Audiologic Outcome of Cochlear Implantation in Children With Cochlear Nerve Deficiency

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ABSTRACT

Objective/Hypothesis: The aim of this study was to investigate cochlear implantation (CI) outcome in children with nerve deficiency.

Study Design: Retrospective chart review.

Methods: A total of seven children with prelingual profound deficiency (hypoplasia or aplasia) were included. A control group of 10 CI children with no cochlear nerve anomalies was also included. In addition to implant stimulation levels, children's performance on pure-tone audiometry, speech reception measure, and auditory and speech skills ratings were compared across groups. Additionally, pre- and postoperative audiologic results were evaluated for the group with nerve deficiency.

Results: In general, children with nerve deficiency performed poorer than those without nerve deficiency on all tested measures. Stimulation levels were considerably higher and more variable than the control group. Results further showed that performance was dependent on the diameter of the internal auditory canal.

Conclusion: Overall, cochlear implantation outcome in children with auditory nerve deficiency is poorer and extremely more variable than those without nerve deficiency. However, three of the patients had a noticeable improvement in auditory performance post implantation suggesting that CI is a viable option in this population but expected benefit can be dependent on the status of the cochlear nerve.

INTRODUCTION

Cochlear nerve deficiency is one of the known causes of congenital sensorineural deafness.[1,2] The prevalence of cochlear nerve deficiency has been reported to be as high as 18% in pediatric patients diagnosed with congenital sensorineural hearing loss.[3] It is generally agreed that a cochlear nerve is considered hypoplastic if it is smaller in diameter than that of the facial nerve in the midportion of the internal auditory canal or aplastic if it is absent as confirmed by imaging findings.[2,4]

Management of hearing loss in children with cochlear nerve deficiency poses a multidimensional challenge. Cochlear nerve deficiency was considered as a contraindication for CI.[5] On the other hand, some implant clinic did not consider it as an absolute contraindication for CI. This was justified based on the limited MRI resolution which may not be sufficient to accurately visualize all hypoplasia cases in addition to the fact that some patients with auditory nerve aplasia responded to electrical stimulation when implanted with a CI.[6,7] The other implant option for rehabilitation of these patients is auditory brainstem implant which is more invasive, critical, expensive, and not available in all implant centers.

During the last two decades more patients with cochlear nerve deficiency have undergone cochlear implantation. However, the reported audiologic outcome in children diagnosed with cochlear nerve deficiency is quite variable with some showing successful implantation outcome[7,8] and others demonstrated poor outcome in this population.[9,10] Therefore, the aim of this study was to investigate the efficacy of CI in patients with cochlear nerve deficiency.

METHODS

Subjects

A retrospective chart review design was used. In order to be included in this study, all children were required to meet the following criteria: 1) Birman grade 0- 4; 2) had a pre-lingual onset of deafness; 3) had severe to profound sensorineural hearing loss; 4) used a hearing aid for a period ranged between 3 and 6 months before implantation; 5) had a minimum of one year of CI use at the time of data collection; 6) underwent rehabilitation for at least one year using auditory-verbal therapy; and 7) had preoperative CT and MRI with documented diagnosis of cochlear nerve deficiency. Those who do not meet these criteria were excluded.

A total of 7 participants (2 males and 5 females) were included (Table 1). A control group of 10 CI children (7 males and 3 females) with no cochlear nerve anomalies were also included. The control group was matched to the study group in terms of implantation age and CI use. All children were implanted in our tertiary CI center using MED-EL SYNCHRONY device (MED-EL, Innsbruck, Austria).

Subject	Gender	Age	Ear	CI use	Cochlear nerve	Nerve grading	Cochlear aperture diameter	IAC diameter	Ear anomaly
1	F	3	Left	1.5	Hypoplastic	I-V	1.2	4.68	None
2	F	7	Left	2.5	Hypoplastic	I-V	1.9	4.79	None
3	M	3	Left	2	Aplastic	II	Absent	5	Absent semicircular canal & dilated vestibule
4	F	6	Left	2.5	Aplastic	II	1	2.3	Absent superior semicircular canal & fused vestibule with lateral semicircular canal
5	M	3	Right	2.5	Aplastic	0	Absent	1.8	None
6	F	5	Left	2.5	Aplastic	I	0.66	2.62	None
7	F	4	Right	2	Aplastic	II-III	Absent	1.9	Absent posterior semicircular canal & enlarged vestibular aqueduct

Assessment

Based on radiologic evidence, all included patients were confirmed with cochlear nerve aplasia or hypoplasia according to the IAC nerve grading system described by Birman et al.[11] The evaluation of nerves within the IAC was performed with the reconstructed parasagittal MR images. Figure 1 shows MRI findings of patient number 6 with cochlear nerve aplasia (a) and patient number 2 with cochlear nerve hypoplasia (b).

Generally, preoperative audiological assessments consisted of objective measures such as auditory brainstem responses (ABR), and otoacoustic emissions (OAEs). All subjects in both groups revealed absent ABR and OAE responses reflecting bilateral severe to profound SNHL. Preoperative assessment further included behavioral audiologic measures such as pure tone audiometry using visual reinforcement audiometry (VRA) or play audiometry, and speech reception thresholds (SRTs) or speech awareness threshold (SAT). Post-operative audiologic assessment included PTA and SRTs/SATs in free field. Additionally, in order to assess patients' performance with their devices in everyday auditory environment, categorical scales were administered by a professional speech therapist. These scales included the administration of the Meaningful Auditory Integration Scale (MAIS) to assess the auditory skills of children and the Meaningful Use of Speech Scale (MUSS) to evaluate children's verbal ability in everyday situations.[12,13]

RESULTS

Subject	500 Hz		1000 Hz		2000 Hz		4000 Hz		SATs/SRT	
	Preop.	Postop.	Preop.	Postop.	Preop.	Postop.	Preop.	Postop.	Preop.	Postop.
1	75	35	80	40	NR	45	NR	40	75	25/40
2	80	35	NR	30	NR	30	NR	30	80	15/25
3	75	50	NR	40	NR	40	NR	45	75	35
4	80	70	85	80	80	NR	90	NR	85	70
5	80	NR	80	NR	NR	NR	NR	NR	80	NR
6	NR	70	NR	NR	NR	NR	NR	NR	NR	70
7	85	80	80	75	115	NR	NR	NR	75	70

TABLE 2. Comparison of preoperative and aided postoperative (using CI processors) audiologic measures. NR indicates no response; SAT, speech awareness threshold; CI, cochlear implantation.

- Audiological benefit has been found in subjects 1, 2 (hypoplasia group). They showed comparable results as regarding aided hearing threshold and SRT.
- Subject 3 showed relative improvement in comparison to the other 4 subjects in the aplasia group.
- There was a significant improvement in threshold levels for speech perception as a result of cochlear implantation and there was no significant effect of group on the results (p> 0.05).

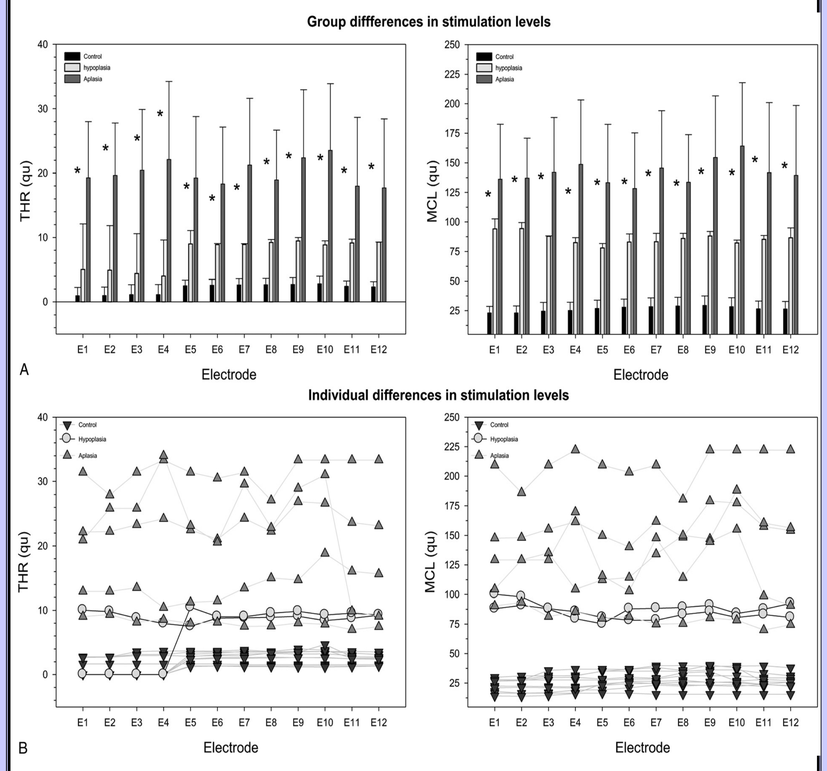
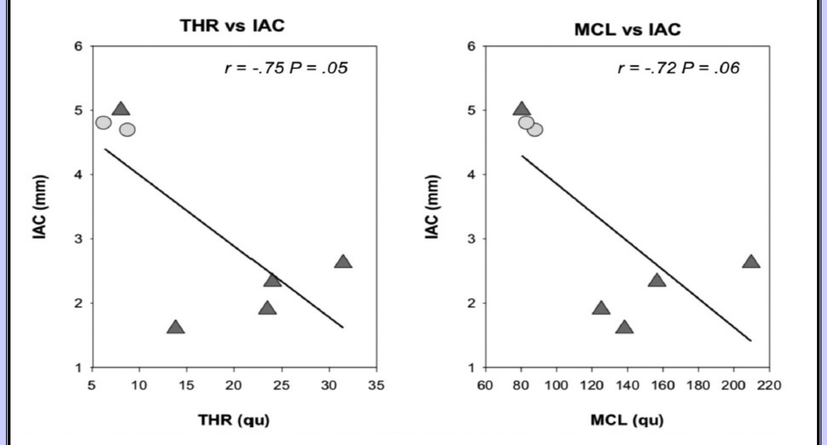


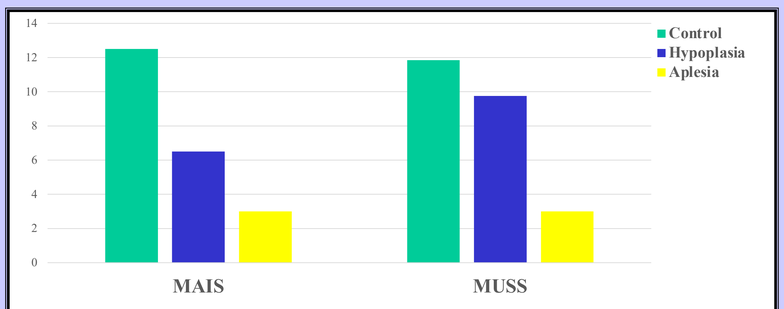
FIG. 3. Average stimulation levels for THR (left panel) and MCL (right panel) are shown for three groups of patients. Individual stimulation levels are also shown for THR and MCL in lower panels. THR indicates thresholds; MCL, maximum comfortable level; SD,standard deviation.

- Results revealed a significantly higher stimulation levels for the aplasia in comparison to the control group (P<.01).
- On the other hand, stimulation levels for the hypoplasia group were not significantly different than those obtained in either group (P>.05).
- The group with nerve deficiency exhibited a large variability in stimulation levels in comparison to that in the control group.



- There was a significant negative association between THR and the diameter of the IAC such that larger IAC diameter resulted in lower thresholds (r = -.75, n=7, p = .05).
- This relationship was not found significant for MCL (r = -.72, n=7, p = .06) but a similar trend can be seen.

RESULTS



Average scores on MAIS and MUSS scales are compared for the three groups of patients in panel. MAIS indicates Meaningful Auditory Integration Scale; MUSS, Meaningful Use of Speech Scale; SD, standard deviation.

- The control group achieved significantly better scores on both measures than those obtained in the aplasia group (P<.005) but their scores were comparable to those obtained in the hypoplasia group.
- There were no significant differences in performance between the hypoplasia and the aplasia groups (P> .05).

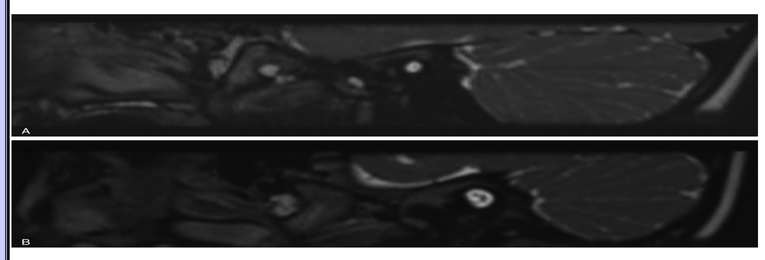


FIG. 1. MRI of subject #6 with cochlear nerve aplasia showing one nerve inside left IAC (A) and MRI of subject #2 with cochlear nerve hypoplasia showing only two nerves inside left IAC (B). MRI indicates magnetic resonance imaging; IAC, internal auditory canal.

SUMMARY and CONCLUSIONS

- Patients with cochlear nerve deficiency had generally poorer implantation outcome than that observed in the control group.
- Outcomes were extremely variable and more dependent on the status of the cochlear nerve.
- Three of the patients had a noticeable improvement in auditory performance post implantation suggesting that CI is a viable option in this population.
- Stimulation levels and SATs were found to be related to the diameter of the IAC; this perhaps suggest that the diameter of the IAC may serve as a prognostic factor in determining the extent of implantation outcome

REFERENCES

1. Casselman JW, Offeciers FE, Govaerts PJ, et al. Aplasia and hypoplasia of the vestibulocochlear nerve: Diagnosis with MR imaging. Radiology 1997;202:773–81.
2. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency.AJNR Am J Neuroradiol 2002;23:635–43.
3. McClay JE, Booth TN, Parry DA, Johnson R, Roland P. Evaluation of pediatric sensorineural hearing loss with magnetic resonance imaging. Arch Otolaryngol Head Neck Surg 2008;134:945–52.
4. Kutz JW Jr, Lee KH, Isaacson B, Booth TN, Sweeney MHPS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotol 2011;32:956–61.
5. Shelton C, Luxford WM, Tonokawa LL. The narrow internal auditory canal in children: A contraindication to cochlear implants.Otolaryngol Head Neck Surg 1989;100:227–31.
6. Thai-Van H, Fraysse B, Berry I, et al. Functional magnetic resonance imaging may avoid misdiagnosis of cochleovestibular nerve aplasia in congenital deafness. Am J Otol 2000;21:663–70.
7. Acker T, Mathur NN, Savy L, Graham JM. Is there a functioning vestibulocochlear nerve? Cochlear implantation in a child with symmetrical auditory findings but asymmetric imaging. Int J Pediatr Otorhinolaryngol 2001;57:171–6.
8. Song MH, Bae MR, Kim HN. Value of intracochlear electrically evoked auditory brainstem response after cochlear implantation in patients with narrow internal auditory canal. Laryngoscope 2010;120:1625–31.
9. Bradley J, Beale T, Graham J, Bell M. Variable long-term outcomes from cochlear implantation in children with hypoplastic auditory nerves. Cochlear Implants Int 2008;9:34–60.
10. 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.
11. Birman CS, Powell HR, Gibson WP, Elliott EJ. Cochlear implant outcomes in cochlea nerve aplasia and hypoplasia. Otol Neurotol 2016;37:438–45.
12. Robbins AM, Renshaw JJ, Berry SW. Evaluating meaningful auditory Integration in profoundly hearing-impaired children. AmJ Otol 1991;12 (suppl):144–50.
13. Robbins AM, Osberger MJ. Meaningful Use of Speech Scale (MUSS). Atlanta, GA: American Speech-Language and Hearing Association; 1991.

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