Original Research Article

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20174338

Cochlear implant in patients with incomplete partition type II: surgical considerations

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Received: 04 August 2017 **Revised:** 02 September 2017 **Accepted:** 04 September 2017

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ABSTRACT

Background: This study presents our experience on surgical aspects, complications and outcome of cochlear implants in incomplete partition type II patients.

Methods: 7 children in the age group of 1 to 5 years with incomplete partition type II, 5 females and 2 males were studied retrospectively from the database of tertiary care centre from February 2015 to December 2016.

Results: 5 patients had conventional transmastoid facial recess approach of which one patient had removal of incudal buttress, incus and stapes suprastructure to facilitate middle turn cochleostomy. 2 patients had subtotal petrosectomy. Complete insertion of electrode was achieved in all patients; by middle turn cochleostomy in 1 patient, cochleostomy in another patient, extended round window in 3 patients and round window in 2 patients. Med-el Synchrony + Form 19 were used in all patients and complete insertion was achieved in all. 4 patients had CSF leak which was sealed with soft tissue. One patient had device failure. 5 children show improvement in CAP and SIR score. One had device failure and was subsequently explanted and re-implanted.

Conclusions: Cochlear implantation in patients with incomplete partition type II is challenging and thorough knowledge of anatomy is essential. One must be competent enough to expedite different surgical approaches and manage the associated complications. Prior counselling for guarded outcome and eventualities are necessary in such patients.

Keywords: Cochlear implant, Incomplete partition, Subtotal petrosectomy, Round window, Cochleostomy

INTRODUCTION

Hearing loss is a common birth defect. One of every 500 newborns have bilateral permanent sensorineural hearing loss 40dB or greater and by adolescence the prevalence increases to 3.5 out of 1000.¹

Carlo Mondini first reported a young deaf boy with 1.5 turns of cochlea and dilated vestibular aqueduct.² Jackler and colleagues in 1987 formulated the classification of

cochlea-vestibular anomalies based on polytomes, CT scans and embryogenesis.³ Recently, Sennaroglu and Saatci in 2002 described the two types of incomplete partitions (IP) which were further extended to include X-linked deafness as IP III. IP type I is characterized by cystic cochlea with absent modiolus and interscalar septum and IP type II is similar to Mondini's malformation with one and half turn of cochlea and enlarged vestibular aqueduct. In IP type III, there is absence of modiolus with presence of interscalar septa.

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There is associated enlarged vestibular aqueduct and deficient bony partition with internal auditory meatus (IAM). 4-6

The first reported cochlear implantation on malformed cochlea was by Pedro Luiz Mangabeira Albernax in 1983. The 22-year-old patient was diagnosed as Mondini dysplasia.⁷

Since then many surgeons had reported cochlear implantations in malformed cochlea with their experience of the challenges faced during these procedures. Cochlear implantation in malformed cochlea poses risk of CSF leak, abnormal course of facial nerve, round window abnormalities and difficult electrode insertions. 8-10

We present our experience on surgical aspects of cochlear implants in incomplete partition type II patients.

METHODS

7 children in the age group of 1 to 5 years with incomplete partition type II, 5 females and 2 males were studied retrospectively from the database of tertiary care centre from February 2015 to December 2016. The patients were evaluated for bilateral severe to profound hearing loss with delayed speech and language development detected during routine health camps and screening programs conducted by the Institute.

Inclusion criteria included children aged 5 years or less with bilateral profound hearing loss and incomplete partition type II on imaging. Children aged more than 5 years and children with normal cochlea were excluded from the study. Children with syndromic deafness associated with incomplete partition type II on imaging were also excluded from the study.

Audiologic evaluation of the children comprised of age appropriate behavioural audiometry, aided audiometry, Impedance audiometry, oto-acoustic emission and Brainstem evoked response audiometry. CT scan temporal bone and MRI of inner ear and brain were performed in all children.

All the CI candidates were thoroughly evaluated by a multi-specialty team comprising of cochlear implant surgeon, paediatrician, cardiologist, ophthalmologist, audiologist, speech and language therapist and child psychologist. The children were provided hearing aids for 6 months prior to the surgery and were regularly followed up for improvement in hearing and speech outcome. The approval for cochlear implant was granted after second level of screening from state board of CI surgeons. The cochlear implants were conducted under the Tamil Nadu state Chief Minister Comprehensive health insurance scheme at Madras ENT Research Foundation, Chennai.

All the children were routinely evaluated for complete haemogram, thyroid function test, kidney function test,

liver function test, urine routine examination, bleeding time, clotting time, ECG, Chest X-ray, serology for hepatitis B, Hepatitis C and HIV. Impedance audiometry was repeated one day prior to the surgery. The surgery was conducted after receiving evaluation and approval from consultant anaesthetist. The surgery was done by senior consultant CI surgeon of the Institute.

The records were maintained in the hospital database in excel format which were analysed. Based on US Department of Health and Human Services criteria 45 CFR 46.101(b4), retrospective study such as ours is qualified for exemption from an institutional review board.

Surgical technique

Retro auricular lazy S incision was employed in all the patients. Anteriorly based palva flap was raised and transmastoid facial recess approach or subtotal petrosectomy with blind sac closure was used depending on the condition of the intraoperative findings. Round window niche was graded according to visibility based on St. Thomas hospital. The Receiver / stimulator of the implant were fixed with permanent prolene sutures to tie down holes after preparing implant bed. Medel Synchrony+Form 19 were used in all the patients. This is a compressed electrode with proximal bulb that has the advantage of providing seal to the CSF leaks. Electrode insertion was by round window, extended round window or cochleostomy or middle turn cochleostomy depending on the visibility and ossification of the round window niche. Soft tissue seal was used in all the patients. Number of electrodes inserted was recorded and intra operative impedance and Auditory response telemetry was checked. The wound was closed in two layers.

The patients were discharged after 2 days and routinely followed up on 10th day for suture removal. Switch on was done on 21st day and all had compulsory 1 year habilitation at Madras ENT Research Foundation Institute of Speech and Hearing centre.

The outcome in terms of Aided audiogram, CAP score and SIR score was recorded on 3rd, 6th and 12th month of habilitation.

RESULTS

7 children, 2 males and 5 females, from 1 to 5 years with mean age of 3.2 years were evaluated. All the children had speech and language delay, prior to implantation and the mode of communication was by signing. There was no improvement after use of hearing aid for 6 months.

1st patient had an attempted failed cochlear implantation elsewhere. 2nd patient had history of 2nd degree consanguinity with history of chicken pox during infancy and 4th child had history of preterm delivery by caesarean section necessitating NICU admission.

Table 1: Audiological and radiological profile of the IP II patients.

	Age at surgery (years)	Sex	Audiometry	Tympanogram	BERA	CT-Scan and MRI finding	Side implanted
Patient 1	4	Male	Severe to profound HL	A type bilateral	No waves	Bilateral IP II	Right
Patient 2	3.4	Male	Severe to profound HL	A type bilateral	No waves	Bilateral IP II	Right
Patient 3	5	Female	Profound HL	B type bilateral	No waves	Bilateral IP II	Right
Patient 4	3	Female	Severe to profound HL	A type bilateral	No waves	Right IP II Left IP I	Right
Patient 5	1	Female	Severe to profound HL	A type bilateral	No waves	Bilateral IP II	Right
Patient 6	5	Female	Right moderately severe HL, left profound HL	A type bilateral	No waves	Bilateral IP II	Left
Patient 7	1	Female	Severe to Profound HL	A type bilateral	No waves	Right IP II with cochlear nerve hypoplasia, Left Cochlear nerve aplasia	Right

HL-Hearing loss, IP-Incomplete Partition, BERA

Table 2: Surgical aspects of cochlear implants in IP II patients.

	Approach	Round window Grade	Insertion	Perilymph/CS F leak	Other Intra-operative complications
Patient 1	Transmastoid facial recess	Ossified RW Niche and basal turn	2nd turn cochleostomy	No leak	Incus buttress, incus and stapes suprastructure removed
Patient 2	Transmastoid facial recess	Grade 2b	Cochleostomy	CSF leak	Facial N sheath exposed
Patient3	Transmastoid facial recess	Grade 2b	Extended RW	CSF leak	Canal breach Repaired with cartilage
Patient 4	Transmastoid facial recess	Grade 2a	RW	CSF leak	
Patient 5	Transmastoid facial recess	Grade 2b	Extended RW	No leak	
Patient 6	Subtotal petrosectomy with blind sac closure	Grade 2a	RW	No leak	Sigmoid sinus anteriorly placed, contracted antrum
Patient 7	Subtotal petrosectomy with blind sac closure	Grade 3	Extended RW	CSF leak	Device failure explanted and reimplanted

CSF-Cerebrospinal fluid

Table 3: Showing skills of Auditory and speech development

	Baseline CAP	Baseline SIR	CAP 6M	SIR 6M	CAP 12M	SIR 12M	Remarks
Patient 1	0	1	4	1	4	1	Completed
Patient 2	1	1	2	1	2	1	Completed
Patient 3	1	2	3	4	5	4	Completed
Patient 4	0	1	2	1	3	1	Completed
Patient 5	0	1					Irregular AVT
Patient 6	0	1	4	1			-
Patient 7	0	1					Device failure, Reimplantation

CAP-Categories of auditory performance, SIR-Speech intelligibility rating, AVT-Audio-verbal therapy, M-Month(s).

The audiological and radiological profile of the patients is shown in (Table 1).

All the children showed severe to profound hearing loss in age appropriate audiogram except 6th patient who had moderately severe hearing loss on right ear and severe to profound hearing loss on left. Aided audiograms in all patients were out of speech spectrum. OAE was absent in all patients showing outer hair cell dysfunction. BERA showed no waves up to 90dB in all the children except 6th patient. Tympanogram showed bilateral B type in 3rd patient but during surgery there was no middle ear fluid. CT scan temporal bone and MRI inner ear showed bilateral IP II in 5 children. 4th patient had IP II in right ear and IP I in left ear. The 7th patient had IP II on right ear with hypoplastic nerve and left cochlear nerve aplasia and she was counselled for ABI in due course, in the eventuality of failure of CI performance.

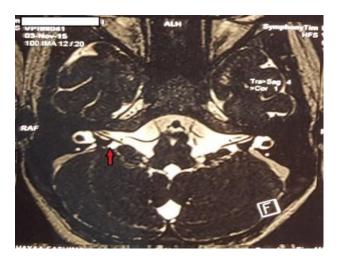


Figure 1: MRI of inner ear showing IP2.
Red arrow showing Enlarged vestibular aqueduct.

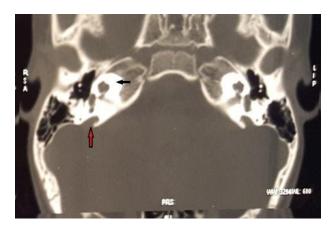


Figure 2: HRCT of temporal bone showing Bilateral IP type 2.

Red Arrow showing Enlarged vestibular aqueduct, Black Arrow showing IP II Cochlea.

All children had cochlear implantation on right ear except one patient who had moderate to severe hearing loss on right side.

Surgical procedures

The surgical aspects of the patients are briefly outlined in the following (Table 2).

5 patients had conventional transmastoid facial recess approach but in one patient; the incus buttress was removed along with incus and stapes suprastructure to facilitate middle turn cochleostomy as the basal turn was completely ossified due to previous surgical attempts at another institute. 2 patients had subtotal petrosectomy with blind sac closure.

In all the patients, post auricular Lazy S incision was given and anteriorly based palva flap was raised. Implant bed and 'tie-down' holes were prepared in all the patients. The implant was fixed with 3-0 prolene suture.

2 patients had cochleostomy. Middle turn cochleostomy was performed in one patient who had surgery elsewhere due to ossification of basal turn and in another patient with grade 2b round window. 3 patients had extended round window technique of insertion. 2 patient had grade 2b round window and one patient had grade 3 round window. Remaining 2 patients with grade 2a round window had round window insertion.

Intra-operative impedance and ART were satisfactory in all the patients.

Complications



Figure 3: CSF leak in a patient with IP 2.

CSF leak was observed in 4 patients and they were sealed intra-operatively with periostium and fascia.

Facial nerve sheath was exposed in one patient with grade 2b round window yet in another patient Canal wall was breached and was subsequently repaired with cartilage. All the patients were uneventful in the post-operative period.

One patient developed device failure with high impedance in all electrodes after 7 months of surgery and

was explanted and re-implanted. Intraoperative auditory response threshold was satisfactory.

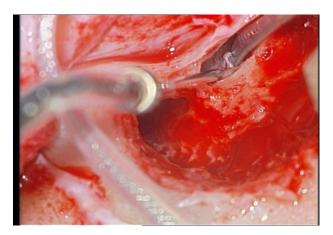


Figure 4: Soft tissue seal in a patient with IP2.

Auditory and speech outcome after cochlear implantation are demonstrated in (Table 3).

There is improvement in the CAP score of the 4 patients and speech intelligibility in one child. One child had completed only 7 months of audio-verbal therapy and there is improvement in the CAP score. In spite of the counselling and motivation from the cochlear implant team, one child had absconded from audio-verbal therapy due to undisclosed personal reasons.

DISCUSSION

The site of cochlear implantation was chosen by convention on the dominant side for easy handling of the implant which in our study is right side in all patients. However, one patient had moderate to severe hearing loss on right side necessitating the implantation to be conducted on the left side. We have chosen the worse ear as the present health scheme covers only one implant and it is our opinion that the better ear may still benefit with hearing aid thus providing the child with an added advantage of bimodal hearing.

Complete insertion was achieved in all the patients including one with the middle turn cochleostomy as in all the cases we used Med-EL Synchrony+Form 19 compressed electrodes. This has the advantage of complete insertion in malformed cochlea as well as prevention of CSF leak when combined with soft tissue seal.

The subtotal petrosectomy with blind sac closure was employed in one patient with completely concealed round window with ill-defined promontory and in another patient who had contracted antrum with forward sinus and low dura. The surgical access to the round window was enhanced following subtotal petrosectomy. Similar approach has been described for 2 malformed cochleae in a study by Free RH et al in 2013. In case of difficult

situations, subtotal petrosectomy provides excellent visibility and access in difficult anatomy or in drill-out procedures. It lowers the risk of repetitive ear infections, CSF leakage, and meningitis by closing off all connection with the external environment. The complication rate of 6% is comparable with normal cochlear implantation.¹¹

None of the patients had CSF leak due to dural tear during the preparation of implant bed and tie-down holes. There was no incidence of device migration in any of the patients. The sub-periosteal pocket technique with no fixation has been reported to have higher incidence of implant migration in a study by Lauria et al in 2015. 12

Assessment of the Status of round window visibility is important in deciding the surgical approach. One patient had grade 3 round window for which subtotal petrosectomy with blind sac closure was performed. Our institute employed St Thomas Hospital (STH) classification of round window. Grade 1 being a fully visible round window while cases with more than 50% round window visible are grade 2a and more than 50% of round window concealed, constitutes grade 2b. A completely concealed round window is grade 3. 13

Wherever possible our technique reserves the cochleostomy and extended round window technique for difficult cases where the visibility of the round window is poor. The very fact that there is less trauma to inner ear structure and new tissue formation after round window insertion was always taken into consideration as a protocol before any cochlear implant surgery in our institute. 14

Chen et al had observed a relatively slow development of auditory skills, especially in the first year in such patients. It takes longer time for habilitation of patients with Mondini type of cochlear malformation then the cochlear implantee child with well-developed cochlea.¹⁵

CONCLUSION

Incomplete partition type II may be associated with hypoplastic cochlear nerve, abnormal course of facial nerve, ill-defined promontory and round window positions. Cochlear implantation in such patients is challenging and thorough knowledge of anatomy and surgical expertise is essential as much as it is necessary to counsel the patients for a guarded outcome and eventualities.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

REFERENCES

1. Morton CC, Nance WE. New-born hearing screening-a silent revolution. N Engl J Med. 2006;354:2151-64.

- 2. Mondini C. Minor works of Carlo Mondini: the anatomical section of a boy born deaf. Am J otol 1997;18:288-93.
- 3. Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. Laryngoscope. 1987;97(40):2-14.
- 4. Sennaroglu L, Saatci I. A new classification for cochleovestibular malformations. Laryngoscope 2002;112:2230-41.
- Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. Otol Neurotol. 2006;27:615-23.
- 6. Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: radiologic differentiation. Otol Neurotol. 2004;25:520-9.
- 7. Pedro LMA. The Mondini dysplasia-from Early Diagnosis to Cochlear Implant. Acta Oto-Laryngologica. 1983;95(5-6):627-31.
- 8. Slattery WH, Luxford WM. Cochlear implantation in the congenital malformed cochlea. The Laryngoscope. 1995;105:1184–7.
- Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N. Case series: Cochlear implant in incomplete partition type I L'impianto cocleare nei pazienti con partizione incompleta tipo. ACTA otorhinolaryngologica Italica. 2013;33:56-62.
- Konstantina MS, Ann MH, Barbara H, Leila AM. Cochlear Implantation in Children with Congenital X-Linked Deafness Due to Novel Mutations in

- POU3F4 Gene. Annals of Otol Rhinol Laryngol 2010;119(12);815-22.
- 11. Free RH, Falcioni M, Di Trapani G, Giannuzzi AL, Russo A, Sanna M. The role of subtotal petrosectomy in cochlear implant surgery-a report of 32 cases and review on indications. Otol Neurotol. 2013;34(6):1033-40.
- Lauria RA, Truzzi GM, Guimarães AC, Curi SB, Castilho AM, Paschoal JR, et al. Device Fixation in Cochlear Implant Surgery: Bone Well vs. Subperiosteal Muscle Pocket Technique. Open Access Library J. 2015;2:e2177.
- 13. Leong AC, Jiang D, Agger A, Fitzgerald-OC. Evaluation of round window accessibility to cochlear implant insertion. Eur Arch Otorhinolaryngol. 2013;270(4):1237-42.
- 14. Céline R, Jose NF, Joni D, Fred HLJ. Round Window versus Cochleostomy Technique in Cochlear Implantation: Histological Findings. Otol Neurotol. 2012;33(7):1181–7.
- 15. Xueqing C, Fei Y, Bo L, Sha L, Ying K, Jun Z, et al. The Development of Auditory Skills in Young Children with Mondini Dysplasia after Cochlear Implantation. PloS One. 2014;9(9):e108079.

Cite this article as: Takhellambam BS, Venkataramani N, Natarajan K, Lairellakpam U, Kumari A, Arumugam SV. Cochlear implant in patients with incomplete partition type II: surgical considerations. Int J Otorhinolaryngol Head Neck Surg 2017;3:1081-6.