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Research Article

OUR EXPERIENCE OF DIFFICULT CASES IN COCHLEAR IMPLANT SURGERY

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ABSTRACT

Introduction : This retrospective study is based on our experience of difficult cases in 400 implant surgeries at late Dr S N Mehrotra charitable ENT foundation between 2016 to 2019. **Materials and methods:** 30 children who presented with different challenges during implantation due to some preexisting abnormalities were included in the study. Outcomes of all the implanted children were analyzed. Scoring system for auditory performance, speech rehabilitation and quality of life were also taken into account for every child implanted taking into consideration practical issues in Indian set up**.** The results were compared with 30 children implanted having no preexisting abnormality. **Results:** Out of 30 cases, 22 had malformed cochlea, 3 had ossified cochlea, 3 children presented with chronic suppurative otitis media, 1 had auditory neuropathy and 1 had iatrogenic complication. Mondini dysplasia was the commonest abnormal cochlea finding.10% attained CAP of 5 at end of 1 year. 20% had maximum SIR score of 5 and 43.33%cases were maximally benefited in terms of quality of life. **Conclusion :** Outcome in terms of auditory perception, speech and quality of life was good taking into consideration the difficult anatomy of cochlea and other abnormal findings but were less when compared to children with no preexisting abnormalities. Cochlear implant must be done in all difficult cases as it definitely improves overall outcome.

INTRODUCTION

Over the last few years cochlear implantation has become the important treatment modality for children with severe to profound sensory neural hearing loss[1,2,3,4]. Inspite of dramatic improvement in cochlear implantation, various other factors decide the final result and outcome of implantation[5].

Variables affecting outcome of implant[6,7] are duration of disease, etiology of disease, age at onset of deafness, pre implant hearing aid use, communication mode, age at implantation[4], type of speech processor, duration of implant usage, family support and financial status, expertise provided, facilities for rehabilitation.

Inner ear anatomy plays a significant role in determining the outcome of implant. Malformed cochlea posts a major challenge for the surgeon for a successful implantation and outcome. Identification of any abnormality in cochlea before the surgery through radiology prepares the surgeon for the challenge to expect during surgery.

There are many classifications when it comes to inner ear malformations. The most widely accepted categorization is SENNAROGLU classification. It is based on embryogenesis. Every malformation is the consequence of an interruption in development at one time or another. Generally speaking it is useful and the more serious the malformation, the more surgical complications we will find and worse the outcome of cochlear implant will be.[8,9,10,11]

Sennaroglu Classification

Type of IEM	Radiology	Audiology	Gusher	FN anomaly	Treatment modality	Electrode choice
Complete labyrinthine aplasia	Absent labyrinth	Profound SNHL		Yes	ABI	ABI
Rudimentary otocyst	Incomplete milimetric otic capsule remnant	Profound SNHL		Yes	ABI	ABI
Cochlear aplasia	Absent cochlea	Profound SNHL		Yes	ABI	ABI
Common cavity	Round or ovoid cystic structure for cochlea and vestibule	Profound SNHL	Rarely	Yes	CI or ABI	Transmastoid labyrinthotomy or double labyrinthotomy, AVOID MODIOLAR HUGGING ELECTRODE
Cochlear hypoplasia	Cochlear size small, four types+	Conductive, mixed, SNHL	In CH-II possible	Yes	HA, Stapedotomy, CI or ABI	Thin and short electrode
Incomplete partition-I	Cystic cochlea	Profound SNHL	50% of the cases	Possible	CI or ABI	Electrode with stopper
Incomplete partition-II	Cystic cochlear apex	Normal to profound mixed or SNHL, progressive	Always pulsation but gusher <10% of the cases	Not expected	HA or CI, NO ABI	Any electrode but electrode with stopper preferred
Incomplete partition-III	Modiolus absent, interscalar septa present	Mixed or SNHL	100% of cases	Yes	HA or CI, NO Stapedotomy, NO ABI	Electrode with stopper. AVOID MODIOLAR HUGGING ELECTRODES
Enlarged vestibular aqueduct	Normal cochlea with enlarged VA	Normal to profound mixed or SNHL, progressive	Always pulsation	Not expected	HA or CI, NO ABI	Any electrode but electrode with stopper preferred
Cochlear aperture abnormalities	Narrow or absent cochlear aperture	Profound SNHL, OAE may be normal, profound SNHL	none	Not expected	CI for CN hypoplasia, or ABI if CN is absent	Standard CI or ABI

IEM: inner ear malformation; SNHL: sensorineural hearing loss; FN: facial nerve; VA: vestibular aqueduct; CN: cochlear nerve; HA: hearing aid; CI: cochlear implantation; ABI: auditory brainstem implantation; OAE: otoacoustic emissions

Every complicated cases present with different challenges . MRI(magnetic resonance imaging) brain and HRCT(high resolution computed tomography) temporal bone are prerequisite for all implant cases to identify all the complicated cochlear cases pre operatively and plan accordingly.

MATERIALS AND METHODS

Study was carried out at late Dr S N Mehrotra Charitable ENT foundation from 2016 to 2019. 400 children implanted during this period, out of which 30 having difficult implantation were taken into consideration. Study was done by collecting data through fully completed clinical records and information regarding present performance of implantees from our team. The discussion also includes feedback from rehabilitation team about performance of each implantee, the

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duration at which patients attained speech abilities and reach the respective categories of CAP(category of auditory perception), SIR(speech intelligibility rating) score and their GCBI(glasgow children benefit inventory) index.

The following inclusion criteria were applied to all the selected subjects in the study.

Inclusion criteria

- Children with bilateral severe to profound sensori-neural hearing loss.
- had hearing aid trial for minimum of 3 months
- psychological assessment was normal.

Evaluation protocol at Mehrotra Ent Hospital

a. Informed written consent was taken from the parents for the study and follow-ups required during the study.
b. A detailed history and thorough physical and ENT examination was carried out.
c. The subjects then underwent pediatric examination to rule out any neurological condition, which may hamper the child postoperative performance. Prior to implantation a basic workup including hematological, chest X-ray, ECG(electrocardiogram), TORCH(toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and HIV) screen (if require). The general physical condition will be evaluated by anaesthetist. A specialist opinion was sought in patients with syndromic etiology of deafness. In children pre implant vaccination will be carried out.
d. Behavioral observational audiometry, impedance, OAE(otoacoustic emissions), auditory brainstem response thresholds and auditory steady-state response was determined to evaluate the degree of hearing loss.
e. Each child was subjected to undergo a high resolution CT(computed tomography) scan and MRI (magnetic resonance imaging)scan of Temporal bones.
f. Speech perception was also assessed by SIR score before implant
g. The child was also evaluated by a child psychologist to determine the IQ(intelligent quotient).
h. Counseling of parents was done regarding regular follow-ups and therapy/support to the child at home. They were also made to realize the realistic expectations about the cochlear implant. Also the parents were made to realize that they are integral part of our rehabilitation team which requires consistent hard work and patience

Cochlear implantation was done and the impedance checked. NEURAL RESPONSE TELEMETRY (NRT) was done in nucleus implants and effectiveness assessed in children. Post operatively x ray was mandatory.

The switch on and speech processor tuning done at 1-2 weeks after surgery. Mapping is done at periodic intervals till a stable map is achieved. The rehabilitation program was started out based on baseline skills of child, periodical assessment of outcomes was done in terms of environmental sound speech discrimination and telephonic conversation. The recommended period for rehabilitation is 2 years. All 30 children are using the implant. There are no non users.

Outcome Measures

The subjects were followed up for a maximum period of 1 year at intervals of 3 months, 6 months and 12 months after implantation. Outcome measures were followed as under category of auditory performance (CAP), Speech intelligibility rating (SIR) and Glasgow children benefit inventory (GCBI)[12,13,14]

RESULTS

Out of 30 children, 12(40%) were male and 18(60%) were females.

12 children belong to age group of 5-6 years, 3 belong to 4-5 year group, 12 to 3-4 year group and 3 were in age group of 1-3 years.

All children were pre lingual deaf .

18(60%) were implanted with Nucleus implant and 11(36.6%) with Digisonic. One was implanted with Medel implant.

Table 1 Showing Findings of 30 Patients Of Abnormal Cochlea

Types of complicated cases	Number of children
Malformed cochlea	22
Ossified cochlea	03
Chronic suppurative otitis media	03
Auditory neuropathy	01
Iatrogenic	01

Table 2 Malformed Cochlea Findings

Malformed Cochlea Findings	Number of children
Common cavity	01
IP_1	04
IP_2	15
IP_3	02

- MONDINI DYSPLASIA (type 2 incomplete partition) contribute to maximum cases of malformed cochlea.(50%). The dysplasia is characterized by presence of only one and half cochlear turns. During implantation mild CSF gusher was present.
- 1 patient of common cavity presented with bilateral abnormal cochlea with large cystic cochlea and large vestibular aqueduct. Internal auditory canal widened. CSF GUSHER was the prominent finding.
- CSF gusher was also prominent in patient with type 1 and type 3 incomplete partition.
- In 3 patients of chronic suppurative otitis media who were implanted, subtotal petrosectomy with blind sac closure with abdominal fat was done.
- In 3 patient of ossified cochlea, 2 had basal turn cochleostomy while 1 had middle turn cochleostomy.
- 1 patient had iatrogenic complication. In this patient, right ear cochlear implantation was abandoned due to presence of 1.0 mm diamond burr which broke in basal turn of cochlea. Revision surgery done on next day. Foreign body (burr) was removed and left ear cochlear implantation was done.

HEARING(CAP), SPEECH(SIR) AND QUALITY OF LIFE(GCBI) index findings of our complicated cases after 1 year of extensive rehabilitation and comparison with cases with no preexisting abnormality.

Table 3 Cap Score for Complicated Cochlear Cases

CAP at end of 1 year	Number Of Children
2	09(30%)
3	06(20%)
4	12(40%)
5	03(10%)

Out of 30 abnormal cochlear patients, 10% patients had attained highest CAP of 5 at end of 1 year. Maximum (40%) had CAP 4at end of 1 year.

Table 4 Cap Score of 30 Cases Implanted With No Pre Existing Abnormality

Cap score at end of 1 year	Number of children
11	01(3.33%)
10	01(3.33%)
09	02(6.67%)
08	03(10%)
07	05(16.67%)
06	05(16.67%)
05	09(30%)
04	03(10%)
03	01(3.33%)

In these cases average highest CAP score is 11 and lowest attained is 3 with maximum (30%) cases attaining score of 5.

Table 5 Comparison of Sir Score of Complicated and Normal Cases Implanted

SIR at end of 1 year	Number of children with complicated ear	Number of children with no pre existing abnormality
01-03	10(33.33%)	03(10%)
04	14(46.67%)	18(60%)
05	06(20%)	09(30%)

20% children had attained maximum SIR of 5 and majority(46%) has attained score of 4 in complicated cases whereas nearly 60%attained score of 4 and 30%with maximum speech benefit.

Table 6 Comparison of Gcbi Index of Complicated And normal cases

gcbi at end of 1 year	number of children with complications	number of children with no complications
>70	13(43.33%)	20(66.67%)
60-70	06(20%)	06(20%)
50-60	07(23.33%)	03(10%)
40-50	04(13.33%)	01(3.33%)

43.33% of abnormal cochlear children had >70 improved quality of life index where as 66.67% showed highest level of GCBI in cases with no pre existing complications.

Overall children with difficult cases show inferior results as compared to cases with no pre existing cases with there highest CAP, SIR score and GCBI index lower compared to normal ones.

Complications

- CSF gusher was the prominent complication seen in malformed cochlea patients. All patients except one were managed conservatively by head end elevation and IV fluids. 1 required revision surgery. Subtotal petrosectomy with blind sac closure was done.
- 4 children had facial paresis which recovered conservatively in 4 weeks.
- 3 had hematoma, recovered conservatively in 2 weeks.

CONCLUSION

- Abnormal cochlea was present in 30 cases out of 400 implanted.(7.5%)
- MALFORMED cochlea seen in 22(73.33%) out of 30 total complicated cases. Ossified in 3(10%) cases.
- MONDINI'S DYSPLASIA (type 2 IP) was the most common findings in cases of abnormal cochlea.
- CSF GUSHER was the commonest finding of all the complicated cases operated.
- CSOM contributed to 10% cases of our complicated cases.
- Hearing results were significantly improved in all of our abnormal cochlear cases with average CAP score of 4 in 40% of our cases, minimum CAP being 2 attained by 30% of our cases and maximum CAP attained was level 5 whereas in normal cochlear cases, average CAP being between 5-7 and maximum level being level 11.
- 66.67% showed significant higher improvement in speech(SIR SCORE OF 4 and 5) of our abnormal cochlear cases with remaining 33.33%% achieved lowest score of 4 whereas in normal cochlea cases 90% showed significant higher speech score of 4 and 5.
- Nearly 43.33% had maximum benefit and further 43% with moderate benefit in improved quality of life compared to 67% with maximum benefit of normal cochlear cases.
- Cochlear implant must be done in all difficult cases as it definitely improves outcome.

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