



# A Study of Anomalies of Cochlea and Cochlear Nerve in Children with Congenital Profound Hearing Loss – An Indian Perspective

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## Authors' contributions

*This work was carried out in collaboration between all authors. Author GN designed the study, performed the statistical analysis, wrote the protocol, wrote the first draft of the manuscript and managed the literature searches. Authors SV, RS and RBP managed the analyses of the study. The study was done under the guidance of author MK. All authors read and approved the final manuscript.*

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## ABSTRACT

**Background:** Congenital Profound hearing loss is one of the leading handicaps that is encountered among children in the Indian subcontinent. There is a great demand for cochlear implantation in the Indian clinical scenario due to the high incidence of congenital hearing loss, due to a multitude of etiological factors. Such factors may also lead to anomalies in the cochlea and cochlear nerve, thus leading to challenges for cochlear implantation. With advancements in high resolution radio-imaging, the selection criteria for cochlear implantation has today become precisely defined. In children born with cochlear nerve aplasia and Michel deformity, auditory brain stem implantation is the only other alternative for auditory rehabilitation. There exists some "grey zones" like cochlear nerve hypoplasia, where the implant of choice cannot be clearly defined. Prevalence of anomalies of cochlea and cochlear nerve in relation to congenital severe to profound

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hearing loss has evoked much interest and since there is no available data on this subject, considered worthwhile to undertake this study.

**Aims:** This study focuses on our experience with identifying the prevalence of inner ear malformations, the possible etiologies for these malformations and planning the appropriate management protocols for the same.

**Study Design:** Retrospective study. Outcome measures compared using student t-test (paired).

**Place and Duration of Study:** Madras ENT Research Foundation, Chennai, India during the period 2005-2014.

**Methodology:** The study assessed the prevalence & distribution of Cochlear and Cochlear Nerve malformations in 82 children with profound hearing loss (aged between 9 months – 6 years) presented at Madras ENT Research Foundation, a premier auditory implant institute in South India during the period 2005-2014. The outcomes of CI/ABI in children with inner ear malformations were compared by using standard measures like CAP, SIR, MAIS and MUSS scores at sequential intervals of 6 months and 12 months post implantation.

**Results:** Eighty-two patients with cochlear and cochlear nerve malformations were identified among a consecutive series of 926 children with congenital severe to profound hearing loss. Of these, bilateral malformations were seen in 74 patients and unilateral malformations in 8 patients. Out of 74 patients, 66 had undergone surgery and among these 66 patients, 56 had CI and 10 had ABI. The study showed that there were significant statistical differences in the results as measured by outcome scores during the study, between 6 months and 12 months post implantation. Complications of surgery, though more common with anomalous anatomy, were not very morbid and could be overcome with appropriate measures.

**Conclusion:** Improvement in auditory perception and speech development skills among the different group of children with inner ear malformations were good both with CI and ABI. We infer that auditory implants have paved the way forward for children with inner ear malformations to acquire auditory verbal skills, but a judicious selection of cases for appropriate implantation is vital to achieve best outcomes.

*Keywords: Cochlear implant; auditory brainstem implant; category of auditory performance; speech intelligibility rating; meaningful auditory integration scale; meaningful use of speech scale.*

## 1. INTRODUCTION

The sense of hearing is perhaps the most important of all the five special senses which we humans possess. Unfortunately, hearing loss is a silent handicap, with so many people in India suffering from varying degrees of hearing impairment. Loss of hearing in childhood leads to delayed speech and language development. The result of congenital hearing loss is a "Deaf and Dumb" individual living within his deaf world, with lack of communication and remains aloof from the society. In contrast to loss of vision, which is an easily recognizable handicap and evokes sympathy from the public, hearing loss is often a hidden handicap and is often ridiculed. Hearing loss is the most common sensory deficit in humans today. The prevalence of hearing loss in India is fairly significant. It is the second most common cause of disability. Approximately 63 million people (6.3%) in India suffer from significant auditory loss [1]. As per the national sample survey organization (NSSO) survey, currently there are 291 persons per lakh population who are suffering from severe to

profound hearing loss (NSSO, 2001). Of these, a large percentage is from children aged between 0 and 14 years. Such a large number of hearing impaired young Indians accounts for a severe loss in productivity, both physically and economically [2]. This scenario reflects the severity of hearing impairment which occurs due to abnormalities in the structure or functioning of the outer ear, middle ear, inner ear or the auditory pathways.

Congenital Profound hearing loss is one of the leading handicaps among children in India. Out of every 1000 children born in India, there may be 5–6 children who cannot hear properly [3]. They may not be identified till they attain at least 2 years of age, by then irreversible damage would have been done. The general awareness level among our public to recognize loss of hearing at the earliest and seek appropriate medical help is still low, in spite of the best efforts of our Government's Health Department, to propagate knowledge regarding hearing loss through the 'National Program for Prevention and Control of Deafness (NPPCD) With the advent of

this programme (NPPCD) in 2006, there is renewed interest in tackling this public health disaster [4].

A good history, clinical examination and full audiological and radiological evaluation are essential to determine the cause of deafness. There is a great demand for cochlear implantation in the Indian clinical scenario due to the high incidence of congenital hearing loss, due to a multitude of etiological factors. The selection criteria for cochlear implantation has today become much precise, thanks to high resolution radio-imaging. In children born with cochlear nerve aplasia and Michel deformity, auditory brain stem implantation is the only other alternative for auditory rehabilitation. There exists some "grey zones" like cochlear nerve hypoplasia, where the implant of choice cannot be clearly defined. This study focuses on our experience with identifying the prevalence of inner ear malformations and planning the appropriate management protocols for the same. The management of hearing impairment in children depends on the cause and requires a multidisciplinary team including the ENT surgeon, neurosurgeon, child psychologist, ophthalmologist, occupational therapist, paediatrician, audiologist & habilitatationist.

Congenital malformations of the outer and middle ear are predominantly unilateral (70-90%) and mostly involve the right ear. Inner ear malformations may be unilateral or bilateral [5]. The incidence of ear malformations is approximately 1 in 3800 newborns [6]. Cochlear malformations have been reported to occur in approximately 20% of children with congenital sensorineural hearing loss [7]. Unfortunately, there is no available data so far on the prevalence of congenital malformations of the inner ear in Indian population. The study aimed to evaluate the prevalence of cochlear and cochlear nerve malformations in children with severe to profound hearing loss in Indian population. The study also looked at any possible etiology for the malformations. A management protocol was formulated for implantation in these malformations and the outcomes among these children were analyzed in detail using CAP, SIR, MAIS and MUSS scores at 6 and 12 months.

## 2. MATERIALS AND METHODS

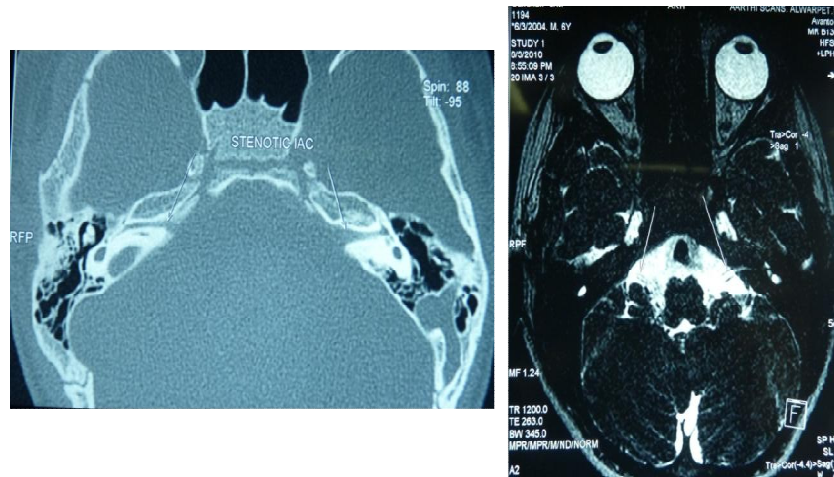
This clinical retrospective study was based on non-randomized longitudinal data, collected from a large database of 926 children who consecutively received implants at our institute

over 9 years from 2005 to 2014. The sample analyzed comprised of eighty two cases of age group ranging from 9 months to 6 years.

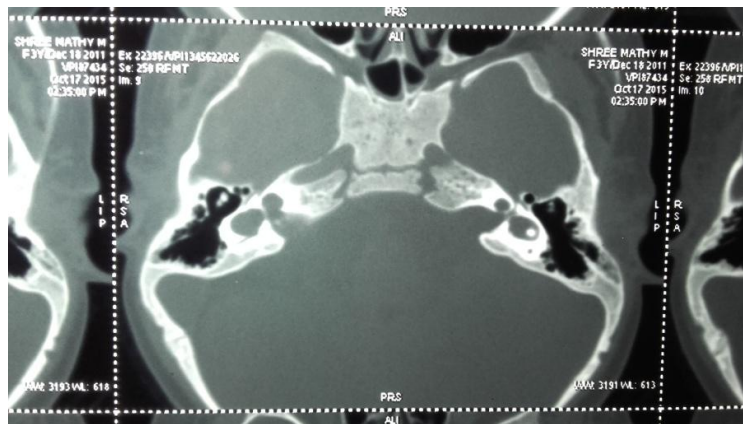
Patients with severe to profound deafness with cochlear anomalies such as common cavity, incomplete partition type I & II with or without large vestibular aqueduct, cochlear nerve anomalies such as aplasia and hypoplasia were included in the study. Those with congenital profound deafness with normal cochlea and cochlear nerve, acquired profound deafness, labyrinthitis ossificans post meningitis, central auditory processing disorder and syndromic association with profound deafness were excluded from the study. The study was approved by the institutional ethical review board on 26<sup>th</sup> Dec 2013.

Patients with hearing loss were assessed based on detailed clinical history including consanguinity among parents, detailed antenatal, perinatal and postnatal history, history of vaccination, neonatal infections, NICU admission, childhood infections, history suggestive of other system involvement to know syndromic associations, developmental milestones etc. and detailed clinical examination, battery of audiological tests including Pure Tone Audiometry, Impedance audiometry, Otoacoustic Emission and Brainstem Evoked Response Audiometry and radiological investigations included High Resolution imaging CT of 0.5 mm thickness and MRI (0.8 mm thickness, matrix 320 X 224 and field of view 18 X 14 cms) of the temporal bone and inner ear (Figs. 1, 2, 3). There was no financial implication to the candidates included in the study.

An algorithm of the appropriate management of the selected cases for cochlear implantation and auditory brain stem implantation were clearly defined and appropriate counseling were given to parents of children before proceeding on to the implantation. Among 82 children included in study group, 66 had undergone surgery, CI (cochlear implantation) and ABI (auditory brain stem implantation) uneventfully. All these children were switched on as per the standardized protocols and they received auditory habilitation under meticulous supervision by the same team of professionals. The outcome measures like CAP(Categories of Auditory performance, SIR (Speech Intelligibility Ratings), MAIS (Meaningful Auditory Integration Scale) and MUSS (Meaningful use of Speech scale) were assessed periodically at 6 and 12 months. Data collected were analyzed with regards to the



**Fig. 1. CT Scan showing stenotic internal auditory canal and MRI showing bilateral cochlear nerve aplasia**



**Fig. 2. CT scan showing Incomplete partition 1**

demographic distribution, management protocols applied and comparison of outcome measures using SPSS software 17.0. Outcome measures were compared by using the student 't' test and values considered statistically significant or not.

### 3. RESULTS AND DISCUSSION

#### 3.1 Patient Characteristics

Eighty-two patients with cochlea and cochlear nerve malformations (8.85%) were identified among a consecutive series of 926 children with congenital severe to profound hearing loss, during the period 2005-2014, which constituted the study group. Mean age of study group was 3.2 years (nine months to six years), among which 54 (65.8%) were males and 28 (34.2%) were females.

The etiology of hearing loss were varied. Among group of eighty two children, thirty (36.5%) patients had history of second degree consanguinity among parents, ten (12.4%) patients with history of prematurity, birth asphyxia among 8 (9.75%) patients, antenatal infections in 7 (8.5%), combined factors i.e. birth asphyxia, prematurity and consanguinity in 4 (4.83%) patients and unknown cause in 23 (28.05%) patients.

#### 3.2 Prevalence of Inner Ear Anomalies

For data analysis, the study population of 82 candidates were divided into two groups – Group I were those with bilateral anomalies which comprised of seventy-four patients (90.2%) and Group II were those with unilateral anomalies which comprised of eight patients (9.8%). These

groups were again divided into three subgroups as below:

**Group I**

*Congenital profound deaf with bilateral anomalies = 74/82 (90.2%).*

- A) Bilateral cochlear anomaly with nerve anomaly =2 (2.6%).
- B) Bilateral cochlear anomaly but normal nerve =50 (61%).
- C) Bilateral normal cochlea & bilateral nerve anomaly = 22 (26.8%).

**Group II**

*Congenital profound deaf with unilateral anomalies =8/82 (9.8%).*

- A) Cochlear anomaly with nerve anomaly = 4 (4.8%).
- B) Cochlear anomaly with normal nerve = 2 (2.4%).
- C) Normal Cochlea with nerve anomaly = 2 (2.4%).

Overall, the total number of cochlear malformations were 58 (70.7%) of which bilateral malformations were 52 (63.4%) and unilateral malformations were 6 (7.3%). Similarly the total nerve malformations were 30 (41.4%) of which bilateral were 24 (29%) and unilateral were 6 (7.3%). All together the isolated cochlear malformation was seen in 52 (63.4%) patients, isolated Nerve malformation were 24 (29.2%)

and cochlear with cochlear nerve malformations were seen in 6 patients (7.3%). The distribution and types of various anomalies are shown in Tables 1 and 2.

**Table 1. Bilateral cochlear malformations (n==52)**

Cochlear anomaly	No. of pts.	Percentage
IP 1	5	9.6%
IP11	39	75%
Common cavity	6	11.5%
Michel deformity	2	3.8%

*Number of patients with each cochlear malformation (bilateral) with percentage*

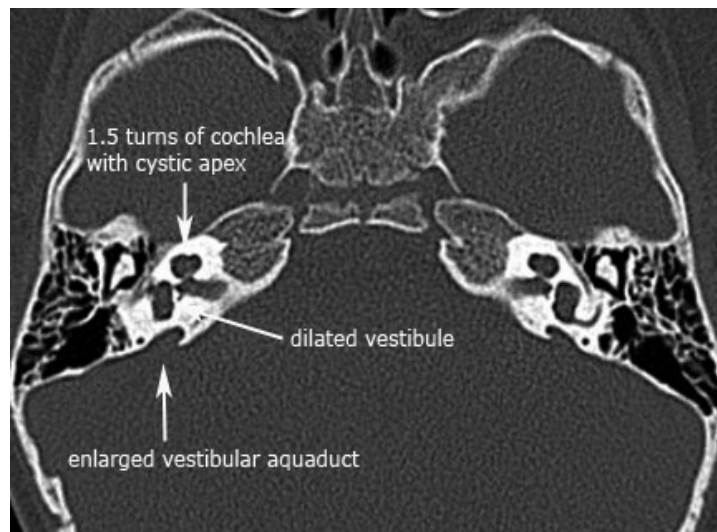
**Table 2. Unilateral cochlear malformations (n=6)**

Cochlear anomaly	No of patients	Percentage
IP II	3	50%
Common cavity	1	16.6%
IP I	1	16.6%
Michel deformity	1	16.6%

*Number of patients with unilateral cochlear malformation with percentage*

**3.3 Management Protocol**

Based on the spectrum of inner ear malformations identified, the implant team devised the algorithm as shown in Table 3 for appropriate management.



**Fig. 3. CT scan showing mondini deformity**

**Table 3. Type of malformations and management protocols**

Side	Cochlear anomaly	Cochlear nerve		Combined
		Hypoplasia	Aplasia	
Unilateral	CI	CI	CI (opp.ear)	CI
Bilateral	CI	CI/ABI	ABI	ABI

**3.4 Summary of Treatment**

In Group I, out of 74 bilateral anomalies, both 2 patients in sub group A had undergone ABI. In B subgroup out of 50, 44 patients had CI and 6 patients are awaiting CI. In subgroup C out of 22, 6 had CI, 8 had ABI and 8 are awaiting ABI. In Group II, out of 8 patients with unilateral malformations, only 6 had surgery and all had cochlear implantation on normal side (Table 4).

**Table 4. Total number of patients for CI and ABI**

Treatment	Advised	Undergone surgery	Surgery planned
CI	64	56 (68.2%)	8 (9.75%)
ABI	18	10 (12%)	8 (9.75%)
Total	82	66 (80.4%)	16 (19.5%)

**3.5 Complications**

Complications following cochlear implantation and auditory brain stem implantation were relatively few. Among 56 patients who underwent cochlear implantation, CSF leak was noticed in ten (17.8%) patients, untoward bleeding in three (5.3%) patients, incomplete insertion in three (5.3%) patients and difficult insertion in four (7.1%) patients. Among postoperative complications, out of 56 patients, six (10.7%) patients developed wound infection, flap necrosis in two (3.5%) patients, bio film in two (3.5%) patients and trauma and device failure in one patient (1.8 %) each. Among ten patients who have undergone ABI, intra operative complications were bleeding in one (10%) patient and non auditory stimulation in eight (80%) patients. Post-operative complications were

wound infection and flap necrosis in one patient (10%) each.

**3.6 Measurement of Outcomes**

Out of 56 patients who underwent CI, 50 patients were from group I and 6 patients from group II. In group I, 44 patients were from subgroup B and 6 patients were from subgroup C. As group II had unilateral malformations, they underwent CI on the opposite normal ear. Thus the outcome of group I was compared with that of group II.

The mean average values of CAP, SIR, MAIS and MUSS scores at six and twelve months were as shown in Table 5.

The graphical representation with error bar of the outcomes are as shown in Figs. 4 and 5.

CAP, SIR, MAIS and MUSS scores of CI group were found to be statistically significant between 6 and 12 months at *P* values of .034, .014, .005 and .018 respectively. The improvement in outcome of group I with CI is not statistically significant (*P*=.082) with as compared with outcomes of group II patients.

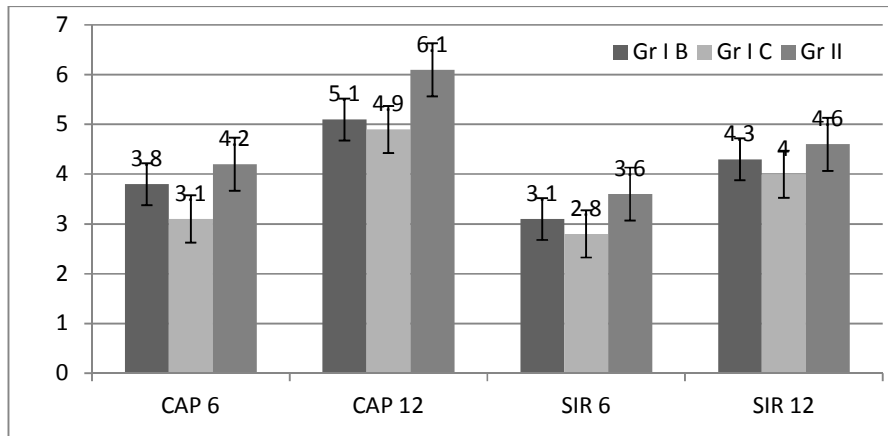
Post ABI comparison of CAP, SIR, MAIS and MUSS scores were found to be statistically significant between 6 and 12 months at *P* values of .004, .012, .010 and .04 respectively.

**3.7 Discussion**

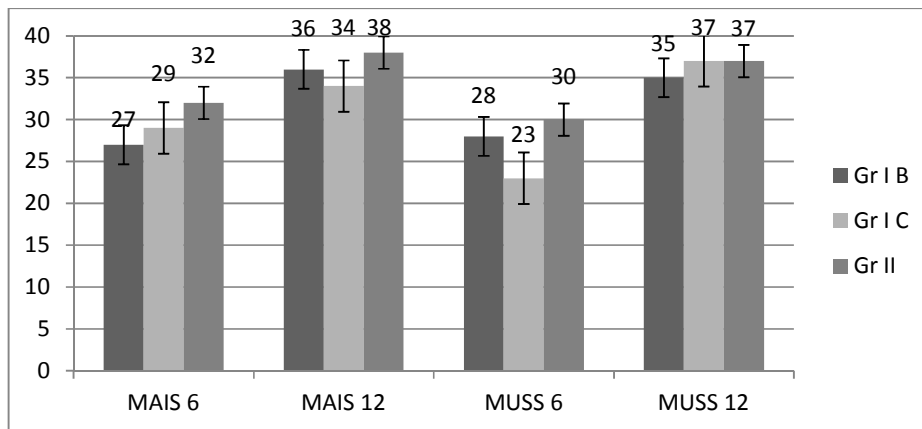
The success of an implant program is directly dependent on its ability to address the issue of patient expectations and balance it with the outcomes. A multidisciplinary approach along with motivation of the patient and family is very

**Table 5. Outcomes of CI at 6 and 12 months**

Group	No. of patients	Mean CAP		Mean SIR		Mean MAIS		Mean MUSS	
		6 mth	12 mth	6 mth	12 mth	6 mth	12 mth	6 mth	12 mth
I B	44	3.8	5.1	3.1	4.3	27	36	28	35
I C	6	3.1	4.9	2.8	4.0	29	34	23	37
II	6	4.2	6.1	3.6	4.6	32	38	30	37
Total	56								



**Fig. 4. Mean CAP and SIR scores at 6 and 12 months**



**Fig. 5. Mean MAIS and MUSS scores at 6 and 12 months**

important in this regard. Variables affecting the outcome of CI in children are the duration and etiology of deafness, residual hearing, age of onset, antenatal, perinatal and postnatal history, age at implantation, type of speech processor used, rehabilitation process and duration of implant usage. In very young children, language acquisition is easier and hence the need for early implantation. Cost of the implant seems to be a major limiting factor in a developing country like India. The dilemma of balancing an advanced technology with the requirements of a developing country still remains. Problems unique to the Indian context are the distances between CI facilities and the multi-lingual society forming a language barrier for rehabilitation. These can be overcome by having satellite centers, trained surgeons with adequate theatre facilities and a well equipped audiology unit with access to good schools for hearing challenged, which believe in an auditory verbal approach along with long term

habilitation programme. In general, inner ear deformities are no longer an absolute contraindication to cochlear implantation [8].

Among etiological factors, consanguinity was seen in parents of thirty (36.5%) patients. A study conducted by Lofti et al. [9] showed that the prevalence of hearing impairment in children of parents who had consanguineous marriage was significant, which almost correlate with our study. Final results of their study showed that the parents of 58.2% of children had consanguineous marriage whereas prevalence of consanguinity was 22.3% in society. Extensive studies carried out in India from Andhra Pradesh, Karnataka and Tamilnadu have shown high percentage of consanguineous marriages leading to high percentage of children with amino acid disorders, congenital anomalies and genetic diseases [10]. Other factors noted were prematurity, birth asphyxia, antenatal infections

and combined causes which include co-existing consanguinity and birth asphyxia.

Among the type of bilateral cochlear malformations, in 52 patients, IP II was found in 39 (75%) patients, common cavity in 6 patients (11.5%), IP I in 5 (9.6%) patients, Michel deformity in 2 (3.8%) patients. This correlates well with the classification system proposed by Jackler et al. [11]. In their study, IP II was encountered in 55% of the cases followed by common cavity in 22%. Among unilateral cochlear malformations which were 8 (19.8%) in number, IP II was seen in 3 (50%) patients, Michel, common cavity and IP I in one patient each (16.6%). In a study by Sennaroglu et al. [12] of 20 patients with inner ear malformations the age range was between 2 and 37 years (average, 8.8 yr). There were two patients with common cavity deformity, four cases of incomplete partition (IP) type I (cystic cochleovestibular malformation), four cases of IP type II (classical Mondini deformity), nine patients with large vestibular aqueduct (LVA) syndrome. In another study by Papcin et al (2005) carried out to review the cochleovestibular anomalies among 298 children implanted over the decade ending in January 2002 the children were grouped based on cochleovestibular anatomy as follows: normal (n=195), common cavity deformity (n = 8), hypoplastic cochlea (n = 16), incomplete partition (n = 42), and vestibular aqueduct enlargement (n=37). Concomitant anomalies of the posterior labyrinth (n=26) and internal auditory canal/cochlear canal (n = 11) and one patient with X-linked deafness were also identified [13].

All patients with unilateral malformation underwent cochlear implantation on the opposite side. Complete labyrinthine aplasia also known as Michel deformity was seen in 3 of our patients of which it was bilateral in 2 patients and unilateral in 1 patient. This bilateral abnormality was an absolute contraindication for CI on the affected side and ABI was done in these two patients.

Of the 82 patients, 64 patients were advised CI. This included 50 patients with bilateral cochlear malformation and normal nerve, 8 patients with unilateral cochlear malformation and 6 patients with bilateral normal cochlea and nerve malformation. These 6 were chosen from 22 patients with bilateral nerve malformation and normal cochlea (Group I C). Though these 6 patients were candidates for ABI, they eventually underwent CI since they were in the "grey zone"

(one sided thin nerve and aplastic nerve on the other side). Out of 56 cochlear implantees, 2 cases had to undergo revision implantation, one due to device failure and other due to trauma sustained.

Isolated nerve malformation was seen in 24 (29.2%) patients, among which bilateral nerve malformation in 22 (26.08%) patients and unilateral nerve malformation noted in 2 (11%) patients. Eighteen patients with nerve malformations were advised ABI. This includes 2 patients with Michel deformity in group I A and 16 patients from group I C. Ten of these patients underwent ABI and eight patients are awaiting surgery.

The intra operative complications in CI we encountered were CSF leak in 10 (17.8%) patients, bleeding in 2 (3.5%) patients, incomplete insertion in 3 (5.3%) and difficult insertion in 4 (7.1%) patients. The incidence of CSF leak in our study is consistent with that of Sennaroglu et al. (22%) [12]. In a study conducted by Dettman et al. [14], 23% had CSF gusher during surgery. In our series we had complete insertion in 59 patients (95.16%). This percentage is slightly higher than that of Luntz et al. [15] who reported a 90% rate for full insertion.

Statistical analysis of the outcome measures were compared using student t-test (paired) and inferences were derived. We found statistical significance in the improvement of CAP scores between 6 and 12 months among the sub groups A, B and C. (*P* value .0384 and .014). All the subgroups who underwent CI showed statistical significance in the CAP and SIR scores when compared at 6 and 12 months (*P* value=.0384). The above results helped to infer that children with inner ear malformation in general tend to have lower auditory perception skills which gradually improve over time but may not achieve the levels which are noted in an implantee with normal cochlear anatomy (group II). Cochlear implantation significantly benefits such children in aiding their speech development skills which is also reflected in the findings above.

Comparative analysis of MAIS and MUSS scores between the sub groups were statistically significant at *P* values .005 and .018 respectively when compared at 12 months of rehabilitation. The study results correlates well with that of Colletti et al. [16] in which post ABI, the CAP scores ranged from 1 to 7 (average 4), MAIS at



an average of 38%, Muss 49%. This implies that intensive auditory rehabilitation significantly helps in providing auditory and speech skills to all children using cochlear implants and ABI in a similar manner irrespective of the presence or absence of inner ear malformations.

Even though children with inner ear malformations may not be up to the mark in comparison to implantees with no cochlear malformation, this difference does exist, more so in earlier part of the rehabilitation. Over a period of time, children learn to use the implant which bypasses the anomaly they were born with. However the final outcome in paediatric implantation is not entirely predictable as there are a large number of factors which alone or in combination will decide the outcome of cochlear implantation. In a study by Govaerts et al. [17], they concluded that patients with aplasia or hypoplasia of the cochleovestibular nerve should be counselled with caution with respect to cochlear implantation but particular circumstances may justify their intervention.

Eventually, the children that we followed up in the various study groups have all become good CI users with appropriate communication skills which have helped them to integrate into normal schools and lead a productive life.

Of 10 ABI recipients whose outcome measures were analyzed over time, we found the mean average CAP score was 2.1 (range 1–3) at 6 months and 3.1 (range 2–4) at 12 months respectively. Similarly, the mean average SIR score was 1.8 (range 1–4) at 6 months and 2.9 (range 1-5) at 12 months respectively. Mean average MAIS score was 21 (14-26) at 6 months and 26 (22-31) at 12 months and MUSS mean average score was 18 (14-23) at 6 months and 22 (18-26) at 12 months respectively as shown in Table 6.

The inferences derived from the above study showed that there is significant advantage of CI over ABI since better outcomes are achieved in CI rather than ABI patients. The underlying reason for such an observation may be due to the preservation of tonotopic integrity and temporal integration of sound within the cochlea in a cochlear implantee in comparison to a direct

surface stimulation of the cochlear nucleus in an ABI recipient. The results have also shown that ABI does provide good auditory perception to patients and may help in aided development of speech skills in those patients who otherwise do not have an alternative for acquiring a natural way of communication.

Rehabilitation in ABI is extremely challenging and needs to be customized according to the needs of the recipient unlike the usual protocols followed for a cochlear implantee. As we followed up our 10 ABI children, we found significant improvement in their communication skills over time. Even though they were slow learners in comparison to cochlear implantees, these children also became good ABI users and have been able to integrate into the normal society through normal communication mode.

Preoperative evaluation, surgical approach and postoperative follow up can be challenging. CT and MRI plays important role in selecting candidates for cochlear implantation and circumvent potential difficulties and complications at cochlear implantation. They also help to choose candidates for ABI (especially paediatric cases with cochlear nerve aplasia). Anticipation of potential problems helps greatly in planning the surgical technique and predicting postoperative outcome. In summary, this study has reflected the panorama of inner ear malformations that occur in congenital hearing impaired children. The management protocol for hearing restoration in these various anomalies has been highlighted. The outcome measures between groups have been evaluated and inferences on the eventual success of CI / ABI in such cases have been highlighted.

Overall, the results of this study have been in comparison with existing world literature even though the present study has provided an insight into the Indian perspective with indigenous data for the first time. Such studies are also evolving in other reputed CI centers across India and meta-analysis of the results from multi-centric research work would pave the way forward for refining the management protocols and predicting the habilitation outcomes of CI and ABI recipients in India in the near future.

**Table 6. Outcomes of ABI at 6 and 12 months**

	No of Pts	Mean CAP		Mean SIR		Mean MAIS		Mean MUSS	
		6 m	12 m	6 m	12 m	6 m	12 m	6 m	12 m
ABI	10	2.1	3.1	1.8	2.9	21	26	18	22

#### 4. CONCLUSION

The importance of early detection and management of hearing loss in children cannot be overemphasized. Identification of the type and cause of hearing loss has to be done meticulously, because the management depends on the etiology of hearing impairment. However, of all the handicaps, hearing loss is the only truly remediable handicap and the mantra is "Early detection and management". Children born deaf must be brought to the ENT surgeon at the first suspicion of hearing loss to enable timely intervention.

The problem of consanguinity resulting in children being born with hearing loss due to admixture of defective gene pool running in families also needs to be addressed and appropriate genetic counselling needs to be done.

This study has for the first time in South India provided objective evidence of the alarmingly high incidence of inner ear malformations in children with hearing loss possibly due to the genetic constitution induced by consanguinity. Even though the prevalence and the spectrum of malformations described in this study are from a single institution, the pattern is expected to be the same for the rest of Indian population. Hence a multicentric large scale prospective epidemiological research has been initiated with active participation from our institution to establish the severity of this entity and to plan appropriate management protocols in future.

#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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