PREDICTORS OF PEDIATRIC COCHLEAR IMPLANTATION OUTCOMES IN SOUTH AFRICA

Talita le Roux [a], Bart Vinck [a,b], Iain Butler [c], Nicolize Cass [d], Leone Nauta [e], Liebie Louw [f], Dani Schlesinger [g], Maggi Soer [a], Mashudu Tshifularo [h], De Wet Swanepoel [a, i, j],

[a] *Department of Speech-Language Pathology and Audiology, University of Pretoria, South Africa*

[b] *Speech-Language Audiology Department, Ghent University, Belgium*

[c] *Department of Otorhinolaryngology, University of the Free State, Bloemfontein, South Africa*

[d] *Nicolize Cass, Pretoria Cochlear Implant Unit, South Africa*

[e] *Leone Nauta, Johannesburg Cochlear Implant Program, South Africa*

[f] *Liebie Louw, Department of Statistics, University of Pretoria, South Africa*

[g] *Dani Schlesinger, Department of Speech Therapy and Audiology, Chris Hani Baragwanath Academic Hospital, Soweto, South Africa*

[h] *Department of Otorhinolaryngology, Steve Biko Academic Hospital, University of Pretoria, South Africa*

[i] *Ear Sciences Centre, School of Surgery, University of Western Australia, Nedlands, Australia*

[j] *Ear Science Institute Australia, Subiaco, Australia*

Corresponding author:

Talita le Roux

Department of Speech-Language Pathology and Audiology

University of Pretoria

c/o Lynnwood and University Road

Hatfield

South Africa

0002

talita.leroux@up.ac.za

ABSTRACT

Objective: To identify and describe predictors of pediatric cochlear implantation outcomes in a South African population.

Methods: A retrospective study of 301 pediatric CI recipients from five cochlear implant programs was conducted and cross-sectional outcome data were added at the time of data collection. Twenty potential prognostic factors were identified from the retrospective dataset, including biographical, cochlear implant (CI), family and risk factors. Multiple regression analyses was performed to identify predictor variables that influence outcomes in terms of auditory performance (CAP scores), speech production (SIR scores), communication mode and educational placement.

Results*:* Although implanted children within this sample did not have equal opportunity to access a second implant, bilateral implantation was strongly predictive of better auditory performance and speech production scores, an oral mode of communication and mainstream education. NICU admittance/ prematurity were associated with poorer auditory performance and speech production scores, together with a higher probability for non-oral communication and non-mainstream education. The presence of one or more additional developmental condition was predictive of poorer outcomes in terms of speech production and educational placement, while a delay between diagnosis and implantation of more than one year was also related to non-mainstream education. Ethnicities other than Caucasian were predictive of poorer auditory performance scores and a lower probability for mainstream education.

Conclusion: An extensive range of prognostic indicators were identified for pediatric CI outcomes in South Africa. These predictive factors of better and poorer outcomes should guide pediatric CI services to promote optimal outcomes and assist professionals in providing evidence-based informational counselling.

Keywords: pediatric cochlear implantation, prognostic factors, cochlear implant, children, outcomes

Abbreviations: CAP, Categories of Auditory Performance; CI, cochlear implant; HL, hearing loss; NICU, neonatal intensive care unit; SASL, South African Sign Language; SIR, Speech Intelligibility Rating

INTRODUCTION

In recent years, significant improvement has been demonstrated in pediatric cochlear implant (CI) outcomes due to technological advances, earlier implantation and earlier intervention [1-3]. Speech and language skills comparable to normal hearing children can be achieved in some prelingually deaf children implanted within the first year of life, as indicated by recent reports [4-6]. Understandably, expectations for pediatric cochlear implantation are high [1]. However, outcomes vary as multiple internal and external factors have the potential to affect clinical outcomes [7-9]. As a result many pediatric cases present with sub-optimal outcomes. In order to counsel families pre-operatively about the range of possible outcomes and to plan for post-implantation intervention, accurate prognostic information is required [10,11].

Indications for pediatric cochlear implantation are becoming more complex with an increase in bilateral implantation and a growing number of children with less severe hearing losses being implanted [10,12-15]. Also, children with multiple medical conditions resulting from prematurity or perinatal etiologies are more likely to be considered as candidates, expanding the criteria for implantation even more [16]. Consequently the number of pediatric cochlear implantation surgeries has increased significantly since 1990 [17], which necessitates a clear understanding of potential threats to overall outcomes in this population [9].

In a recent systematic literature review on prognostic indicators in pediatric CI surgery, Black et al. [10] identified only four factors influencing pediatric CI outcomes consistently, namely age at implantation, presence of inner ear malformations, as well as occurrence of meningitis and Connexin 26 GJB2 gene-related deafness. Firstly, early implantation is indisputably considered as a strong positive predictor of expressive and receptive language skills, as confirmed by a plethora of published studies [9,18-25]. Secondly, inner ear malformations are strongly associated with pediatric CI outcomes in terms of speech perception and expressive language skills, with children who have more severe cochlear malformations (e.g. cochlear dysplasia and common cavity) performing worse than children with less severe malformations (e.g. incomplete partition or enlarged vestibular aqueduct) [9,26-29]. Thirdly, despite the fact that the central effects associated with meningitis may impact language learning potential [30], children with postmeningitic hearing loss do appear to benefit from CIs in terms of auditory receptive abilities, provided they receive an implant early [31]. However, for children with ossified cochleae as a result of meningitis, speech perception is frequently poorer than children with non-ossified cochleae [32]. Lastly, Connexin 26 GJB2-

related deafness in children with CIs appear to have lesser impacts predicting better speech intelligibility, speech discrimination and communication abilities when compared to implanted children with other etiologies of hearing loss [33-35].

Many other prognostic factors are described in literature, but only anecdotally, mostly due to small sample sizes [10]. Likewise, emerging trends in pediatric CI such as multiple disabilities, family influences and the impact of prematurity still require further evaluation as prognostic indicators [9]. The presence of additional disabilities negatively effects the language development of implanted children [1,23,35,36]. Yet outcomes after cochlear implantation for these children with associated disabilities, even if variable, show a positive evolution in speech perception, communication abilities, social engagement and quality of life [3,37]. Problematic family environments are significantly associated with poorer speech and language outcomes [9,38]. Then again, family factors such as a high socioeconomic level [5,35,39], sufficient parental involvement in the rehabilitation process [23,40,41] and higher levels of maternal education [42] are all related to improved language outcomes. Prematurity is considered as an anecdotal prognostic factor often described in pediatric CI literature, but has not been consistently proven [43]. The same holds for other likely etiological factors or risk indicators associated with permanent childhood hearing loss, such as neonatal intensive care unit (NICU) admittance, low birth weight and assisted ventilation [44].

In recent years there has been increasing interest in outcomes of bilateral cochlear implantation, since it has become the standard of care for children with severe to profound hearing loss in developed countries [14,45]. The benefits of bilateral implantation in children are well documented in terms of improved localization [46-48] and enhanced speech recognition in quiet [49,50] and in noise [46,51,52] when compared to listening with a unilateral CI. Also recently confirmed, children with bilateral CIs have significantly better language outcomes compared to children with unilateral CIs [45,53]. However, there is still a lack of evidence regarding the effect of bilateral cochlear implantation on broader outcomes such as literacy, academic skills and overall quality of life, particularly concerning long-term outcomes [45,48,54].

Prognostication is considered as a key component in pediatric cochlear implantation. Parents will only be able to set evidence-based and achievable expectations for their children if they are guided by professionals who are able to discern the factors that will exert an adverse effect on outcomes [3,43]. Given the paucity of proven prognostic factors in pediatric cochlear implantation [43], this current work aims to identify possible predictors of outcomes and to investigate the prognostic significance of these factors, in a large caseload of pediatric CI recipients in South Africa. Since the first multichannel cochlear implantation took place in South Africa in 1986, more than 1500 individuals has been implanted at nine respective CI programs [55,56]. Therefore, this study also provides a broad depiction of the current status of pediatric cochlear implantation in South Africa and reports on an extensive range of prognostic indicators identified in an unselected group of pediatric CI recipients.

MATERIALS AND METHODS

A retrospective study of 301 pediatric CI recipients was conducted. Institutional ethics committee approval was obtained before data collection commenced.

Study population

Five South African CI programs participated in this multicentre study, from which four programs are situated in the Gauteng Province (University of Pretoria Cochlear Implant Unit, Johannesburg Cochlear Implant Program, Chris Hani Baragwanath Academic Hospital Cochlear Implant Program, Steve Biko Academic Hospital Cochlear Implant Program) and one program in the Free State Province (Bloemfontein Cochlear Implant Program). Patient files of pediatric CI recipients at participating programs were reviewed retrospectively and cross-sectional outcome measures were added during an eight month data collection period. All children (≤18 years), implanted between 1996 and 2013 with a minimum of six months implant use at the time of data-collection and with data available on at least one outcome measure, were considered as eligible participants for this study. No case selection occurred and children from the complete range of educational and communication environments were included. The final sample consisted of 301 children, including eight (2.7%) children who were non-users of their CI devices (n=301). Of the total sample, 190 (63.1%) children were implanted unilaterally and 111 (36.9%) were implanted bilaterally at the time of data collection (n=301). All bilateral implants were performed sequentially, except for two children who were implanted simultaneously (2/111, 1.8%). The mean interval between first and second implant was 35 months (range: 1 - 156 months; 34.6 SD; n=107). Characteristics of the study population are presented in Table 1. Most children (94%) were implanted with Cochlear© devices and 18 children (6%) with Med-el© devices (n=301). With the exception of 13 children (5.3%), all children had a fully inserted electrode array in at least one cochlea (n=243). Nine children (9/301, 3%) had explant/re-implant procedures of their 1st/only implant, while 4 children (4/113, 3.5%) with bilateral implants were reimplanted in their 2nd ear. Of the children implanted unilaterally, most (81.8%, 108/132) used bimodal amplification. Less than a third of the children (29%, 77/265) made use of assistive listening devices. Almost all children had normal hearing parents (96.4%, 268/278).

Table 1: Characteristics of study population

Demographics	*% (n)*	Hearing loss and CI characteristics	*% (n)*
Gender		**Onset of hearing loss**	
Male	52.5 (158/301)	Congenital/ early onset	73.2 (188/257)
Female	47.5 (143/301)	Progressive	10.9 (28/257)
		Sudden	14.4 (37/257)
		Unknown	1.6 (4/257)
Ethnic category		**Age at diagnosis of hearing loss (months):**	
Caucasian	61.8 (186/301)	Congenital/ early onset (n=122)	
Black	24.3 (73/301)	Mean (SD)	16.1 (10.0)
Indian/ Asian	8.6 (26/301)	Range	1 - 60
Coloured	5.3 (16/301)	Post-natal (sudden/progressive) onset (n=51)	
		Mean (SD)	30.8 (31.2)
		Range	3 - 180
Home language		**Age at implantation (months):**	
Afrikaans	46.4 (129/278)	Congenital/ early onset (n=187)	
English	42.8 (119/278)	Mean (SD)	45.6 (32.5)
African language	4.0 (11/278)	Range	5 - 188
Other	6.8 (19/278)	Post-natal (sudden/progressive) onset (n=65)	
		Mean (SD)	64.9 (42.5)
		Range	9 – 193
Health sector		**Delay from diagnosis to implantation (months) in 1st ear** (n=188)	
Private	95.0 (286/301)	Mean (SD)	28.7 (28.5)
Public	5.0 (15/301)	Range	0.6 - 164.1
South African citizen		**Type of hearing loss**	
Yes	91.7 (276/301)	Sensory-neural	96.5 (275/285)
No	8.3 (25/301)	Auditory Neuropathy Spectrum Disorder	3.5 (10/285)

Description of variables

Regression modelling was performed to determine prognostic factors that will influence outcomes in terms of auditory performance (CAP scores), speech production (SIR scores), communication mode and educational placement.

Outcome variables

Both “auditory performance” and “speech production” were used as continuous outcome variables in this study. Auditory performance was rated by the Categories of Auditory Performance (CAP) [57] - a language- and age-independent hierarchical scale of auditory receptive abilities. The CAP has 8 categories, ranging from 0 (unaware of environmental sounds) to 7 (use of telephone with a familiar person). A revised version, referred to as the CAP_R [58] was used, in which a ninth category was added (use of telephone with an unfamiliar person). The Speech Intelligibility Rating (SIR) [59] was used for the assessment of speech production to classify children's speech production according to one of five hierarchical categories, ranging from Category 1 (connected speech is unintelligible) to Category 5 (connected speech is intelligible to all listeners). Validity, reliability and inter-tester reliability of both the CAP and SIR scales has been confirmed [60-62].

The research also included "communication mode" and "educational placement" as categorical outcome variables. Children's mode of communication included oral communication, South African Sign Language (SASL), total communication, and other alternative modes of manual communication (such as informal gestures or augmentative communication devices). Oral communication refers to the use of spoken language, with primary reliance on auditory cues for communication [63]. The children in this study, who were communicating orally, received auditory-oral or auditory-verbal style intervention. SASL is a system of manual communication using visual gestures and signs used by the Deaf community in South Africa, while total communication implies the combined use of oral speech, a formal sign language system, speech reading and audition for communication [64]. Educational placement of implanted children involved mainstream schooling (normal hearing educational setting), school for the Deaf (SASL mode of communication), school for the hard of hearing (oral mode of communication), special school (following either a mainstream or adapted special syllabus), home school, or no school if children did not attend school for some reason (e.g. placement challenges as a result of multiple disabilities) or were too young to attend school.

Explanatory variables

The collected retrospective data included demographical, CI and hearing loss data (Table 1), as well as family and risk factor data (Appendix A, Table A.1). From this retrospective dataset, 20 potential prognostic factors were identified and defined as categorical variables in two-way categories. These categorical predictors are presented in Appendix A, Table A.2 in terms of biographical and hearing loss factors (gender, ethnicity, age of diagnosis of hearing loss), CI factors (choice of ear for first implant, age at implant, delay from diagnosis to implant, bilateral implantation), family factors (family history of permanent childhood hearing loss, parental marital status, highest educational qualification of father, highest educational qualification of mother, employment status of mother) and risk factors (additional developmental conditions, prenatal risk factors, admittance to NICU, prematurity, natal risk factors, post-natal risk factors, meningitis, risk factors in general).

Data collection

All pediatric CI recipients who met the inclusion criteria were identified at each of the five participating CI programs. After data capturers were identified and trained for each participating program, the clinical files of all eligible children were reviewed retrospectively. An electronic database was developed for the capturing of the retrospective data (Table 1; Appendix A, Table A.1) amongst the participating programs. Cross-sectional outcome data in terms of auditory performance, speech production, communication mode and educational placement were added to the database at the time of data collection. CAP and SIR scores were allocated by experienced audiologists/ speech-language therapists involved in the rehabilitation of the children at the respective CI programs. These professionals also provided the outcome data on the communication mode and educational placement of the implanted children.

Statistical analysis

Simple descriptive statistics were utilized to define the study population in terms of demographical, CI and hearing loss characteristics (Table 1), as well as family and risk profiles (Appendix A, Table A.1). From these characteristics, 20 suspected prognostic factors were identified (Appendix A, Table A.2).

For age of hearing loss diagnosis and age at implantation, only the children with congenital/ early onset hearing loss were considered and categorized into either an early diagnosis/implantation (<36 months) or late diagnosis/ implantation (≥36 months) category. For bilateral implantation, only the children who had at least 6 months experience with their bilateral implant at the time of data collection were considered as bilateral implant users (78.4%, 87/111).

Children were categorised into performance groups for auditory receptive abilities (CAP scores). Thus, a low score was defined as CAP category 0-4 and a high score as CAP category 5-8. Children's speech intelligibility was also categorised into performance groups according to SIR scores, indicating whether a child's connected speech is intelligible or not to a listener who concentrates and lip-reads. SIR category 1-2 was defined as a low score and SIR category 3-5 as a high score. Furthermore, children's hearing age with a CI (i.e. length of device use from the day of initial stimulation of 1st implant) at the time of the scoring of the CAP and SIR was defined in months and is hence referred to as hearing age at CAP/SIR. Children's mode of communication was described as being either oral or non-oral, with non-oral referring to children utilising SASL, total communication or any alternative mode of manual communication. For educational placement children were divided in 2 groups: mainstream education and non-mainstream education.

For the purpose of variable selection for regression modelling, bivariate data analyses were undertaken to determine the existence of a possible association between a potential predictor

(Appendix A, Table A.2) and a categorical outcome variable, in two categories using the Pearson Chi-Square test. The p-values of the Pearson Chi-Square test on these 2x2 tables appear in Appendix A, Table A.3.

For the main prediction analysis, two types of multiple regression were used: For continuous outcome variables (auditory performance and speech production), linear regression models in the form of multiway analysis of variance were constructed to investigate the influence of categorical predictors on the mean auditory performance (CAP scores) as well as the mean speech production (SIR scores).

Loglinear models were constructed for categorical outcome variables (communication mode and educational placement) to model the log odds of children's mode of communication to be non-oral and the log odds of educational placement to be non-mainstream in terms of the categorical predictors. An index for each category of a predictor can be calculated as the exponent of the regression coefficient of that category, obtained from the loglinear model. The odds for any combination of categories of predictors can be found by multiplication of the overall mean odds (the exponent of the intercept term in the log odds model) with the indices of the specified categories. Based on these odds the probability for non-oral communication or non-mainstream educational placement was estimated by dividing the odds outcome by the factor (1 + odds).

Throughout the process two factors were additionally forced into the models. For the linear regression models, the hearing age at CAP/SIR factor (being either ≤36 months or ≥37 months) was added. The onset of hearing loss (being either congenital/ early onset or post-natal) was forced into both the linear regression models, as well as in the loglinear models to ensure that a clear distinction was made statistically between children with congenital/ early onset (pre-lingual) hearing loss and children with post-natal (sudden or progressive) onset hearing loss.

RESULTS

General clinical and outcome profile

The demographical and CI profile of the study sample are presented in Table 1. CAP and SIR scores were obtained for 240 children at the time of the study (240/301 or 79.7%). Overall, most children (164/240 or 68.3%) achieved high CAP scores (category 5-8), while 76 children (76/240 or 31.7%) achieved low CAP scores (category 0-4). For the total sample, high SIR scores (category 3-5) were attained by 171 children (171/240 or 71.2%), with 69 children (69/240 or 28.8%) attaining low SIR scores (category 1-2). Average hearing age at CAP/SIR for this study sample was 67.4 months (range: 6 - 88 months; 43.6 SD; n=236).

This hearing age at CAP/SIR was divided into two groups: children with a hearing age with CI of ≤36 months (73/236 or 30.9%) and children with a hearing age with CI ≥37 months (163/236 or 69.1%). Taking this hearing age with CI into account, for children wearing their implants ≥37 months, high CAP scores (128/163 or 78%) and high SIR scores (128/163 or 78%) were achieved for even more children.

Data on children's current mode of communication were obtained for 96.3% (290/301) of the total sample. Most children (74.5%, 216/290) were oral communicators, while 13.1% (38/290) utilized TC and 6.2% (18/290) used SASL. The remaining 6.2% (18/290) were using other alternative modes of manual communication. All children who were not oral communicators were grouped together as non-oral communicators (25.5%, 74/290).

For almost the entire sample (99%, 298/301), data were available on the educational placement of children. Just more than half of the children were in mainstream schools

(52.3%, 156/298), while 15.1% (45/298) were in schools for the deaf (SASL mode of communication) and 7.4% (22/298) were in schools for the hard-of-hearing (oral mode of communication). A significant proportion of children (17.4%, 52/298) attended special schools where in half of the cases (50%, 26/52) a mainstream syllabus was followed and the other half of the cases (50%, 26/52) an adapted special syllabus was followed. Fourteen children (4.7%, 14/298) did not go to school, and another 9 children (3%, 9/298) were home-schooled. All children not attending mainstream schools were grouped together as being placed in non-mainstream education (47.7%, 142/298).

Linear regression analysis: Auditory performance (CAP scores) and Speech production (SIR scores)

Only the predictor variables that appeared to be associated with the outcome variables in the bivariate analysis were included in the regression models (Appendix A, Table A.3). For the linear regression models, all associated predictor variables with a significance level of $p < 0.1$, as well as the two forced factors, were randomly fed into the model. During the model building process, the best predictors of the two continuous outcome variables (CAP and SIR scores) were identified.

The two resulting linear regression models showing the best predictors of outcomes in terms of auditory performance (model 1) and speech production (model 2) are presented in Table 2.

Both linear regression models (model 1 and 2) were highly significant ($p = < 0.0001$) and present with determination coefficients (R^2) of 28% and 26% respectively, giving an indication that less than 30% of the variation in the outcomes observed in the data was accounted for by the models. Accordingly, 72% of the variation in auditory performance outcomes and 74% of the variation in speech production outcomes was not explained by the selected factors in the linear regression models.

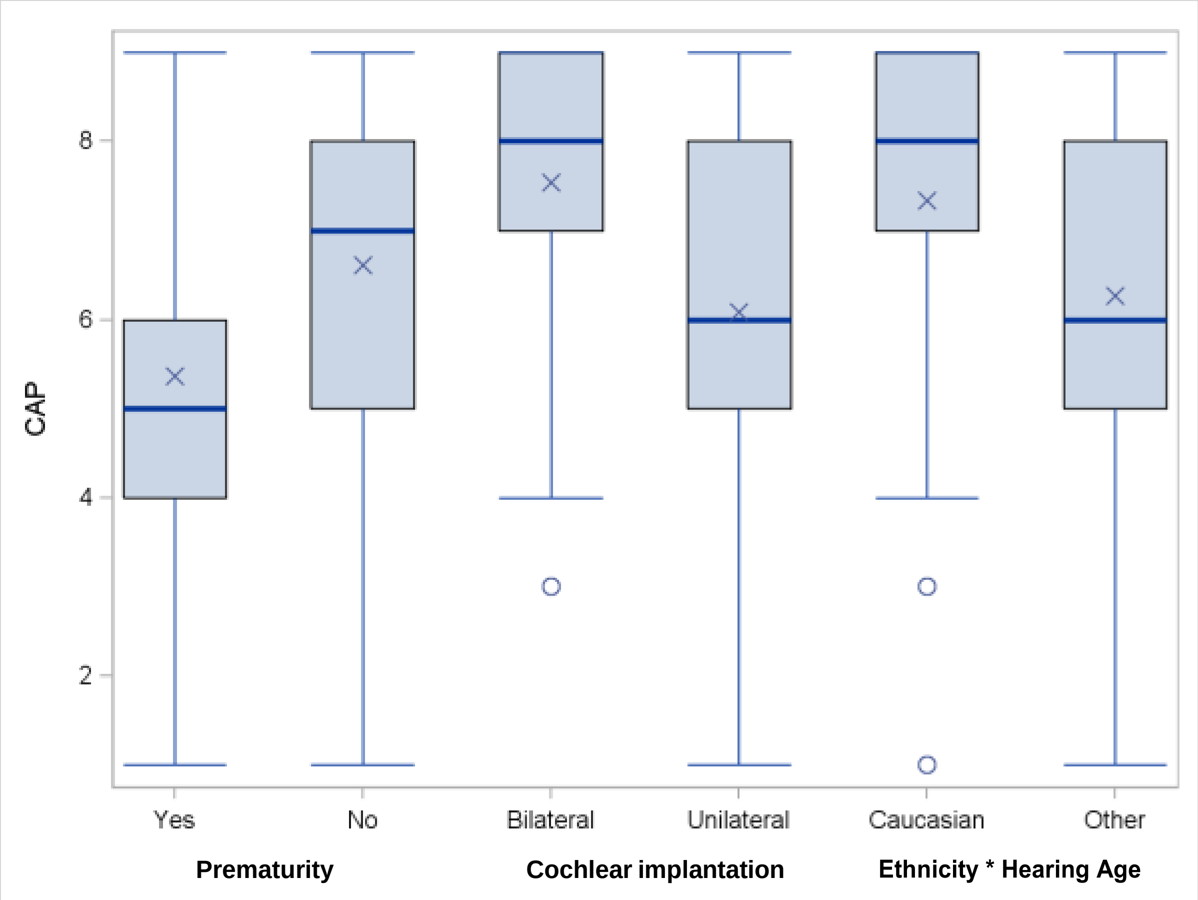

Figure 1: Bilateral implantation, prematurity and ethnicity*hearing age as predictors of auditory performance (CAP scores) n=193 The box plots represent the smallest observation, lower quartile, median (bold line), mean (x), upper quartile, largest observation, and outliers (>1.5 times interquartile range) (○). * interaction

The boxplots in Figure 1 illustrate bilateral implantation, prematurity and ethnicity as predicting factors for the auditory performance outcome (regression model 1). It shows that children implanted unilaterally have significantly lower average CAP scores (minus 2 units) compared to children who are implanted bilaterally ($p = 0.0003$). The same results are observed for the prematurity factor where on average children born prematurely (≤34 weeks gestation) also score 2 CAP units lower ($p = 0.0075$). A third factor that was identified in model 1 was the interaction between the hearing age at CAP/SIR and ethnic category for the group of children with a hearing age with CI ≥37 months , showing a lower average CAP score (minus 2 units) for children with “other” ethnicities when compared to Caucasian children ($p < 0.0001$).

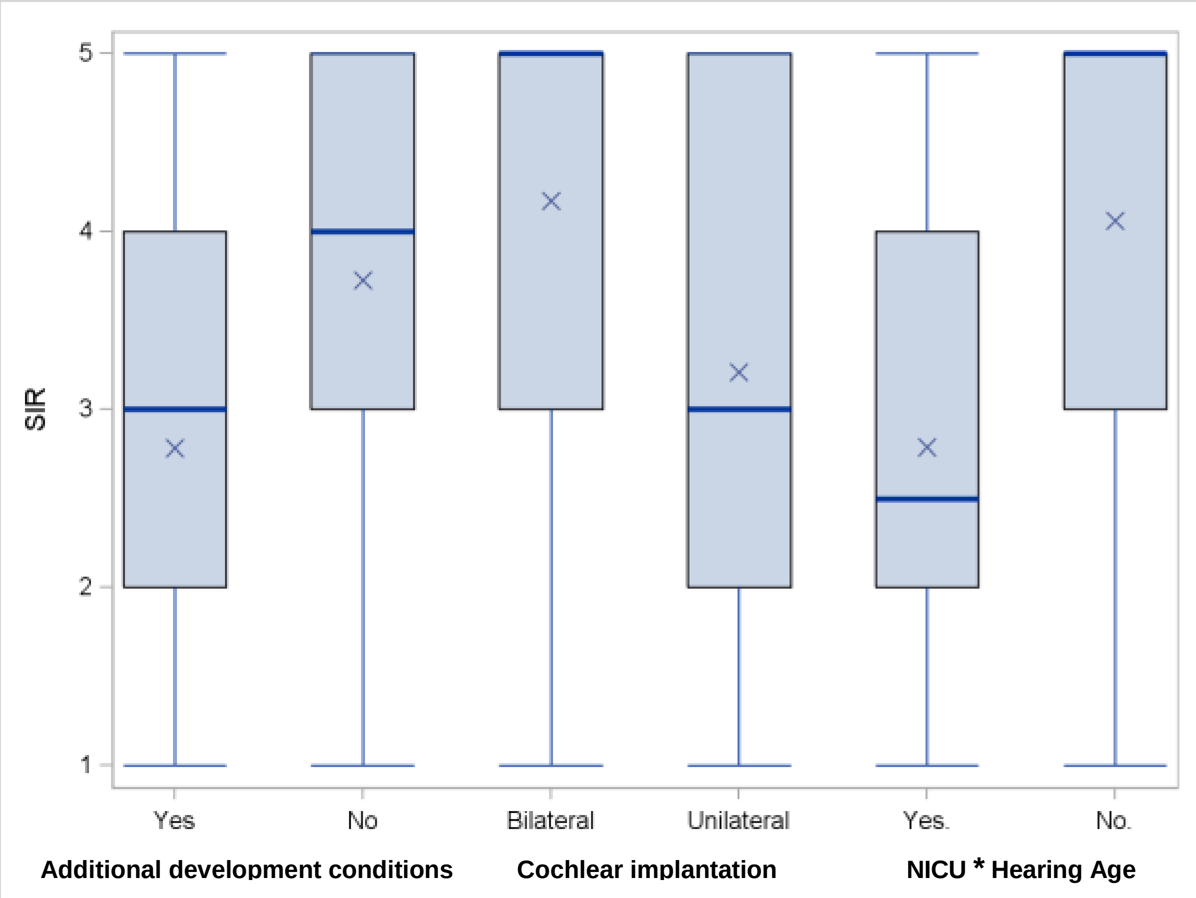

Figure 2: Bilateral implantation, presence of additional developmental conditions and admittance to NICU*hearing age as predictors of speech production (SIR scores) n=213 The box plots represent the smallest observation, lower quartile, median (bold line), mean (x), upper quartile, largest observation. *interaction

The identified relevant predictors for speech productions (regression model 2) (Table 2), illustrated by the boxplots in Figure 2, show that children with bilateral implants are expected to have an average SIR score of 5, compared to an average score of 3 for children with

Table 2: Linear regression analysis results

Model	Outcome variables	Explanatory variables	DF*	Sum of Squares	F Value	Pr > F** (P value)	R^2
1	Auditory performance (CAP)	Bilateral implantation Prematurity Hearing age at CAP/SIR & ethnic category interaction	5	202.221	14.39	<0.0001	0.28
2	Speech production (SIR)	Bilateral implantation Additional developmental conditions Hearing age at CAP/SIR & NICU interaction	5	113.083	14.81	<0.0001	0.26

**DF: Degrees of freedom; **Pr>F: p-value of the F-test (with F-test testing the significance of the model)*

unilateral implants (p = 0.0038). Secondly, children with additional developmental conditions are expected to have a lower average SIR score of 3 when compared to children without any additional developmental conditions (average SIR score of 4) (p = 0.0002). Lastly, the hearing age at CAP/SIR and NICU admittance interaction shows that for children with a hearing age with CI ≥37 months, those who were admitted to the NICU have lower SIR scores (minus 2.5 units) than those who did not have a history of NICU admittance (p = < 0.0001).

Log Linear analysis: non-oral mode of communication and non-mainstream educational setting

Log linear modelling is used to determine the influence of a set of categorical explanatory factors on a categorical outcome. The cell frequencies within a combination of categories of predictors must be large enough. Therefore only a limited number of predictor variables in the categorical modelling can be considered for the sample sizes of n=139 and n=151 for the

Table 3: Log linear analysis results (maximum likelihood estimates)

Model	Parameter (combined factors)	Categories	Estimate from log-linear model	Index*
3	Overall mean odds		-1.2025	0.30
	Bilateral/ unilateral CI and natal risk factors	Bilateral CI	-0.3465	0.71
		Unilateral CI	0.3465	1.41
	Onset of HL, NICU admittance, risk factors (general) and additional developmental conditions	Congenital onset HL, NICU	0.5411	1.72
		Congenital onset HL, no NICU	-0.4027	0.67
		Post-natal onset HL	-0.1384	0.87
4	Overall mean odds		-0.0772	0.93
	Onset of HL, ethnicity, delay between diagnosis and implantation, bilateral/ unilateral implantation	White, congenital onset HL, <12 months, bilateral CI	-1.5484	0.21
		Post-natal onset HL	0.2921	1.34
		Non-white, congenital onset HL	0.6383	1.89
		White, congenital onset HL, >12 months, bilateral CI	-0.4461	0.64
		White, congenital onset HL, <12 months, unilateral CI	0.4095	1.51
		White, congenital onset HL, ≥12 months, unilateral CI	0.6546	1.92
	Additional developmental conditions and NICU admittance	Additional developmental condition, NICU	0.9102	2.49
		Additional developmental condition, no NICU	-0.1756	0.84
		No additional developmental condition	-0.7346	0.48

**Index is the exponent of the estimate. An index lower than 1 indicates an odds (to be non-oral/ to be placed in non-mainstream education) that is lower than the average odds of 1 (low-risk category), while an index higher than 1 indicates an odds higher than average (high-risk category).*

two odds models. In this study, only the predictor variables that were significantly associated with the categorical outcome variables were included ($p < 0.05$) (Appendix A, Table A.3). Log linear model analysis was used to model the log of the odds to be a non-oral CI user (model 3) and the log of the odds to be a CI user in a non-mainstream educational setting (model 4). The statistical outcome of the log linear analysis is summarized in Table 3 in terms of indices. By using the indices, the odds to be non-oral and non-mainstream were calculated by multiplying the overall main effect (index of the intercept) with the indices of any combination of categories of predictors. The odds and percentage chance for models 3 and 4 are presented in Figure 3 and Figure 4 respectively.

For unilaterally implanted children with a congenital/ early onset hearing loss and a history of admittance to the NICU, there was a 42% probability to be a non-oral communicator, with the probability being almost half times less (22%) if children were not admitted to NICU. In contrast, should bilaterally implanted children with a congenital/ early onset hearing loss have a history of NICU admittance, the chance to be a non-oral communicator was less (27%). For children with a post-natal onset of hearing loss, those implanted unilaterally had a

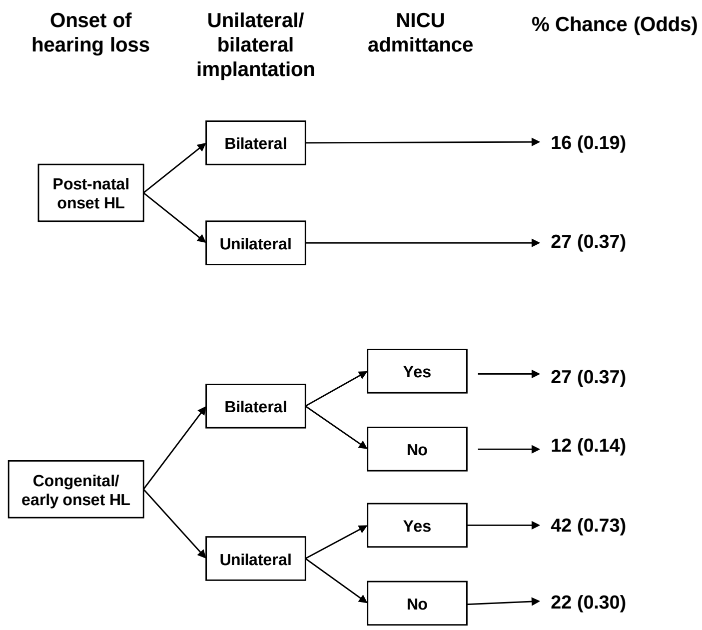

Figure 3: Associated probability predisposing non-oral mode of communication (model 3) n=139

higher probability (27%) to be non-oral communicators, in contrast to children with bilateral implants (16%).

For model 4 (Figure 4), a very high probability for non-mainstream educational placement (82%) was indicated for Caucasian, unilaterally implanted children with a congenital/ early onset hearing loss, with a delay of more than one year between diagnoses and implantation, who presented with a history of NICU admittance and at least one additional developmental condition. Similarly a high chance for non-mainstream educational placement (81%) was indicated for all Caucasian children with a congenital/ early onset hearing loss, who were admitted to NICU and presented with at least one additional developmental condition. For Caucasian children with a congenital/ early onset hearing loss, with a delay of less than one year between diagnosis and implantation, who presented with a history of NICU admittance

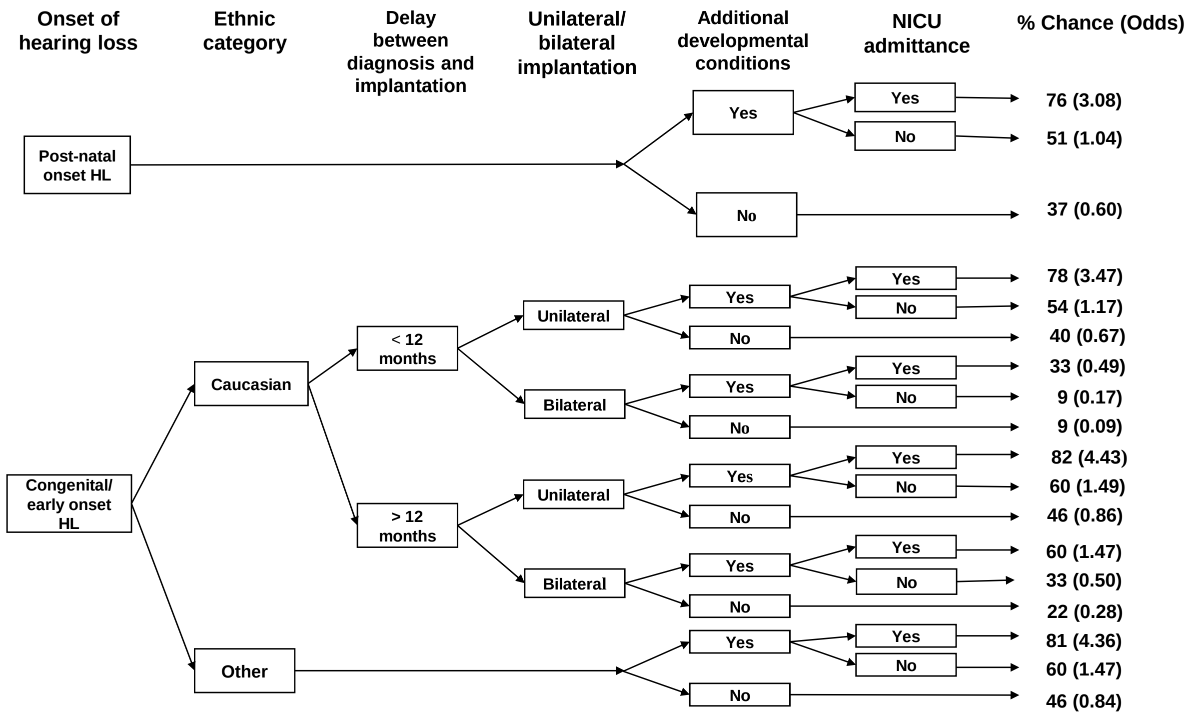

Figure 4: Associated probability predisposing non-mainstream educational setting (model 4) n=151

and the presence of at least one additional developmental condition, the difference in probability for non-mainstream education was significant between those implanted unilaterally (78%) and bilaterally (33%). Even if these children were not admitted to NICU, the difference in probability for unilateral implanted children (54%) and bilateral implanted children (9%) was still substantial. For children with a post-natal hearing loss, the probability to be placed in a non-mainstream educational setting were twice as high (76%) for those who were admitted to NICU and presented with at least 1 additional developmental condition, in contrast to those with no history of NICU admittance or additional developmental conditions (37%).

DISCUSSION

A broad range of predictors for pediatric cochlear implantation outcomes in South Africa were identified. The vast majority of children (78%) implanted for more than three years achieved high CAP scores for auditory performance, and high SIR scores for speech production, suggesting they can understand spoken conversation with a familiar person and have connected speech that is intelligible for at least an experienced listener. Almost 75% of children in this study were oral communicators and more than half (56%) of children who used their implants for longer than three years were placed in mainstream educational settings. However, mainstream education as a measure of success in cochlear implantation should be used with caution, since the emphasis should rather be on the appropriateness of educational placement to each child’s specific needs. Current educational policy in South Africa has the long-term goal to develop an inclusive education system, also for children with severe-profound hearing loss, which will address barriers to learning such as socio-economic barriers, language and communication and inflexible curriculums [65]. Nonetheless, various persistent challenges, such as disparities in resourcing inclusive education across provinces and limited access to specialist support in public ordinary schools, currently impede the progress that is being made towards an inclusive education system [66].

Predictive factors for pediatric CI outcomes in this study were bilateral implantation, admittance to the NICU, prematurity, additional developmental conditions, ethnicity and the delay between diagnosis and implantation. Clear distinction was made in the statistical analysis of data between children with congenital/ early onset (pre-lingual) hearing loss and children with post-natal (sudden/progressive) onset hearing loss. This distinction is important when a heterogeneous caseload is considered, since it is expected that children with post-lingual onset hearing loss will mostly perform well after cochlear implantation as a result of more mature auditory pathways and early foundations for speech and language [24,67-69].

Bilateral implantation was a strong predictor for better auditory performance and speech production scores, and was associated with a lower probability for a non-oral mode of communication and a non-mainstream educational setting. Scherf at al.[70] also used the CAP as outcome measure in a group of 35 children with bilateral CIs, showing that after three years of bilateral implant use, higher CAP scores were obtained for significantly more children than before their second implant. Although the positive effect of bilateral implantation on spoken language development has recently been demonstrated [45,53], the influence of bilateral implantation on speech production remains to be demonstrated [48]. The strong association between bilateral implantation and the increased ability of children to produce intelligible speech in the current study, could be the direct result of the improved auditory input from a second CI, since speech perception and speech production skills are highly correlated with each other [71].

Evidence for bilateral cochlear implantation affecting educational outcomes is lacking [48,54]. The current study provides preliminary evidence that children with bilateral CIs, in at least a subgroup of Caucasian children with congenital/ early-onset hearing loss, have a lower associated probability for non-mainstream education, compared to those children with unilateral CIs. Also, the probability for non-oral communication was greater for unilaterally implanted children, irrespective of onset of hearing loss. However, it is imperative that the association between bilateral implantation and better outcomes in this study should be viewed against the background that implanted children in South Africa do not have equal opportunity to access a second CI. With 95% of this sample representing the private health care sector, family financial resources remains to be a significant determining factor for bilateral implantation in South Africa, implying that a bilateral CI will only be accessible to children who’s caregivers have adequate finances. As a result, it is more likely that unilaterally implanted children from affluent families, who communicates orally and already functions in mainstream educational environments, would be considered for bilateral implantation.

NICU admittance was associated with poorer speech production scores and a higher probability for non-oral communication and non-mainstream school placement, while prematurity was associated with lower auditory performance scores. To the authors’ knowledge, NICU admittance, together with prematurity, has not yet been demonstrated as prognostic factors in pediatric CI. Robertson et al. [72] reported that in a group of 1279 children admitted to NICU because of extreme prematurity, 3.1% presented with permanent childhood hearing loss, of whom 73% had more than one other major developmental disability. The outcomes of NICU graduates with permanent childhood hearing loss who eventually receive CIs are likely to be affected by the increased incidence of additional developmental conditions. NICU admittance and prematurity are therefore risk factors for poorer CI outcomes, likely related to the comorbidities that accompany these perinatal developmental challenges.

The presence of one or more additional developmental condition was found to be strongly predictive of poorer speech production scores, and was associated with a higher probability for non-mainstream education. It is estimated that 30 to 40% of children with profound deafness have additional disabilities [1,73,74], which is slightly higher than the 24% prevalence in this study population. In an outcome study of 119 three year old children with hearing loss and additional disabilities, of whom 29% were CI users, speech intelligibility ratings revealed relatively poor outcomes, with a mean rating of 4.2 on a scale from 1 to 6, where 1 represents 100% intelligibility [42]. Not only does the presence of additional developmental conditions negatively impact language development in pediatric CI recipients [1,23,36], but it may also prevent them from reaching their full potential cognitively, socially and educationally [42].

Ethnicity was found to be a predictor of auditory performance and educational placement, with ethnicities other than Caucasian achieving lower auditory performance scores and having a higher associated probability for non-mainstream education. Not only in South Africa, but all over the world many areas of healthcare are replete with evidence of socioeconomic status and ethnicity related disparities, with pediatric cochlear implantation being no exception [75-77]. The current study population consisted of predominantly Caucasian (62%) children from the private health care system (95%), speaking either Afrikaans (46%) or English (43%). This sample could be considered as representative of pediatric CI recipients in South Africa and reflects the current health care inequalities for advanced interventions such as CIs. However, this sample does not represent the larger South African population, with 79.8% of the population being of African ethnicity, 74.9% speaking an African first language and 75% relying on public health care for health services [78-80].

Ethnicity as a prognostic indicator in this study is most likely a proxy for social and health inequality.

A delay between diagnosis and implantation of more than 12 months was strongly associated with a higher probability of non-mainstream school placement in at least a subgroup of Caucasian children with congenital/ early-onset hearing loss. With the recent emphasis on early access to sound through early implantation, late implantation is now defined as more than 12 months after diagnosis of hearing loss [81]. Early implantation during periods of optimal neural plasticity maximizes early auditory experience and leads to more age-appropriate speech and language skills [67,82,83], which may also increase the likelihood for mainstream education from earlier ages onwards. Likely contributing factors for this delay between diagnosis and implantation include funding constraints, lack of prompt referral to specialized CI services, parental barriers such as delayed/missed appointments, complex medical conditions, family indecision and geographical location [81,84,85].

Within this relatively large dataset, various factors were identified to be predictive of outcomes, however the determination coefficients of the linear regression models were less than 30% and do not account for two-thirds of the remaining variation in auditory performance and speech production outcomes. This implies that both outcomes are in reality determined by many more single or interacting factors not included in the different models used herein.

Unlike many other studies, age of implantation was not confirmed as a prognostic factor for this dataset. A possible explanation for this could be the fact that this study examined outcomes at a single point in time, rather than longitudinally, as also reported by other studies [39,86]. It might be that some of the advantages for early implantation are more evident at younger ages, becoming less apparent when children become older.

CONCLUSION

Bilateral implantation was a strong predictor of better auditory performance and speech production outcomes, and was strongly related to an oral communication mode and mainstream education. However, since family financial resources remains a decisive factor for bilateral implantation in South Africa, not all implanted children in this dataset had the opportunity to access a second CI. NICU admittance/ prematurity were predictive of poorer auditory performance and production outcomes, together with a higher probability for non-oral communication and non-mainstream education. The presence of one or more additional developmental conditions was associated with poorer outcomes in terms of speech production and educational placement, with a delay between diagnosis and implantation of more than 12 months also being associated with non-mainstream schooling. Ethnicity was validated to be predictive of auditory performance outcomes and educational setting, with ethnicities other than Caucasian having lower auditory performance outcomes and a lower probability for mainstream education. The challenges associated with multicentre retrospective data collection in this study, such as unsystematic, missing and inconsistently recorded data, highlighted the need for the implementation of a shared data recording methodology across programs in South Africa. Only within such an agreed standardized framework, with universal standardized outcome measures, can compatible patient and outcome data be captured and utilized for the purpose of collaborative multicentre research [9]. Irrespective, findings from this study provide valuable guidance and understanding into the causes of variation of pediatric CI outcomes, and also contribute to evidence-based pediatric CI services that promote optimal outcomes.

ACKNOWLEDGEMENTS

The authors gratefully acknowledge the support of involved professionals from the five participating cochlear implant programs (University of Pretoria Cochlear Implant Unit, Johannesburg Cochlear Implant Program, Chris Hani Baragwanath Academic Hospital Cochlear Implant Program, Bloemfontein Cochlear Implant Program, Steve Biko Academic Hospital Cochlear Implant Program) who assisted with data collection for this study. The first author would also like to thank Aaqilah Bhamjee, Sabeehah Dawood and Mahmooda Noor Mohamed for their assistance with data capturing, and Andries Masenge for his valuable support with the data analysis.

REFERENCES

[1] C.S. Birman, E.J. Elliott, W.P.R. Gibson, Pediatric cochlear implants: additional disabilities prevalence, risk factors, and effect on language outcomes., Otol. Neurotol. 33 (2012) 1347–52. doi:10.1097/MAO.0b013e31826939cc.

[2] S. Amirsalari, J. Yousefi, S. Radfar, A. Saburi, S.A. Tavallaie, M.J. Hosseini, et al., Cochlear implant outcomes in children with motor developmental delay, Int. J. Pediatr. Otorhinolaryngol. 76 (2012) 100–103. doi:10.1016/j.ijporl.2011.10.011.

[3] A. Kral, G.M. O’Donoghue, Profound deafness in childhood., N. Engl. J. Med. 364 (2011) 1438–1450. doi:10.1056/NEJMra0911225.

[4] O.B. Wie, Language development in children after receiving bilateral cochlear implants between 5 and 18 months, Int. J. Pediatr. Otorhinolaryngol. 74 (2010) 1258–1266. doi:10.1016/j.ijporl.2010.07.026.

[5] J.K. Niparko, E.A. Tobey, D.J. Thal, L.S. Eisenberg, N.-Y. Wang, A.L. Quittner, et al., Spoken language development in children following cochlear implantation., JAMA. 303 (2010) 1498–1506. doi:10.1001/jama.2010.451.

[6] T.Y. Ching, H. Dillon, J. Day, K. Crowe, L. Close, K. Chisholm, et al., Early language outcomes of children with cochlear implants: interim findings of the NAL study on longitudinal outcomes of children with hearing impairment, Cochlear Implant. Int. 10 Suppl 1 (2009) 28–32. doi:10.1002/cii.382.

[7] K. Hawker, J. Ramirez-Inscoe, D.V.M. Bishop, T. Twomey, G.M. O’Donoghue, D.R. Moore, Disproportionate language impairment in children using cochlear implants., Ear Hear. 29 (2008) 467–471. doi:10.1097/AUD.0b013e318167b857.

[8] T. Ching, K. Crowe, V. Martin, Language development and everyday functioning of

children with hearing loss assessed at 3 years of age, J. Speech-Language Pathol. 12 (2010) 124–131. http://informahealthcare.com/doi/abs/10.3109/17549500903577022.

[9] J. Black, L. Hickson, B. Black, A. Khan, Paediatric cochlear implantation: Adverse prognostic factors and trends from a review of 174 cases, Cochlear Implants Int. 15 (2014) 62–77. doi:10.1179/1754762813Y.0000000045; 10.1179/1754762813Y.0000000045.

[10] J. Black, L. Hickson, B. Black, C. Perry, Prognostic indicators in paediatric cochlear implant surgery: a systematic literature review., Cochlear Implants Int. 12 (2011) 67–93. doi:10.1179/146701010X486417.

[11] L. Graham O'Brien, C. Valim, M. Neault, B. Kammerer, T. Clark, J. Johnston, et al., Prognosis tool based on a modified children's implant profile for use in pediatric cochlear implant candidacy evaluation, Ann. Otol. Rhinol. Laryngol. 121 (2012) 73–84.

[12] S.J. Dettman, W.A. D'Costa, R.C. Dowell, E.J. Winton, K.L. Hill, S.S. Williams, Cochlear implants for children with significant residual hearing., Arch. Otolaryngol. Head. Neck Surg. 130 (2004) 612–618. doi:10.1001/archotol.130.5.612.

[13] E. Fitzpatrick, J. Olds, A. Durieux-Smith, R. McCrae, D. Schramm, I. Gaboury, Pediatric cochlear implantation: how much hearing is too much?, Int. J. Audiol. 48 (2009) 91–97. doi:10.1080/14992020802516541.

[14] M. Tait, T.P. Nikolopoulos, L. De Raeve, S. Johnson, G. Datta, E. Karltorp, et al., Bilateral versus unilateral cochlear implantation in young children, Int. J. Pediatr. Otorhinolaryngol. 74 (2010) 206–211. doi:10.1016/j.ijporl.2009.11.015.

[15] M. Sparreboom, A.R. Leeuw, A.F.M. Snik, E.A.M. Mylanus, Sequential bilateral cochlear implantation in children: Parents' Perspective and device use, Int. J. Pediatr. Otorhinolaryngol. 76 (2012) 339–344. doi:10.1016/j.ijporl.2011.12.004.

[16] H.F.B. Teagle, Cochlear Implantation for Children: Opening Doors to Opportunity, J. Child Neurol. 27 (2012) 824–826. doi:10.1177/0883073812442590.

[17] S. Özdemir, Ü. Tuncer, Ö. Tarkan, M. Kiroĝlu, F. Çetik, F. Akar, Factors contributing to limited or non-use in the cochlear implant systems in children: 11 years experience, Int. J. Pediatr. Otorhinolaryngol. 77 (2013) 407–409. doi:10.1016/j.ijporl.2012.11.041.

[18] P.J. Govaerts, C. De Beukelaer, K. Daemers, G. De Ceulaer, M. Yperman, T. Somers, et al., Outcome of cochlear implantation at different ages from 0 to 6 years., Otol. Neurotol. 23 (2002) 885–890.

[19] M.G. Habib, S.B. Waltzman, B. Tajudeen, M.A. Svirsky, Speech production intelligibility of early implanted pediatric cochlear implant users, Int. J. Pediatr. Otorhinolaryngol. 74 (2010) 855–859. doi:10.1016/j.ijporl.2010.04.009.

[20] M. Manrique, F.J. Cervera-Paz, A. Huarte, M. Molina, Advantages of cochlear implantation in prelingual deaf children before 2 years of age when compared with later implantation., Laryngoscope. 114 (2004) 1462–1469. doi:10.1097/00005537-200408000-00027.

[21] T.A. Zwolan, C.M. Ashbaugh, A. Alarfaj, P.R. Kileny, H.A. Arts, H.K. El-Kashlan, et al., Pediatric cochlear implant patient performance as a function of age at implantation., Otol. Neurotol. 25 (2004) 112–120. doi:10.1097/00129492-200403000-

00006.

[22] B. May-Mederake, Early intervention and assessment of speech and language development in young children with cochlear implants, Int. J. Pediatr. Otorhinolaryngol. 76 (2012) 939–946. doi:10.1016/j.ijporl.2012.02.051.

[23] T. Boons, J.P.L. Brokx, I. Dhooge, J.H.M. Frijns, L. Peeraer, A. Vermeulen, et al., Predictors of Spoken Language Development Following Pediatric Cochlear Implantation, Ear Hear. 33 (2012) 617–639. doi:10.1097/AUD.0b013e3182503e47.

[24] J.G. Nicholas, A.E. Geers, Will they catch up? The role of age at cochlear implantation in the spoken language development of children with severe to profound hearing loss., J. Speech. Lang. Hear. Res. 50 (2007) 1048–1062. doi:10.1044/1092-4388(2007/073).

[25] M.A. Svirsky, S. Teoh, H. Neuburger, Development of language and speech perception in congenitally, profoundly deaf children as a function of age at cochlear implantation., Audiol. Neurootol. 9 (2004) 224–33. doi:10.1159/000078392.

[26] S.J. Broomfield, I.A. Bruce, L. Henderson, R.T. Ramsden, K.M.J. Green, Cochlear implantation and congenital inner ear anomalies., Cochlear Implants Int. 11 Suppl 1 (2010) 166–168. doi:10.1179/146701010X12671177818542.

[27] D.J. Eisenman, C. Ashbaugh, T.A. Zwolan, H.A. Arts, S.A. Telian, Implantation of the malformed cochlea., Otol. Neurotol. 22 (2001) 834–841. doi:10.1097/00129492-200111000-00020.

[28] L. Kim, S. Jeong, M. Huh, Y. Park, Cochlear Implantation in Children With Inner Ear Malformations, Ann. Otol. Rhinol. Laryngol. 115 (2006) 205–214.

[29] D. Rachovitsas, G. Psillas, V. Chatzigiannakidou, S. Triaridis, J. Constantinidis, V. Vital, Speech perception and production in children with inner ear malformations after cochlear implantation, Int. J. Pediatr. Otorhinolaryngol. 76 (2012) 1370–1374. doi:10.1016/j.ijporl.2012.06.009.

[30] H.W. Francis, M.B. Pulsifer, J. Chinnici, R. Nutt, H.S. Venick, J.D. Yeagle, et al., Effects of central nervous system residua on cochlear implant results in children deafened by meningitis., Arch. Otolaryngol. Head. Neck Surg. 130 (2004) 604–611. doi:10.1001/archotol.130.5.604.

[31] T.P. Nikolopoulos, S.M. Archbold, G.M. O'Donoghue, Does cause of deafness influence outcome after cochlear implantation in children?, Pediatrics. 118 (2006) 1350–1356. doi:10.1542/peds.2006-0502.

[32] H.K. El-Kashlan, C. Ashbaugh, T. Zwolan, S.A. Telian, Cochlear implantation in prelingually deaf children with ossified cochleae., Otol. Neurotol. 24 (2003) 596–600. doi:10.1097/00129492-200307000-00011.

[33] A.R. Sinnathuray, J.G. Toner, J. Clarke-Lyttle, A. Geddis, C.C. Patterson, A.E. Hughes, Connexin 26 (GJB2) gene-related deafness and speech intelligibility after cochlear implantation., Otol. Neurotol. 25 (2004) 935–942. doi:10.1097/00129492-200411000-00013.

[34] A.R. Sinnathuray, J.G. Toner, A. Geddis, J. Clarke-Lyttle, C.C. Patterson, A.E. Hughes, Auditory perception and speech discrimination after cochlear implantation in patients with connexin 26 (GJB2) gene-related deafness., Otol. Neurotol. 25 (2004) 930–934. doi:10.1097/00129492-200411000-00012.

[35] J.M. Gérard, N. Deggouj, C. Hupin, A.L. Buisson, V. Monteyne, C. Lavis, et al., Evolution of communication abilities after cochlear implantation in prelingually deaf children, Int. J. Pediatr. Otorhinolaryngol. 74 (2010) 642–648. doi:10.1016/j.ijporl.2010.03.010.

[36] K. Rajput, T. Brown, D.E. Bamiou, Aetiology of hearing loss and other related factors versus language outcome after cochlear implantation in children, Int. J. Pediatr. Otorhinolaryngol. 67 (2003) 497–504. doi:10.1016/S0165-5876(03)00006-5.

[37] S. Berrettini, F. Forli, E. Genovese, R. Santarelli, E. Arslan, A.M. Chilosi, et al., Cochlear implantation in deaf children with associated disabilities: challenges and outcomes., Int. J. Audiol. 47 (2008) 199–208. doi:10.1590/S1516-80342008000200017.

[38] R.F. Holt, J. Beer, W.G. Kronenberger, D.B. Pisoni, K. Lalonde, Contribution of family environment to pediatric cochlear implant users' speech and language outcomes: Some preliminary findings, J. Speech, Lang. Hear. Res. 55 (2012) 848–864. doi:10.1126/science.1151148.\n10.1076/chin.6.3.235.3152\n10.1111/1467-8624.00612\n10.1037/0012-1649.43.6.1447\nhttp://dx.doi.org/10.1044/1092-4388(2011/11-0143.

[39] A.E. Geers, J.G. Nicholas, A.L. Sedey, Language skills of children with early cochlear implantation., Ear Hear. 24 (2003) 46S–58S. doi:10.1097/01.AUD.0000051689.57380.1B.

[40] J.Z. Sarant, C.M. Holt, R.C. Dowell, F.W. Rickards, P.J. Blamey, Spoken language development in oral preschool children with permanent childhood deafness, J. Deaf Stud. Deaf Educ. 14 (2009) 205–217. doi:10.1093/deafed/enn034.

[41] P.E. Spencer, Individual differences in language performance after cochlear implantation at one to three years of age: child, family, and linguistic factors., J. Deaf Stud. Deaf Educ. 9 (2004) 395–412. doi:10.1093/deafed/enh033.

[42] L. Cupples, T.Y.C. Ching, K. Crowe, M. Seeto, G. Leigh, L. Street, et al., Outcomes of 3-Year-Old Children With Hearing Loss and Different Types of Additional Disabilities, J. Deaf Stud. Deaf Educ. 19 (2014) 20–39. doi:10.1093/deafed/ent039.

[43] J. Black, L. Hickson, B. Black, Defining and evaluating success in paediatric cochlear implantation - An exploratory study, Int. J. Pediatr. Otorhinolaryngol. 76 (2012) 1317–1326. doi:10.1016/j.ijporl.2012.05.027.

[44] P. Statement, Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs., Pediatrics. 120 (2007) 898–921. doi:10.1542/peds.2007-2333.

[45] J. Sarant, D. Harris, L. Bennet, S. Bant, Bilateral versus unilateral cochlear implants in children: a study of spoken language outcomes., Ear Hear. 35 (2014) 396–409. doi:10.1097/AUD.0000000000000022.

[46] R.E.S. Lovett, P.T. Kitterick, C.E. Hewitt, A.Q. Summerfield, Bilateral or unilateral cochlear implantation for deaf children: an observational study., Arch. Dis. Child. 95 (2010) 107–112. doi:10.1136/adc.2009.160325.

[47] L. Van Deun, A. Van Wieringen, F. Scherf, N. Deggouj, C. Desloovere, F.E. Offeciers, et al., Earlier intervention leads to better sound localization in children with bilateral cochlear implants, Audiol. Neurotol. 15 (2009) 7–17. doi:10.1159/000218358.

[48] M. Sparreboom, J. van Schoonhoven, B.G.A. van Zanten, R.J.P.M. Scholten, E.A.M. Mylanus, W. Grolman, et al., The effectiveness of bilateral cochlear implants for severe-to-profound deafness in children: a systematic review., Otol. Neurotol. 31 (2010) 1062–1071. doi:10.1097/MAO.0b013e3181e3d62c.

[49] F. Scherf, L. van Deun, A. van Wieringen, J. Wouters, C. Desloovere, I. Dhooge, et al., Hearing benefits of second-side cochlear implantation in two groups of children, Int. J. Pediatr. Otorhinolaryngol. 71 (2007) 1855–1863. doi:10.1016/j.ijporl.2007.08.012.

[50] D.M. Zeitler, M.A. Kessler, V. Terushkin, T.J. Roland, M.A. Svirsky, A.K. Lalwani, et al., Speech perception benefits of sequential bilateral cochlear implantation in children and adults: a retrospective analysis., Otol. Neurotol. 29 (2008) 314–325.

[51] L. Van Deun, A. van Wieringen, J. Wouters, Spatial speech perception benefits in young children with normal hearing and cochlear implants., Ear Hear. 31 (2010) 702–713. doi:10.1097/AUD.0b013e3181e40dfe.

[52] R.Y. Litovsky, P.M. Johnstone, S.P. Godar, Benefits of bilateral cochlear implants and/or hearing aids in children., Int. J. Audiol. 45 Suppl 1 (2006) S78–S91. doi:P780766523R0572V [pii]\n10.1080/14992020600782956.

[53] T. Boons, J.P.L. Brokx, J.H.M. Frijns, L. Peeraer, B. Philips, a. Vermeulen, et al., Effect of Pediatric Bilateral Cochlear Implantation on Language Development, Arch. Pediatr. Adolesc. Med. 166 (2012) 28–34. doi:10.1001/archpediatrics.2011.748.

[54] J.C. Johnston, A. Durieux-Smith, D. Angus, A. O'Connor, E. Fitzpatrick, Bilateral paediatric cochlear implants: a critical review., Int. J. Audiol. 48 (2009) 601–617. doi:10.1080/14992020802665967.

[55] G. Kerr, S. Tuomi, A. Müller, Costs involved in using a cochlear implant in South Africa., S. Afr. J. Commun. Disord. 59 (2012) 16–26. doi:10.7196/SAJCD.117.

[56] South African Cochlear Implant Group (SACIG), Annual Report 2014, Unpublished Society Document, 2015.

[57] S. Archbold, M.E. Lutman, D.H. Marshall, Categories of auditory performance, Ann. Otol. Rhinol. Laryngol. 104 (1995) 312–314. doi:10.3109/03005364000000045.

[58] P.C. Stacey, H.M. Fortnum, G.R. Barton, A.Q. Summerfield, Hearing-impaired children in the United Kingdom, I: Auditory performance, communication skills, educational achievements, quality of life, and cochlear implantation., Ear Hear. 27 (2006) 161–186. doi:10.1097/01.aud.0000202353.37567.b4.

[59] M.C. Allen, T.P. Nikolopoulos, G.M. O'Donoghue, Speech intelligibility in children after cochlear implantation, Am J Otol. 19 (1998) 742–746. http://www.ncbi.nlm.nih.gov/pubmed/9831147.

[60] C. Allen, T.P. Nikolopoulos, D. Dyar, G.M. O'Donoghue, Reliability of a rating scale for measuring speech intelligibility after pediatric cochlear implantation., Otol. Neurotol. 22 (2001) 631–633.

[61] S. Archbold, M.E. Lutman, T. Nikolopoulos, Categories of auditory performance: Inter-user reliability, Br. J. Audiol. J. Audioly. 32 (1998) 7–12.

[62] B. Philips, P. Corthals, L. De Raeve, W. D'haenens, L. Maes, A. Bockstael, et al., Impact of newborn hearing screening: Comparing outcomes in pediatric cochlear

implant users, Laryngoscope. 119 (2009) 974–979. doi:10.1002/lary.20188.

[63] C.L. Budenz, S.A. Telian, C. Arnedt, K. Starr, H.A. Arts, H.K. El-Kashlan, et al., Outcomes of cochlear implantation in children with isolated auditory neuropathy versus cochlear hearing loss, Otol. Neurotol. 34 (2013) 477–483.

[64] C.E. Johnson, Introduction to auditory rehabilitation – A contemporary issues approach, Pearson Education, 2012.

[65] Department of Education, Republic of South Africa, Education White Paper 6 on Special Needs Education: Building an inclusive education and training system, Pretoria, 2001. http://www.education.gov.za/LinkClick.aspx?fileticket=gVFccZLi/tI= (accessed 14.01.16)

[66] Department of Education, republic of South Africa, Report on the implementation of Education White Paper 6 on Inclusive Education: An overview for the period 2013-2015, Pretoria, 2015. http://www.thutong.doe.gov.za/Default.aspx?alias=www.thutong.doe.gov.za/inclusiveeducation (accessed 14.01.16)

[67] A. Sharma, M.F. Dorman, A.J. Spahr, A sensitive period for the development of the central auditory system in children with cochlear implants: implications for age of implantation., Ear Hear. 23 (2002) 532–539. doi:10.1097/00003446-200212000-00004.

[68] R.F. Gray, S.E.M. Jones, I. Court, Cochlear implantation for progressive hearing loss., Arch. Dis. Child. 88 (2003) 708–711. doi:10.1136/adc.88.8.708.

[69] F.I. Ahmad, C.E. Demason, H.F. Teagle, L. Henderson, O.F. Adunka, C. a Buchman, Cochlear implantation in children with postlingual hearing loss, Laryngoscope. 122 (2012) 1852–1857. doi:10.1002/lary.23362; 10.1002/lary.23362.

[70] F.W.A.C. Scherf, L. van Deun, A. van Wieringen, J. Wouters, C. Desloovere, I. Dhooge, et al., Functional outcome of sequential bilateral cochlear implantation in young children: 36 months postoperative results, Int. J. Pediatr. Otorhinolaryngol. 73 (2009) 723–730. doi:10.1016/j.ijporl.2009.01.009.

[71] A.E. Geers, M.J. Strube, E.A. Tobey, D.B. Pisoni, J.S. Moog, Epilogue: factors contributing to long-term outcomes of cochlear implantation in early childhood., Ear Hear. 32 (2011) 84S–92S. doi:10.1097/AUD.0b013e3181ffd5b5.

[72] C.M.T. Robertson, T.M. Howarth, D.L.R. Bork, I.A. Dinu, Permanent bilateral sensory and neural hearing loss of children after neonatal intensive care because of extreme prematurity: a thirty-year study., Pediatrics. 123 (2009) e797–e807. doi:10.1542/peds.2008-2531.

[73] A. Van Wieringen, J. Wouters, What can we expect of normally-developing children implanted at a young age with respect to their auditory , linguistic and cognitive skills ?, Hear. Res. (2014) 1–9. doi:10.1016/j.heares.2014.09.002.

[74] I. Cruz, I. Vicaria, N. Wang, J. Niparko, A.L. Quittner, Cd.I. Team, Language and behavioral outcomes in children with developmental disabilities using cochlear implants, Otol. Neurotol. 33 (2012) 751–760.

[75] E. Kirkham, C. Sacks, F. Baroody, J. Siddique, M.E. Nevins, A. Woolley, et al., Health disparities in pediatric cochlear implantation: an audiologic perspective., Ear

Hear. 30 (2009) 515–25. doi:10.1097/AUD.0b013e3181aec5e0.

[76] E.A. Tobey, A.E. Geers, C. Brenner, D. Altuna, G. Gabbert, Factors associated with development of speech production skills in children implanted by age five., Ear Hear. 24 (2003) 36S–45S. doi:10.1097/01.AUD.0000051688.48224.A6.

[77] R.E. Stern, B. Yueh, C. Lewis, S. Norton, K.C.Y. Sie, Recent Epidemiology of Pediatric Cochlear Implantation in the United States: Disparity Among Children of Different Ethnicity and Socioeconomic Status, Laryngoscope. 115 (2005) 125–131. doi:10.1097/01.mlg.0000150698.61624.3c.

[78] Statistics South Africa, Mid-year population estimates, 2013 http://www.stats-sa.gov.za/publications/P0302/P03022013.pdf, 2013 (accessed 01.11.15).

[79] Statistics South Africa, Census 2011, 2011. http://mobi.statssa.gov.za/census2011/ First Language.html (accessed 01.11.15).

[80] M. Blecher, S. Harrison, Healthcare financing, in: P. Ijumba, A. Padarath (Eds.), South African Health Review 2006, Health Systems Trust, Durban, 2006. http// www.healthlink.org.za/uploads/files/chap3_06.pdf (accessed 01.11.15).

[81] E.M. Fitzpatrick, J. Ham, J. Whittingham, Pediatric Cochlear Implantation: Why Do Children Receive Implants Late?, Ear Hear. (2015). doi:10.1097/AUD.0000000000000184.

[82] J. Leigh, S. Dettman, R. Dowell, R. Briggs, Communication development in children who receive a cochlear implant by 12 months of age., Otol. Neurotol. 34 (2013) 443–50. doi:10.1097/MAO.0b013e3182814d2c.

[83] T.Y.C. Ching, H. Dillon, V. Marnane, S. Hou, J. Day, M. Seeto, et al., Outcomes of early- and late-identified children at 3 years of age: findings from a prospective population-based study., Ear Hear. 34 (2013) 535–52. doi:10.1097/AUD.0b013e3182857718.

[84] M. Armstrong, A. Maresh, C. Buxton, P. Craun, L. Wowroski, B. Reilly, et al., Barriers to early pediatric cochlear implantation, Int. J. Pediatr. Otorhinolaryngol. 77 (2013) 1869–1872. doi:10.1016/j.ijporl.2013.08.031.

[85] T. le Roux, D.W. Swanepoel, A. Louw, B. Vinck, M. Tshifularo, Profound childhood hearing loss in a South Africa cohort: Risk profile, diagnosis and age of intervention, Int. J. Pediatr. Otorhinolaryngol. 79 (2015) 8–14. doi:10.1016/j.ijporl.2014.09.033.

[86] L. Percy-Smith, P. Cayé-Thomasen, N. Breinegaard, J.H. Jensen, Parental mode of communication is essential for speech and language outcomes in cochlear implanted children., Acta Otolaryngol. 130 (2010) 708–715. doi:10.3109/00016480903359939.

Appendix A Supplementary data

Table A.1: Risk and family factor prevalence

Syndromes and additional developmental conditions identified	% (n)	Risk factors identified	% (n)	Family factors identified	% (n)
Syndromes		**Prenatal risk factor**		Family history of permanent childhood	
Any syndrome diagnosed (including syndromes listed below)	9.5 (24/252)	Rubella	6.2 (14/225)	hearing loss	20.1 (44/219)
		Cytomegalovirus	3.6 (8/225)		
		Twin/triplet	3.1 (7/225)	Parental marital status	
Waardenberg Syndrome	5.2 (13/252)	Syphilis	0.4 (1/225)	Married	74.2 (196/264)
Ushers Syndrome	1.2 (3/252)	Toxoplasmosis	0.4 (1/225)	Divorced	15.9 (42/264)
Pierre Robin Syndrome	0.8 (2/252)			Single	8.7 (23/264)
Leopard Syndrome	0.8 (2/252)			Partner, not married	1.1 (3/264)
				Parental hearing status	
				Both hearing	96.4 (268/278)
				One/both hearing loss	2.5 (7/278)
Additional developmental conditions		**Natal risk factor**			
		Admittance to NICU	26.9 (43/160)	Communication mode of mother	
1 or more condition present	24.4 (64/262)	Prematurity (≤34 weeks gestation)	13.9 (32/230)	Oral	97.8 (266/272)
Visual impairment	7.6 (20/262)	Low birth weight (<2500g)	9.1 (21/230)	Sign Language	1.5 (4/272)
Cerebral palsy	5.3 (14/262)	Extremely low birth weight (<1500g)	4.8 (11/230)	Total communication	0.7 (2/272)
ADHD	4.6 (12/262)	Birth asphyxia	1.7 (4/230)		
Mobility impaired	3.1 (8/262)	Maternal hypertensive disorder in		Communication mode of father	
Learning disabilities	2.7 (7/262)	pregnancy	1.3 (3/230)	Oral	97.5 (274/281)
Autism	1.9 (5/262)	Rupture of membranes	1.3 (3/230)	Sign Language	1.4 (4/281)
Apraxia	1.9 (5/262)	Birth trauma	0.9 (2/230)	Total communication	1.1 (3/281)
Developmental motor delay	1.5 (4/262)	Rh incompatibility	0.4 (1/230)		
Epilepsy	1.1 (3/262)			Highest educational qualification: mother	
Cleft lip and/or palate	1.1 (3/262)	**Postnatal risk factor**		Tertiary qualification (University)	40.4 (38/94)
		Meningitis	13.6 (31/228)	Tertiary qualification (other)	19.1 (18/94)
		Neonatal jaundice/ hyperbilirubinemia	7.9 (18/228)	Matric completed	33.0 (31/94)
		Blood transfusion	1.8 (4/228)	High school (Grade 8-11)	7.4 (7/94)
		Viral infection (unspecified)	5.3 (12/228)		
		Ototoxic drugs	3.1 (7/228)	Highest educational qualification: father	
		Mumps	0.9 (2/228)	Tertiary qualification (University)	58.5 (48/82)
		Measles	0.4 (1/228)	Tertiary qualification (other)	15.9 (13/82)
		Tuberculosis	0.4 (1/228)	Matric completed	23.2 (19/82)

High school (Grade 8-11)	2.4 (2/82)
Mother employment status	
Employed	76.6 (98/128)
Not employed	23.4 (30/128)
Father employment status	
Employed	99.3 (146/147)
Not employed	0.7 (1/147)

Table A.2: Suspected prognostic factors

Prognostic factors	Two-way categories	*% (n)*
Demographical and hearing loss factors		
Gender	Male	52.5 (158/301)
	Female	47.5 (143/301)
Ethnic category	Caucasian	61.8 (186/301)
	Other	38.2 (115/301)
Age of diagnosis of hearing loss (congenital/early	Early diagnosis (<36 months)	93.4 (114/122)
onset only)	Late diagnosis (≥ 36 months)	6.6 (8/122)
Cochlear implant factors		
1st ear left/ right	Left	35.8 (106/296)
	Right	64.2 (190/296)
Age at implant 1st ear (congenital/early onset only)	Early implantation (<36 months)	49.2 (92/187)
	Late implantation (≥36 months)	50.8 (95/187)
Delay from diagnosis to 1st implant	<12 months	29.3 (55/188)
	≥12 months	70.7 (133/188)
Bilateral implant (including only cases with at least 6	Yes	29.0 (87/301)
month experience with bilateral implant)	No	71.0 (214/301)
Family factors		
Family history of permanent childhood hearing loss	Yes	20.1 (44/219)
	No/ unsure	79.9 (175/219)
Parental marital status	Married	74.2 (196/264)
	Single/ divorced	25.8 (68/264)
Highest educational qualification: Mother	High school	40.4 (38/94)
	Tertiary education	59.6 (56/94)
Highest educational qualification: Father	High school	25.6 (21/82)
	Tertiary education	74.4 (61/82)
Mother employment status	Employed	76.6 (98/128)
	Not employed	23.4 (30/128)
Risk factors		
Additional developmental conditions	Yes (1 or more)	
	None	24.4 (64/262)
Admittance to NICU	Yes	75.6 (198/262)
	No	26.9 (43/160)
Prematurity (≤34 weeks gestation)	Yes	73.1 (117/160)
	No	13.9 (32/230)
Prenatal risk factors	Yes (1 or more)	86.1 (198/230)
	None	15.6 (35/225)
Natal risk factors	Yes (1 or more)	84.4 (190/225)
	None	23.5 (54/230)
Post-natal risk factors	Yes (1 or more)	76.5 (176/230)
	None	36.4 (83/228)
Meningitis	Yes	63.6 (145/228)
	No	13.6 (31/228)
Risk factors present (pre-natal, natal, post-natal	Yes (1 or more)	86.4 (197/228)
combined)	None	55.6 (133/239)
		44.4 (106/239)

Table A.3: Predictors having a possible association with outcome variables

Potential predictors	CAP score (in two categories)	SIR score (in two categories)	Communication Mode	Educational placement
Ethnic category (n=301)	0.095*	0.031**	0.096*	0.015**
Age at implant 1st ear (congenital/early onset hearing loss only) (n=187)	-	-	-	0.054**
Delay from diagnosis to 1st implant (n=188)	-	-	-	0.005**
Bilateral implantation (n=301)	0.000**	0.000**	0.000**	0.000**
Highest educational qualification of mother (n=94)	-	-	-	0.005**
Additional developmental conditions (n=262)	-	0.002**	0.027**	0.002**
Admittance to NICU (n=160)	0.002**	0.022**	0.008**	0.037**
Prematurity (≤34 weeks gestation) (n=230)	0.053*	-	-	-
Natal risk factors (n=230)	0.005**	0.058*	0.011**	0.072*
Post-natal risk factors (n=228)	0.003**	0.078*	0.085*	-
Meningitis (n=228)	0.066*	-	-	-
Risk factors present (in general) (n=239)	0.003**	-	0.031**	0.036**

**possible significance (0.05 < p < 0.1); **significance (p < 0.05)*