A Study of Outcome of Pediatric Cochlear Implantation in Patients with Cochleovestibular Nerve Deficiency

Senthil Vadivu Arumugam, Geetha Nair, Vijaya Krishnan Paramasivan, Sunil Goyal, Sathiya Murali, Mohan Kameswaran

Madras ENT Research Foundation (Pvt) Ltd, Chennai, India (SVA, VKP, SM, MK)
Department of ENT, Government Medical College, Kerala, India (GN)
Department of ENT-HNS, Army Hospital (R&R), Delhi, India (SG)

OBJECTIVES: A cochleovestibular nerve deficiency (CVND) could compromise stimulation of nerve by electrical pulses delivered from a cochlear implant, thereby hindering activity along auditory pathway. The evaluation of children with congenital hearing loss with a high-resolution magnetic resonance imaging is presently the investigative modality of choice to diagnose CVND. The objectives included (1) to study the prevalence of CVND among children with prelingual congenital severe to profound hearing loss; (2) to assess post cochlear implantation (CI) outcomes in children with CVND using categories of auditory performance (CAP), speech intelligibility rating (SIR), and cortical auditory evoked potentials (CAEPs); and (3) to propose a management protocol for these children.

MATERIALS and METHODS: All CI procedures performed during the study period in children 5 years or younger were included in study. All patients who were older than 5 years or had syndromic associations, multiple disabilities, second side or revision CI were excluded from the study. Children with unilateral cochleovestibular nerve aplasia and all other cases of CVND (type Ila and Iib) were advised to undergo CI on side with more radiologically robust nerve and/or cochlea anatomy. Children with bilateral CVND were included in group A, and age-matched cochlear implant candidates with normal cochleovestibular nerve anatomy were included in group B for statistical comparison of outcomes.

RESULTS: In group A, post CI CAP and SIR, CAEP amplitude and latency at 12 months showed statistically significant difference (p<0.05) compared with preoperative values. However, mean score of CAEP latency and amplitude and SIR score was worse for group A compared with group B at 12 months, which was statistically significant (p<0.05).

CONCLUSION: This study supports the fact that CI is a viable option to be offered in children with CVND (type Ila and Iib) for the development of auditory perception and speech.

KEYWORDS: Cochlear nerve hypoplasia, cochlear nerve aplasia, cortical auditory evoked potential, category of auditory perception, speech intelligibility rating, objective outcomes

INTRODUCTION
Cochlear implantation (CI) is an established modality of treatment in providing auditory perception and speech development for children with congenital severe to profound hearing-impairment. However, the stimulation of nerve by electrical impulses delivered from cochlear implant electrodes, around the modiolus through spiral ganglion, would be limited in patients with cochleovestibular nerve deficiency (CVND). This is turn would limit neurological activity generated at higher center along the auditory pathway and its associated areas.

The term CVND was coined to refer to the situation in which the cochlear nerve or its osseous conduit is not visible on imaging [1-3]. However, the diagnosis of CVND is decidedly more challenging and makes high-resolution magnetic resonance imaging (MRI)
The cortical auditory evoked potentials (CAEPs) is a cortically generated potential that provides information about the integrity of the auditory system and the neural processing of sound beyond the auditory brainstem. It is a biomarker of maturation of the auditory cortex in response to sound stimulus. Responses from normal hearing children younger than 5 years show only a large, broad P1 (around 100 milliseconds) followed by the N2 component \[6, 7\]. The emergence of the P1-N1-P2 complex as seen in adults occurs by the age of 9-12 years \[7\]. In children with a cochlear implant, CAEPs can be reliably recorded in response to the sound field stimuli transduced by the implant’s speech processor or by electrical pulse trains delivered directly to the specific implant electrodes \[8-11\]. Although CAEPs in children who receive the implant at an early age are of similar morphology to those of age-matched peers with normal hearing, children who receive implants at a later age may not show age-appropriate changes \[8\]. However, the use of CAEPs as a measure of objective outcome in CVND is not clearly defined in the literature.

The aim of this study was to determine the outcomes of pediatric CI in children with CVND. The objectives were to: (1) study the prevalence of CVND among children with prelingual congenital severe to profound hearing loss; (2) assess post-CI outcomes in children with CVND using categories of auditory performance (CAP), speech intelligibility rate (SIR), and CAEP; and (3) propose a management protocol for these children with CVND.

MATERIALS AND METHODS

Our study is a prospective case-control study conducted at a high-volume cochlear implant center in India between July 1, 2015, and June 30, 2018. All CI performed in children aged 5 years or younger were included in the study. All patients who were older than 5 years or had syndromic associations, multiple disabilities, and second side implant or revision CI were excluded from the study to reduce bias. Informed consent was taken, and the institutional ethical committee clearance was obtained.

A detailed medical history, clinical examination, and investigations were conducted per our institutional protocol. A battery of comprehensive evaluations including the opinions of the pediatrician, neurologist, ophthalmologist, speech pathologist, clinical psychologist, and occupational therapist were sought for all children.

Audiological battery of tests performed included pure tone audiometry, impedence audiometry, oto-acoustic emission, brain stem evoked response audiometry, hearing aid trial, and CAEP. 3 Tesla MRI of inner ear and brain was conducted to assess the anatomy of the inner ear and cochleovestibular nerve. CVND was classified based on the types as described by Casselman and Govaerts. The diagnosis of hypoplastic cochlear nerve was based on oblique sagittal MRI images through the middle portion of internal auditory canal (IAC) and comparison of the diameter of cochlear nerve to that of the facial nerve. If the diameter of cochlear nerve was less than or equal to that of the facial nerve, it was considered as hypoplastic cochlear nerve. Figure 1 depicts 3 Tesla MRI oblique sagittal view through middle part of IAC of a 2-year-old boy with bilateral CVND (Casselman type IIa) and cochlear nerve aplasia.

As per our institutional protocol, all children with bilateral cochleovestibular nerve aplasia (Casselman type I) and/or cochlear aplasia were advised auditory brainstem implantation (ABI). Children with unilateral cochleovestibular nerve aplasia and all other cases of CVND (Casselman type IIa and type IIb) were advised to undergo CI on side with more radiologically robust nerve and/or cochlea anatomy. The distribution and management protocol followed at our center for implantation in children with CVND is shown in Figure 2.

Children with bilateral CVND were included in group A, and age matched cochlear implant candidates having normal cochleovestibular nerve defect were included in group B.

MAIN POINTS

- Cochleovestibular nerve deficiency (CVND), seen in congenital sensorineural hearing loss, is an uncommon anomaly encountered by a neuro-otologist or implant otologist.
- Choice of treatment in CVND is a gray zone. Cochlear implantation is a viable option and should be offered to this group of patients for development of audition and speech.
- Parents need to be adequately counselled about variable outcomes and possibility of limited benefit and informed about the option of auditory brain stem implantation as an alternative.
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CI was performed under general anesthesia, and their outcomes were evaluated. All children underwent CI through transmastoid posterior tympanotomy approach using round window insertion by a senior surgeon. Med EL Pulsar device (Innsbruck, Austria) with Opus II speech processor was used in all cases. Switch-on was done on postoperative day 21 as per our institutional protocol.

Regular mapping was carried out, and objective audiological outcomes were assessed periodically using CAEP amplitude and latency, CAP, and SIR.

The values of CAEP amplitude and latency at 65 dB sound pressure level (SPL) in “ta” sound stimuli was used for our study because normal conversation falls at 65 dB SPL and “ta” is the most common speech sound across different languages. The typical CAEP wave morphology is depicted in Figure 3.

**Table 2. Audiological outcomes: Cortical Auditory Evoked Potentials (CAEPs)**

| Serial no. | Age at cochlear implant, y | CAEP amplitude | CAEP latency |
|------------|----------------------------|----------------|--------------|
|            | Group A at 12 months | Group B at 12 months | Group A at 12 months | Group B at 12 months |
| 1          | 3                         | 6.7             | 7.56         | 94            | 82            |
| 2          | 5                         | 3.73            | 5.94         | 101           | 92            |
| 3          | 4.6                       | 5.94            | 6.66         | 95            | 83            |
| 4          | 4                         | 5.72            | 7.04         | 97            | 87            |
| 5          | 4.2                       | 7.56            | 7.64         | 83            | 78            |
| 6          | 4.8                       | 6.14            | 7.52         | 88            | 76            |
| 7          | 3.8                       | 6.04            | 6.14         | 92            | 84            |
| 8          | 3.01                      | 5.94            | 6.8          | 94            | 84            |
| Mean       | 3.9                       | 5.6             | 6.8          | 94.5          | 84.5          |

CAEP: cortical auditory evoked potential

**Table 3. Distribution of cochleovestibular nerve deficiency based on Casselman and Govaerts’ classification**

| Type | Affected nerve on imaging | Remarks | Number (n) |
|------|---------------------------|---------|------------|
| I    | Aplasia of CVN            | Labyrinth may be normal or dysplastic; internal auditory canal is stenotic | 15 ears |
| IIa  | Common CVN with aplasia/hypoplasia of cochlear nerve | Minor labyrinthine dysplasia to common cavity | 6 ears |
| IIb  | Common CVN with aplasia/hypoplasia of cochlear nerve | Normal labyrinth | 14 ears |
| III  | Common CVN with aplasia/hypoplasia of vestibular nerve | Not reported yet | Nil |

CVN: cochleovestibular nerve

All implantees underwent regular habilitation on a one-to-one basis during biweekly sessions. The evaluation of habilitation was performed at our habilitation center, and those making evaluations were blinded about nerve status of these children.

**Statistical Analysis**

Appropriate statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS) version 20.0 (IBM Corp.; Armonk, NY, USA).

**RESULTS**

A total of 405 CI procedures were performed at our center during the study period for children with bilateral congenital severe to profound hearing loss. Of these 20 children received a diagnosis of CVND. This made a prevalence of CVND among cochlear implantees at our center to be approximately 5. Of the 20 pediatric patients with CVND, 12 children were boys and 8 were girls (M:F ratio of 3:2).

Among children with CVND, 5 (25%) had unilateral involvement, whereas the remaining 15 (75%) had bilateral involvement (i.e., total of 35 ears with CVND). Of the 15 patients with bilateral CVND, 7 had bilateral aplasia of cochleovestibular nerve and the remaining 8 had bilateral hypoplasia of cochlea nerve. Isolated vestibular nerve hypoplasia was not seen in any of our patients.

The distribution and management of CVND in our study is shown in Figure 2.

The CVND is classified by Casselman and Govaerts into types I, II, and III. The distribution of CVND in our study population based on Casselman and Govaerts’ classification is as shown in Table 1 and Figure 2.

The one child with bilateral hypoplastic cochlear nerve (group A) with bilateral cochleovestibular inner ear anomaly (type IIa CVND) who underwent CI developed cerebrospinal fluid gusher, which was successfully managed intraoperatively (waiting for the leak to reduce, intravenous mannitol, dexamethasone, and tissue plug). Postoperative period was uneventful.
The parents of all seven children with bilateral aplasia of cochlear nerve were counseled for ABI; however, only five opted for an ABI. The parents of the remaining 2 children did not opt for surgery because of financial constraints and risk of surgery; they opted for total communication. While the five children with unilateral CND, CI was done on the side with normal anatomy. The management protocol followed is as given in Figure 2.

The objective outcomes of patients in group A and group B (age-matched control) with respect to CAP latency and amplitude are presented in Table 2, whereas the CAP and SIR score are presented in Table 3. Statistical analysis was performed for postoperative outcomes at 12 months between the two groups. Outcomes at 12 months were also compared with preoperative values in group A. Because our data was nonparametric data, Wilcoxon signed-rank test was performed for statistical analysis.

In group A, post-CI CAP and SIR, CAEP amplitude and latency at 12 months showed statistically significant difference (p<0.05) compared with preoperative values. However, the mean score of CAEP latency and amplitude and SIR score was worse for group A compared with that of group B at 12 months, which was statistically significant (p<0.05). CAP score at 12 months was not found to be statistically significant (p=0.059) between the two groups.

DISCUSSION

Cochlear nerve hypoplasia is radiologically defined as a cochlear nerve that is smaller in diameter than the adjacent facial nerve in the mid-portion of IAC [15]. While CVND has been classified by Casselman et al. into three types [15].

The reported incidence of CVND using MRI of the brain and IAC has been reported between 12% and 21.2% [17,18] in children with bilateral to profound hearing loss. Wu et al. [18] used high-resolution MRI and reported an incidence of 21.2% of CVND with bilateral CVND comprising 4.3%, which constituted a fifth of the total children with CVND. However, in our study population of bilateral severe to profound congenital deafness, the overall incidence of CVND was only 5%, of which 75% had bilateral CVND using 3 Tesla MRI. This variation in the incidence of CVND in this study compared with other published studies may be because of the small sample size, ethnic differences in study population, and use of 3 Tesla MRI [16].

The neural responses generated by the electrical impulses delivered from cochlear implant are likely to be limited in patients with CVND. This is turn would limit the neurological activity generated at higher centers along the auditory pathway. Valero et al. [19] studied electrophysiological and behavioral response in cochlear implantees with CVND. On the basis of their outcomes, it was suggested that children with CVND are unsuitable candidates for CI because of poor auditory development and higher incidence of nonauditory activity in his group of children. Several other studies on CI in children with CVND [14,20-23] have suggested that most children with CVND can gain sound awareness from their device, albeit using higher levels of electrical stimulation. However, the achievement of open-set speech perception for purposes of speech and language development has been largely unsuccessful, probably as a result of a poor electrode neural interface and other associated conditions.

In this study of CI in patients with CVND, statistically significant improvement was found in outcomes at 12 months post implantation in terms of CAP, SIR, CAEP amplitude and latency compared with pre-implant children, and none of them reported nonauditory activity such as facial nerve stimulation.

Auditory perception (CAP) at 12 months was comparable in cochlear implantees with CVND and without CVND, but there was statistically significant worse outcome in children with CVND in terms of speech intelligibility and CAEP latency and amplitude. The possible explanation could be that patients with CVND, although able to receive the sound stimulus (comparable CAP), are not able to integrate them centrally, which in turn results in poor speech outcomes and poorer waveform of CAEPs.

A study conducted by Wu et al. [18] suggested that the type of CVND (aplasia/hypoplasia) may affect CI outcomes. Patients who received the implant on the side with cochlear nerve hypoplasia are more likely to exhibit favorable results compared with those who received the

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**Table 3. Habilitation outcomes: CAP and SIR at 12 months post cochlear implantation for group A (cases) and group B (age-matched control)**

| Serial no. | CAP | SIR |
|-----------|-----|-----|
|           | Baseline (group A and B) | 12 months (group A) | 12 months (group B) | Baseline (group A and B) | 12 months (group A) | 12 months (group B) |
| 1         | 1 | 4 | 5 | 1 | 3 | 4 |
| 2         | 0 | 3 | 5 | 1 | 2 | 4 |
| 3         | 2 | 5 | 5 | 2 | 2 | 3 |
| 4         | 1 | 4 | 4 | 1 | 3 | 5 |
| 5         | 2 | 6 | 5 | 1 | 3 | 4 |
| 6         | 1 | 5 | 5 | 2 | 4 | 4 |
| 7         | 1 | 4 | 5 | 1 | 3 | 3 |
| 8         | 1 | 4 | 5 | 1 | 3 | 4 |
| Mean      | 1.125 | 4.375 | 5 | 1.25 | 2.875 | 3.875 |

CAP: category of auditory perception; SIR: speech intelligibility rating.
implanted on the side with cochlear nerve aplasia. No significant difference was noted between the hypoplasia group and the non-CVND group in their study in terms of CAP and SIR scores. As per our institute management protocol (Figure 2) for children with type I CVND (aplasia of cochleovestibular nerve), we did not perform CI in any of the patients.

In a study performed by Vincenti et al. [24] based on the variable outcomes of CI in five children with CVND, ranging from improved awareness to environmental sounds to open-set speech perception and acquisition of spoken language, the study findings suggested that careful counseling of family is essential regarding possibility of limited benefit. The outcomes of our study corroborates with existing literature and firmly agree with others in that the family of the patient needs to be intensively counseled about the possibility of limited benefit, especially with regard to the development of speech intelligibility.

In a study conducted by Colletti et al. [21] in children with CVND, patients fitted with cochlear implants did not develop speech understanding and production. Those fitted with ABIs had the opportunity to develop open-set speech perception, acquiring verbal language competence using oral communication exclusively and participating in mainstream education. However, the results of our study show that CI in patients with type II CVND had good auditory perception and satisfactory speech intelligibility.

**Limitations of the Study**

There are two major limitations of this study. First, our outcome data cannot be generalized in view of the small sample size. Second, because long-term outcome data are lacking in this study, we cannot comment whether these children are able to catch up with cochlear implantees without CVND.

**CONCLUSION**

CVND is an uncommon entity seen in congenital hearing loss that a neuro-otologist will encounter. The diagnosis can be easily made with high resolution MRI. CVND is a gray zone when it comes to choosing the treatment option. Our study supports the fact that CI is a viable option to be offered to this group (type Ila and type Ilb) of children for the development of auditory perception and speech with a guarded prognosis. Parents need to be adequately counseled about the possibility of no benefit or limited benefit. We suggest that CI should be the primary modality of surgical treatment in these patients, especially in view of the increased financial burden and higher complication rate with ABI. In addition, ABI should be considered as an option where CI is not providing satisfactory benefit both in terms of audition and speech development.

**Ethics Committee Approval**: Ethics committee approval was received from the Ethics Committee of MERF, Chennai, Tamil Nadu India.

**Informed Consent**: Written informed consent was obtained from the patients who participated in this study.

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**REFERENCES**

1. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kites, Shelton C. Imaging findings of cochlear nerve deficiency. Am J Neuroradiol 2002; 23: 635-43.

2. Adunka OF, Roush PA, Teagle HFB, Brown CJ, Zdanski CJ, Jewels V, et al. Internal auditory canal morphology in children with cochlear nerve deficiency. Otol Neurotol 2006; 27: 793-801. [Crossref]

3. Adunka OF, Jewels V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. Otol Neurotol 2007; 28: 597-604. [Crossref]

4. Buchman CA, Roush PA, Teagle HFB, Brown CJ, Zdanski CJ, Gross JH. Auditory neuropathy characteristics in children with cochlear nerve deficiency. Ear Hear 2006; 27: 399-408. [Crossref]

5. Ponton CW, Eggermont JJ, Kwong B, Manuel Don. Maturation of human central auditory system activity: evidence from multi-channel evoked potentials. Clin Neurophysiol 2000; 111: 220-36. [Crossref]

6. Wunderlich JL, Cone-Wesson BK, Shepherd R. Maturation of the cortical auditory evoked potential in infants and young children. Hear Res 2006; 212: 185-202. [Crossref]

7. Ponton C, Eggermont JJ, Khosla D, Kwong B, Don M. Maturation of human central auditory system activity: separating auditory evoked potentials by dipole modeling. Clin Neurophysiol 2002; 113: 407-20. [Crossref]

8. Sharma A, Dormann MF, Spahr AJ. A sensitive period for the development of the central auditory system in children with cochlear implant: implications for age of implantation. Ear Hear 2002; 23: 532-9. [Crossref]

9. Ponton CW, Eggermont JJ. Of kittens and kids: altered cortical maturation following profound deafness and cochlear implant use. Audiol Neurootol 2001; 6: 363-80. [Crossref]

10. Gordon KA, Tanaka S, Wong DDE, Papsin BC. Characterizing responses from auditory cortex in young people with several years of cochlear implant experience. Clin Neurophysiol 2008; 119: 2347-62. [Crossref]

11. Gordon KA, Tanaka S, Wong DDE, Stockley T, Ramsden JD, Brown T, et al. Multiple effects of childhood deafness on cortical activity in children receiving bilateral cochlear implants simultaneously. Clin Neurophysiol 2011; 122: 823-33. [Crossref]

12. Archbold S, Lutman M, Marshall D. Categories of Auditory Performance. Ann Otol Rhinol Laryngol Suppl 1995; 166: 312-4.

13. Archbold S, Lutman ME, Nikolopoulos T. Categories of auditory performance: inter-user reliability. Br J Audiol 1998; 32: 7-12. [Crossref]

14. Allen MC, Nikolopoulos TP, O’Donoghue GM. Speech Intelligibility in Children After Cochlear Implantation. Otol Neurotol 1998; 19: 742-6. [Crossref]

15. Govaerts PJ, Casselman J, Daemers K, DeBeukelaer C, Yperman M, DeCeulaer G. Cochlear Implants in Aplasia and Hypoplasia of the Cochleovestibular Nerve. Otol Neurotol 2003; 24: 887-91. [Crossref]

16. Kutz Jr JW, Lee KH, Isaacson B, Booth TN, Sweeney MH, Roland PS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotrol 2011; 32: 956-61. [Crossref]

17. Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. Otol Neurotrol 2005; 26: 976-82. [Crossref]

18. Wu CM, Lee LA, Chen CK, Chan KC, Tsou YT, Ng SH. Impact of cochlear nerve deficiency determined using 3-dimensional magnetic resonance imaging on hearing outcome in children with cochlear implants. Otol Neurotrol 2015; 36: 14-21. [Crossref]

19. Valero J, Blaser S, Papsin BC, James AL, Gordon KA. Electrophysiologic and behavioral outcomes of cochlear implantation in children with auditory nerve hypoplasia. Ear Hear 2012; 33: 3-18. [Crossref]
20. He S, Grose J, Hang AX, Buchman CA. Cochlear Implant-Evoked Cortical Activation in Children with Cochlear Nerve Deficiency. Otol Neurotol 2012; 33: 1188-96. [Crossref]

21. Buchman CA, Teagle HF, Roush PA, Park LR, Hatch D, Woodard J, Zdanski C, Adunka OF. Cochlear implantation in children with labyrinthine anomalies and cochlear nerve deficiency: implications for auditory brainstem implantation. Laryngoscope 2011; 121: 1979-88. [Crossref]

22. Kutz JW Jr, Lee KH, Isaacson B, Booth TN, Sweeney MH, Roland PS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotol 2011; 32: 956-61. [Crossref]

23. Warren FM 3rd, Wiggins RH 3rd, Pitt C, Hamsberger HR, Shelton C. Apparent cochlear nerve aplasia: to implant or not to implant Otol Neurotol 2010; 31: 1088-94. [Crossref]

24. Vincenti V, Ormitti F, Ventura E, Guida M, Piccinini A, Pasanisi E. Cochlear implantation in children with cochlear nerve deficiency. Int J Pediatr Otorhinolaryngol 2014; 78: 912-7. [Crossref]

25. Colletti L, Colletti G, Mandalà M, Colletti V. The Therapeutic Dilemma of Cochlear Nerve Deficiency: Cochlear or Brainstem Implantation. Otolaryngol Head Neck Surg 2014; 151: 308-14. [Crossref]