The Journal of International Advanced Otology

J Int Adv Otol 2021; 17(1): 19–22 • DOI: 10.5152/iao.2020.8983

Original Article

Audiological and Surgical Outcomes of Pediatric Cochlear Implantation in Mondini's Dysplasia: Our Experience

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Cite this article as: Kumari A, Arumugam SV, Malik V, Goyal S, Kameswaran M. Audiological and Surgical Outcomes of Pediatric Cochlear Implantation in Mondini's Dysplasia: Our Experience. J Int Adv Otol 2021; 17(1): 19-22.

OBJECTIVES: Aim of present study is to compare audiological and surgical outcomes in prelingual deaf children with Mondini's dysplasia (MD) and those with normal inner ear anatomy.

MATERIALS and METHODS: Retrospective data was collected from Jan 2008 to Dec 2016. Children with bony IEM other than MD, syndromic association, multiple disabilities, those lost to follow up, and perilingual or postlingual deafness were excluded from study. Audiological outcomes for auditory perception (CAP score) and speech intelligibility (SIR score) was noted for a follow up period of 1 year.

RESULTS: Mean age at implantation was 2.8 years (Range of 2 to 6 years). 2 patients had intraoperative CSF ooze which was controlled intraoperatively by conservative measures. Post operative facial nerve function was normal in all patients. None of the patient in either group had any complications at one year of follow up period. There was statistically significant improvement in CAP & SIR score in Group A at 6 & 12 months compared to pretreatment. There was no statistically significant difference between the 2 groups in terms of CAP & SIR score at 6 & 12 months.

CONCLUSION: The study stresses the fact that cochlear implantation can be safely performed in children with MD although there is a risk of intraoperative CSF leak which can be controlled intraoperatively. Cochlear implantation in children with MD has good surgical, auditory and speech outcomes at par with children with normal bony inner ear anatomy.

KEYWORDS: Prelingual deafness, category of auditory perception, speech intelligibility rating, objective outcomes, cerebrospinal fluid leak, facial nerve trauma

INTRODUCTION

Approximately 20% cases of sensorineural hearing loss (SNHL) have associated inner ear malformation (IEM) [1]. Advances in imaging have significantly contributed to changes in management of congenital hearing loss and associated IEM in children. In its infancy, cochlear implantation (CI) was contraindicated in cases with IEM. The first cochlear implant in a patient with IEM was performed successfully in 1983 for Mondini's dysplasia (MD) [2].

Mondini's dysplasis is the most common IEM and results from arrest of development in 7th week of gestation. It is presently classified as incomplete partition type 2 (IP-2) and is characterized by 1.5 turns of cochlea with normal basal turn, cystic apex due to fused middle & apical turns, enlarged vestibular aqueduct, and dilated vestibule [3-5]. As the basal turn is normal with intact modiolus and spiral ganglion, results of CI can be expected to be at par with normal cochlea [5]. Several studies have shown good outcome of CI in these subgroup of patients [6-13]. This study aims to compare audiological and surgical outcomes in prelingual deaf children with MD and those with normal inner ear anatomy in a group of children who got implanted in a tertiary care cochlear implant center in India.

MATERIALS AND METHODS

A retrospective study was conducted at a tertiary care hospital in southern India. The data were collected over a period of 6 years (from Jan 2011 to Dec 2016) from hospital medical record database. Institutional research ethics board approval was obtained. Chil-

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Submitted: 06.18.2020 • **Revision Received:** 07.28.2020 • **Accepted:** 08.24.2020
Available online at www.advancedotology.org

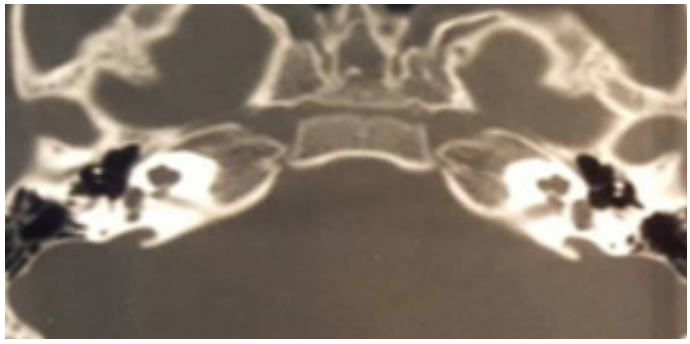

Figure 1. Axial section HRCT temporal bone of a child with Mondini dysplasia showing cystic apical turn and enlarged vestibular aqueduct.

dren with prelingual deafness with MD and normal inner ear anatomy were included in the study. Children with other bony IEM, syndromic association, multiple disabilities, those lost to follow-up, and perilingual or postlingual deafness were excluded from the study.

Inner ear anatomy was diagnosed based on radiological findings that included magnetic resonance imaging (MRI) of brain and inner ear and high-resolution computed tomography (HRCT) scan of temporal bone. Figure 1 shows MD in imaging. Children with MD were included in Group A. Controls were selected from age and sex matched children with profound hearing loss and normal inner ear anatomy and included in Group B.

Intraoperative surgical details and complications were noted. All patients underwent standard transmastoid posterior tympanotomy approach for CI. Multichannel straight electrodes from the same company (MED EL, Austria) were used, and in all cases, the insertion of electrodes was achieved via round window or cochleostomy. Intraoperative impedance and evoked cochlear action potentials (eCAP) were measured. Any Cerebrospinal fluid (CSF) ooze was managed and controlled intraoperatively by conservative measures including wait and watch, iv dexamethasone, hyperventilation, and tissue seal and tissue glue to plug the opening into cochlea after electrode insertion.

The switch on and mapping was done 3 weeks after the surgery. The same model of speech processor (OPUS 1) was used in all the cases. Mapping was repeated at 1, 3, 6, and 12 months and when patient reported for troubleshooting. The post-operative habilitation programme consisted of intensive auditory verbal habilitation therapy (AVHT) with at least 2 therapy sessions per week at our center by an expert team of speech language pathologist and auditory habilitationalist. Duration of post-operative AVHT given to all the children in both the groups was a minimum period of 1 year. Parents were also effectively trained to help the child at home.

Category of Auditory Perception (CAP) rating and Speech Intelligibility Rating (SIR) are used to record subjective outcomes in implanted children at our center. Where CAP is a hierarchical scale of auditory perceptive ability ranging from 0 (i.e., no awareness of environmental sounds) to 7 (i.e., can use the telephone with a familiar talker). SIR consists of five performance categories ranging from "pre recognizable words in spoken language" to "connected speech is intelligible to all listeners". The audiological progress for audition (CAP) and SIR were recorded at 6-month intervals (6 and 12 months).

Statistical Analysis

Appropriate statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS) version 20.0 (IBM Corp.; Armonk, NY, USA). As the data were nonparametric and qualitative in nature, Friedman X^2 test and Mann Whitney U test were applied for analysis.

RESULTS

Mean age at implantation was 2.8 years with range of 2 to 6 years. Male to female ratio was 3:2 in both the groups as our groups were age and sex matched.

Intraoperative impedance and auditory response telemetry (ART) were satisfactory for all cases. Two patients had intraoperative CSF ooze which was managed and controlled intraoperatively by conservative measures. Both the patients belonged to Group A**.** Post-operative facial nerve function was normal in all patients. None of the patient in either group had any post-operative complications at one year of follow-up period.

The auditory and speech outcomes 6 months and 12 months after CI are given in Table 1.

The mean CAP score in Group A increased from 0.9 to 3.3 to 4.3 at 0, 6, and 12 months, respectively. In contrast, in Group B, it increased from 0.9 to 4.3 to 5.2 over the same period. The mean SIR score in Group A increased from 1 to 1.3 to 2.1 at 0, 6, and 12 months, respectively, whereas in Group B, it increased from 1 to 1.7 to 2.4 over the same period.

All patients in both groups showed progressive improvement in CAP and SIR score, except 2 patients in Group A, where the speech intelligibility rating did not change even at 12 months (patient number 7 and 8 in Group A) although there was improvement in auditory perception. As the data were nonparametric and qualitative in nature, Friedman χ^2 test and Mann Whitney U test were applied as given in Table 2.

Pretreatment CAP and SIR scores were compared with scores at 6 months and 12 months in Group A. In addition, CAP and SIR scores at 6 months and 12 months were compared between the two Groups**.** A statistically significant improvement in CAP and SIR scores was observed in Group A at 6 months and 12 months compared to pretreatment. However, there was no statistically significant difference between the 2 groups in CAP and SIR scores at 6 months and 12 months follow-up period.

MAIN POINTS

- Cochlear implantation can be safely performed in Mondini dysplasia with low complication rate.
- There is increased risk of CSF leak in the perioperative period which can usually be controlled intraoperatively.
- The auditory perception and speech development outcomes are at par in prelingual deaf children with normal bony inner ear anatomy.

Table 1. Audiological outcomes in terms of CAP (audition) and SIR (speech intelligibility) in Group A and B

	Category of Auditory Perception (CAP)					Speech Intelligibility Rating (SIR)				
	Baseline	Group A		Group B (Control)		Baseline	Group A		Group B (Control)	
Serial number	0 month	6 m	12 m	6 m	12 m	0 month	6 m	12 m	6 m	12 m
1	1	4	5	4	5	1	1	3	2	3
2	1	4	5	4	5	1	1	2	1	2
3	0	2	5	3	5	1	2	2	2	3
4	1	2	2	3	5	1	1	2	2	2
5	1	4	5	4	5	1	1	3	2	2
6	1	4	5	3	5	1	1	2	2	2
7	1	4	5	2	4	1	1	1	1	2
8	0	2	2	4	6	1	1	1	2	3
9	1	4	5	3	6	1	2	2	1	2
10	2	3	4	3	6	1	2	3	2	3
Median	1	4	5	3	5	1	1	2	2	2
Mean	0.9	3.3	4.3	3.3	5.2	1	1.3	2.1	1.7	2.4
SD	0.56	0.94	1.25	0.67	0.63	0	0.48	0.73	0.48	0.51

Table 2. Statistical comparison of Category of auditory perception (CAP) and Speech intelligibility rating (SIR) between the 2 groups

Group A (n=10)		Group A Vs Group B (n=10)	
Outcome measures	**Friedman χ^2**	**Outcome measure**	**Mann Whitney U**
CAP 0 month, 6 months, and 12 months	18.2; p<0.05	CAP at 6 months	U: 47; p>0.05
		CAP at 12 months	U: 28.5; p>0.05
SIR 0 month, 6 months, and 12 months	8.15; p<0.05	SIR at 6 months	U: 30; p>0.05
		SIR at 12 months	U: 39; p>0.05

DISCUSSION

The practice of universal neonatal hearing screening, advances in hearing assessment, and imaging have facilitated diagnosis of congenital hearing loss at an early age, and hence, early CI is performed when indicated. As a result, today, we are able to identify congenital bony IEM including MD at an early age and therefore manage them early. The mean age of CI in this study was 2.8 years in both groups.

This study overviews surgical and audiological outcomes in prelingual deaf children with MD compared with a similar group of age and sex matched children with prelingual deafness with normal inner ear anatomy at a tertiary care referral center in South India.

With regard to facial nerve course, none of our patient had an abnormal facial nerve course in either group, and we were able to perform CI via standard transmastoid posterior tympanotomy approach. A recent radiological study of facial nerve anomalies in pediatric cochlear implant candidates reported 7% incidence of abnormal facial nerve course involving mastoid segment [14]. In another study, it was observed that aberrant shape or course involving vertical segment of facial nerve was seen in 0.7% of candidates during pediatric CI [15]. Both the studies mentioned that all cases of abnormal facial nerve course were associated with inner ear malformation [14, 15]. In a radiological study by Romo and Curtin [16] in 2001, it was revealed that abnormal first segment of facial nerve course was associated with cochlear abnormalities of non-MD variety whereas those with MD had a normal facial nerve course. In this study, of the 10 cases of MD, no variation in facial nerve course was noted radiologically or surgically.

The incidence of facial nerve palsy without the use of facial nerve monitor in CI via transmastoid posterior tympanotomy approach has been reported to range from 0.71% to 2.1% [17,18]. No post-operative facial nerve palsy was observed in our series of 10 patients in each group.

Out of the 10 patients, 2 patients in Group A had CSF ooze that was managed and controlled intraoperatively by conservative measures. Two types of CSF leaks have been described. Ooze is a gentle flow of clear CSF fluid whereas gusher is a profuse flow. CSF oozing may be encountered in cases with a minor defect (e.g., between IAC and malformed inner ear) and is easily controlled with conservative measures [19,20]. CSF gusher rarely occurs in cases with MD [21]. None of the patients in this study had CSF gusher.

The results of this study show that auditory perception CAP and SIR scores show a statistically significant improvement post CI; the scores

are at par with CI in children with normal inner ear bony anatomy. Our result are similar to audiological outcomes in the existing literature for children with MD or inner ear anomalies [22,23].

According to Otte et al. [24], there are approximately 36,000 spiral ganglion cells in normal cochlea during first decade of life. A minimum of 10,000 of these ganglion cells (with at least 3000 at apical 10 mm of the organ of Corti) are required for speech discrimination. Spiral ganglion cells are the main neural elements that are electrically stimulated by the cochlear implant, and the number of spiral ganglion cells required for electrical stimulation through cochlear implant is thought to be much less than that required for acoustic stimulation [25, 26]. This may explain why auditory and speech outcome were not significantly different between the two groups in our study.

The limitation of this study was the small sample size and relatively shorter follow-up period. A longer follow-up period would provide further insight into the treatment benefit.

CONCLUSION

This study stresses the fact that although there is a risk of intraoperative CSF leak, CI can be safely performed in children with MD as the CSF leak can be controlled intraoperatively. CI in children with MD has good surgical, auditory, and speech outcomes, which are at par with children with normal bony inner ear anatomy.

Ethics Committee Approval: Ethical committee approval was received for this study from the Ethics Committee of Madras ENT Research Foundation, Chennai, India.

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

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.K., A.K.; Design – A.K., S.G.; Supervision – A.K., S.V.A.; Resource – S.V.A.; Materials – S.V.A., V.M.; Data Collection and/or Processing – A.K., S.V.A.; Analysis and/or Interpretation – A.K., S.G.; Literature Search – A.K., V.M.; Writing – A.K., S.G.; Critical Reviews – M.K.

Acknowledgements: MERFISH, Chennai, India for all the data and Dr Vipra Mangla, BHDC for all the help with the statistical analysis.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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