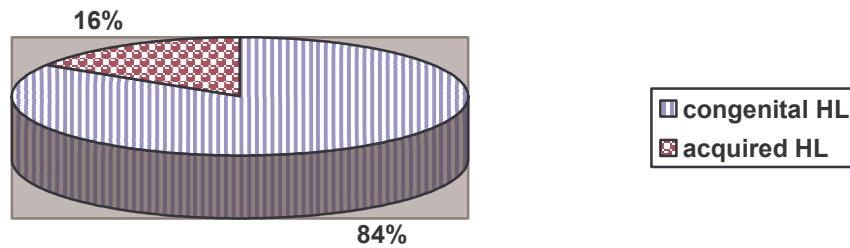
Of the children whose mothers contracted CMV during pregnancy, one participant's hearing loss followed the progressive course typical of CMV infection, while in the other participant it is not clear whether the hearing loss was progressive or not, although some degree of hearing impairment from birth is suspected by the mother. This child presented with additional disabilities resulting from a sensory integration disorder, requiring intensive therapy. He has good residual hearing in the non-implanted ear and benefits greatly from the use of a hearing aid on this side, which

leads back to the questions of shifting selection criteria for cochlear implantation, as well as the choice of ear to implant (Ching, *et al.*, 2002). (Children such as this one began to show significant benefit from the use of a hearing aid only after cochlear implantation in the ear with least residual hearing, as reported as additional information on the questionnaire by participants.) One could argue that stimulation of the auditory pathway and cortex following cochlear implantation, with time, better enabled the child to process the sound and auditory information received via the hearing aid (Ching, *et al.*, 2002). This may be an argument for implanting the ear with least residual hearing and recommending the use of a hearing aid on the non-implanted side. Further in-depth research on this topic would yield useful results.

#### *4.2.1.2) Nature of the hearing loss*

Only 16% of children in the current study had an acquired hearing impairment, while the hearing impairment was congenital in 84% of cases. Data revealed that 28% of children (including those with congenital and acquired hearing impairments) reported a progressive hearing loss, and in 7% of cases this was of a fluctuating nature. The reasons for the progression were the presence of renotubular Fanconi syndrome, hereditary hearing loss, unknown causes and CMV. The majority of children, with the exception of those affected by meningitis and perinatal factors, had congenital hearing impairments which emphasizes age at implantation as a crucial factor in language development.

### Nature of hearing loss



**Figure 2: Relative percentages of congenital and acquired hearing loss in children (n=45).**

Of the 7 children (16%) in Figure 2 whose hearing loss was acquired, 85% of parents reported that development prior to onset of hearing loss was normal, indicating the absence of early developing additional conditions such as a condition affecting cognition, motor or sensory development, social development or behaviour. Children with acquired hearing loss have the advantage of early exposure to sound and possibly some linguistic development, and generally develop language at a faster rate post-implant than children with congenital hearing loss (Katz, *et al.*, 2002).

These characteristics of the children added to the complexity and heterogeneity of the data, and imply an additional dimension to the planning, expectations and outcomes of cochlear implant programmes.

4.2.1.3) *Biological and medical variables*

Table 8 compares the medical histories of the participants' children with cochlear implants when they are divided into two distinct groups: those in inclusive education (IE), and those being educated in specialised settings (SE). Variables which may have affected this outcome, including co-existing disorders and conditions in addition to the hearing impairment, are described and discussed. Attention deficit disorder and attention deficit and hyperactivity disorder are abbreviated as ADD/ ADHD respectively and mentioned in one category.

**Table 8: Comparison of medical histories of children in inclusive and specialised educational settings.**

<b><u>Medical condition from parent questionnaire</u></b>	<b><u>Group in inclusive education n=19</u></b>	<b><u>Group in specialised education n=24</u></b>
Otitis media	15	12
<b>Childhood illnesses: measles, mumps, chicken pox</b>	<b>4</b>	<b>8</b>
Allergies	4	6
<b>ADD/ADHD</b>	<b>1</b>	<b>5</b>
Chronic upper respiratory tract infection	5	5
Physical/muscle disorder (condition affecting muscle tone, strength or co-ordination / mobility)	3	3
Genetic syndrome	2	2
Asthma	2	1
Disorder of vision/eyes	2	2
Meningitis	2	2
Kidney disorder	1(reflux: surgically corrected)	1
Behavioural problems	1	1
Chronic illness	0	1 (kidney disorder)
Epilepsy	0	0
Other	- Rota virus 1 - Balance problems 1 - Obesity 1	- High fever 2 - Reaction to inoculation 1 - Accidental poisoning 1

As seen in Table 8, the only significant difference highlighted by this comparison is the greater number of children in the SE group who suffered from ADD/ADHD

(reported by parents), namely 5% of the IE group as opposed to 21% of the SE group. This finding corresponds with that of Pisoni (2000) and Pisoni and Cleary (2003), calling for more emphasis in cochlear implant research on issues of information processing such as attention; memory; perception and learning to explain the individual differences in outcome after paediatric cochlear implantation. The group in IE also seemed to suffer from fewer bouts of general childhood illnesses such as measles, mumps, chicken pox etc., which could be interpreted to indicate a generally higher resistance to infectious illness and possibly better general immunity and health leading to fewer absences from school and missed learning opportunities. Both groups indicated that middle ear infections as well as upper respiratory tract infections were frequent occurrences, which would have a temporary negative impact on auditory functioning. The JCIH (2000) reported otitis media to have an especially negative effect in children with sensorineural hearing loss.

As depicted in Table 8, none of the children in the inclusive group suffered from a chronic illness, as opposed to one participant in the specialised education group. The severity of the disorders of vision, not indicated by these broad categories, revealed itself to be significant on closer inspection. The children in inclusive settings had mild visual acuity problems, corrected with lenses, while those in the second group had more severe visual complications including retinitis pigmentosa. The Birmingham Paediatric Cochlear Implant Programme (BPCIP) (2000) reports having provided cochlear implants to two children with blindness, and 5 with Usher's syndrome (resulting in retinitis pigmentosa and blindness). Slightly more cases of children with genetic syndromes are reported by the BPCIP in relation to the larger

number of children implanted (11%) of which only one case has Waardenburg syndrome.

It is interesting to note that the two groups in Table 8 (namely SE and IE) in the present comparison had equal numbers of children who had suffered meningitis, as well as children with a genetic syndrome indicating that in this analysis this was not a determining factor in school placement. This is in contrast to a study by Nikolopoulos, *et al.* (1997) who found that post-meningitically hearing impaired children with cochlear implants scored lower than their peers with congenital hearing loss on measures of the development of audition one year after implantation. However, the children in that study lost their hearing to meningitis pre-lingually, while some in the PCIP group had some early linguistic development prior to onset of hearing loss.

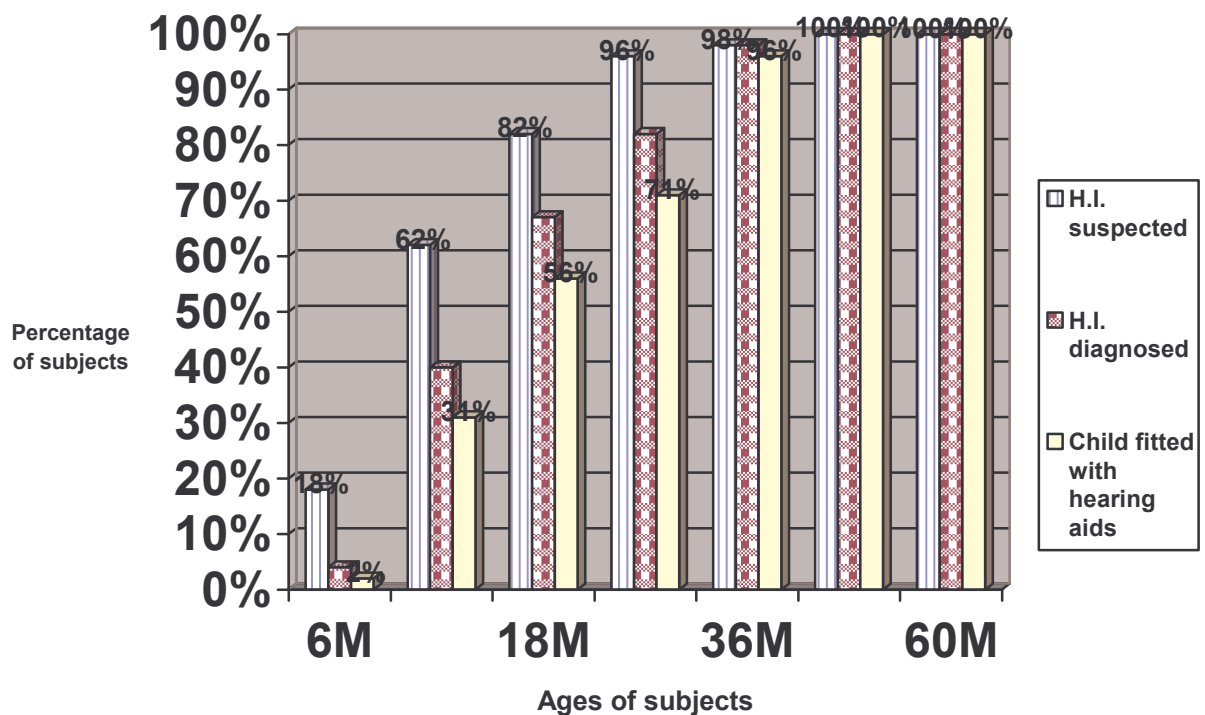
Asthma and allergies occurred in both groups, and can be correlated with the upper respiratory tract infections and otitis media. These factors may be climate and region-specific to some degree.

#### *4.2.1.4) History of diagnosis of hearing loss and intervention*

This variable is discussed in accordance with the first sub-aim of the study. As illustrated by Figure 5, hearing loss was generally suspected by mothers/parents by 12 months of age (62%), although 18% of parents suspected that their infants could not hear by 6 months. The percentage increases to until all children's hearing losses were suspected by 44 months. In 44% of cases it was the mother who noticed the child's lack of response to sound, in 42% of cases both parents, and in only 2% of children was it a teacher or therapist, although this may be due to the fact that prior to



diagnosis these team members were not yet involved. Hearing loss was never suspected or identified by the family doctor, paediatrician or nursing sister. Lemmer (2002) found that pregnant mothers in Pretoria, South Africa would not necessarily consider consulting a speech-language pathologist and audiologist first due to suspicion of hearing loss in their infants. The mothers in that study were unsure who to consult regarding their infants' hearing. A comparison of the percentages at each age group for suspicion of hearing loss, diagnosis and fitting of hearing aids for children with cochlear implants in the present study follows in Figure 3.



**Figure 3: Ages at which hearing loss was suspected, diagnosed and children fitted with hearing aids. (n=45).**

As depicted in Figure 3, while all children's hearing losses were diagnosed before 48 months and 98% before 36 months, only 40% of these mostly congenital hearing losses were diagnosed before 12 months. Only 4% were diagnosed by 6 months, a figure which lends support to the idea of compulsory hearing screening after birth as

implemented in the USA (Mayne *et al.*, 2000). In five cases, it was reported by parents through the addition of information not directly asked in the questionnaire, that doctors did not react immediately to mothers' concerns regarding their children's development, despite findings indicating the high reliability of parental report of concern (Rossetti, 1996). Although this result was not part of the questionnaire, it was decided to include the information as it may be relevant to later studies involving medical professionals and cochlear implantation, as well as being an indicator of the need for more widespread information on the identification and referral protocol of children with hearing loss.

Figure 3 illustrates a substantial delay between suspicion; diagnosis and treatment of hearing loss in the children, which decreases towards 36 months of age. At age 12 months, only 40% of children's hearing losses had been diagnosed while 62% were suspected. The role of the child's doctor or paediatrician at this stage, in referring promptly to an audiologist at the first sign of doubt or of parental concern, is crucial (JCIH, 2000) and should be targeted (Lemmer, 2002).

The slightly shorter but still important delay between diagnosis and fitting of hearing aids was found. This could be a result of the financial implications of fitting a child with bilateral hearing aids, capable of producing the large degree of amplification required for the degree of hearing losses of the children (98% were fitted bilaterally), or possibly of seeking second opinions regarding diagnosis and coming to terms with the diagnosis. Harrison, Roush and Wallace (2003) cited financial issues, problems with the scheduling of appointments, further testing including second opinions and the suspicion of auditory neuropathy as being the main causes for the delay between

diagnosis of hearing loss and fitting of hearing aids. Harrison *et al.* (2003) continue to emphasize the need for priority to be given by audiologists to appointments made for first-time hearing aid fitting of newly diagnosed infants and children, to speed up the process.

The 28% of children with progressive hearing loss, as well as the 7% whose hearing losses fluctuated (see 4.2.1.2), may also have affected these numbers as the time between the initial diagnosis of the hearing loss and the decision that the severity of the impairment warranted cochlear implantation, would be longer. Counseling by audiologists and when necessary, other team members such as psychologists, as well as prompt action by medical aid schemes is required at this stage to shorten the time between diagnosis and fitting of amplification. Referral to a cochlear implant programme at this early stage, when profound bilateral sensorineural hearing loss is diagnosed, would increase the chances of early implantation where appropriate candidacy is established. A shorter time delay between these three steps in the process prior to cochlear implantation would necessarily imply a younger age of implantation and should be a goal of healthcare professionals (Mayne *et al.*, 2000) and systems in South Africa.

A related question in the questionnaire asked who referred the family to the cochlear implant programme. The responses are presented in Table 9.

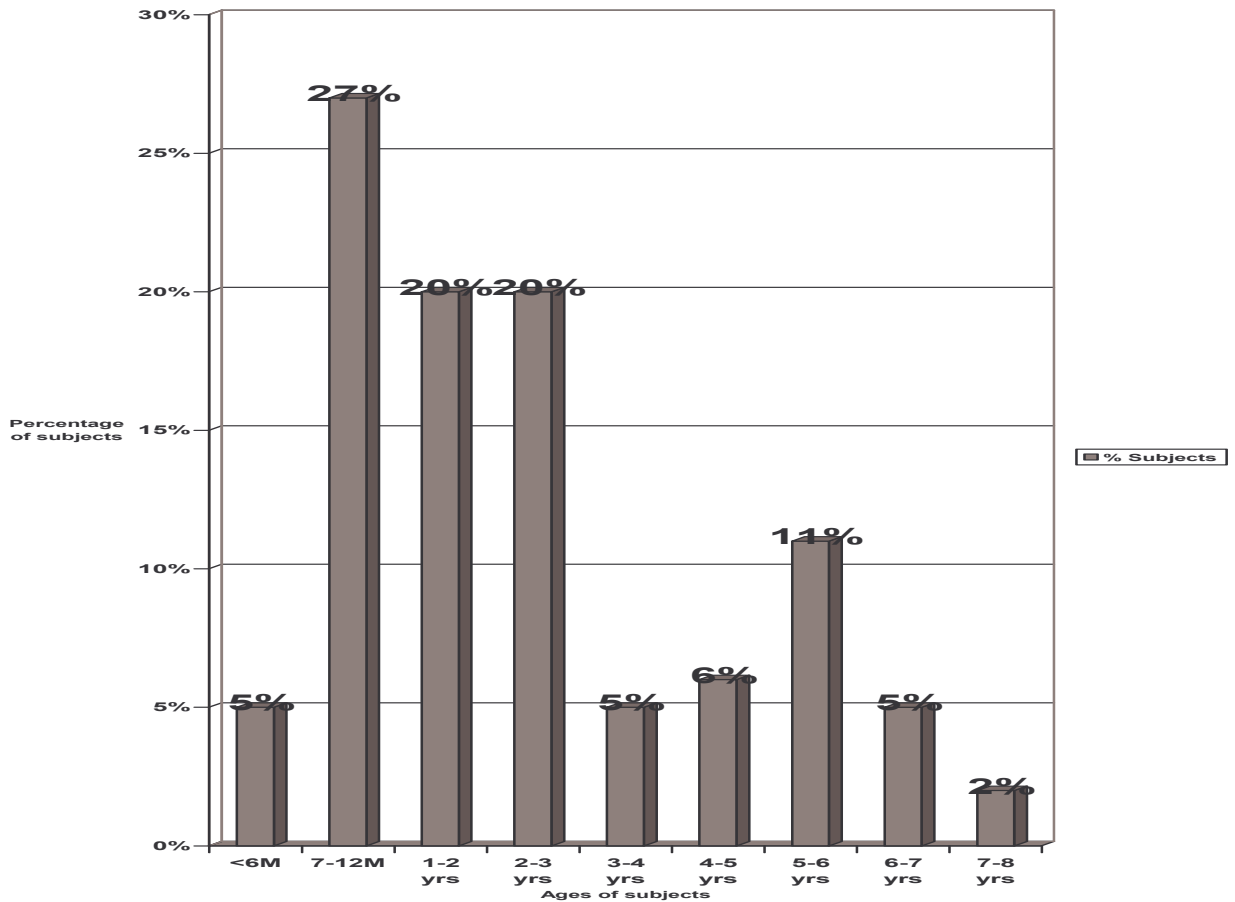
**Table 9: Source of referral to the PCIP**

<b>Person referring family to the PCIP</b>	<b>Percentage</b>
Audiologist	64%

Mother	11%
Teacher	7%
Family doctor	5%
ENT specialist	2%
Other	11%

As indicated in Table 9 it appears, once more, that the doctors who were involved in the children's medical management need to be informed more extensively of the referral guidelines for cochlear implantation, as well as audiologists who are the first step in the process starting at diagnosis. As the majority of participants resided in Gauteng, this is a manageable goal of the PCIP for consideration in terms of marketing and awareness campaigning for cochlear implantation.

A further audiological variable related to the study's first sub-aim involves the length of time that the child wore hearing aids, implying the time between fitting of hearing aids and cochlear implantation. This duration varied as depicted by Figure 4.



**Figure 4: Period (in years) for which children wore hearing aids prior to C.I.**

In Figure 4, the x-axis depicts the time period in months or years for which children wore hearing aids, while the y-axis represents the percentages of children in each time group.

From Figure 4 it can be seen that, while the required period for wearing hearing aids prior to establishing candidacy for cochlear implantation is 6 months, most participants' children wore hearing aids for longer than this period prior to cochlear implantation. It is an important requirement of the PCIP that while the child is wearing hearing aids during the trial period, he should either be included in a

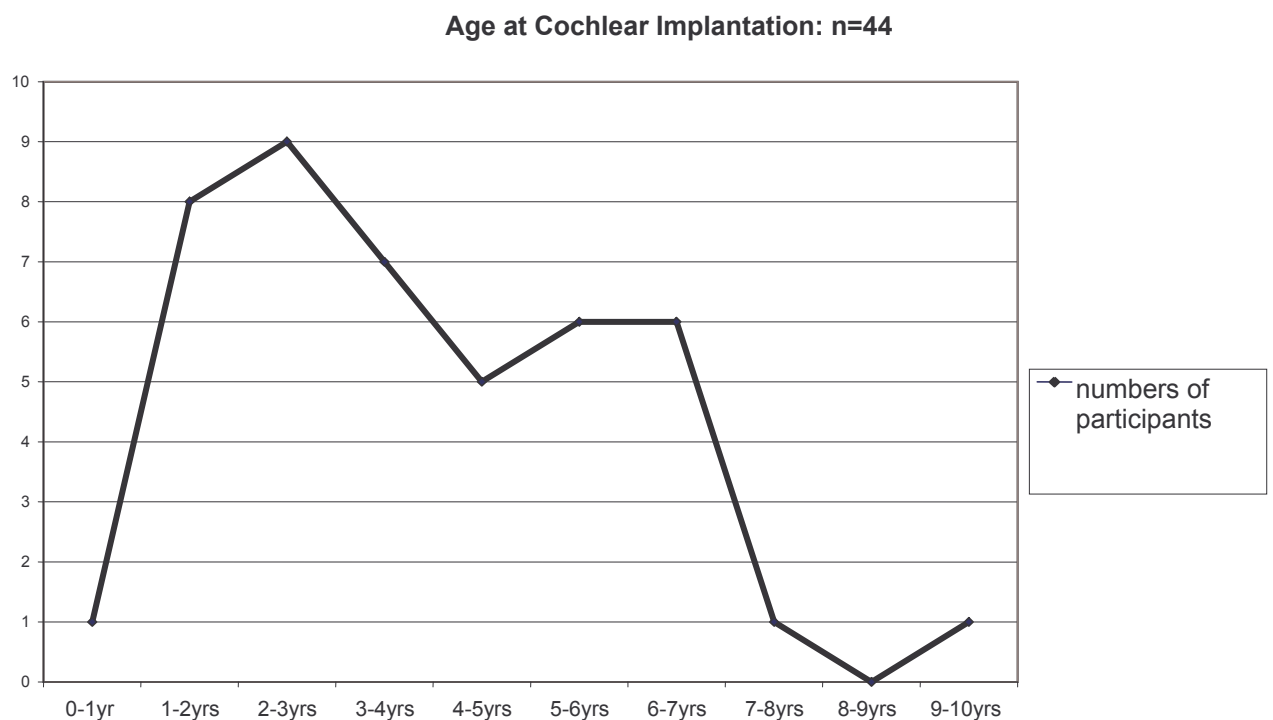
specialised classroom or receive intensive, appropriate auditory stimulation (Mrs Nellie Venter, 2005: personal communication). The highest percentage of children (27%) wore hearing aids for between 7 and 12 months, with a large percentage (40%) wearing hearing aids for 13-36 months. These children received little or no benefit from their hearing aids in this time, as this is a candidacy criterion for cochlear implantation.

Several factors may play a role in affecting the duration of hearing aid use prior to cochlear implantation, such as late referral to the cochlear implant programme; difficulty obtaining the necessary funding for the cochlear implantation; delays in the process of making the decision to operate as well as in the process of establishing candidacy, and the health of the child in order to make the surgery viable. All of these variables may play a greater or lesser role in determining the speed with which a child is implanted, although in the PCIP delays are not generated by an annual quota of cochlear implants or a lengthy waiting list as it is largely still an elective procedure and thus part of the private health care sector. In order for greater efficiency and shorter time between diagnosis and implantation of the child, all of these steps need to be optimized. This involves careful and efficient co-ordination between all of the team members involved: the family, audiologist and medical practitioners, healthcare professionals, the family's medical aid scheme and the cochlear implant programme itself (JCIH, 2000). Of these variables, one in particular, namely funding, can pose a large barrier to early implantation for many South African children. A question inquiring as to how the funds were raised for the cochlear implant revealed that of the 45 children, only one received a small amount of state funding for the procedure. The rest were, in 84% of cases, partly funded by the family's medical aid scheme, and

partly (41%) funded privately and through donations (39%). Most cases involved a combination of these three methods, with very few children’s cochlear implants being completely funded by medical aid schemes. This implies a waiting period before cochlear implantation as families raise the necessary funds themselves. A similar problem will be experienced by the other cochlear implant programmes in South Africa.

4.2.1.5) Age at cochlear implantation

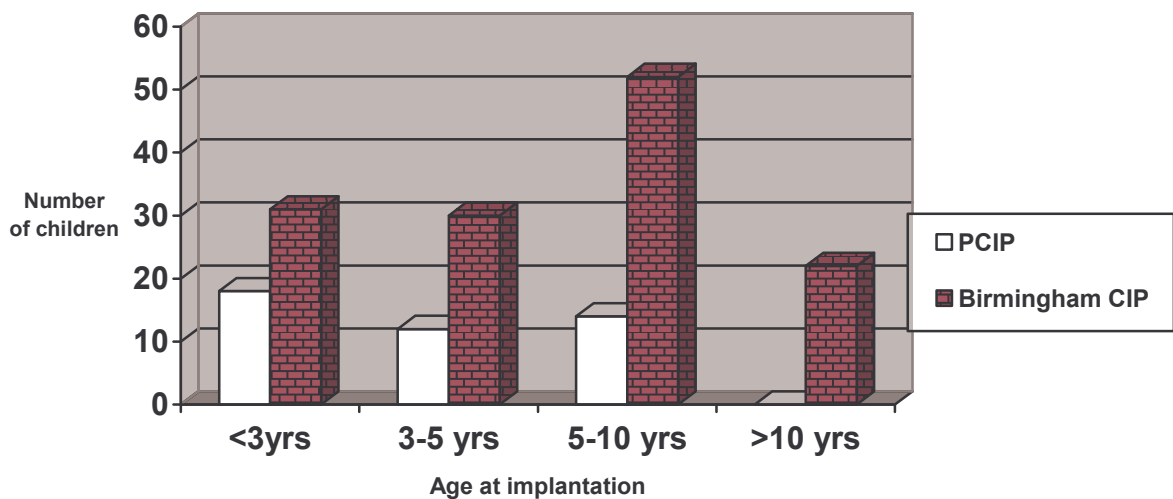
Figure 5 illustrates the distribution of ages at which children in the study received their cochlear implants.



**Figure 5: Age at cochlear implantation (n=44)**

It should be noted that the participant mentioned in the 0-1 year group in Figure 5 was implanted at the age of 12 months, thus no implants prior to age 12 months had yet

been performed at the time of data collection. The largest groups, in descending order, were those implanted at age 2-3, age 1-2, and age 3-4 respectively. Groups remain large until age 6-7, and only two children older than 7 years of age were implanted. A world-wide trend toward earlier cochlear implantation exists (Archbold *et al.*, 2000). Results therefore indicate that the PCIP data are in accordance with this trend, with more children being implanted prior to age 5 years than later. Since the study was cross sectional and not longitudinal in nature, comparison of the success of children implanted at various ages would prove difficult. When the numbers are divided into categories comparable to those used by the Birmingham Paediatric Cochlear Implant programme (BPCIP) (BCIP Outcomes Report 2001), the following chart can be drawn.



**Figure 6: Comparison of age at implantation for children in PCIP and BPCIP Outcomes Reports.**

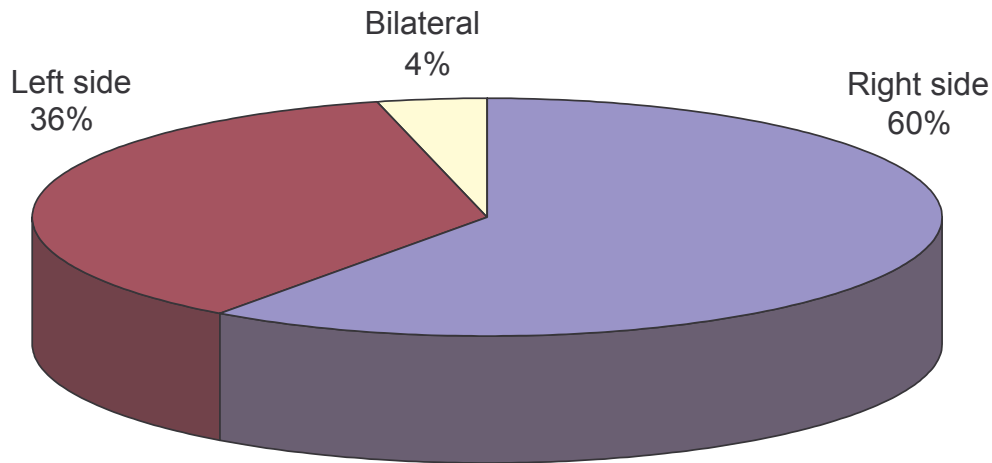
In Figure 6, age at implantation (x-axis) is divided into four categories as a function of the numbers of children (y-axis) in the PCIP (white) and the BPCIP (shaded brick pattern) in each age group.



While the majority of children in the BCIP (with congenital hearing loss) implanted between 1992 and 2000 received their implants between 5 and 10 years of age (see Figure 6), the majority of children in the present study of the PCIP (1992-2003) were implanted before the age of three. This represents a significant time advantage available to the majority of children in the PCIP study. Although the study does not include all the children enrolled in the PCIP and the numbers of children are not equivalent to the BCIP, sufficient data is available to make the comparison. The Nottingham Paediatric Cochlear Implant Programme's 1997 outcomes report (NPCIP Outcomes report, 1997) indicated that the majority of children in their programme were implanted between 3 and 5 years of age, which is slightly older than the PCIP group but younger than those in the BPCIP. Thus, despite a lack of universal neonatal hearing screening and efficient identification and referral systems in South Africa, the PCIP seems to be achieving a relatively early age at implantation. A possible reason for the BCIP's later age at implantation could be the presence of long waiting lists for cochlear implantation by the National Health System, with children being implanted in order of these lists and according to restrictive annual quotas of cochlear implants (Mrs. Nellie Venter, 2005: personal communication)

**Audiological variables**

*4.2.1.6) Side of implant and reasons*



**Figure 7: Side chosen for cochlear implantation: percentages of children (n=45)**

As illustrated by the Figure 7, in most children (60%) the right side was chosen for cochlear implantation, while 36% had their left ear implanted and 4% received bilateral implants. When the hearing loss is similar in both ears, the dexterity of the child is taken into account. As most people are right handed, this could explain the high percentage of right side implants.

The reasons for the ear chosen, in the case of unilateral implants, are next reported in Table 10.

**Table 10: Reasons for choice of side for cochlear implantation**

Reason for choice of ear to implant	Percentage of participants
Least residual hearing ('poorer ear')	38%
Handedness	21%
Most residual hearing or most normally formed cochlea ('better ear')	13%
Unsure	8%
Other ear not useable due to cochlear malformation	8%
Other ear had frequent middle ear infections	8%
Facial nerve implications	2%
Child's own choice	2%

As shown in Table 10, the largest group (37%) chose the poorer ear (with less residual hearing). Side of implant may be affected by many factors where there is not a clear structural or surgical choice of side. Cowan, *et al.* (1997) demonstrated that children with some residual hearing were more consistent, as a group, in achieving high levels of open set speech perception after cochlear implantation than those with profound congenital hearing losses and cochlear implants. As selection criteria shift to include children with greater levels of residual hearing, this decision becomes more complex as the possibility of 'losing' the residual hearing in the implanted ear due to the effects of surgery, including cochlear trauma and connective tissue growth, become relevant (Kiefer, *et al.*, 1998). In general, the side with the least residual hearing is then implanted, affording two advantages to the child and the family, namely the psychological sense of security that his/her hearing sense will not be obliterated completely, as well as the possibility of making use of the binaural/bimodal advantage with a well fitted hearing aid on the non-implanted ear. Kiefer, *et al.* (1998) reported increases in levels of residual hearing in the non-implanted ears of children tested 6 months post-operatively, which was attributed to maturation of the auditory pathway induced by the sensory experience provided by the cochlear implant. This lends support to the practice of wearing a hearing aid on the non-implanted ear. However,

the implanted ear, although not showing loss of residual hearing at the same test interval, did show deterioration in non-aided thresholds when tested at a later stage after more connective tissue and new bone growth in the cochlea despite surgical techniques which aimed to minimize insertion trauma of the electrode array. Lenarz (1998) reported similarly that it cannot be assumed that hearing can be preserved in the implanted ear, even using soft surgical techniques, due to the possibility of a direct mechanical dysfunction of the inner ear or a dysfunction of the remaining hair cells. This brings the decision regarding side of implantation to the following criteria, namely degree of hearing loss on each side; duration of deafness on both sides, comparison of auditory performance with hearing aids of the two ears and aetiology of hearing loss.

In general these criteria imply that the ear with least residual hearing and weaker performance with hearing aids be implanted, provided the duration of deafness of that side has not been too long (e.g. 10 years or longer) which might imply functional reorganization in the auditory cortex based on a lack of input from this ear. The brain would then possibly not register sound stimuli from this side, resulting in a disappointing outcome of cochlear implantation. In considering the aetiology of deafness, the functional and structural status of the cochlea and hearing nerve must be examined on both sides, with the more intact side being selected (Lenarz, 1998). Philosophies and viewpoints regarding the choice of ear to be implanted have changed in the PCIP as a function of time, new research findings, shifting selection criteria and changing surgical techniques. As a rule, the largest number of participants in the current study had chosen their children's 'weaker' ear for implantation. This raises the next issue, namely the use of a hearing aid on the non-implanted side.

4.2.1.7) *Hearing aid use on the non-implanted ear.*

Only 44% of children wore a hearing aid on the contra lateral ear. Reasons for not wearing a hearing aid included stating that the ear had too little residual hearing to benefit, not knowing that it was advisable, and the child's refusal to wear the hearing aid. The reasons for wearing the hearing aid included better sound, better localization, stimulation of the acoustic nerve, the school's policy, more balanced hearing, making use of residual hearing, and recommendation by the audiologist. In Pretoria, the majority of specialised schools advocate the wearing of a hearing aid on the non-implanted side, at least during school hours. This, together with the audiologists of the PCIP's own frequent recommendations to encourage this practice, may account for a large percentage of the participants' motivation to do so, although this is a fairly recent development based on more recent research and the advent of bilateral cochlear implants as a viable option. However, many children implanted in earlier years stopped wearing their hearing aids after cochlear implantation, as parents were not at that stage counseled regarding the benefits of binaural/bimodal stimulation, and have since rejected the idea or lost much of the residual hearing in the ear due to lack of stimulation (Ching, *et al.*, 2001). They may only have experience of linear, analogue models of hearing aids, which were not as compatible with cochlear implants as digital hearing aids, and were not well suited to balancing with the sound of a cochlear implant. Another contributing factor to children's rejection of the hearing aid may be incorrect or inadequate balancing of the acoustic input from the cochlear implant and hearing aid together by an audiologist. This could lead to the hearing aid proving a hindrance to listening with the cochlear implant instead of providing a binaural advantage. Armstrong, *et al.* (1997) reported

that some children in his study found that the hearing aid dominated the cochlear implant prior to loudness balancing, since loudness between the two devices varied as a function of frequency with low frequency information being louder with the hearing aid, causing competition between electrical and acoustic signals. This negatively affected both speech perception and comfort. However, in the same study by Armstrong, *et al.* (1997) significant binaural advantage in background noise, as well as localization and a sensation of balanced hearing, all reasons for wearing a hearing aid stated by participants in the present PCIP study, were found after balancing. This especially occurred in the case of children with significant residual hearing, which may be stimulated by the subsequent maturation of the auditory cortex brought about by stimulation via the cochlear implant (Kiefer, *et al.*, 1998). It is important that the non-implanted ear be fitted appropriately with a digital hearing aid. Ching, *et al.* (2001) recommended the NAL-RP protocol, used in their study to balance loudness of hearing aid gain to match the cochlear implant signal, to aid integration of signals from both ears leading to better speech perception. These authors further urge professionals to encourage the use of a hearing aid with a cochlear implant after a stable map has been established with the cochlear implant alone.

A small number of children in the present study found it too difficult to integrate the two different sounds, and thus rejected the hearing aid. These may be children who have more difficulty in general with the processing of auditory input, as was the case with two children. Closer, individual and anecdotal questioning of some participants revealed one child who remained reliant on his hearing aid after cochlear implantation, to the detriment of learning to listen to the new range of high frequency information to which he had access via his cochlear implant. In this case the hearing

aid had to be removed and then re-introduced and balanced with the sound from the cochlear implant once the child was accustomed to listening with the cochlear implant alone.

Armstrong, *et al.* (1997) reported on the development of a ‘combionic’ device, combining hearing aid and cochlear implant processing, to overcome the problems inherent to binaural/bimodal fitting. This development is, however, still in the experimental stage.

#### *4.2.1.8) Bilateral cochlear implants*

Only two children have received bilateral cochlear implants, both in successive operations as opposed to simultaneous operations. According to the participants, of the two, one wears both his devices well and the other refuses to wear the second device. Studies demonstrating the advantages of bilateral implants are growing in number (Ertmer, 2002). Further studies should report on this section of the population once its numbers have grown sufficiently to allow for a measurable sample.

4.2.2) *Results and discussion relating to Sub-aim Two*

**SUB-AIM 2: COMPILATION OF A DESCRIPTIVE PROFILE OF THE FAMILIES OF CHILDREN WITH COCHLEAR IMPLANTS IN THE PCIP**

In this section a profile of the families of children with cochlear implants in the study is created, including information on the prevalence of hearing loss or additional disorders of speech, language or learning in families; educational level and employment status of parents; time spent with children by parents; and socioeconomic status of families.

4.2.2.1) *Family history of disorders.*

In eight children's families there existed a history of childhood hearing loss. Two participants responded that there was a family history of a genetic syndrome, namely Waardenburg Syndrome and retinitis pigmentosa, in the participant's mother and sibling respectively. Participants were further asked to report on a family history of speech/language or academic difficulties not related to hearing loss. The presence of these difficulties in the parent might affect their level of stimulation of the child (Dollaghan, *et al.*, 1999), while the presence of these hereditary traits in the children themselves would hamper their success in acquiring language and academic progress. For comparison purposes, the group was again divided into those children in specialised educational settings (SE), and those in inclusive education (IE). In the SE group, 17% of families had a history of learning/academic and serious speech/language difficulties in childhood. This is higher than the 5% reported in the IE group, and may be a factor contributing to the need for specialised education in some members of the SE group. For the respondent sample as a whole (n=45), the



figure is just under 16% for a family history of either a genetic syndrome, speech/language difficulty or academic difficulty in either biological parent or siblings. These results relate to the small number of participants' children who displayed hereditary hearing loss and presented with a genetic syndrome associated with hearing loss (see Table 7: Causes of hearing loss in children).

### **Environmental variables related to the family unit**

#### *4.2.2.2) Parental education and employment status*

In order to explore the role that the educational status of the parents may play in the academic success of a child with a cochlear implant, the group of children enrolled in educational settings was again divided into two groups, the first being those in inclusive educational settings and the second being those in specialised educational settings. These results are presented in Table 11. The employment status of the parents of children in these two groups was also investigated in order to give an indication of socio-economic status and the amount of time spent at home with the children by the maternal and paternal parents/caregivers respectively. It follows that the parent spending the most time with the children will play a greater role in academic development, and this parent's own educational status may then be of greater relevance.

**Table 11: Parental education and employment for children in inclusive and specialised educational settings.**

<u>Characteristic:</u>	<u>Parents of children in inclusive education(n=19)</u>	<u>Parents of children in specialised education (n=24)</u>
<u>Maternal education level: highest level attained</u>	Standard 8 =1 (5%) Matric =6 (32%) Tertiary diploma = 7 (37%) University degree= 5 (26%)	Standard 8=4 (17%) Matric =10 (42%) Tertiary diploma =8 (33%) University degree =2 (8%)
<u>Paternal education level: highest level attained</u>	Standard 8=3 (16%) Matric =2 (10%) Tertiary diploma =7 (37%) University degree =7 (37%)	Standard 8=3 (14%) Matric =6 (27%) Tertiary diploma=8 (36%) University degree=5 (23%) <b>(n=22: 2 fathers' data absent)</b>
<u>Both parents employed</u>	12 = 63%	16 = 67%
<u>One parent employed</u>	Only maternal parent/caregiver =0 Only paternal parent/caregiver =7 (37%)	Only maternal parent/caregiver =4 (16.5%) Only paternal parent/caregiver =4 (16.5%)

The results of Table 11 show a number of differences between the two groups of families. Mothers/maternal caregivers of the children in inclusive educational settings showed a higher level of educational status in general. They were more likely to have a tertiary diploma or university degree, and less likely to have a maximum educational level of matric or standard 8. The most pronounced differences were seen in the highest level, namely university degree which included 26% of mothers in the first group and only 8% of mothers in the second group. The lowest level measured, namely standard 8, included only 5% of mothers of children in inclusive settings and 17% of mothers in the second group.

A similar trend was seen with paternal parents/caregivers, where 37% of fathers of children in inclusive education had a university degree as opposed to 23% of fathers of children in specialised settings. Thus it would appear from this data that the level

of maternal and paternal educational status may play a role in the academic success of a child with a cochlear implant. In a study by Dollaghan, *et al.* (1999) it was found that the vocabulary and general language development of normal hearing preschoolers increased as a function of their mothers' level of education. This was attributed to factors such as availability of non-material resources (as opposed to material resources such as books and appropriate educational toys, which was more linked to socio-economic status) such as the parents' attitude toward education and academic competence; quality of language use; knowledge of child care, development and stimulation and their overt behaviour in this regard (Dollaghan, *et al.*, 1999). The quality and quantity of child-directed language is expected to be affected, as well as the quality of assistance with academic tasks and concepts. The same, or greater, effect can reasonably be expected with children with hearing loss and cochlear implants. Mayne, *et al.* (2000) reported that mothers with a lower level of education and lower socio-economic status spoke less often to their infants with hearing loss, and used fewer and less diverse words resulting in lower levels of receptive vocabulary development.

As depicted in Table 11, when employment status of parents is investigated, it is clear that in a large percentage of families in both groups (between 63% and 67%), both parents worked. In all families at least one parent/caregiver worked, thus there was no unemployment. However, mothers of children in inclusive settings were less likely than mothers in specialised settings to be the sole provider of income, and more likely to be the parent not working and, presumably, tending to the children and assisting with academic tasks at home as well as providing learning opportunities and stimulation. Mothers' levels of education and ability to provide stimulating learning

situations and a rich language model to their children can thus be expected to have an influence on the educational level achieved by the child with a cochlear implant.

*4.2.2.3) Parental employment: socioeconomic status and home life*

A total of 75% of mothers were employed, however it was also reported that 64% of children spent the majority of their time with their mothers (see Table 11). This indicates that mothers generally worked from the home, flexible hours or part time only, since school was also listed as an option for where the child spent the most time. This is hypothesized to be especially true of mothers with higher educational status, who might thus have jobs involving more flexibility and choice in terms of own hours and work venue. In general, as seen in an earlier analysis, these mothers would be more likely to be in the group whose children are in inclusive educational settings. Alternately, it could indicate that mothers simply meant that their children spent evenings and weekends with them and were not in a school residence facility. This question should thus be interpreted cautiously as it proved to be ambiguous. A larger percentage (98%) of fathers / parental caregivers was employed.

Another factor which could indicate the socio-economic status of the family and ability to provide adequate financial support for a child with a cochlear implant, is the question of whether the pregnancy was planned or not, to which 62% of parents answered that it was.

In families where only one parent worked, the parent was more likely to be the paternal than the maternal parent. Socioeconomic status was not questioned directly in the questionnaire used for the present study.

*4.2.3) Results and discussion relating to Sub-aim Three*

**SUB-AIM 3: DESCRIPTION OF CHILDREN'S CURRENT PERCEIVED OUTCOMES IN TERMS OF EDUCATIONAL, AUDITORY AND COMMUNICATIVE FUNCTIONING**

In this section, the following sub headings will be used to guide the reader towards a description of outcomes.

-Auditory outcomes (including device use, auditory performance and musical enjoyment)

-Linguistic outcomes

-Educational placement outcomes and variables surrounding this outcome. Here

Tables 14 and 15 are used for comparison of the group of children in the study currently attending specialised schools, with the group in inclusive education.

Although many variables affecting the children in these groups are listed and discussed, seeming to be more appropriate to the preceding two sub-aims, these are used here as a means by which the outcomes can be understood and discussed. This sub-aim, including this comparison, leads directly to the next sub-aim in which the specific variables leading to the successful outcomes of a child with a cochlear implant in the PCIP, are highlighted and explained.

**Auditory outcomes**

*4.2.3.1) Use of the device*

All participants reported that the cochlear implant device was worn at all times (all waking hours), at home and at school. This figure is significantly higher than that

quoted by both the Birmingham Paediatric Cochlear Implant Programme (2000) which reported 84%, and the Nottingham Paediatric Cochlear Implant Programme (1997) which reported 87,5% of children using their cochlear implants at all times. A full comparison cannot, however, be made since the present study does not include all clients of the PCIP, but only those who responded to the request to participate in the study. This may have inadvertently discouraged those who are less optimal users of their cochlear implants.

#### 4.2.3.2) Auditory performance

In order to gain insight into the children' hearing with their cochlear implants, participants were asked to state, for a number of auditory stimuli, whether children could hear (detect) and/or identify the sounds (know what they are).

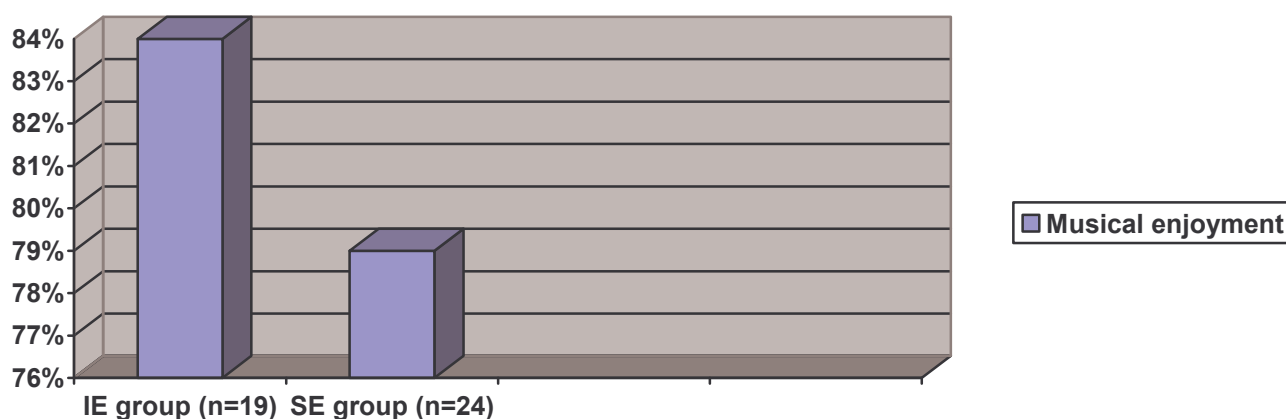
**Table 12: Detection and Identification of various auditory stimuli by children as reported by participants (n=45).**

<b>Auditory stimuli</b>	<b>Percentages of children who could <u>detect</u> the sound</b>	<b>Percentages of children who could <u>identify</u> the sound</b>
Environmental sounds	100%	82%
Music	100%	71%
Own name and family names	100%	87%
Speech	100%	91%
Soft sounds (water running, leaves rustling)	96%	60%
Whispered speech	93%	53%

As seen in Table 12, speech was most easily detected and identified, which is probably due to the cochlear implant's bias toward providing access to sounds in the speech frequency (Gibson, Rennie & Psarros, 2000). These reports indicate that cochlear implantation made most sounds accessible to the sample children with a

severe to profound hearing loss, and that more than half of this sample could now identify even very soft sounds. Many of these sounds and loudness levels would not have been detected by these children with the use of hearing aids instead of cochlear implants, thus access to sound was greatly improved by cochlear implantation.

#### 4.2.3.3) Musical enjoyment



**Figure 8: Percentages of children in inclusive (IE) and specialised (SE) education who enjoy music (n=43).**

(As two children do not attend any educational setting, their data was excluded and the number of children is thus 43.)

According to Figure 8, slightly more children in IE (84%) were reported to enjoy music than the children in SE (79%). As a group (n=43), 80% of the children with cochlear implants were reported to enjoy music. Musical appreciation may give an indication of the brain's central auditory processing of sound via the cochlear implant as well as of auditory closure skills required to make sense of the lyrics and melody together. Music appreciation may also be stimulated in an inclusive setting by normal hearing peers. At present, speech processors of cochlear implant systems are

designed to obtain the maximal reception of the speech frequencies (Kong, Cruz, Jones & Zeng, 2004) although future processing strategies may include programmes for music specifically.

### **Linguistic outcomes**

#### *4.2.3.4) Mode of communication*

Although the question was graded into a ‘never, sometimes, often’ scale, for the purposes of this analysis only the ‘often’ group has been reported. The percentages shown in Table 13 indicate how many participants marked the various levels of communication development and mode of communication as being used ‘often’ by the child.

**Table 13: Mode of communication and level of language development for all children (n=45).**

<b>Mode/level of communication development</b>	<b>Percentage of children using this mode/level ‘often’</b>
Sounds	36%
Natural gestures	38%
Word approximations	31%
<b>AVERAGE FOR PREVERBAL LEVEL</b>	<b>35%</b>
Single words	36%
<b>AVERAGE FOR TRANSITIONAL LEVEL</b>	<b>36%</b>
Two word combinations	29%
Three word combinations	29%
<b>AVERAGE FOR LEVEL ACQUIRING FUNCTIONAL LANGUAGE</b>	<b>29%</b>
Incomplete sentences	36%
Complete sentences	47%
<b>AVERAGE FOR FUNCTIONAL LANGUAGE LEVEL</b>	<b>42%</b>
<b>SIGN LANGUAGE</b>	<b>11%</b>
<b>SPEECH PLUS SIGN: TOTAL COMMUNICATION</b>	<b>22%</b>



From the breakdown in Table 13 it is evident that the majority of children was using the auditory-oral mode of communication as opposed to signing, and were in various stages of development of spoken language. The largest number of children were categorized in the 'functional language' column, which in this context means that they are using connected speech in complete or incomplete sentences more often than 3-word utterances or shorter units of language. A total of 32% of parents indicated that they were using formal sign 'often' to some extent, either in conjunction with speech or in isolation. This implies that 68% of children were not using any form of manual communication to supplement listening and speaking. This is in contrast to the findings of the Birmingham Paediatric Cochlear Implant Programme (2000), who reported a total of 16% of children using speech only to communicate. This may indicate a difference in philosophy rather than outcome after implantation, as total communication is used widely as opposed to the PCIP's bias toward auditory/oral communication modes. It may also reflect, in part, the influence of the BPCIP's generally higher age at implantation and levels of device use, resulting in lower speech perception and production skills of children.

The three categories included in Table 13 in the preverbal level, namely sounds, natural gestures and word approximations, generally indicate those children who have been implanted recently or who are very young and in the process of developing speech in an auditory/oral setting without the use of sign. One participant in this category was already a proficient user of sign language, coming from a bilingual-bicultural household where both parents were Deaf, and so her language level was considerably higher than her use of verbal communication implied. This child's sister, from the same home, was fully bilingual and functioned in the category

‘functional language level’ with normal speech intelligibility and in a regular classroom, having been exposed to sign initially by her parents and her parents’ Deaf friends, and spoken language by her grandmother, the school and the rest of the community. Tait, *et al.* (2000) described this situation where-in the age at implantation and the level of communicative and linguistic functioning, albeit using manual communication, were more relevant to later language outcomes than the communication mode used pre-implantation. However, post-implantation the children in the study by Archbold, *et al.* (2000) using total communication showed lower levels of speech perception and intelligibility 5 years later than those in oral settings. Kirk, *et al.* (2002) found that expressive language lagged behind receptive language for children in oral and in total communication settings, which could imply that the children in the present study had a higher level of receptive language (not measured by the study due to the difficulty using parental report for this variable) than the reported level of expressive language represented by this data set. Kirk, *et al.* (2002) further found that children of parents with a hearing loss were exposed to an impoverished linguistic environment, and that their language development was slower and reached lower ultimate levels. In this study, the two children with Deaf parents had the extenuating variable of a hearing, involved grandparent and thus this effect on their development was probably lessened. This highlights the value of descriptive research including in-depth analysis of the variables involved in individual children and families’ situations.

As seen in Table 13, the category ‘transitional level’, indicating single word use, was separated from the category ‘acquiring functional language’, indicating two and three word utterances, for a greater degree of specificity. More children in this study were

using single words than more complex word combinations, however an even larger group was using mature sentence forms. This indicated that in the present cross sectional study, more children were in either the earlier, or the more advanced stages of language acquisition than in the middle stages. This result does not necessarily imply that these children are of a certain age group, since the linguistic outcome of paediatric cochlear implantation is highly variable. The population is a “notoriously heterogenous population with great variation in their auditory, cognitive and linguistic maturity which may be impossible to quantify before intervention.” (O’Donague, *et al.*, 2000: p.467), however duration of device use would play a role. A possible cause of the variation in speed and extent of linguistic outcome is the central auditory processing mechanisms that are crucial to the perception of speech through a cochlear implant, and for which there are as yet no reliable measures (O’Donague, *et al.*, 2000).

Moog and Geers (2003) further add that children in inclusive educational settings are more likely to have well developed reading abilities, a later developing linguistic skill highly correlated to academic success, and dependent on good receptive and expressive language and verbal abstract reasoning abilities. These skills in turn, develop in the presence of good speech perception with the cochlear implant (Moog & Geers, 2003).

From this discussion it is evident that cochlear implantation resulted in access to a wide range of sounds, including speech; soft environmental sounds and music for most of the children in the PCIP, although children in the IE group showed slightly

better outcomes. All the children wore their cochlear implants, and the majority were acquiring, or had acquired, speech through listening.

*4.2.3.5) Educational placement outcomes: Comparison of variables and outcomes for children in inclusive education and those in specialised educational settings.*

The third sub-aim of the study requires a description of the audiological, educational and communicative outcomes of the children, which will become apparent through presentation and discussion of the following results.

For most cochlear implant programmes the goal of early implantation is to allow the child to be placed into an inclusive educational setting and to function among his/her normal hearing peers. The Nottingham Paediatric Cochlear Implant Programme (1997) reported placing 53% of their preschool age children in inclusive settings at two years post-implant, while the Birmingham Paediatric Cochlear Implant Programme (2000) reported 17% of their total population, of all ages, in regular classrooms.

Tables 14 and 15 aim to profile various characteristics and outcomes as they pertain to children in inclusive education, and those in specialised educational settings. Thereafter follows a discussion of the key similarities and differences between the profiles of these two groups.

Hearing loss is abbreviated as H.L, and cytomegalovirus as CMV. Two children are at home with their mothers and not in any educational setting, therefore the total for

the two groups is 43 children. Only two children attended schools for the Deaf where sign language was a medium of tuition.

**Table 14: Description of characteristics of children in inclusive educational settings (n=19)**

Characteristics	Division of group into age categories		
	Preschool n=6	Primary school n=10	High school n=3
Age at diagnosis of H.L.	6-16M	6-18M	10-33M
Age at fitting of hearing aids	7-18M	9-19M	10-34M
Age at cochlear implantation	19-24M: n=4 32M: n=1 42M: n=1	22-75M	52-140M
Cause of H.L.	- Unknown:3 - Hereditary:1 - Syndrome (Waardenburg): 1 - CMV: 1	- Unknown:3 - Hereditary:2 - Meningitis:1 - CMV: 1 - Complications in pregnancy:1 (placenta) - Syndrome: 1 (unspecified: reported) - Myelin sheath disorder: 1	- Unknown:1 - Maternal medication:1 - Meningitis: 1
Congenital H.L.	All	n=7	n=2
Progressive H.L.	None	n=2	n=1
Married parents	All	n=6	n=2
Older siblings	All	n=5	n=2
No. of languages spoken in home	1-2	1-2	2
SIR* category of speech intelligibility	2-5	5: n=7 4: n=3	5
No. currently enrolled in speech therapy	All (Previously & currently)	n=5 (all previously)	n=1 (all previously)
No. currently enrolled in occupational therapy	N=1 (2 previously)	n=1 (5 previously)	n=0 (2 previously)
Able to use telephone	n=2	n=8	n=3
Use of FM system	None	n=6	n=1
Prenatal complications	None	n=3	None
Perinatal complications	None	n=6	n=1
Feeding problems	n=2	n=2	n=0
Developmental milestones excluding speech	Delayed: n=2	Delayed: n=1	Delayed: n=1
Hospitalizations and reasons	- Insertion of grommets for otitis media - respiratory tract infections - digestive tract illness	- Insertion of grommets for otitis media - respiratory tract infections - digestive tract illness - meningitis - trauma - blood transfusions	- Insertion of grommets for otitis media - meningitis

\* SIR: Speech Intelligibility Rating Scale

**Table 15: Description of children in specialised educational settings (n=24)**

Characteristics	Division of group into age categories		
	Preschool n=11	Primary school n=9	High school n=4
Age at diagnosis of H.L.	7-26M	10-48M	23-36M
Age at fitting of hearing aids	11-36M	11-48M	26-36M
Age at cochlear implantation	<2yrs:n=1 2-3:n=7 3-5:n=3	40-84M <3yrs:n=0 3-5:n=2	96-120M <3yrs: n=0 3-5: n=0
Cause of H.L.	- Birth trauma:1 - Unknown:6 - Hereditary:1 - Rubella:1 - Syndrome (Waardenburg): 1 - Complications in pregnancy:1	- Birth trauma:1 - Unknown:2 - Hereditary:1 - Rubella:2 - Meningitis:1 - Otitis media: 1 - Maternal CMV: 1	- Birth trauma:1 - Unknown:1 - Syndrome:1(with kidney disease) - Maternal medication: 1
Congenital H.L.	n=10	n=8	n=3
Progressive H.L.	n=3	n=1	n=2
Married parents	n=9 divorced =1 live together = 1	n=8 remarried =1	n=2 widowed=1 divorced=1
Older siblings	n=8 2=middle 7=youngest	n=5	n=2 (1=only child, 1=youngest of 3)
No. of languages spoken in home	1-2 language: n=10 3 languages: n=1	1-2 languages	1-2 languages
SIR category of speech intelligibility	1:n=3 2:n=3 3:n=1 4:n=3 5:n=1	1:n=1 2:n=0 3:n=3 4:n=2 5:n=3	1:n=0 2:n=0 3: n=1 4: n=2 5: n=1
No. currently in speech therapy	6 (9 previously enrolled)	n=5 (6 previously)	n=3 (4 previously)
No. currently in occupational therapy	n=0 (4 previously)	n=2 (6 previously)	n=0 (1 previously)
Able to use telephone	n=5	n=4	n=3
Use of FM system	5	n=2	n=0
Prenatal complications	n=2	n=3	n= 0
Perinatal complications	n=12	n=9	n= 2
Feeding problems	n=5	n=0	n=0
Developmental milestones excluding speech	Delayed: n=6	Delayed: n=2	Delayed: n=1
Hospitalizations and reasons	- Insertion of grommets - fever - failure to thrive - reaction to vaccination - poisoning - respiratory tract infections - digestive tract illness - meningitis - trauma - blood transfusions	- Meningitis - Insertion of grommets for otitis media - respiratory tract infections	- Respiratory tract infections - skeletal fractures

The two tables (Tables 14 and 15) will be discussed concurrently, and the specific age group of children in each of the two educational settings compared for each variable.

For the purposes of this discussion, the group in specialised education will be referred to as SE, and those in inclusive settings as IE. Sub-headings given in italics relate to the relevant sections of the tables, and the reader is referred to the tables throughout the discussion of these sections.

- *Age at diagnosis of hearing loss:*

The children in the preschool IE group received a diagnosis slightly earlier than those in the SE group, with an average difference of 6 months, as illustrated by Tables 14 and 15. The same trend was seen in the primary school (average difference 17 months) as well as the high school (average difference 9 months) groups. Since diagnosis is the first step toward amplification and cochlear implantation, it seems that the children in the SE group were, in general, at a slight time disadvantage from the start (Yoshinaga-Itano, *et al.*, 1998).

- *Age at fitting of hearing aids:*

In general, children in the three groups in IE were fitted with hearing aids earlier and younger than those in SE (for specific age ranges see the tables). The average differences in age at hearing aid fitting for the preschool, primary school and high school groups respectively were 9, 11 and 16 months in favour of the IE group. The IE group thus had access to sound, prior to cochlear implantation, at a younger age than the SE group, allowing important neurological development to take place in the auditory cortex (Katz, *et al.*, 2000).



- *Age at cochlear implantation:*

The largest group of IE preschoolers was implanted between the ages of 1 and 2, while the majority of SE preschool children received their cochlear implants between the ages of 2 and 3 years of age. This represents a significant difference in the children's groups in terms of neural plasticity and capacity to acquire age-appropriate language, although it is positive that the majority of children in this age range were implanted at or before 3 years of age. Ruben (1995) reported the critical sensitive period for the ultimate achievement of maximal linguistic proficiency to extend from the sixth week of foetal life to between the second and third year of life. The difference in subsequent communication outcomes between children implanted before 2 years, and those implanted between 2 and 3 years, was reported by Kirk, *et al.* (2002) to be significant. As seen in Tables 14 and 15 of the current study, in the primary school SE group, however, no children were implanted prior to age 3 years, and this group received their cochlear implants on average 14 months later than their peers in the IE group. The high school group was generally implanted later, partly due to their age in relation to the history of the PCIP, but again the SE group received their cochlear implants an average of 12 months later than the IE group. The trend seen in the previous two variables is thus continued, and in general it can be said that children in the SE group gained access to sound at a later age than children in the IE group. Since age has been identified through-out the literature as the major determining factor in successful paediatric cochlear implantation (Kirk, *et al.*, 2002; Mayne, *et al.*, 2000), these results are significant and point to the urgency for more efficient systems of diagnosis, referral to cochlear implant teams, establishing of candidacy and funding for cochlear implantation in the PCIP.

- *Cause of hearing loss:*

Birth trauma was given by participants as the cause of hearing loss for three children (all three age groups) in the SE group, but for none of the IE group. These children might be expected to have suffered further developmental consequences of this early traumatic experience (Merenstein & Gardner, 1998), including increased risk for developmental delays in all areas including sensory; motor; cognitive and speech/language development (Rossetti, 1996). Two children in each group had a genetic syndrome, one of which was Waardenburg Syndrome in each group. Meningitis was the cause for two IE children and one SE participant, with the same figures applying to maternal CMV infection. Of the group in IE, there were no reports of maternal rubella being the cause of hearing loss, while this was true for three children in the SE group. It follows that children of mothers who contracted rubella while pregnant would have additional complications and developmental implications, and thus possibly be more likely to require specialised education (Clark, *et al.*, 1997). Seven of the children in IE had hearing losses due to unknown causes, and nine of those in SE while hereditary, non syndromic hearing losses occurred in 3 IE and 2 SE children. Hereditary, non-syndromic hearing loss is more likely to go unaccompanied by additional disabilities (Angeli, *et al.*, 2000; Matsushiro, *et al.*, 2002), implying that this specific cause of hearing loss may be related to higher levels of success with a cochlear implant.

The etiologies of hearing loss for which the numbers are comparable between the two groups are genetic syndromes and unknown causes. More SE than IE children lost their hearing due to congenital rubella syndrome and birth trauma, while more children in IE had hereditary hearing loss. The children in IE also had more

incidences of maternal CMV and meningitis being the cause of their hearing loss than those in SE, which is not what might be expected since these two etiologies hold the potential for complications and disabilities additional to the hearing loss (Clark, *et al.*, 1997). However, in the children with meningitis, an advantage may have been their early exposure to sound as well as their swift cochlear implantation following the illness to reduce the effects of cochlear ossification (Bredberg, *et al.*, 1997). Closer inspection of the data revealed that the two children with a history of maternal CMV infection in IE were a part of families where parental level of education was very high, as was socio-economic status and early as well as frequent access to therapeutic services and support within the inclusive setting and in addition to it. The other causes listed in Tables 14 and 15 affected single children, and are thus not useful to a comparative discussion.

- *Nature of hearing loss: progressive and congenital hearing loss*

In Tables 14 and 15 it can be seen that within the IE group, hearing loss was congenital in 79% of cases, while this was true in 88% of those children in the SE group. This implies that slightly more children in IE had exposure to sound prior to the onset of their hearing losses, placing them at an advantage in terms of neurological maturation of the auditory cortex, even if exposure to sound was brief (Ponton, *et al.*, 1996). Progression of the severity of the hearing loss was present in 16% of children in IE and in 25% of the SE group. The majority of children in both groups thus had congenital hearing losses, which were not progressive in nature, implying that very little access to sound and language development occurred prior to cochlear implantation in the sample as a whole. This increases the importance of the variables

of age at diagnosis of hearing loss (see Figure 3) and age at cochlear implantation (see Figure 5)

- *Marital status of parents*

Most of the parents of children with cochlear implants in both groups were married. Most children thus benefited from two parent, nuclear family contexts. Calderon and Low (1998) found intact, two parent families to be positively correlated with significantly better academic and language outcomes. In a study by Geers and Brenner (2003), intact families in which parents regularly engaged their children with cochlear implants in family activities had a positive effect on the child's educational outcome 3 to 4 years after cochlear implantation.

- *Presence of older siblings*

This variable was explored due to the possible advantages to language acquisition of having an older sibling as a peer language model. Results indicated that slightly more of the children in the IE group had the advantage of an older sibling in the home. In general, most children in this study were the youngest and had older siblings (62-68%). This may indicate that parents were cautious about having another child, possibly due to genetic counseling or the financial, emotional and logistical demands placed on the parent by the needs of the child with hearing loss (Horsch, *et al.*, 1997). It may also indicate that, in general, the child was conceived at a higher maternal age than normal hearing siblings and thus at increased risk for congenital developmental difficulties. This possibility could be further explored by a study which includes maternal age in its questionnaire.

- *Number of languages spoken in the home*

The majority of families in both groups spoke one or two languages in the home, with the exception of one child who had emigrated from a neighbouring country. The first language was Afrikaans in the most families in the current study, followed by English. The second language was most often English, and less often Afrikaans. Sign language was included as a possible home language and was used in one home. Although South Africa is a multicultural and multilingual country, with many languages often spoken in a home, the present study did not reflect this as the clients of the PCIP tended to be of a smaller section of the population. The possible effects of language confusion on the language acquisition of a child with a cochlear implant were thus not significant in either group. Closer inspection revealed that in both groups, bilingual families attempted to speak only one language in the home to accommodate the participant. There was thus no difference in the two groups regarding the number of languages spoken in the home.

- *Speech intelligibility according to the Speech Intelligibility Rating Scale (SIR)*

The rating scale used, namely Speech Intelligibility Rating Scale (Allen, *et al.*, 1998) (SIR), grades speech intelligibility in 5 levels as follows.

1 indicates unintelligible speech; 2 indicates that intelligible speech is developing in single words in a known context; 3 means speech is intelligible to a listener who concentrates and lip-reads; 4 indicates speech which is intelligible to a listener without much prior experience of deaf speakers; and 5 means speech is intelligible to all listeners. In general, the SIR level reported was higher for preschool children in IE than in SE, and no children in IE had a rating indicating unintelligible speech. For the

primary school children, the difference became more pronounced with children in IE reported as having normal or near-normal speech, as opposed to the SE children whose ratings varied widely and tended to be lower. IE children in high school were all rated at the maximum score as being normally intelligible, where-as their peers in SE were generally rated as being more difficult to understand with ratings indicating that the listener would have to lip-read, be familiar with the speech of a deaf individual, and at times know the context. This variable showed a marked difference between the two groups, indicating that speech intelligibility is affected by, and affects, decisions regarding educational placement. Speech intelligibility may also be affected differently in these two groups by multiple variables such as auditory sensory input; survival of neurological elements in the cochlea and auditory tract; central auditory processing abilities; cognition and social pressure to produce intelligible speech (O'Donoghue, *et al.*, 1999). The nature of the linguistic environment and mode of communication used most frequently (Kirk, *et al.*, 2002) such as in bilingual-bicultural homes, and the quality of the input heard as well as the opportunities provided to practice intelligible speech (Moog, 2002) may all play a role.

- *Speech-language and occupational therapy: children currently and previously enrolled in therapy*

Of the preschoolers depicted in Tables 14 and 15, a larger percentage in IE was enrolled in speech-language therapy than those in SE. This could be due to the greater support given in class by specialised settings, or due to the increased pressure on children in IE to keep up with their normal hearing peers, requiring support services outside the school. However, as the ages of children increased, more children in SE were still making use of the services of a speech-language therapist in

primary and high school than those in IE. This appears to indicate that inclusive education required greater levels of support services initially, but that this tends to decrease as the child gets older and becomes fully integrated into the system. Most children in both groups indicated that they had previously been enrolled in speech-language therapy, regardless of whether they were currently enrolled in services or not. Closer inspection, however, revealed that all of the children in the IE group had received speech therapy at some stage previously while many, but not all, children in the SE group had been given access to this service. Since early communication intervention (ECI) has been shown to be crucial in later academic success (Blair, *et al.*, 1995; Mayne, *et al.*, 2000), this would appear to be a contributing variable in at least some of the children's eventual school placement. Access to ECI for some of this group may have been curbed due to the financial restraints related to the family's socio-economic status.

Children in both groups were making little use of the services of an occupational therapist at the time of the study, although many had in the past received these services. Occupational therapy seems, in general in this sample, to have played its major role in the earlier years of the child's development.

- *Telephone use*

As shown in Tables 14 and 15, in both the primary and high school groups, the vast majority of children in IE ( 80% and 100% respectively) could use a telephone to communicate successfully, while the percentages are smaller for the SE group (44% and 75% for the age groups respectively). This may give an indication of hearing with the cochlear implant, as well as central auditory processing of a less than ideal

signal such as that heard over a telephone line. A telephone conversation is an auditory-only situation, with no visual clues such as situation, non-verbal cues or lipreading and relies heavily on open set auditory discrimination and closure skills. This variable indicated that these skills were better developed in children in IE, perhaps allowing them to cope with the more difficult listening situation of an unadapted, non-specialised classroom. The Birmingham Paediatric Cochlear Implant Programme's 2000 outcomes report, 42% of the children were reported to be able to use a telephone with a known speaker after 6 years of implant use. Many of this group were reportedly 'borderline cases' with more residual hearing and open set speech perception prior to implantation (Birmingham Paediatric Cochlear Implant Programme, 2000). The questionnaire did not, however, distinguish between cellular/mobile telephones and land-line telephones.

- *Use of an FM system at school*

Of the preschool age children, almost half (45%) of those in SE used an FM system at school, while none of the IE group did. However, at primary school age only 22% of SE children were using an FM system as opposed to 60% of children in IE, presumably as the listening demands in an inclusive classroom increase in the formal education system where-as more adaptations are made for listening in SE classes. Assistive listening devices such as an FM system would be largely funded privately by parents, thus again a financial constraint may be operational in this variable. Of the high-school group, only one child (a participant in IE) was making use of an FM system. The results from the SE preschool group, however, are largely situation-specific since the specialised setting attended by many of the children has a strict policy regarding the use of standard school-issued FM systems in the classroom.



Thus the only reliable trend from this data is the large percentage of primary school children in IE (60%) making use of FM systems, as opposed to any other age or group.

The conclusion can be drawn that FM systems are still important after cochlear implantation in order to optimize the listening environment. This is true especially in inclusive primary school classrooms where no special acoustic adaptations are made, class sizes may be large, and children's auditory processing skills are still developing in order to cope with listening in background noise.

#### **Biological Contributing factors**

- *Prenatal and perinatal complications or conditions*

In general, more children in the SE group presented with prenatal or perinatal complications or conditions than children in the IE group: 39% of the SE group as opposed to 17% of the IE group (see Tables 14 and 15). Low birth weight, a significant risk factor for developmental delays (Gonzales, *et al.*, 1997), was present in only one participant. In the SE group, perinatal conditions were more prevalent than prenatal complications, while in the IE group the small number of cases was evenly distributed. These variables may have played a role in placing the children in the SE group at a developmental disadvantage due to their far-reaching implications (Rossetti, 1996), resulting in their placement in specialised educational settings. According to the JCIH (2000), any condition requiring NICU admission for 48 hours or longer is already a risk factor for sensorineural hearing loss, and is associated with additional disabilities such as learning, intellectual, attention, visual, motor (cerebral

palsy) and emotional disorders or conditions (Roush, Holcomb, Roush & Escolar, 2004).

- *Feeding difficulties in neonatal and early childhood stage*

According to Tables 14 and 15, the group which reported the largest incidence of feeding difficulties was the preschool SE group, although a small number of children in the IE preschool and primary school group also reported feeding difficulties. Early feeding is both indicative of sensory and motor functioning of the oral digestive and respiratory tract, as well as critical to adequate nutrition required for optimal neurological development. It also plays a vital role in infant-mother attachment which has an influence on communication development. The group experiencing these difficulties was thus at increased risk for developmental delays (Wooster, 1999). Closer inspection of the data revealed the majority of problems experienced by children as being related to oral motor weakness or inco-ordination of the suck-swallow-breathe mechanism, or sensory (tactile) defensiveness in the oral area leading to an aversion to feeding or rigidity in the types of foods that would be tolerated. Although only one child suffered from failure to thrive in the study, in some cases feeding difficulties may have led to some degree of under-nourishment. In the extreme degree, Wooster (1999) states that children who have been malnourished will limit their exploration and interaction and display social, cognitive, behavioral and physical developmental delays. These children may never fully recover from the effects of under-nutrition on the developing nervous system. When seen together with perinatal complications or conditions, children in this group would have a significant developmental disadvantage to the development of communicative and cognitive skills.

- *Developmental milestones (excluding speech and language milestones)*

Significantly more of the preschoolers and primary school children in the SE group were reported to have presented with delays in achieving developmental milestones than their peers in IE, as measured by the guidelines for milestones set by the Developmental Assessment Schema (Anderson, Nelson & Fowler, 1978). This may indicate the presence of additional delays in development of the children who were subsequently placed in special educational settings, in areas not directly related to the hearing loss such as motor; sensory and self help skill development. The presence of perinatal and feeding problems could have been causative factors in developmental delays, thus these variables should be seen as a cluster and interpreted together, with the presence of any one factor leading to closer inspection of the other related factors.

- *Hospitalisations*

Reasons for hospitalization were similar for the SE and IE groups, including both the causes of acquired hearing loss (meningitis) and most frequently illnesses affecting the respiratory and digestive systems. Many children in both groups received grommets due to chronic middle ear infections. The ‘trauma’ listed in the table refers to accidents excluding head injuries or skull fractures. The child with renotubular Fanconi syndrome had frequent skeletal fractures related to the disorder. One child in both groups required blood transfusion for infectious conditions. Items listed only in the SE group include failure to thrive (one child) mentioned earlier and strongly related to feeding difficulties in the perinatal and infant period; hospitalization due to high fever (one child) and accidental poisoning (one child).

4.2.4) Results and discussion relating to Sub-aim Four

**SUB-AIM 4: THE IDENTIFICATION OF VARIABLES AFFECTING THE SUCCESS OF CHILDREN WITH COCHLEAR IMPLANTS IN THE PCIP**

This sub-aim is fulfilled using the results and discussions arising from the division of the participant group into two groups based on educational outcomes. This is in order to provide a set of variables which can be linked to outcome measure of fully inclusive educational placement, which implies positive speech, language and auditory outcomes as demonstrated in the preceding results and discussions.

In general it can be seen that, in the children of this study, the variables listed in 4.2.4.1 are linked to positive outcomes.

*4.2.4.1: Variables associated with success with a cochlear implant for children in the PCIP: (as measured by placement in IE settings).*

These variables are represented in Table 16.

**Table 16: Ten most important variables associated with successful outcomes of children with cochlear implants in the PCIP**

- **Early age at diagnosis, intervention with hearing aids, and cochlear implantation**
- **Absence of prenatal and perinatal complications**
- **Higher level of maternal education**
- **Developmental milestones achieved at normal ages**
- **Absence of early feeding difficulties**
- **Use of an FM system in early schooling (primary school level)**
- **Access to ECI**
- **Presence of an older sibling**
- **Absence of birth trauma and congenital rubella syndrome as causes of hearing loss. (More successful outcomes when cause of hearing loss was hereditary and non-syndromic, or unknown)**
- **Later onset of hearing loss: acquired (few children)**

In order to provide clarity and structure, these variables can be integrated into a theoretical framework for early intervention. The framework of Guralnick (2001) was the conceptual model adapted for this goal, as illustrated in Figure 9 further on in the chapter. It should be noted that these factors can only be said to apply to the specific population of children in the PCIP. They can therefore be used by the PCIP's team of professionals to guide programme planning in terms of selection criteria, intervention decisions and counseling of realistic expectations in parents of children with cochlear implants. Each child and family's individual profile of risks, strengths and needs can be ascertained, and planning can be optimised.

#### **4.3 Implications from the study for universal newborn or infant hearing screening in South Africa: the limited use of high risk registers and the need for universal newborn/infant hearing screening and follow-up programmes**

The JCIH's position on universal newborn hearing screening as being the only reliable measure of targeting congenital and early acquired hearing loss, as opposed to the use of high risk registers, applies to all contexts where this level of technology is available. The JCIH found high risk registers to exclude from referral for hearing testing approximately half of all infants with sensorineural hearing loss. High risk registers, involving the list of risk factors set out by the JCIH (1990) have been reported to only identify approximately 50% of infants and neonates with severe to profound congenital or acquired sensorineural hearing loss (Mauk, *et al.*, 1995). The high risk register was, however, useful in deciding which infants to monitor over time for the possibility of a later progressing hearing loss. In South Africa, lobbying for the use of universal newborn hearing screening as opposed to the less expensive use of a high risk register requires the gathering of data to support this position. Table 17 demonstrates data gathered from the participants of the present study.

Although the JCIH released an updated list of factors recognized as putting neonates and infants at high risk for sensorineural hearing loss in 2000, the previous edition was used for this comparison as it is more descriptive and lists conditions more specifically.

**Table 17: The limited use of high risk registers: the PCIP data.**

<b>Risk criteria for neonates (0-28 days)</b>	<b>Number of children from the PCIP study with each risk indicator (n=45)</b>
• Family history of childhood hearing loss	n=8
• Congenital perinatal infection (CMV, rubella, herpes, toxomplasmosis, syphilis):	n=5
• Anatomical malformations of the head or neck	n=2
• Birthweight less than 1500g	n=1
• Hyperbilirubinaemia requiring blood transfusion:	n=2
• Bacterial meningitis:	n=0
• Severe asphyxia (Apgar 0-3):	n=1
• Ototoxic medications:	n=1
• Stigmata associated with relevant genetic syndromes:	n=4
• Prolonged mechanical ventilation (10 days or longer):	n=0
<b>TOTAL FOR ALL RISK INDICATORS:</b>	<b>n=24 (53%)</b>

Of the children in the present PCIP study, 24 (53%) met criteria on the neonatal high risk register, which corresponds with the percentages reported by the JCIH (1990 and 2000) and lends support to the use of neonatal, and follow-up, hearing screening in addition to the use of high risk registers. High risk registers are, however, important in providing crucial information in addition to the results of universal newborn hearing screening, including variables which would impact on the infant's later hearing and could be cause for monitoring for cochlear implant candidature at a later stage, as well as the need for more thorough testing for auditory neuropathy (Yoshinaga-Itano, 2000). The use of a high risk register with follow-up screening and monitoring is thus advocated. Swanepoel (2004) suggests that the Maternal and Child Health clinics, situated all over South Africa and providing free healthcare to children up to the age of six years, would be a more relevant and immediately available platform for the

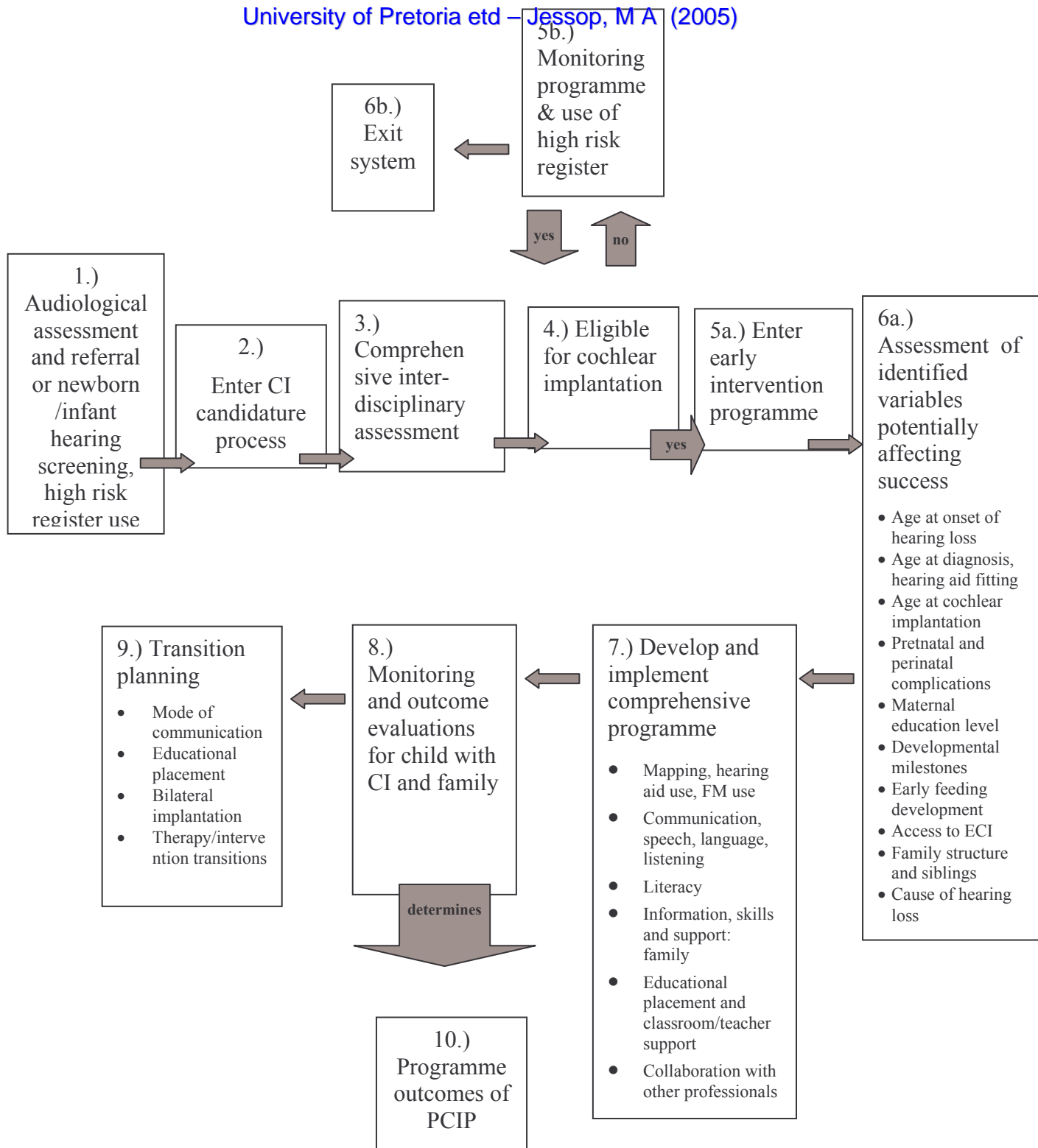
introduction of a universal infant hearing screening programme. These clinics are visited by mothers/caregivers and their infants from the six week immunisation onward, and have the advantage of local nurses, tasked by the Department of Health with the identification of children with special needs as well as health surveillance, who can be trained as sources of information and referral. Swanepoel (2004) further adds that the high risk register can be used in this context to document risk indicators in order to identify infants requiring audiological surveillance due to an increased risk of late onset or progressive hearing loss. In this model, the audiologist is seen as the agent of change, and the person carrying out the two stage oto-acoustic emission testing and immittance testing, while nurses and community volunteers are utilised as sources of information to mothers as well as the team members responsible for monitoring infants and completing high risk registers after training by the audiologist. Through information sharing and the acknowledgement of parent anxiety as a significant risk indicator, parents/caregivers are seen as essential and equal partners and are active, responsible participants in the process (Swanepoel, 2004). The universal screening thus changes from newborn to infant screening, and is carried out initially at a primary healthcare level (number 1 in Figure 9 to follow). Infants who do not pass the second screening are moved onto the secondary healthcare level, namely the regional hospital where diagnostic audiological facilities are available, and if need be to the tertiary level where the services of an ENT as well as specialised audiology services, such as auditory brainstem response testing, can be utilized (Swanepoel, 2004). It is from this level that the referral for cochlear implant candidacy assessment, the second step in the proposed developmental systems model (Figure 9 to follow), is made.



#### **4.4 A Developmental Systems Model for guiding the practical application of the results of the study in the PCIP**

Guralnick's 2001 Developmental Systems Model was designed to "guide early intervention programmes for vulnerable children and their families" (Guralnick, 2001: p.1). Key elements of the model were the emphasis on the family and on maximizing patterns of interaction in the family and minimizing child and family stressors, as well as on guiding research into practice (Guralnick, 2001). As these are all ideals of the implications of the present study for the PCIP, the model was selected for adaptation and application to the group of equally vulnerable children with cochlear implants and their families in the PCIP.

As a conclusion to the present study, the developmental systems model of Guralnick (2001) was thus adapted in order to design a proposed guideline for the process of identification, candidate selection, assessment, intervention and transition planning for children with cochlear implants and their families in the PCIP. The variables identified in this study to have the most direct bearing on the success of children, were incorporated in the model as they are unique and specific to the PCIP.



**Figure 9: Developmental Systems Model for identification, selection, assessment, intervention and monitoring of clients of the PCIP. Adapted from Guralnick (2001).**

As illustrated in the model (Figure 9), the outcomes of the child and family have a direct influence on the outcomes of the programme, thus the model provides a framework in which to measure programme outcomes. Since the profile of clients

(children and families) is likely to change in years to come as candidate selection criteria, funding variables and technology develop and shift, the specific assessment variables listed are expected to be different, however the elements of the model will remain unchanged. The model thus provides a standard framework for tracking the dynamic and expanding nature of cochlear implantation, and a cochlear implant programme and can support ongoing research in the PCIP. Equally important, it provides a sound theoretical basis for assessment, intervention and planning for each individual child and family. Furthermore, the model's implementation ensures that the group of infants and children who do not qualify for cochlear implantation but whose risk profiles indicate that they should be monitored for later re-assessment, do not exit the system prematurely or without a thorough review (Guralnick, 2001).

Such a model also provides the framework for a shared way of thinking and a shared vision by collaborating professionals from diverse fields (Guralnick, 2001), of early intervention for children with cochlear implants in the PCIP. The first stage of the system, namely hearing testing or screening, would be optimized in the case of congenital hearing loss or hearing loss with onset in the perinatal period, if it were newborn or infant hearing screening as well as the use of high risk registers (Swanepoel, 2004). These ideals are thus listed in the first step.

As illustrated in Figure 9, an infant or child enters the process after identification of his/her hearing loss and referral by an audiologist or other professional. In the ideal situation, which must be aimed toward, this first step would be characterized by universal newborn or infant hearing screening together with ongoing identification of all late-onset risk factors for later developing or progressive hearing loss (Swanepoel,

2004) as well as auditory neuropathy using a high risk register (JICH, 2000; Yoshinaga-Itano, 2000). The reason for this is that auditory neuropathy will not be identified by the use of screening oto-acoustic emission testing (OAE). As a group, these infants should be monitored for communication developmental milestones (JCIH, 2000) as they may not be immediate candidates for cochlear implantation, but will follow the route indicated by step 5b in Figure 9.

The second step in Figure 9 involves entry into the decision process regarding candidacy for cochlear implantation, which is a multi-disciplinary decision made after the next step, namely comprehensive assessment. In the PCIP this involves audiological, medical, radiological and social/emotional assessments. Should the child be a successful candidate for a cochlear implant (step 4), the child should then automatically enter the early intervention process (step 5a). The assessment of the child and family variables (step 6a) identified by the current study to be the most significant in leading to successful outcome after cochlear implantation and inclusion in a regular educational setting, and described in section 4.2.4.1 (relating to sub-aim four of the study), is then undertaken. The aim is to provide guidelines for the next step, namely the development and implementation of a comprehensive intervention programme for each individual child and family, based on their specific profile of variables and thus their needs (step 7) (Guralncik, 2001). Monitoring of the intervention process by the PCIP and regular outcome evaluations (step 8) allow planning for transitions (step 9) in educational placement; mode of communication, the intervention process including therapies and level of support; and decisions such as bilateral cochlear implantation. Steps 8 and 9, namely monitoring and outcome evaluations and the planning of transitions, are ongoing and dynamic in nature.

Outcome evaluations directly determine the outcomes of the cochlear implant programme as a whole, and can be used for programme evaluation and decision making. Should the child not be a candidate for a cochlear implant, then step 5b ensures regular monitoring and re-assessment of this status prior to exiting of the system (step 6b) should hearing loss not progress sufficiently to indicate the need for a cochlear implant. The use of high risk registers is important at this point, to identify factors which may impact on the later development of the child (JCIH, 2000) and indicating the possibility of later implantation.

#### **4.5 Summary**

The results and discussion of the current study explored the myriad of variables surrounding paediatric cochlear implantation as reported by parents in the PCIP, and identified a set of variables specifically common to the group of children attending regular, inclusive educational settings at the time of data collection, and their families. This educational placement was interpreted to indicate success with the cochlear implant, as inclusion in the present South African context requires a level of speech perception and production which enables the child with a cochlear implant to compete with his/her normal hearing peers at school. These variables were then used in a developmental systems model, intended to guide informed practice within the PCIP's protocol from the identification of hearing loss to exiting of the system. The study was nested in a family centred approach focusing on early intervention principles, and highlighted the need for early identification of hearing loss and prompt intervention efforts.

## CHAPTER FIVE

### CONCLUSIONS AND IMPLICATIONS

#### **5.1 Introduction**

In this chapter, the implications of the study to the theoretical database of the PCIP as well as its clinical practice will be discussed. The current study, although yielding much data on the children enrolled in the programme and their families, had limitations and these will be reported through a critical evaluation of the study. This is followed by recommendations and indicators of possible directions for future researchers interested in continuing with the important research in this area. The chapter's conclusions then close off the discussions regarding this study.

#### **5.2 Conclusions: Theoretical and clinical implications**

The children of this study, although differing widely in age, educational level and setting, and circumstances leading to cochlear implant, cannot be said to be representative of the South African population or the population of South African children with severe to profound bilateral hearing loss as a whole. In general, they are white, middle class children from nuclear, two-parent families where one or two languages, mainly Afrikaans and English, are spoken. Most live in and around Gauteng, and near to good infrastructure and services including medical services and rehabilitative services such as audiology, speech-language therapy, occupational and physiotherapy and counseling. In general they received their cochlear implants by the age of 5 years, and were in supportive educational settings which tend to be auditory-oral in philosophy (refer to Table 14). Most have parents and families with normal hearing, and older siblings. However, some important differences were seen

within the group, when one contrasted those in inclusive education, generally believed to be a major goal of early cochlear implantation, and those in specialised educational settings. Those in specialised settings were more likely to have started out with pre-, peri- or neonatal complications, and were more likely born to parents of lower educational status and socio-economic status. In general, their hearing losses were more likely to be the result of maternal rubella or birth trauma than those in the inclusive education group. They had less access to early services such as ECI and occupational therapy, and in general their hearing loss was diagnosed later, resulting in later hearing aid fitting and cochlear implantation than the other, more successfully integrated group. These children's parents were slightly more likely to have experienced academic and speech/language difficulties as children. Children in the specialised education group were less likely to have normally intelligible speech, less likely to be able to communicate using a telephone, and reportedly enjoyed music slightly less than the group in regular education.

### **5.3 Critical evaluation of the current study**

While this study produced large volumes of valuable data on 63% of the total number of children and families enrolled in the PCIP, it is only the first step in establishing an extensive and exhaustive database on the programme's entire population. The study further relied on parental report in some areas which would have been better assessed by a professional, such as speech intelligibility and mode of communication. Further investigation into the history surrounding the group of children whose cause of hearing loss is 'unknown', as well as the influence of maternal age at pregnancy and more extensive prenatal; perinatal and neonatal information would be beneficial in future studies of this nature, as these factors would have shed more light on the

influence of biological and medical variables in the study. More thorough analysis of the role of socio-economic status, which was not extensively analysed in the present study, may yield valuable results in further research. Possible shifts in educational placement were not investigated, and thus the current educational status of the participant may in some cases have been misleading. The scale and nature of support systems offered by the various schools were not determined, which may have caused variability within the groups used for comparative purposes namely those in inclusive education and those in specialised settings.

#### **5.4 Recommendations for future research**

The issue of side of implantation as well as binaural/bimodal stimulation requires further investigation as more research becomes available on this issue; and the population of children in the PCIP with bilateral cochlear implants should be closely followed as their numbers increase. These studies should be closely linked to issues of central auditory processing, and how this is affected by, or affects, bilateral stimulation as opposed to unilateral stimulation. Central auditory processing in the various syndromes that affect the population, such as Waardenburg Syndrome (Keats, 2002), could hold important information for rehabilitation and expectations. The ‘process variables’ of learning, attention and memory and their role in the success of cochlear implantees should receive attention.

Further, longitudinal studies are required to more closely examine the influence of age at implantation on the success of a child with a cochlear implant, as this variable was not adequately explored with the cross sectional nature of the present study. The success of the children in the inclusive education group within the school system



should be researched as much crucial information, such as the level of support offered by the school system in question, can be gained. Analysis of the audiological information, in particular amount of residual hearing, in relation to time (year in which the child was implanted as selection criteria shift over time) and success with cochlear implantation may yield guidelines for participant selection criteria. Lastly, cost effectiveness studies based on the information regarding funding for cochlear implant, use of support services and choice of educational placement would aid in motivating for earlier, more subsidised and more widespread cochlear implantation.

Together with evidence of the limited use of high risk registers and the need for mandatory newborn or infant hearing screening programmes (Swanepoel, 2004), this information may help to speed up the rate of early diagnosis of hearing loss, leading to earlier cochlear implantation. This is a world-wide phenomenon, reiterated by O'Donoghue, *et al.* (2000) in their statement that implementation of universal neonatal hearing screening programmes are imperative to ensure timely referral to cochlear implant centres. Summerfield & Marshall (1999) predicted that society could benefit from a net saving from cochlear implantation only if children with congenital hearing losses were implanted before their second birthday, leading to a vast increase in numbers of these children in regular education and later in successful employment.

## 5.5 Conclusion

In order to utilise the wealth of information rendered by this study, it is imperative that follow-up studies scrutinize key issues raised. In this way information can be transformed into educated action. The profile of a child with a cochlear implant in the PCIP, although not homogenous, does not represent the majority of children in South Africa, and as such the majority of children with severe to profound sensorineural hearing loss in this country. As cochlear implantation is still an expensive, elective procedure falling almost exclusively in the realm of the private health care sector in this country, this profile of clients is unlikely to change until state funding for cochlear implantation is secured. This can only be achieved by the concerted efforts of clinicians and researchers to generate similar studies on which to base strong motivation for cochlear implantation, neonatal hearing screening and follow-up programmes for all South African hospitals in addition to the use of high risk registers, and early intervention. As the clients of cochlear implant programmes change, so then will the variables affecting their success with a cochlear implant be affected, and therefore research of this nature would have to be dynamic to remain useful to the ongoing modification of selection criteria, individualised intervention plans and expected outcomes of children with cochlear implants in the PCIP. The study has provided descriptions and guidelines for putting theory into action within the framework of a developmental systems model for early intervention with children with cochlear implants in the PCIP. This model, however, requires an “unprecedented level of commitment and cooperation by all those involved: state and community agencies, professional groups and practitioners, parents, advocacy organizations, researchers, and institutions of higher education” (Guralnick, 2001: p.18). With the challenge of early identification of hearing loss, access to early intervention services

for all, and state funding enabling earlier implantation and the reaching of a more representative section of the population realised, the outcomes of cochlear implantation in South Africa may exceed the most optimistic expectations.

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**APPENDIX A:**  
**Consent form for Pilot Study**



Ingeligte toestemming: Loodsstudie

**Toestemmingsvorm**

Ek, (*naam in drukskrif*) \_\_\_\_\_ verstaan dat die inligting wat ek verskaf deur die vraelys te voltooi, as streng vertroulik hanteer sal word en dat dit gebruik sal word om die vraelys aan te pas en te verbeter vir die finale navorsingsstudie. Ek verstaan ook dat ek ten alle tye die volle reg het om te onttrek van die studie, sonder enige negatiewe gevolge.

Ek gee dus my toestemming dat die inligting in my lêer by die kogleêre inplantingsprogram sowel as die inligting deur my verskaf in die vraelys, vir die bogenoemde doel gebruik mag word. Ek verstaan ook dat Professor J.G. Swart, Hoof van die Pretoria Kogleêre Inplantingspan, sy toestemming hiervoor gegee het en dat hy die navorsing goedkeur en toegang tot die lêers van die Program toelaat.

Handtekening:

\_\_\_\_\_

**APPENDIX B:**  
**Letters relating to Ethics Committee**

(see bound hard copy)

**APPENDIX C:**  
**English and Afrikaans Questionnaires**

**Clients of the Pretoria Cochlear Implant Programme:  
Characteristics and perceived outcomes of children and their families.**

Column for office use

*Parent questionnaire: English version.*

*In this questionnaire, 'maternal parent or caregiver' refers to the child's mother or the primary female person taking care of the child (e.g. grandmother, stepmother, foster mother, female legal guardian etc.). 'Paternal parent or caregiver' refers to the child's father or primary male caregiver (e.g. foster father, grandfather, stepfather, male legal guardian etc). Some information can only be supplied by the child's biological mother (Section B.). 'Siblings' refers to brothers and sisters.*

Respondent number		
V1		1-2

**Section A) Biographical information**

**1) Name/s of maternal and paternal parents/caregivers:**


**2) In case of caregiver, state relationship to the child.**

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V2

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3

**3) Residential address of parents/caregivers:**


**4) Are the parents/ maternal & paternal caregivers:**

<b>Married</b>	
<b>Divorced</b>	
<b>Other, please specify:</b>	

V3

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4

**5) If the child has biological siblings, please fill in how many are older, younger or of the same age (e.g. twins).**

Number of older children	
Number of younger children	
Number of children of the same age (e.g. twins)	

V4

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5-6

V5

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7-8

V6

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9-10

**6) Who lives in the home with the child? Please tick all and state the number of people in each case.**

Maternal parent/caregiver	
Paternal parent/caregiver	
Brothers and sisters	
Other adults	
Other children	

V7

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11

V8

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12

V9

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13-14

V10

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15-16

V11

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17-18

Column for office use

**7) With whom does the child spend the most time? (tick one)**

Maternal parent or caregiver	
Paternal parent or caregiver	
Grandparent/s	
School/crèche	
Brothers and/or sisters	
Domestic helper	
Other- please specify	

V12 

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 19-20

**8) Which languages are spoken in the home? Please tick all applicable languages and indicate the main language with an 'M'.**

English	
Afrikaans	
Sotho	
Zulu	
Tswana	
Other-please specify	

V13	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			21
V14	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			22-23
V15	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			24-25
V16	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			26-27
V17	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			28-29
V18	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			30-31
V19	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			32-33

**9) Has the maternal parent/caregiver or paternal parent/caregiver been employed in the last 6 months? Please tick yes/no for each person.**

Maternal parent/caregiver	Yes	No
Paternal parent/caregiver	Yes	No

V20	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			34
V21	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			35

**10) What is the highest educational qualification achieved by the following people? Please tick one for each person.**

Highest educational achievement	Maternal parent or caregiver	Paternal parent or caregiver
No formal schooling		
Primary school		
Std 6-7		
Std 8		
Matric		
Tertiary diploma		
University degree		

V22	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			36
V23	<table border="1" style="display: inline-table; vertical-align: middle;"><tr><td style="width: 20px; height: 20px;"></td><td style="width: 20px; height: 20px;"></td></tr></table>			37

**Section B: history of pregnancy and birth**

**Please note that information for this section should be gathered**

**from the child’s biological mother where possible.**

**11) How would you describe your pregnancy?**

Normal progression, no complications		V24		38
Minor complications: please specify		V25		39-40
Severe complications or illness: please specify		V26		41-42

**12) Were you worried about anything during your pregnancy?**

Yes	If yes, please specify:	V27		43
		V28		44-45
No		V29		46-47

**13) Was the pregnancy planned?**

Yes	No	V30		48
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**14) How would you describe the birth of your child?**

Normal progression, no complications		V31		49
Minor complications: please specify		V32		50-51
Severe complications or illness: please specify		V33		52-53

**15) Describe the baby's condition after birth:  
Circle yes/no and specify as indicated.**

Apgar score	/10	V34			54-55
Birth weight	kg	V35			56-59
General condition	Good / moderate / poor	V36		60	
Oxygen required	Yes / No	V37		61	
Incubator	Yes / No	V38		62	
Mechanical ventilation	Yes / No Number of days:	V39			63-66
Time spent in neonatal ICU	Yes / No Number of days:	V40			67-70
Blood transfusion	Yes / No	V41		71	
Infections	Yes / No	V42		72	
Convulsions	Yes / No	V43		73	
High fever	Yes / No	V44		74	
Physical abnormalities of face or head	Yes / No	V45		75	
Abnormalities of body	Yes / No	V46		76	
Abnormalities of internal organs	Yes / No	V47		77	
Kidney disorder	Yes / No	V48		78	
Neonatal jaundice	Yes / No	V49		79	
Phototherapy	Yes / No Number of days:	V50			80-82
Other conditions: please describe		V51			83-84
		V52			85-86
		V53			87-88





**Section D: Developmental history**

**18) At what ages did the child achieve the following milestones? ( Please give ages in months.)**

Sit		V77			154-155
Crawl		V78			156-157
Stand		V79			158-159
Walk		V80			160-161
Dress self		V81			162-163
Feed self		V82			164-165
Use toilet		V83			166-167
Play with others		V84			168-169

**19) Did the child experience any feeding problems during early development (e.g. sucking, swallowing, chewing, or oversensitive mouth) ?**

Yes	Type of problem and age of child at the time:	V85		170	
No		V86			171-172
		V87			173-174

**20) If the hearing loss was not present from birth, would you describe your child's development prior to the onset of hearing loss as normal? ?**

Yes	No	V88		175
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**Section E: Family history**

**21) Does any other biological family member have a diagnosed hearing loss? Please tick.**

Mother		V89		176
Father		V90		177
Grandparent		V91		178
Brothers/sisters		V92		179
Other: specify		V93		180-181

**22) Do either maternal or paternal parent/caregiver consider themselves as part of the Deaf Community?**

Yes	No	V94		182
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**23) Is there a biological family history of the following? Please tick appropriate blocks to complete the table.**

	Mother	Father	Child's siblings		
Genetic syndrome: specify				V95	183-185
Academic difficulties				V96	186-188
Speech/language problems				V97	189-191

**Section F: History of child's hearing loss**

**24) At what age was the hearing loss suspected?**

months
--------

V98 

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 192-194

**25) Who first suspected the hearing loss? Tick one.**

Parents/caregivers	
Maternal parent/caregiver	
Paternal parent/caregiver	
Paediatrician	
Clinic sister	
Other: specify	

V99 

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 195-196

**26) At what age was a hearing loss diagnosed by an audiologist?**

Months:
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V100 

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 197-199

**27) What is the suspected cause of the hearing loss?**

**Please tick one**

Genetic syndrome	
Viral infection	
Meningitis	
Complications during pregnancy	
Birth trauma	
Hereditary hearing loss	
Hearing loss associated with kidney disorder	
Unknown	
Other: specify	

V101 

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 200-201

**28) Was the hearing loss progressive (i.e. did it worsen over time) ?**

Yes	No
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V102 

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 202

**29) Did the hearing loss fluctuate (i.e. improve and worsen over time)?**

Yes	No
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V103 

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 203

**30) At what age did the child first receive hearing aids?**

Months
--------

V104 

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 204-206

**31) What type of hearing aids were fitted?**

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**32) Were hearing aids fitted for both ears?**

Yes	No
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V105 



 207

**33) For how long did the child wear hearing aids?**

Months
--------

V106 



 208-210

**34) Who referred you to the cochlear implant programme?**

Relationship of referring person (e.g. mother, doctor, audiologist.)	
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V107 



 211

**35) When were you referred to the programme?**

Date :
--------

V108 



 212-217

**36) At what age (in months) did the child receive a cochlear implant?**

Months
--------

V109 



 218-220

**37) How were the funds raised for the cochlear implant?**

Funded by medical aid	
Private	
Donations	
Funded by state	
Other: please specify	

V110 



 221

V111 



 222

V112 



 223

V113 



 224

V114 



 225-226

V115 



 227-228

**38) Which ear was chosen for the implant?**

Left		State reason
Right		State reason
Bilateral		State reason

V116 



 229

V117 



 230-231

**39) Does the child wear a hearing aid on the non-implanted ear?**

Yes	No
-----	----

V118 



 232

**40) Please state the reason why the child does or does not wear a hearing aid on the non-implanted ear.**

--

V119 



 233-234

V120 



 235-236

**41) If the child has bilateral cochlear implants, were the two ears implanted simultaneously (in one operation) or in two consecutive operations? Tick one.**

Simultaneous (one operation)	
Consecutive	

V121  237

**42) Please supply the dates of the first and second cochlear implant operations (for bilateral implants).**

Date of first operation:	
Date of second operation:	

V122       238  
 V123       243  
 244  
 249

**43) In the case of bilateral implants, does the child wear both devices equally well?**

Yes	No
-----	----

V124  250

**44) Does the child use an FM system?**

Yes	Type:	
No		

V125  251

**Section G: Current level of functioning**

**45) Please describe the child's current educational setting: (Tick one.)**

At home with mother	
At home with nanny/ caregiver	
Mainstream playgroup or kindergarten	
Special playgroup or kindergarten for hearing impaired children: oral approach	
Special playgroup or kindergarten for hearing impaired children: signing or total communication approach	
Integrated playgroup or kindergarten: hearing and hearing impaired children.	
Mainstream primary school	
Integrated primary school for hearing impaired and normal hearing children: oral approach	
Primary school for children who are hard-of-hearing	
Mainstream high school	
High school for children who are hard-of- hearing	
School for the Deaf: signing approach	
Other, please specify	

V126   252-253

46) If the child attends a school, please supply the name of the school and state how long the child has been in the school:

	V127	<input style="width: 80%; height: 20px;" type="text"/>	254
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47) What portion of the day does the child wear his/her cochlear implant? (Tick one.)

All waking hours		V128	<input style="width: 80%; height: 20px;" type="text"/>	255
Only at school				
Only at home				
Infrequently/seldom				
Never				

48) How often does the child communicate using each of the following means of communication? Tick the appropriate column for each one.

	<i>Never</i>	<i>Sometimes</i>	<i>Often</i>			
Sounds				V129	<input style="width: 80%; height: 20px;" type="text"/>	256
Natural gestures				V130	<input style="width: 80%; height: 20px;" type="text"/>	257
Word approximations				V131	<input style="width: 80%; height: 20px;" type="text"/>	258
Single words				V132	<input style="width: 80%; height: 20px;" type="text"/>	259
Two word combinations				V133	<input style="width: 80%; height: 20px;" type="text"/>	260
Three word combinations				V134	<input style="width: 80%; height: 20px;" type="text"/>	261
Incomplete sentences				V135	<input style="width: 80%; height: 20px;" type="text"/>	262
Complete sentences				V136	<input style="width: 80%; height: 20px;" type="text"/>	263
Signs				V137	<input style="width: 80%; height: 20px;" type="text"/>	264
Speech plus signing				V138	<input style="width: 80%; height: 20px;" type="text"/>	265

49) How does the child react to the following sounds with his/her cochlear implant? (Tick one and indicate whether the child can simply hear the sound, or whether he/she can also identify what the sound is.)

Sound	Can hear	Can identify			
Music			V139	<input style="width: 80%; height: 20px;" type="text"/>	266
Environmental sounds outside (e.g. airplane, dog)			V140	<input style="width: 80%; height: 20px;" type="text"/>	267
Sounds in the home (e.g. vacuum cleaner)			V141	<input style="width: 80%; height: 20px;" type="text"/>	268
Speech			V142	<input style="width: 80%; height: 20px;" type="text"/>	269
Own name			V143	<input style="width: 80%; height: 20px;" type="text"/>	270
Family names			V144	<input style="width: 80%; height: 20px;" type="text"/>	271
Soft sounds (e.g. water)			V145	<input style="width: 80%; height: 20px;" type="text"/>	272
Whispered speech			V146	<input style="width: 80%; height: 20px;" type="text"/>	273

**50) Please indicate how well unfamiliar people can understand the child's speech (tick one option).**

Intelligibility	Tick block
The child's speech is understood by all listeners.	
Connected speech is understood by a listener with little experience of the speech of a hearing impaired person.	
Connected speech is understood by a listener who concentrates and lip-reads.	
Connected speech is not understood by others. (Intelligible speech is developing on single words when the listener lip-reads and understands the context.)	
Connected speech is unintelligible/not understood.	

V147  274

**51) Does the child currently receive any additional support services at the school (e.g. additional therapy, remedial teaching) ?**

Yes	If yes, describe type of service:
No	

V148  275  
 V149  276-277

**52) Has the child received speech and/or occupational therapy in the past?**

Speech therapy		
Yes		No
For how long was therapy attended? (e.g. 6 months)		
At what age did the child receive therapy?		
Occupational therapy		
Yes		No
For how long was therapy attended?		
At what age did the child receive therapy?		

V150  278  
 V151  279-280  
 V152  281-282  
 V153  283  
 V154  284-285  
 V155  286-287

**53) Does the child currently receive speech therapy?**

Yes
Name of therapist:
No

V156  288

**54) Does the child currently receive occupational therapy?**

Yes
Name of therapist:
No

V157  289

**55) Do you make use of the services of a psychologist for the child? (You may choose not to answer the question if you find it to be intrusive.)**

Yes
Name of psychologist:
No

V158  290

**56) Can the child communicate using a telephone?**

Yes	No
-----	----

V159  291

**57) Does the child enjoy music?**

Yes	No
-----	----

V160  292

*Thank you for completing the questionnaire.*

**Kliënte van die Pretoria Kogleêre Inplantingsprogram: Kenmerke en persepsies van uitkomst van kinders en hulle families.**

Kolom vir kantoorgebruik

***Ouervraelys: Afrikaanse weergawe***

*In hierdie vraelys verwys 'vroulike ouer of sorggewer' na die kind se moeder of die primêre vroulike persoon verantwoordelik vir die versorging van die kind (bv ouma, stiefma, wettige voog, pleegmoeder ens.). "Manlike ouer of sorggewer" verwys dan na die kind se vader of die primêre manlike persoon verantwoordelik vir die kind (bv oupa, stiefpa, pleegvader, wettige voog ens.). Sommige inligting kan slegs deur die kind se biologiese moeder verskaf word (bv. die in Afdeling B).*

Respondent nommer		
V1		1-2

**Afdeling A: Biografiese Inligting**

**1) Name van vroulike en manlike ouers of sorggewers:**


**2) Indien sorggewer, wat is u verwantskap met die kind?**

--

V2 

--

 3

**3) Woonadres van vroulike/manlike ouers of sorggewers:**


**4) Is die ouers / sorggewers: (merk asb.)**

Getroud	
Geskei	
Ander: spesifiseer asb.	

V3 

--

 4

**5) Indien die kind biologiese broers/susters het, dui asb. aan hoeveel is ouer, jonger of dieselfde ouderdom.**

Aantal ouer kinders	
Aantal jonger kinders	
Aantal kinders van dieselfde ouderdom (bv. 'n tweeling)	

V4 

--	--

 5-6

V5 

--	--

 7-8

V6 

--	--

 9-10



**6) Wie bly by die kind in die huis? Merk asb. alle toepaslike opsies en dui aan hoeveel mense in elke geval.**

Vroulike ouer of sorggewer	
Manlike ouer of sorggewer	
Broers en/of susters	
Ander volwassenes	
Ander kinders	

V7	<input type="text"/>	11	
V8	<input type="text"/>	12	
V9	<input type="text"/>	<input type="text"/>	13-14
V10	<input type="text"/>	<input type="text"/>	15-16
V11	<input type="text"/>	<input type="text"/>	17-18

**7) Met wie spandeer die kind die meeste tyd? (Merk een.)**

Vroulike ouer/sorggewer	
Manlike ouer/sorggewer	
Ouma/oupa	
Skool/creche	
Broers en susters	
Huishulp	
Ander, spesifiseer asb.	

V12	<input type="text"/>	<input type="text"/>	19-20
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**8) Watter tale word in die huis gepraat? Merk asb. alle toepaslike antwoorde en dui die hooftaal aan met 'n H.**

Engels	
Afrikaans	
Sotho	
Zulu	
Tswana	
Ander, spesifiseer asb.	

V13	<input type="text"/>	21	
V14	<input type="text"/>	<input type="text"/>	22-23
V15	<input type="text"/>	<input type="text"/>	24-25
V16	<input type="text"/>	<input type="text"/>	26-27
V17	<input type="text"/>	<input type="text"/>	28-29
V18	<input type="text"/>	<input type="text"/>	30-31
V19	<input type="text"/>	<input type="text"/>	32-33

**9) Het die vroulike/manlike ouer/sorggewer binne die afgelope 6 maande gewerk en 'n inkomste verdien? Merk asb.**

Vroulike ouer/sorggewer	Ja	Nee
Manlike ouer/sorggewer	Ja	Nee

V20	<input type="text"/>	34
V21	<input type="text"/>	35

**10) Wat is die hoogste opvoedkundige kwalifikasie wat deur die volgende mense bereik is? Merk asb. een.**

Hoogste opvoedkundige kwalifikasie	Vroulike ouer of sorggewer	Manlike ouer of sorggewer
Geen formele onderrig		

V22	<input type="text"/>	36
V23	<input type="text"/>	37

Laerskool		
St 6-7		
St 8		
Matriek		
Tersiêre diploma		
Universiteitsgraad		

**Afdeling B) Geskiedenis van swangerskap en geboorte**

**Neem asb kennis dat inligting vir hierdie afdeling verkieslik by die kind se biologiese moeder verkry moet word.**

**11) Hoe sou u die swangerskap beskryf?**

Normale verloop, geen komplikasies		V24		38
Geringe komplikasies: spesifiseer		V25		39-40
Ernstige komplikasies of siekte: spesifiseer		V26		41-42

**12) Was u oor iets bekommerd tydens u swangerskap?**

Ja	Indien ja, spesifiseer:	V27		43
		V28		44-45
Nee		V29		46-47

**13) Was dit 'n beplande swangerskap?**

Ja	Nee	V30		48
----	-----	-----	--	----

**14) Hoe sou u die geboorte van u kind beskryf?**

Normale verloop, geen komplikasies		V31		49
------------------------------------	--	-----	--	----

Geringe komplikasies: spesifiseer		V32			50-51
Ernstige komplikasies of siektes: spesifiseer		V33			52-53

**15) Beskryf asb. die baba se toestand na geboorte: omkring ja/nee en spesifiseer soos aangedui.**

Apgartelling	/10	V34			54-55
Geboortegewig	kg	V35			56-59
Algemene toestand	Goed / gemiddeld / swak	V36		60	
Suurstof ontvang	Ja/nee	V37		61	
Broeikas	Ja/nee	V38		62	
Meganiese ventilasie	Ja/nee Hoeveel dae:	V39			63-66
Tyd in Neonatale Intensiewe Sorgseenheid	Ja/Nee Hoeveel dae:	V40			67-70
Bloedoortapping	Ja/nee	V41			71
Infeksies	Ja/nee	V42			72
Konvulsies	Ja/nee	V43			73
Hoë koors	Ja/nee	V44			74
Fisiese abnormaliteite van kop/gesig	Ja/nee	V45			75
Abnormaliteite van liggaam	Ja/nee	V46			76
Abnormaliteite van interne organe	Ja/nee	V47			77
Nier-afwykings	Ja/nee	V48			78
Neonatale geelsug	Ja/nee	V49			79
Fototerapie	Ja/nee Hoeveel dae:	V50			80-82
Ander toestande: beskryf asb		V51			83-84

		V52
		V53

		85-86
		87-88

**Afdeling C: Mediese geskiedenis van kind**

**16) Het die kind 'n geskiedenis van die volgende toestande? Merk en spesifiseer asb. soos aangedui.**

Middeloor-infeksies	Hoeveel keer: Spesifiseer behandeling:	V54	89				
		V55	90				-91
		V56	92				93
Epilepsie		V57	94				
Genetiese sindroom	Spesifiseer	V58	95-	96-	97-		
Asma		V59	98				
Allergieë	Spesifiseer	V60	99-	100-	101-	102-	
Nier-probleme		V61	104				
Herhaaldelike boonste lugweg infeksies		V62	105				
Fisiese/spier-afwyking	Spesifiseer	V63	106-	107-	108-	109-	
Aandagafleibaarheid /hiperaktiwiteits afwyking	Spesifiseer behandeling:	V64	111-				113
Gedragsprobleme		V65	114				
Afwyking van oë/visie	Spesifiseer	V66	115-	116-	117-	118-	
Kroniese siekte	Spesifiseer	V67	120-	121-	122-	123-	
Masels		V68	125				
Meningitis		V69	126				
Pampoentjies		V70	127				
Ander	Spesifiseer	V71	128-				129
		V72	130-				131

**17) Was die kind al in die hospitaal opgeneem?**

Ja	Nee
----	-----

V73  132

**Indien ja, voltooi asb. die tabel:**

Ouderdom tydens hospitalisasie	Rede vir hospitalisasie	Aantal dae in hospitaal spandeer

V74							133-139
V75							140-146
V76							147-153

**Afdeling D) Ontwikkelingsgeskiedenis**

**18) Op watter ouderdomme het die kind die volgende mylpale bereik? (Verskaf ouderdomme in maande asb.):**

Sit	
Kruip	
Staan	
Loop	
Self aantrek	
Self voer	
Toilet gebruik	
Speel met ander	

V77			154-155
V78			156-157
V79			158-159
V80			160-161
V81			162-163
V82			164-165
V83			166-167
V84			168-169

**19) Het die kind voedingsprobleme ondervind tydens vroeë ontwikkeling (bv. suig, sluk, kou of oorsensitiewe mond)?**

Ja	Tipe probleem en ouderdom van kind
Nee	

V85		170	
V86			171-172
V87			173-174

**20) Indien die gehoorverlies later ontstaan het, sou u die kind se ontwikkeling voor die gehoorverlies as normaal beskryf?**

Ja	Nee
----	-----

V88		175
-----	--	-----

**Afdeling E) Familiageskiedenis**

**21) Het enige ander biologiese familielid 'n gediagnoseerde gehoorverlies? Merk asb.**

Moeder	
Vader	
Oupa/Ouma	
Broers/susters	
Ander, spesifiseer	

V89		176	
V90		177	
V91		178	
V92		179	
V93			180-181

**22) Beskou een/albei ouers of sorggewers hulleself as deel van die Dowe Gemeenskap?**

Ja	Nee
----	-----

V94		182
-----	--	-----

**23) Is daar 'n biologiese familiegeskiedenis van een van die volgende? Voltooi asb. die tabel deur die toepaslike blokkies te merk.**

	Moeder	Vader	Kind se broers/susters
Genetiese sindroom: spesifiseer			
Akademiese probleme			
Spraak/taal probleme			

V95	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	183-185
V96	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	186-188
V97	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	189-191

***Afdeling F) Geskiedenis van die kind se gehoorverlies***

**24) Op watter ouderdom is die gehoorverlies vermoed?**

maande
--------

V98	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	192-194
-----	--------------------------	--------------------------	--------------------------	---------

**25) Wie het eerste die gehoorverlies vermoed? Merk een.**

Ouers/sorggewers	
Vroulike ouer/sorggewer	
Manlike ouer/sorggewer	
Pediater	
Klinieksuster	
Ander, spesifiseer	

V99	<input type="checkbox"/>	<input type="checkbox"/>	195-196
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**26) Op watter ouderdom is die gehoorverlies deur 'n audioloog gediagnoseer?**

Maande :
----------

V100	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	197-199
------	--------------------------	--------------------------	--------------------------	---------

**27) Wat is vermoedelik die oorsaak van die gehoorverlies? Merk een asb.**

Genetiese sindroom	
Virale infeksie	
Meningitis	
Komplikasies tydens swangerskap	
Trauma gedurende geboorte	
Oorerflike gehoorverlies	
Gehoortverlies gekoppel aan nierafwyking	
Onbekend	
Ander, spesifiseer asb.	

V101	<input type="checkbox"/>	<input type="checkbox"/>	200-201
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**28) Was die gehoorverlies progressief  
(m.a.w. het dit met die tyd vererger) ?**

Ja	Nee
----	-----

V102  202

**29) Het die gehoorverlies gefluktueer  
(m.a.w. verbeter en verswak met die tyd) ?**

Ja	Nee
----	-----

V103  203

**30) Op watter ouderdom het die kind die eerste keer  
gehoorapparaat ontvang?**

Maande
--------

V104  204-206

**31) Watter tipe gehoorapparaat is gepas?**

--

**32) Is albei ore met gehoorapparaat gepas?**

Ja	Nee
----	-----

V105  207

**33) Hoe lank het die kind gehoorapparaat gedra?**

Maande
--------

V106  208-210

**34) Wie het u verwys na die kogleêre inplantingsprogram?**

Verwantskap van verwysende persoon (bv. ma, dr, oudioloog, ens.)	
--	--

V107  211

**35) Wanneer is u na die program verwys?**

Datum :
---------

V108  212-217

**36) Op watter ouderdom (in maande) het die kind 'n kogleêre inplanting ontvang?**

Maande
--------

V109  218-220

**37) Hoe is die fondse ingesamel vir die inplanting?**

Befonds deur mediese fonds	
Privaat	
Donasies	
Deur staat befonds	
Ander, spesifiseer asb.	

V110	<input type="text"/>	221
V111	<input type="text"/>	222
V112	<input type="text"/>	223
V113	<input type="text"/>	224
V114	<input type="text"/> <input type="text"/>	225-226
V115	<input type="text"/> <input type="text"/>	227-228

**38) Watter oor is gekies vir die inplanting?**

Links	Noem rede
Regs	Noem rede
Bilateraal	Noem rede

V116


229

V117


230-231

**39) Dra die kind 'n gehoorapparaat op die nie-geïnplanteerde oor?**

Ja	Nee
----	-----

V118

--

232

**40) Noem asb. die rede waarom die kind 'n gehoorapparaat op die nie-geïnplanteerde oor dra of nie dra nie.**

--

V119

--	--

233-234

V120

--	--

235-236

**41) Indien die kind bilaterale kogleêre inplantings het, is die twee ore gelyk geïnplanteer, of in twee afsonderlike operasies (opeenvolgend)? Merk een.**

Gelyk (een operasie)	
Opeenvolgend	

V121

--

237

**42) Gee asb. die datums van die eerste en tweede kogleêre inplantings-operasies (vir bilaterale inplantings):**

Datum van eerste operasie:	
Datum van tweede operasie	

V122

--	--	--	--	--	--

238

243

V123

244

249

**43) In die geval van bilaterale inplantings, dra die kind albei toestelle ewe goed?**

Ja	Nee
----	-----

V124

--

250

**44) Gebruik die kind 'n FM sisteem?**

Ja	Tipe:
Nee	

V125

--

251

***Afdeling G) Huidige vlak van funksionering***



**45) Beskryf asb. die kind se huidige opvoedkundige plasing: (merk een)**

Tuis by moeder		V126	<table border="1"> <tr> <td></td> <td></td> <td>252-253</td> </tr> </table>			252-253
				252-253		
Tuis by oppasser/sorggewer						
Hoofstroom speelgroep of kleuterskool						
Spesiale speelgroep of kleuterskool vir hardhorende kinders: orale benadering						
Spesiale speelgroep of kleuterskool vir hardhorende kinders: gebaretaal of ' <i>total communication</i> ' benadering (gebare plus spraak)						
Geïntegreerde speelgroep of kleuterskool: horende en gehoorgestremde kinders.						
Hoofstroom laerskool						
Geïntegreerde laerskool vir gehoorgestremde leerders en horende kinders: orale benadering						
Laerskool vir hardhorende kinders						
Hoofstroom hoërskool						
Hoërskool vir hardhorende kinders						
Skool vir Doves: gebaretaal benadering						
Ander, spesifiseer asb						

**46) Indien die kind in 'n skool is, verskaf asb. die naam van die skool en hoe lank die kind al in die skool is:**

	V127		254
--	------	--	-----

**47) Watter gedeelte van die dag dra die kind sy/haar kogleêre inplanting? (Merk een.)**

Ten alle tye		V128	<table border="1"> <tr> <td></td> <td>255</td> </tr> </table>		255
	255				
Slegs by die skool					
Slegs by die huis					
Selde / ongereeld					
Nooit					

**48) Hoe dikwels kommunikeer die kind deur gebruik te maak van die volgende wyses van kommunikasie? Merk asb. die toepaslike kolom.**

	<i>Nooit</i>	<i>Soms</i>	<i>Dikwels</i>			
Klanke				V129		256
Natuurlike gebare				V130		257
Woordbenaderings				V131		258

Enkelwoorde				V132		259
Twee-woordkombinasies				V133		260
Drie-woordkombinasies				V134		261
Onvolledige sinne				V135		262
Volledige sinne				V136		263
Gebare				V137		264
Spraak plus gebare				V138		265

**49) Hoe reageer die kind op die volgende klanke met sy/haar implanting? (Merk en dui aan of die kind slegs die klank hoor, en of hy/sy dit ook kan identifiseer en weet wat dit is.)**

Klank	Kan hoor	Kan Identifiseer			
Musiek			V139		266
Omgewingsklanke buite (bv. vliegtuig, hond wat blaf)			V140		267
Klanke binne die huis (bv. stofsuier, mikrogolf)			V141		268
Spraak			V142		269
Eie naam			V143		270
Familie se name			V144		271
Sagte klanke (bv. blare, water)			V145		272
Fluisterspraak			V146		273

**50) Dui asb. aan hoe goed vreemde persone die kind se spraak kan verstaan. (Merk een.)**

Verstaanbaarheid	Merk blokkie			
Die kind se spraak word deur alle luisteraars verstaan.		V147		274
Aaneenlopende spraak word deur 'n luisteraar met min ervaring van die spraak van 'n gehoorgestremde persoon verstaan.				
Aaneenlopende spraak word verstaan deur 'n luisteraar wat konsentreer en liplees.				
Aaneenlopende spraak word nie deur ander verstaan nie. (Verstaanbare spraak ontwikkel op enkelwoorde wanneer die luisteraar liplees en die konteks verstaan.)				
Aaneenlopende spraak is onverstaanbaar.				

**51) Ontvang die kind tans enige hulpdienste by die skool (bv. addisionele terapie, remediërende onderrig)?**

Ja	Indien ja, beskryf die tipe diens:	V148	<input type="text"/>	275	
Nee		V149	<input type="text"/>	<input type="text"/>	276-277

**52) Het die kind in die verlede spraakterapie en/of arbeidsterapie ontvang?**

<b>Spraakterapie</b>					
Ja		Nee			
Vir hoe lank (bv. 6 mnde) is terapie bygewoon?	<input type="text"/>		V150	<input type="text"/>	278
Op watter ouderdom het die kind terapie ontvang?	<input type="text"/>		V151	<input type="text"/>	279-280
			V152	<input type="text"/>	281-282
<b>Arbeidsterapie</b>					
Ja		Nee			
Vir hoe lank is terapie bygewoon? (bv. 6 mnde)	<input type="text"/>		V153	<input type="text"/>	283
Op watter ouderdom het die kind terapie ontvang?	<input type="text"/>		V154	<input type="text"/>	284-285
			V155	<input type="text"/>	286-287

**53) Woon die kind tans spraakterapie by?**

Ja	Naam van terapeut:	V156	<input type="text"/>	288
Nee				

**54) Woon die kind tans arbeidsterapie by?**

Ja	Naam van terapeut:	V157	<input type="text"/>	289
Nee				

**55) Maak u gebruik van die dienste van 'n sielkundige vir die kind? (U kan kies om die vraag te ignoreer.)**

Ja	Naam van sielkundige	V158	<input type="text"/>	290
Nee				

**56) Kan die kind met behulp van 'n telefoon kommunikeer?**

Ja	Nee
----	-----

V159  291

**57) Geniet die kind musiek?**

Ja	Nee
----	-----

V160  292

*Dankie dat u die vraelys voltooi het.*

**APPENDIX D:**  
**Covering Letters of Questionnaires**

Dear Parents

My name is Marguerite Jessop. I am completing a research project for the Pretoria Cochlear Implant Programme and as part of my Masters degree, gathering information about all the children with cochlear implants in the Pretoria Cochlear Implant Programme. The aim of the project is to profile and describe the defining characteristics of the children implanted at the Pretoria Cochlear Implant Programme and their families. With this information we can determine the success of our programme compared to programmes world wide, and can begin to understand which factors make a programme more successful for the children involved and which factors affect the success of a child with a cochlear implant. We can then plan for the future more effectively.

With the new data we can also update and complete each of your files, so that our service to you can be faster and more efficient and informed. Further, your data will help us to write motivational letters for funding substantiated by facts about the success of early cochlear implantation in children. The information will be treated as strictly confidential, and will be used only by the cochlear implant team.

Enclosed please find a questionnaire concerning your child with a cochlear implant. I would be very grateful if you would complete the questionnaire and return it by post, using the postage paid return addressed envelopes. No further involvement will be expected from you. Upon completion of my study, the results of the processing of this information will be made available to you should you be interested

**Please note that all the information will be treated as confidential and no names will be used in presentation of the results of the study. Names will be used purely to update the records of the Pretoria Cochlear Implant Programme. Although complete sets of information are preferable to the study, you retain the right to choose not to answer questions with which you are not comfortable. You also have the right to withdraw from the study at any time. You are kindly requested to sign the consent form enclosed after you have read this letter and are comfortable with participating in the study. Please send the tear-off slip of the signed consent form back to me with the completed questionnaire.**

Instructions are given at the start of each question regarding how to answer it. Most questions are answered by simply ticking the appropriate block provided. When asked to 'specify', please provide more information regarding the question (e.g. the name of a specific genetic syndrome, the specific complicating factors during the birth of your child). If the child is not living with his/her biological mother I ask, if possible, that she please be consulted regarding the questions on pregnancy and birth in Section B of the questionnaire for accuracy's sake. Although I have attempted to make the questionnaire as time-efficient and user friendly as possible, I do realise that most of us have little time available and I sincerely appreciate the time you take to provide us with this valuable information.

Kind regards,

Marguerite Jessop

Speech/language therapist & Audiologist: University of Pretoria

Tel (012) 667 3998

Geagte Ouers

My naam is Marguerite Jessop. Ek is besig met 'n navorsingsprojek vir die Pretoria Kogleêre Inplantingsprogram, as deel van my Meestersgraad. Dit behels dat ek inligting aangaande al die kinders met kogleêre inplantings in die Pretoria Kogleêre Inplantingsprogram insamel. Die doel van die projek is om 'n profiel saam te stel, en om die kenmerke van die kinders met kogleêre inplantings in die program, asook hulle gesinne, te beskryf. Hierdie inligting sal ons help om die sukses van ons program te vergelyk met die van programme wêreldwyd, en om die faktore te begin verstaan wat 'n program meer suksesvol kan maak asook die faktore wat 'n invloed het op die sukses van 'n kind met 'n kogleêre inplanting. Ons sal dus in staat wees om meer effektief vir die toekoms te beplan.

Met die nuwe data kan ons ook elkeen van ons kliënte se lêers op datum bring, sodat 'n meer ingeligde, vinniger en meer effektiewe diens aan julle gelewer kan word. Julle data sal verder gebruik word om motiverende briewe te skryf vir befondsing vir kogleêre inplantings, gesteun deur feite oor die sukses van kogleêre inplantings in kinders. Die inligting sal as streng vertroulik hanteer word, en sal slegs deur die kogleêre span gebruik word. Ingesluit vind asseblief 'n vraelys aangaande u kind met 'n kogleêre inplanting. Ek sal baie dankbaar wees indien u die vraelys voltooi en terugstuur per pos, in die kovert wat ingesluit is. Geen verdere betrokkenheid sal van u verwag word nie. Die resultate van die studie sal aan u bekend gemaak word indien u sou belangstel.

Neem asseblief kennis dat die inligting as streng vertroulik hanteer sal word, en dat geen name gebruik sal word in die bekendstelling van die resultate nie. Name sal slegs gebruik word om die lêers van die Pretoria Kogleêre Inplantingsprogram op datum te bring. Alhoewel volledige stelle inligting verkieslik is vir die studie, het u die reg om enige vrae te ignoreer en uit te laat. U het ook die reg om ten alle tye te onttrek van die studie, sonder enige negatiewe gevolge. U word vriendelik versoek om die toestemmingsvorm te teken nadat u hierdie brief gelees het, en indien u gemaklik voel om aan die studie deel te neem. Skeur asseblief dan die strokie af en pos dit saam met die vraelys terug.

Instruksies is aan die begin van elke vraag gegee oor hoe om die vraag te beantwoord. Die meeste vrae word beantwoord deur slegs die toepaslike blokkie te merk. Wanneer u gevra word om te 'spesifiseer', moet u asseblief meer inligting verskaf (bv. die naam van 'n genetiese sindroom, of die spesifieke kompliserende faktore gedurende die geboorte van u kind). Indien die kind nie by die biologiese moeder woon nie, vra ek dat die moeder sover moontlik gekonsulteer word om die vrae oor swangerskap en geboorte in Afdeling B te voltooi, vir volledigheid. Alhoewel ek probeer het om die vraelys so tydseffektief en gebruikersvriendelik as moontlik te hou, verstaan ek dat die meeste van ons min tyd tot ons beskikking het en ek waardeer die tyd wat u bestee om die vraelys te voltooi.

Vriendelike groete  
Marguerite Jessop  
Spraak/taal-Terapeut en Oudioloog: Universiteit van Pretoria  
Tel (012) 667 3998

APPENDIX E:  
Informed Consent Forms



**Consent Form**

I, (*print name*) \_\_\_\_\_ understand that the information which I supply in completing this questionnaire will be treated as strictly confidential, and will be used for the purposes of research, updating files, and contributing to the efficacy of motivating for the funding of cochlear implantation. I also understand that I have the right to withdraw from the study at any time, with no negative consequences.

I thus give my permission that the information in my client file at the Pretoria Cochlear Implant Programme and the information which I provide by completing this questionnaire may be used for the purposes stated above. I also understand that Professor J.G. Swart, Head of the Pretoria Cochlear Implant Programme, has given his consent that the research may be conducted and records may be accessed.

Signature:

\_\_\_\_\_

*(Tear return slip off here)*

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**Please tear off this section and return in the envelope with the questionnaire.**

I, (*print name*) \_\_\_\_\_

**do / do not give my informed consent to participate willingly in the study by completing the questionnaire.**

**Signature of parent/caregiver:** \_\_\_\_\_

Toestemmingsvorm

Ek, (*naam in drukskrif*) \_\_\_\_\_ verstaan dat die inligting wat ek verskaf deur die vraelys te voltooi, as streng vertroulik hanteer sal word en dat dit gebruik sal word vir navorsing, om lêers op datum te bring, en om by te dra tot motivering vir befondsing van kogleêre inplantings. Ek verstaan ook dat ek ten alle tye die reg het om te onttrek van die studie sonder enige negatiewe gevolge.

Ek gee dus my toestemming dat die inligting in my kiëntelêr by die Pretoria Kogleêre Inplantingsprogram asook die inligting wat ek verskaf deur die vraelys te voltooi, gebruik mag word vir die bogenoemde doel. Ek verstaan ook dat Professor J.G. Swart, Hoof van die Pretoria Kogleêre Inplantingsprogram, sy toestemming gegee het vir toegang tot die lêers en dat die navorsing uitgevoer mag word.

Handtekening:

\_\_\_\_\_

*(skeur strokie hier af)*

**Skeur asb. hierdie gedeelte af en stuur dit saam met die vraelys terug.**

Ek, (*naam in drukskrif*) \_\_\_\_\_  
**gee ingeligte toestemming / gee nie toestemming nie om gewillig aan die navorsing deel te neem deur die vraelys te voltooi.**

**Handtekening van ouer/sorggewer:** \_\_\_\_\_