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**RESEARCH ARTICLE**

# A STUDY OF UNILATERAL COCHLEAR IMPLANT IN PRELINGUAL DEAF CHILDREN

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**ABSTRACT**

**Introduction:-** Hearing impairment, is defined as a partial or total inability to hear. Hearing Loss is broadly divided into Sensorineural Hearing Loss and Conductive Hearing Loss. Spoken language development is often delayed in children with unaddressed hearing loss. A **Cochlear implant** (**CI**) is surgically implanted electronic device that provides a sense of sound to a person with severe to profound sensorineural hearing loss. The process of cochlear implantation starts with early identification of hearing impairment by neonatologist-paediatrician. Subsequently the physicians, paediatrician, psychologist and radiologist evaluate the patient. Post implantation rigorous therapy is required to enable the child to join the mainstream. The whole process is a team effort and it requires constant motivation on the part of parents. Cochlear implant has to be individualized because every child is unique. Generally, at the functional level, children with inner ear damage are considered for cochlear implantation. The present study aims to assess the experience of the Department of Otorhinolaryngology of R.N.T. Medical College, Udaipur with patients who will undergo CI surgery. **Materials and Methods:** it is a prospective cohort study. The study population in our study were patients attending the Department of ENT in RNT Medical College, Udaipur, Rajasthan during Two and half years from June 2016 to December 2018. A total of 10 cochlear Implants were done at our institute. Data was recorded on a Performa. The data analysis was computer based; SPSS-22 will be used for analysis. Data were shown as Mean±SD. For *p*-value <0.05 was considered as significant. **Results:-** cochlear implantation definitely provides a satisfactory hearing and speech ability to a hearing disabled child.



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## INTRODUCTION

Hearing loss, deafness, hard of hearing, anacusis, or hearing impairment, is defined as a partial or total inability to hear. Hearing loss is caused by many factors, including: genetics, age, exposure to noise, illness, chemicals and physical trauma. Hearing Loss is broadly divided into Sensorineural Hearing Loss and Conductive Hearing Loss. Conductive Hearing Loss Occurs when the parts of Ear responsible for transmission of Sound malfunctioned while the sensorineural Hearing Loss occurs when neurological aspect of Hearing is affected. As a result of the hearing loss the patient ability to communicate is affected as hearing and speech forms backbone of communication. Spoken language development is often delayed in children with unaddressed hearing loss (WHO, 2008). Early intervention in the form of surgical management can improve the linguistic and educational outcomes for the child (About Hearing Loss, 2016).

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A Cochlear implant (CI) is surgically implanted electronic device that provides a sense of sound to a person with severe to profound sensorineural hearing loss. Cochlear implants, like the human hair cell, receive mechanical sound energy and convert it into a series of electrical impulses. The brain adapts to the new mode of hearing, and eventually can interpret the electric signals as sound and speech. The implant has two main components. The outside component is generally worn behind the ear, but could also be attached to clothing, for example, in young children. This component, the sound processor, contains microphones, electronics that include DSP chips, battery, and a coil which transmits a signal to the implant across the skin. The inside component, the actual implant, has a coil to receive signals, electronics, and an array of electrodes which is placed into the cochlea, which stimulate the Cochlear nerve (Yawn, 2015; Balkany, 2018). The process of cochlear implantation starts with early identification of hearing impairment by neonatologist-paediatrician. Subsequently the physicians, paediatrician, psychologist and radiologist evaluate the patient. After evaluation potential Cochlear Implant recipient are identified based on guidelines by FDA.

Post implantation rigorous therapy is required to enable the child to join the mainstream. The whole process is a team effort and it requires constant motivation on the part of parents. Multiple Studies shows that the acquisition of language and communication skills in individuals with deafnessthrough cochlear implant (CI) confers greater benefits compared to conservativehearing aids (Niparko, 2000). Cochlear implant has to be individualized because every child is unique. Generally, at the functional level, children with inner ear damage are considered for cochlear implantation. The present study aims to assess the experience of the Department of Otorhinolaryngology of R.N.T. Medical College, Udaipur with patients who will undergo CI surgery.

## MATERIALS AND METHODS

**Study population:** The study population in our study were patients attending the Department of ENT in RNT Medical College, Udaipur, Rajasthan.

**Duration of study:** Two and half years from June 2016 to December 2018. A total of 10 cochlear Implants were done at our institute.

**Inclusion criteria**

- Severe to Profound Hearing loss in Bilateral Ear.
- Below 6 years of age
- Patient consenting to participate in the study.

**Exclusion criteria**

- Agenesis of Cochlea
- Absent cochlear Nerve
- Age above 6 years
- Active infection in ear
- Patient not consenting to participate in the study.

**Methods of collection of data:** The study was a prospective cohort study and it was carried out by means of information obtained from patients who underwent CI Surgery in the Department of Otorhinolaryngology of RNT Medical College, Udaipur. The various parameters that were analysed were duration of deafness, type of implant, classification of the deafness, age, and gender. The patients were selected for cochlear implantation after thorough evaluation of their type and degree of hearing loss, lack of benefit with hearing aids, radiological analysis of the temporal bone–cochlear anatomy for feasibility for implantation, adequate parental motivation for habilitation and the possibility/accessibility of pre-and post implantation habilitation/therapy and programming centers.

**Data analysis**: Data was recorded on a Performa. The data analysis was computer based; SPSS-22 will be used for analysis. Data were shown as Mean±SD. For *p*-value $<0.05$ was considered as significant.

**Selection and evaluation of patients:**

**Audiologic assessment:** Candidacy for cochlear implantation relies heavily upon the audiologic evaluation. In young children a battery of tests are being employed to evaluate the need for cochlear Implant. Some common tests being employed are as follows-

**Auditory Brainstem Response Testing (ABR):** This is the test of choice in children under 6 months of age. The ABR follows neuromaturation of the central auditory nervous system and shows wave latency changes until about 18 months of age.

**Otoacoustic Emission Testing (OAE):** OAEs give us the ability to view the functioning of the cochlea, although not without contribution of the middle ear. OAEs may be absent due to middle-ear dysfunction, resulting in inability of the emission to be transmitted, or to sensory hearing loss affecting production of the emission. Prediction of hearing levels is not possible by measuring OAE. OAFs are classified according to whether there is a stimulus used to record them.

Spontaneous Otoacoustic Emissions (SOAEs) are recorded when no stimulus is present.

Evoked Otoacoustic Emissions (EOAEs) are recorded following a stimulus.

The two clinically useful EOAEs are:

Transient Evoked Otoacoustic Emissions **(TEOAEs)**,

Distortion Product Otoacoustic Emissions **(DPOAEs)**,

**Medical assessment:** A detailed history and examination was conducted to evaluate the cause of the hearing loss, including prenatal and perinatal history to assess for risk factors some of which include TORCH infections (toxoplasmosis, other [syphilis, varicella-zoster, parvovirus B19], rubella, cytomegalovirus [CMV] and herpes infections), teratogens, prematurity, low birth weight, low APGAR scores, hyperbilirubinaemia, sepsis, meningitis and the administration of ototoxic medications.

**Imaging protocols for cochlear implant:** Cochlear implant radiology is an indispensable part of cochlear implant work up. Candidacy can be finalized only after complete radiology of the temporal bone and brain has been performed. Radiological Imaging has immense role in pre-operative workup, surgical planning, implant selection and preparation for any surgical complication. After a definitive audiological diagnosis of bilateral severe to profound sensorineural hearing loss patients were advised to undergo both HRCT scan and MRI of temporal Bone (Mohamad Hasan Alam-Eldeen).

**HRCT Protocol:** Pathologies of mastoid as mastoid sclerosis, middle ear disease, otosclerosis, Paget's disease, post meningitic stenosis of the round window niche and evidence of labyrinthitis ossificans can be very well ascertained by a detailed HRCT of the temporal bone (Mohamad Hasan Alam-Eldeen).

**MRI Protocol:** MRI was useful in evaluating the soft tissue structures especially the status of fluid in the cochlear turns and the vestibular apparatus. MRI also helps in determining size of internal acoustic meatus and the cochlear aperture, the presence or absence of cochlear nerves and the pathological conditions like early stages of labyrinthitis ossificans (Kranti Bhavana and Subhash Kumar, 2000).

**Post–Operative Evaluation:** It was done using the **Revised Category of Auditory Performance (CAP) score** described by The Shepherd Centre based on Nottingham CI Programme, 1995.

**Table 1. Comparision Of Mean Score Of Different Scales At Different Time Duration**

| Scales | Pre-operative | At 6 months | At 12 months |
|---|---|---|---|
| Mean CAP Score | 0.5±0.5 | 3.8±0.6 | 7.875±0.6 |
| Mean MAIS Score | 1.9±1.92 | 25.3±1.27 | 32.25±0.66 |
| Mean SIR Score | 1.0±0.0 | 2.2±0.4 | 2.5±0.5 |

**Table 2. Revised CAP Score At Different Time Duration**

| CAP Score | Pre-op(no. of patients) | At 6 month (no. of patients) | At 12 month (no. of patients) |
|---|---|---|---|
| 0 | 5(50%) | -- | -- |
| 1 | 5(50%) | -- | -- |
| 2 | 0 | -- | -- |
| 3 | -- | 3(30%) | -- |
| 4 | -- | 6(60%) | -- |
| 5 | -- | 1(10%) | -- |
| 7 | -- | -- | 2(20%) |
| 8 | -- | -- | 5(50%) |
| 9 | -- | -- | 1(10%) |
| Not available | -- | -- | 2(20%) |

**Table 3. SIR Scoring At Different Time Duration**

| SIR Score | Pre-op(no. of patients) | At 6 month (no. of patients) | At 12 month (no. of patients) |
|---|---|---|---|
| 1 | 10(100%) | 0(0%) | -- |
| 2 | -- | 8(80%) | 4(40%) |
| 3 | -- | 2(20%) | 4(40%) |
| 4 | -- | -- | -- |
| 5 | -- | -- | -- |
| Not available | -- | -- | 2(20%) |

**Table 4. Complications**

| Categories of Complication | Number of Patients |
|---|---|
| Facial Nerve Paresis | 0 |
| Chorda Tympani Syndrome | 0 |
| Electrode Extrusion | 0 |
| Wound Infection | 0 |
| Tympanomeatal flap rupture | 1 |

The ability to discriminate and understand speech with or without lip reading was assessed and the results were categorized accordingly and a score was given.

Similarly the **Speech Intelligibility Rating (SIR) of O' Donoghue** was utilized to measure the outcome of cochlear implantation with respect to speech, measuring' the intelligibility of speech and the quality, which might be recognizable by the listener (O'Donoghue, 2002; O' Donoghue, 2001).

**Surgical preparation & technique:** All patients were operated Under General Anaesthesia following Standard protocol.

**Post-operative management:** A mastoid dressing was applied after the operation and kept till 10 days. IV antibiotics were given till 10 days. After 10 days stitches were removed and further mastoid dressing was done. The patients were discharged thereafter.

## RESULTS

**Post operative Hearing and speech Assessment:** Post Cochlear Implantation Auditory receptive skills of the patients and subsequently speech developmentere measured. The Scales used were Revised Category of Auditory Performance (CAP) Scores, Speech Intelligibility Rating (SIR) Scale, MAIS Questionnaire.

Out of the 10 Patients that were operated at our institute only one patient had complication i.e. rupture of tympanomeatal flap which is a minor perioperative complication.

## DISCUSSION

For Profound Hearing Loss Cochlear implant is now considered a gold standard treatment. Various studies have taken place at varied universities and medical research centres to prove the efficacy of cochlear implant in treatment of deafness and its rehabilitation. Though the candidacy of cochlear implant has widened to include pre-lingual deaf, post -lingual deaf children and adults and profoundly deaf adults.
However in the present study, the cohort consists of patient of age group below 6 years of age and prelingualy deaf. The mean age of implantation is 4.851±1.265 years. Many authors have shown better outcome from the point of auditory performance and speech intelligibility in congenitally deaf children who had cochlear implantation in early childhood comparing to those operated in adulthood. A study done by Erin Schafer *et al.* 2016 in which a systematic review was performed on peer-reviewed research pertaining to factors influencing speech intelligibility of children with cochlear implants. The study concluded that the age at implantation proved the most important factor influencing a child's speech intelligibility. Earlier the child is implanted better is his speech intelligibility. Currently, the FDA has approved cochlear implantation in 12 months and older children, but some centres are implanting infants as young as 6 months.

In the present study The Torch screening of the cohort for etiology of sensor neural deafness shows Cytomegalovirus IgG in 4 (40 %) and HSV 2 in 2 (20 %) while 4 were of etiology Other than congenital infections. In the present Study the post cochlear implantation evaluation was done using Revised CAP Scale, MAIS Questionnaire and SIR scoring. The Scores for the cohort were obtained preoperatively then at first switch on followed by on 6 months and 12 months respectively. In the Present Study the mean pre op Revised CAP Score was 0.5 ±0.5 which means that that the cohort under study was "unaware of the environmental sounds." At the end of 6 months the revised CAP Score was raised to 3.8±0.6 which means that the cohort started to identify some environmental sounds and understood some words with addition per formatives. At the end of 12 months the revised CAP Score raised to 7.875±0.6 which means that the cohort started to respond appropriately to simple Questions. (Table-1) The percentage of children at level 3 or higher increased from 0 before surgery to 60 % and 80 % at 6 and 12 months respectively after implantation. According to a study by Huiqun Zhou el al there was and progressive increment in the mean CAP Score from 0 preoperatively to 4 at the end of 6 months and 5 at 12 months. The percentage of children at level 3 or higher increased from 0 before surgery to 73.7%, 89.5% at 6 and12 months after implantation (Mariana Cardoso Guedes).

In another study by V Sharvanan *et al.* (2013), The CAP Score which was found to be 3.62 ± 0.983. So on an average child implanted after 3 years are only able to "discriminate between speech sounds" at the end of 1 year after rehabilitation. This however when compared to the child implanted below 3 Years of age in this study showing a mean CAP Score of 5.17± 0.072. It was found to be statistically significant. Implies that earlier implanted children have much better auditory perception compare to late implanted children. However result of the study in earlier age group could not be corroborated with ours as our cohort consists of patients of age above 3 years. In the present study the Mean SIR Score preoperatively was 1.0±0.0which means that the normal speech of the patient was unintelligible to guardians. (Table-3) At the end of 6 months the Mean SIR Score was raised to 2.2±0.4which further increased to 2.5±0.5at the end of 12 months. It means that intelligible speech is developing in single words when context &lip reading cues are available (Table-1).

According to a study by M. Bakhshaee *et al.* (2007) there was gradual increment in the mean SIR Score of his cohort. Preoperatively the cohort has Mean SIR Score of 1 which increased to 2 at the end of 6 months and further increased to 3 at the 12 months. The increase in ratings each year until the third year was statistically significant (from pre-implantation to year 1, p=0.000; from year 1 to year 2, p=0.000; and from year 2 to year 3, p=0.000). The children's auditory performance and speech development under the age of 4 was significantly better than those over 4 at the time of implantation ($p<0.05$). Another study done by Anjan Das et al shows themean SIR score at 0 month, 6 months, 12 months of surgery was 1,1.7 and2.6 respectively (Reddy). In the present study the Mean MAIS score preoperatively was 1.9±1.92 which increased to 25.3±1.27 at the 6 months and 32.25±0.66 at the end of 12 months. In a study by Dr. Rajesh Vishwakarma *et al.* shows pre-op average MAIS questionnaire score was ≤ 8/40 in all implanted age groups with scores increasing over time to attain average score ranging from 34 to 37.44/40 in all implanted age groups after two years of implantation. The score increased significantly ($p<0.05$) in all implanted age groups from pre- to post- CI. There was positive effect of time with scores increasing on every follow-up (Rajesh Vishwakarma, 2015). Another study by Dr. V Sarvanan shows that the average Meaningful Auditory Integration Scale in children implanted before the age of 3 years was 34.88 with a standard deviation of ±2.309 and in children whose age at the time of implantation was 3-6 years showed an average score of 27.38 with a standard deviation of ±6.2686 (Tobey, 2003). The results of these studies are comparable to our study which shows that there is gradual improvement in the MAIS score with gradual time and the implanted patient is using his environmental sounds in more meaningful way after surgery when compared to preoperatively. In our study out of 10 patients, only one patient (10 %) experience perioperative complication of tympanomeatal flap rupture. No major complication like electrode misplacement, electrode extrusion, meningitis was observed over the duration of study (Table-4). A study by Jonas Jeppesen *et al.* shows that three most common major complications were wound infection (1.6%), permanent chorda tympani syndrome (1.6%) and electrode migration/misplacement/accidental removal (1.3%). Permanent facial nerve paresis occurred following one implantation (0.3%). Transient chorda tympani syndrome (30.8%), vertigo/dizziness (29.5%) and tinnitus (4.9%) were the most frequent minor complications (Jonas Jeppesen and Christian Emil Faber, 2013).

**Limitations**

Apart from small sample size and short period of study, result of this study cannot be corroborated on patients with cochlear malformation like common cavity deformity, cochlear hypoplasia, and incomplete partition, vestibular aqueduct enlargement because all the patients that were operated had normal cochlear anatomy.

**Conclusion**

On the basis of the present study it was found that there is a definite improvement in the parameters used to assess hearing and speech at the end of 1 year when compared with preoperative levels. Cochlear implantation provides satisfactory hearing and speech ability to congenitally deaf children who do not benefit from traditional hearing amplification and speech therapy.

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