

Original Article

Auditory Performance in Early Implanted Children with Cochleovestibular Malformation and Cochlear Nerve Deficiency

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OBJECTIVES: This study aimed to report the auditory performance in children with cochleovestibular malformation (CVM)/cochlear nerve deficiency (CND) who were implanted early at the Universiti Kebangsaan Malaysia Medical Centre, using Categorical Auditory Performance (CAP)-II score and Speech Intelligibility Rating (SIR) scales, and to compare the outcome of their matched counterparts.

MATERIALS and METHODS: A total of 14 children with CVM/CND with unilateral cochlear implant (CI) implanted before the age of 4 years old were matched and compared with 14 children with normal inner ear structures. Their improvement in auditory performance was evaluated twice using CAP-II score and SIR scales at 6-month intervals, with the baseline evaluation done at least 6 months after implantation.

RESULTS: The average age of implantation was 31 ± 8 and 33 ± 7 months for the control group and the case (CVM/CND) group, respectively. Overall, there were no significant differences in outcome when comparing the entire cohort of case subjects and their matched control subjects in this study. However, the improvement in CAP-II scores and SIR scales among the case subjects in between the first and second evaluations was statistically significant ($p=0.040$ and $p=0.034$, respectively). With longer duration of CI usage, children with CVM/CND showed significant speech perception outcome evident by their SIR scales ($p=0.011$).

CONCLUSION: Children with radiographically malformed inner ear structures who were implanted before the age of 4 years have comparable performance to their matched counterparts, evident by their similar improvement of CAP-II scores and SIR scales over time. Hence, this group of children benefited from cochlear implantation.

KEYWORDS: Cochlear implant, cochlear nerve, auditory perception, speech intelligibility

INTRODUCTION

As a university-based national cochlear implant (CI) center, we have successfully implanted more than 400 CI devices since the CI team was set up in 1995. The goal of both hearing aids and CIs is to ensure that a child with hearing loss will be able to perceive sound better as soon as possible and subsequently acquire normal speech. This is important for the development of their interpersonal skills and self-confidence. A study by Umat et al. ^[1] showed that children who were better at integrating the sounds they have perceived were also those who were more confident in their speech with others.

Since its first introduction in the 1970s, cochlear implantation has emerged as the most effective surgical technique to restore auditory function in patients with bilateral severe-to-profound sensorineural hearing loss. With improved techniques and more expe-

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rience, surgeons can perform cochlear implantations in challenging cases, including children as young as 12 months.

Nevertheless, performing cochlear implantation surgery in a congenital malformed inner ear still poses a challenge. Major concerns in this group of candidates are cerebrospinal fluid (CSF) gusher, the incomplete insertion of electrodes into the cochlea, unpredictable distribution of spiral ganglions that are able to be stimulated, abnormal facial nerve activation, postoperative meningitis, and uncertainty regarding the postoperative auditory performance^[2]. These risks increase with the degree of cochleovestibular malformation (CVM). Therefore, implant candidacy in this set of patients remains relatively controversial from economic and clinical points of view.

CVM classification was first described by Jackler et al. in 1987^[3]. Sennaroglu and Saatci revised Jackler's system in 2002^[4]. Sennaroglu^[2] then further revised the classification in 2010, which is the classification currently used. Cochlear nerve (CN) classification was first described by Govaerts et al.^[5] in 2003. The classification defined four types of CNs based on the otic, labyrinthine, and internal auditory meatus (IAM) abnormalities. However, this grading did not emphasize the contents of IAM or CN aplasia or hypoplasia. In 2016, Birman et al.^[6] proposed a new grading system for CN classifications.

In this study, the types and severity of CVM among case subjects were graded using the revised Sennaroglu classification in 2010^[2]. A revised CN classification by Birman et al.^[6] was used to categorize the types of CN deficiency (CND).

This study reports the auditory performance outcome in children with CVM and CND after receiving early cochlear implantation, using Categorical Auditory Performance (CAP)-II scores and Speech Intelligibility Rating (SIR) scales.

MATERIALS AND METHODS

Study Design

This study was a cross-sectional, longitudinal, single-center, matched cohort analysis of post-implantation auditory performance of all children with CVM and/or CND who received their implants between 2012 to 2017 in our center. The study was carried out in the Otorhinolaryngology, Head and Neck Surgery clinic, conducted within the proposed time frame of 12 months from March 2018 to March 2019.

All prelingual deafened children with underlying CVM and/or CND who received their first cochlear implantation before the age of 4 years (48 months), with traceable preoperative radiological investigations (computed tomography [CT] and magnetic resonance imaging [MRI]), who have been using the device for at least 6 months, and who regularly attended audiological assessment and a speech rehabilitation program after implantation were recruited. Implant recipients with incomplete data or with underlying brain parenchymal disease were excluded. Those who did not wish to participate in this study were also excluded. To exclude several variables that may affect outcome, these groups of children were matched with their radiographically normal inner ear counterparts.

A detailed medical chart review was executed to record the age of implantation, duration of implant usage, mode of hearing, and type

of device model implanted. Intraoperative notes were reviewed to chart the presence of CSF gusher during cochleostomy or round window puncture, completeness of electrode insertion, and presence of neural response telemetry (NRT) on electrode insertion.

CAP-II score and SIR scale are non-language-based indexes, which can be objectively used across all races irrespective of their acquired language. Given the multiracial pool of our patients, these assessment tools were chosen for this study.

All children who participated in this study were assessed twice at 6-month intervals. Their auditory performances were scored and rated accordingly. The first evaluation was done at least 6 months after CI switch-on for all subjects. The second evaluation was done 6 months after the first evaluation.

The mean scores for both groups of patients during both periods of evaluation were calculated, and the significance in difference between the mean values of CAP-II scores and SIR scales was calculated using paired *t*-test.

Descriptive analysis was used to summarize the preoperative, intraoperative, and postoperative data collected, namely, the CVM and/or CND classifications, age at surgery, device model implanted, presence of CSF gusher during cochleostomy or round window puncture, completeness of electrode insertion, presence of NRT measurement, mode of hearing, and duration of implant usage during the two periods of evaluation. The findings from this study will allow us to understand better the association between CVM and/or CND and the auditory outcome after receiving CI and to predict the long-term speech perception outcome. Thus, by obtaining the data pertaining to the outcome after cochlear implantation in this group of patients, it will assist in the surgeon's decision making regarding surgery and enable us to outline more realistic expectations for CI candidates and their families.

Statistical Analysis

Data was analysed using the Statistical Package for the Social Sciences (SPSS) version 24 (IBM Corp.; Armonk, NY, USA).

Symmetrical testing using the McNemar test was performed and it showed no significant asymmetry of the matched variables between the case and control group, indicating adequacy for matching for statistical analysis. Paired *t*-test was used to analyse the improvement in CAP-II scores among the case and control subjects between the first and second evaluations.

Wilcoxon signed rank test was applied to compare the improvement in SIR scales among the case and control subjects between the first and second evaluations. The Pearson correlation test was used to measure the linear correlation between the CAP-II scores and SIR scales and duration of implant usage after CI switch-on.

RESULTS

A total of 28 patients were included in our study, 14 of whom were case subjects with underlying CVM and/or CND, matched with 14 control implant recipients with radiologically normal inner ear structures.

All subjects were completely matched for mode of hearing (on bimodal hearing using CI in one ear and a hearing aid in the other ear). They were also matched for model of device implanted, completeness of electrode insertion, and presence of NRT measurement intraoperatively.

Although it was not statistically significant ($p=0.504$), subjects with normal inner ear structures were noted to be implanted slightly ear-

Table 1. The CAP-II scores and SIR scales of the children with Cochleovestibular Malformation and Cochlear Nerve deficiency and their matched control subjects during both period of evaluations

	CAP-II Score (1 st data)	CAP-II Score (2 nd data)	SIR Scale (1 st data)	SIR Score (2 nd data)
Case Subjects				
A1	9	9	5	5
A2	5	5	2	3
A3	5	6	3	4
A4	5	6	1	3
A5	3	4	2	3
A6	8	8	4	4
A7	6	6	3	3
A8	5	5	2	3
A9	8	8	4	4
A10	5	5	3	3
A11	8	8	4	4
A12	0	1	1	1
A13	5	5	2	2
A14	4	4	1	1
Mean	5.43±2.34	5.71±2.09	2.64±1.28	3.07±1.14
Control Subjects				
B1	8	8	4	4
B2	5	5	4	4
B3	3	3	1	2
B4	5	5	3	3
B5	6	6	4	4
B6	8	8	5	5
B7	8	8	5	5
B8	3	4	1	1
B9	7	7	4	4
B10	7	7	4	4
B11	6	6	4	4
B12	4	4	2	2
B13	4	4	3	3
B14	3	3	1	1
Mean	5.50±1.91	5.57±1.83	3.21±1.42	3.29±1.33

lier (mean age of implantation, 31 ± 8 months) than those with underlying CVM and/or CND (mean age of implantation, 33 ± 7 months).

Overall, there were no significant differences in outcome when comparing the entire cohort of case subjects and their matched control subjects in this study on the basis of their CAP-II score and SIR scales during both periods of evaluation (Table 1). However, the improvement in CAP-II scores among the case subjects between the first and second evaluations was statistically significant ($p=0.04$, paired *t*-test) (Figure 1). The improvement in SIR scales among the case subjects in between the first and second evaluations also showed a significant difference ($p=0.034$, Wilcoxon signed rank test) (Figure 2).

There was a significant correlation seen between the duration of implant usage and the SIR scales achieved during the second evaluation among the case subjects ($p=0.011$).

DISCUSSION

Numerous studies have concluded that prelingual deafened children who receive CI early (with or without CVM) will have better auditory and speech performance than children implanted at a later age [7, 8]. A study by Govaerts et al. [9] showed that intervention before the age of 4 years seemed critical to avoid irreversible loss of auditory performance. Children who were implanted before the age of 2 years were shown to achieve optimal results, with 90% of them being able to get integrated into mainstream kindergarten and reach good CAP scores.

A cross-sectional study by Goh et al. [10] in 2018 showed that the mean age at implantation for children who were able to use oral communication and attend mainstream education was 38 months. Hence, they concluded that those implanted at the age of less than 4 years would have better functional auditory and oral performance outcomes. In our study, the patients selected for evaluation were children implanted before the age of 4 years.

Traditionally, children with CVM and/or CND were not subjected to CI. However, in recent years, more parents were willing to take the chance and requested their children to undergo CI despite being diagnosed with underlying CVM and/or CND.

Patients with CVM and/or CND account for 20% of pediatric implant candidates [2]. Many studies have shown that patients with inner ear abnormalities can benefit from cochlear implantation with good but varying outcomes. Based on this premise, we conducted this longitudinal study to report the auditory outcome in this set of CI recipients in our medical center.

The mean age of implantation for both our case and control subjects did not differ significantly ($p=0.504$). This demonstrates that all cases were referred and evaluated at about the same age. A retrospective study by Van Wermeskerken et al. [11] also showed similar findings, with no significant difference between the mean age of implantation for those with and without inner ear malformation (3.9 years and 2.8 years, respectively; $p=0.130$). In a large review by Buchman et al., [12] the age of implantation for children with inner ear malformation ranged from 17 to 212 months (17 years, 8 months), with a mean age of 66 months (5 years, 6 months).

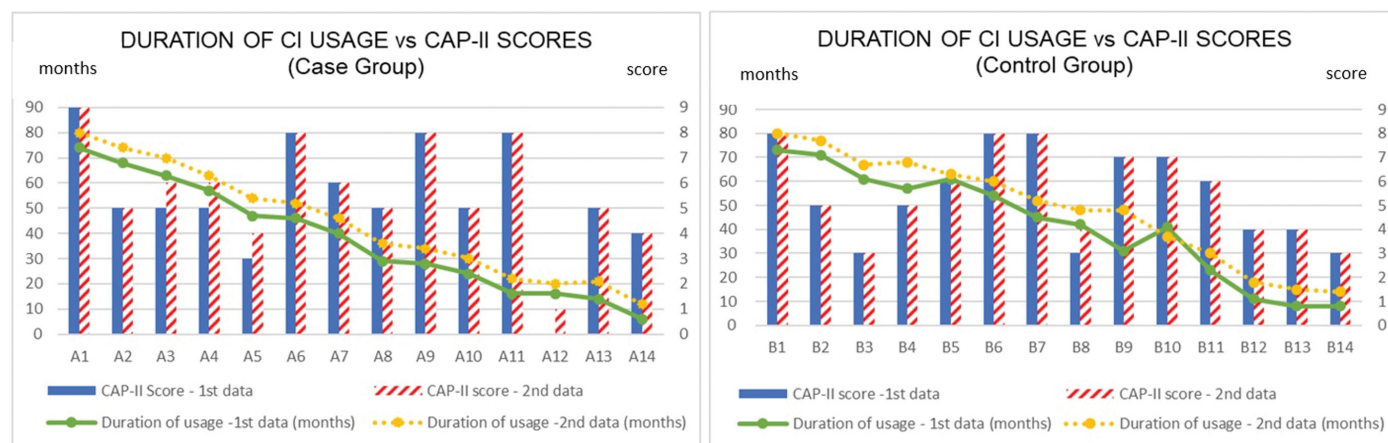


Figure 1. The changes in CAP-II scores among the case and control subjects between the 1st and 2nd evaluations and the relationship of the scores to the duration of CI usage.

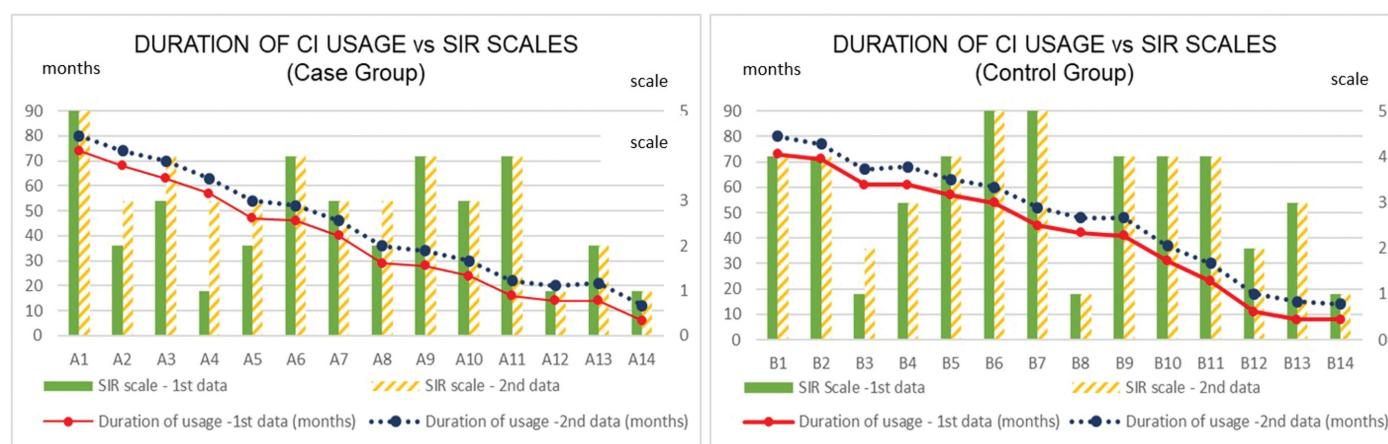


Figure 2. The changes in SIR scales among the case and control subjects between the 1st and 2nd evaluations and the relationship of the scores to the duration of CI usage.

The most common intraoperative concerns for our cases with underlying CVM were CSF gusher and incomplete insertion of electrodes, as well as unpredictable distribution of spiral ganglions that can be stimulated in both groups with CVM and CND.

The presence of CSF gusher was seen in 5 (35.7%) of 14 of our subjects, and one (7.1%) subject with common cavity had incomplete insertion of electrodes. In a similar study by Buchman et al.,^[12] CSF gushers were encountered in 6 (21%) out of their 28 patients, and three (11%) patients had incomplete insertion of electrodes.

In a retrospective study by Adunka et al.,^[13] all their cases of perilymph fluid gushers were managed intraoperatively by packing the cochleostomy site tightly with connective tissue after CI electrode insertion. They also concluded that the presence or absence of a perilymph gusher did not significantly influence results after cochlear implantation. Similar outcomes were also observed in our series of patients.

NRT

NRT is a quick, non-invasive way of recording the electrically evoked compound action potential (ECAP) of the peripheral nerves in situ. It uses a small mobile device in a near-field environment. It records the neural responses to electrical stimulation at discrete sites along

the electrode array, providing direct evidence of the auditory nerve's responsiveness^[14].

The most clinically relevant measure of NRT is the ECAP threshold, which has been found to lie close to or above the postoperative comfort level (C-level)^[15]. By obtaining this information intraoperatively, it can be used as a guide in the first mapping session, especially in young children or difficult-to-test recipients. In a congenitally deaf child, the perceptual markers for comfortable listening level may be absent as the child has no prior auditory experience; however, these markers possibly will develop overtime as the child experiences more auditory stimuli and subsequently is able to respond appropriately.

Four (28.5%) out of 14 of our case subjects had unmeasurable NRT intraoperatively. However, three of these four subjects had favorable auditory outcomes after several years of implantation, based on their high CAP-II scores and SIR scales. De Moura et al.^[16] in 2014 studied the longitudinal outcome in children who received CIs with or without intraoperative response to NRT. The results from their study showed that 49% of those who had no response during surgery would eventually show NRT responses after 4.9 months of stimulation. These findings suggested that there is a tendency for the response to appear after continual use of CI. They also found that there is no significant influence of age at implantation and length of

hearing loss on the absence of response intraoperatively or in later appearance of response.

Patients with malformed inner ear structures tend to demonstrate fluctuating and higher threshold and C-level measurements (noted by their high level of intraoperative NRT and first mapping switch-on session). They required wider modes of stimulation than patients with normal inner ear structures. More frequent mapping sessions and readjustments were usually required in our cases with CVM and/or CNL to achieve the best C-level for better compliance and outcome.

CAP-II Score and SIR Scale

Various tools or questionnaires have been created and used to measure the functional outcome after cochlear implantation. In our center, most CI recipients' auditory performance was assessed and monitored using Meaningful Auditory Integration Scales, Meaningful Use of Speech Scale, and Parents' Evaluation of Aural/Oral Performance of Children.

In this study, the assessment tools used were the CAP-II score and SIR scale. The components in CAP-II score and SIR scale are not lengthy and are easy to understand; thus, assessments are not time consuming. The interviewer may start the session with an open-ended question pertaining to the child's current performance, subsequently categorized to the most appropriate level in the indexes.

CAP was first introduced by Archbold et al.^[17] in 1995. CAP was designed to provide an outcome measure that would be accessible to both parents and health care professionals. It is thought to reflect the implanted child's everyday auditory performance in a more realistic way. Given its subjective form of outcome measures, Archbold et al. in 1997 verified the reliability of CAP for use in CI programs^[18]. In 2010, Louise et al.^[19] revised the CAP score and named it as CAP-II. This revised version is composed of a graded scale ranging from 0 to 9 (which traditionally ranges from 0 to 7).

The SIR scale is a practical and reliable clinical measure of speech intelligibility. It consists of a five-point rating scale that increases in levels of complexity along with the child's speech production. The SIR scale provides a baseline of a child's speech intelligibility skills and can be used as a monitoring tool to assess his or her speech over time. Allen et al.^[20] in 2001 assessed the inter-rater reliability of SIR, and they found a high rate of agreement between observers when they used the SIR scale to evaluate speech intelligibility of deaf children after cochlear implantation.

Our results showed that although there were no significant differences in overall outcome between the entire cohort of children with CVM and/or CNL and their matched control subjects, children with malformed inner ear structures demonstrated significant improvements in their auditory outcomes on the basis of their CAP-II scores over time. In addition, with longer duration of CI usage, these children also developed better speech perception outcome evident by the SIR scales they achieved over time. Numerous other studies also concluded similar outcomes.

Bille et al. in 2015^[21] demonstrated that there were no statistical differences between the CAP and SIR scores after 3 years of implan-

tation between those with inner ear malformation and the control group ($p=0.29$ and 0.40 , respectively).

In their series of 34 children, Eisenman et al.^[22] found that the children with inner ear malformations showed initial delay in improvement after implantation. However, over time, their speech perception outcome became comparable to their matched radiographically normal counterparts.

In 2014, Vincenti et al.^[24] reported the auditory performance after cochlear implantation in five children with CNL. Their results showed that the outcomes of cochlear implantation in these five children with CNL were extremely variable. Therefore, they concluded that cochlear implantation can be an option in children with CNL, but careful family counseling is needed to discuss the expected outcome in view of possible restricted benefits.

Interestingly, a study by Rachovitsas et al.^[23] concluded that apart from structural inner ear abnormality, bilingualism could be a contributing factor that could affect outcomes. In addition, they proposed that cognitive function and developmental delay should be assessed preoperatively because these factors have a possible impact on outcomes and habilitations.

Study Limitations

Because of the relatively small number of subjects in both cohorts and each subgroup of CVM and CNL, the power of this study might be affected. The possibility of a false negative result is inevitable.

We also did not divide the case subjects into separate categories of CVM only, CNL only, or those with both CVM and CNL.

This study did not include the subjects' preoperative CAP-II scores and SIR scales or their baseline pure tone audiogram and brainstem evoked response results to denote the actual performance outcome before and after implantation.

Variables such as socioeconomic background, parent's education level, and child's cognitive function were not discussed in this study. These factors are known to affect speech and language acquisition, especially among pediatric CI users.

CONCLUSION

Successful implantation with favorable outcomes was seen in our series of children with radiographically malformed inner ear structures evident by their comparable improvement of CAP-II scores and SIR scales over time. Therefore, these groups of children inarguably should be given the option of early cochlear implantation to enhance their hearing level and subsequently develop acceptable speech.

Cerebrospinal fluid gusher must be anticipated in cases with CVM, especially those associated with an enlarged vestibular aqueduct. Extra care is essential when faced with such cases, during and after surgery.

Some cases with absence of NRT measurement intraoperatively still show satisfactory speech performance outcome over time, especially when complemented with intensive habilitation.

CAP-II score and SIR scale are deemed good assessment tools to assess speech performance outcome among our CI recipients, regardless of their race and native language.

Ethics Committee Approval: This study was approved by the Research Ethics Committee of UKM Medical Centre on the 30th March 2018 (JEP-2017-768).

Informed Consent: Written informed consent was obtained from the participants' guardians.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – A.A., G.B.S., C.U.; Design – A.A., G.B.S., C.U.; Supervision – A.A., G.B.S., C.U.; Resource – A.A., G.B.S.; Materials – A.A., G.B.S.; Data Collection and/or Processing – I.A.O.; Analysis and/or Interpretation – A.A., I.A.O., G.B.S., C.U.; Literature Search – A.A., G.B.S., I.A.O.; Writing – A.A., I.A.O., G.B.S., C.U.; Critical Reviews – A.A., G.B.S., C.U., R.T.

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