

Outcome of the unilateral cochlear implant in prelingual deaf children in Mewar region of Rajasthan

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Abstract

Introduction: A Cochlear implant (CI) is surgically implanted electronic device that provide a sense of sound to a person with severe to profound sensorineural hearing loss. It helps in social rehabilitation of the patient with 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.

Aim: The present study aims to evaluate the outcomes of cochlear implant in terms of hearing and speech gain, The Department of Otorhinolaryngology of R.N.T. Medical College, Udaipur with patients who will undergo CI surgery.

Materials and Methods: The prospective study (conducted between June 2016 to December 2018), the study population in our study were patients attending the Department of ENT in RNT medical college, Udaipur. A detailed history, including prenatal and perinatal history to assess for risk factors include TORCH infection and the various parameters that were analyzed like Audiological assessment (BERA, Otoacoustic Emission testing) duration of deafness, age, sex, type and degree of deafness, lack of benefit with hearing aids, radiological analysis (HRCT & MRI) of temporal bone and cochlear anatomy for feasibility for implantation.

Surgically cochlear implanted through Veria technique under all aseptic precautions.

Results: In our study mean age of implantation was 4.851 \pm 1.265. The mean revised CAP score at pre op-, 6months, 12months of surgery was 0.5 \pm 0.5, 3.8 \pm 0.6 and 7.875 \pm 0.6 respectively; the mean SIR score at pre-op 6 months, 12 months of surgery was 1.0 \pm 0.0, 2.2 \pm 0.4 and 2.5 \pm 0.5 respectively; the mean MAIS score at pre op-, 6 months, 12 months of surgery was 1.9 \pm 1.92, 25.3 \pm 1.27 and 32.25 \pm 0.66 respectively.

Conclusion: Intelligible speech and auditory performance of children who underwent prelingual cochlear implantation was improved compared to their pre-operative status. Cochlear implantation appears to be beneficial for pre-lingual deaf children in term of quality of life.

Keywords: Cochlear implant (CI), Revised CAP (Categories of auditory perception) scale, speech intelligibility rating (SIR), meaningful auditory integration scale (MAIS)

Introduction

Hearing is one of the basic five senses. It is defined as ability to perceive sound by detecting vibration via an organ such as the ear. Deafness occurs when patient is unable to perceive sound. A hearing impairment or hearing loss is a full or partial decrease in the ability to detect or understand sounds. Hearing Impairment is multifactorial in nature with multitude of environment and genetic factors [1].

As a result of the hearing loss the patient ability to communicate is affected as hearing and speech form backbone of communication. Spoken language development is often delayed in children with unaddressed hearing loss [2].

Impact of Hearing loss can itself express in the form of feeling of loneliness, isolation, and frustration, particularly among older people and if sufficient mental and psychological support is not provided then it can have detrimental effect on the overall health of the patient.

Early detection and timely intervention are very crucial to minimizing the impact of hearing loss on a child's development in infant and young children with hearing loss, it is achieved via infant hearing screening programs. Early intervention in the form of surgical management can improve the linguistic and educational outcomes for the child [3].

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 electrode which is placed into the cochlea, which stimulate the cochlear nerve [4, 5].

There are four manufacturers providing cochlear implants for use: Advanced Bionics, Cochlear, MED-EL and Oticon Medical (previously Neurelec). All of the device shares a similar basic design.

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

The study was conducted between June 2016 to December 2018. The study was a prospective cohort study and it was carried out by means of information obtained from patients who underwent CI Surgery in Department of Otorhinolaryngology of R.N.T. Medical College Udaipur. The various parameters that were analyzed 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.

Inclusion criteria

1. Severe to profound Hearing loss in Bilateral Ear.
2. Below 6 years of age.
3. Patient consenting to participate in the study.

Exclusion criteria

1. Agenesis of Cochlea.
2. Absent cochlear Nerve.
3. Age above 6 years.
4. Active infection in ear.
5. Patient not consenting to participate in the study.

Surgical preparation

Anaesthesia: All patients were operated under general anaesthesia.

Surgical technique

Preparation

To isolate the surgical field hairs were shaved completely over the scalp.

The patient lay in supine position with head turned away from the surgeon, close to the edge of the table on the operating table, so that the operating ear faced upwards. The ear and adjacent area were scrubbed using 7.5% povidone Iodine surgical scrub followed by paints using 10% Povidone Iodine solution and spirit. The process was repeated three times. The patient was then draped using sterile surgical towels.

Infiltration

It was done 10-12 minutes prior to the incision. 2% xylocaine with 1:200000 adrenaline was used for infiltration at the incision site.

The bony cartilaginous junction is identified and an endaural incision was made in the canal. An inverted C shaped incision was made in the region of the squamous part of the temporal bone meeting the endaural incision. A posteriorly based skin flap was elevated which is supplied by the posterior auricular artery. The underlying temporalis muscle flap (anterior based flap) was elevated to expose the underlying temporal bone. The incision for this flap will lie underneath the bulk of the skin flap thereby preventing the exposure of the implant and electrode to the exterior. It has its blood supply from the superficial temporalis artery. Han's flap which is the posteriorly based periosteal flap is located in the suprimeatal junction. This covers the electrode in the suprimeatal well. The flap is supplied individually promoting better healing and the implant lies secure beneath these flaps without crossing the incision ^[6].

Sub-periosteal pocket is made superiorly and anteriorly to house the implant and the ground electrode (if present separate from implant). The bed for the implant is marked in the most stable part of the squamous temporal bone. The bed was drilled to a depth where the endosteum is visualized. Margins of the bed were made at right angles which prevents the migration of the implant.

After identifying the suprimeatal line the suprimeatal well was drilled down not more than 1.5 mm deep keeping the edges of the well undermined. This keeps the electrode in-place. Canal wall incision were made and the tympanomeatal flap elevated. This gives a wider visualization of the round window and the promontory ^[7].

Canaloplasty was done in order to straighten the posterior canal wall. This prevents cholesteatoma occurrence in the implanted children. Drilling at the tympanic plate was avoided as it can obliterated the facial recess. The suprimeatal well and bed are connected by a groove which is undermined at its entry in the well.

The tunnel was made in the posterior canal wall using a specially designed hand piece which has a guide and cutting drill bit at a distance of 4 mm. The tunnel drilled is of 1.4 mm in diameter. While making the tunnel, the entry at the suprimeatal well is at 11 o' clock and exit medially in the middle ear is at 9 o' clock. Slow to and fro motion of the hand piece with rotation of drill bit at slow rotation is done which smoothens the tunnel as it progress. Good irrigation is required at there can be heat dissipation through the mastoid air cell system towards the facial nerve. The electrode passing through this tunnel is safely surrounded by bone on all sides separated from the mastoid air system. The obliqueness of the tunnel moves the line of drilling away from the origin of the chorda tympani nerve and also keep the direction of the tunnel parallel to the long process of incus. A total depth of 2mm of the tunnel from the posterior canal wall (1.4 mm diameter of the tunnel and a depth of <0.5mm) keeps it in the posterior canal wall which keeps the facial nerve safe ^[7].

Identify the round window niche and the basal turn of the cochlea. The junction where these structures meet is marked and cochleostomy is done Antero inferior to the round window niche. The c cell system should not be mistaken for the niche as it can lead to drilling the otic capsule and entry into the carotid canal. Cochleostomy is done using the 1.2mm diamond burr and a 0.8 mm straight diamond burr. The anterior margin of the cochleostomy is undermined and the posterior margin beveled which enables smooth transition of the electrode into the cochlea [6,7].

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.

Observation & Results

Table 1: Gender distribution

Gender	Number of patients	Percentage
Male	7	70%
Female	3	30%
Total	10	100%

In our study out of 10 patients 7 were boys and 3 were girls.

Age distribution: In our study the patients operated were of different ages when operated. The mean age of surgery was 4.851+/-1.265 years. The youngest child was of 4 year and 8 months of age at implantation while the oldest child was of 5 years and 3 months of age.

Table 2: TORCH Screening

Name of the Test	Patient Positive	Percentage
CMV IgG	4	40%
Rubella IgG	0	0%
HSV1 IgG	0	0%
HSV2 IgG	2	20%
Unknown Etiology	4	40%

In present study testing for the etiology of the deafness was done, result indicate that 40% patients were infected with CMV prenatally, 20% were infected with HSV type 2 prenatally, and 40% were of unknown etiology.

Table 3: Revised CAP Scoring

Preoperatively	CAP Score	No of patient in respective score category	Percentage
	0	5	50%
	1	5	50%
	2	0	0%
at 6 months	3	3	30%
	4	6	60%
	5	1	10%
at 12 Months	7	2	20%
	8	5	50%
	9	1	10%
	Not available	2	20%

Before Cochlear Implant Revised CAP Score was 0 in 50% patients, and was 1 in 50% patients.

6 months after surgery, revised CAP Score was 3 in 3 patients (30%), 4 in 6 patients (60%) and was 5 in 1 patient (10%).

12 months after surgery, revised CAP Score was 7 in 2 patients (20%), 8 in 5 patients (50%) and was 9 in 1 patient (10%). Data was not available for 2 patients at the end of 12 month.

Table 4: SIR Score

Preoperatively	Sir Score	No. of Patient in Respective Category	Percentage %
		1	10
At 6 month	1	0	0
	2	8	80
	3	2	20
At 12 month	2	4	40
	3	4	40
	NA	2	20

6 months after surgery, SIR Score was 2 in 8 patients (80%) and was 3 in 2 patients (20%).

12 months after surgery, SIR Score was 2 in 4 patients (40%), 3 in 4 patients (40%) and was not available in 2 patients (20%). Non availability was due to absence of child at that scoring time.

Table 5: Mean Score

Mean score	Pre op	6 month	12 month
Mean revised 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

As the above table shows that the mean pre op Revised CAP Score was 0.5+/-0.5 which means that the cohort under study was “unaware of the environmental sounds”. At the end of 6 months the revised CAP Score raised to 3.8+/- 0.6 which means that the cohort starts to identify some environmental sounds and understand some words with addition performative. At the end of 12 months the revised CAP Score raised to 7.875+/-0.6 which means that the cohort starts to respond appropriately to simple question. It clearly indicated that the auditory perception skill is gradually increasing over time duration with the aid of Cochlear implantation.

The Mean MAIS Score preoperatively was 1.9+/- 1.92 which increased to 25+/- 1.27 at the 6 months and 32.25+/- 0.66 at the end of 12 months. The gradual improvement in the MAIS Score shows that with gradual time the implanted patient is using his environment sound in more meaningful way than earlier. The Mean SIR Score preoperatively was 1.0+/- 0.0 which means that the normal speech of the patient was unintelligible to guardians. At the end of the months the Mean SIR Score raised to 2.2+/- which further increased to 2.5+/- 0.5 at the end of 12 months. It means that intelligible speech is developing in single words when context and lip-reading cues are available.

Discussion

For Profound Hearing Loss Cochlear implant is now considered a gold standard treatment. It has taken several decades of research and hard work of scientist to reach this stage and it is continuing till this day. Various studies have taken place at varied universities and medical research centers 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 patients of age group below 6 years of age and

prelingually 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.

In the present study the number of female patients was 3 (30%), and the number of male patients was 7 (70%).

In a study by Dr. Ayse Sanem Sahli, 60% (N=9) of the children were boys and 40% (N=6) were girls. Average chronological age of the children was approximately 44 months (range 12-56 months).

In the present study the Torch screening of the cohort for etiology of sensorineural deafness shows Cytomegalovirus IgG in 4 (40%) and HSV 2 in 2 (20%) while 4 were of etiology other than congenital infections.

M.V.V. Reddy *et al.* 2004, conducted Interview based prospective study in children below 14 years of age with hearing loss which showed the results on the type of the hearing impairment are presented in their study; Out of 743 children with hearing loss 18.57% were found with syndromic hearing impairment and 81.73% had isolated (non-syndromic) deafness. The results on etiology of hearing loss in children with deafness shows that in 15.22% of children, deafness was inherited, in 13.77% it was acquired, and in 71.01% the etiology was unknown^[8].

In the present study all of 10 patients had complete insertion of electrodes and showed a positive intraoperative neural response, which means that the auditory nerve responded to the first electrical stimulation of the cochlear implant.

A study by Guedes *et al.* showed that adult patients who showed intraoperative telemetry responses had better results in their pre-operative speech perception tests, but this relationship was not statistically significant among children. In the current study, all patients were category 0 in pre-operative SIR but all of them showed positive neural response test intra-operative^[9].

In the present study the post cochlear implantation evaluation was done using Revised CAP Scale, MAIS Questionnaire and SIR Scoring. The score 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 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 performatives. 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 question. 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 *et al.* there was and progressive increment in 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 and 12 months after implantation.

Yang *et al.* studied 26 children in 2004 to compare results of cochlear implantation between normal deaf children and those with other disorder such as cerebral palsy. The mean CAP Score 1 and 2 years after surgery was 3.93 and 5.86 in the normal group and 2.5 and 4.17 in those with cerebral palsy or other mental anomalies, respectively^[10, 11].

In the present study the Mean SIR Score preoperatively was 1.0+/- 0.0 which means that the normal speech of the patient was unintelligible to guardians. At the end of 6 months the mean SIR Score was raised to 2.2 +/- 0.4 which further increased to 2.5 +/- 0.5 at the end of 12 months. It means that intelligible speech is developing in single words when context & lip-reading cues are available.

According to a study by M. Bakhshae *et al.* 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 rating each year until the third year was statistically significant (from pre-implantation to year 1, $p=0.00$; from year 1 to year 2, $p=0.00$ and from year 2 to year 3, $p=0.00$). 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 ($p<0.05$). Study also indicated that age is a major factor in the development of auditory skills after surgery and better result were seen in those children who underwent surgery before the age of four while poor results were reported in those who were operated after that [12].

Another study done by Anjan Das *et al.* shows the mean SIR Score at 0-month, 6 month, 12 months of surgery was 1, 1.7 and 2.6 respectively [13].

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 [14].

Another study by Dr. V Sarvnan 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 [15].

The result of these studies is 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.

Summary & Conclusion

In our study a total of 10 children were implanted with cochlear implant. This is a prospective study which followed the implanted children over a period of 1 year following implantation and their auditory perception and speech intelligibility was assessed over this period.

All children had undergone a thorough preoperative evaluation of general health and audiological parameters.

The mean age of implantation was 4.851 ± 1.265 .

Post operatively the variables recorded during the follow up for the purpose of our study included Revised CAP Scale, SIR Scale, MAIS Scale.

The Mean Revised CAP Score raised from 0.5 ± 0.5 preoperatively to 3.8 ± 0.6 at 6 months and 7.875 ± 0.6 at 12 months i.e. at the start of study the cohort under study was unaware of the environmental sound and with gradual use of cochlear implant at the end of 12 months the cohort starts to respond appropriately to simple question.

The mean SIR Score raised from 1.0 ± 0.0 preoperatively to 2.2 ± 0.4 at 6 months and 2.5 ± 0.5 at 12 months i.e. at the start the speech of the patient was intelligible to general population and with gradual use of cochlear implant intelligible speech start developing in single words when context & lip-reading cues are available.

Similarly, the mean MAIS Score raised from 1.9 ± 1.92 preoperatively to 25.3 ± 1.27 at 6 months and 32.25 ± 0.66 at 12 months i.e. the patient start using his environmental sounds in more meaningful way when compared to start of speech therapy.

Statistically significant difference was found between the preoperatively and postoperatively Revised CAP, SIR and MAIS Score.

Cochlear implantation provides satisfactory hearing and speech ability to congenitally deaf children who do not benefit from traditional hearing amplification and speech therapy.

Thus, Cochlear implant surgery provides a hearing disabled child to lead a near normal life at par with peers of his age with normal hearing.

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