Long term outcomes of Cochlear Implantation in inner ear malformations – surgical and auditory perspectives.

Poonam Sagar, Kapil Sikka, Prem Sagar, Rakesh Kumar, Alok Thakar

Department of Otorhinolaryngology & Head Neck Surgery, All India Institute of Medical Sciences, New Delhi, India.

Introduction

- Inner ear malformations (IEM) constitutes for around 20% of congenital severe to profound hearing loss[1]. Cochlear implantation in these cases have shown benefit in existing literature[2]. However, type of inner ear malformation have significant impact on surgical, auditory and speech outcomes and long-term impact of these needs to be studied.
- CSF leaks, injury to facial nerve and complete electrode insertion are major concerns[3]. Present study also compares these parameters in IEM cases and normal inner ear implanted ears (control) group.

Objectives

- To study auditory and speech outcomes of cochlear implantation in patients with Inner ear malformation (IEM)
- To study surgical difficulties and complications of cochlear implantation in patients with inner ear malformation.

Methodology

- Study design- Retrospective study.
- Medical records of patients undergone cochlear implantation for bilateral severe to profound sensorineural hearing loss during 2018-2023 at tertiary care centre were reviewed. Group I (cases) included inner ear malformations (IEM) (n=37); group II (controls) included normal inner ear anatomy (n=42).
- Data was analysed for demographic details; types of IEM, electrode insertion, intra-operative neural response telemetry (NRT), complications, auditory outcomes with Categories of auditory performance (CAP) score and Speech intelligibility rating (SIR) using Stata 18.0 (StataCorp LP) software. Chi square/Fischer's Exact and t-test were used for categorical and quantitative variables respectively.
- Probability of auditory improvement was compared between two groups using Kaplan Meier survival analysis followed by Log rank test; (CAP – 0,1, 2 Scores are taken as poor outcome = 1; SIR- 1,2 scores are taken as poor outcome =1). Inner ear malformation, normal inner ear anatomy were independent variables.

Results

- **Case (group 1) IEM** group consisted of n= 37 patients, control (group 2) consisted of patient with normal inner ear anatomy (n=42).
- **Gender** -The number of male(M) and female(F) patients in cases (M:F, 15:22) and in controls (M:F, 18:24) (*p value= 0.835*)
- **Age** – Median age at presentation is 36 months in both the groups.

Age at implantation

- Median age at implantation is 44.9 months (P_{25}-P_{75}:36.4-60.6 months) in cases and 46 months (P_{25}-P_{75}: 31-58 months) in controls, (*p value= 0.8544*)

Social quotient (SQ)

- Mean SQ in cases and controls is 77.42 ± 8.91 and 76.34±5.606 respectively (*p value =0.2461*) . Median SQ in cases and controls is 77.5 and 75 respectively.

Hearing aid usage duration

- Median hearing aid usage duration in both cases and controls is 6 months (*p value =0.48*)

Surgical Approach

- All cases underwent cochlear implantation with standard cortical mastoidectomy with posterior tympanotomy. Approach to cochlea was either through round window or via cochleostomy. One patient in case group with IP-2 malformation required extended facial recess approach for better visualization.

Results

Operated side

- Right ear, n=71 (cases= 32, controls= 39); left ear, n=6 (cases= 5, controls =1). Sequential implantation, n=2 in control group.

Intraoperative neural response telemetry (NRT)

- NRT-all electrodes present in 19/36 in cases; 33/38 in controls.
- Number of electrodes showing absent NRT was found significant between cases and control group with *p value* of *0.0004* with case group obtaining poor NRT response rate.

Intraoperative insertion of electrodes

- Complete insertion of electrodes was obtained in 32 patients in cases and 38 patients in control group. Incomplete insertion in 4 patients in cases group and 3 in control group (One patient data not mentioned in case group), *p value= 0.699*

Table: 1 Electrode insertion specific to cochlear malformation

Electrode Insertion	IP-1(n=4)	IP-2 (n=15)	CH-II (n=1)	CH-III (n=2)
Incomplete	2	2	0	0
Complete	2	13	1	2

Electrode insertion route- Cochleostomy Vs Round window insertion

- Cochleostomy was done in 11 patients in cases and 5 in controls. Round window insertion was done in 23 patients in cases and 35 in controls with *p value 0.05,* reaching significance. In one patient, round window insertion was converted to cochleostomy. 4 cases had difficult round window insertion.

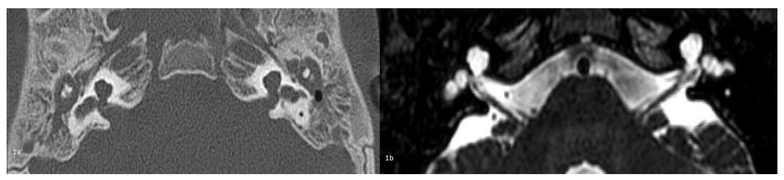

Fig 1(a): CT- Cystic cochlea bilaterally with absent modiolus and interscalar septa, (Incomplete Partition, IP-1). Enlarged and dysplastic bilateral vestibule and enlarged IAC. 1(b)MRI – hypoplasia of cochlear nerves; good outcome.

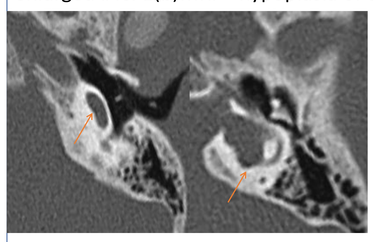

Fig 2(a): Small cystic cochlea (Cochlear hypoplasia, CH-II) Left ear (orange arrows) with Fig: 2(b) enlarged and dysplastic vestibule; poor outcome.

Cochlear nerve hypoplasia

10 IEM cases associated with cochlear nerve (CN) hypoplasia (10/37; 27.02%) while 1 case had isolated cochlear nerve hypoplasia.

Complications

- 12/37 in cases and 0/42 in controls, *(p value= 0.000).*

Table2: Complications in IEM cases. (* Transient Facial nerve palsy was present in two cases. In one case facial nerve was dehiscent and in other case facial nerve canal was intact on postoperative CT scan, probable cause thermal injury.)

Complications	Number (n)
CSF leak (16.21%)	6
Meningitis	01
Facial Nerve palsy (Transient)*	02
Vestibular	3
Others	2

CSF Leaks

Table 3: CSF leak occurrence in different IEM cases

IEM (n, %)	Major CSF	CSF Gusher	CSF Oozer
IP-1(1/4, 25%)	0	1	0
IP-2 (4/15, 26.6%)	0	3	1
CH-II (1/1)	1	0	0
CH-III (0/2)	0	0	0

Results

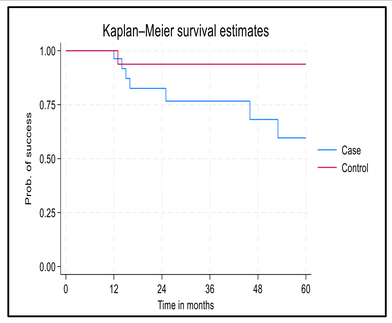

Fig 3: Kaplan Meier analysis of SIR scores

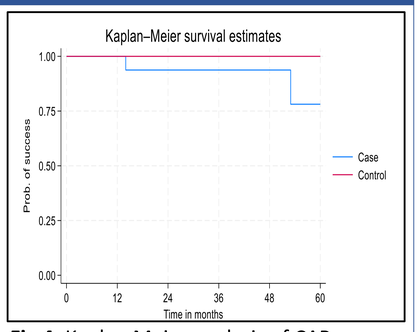

Fig 4: Kaplan Meier analysis of CAP scores

Auditory and speech outcomes

Speech outcomes: SIR score.

- Probability of having poor speech outcome in cases is 32% and 19% in controls; *p value =0.304.*
- The hazard of having poor speech outcome is 1.2 times (0.37, 4.22) higher in cases as compared to controls; *p value = 0.721.*

Table 4: Probability of getting better speech outcome SIR score; *p value= 0.721*

Time (years)	Probability Cases (%)	Probability Controls (%)
1	96	100
2	82.5	93.8
5	59.6	93.7

Auditory outcomes

- Categories of auditory performance (CAP) score

  Probability of having poor auditory outcome in control is 0% and 10.7 % in cases; *p value = 0.242.*

Table 5: Probability of getting better auditory outcome; *p value = 0.277.*

Time (years)	Probability in Cases	Probability in Controls
1	100%	100
2	93.7%	100
5	62.5%	100

Discussion

- Auditory and speech outcomes did not differ significantly between two groups in coherence with the existing literature[1-3]. However, the probability of getting better auditory & speech outcomes decreases as time progresses though not statistically significant.
- In present study, IP-1,IP-2 & CH-III malformation showed good auditory & speech outcomes. Cochlear hypoplasia of severe nature such as CH-II was associated with poor outcomes.
- Incidence of CSF leaks is 16.21%; IP-2 cases were more associated with CSF leaks as compared to IP-1 cases.
- Intraoperative electrical neural response telemetry was significantly less responsive in inner ear malformation group than controls.
- Associated status of cochlear nerve hypoplasia did not alter the prognosis though severity should be taken into consideration.
- Implanting inner with malformation carries significant risk of complications as compared to normal inner ear implantation.

Conclusion

- Majority of patients with IEM benefit from cochlear implantation in terms of auditory and speech outcomes. Decision of implanting severe IEM is guarded against increased risk of complications, poor speech and auditory outcomes.
- Poor outcomes in the present study may be affected by confounding factors[4] and retrospective nature of the study.

Contact

Dr. Poonam Sagar
AIIMS, New Delhi, India
poonamsagar249@gmail.com

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[1]Department of Otorhinolaryngology
[2]Department of Radiodiagnosis and Interventional radiology.
[3] Rehabilitation Unit of Auditory and Speech