# COCHLEAR IMPLANTATION IN PATIENTS WITH INNER EAR MALFORMATIONS

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**Abstract-** Performing cochlear implantation in patients with inner ear malformation has always been a matter of dispute. This study was designed to analyze the operative findings, complications, and post-operative performance of patients with inner ear anomalies who underwent cochlear implantation. Six patients with inner ear malformations underwent implantation in our academic tertiary referral center from 1997 to 2002. The average follow-up period was 27 months. Malformations included one incomplete partition, one common cavity, one narrow internal acoustic canal (IAC) in a patient with Riley-Day syndrome and 3 cases of large vestibular aqueduct. All received multi-channel implants either Nucleus 22 or Clarion device. Facial nerve was anomalous in 2 cases. CSF gusher occurred in 4 patients, which was controlled with packing the cochleostomy site. In all cases, the full length of electrode array was inserted, except one with Mondini's dysplasia where insertion failed in the first operation and was referred to another center for a successful surgery on the opposite ear. No other surgical complications were encountered. In 4 cases, all the 22 electrodes could be activated. All patients showed improved hearing performance after implantation. Four showed open-set speech recognition. The one with narrow IAC showed improved awareness to environmental sounds. In the other case (common cavity), the perception tests could not be performed because of very young age. Cochlear implantation in patients with inner ear malformations is a successful way of rehabilitation, although complications should be expected and auditory responses may be highly variable and relatively moderate.

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**Key words:** Cochlear implant, inner ear malformation, Riley-Day syndrome, speech perception tests

## INTRODUCTION

Cochlear implantation is an accepted method of auditory rehabilitation for profoundly hearing-impaired children who do not derive benefit from amplification. Cochlear malformations have been reported to occur in approximately 20% of children with congenital sensorineural hearing loss (1). Children with inner ear developmental malformations present a significant challenge to even the most experienced clinician. In fact, until recently, many cochlear implant centers deferred implantation in these children because of uncertainty regarding surgical feasibility and performance expectations (2). Surgeons operating on a malformed inner ear should not only be aware of an aberrant facial nerve, but also of a possible CSF fistula. Whereas the former can be diagnosed preoperatively on CT and MRI studies, the latter will become noticeable just at the moment when the cochleostomy is performed: CSF that has traversed the peri-lymyphatic space will 'gush' out of the drilled holes, hence the name 'Gusher ear' (3). Expectations from auditory performance after cochlear implant in patients with inner ear malformations are relatively less than those without. This may be due to the substantially reduced

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population of spiral ganglion cells (4-6). Jackler *et al.* (7) cautioned that a narrow IAC suggests absence or severely decreased nerve fibers of the auditory nerve and thus a relative contraindication to cochlear implantation, a recommendation reiterated by Shelton *et al*. (8). Detailed data regarding performance could not be abstracted from the literature. However, it is clear that the performance has a direct relationship with the duration of device use and the time period that has elapsed from implantation. The overall hearing results of patients with inner ear malformations are slightly less but comparable with those of same age and experience who have no inner ear dysplasia. Between 1997 and 2002, more than 320 patients have received cochlear implant at the Amir Alam Hospital of Tehran University of Medical Sciences. Of this considerable number of patients, only 6 had congenital malformations of the inner ear. The purpose of this descriptive multi-case series and interventional study (before and after trial) was to analyze the surgical and audiologic outcome of patients with inner ear malformations who underwent cochlear implantation.

# MATERIALS AND METHODS

## Patients

The patient population included six children with varying degrees of inner ear malformations who underwent implantation at ages ranging from 10 months to 12 years. The mean age of operation was 51 months. Of the six children who were operated 5 were girls and one was a boy. The average follow-up period was 27 months. Informed consent was otained from both of the children's parents before the operation. A separate consent was taken for participating in this study from the parents as well.

Basic demographic data of all children are presented in table 1.

Preoperative evaluation included otologic examination, audiologic evaluation and a high resolution computed tomography (CT) of temporal bone as well as psychological assessment. Roentgenographic abnormalities were classified according to the scheme of Jackler *et al*.(1).

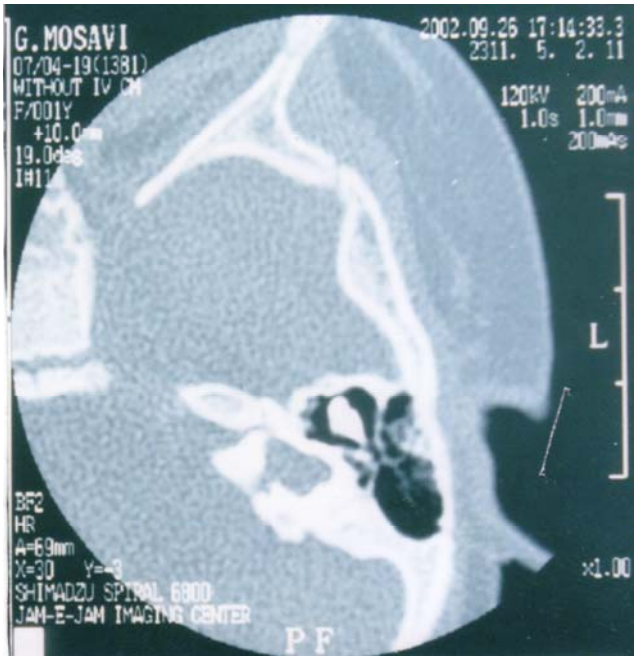
Inner ear anomalies included one common cavity deformity (Fig. 1), one incomplete partition or classic Mondini's cochlear dysplasia, 3 patients with enlarged vestibular aqueducts (Fig. 2), and one narrow IAC (Fig. 3) in a patient with Riley-Day syndrome or familial dysautonomia who had a positive promontory test.

All children were prelingually deaf, except one (patient 3) who had classic enlarged vestibular aqueduct syndrome; including a mild hearing loss noted at birth that progressed over many years to complete deafness.

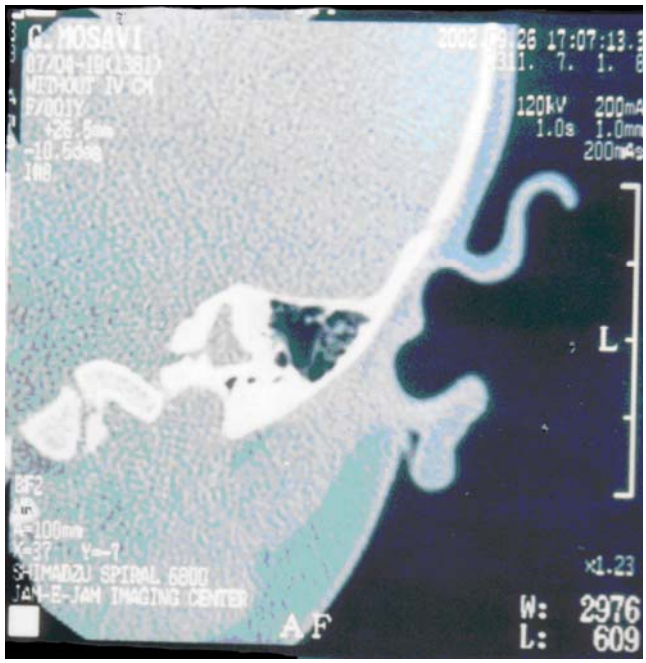
**Table 1.** Basic information and surgical findings of the patients

| No | Sex | Inner Ear Anomaly | Age at Sx | Date of Surgery | Side of Sx & Prosthesis | Approach | Surgical Findings & Complications |
|---|---|---|---|---|---|---|---|
| 1 | F | LVA | 26m | Apr. 1999 | L Clarion | Facial recess, cochleostomy | CSF Gusher: controlled with packing |
| 2 | F | Incomplete partition | 27m | Dec. 2000 | L | Facial recess, cochleostomy | Electrode array didn't go into the cavity through the cochleostomy site |
| | | (Mondini's dysplasia) | 33m | Jun. 2000 | R Nucleus 22 | Facial recess, cochleostomy | Was done in Belgium |
| 3 | F | LVA | 12y | Apr. 2000 | R Nucleus 22 | Facial recess, cochleostomy | Chorda tympani was placed more medial. CSF Gusher: controlled with packing |
| 4 | M | Narrow IAC | 5.5y | Jan. 2001 | R Clarion | Facial recess, cochleostomy | Anomalous stimulation of facial nerve. Normal facial nerve anatomy |
| 5 | F | LVA | 30m | Feb. 2001 | R Clarion | Facial recess, cochleostomy | CSF Gusher: controlled with packing |
| 6 | F | Common Cavity | 10m | Nov. 2002 | L Nucleus 22 | Trans-mastoid labyrinthotomy | Facial nerve was placed much more anteriorly & was not exposed during the procedure. Stapedial crura were directed vertically. Persistent stapedial artery present. A round bony capsule protruding from middle ear to the mastoid cavity. CSF Gusher: controlled with packing. Anomalous stimulation of facial nerve. |

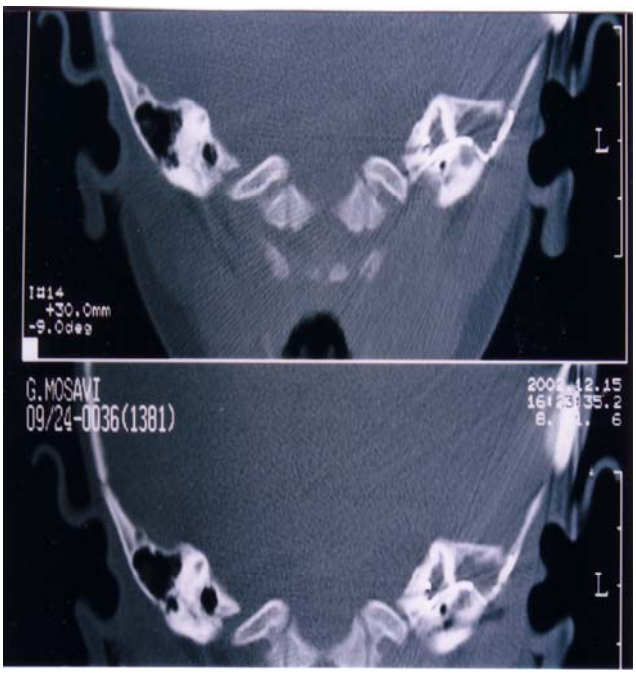
Abbreviations: LVA, enlarged vestibular aqueduct; IAC, internal acoustic canal; Sx, surgery; CSF, cereberospinal fluid.



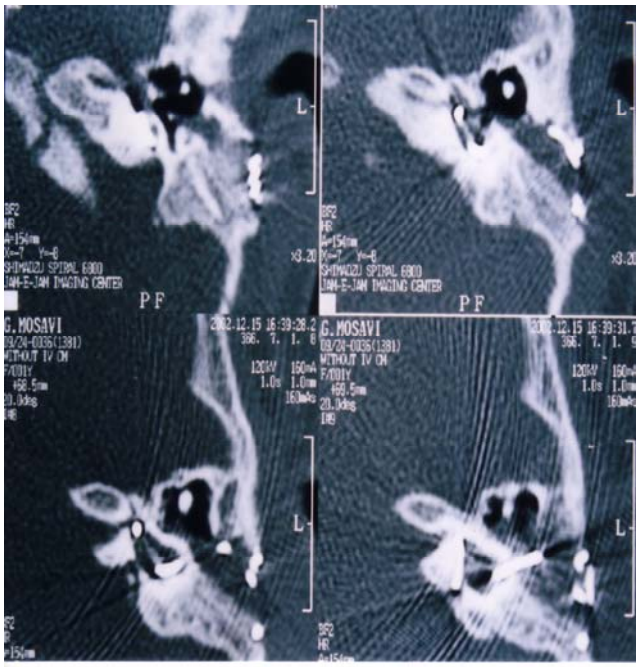

Preoperative axial CT scan.




Preoperative coronal CT scan.




Postoperative coronal CT scan.




Postoperative axial CT scan.

**Fig. 1.** Common cavity

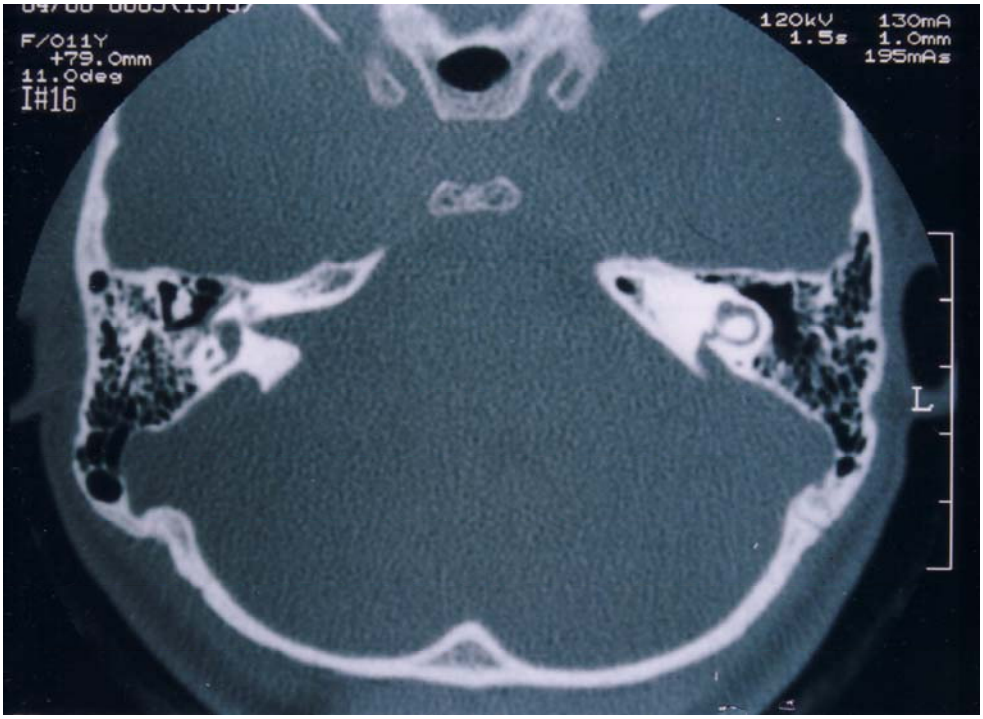



**Fig. 2.** Enlarged vestibular aqueduct. Axial CT-scan

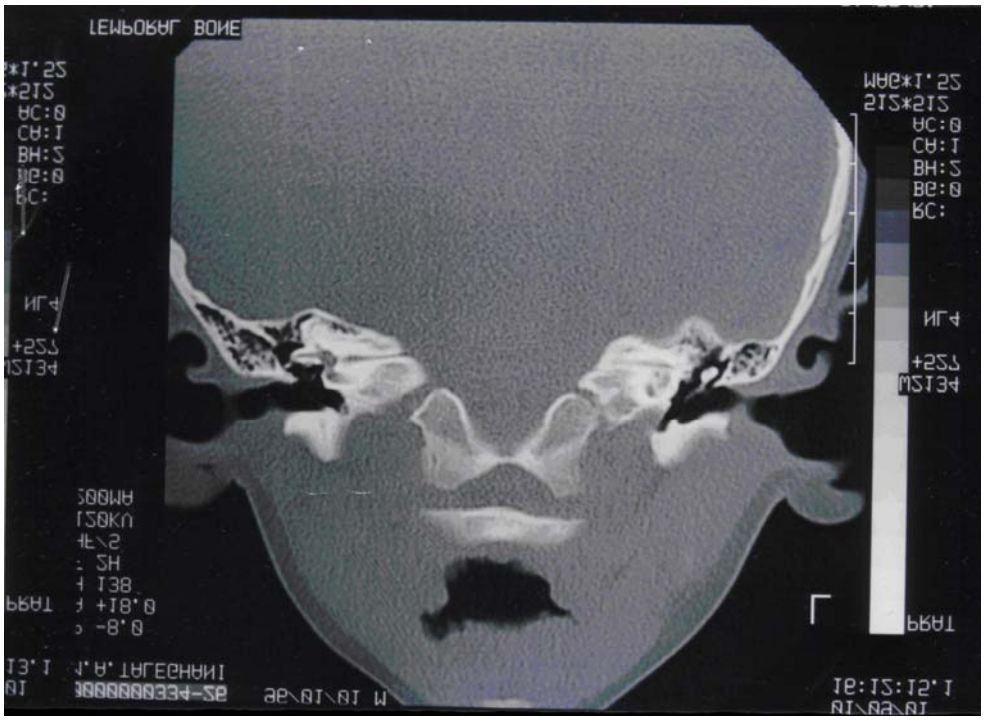


**Fig. 3.** Narrow IAC Riley-Day syndrome. Coronal CT scan

Informed consent was obtained for each patient before the surgical procedure. Decision for surgery of forth patient with narrow IAC and Riley-Day syndrome (familial dysautonomia) was taken in the view that the child had severe corneal ulcers as well and his vision had been severely compromised by the time of surgery.

Having a positive promontory test despite knowing the risks and limitations and explaining them to the parents, the surgery was done to give the child some awareness of the surrounding environment and its dangers. However a relatively poor performance was expected.

## Surgical Procedure

Overall, 3 left ears and 3 right ears were operated on, but patient number 2 had a second operation on her other ear at another center due to unsuccessful attempt of insertion of the electrode array into the cochleostomy site in our center.

Half of the cases received Nucleus 22 multi-channel, and in the remaining half, a Clarion multi-channel device was used. The standard facial recess approach was chosen in all but one patient. For patient number 6 with the common cavity, trans-mastoid labyrinthotomy approach as described by McElveen *et al.* (9) was performed. This was done to avoid the high risk of injury to the facial nerve which was seen to be anteriorly placed on preoperative CT scan.

Post implantation auditory performance was measured using pure tone threshold audiometry and standard speech perception tests (closed set, GASP and open set) in Persian language. All children were seen at least once a month following initial activation for a total of 6 months and at 3 months intervals thereafter.

At the time of activation, thresholds and comfort levels were measured for each electrode. This information was used to develop an electrode "program" for every patient. This individualized program contains information regarding the appropriate levels of stimulation for each electrode in the array. The average thresholds for 3 mid-frequencies i.e., 500, 1000 and 2000 Hz were measured every 6 months after the activation of device and recorded.

Speech perception tests were carried out every 6 months post-implant using available speech perception tests in Persian language.

The closed set test included a number of questions with 4 choices of phonetically similar written words or their pictures.

Depending on the child's age, exposed vocabulary and reading abilities, a set of either 24 or 62 questions were chosen. Each question was presented twice, using live voice from behind the child. Pre-implant results for all the cases were at or below the chance score of 25%.

The GASP word, an open set speech recognition test having a list of 12 familiar words or pictures, each of them repeated twice (total 24) in a sheet which is given to the candidate just before the test for a glance and then taken away form the child. The 24 words are presented in a different order via a live voice from behind the head and he/she must repeat them following the examiner.

The open set word recognition test is usually administered to slightly older patients, using taped sound and is an auditory test without any visual clues. Child is required to repeat, sign or write the word presented.

A literature review of available articles was also performed to compare the results of this study with those in the literature.

# RESULTS

Surgical findings and postoperative audiologic performance of the patients are shown in tables 1 and 2.

**Case 1.** Her parents noticed delayed speech development of their child at the age of 15 months. Further investigations revealed bilateral profound sensorineural hearing loss. CT scan of the temporal bone showed enlarged vestibular aqueducts on both sides. She did not benefit from hearing aids and therefore cochlear implant was planned and done at the age of 26 months on her left ear. During surgery, CSF gusher occurred. After waiting for its flow to decrease, full length multi-channel Clarion implant was inserted in to the cochleostomy and then packed with a piece of muscle to control the gusher. The course of the facial nerve seemed to be normal.

**Table 2.** Post operative audiologic performance of the patients

| No | Anomaly | Age at Surgery | Follow-up | No. of active electrodes | Average PTA* | SPT† | | |
|---|---|---|---|---|---|---|---|---|
| | | | | | | Closed set | GASP | Open set |
| 1 | LVA | 26 m | 60 m | 22 pairs | 38 dB | 100% | 14 of 24 | 42% |
| 2 | IP | 33 m | 24 m | 22 pairs | 30 dB | 89% | 12 of 24 | 67% |
| 3 | LVA | 12 y | 28 m | 22 pairs | 28 dB | 100% | 24 of 24 | 95% |
| 4 | Narrow IAC | 5.5 y | 27 m | 7 pairs | 42 dB | 46% | 4 of 24 | 0% |
| 5 | LVA | 30 m | 16 m | 22 pairs | 32 dB | 58% | 12 of 24 | 31% |
| 6‡ | CC | 10 m | 6 m | 15 pairs | 53 dB | -- | -- | -- |

Abbreviations: LVA, enlarged vestibular aqueduct; IP, incomplete partition or Mondini's dysplasia; IAC, internal acoustic canal; CC, common cavity; GASP, Glendonald's Auditory Screening Procedure.

* The average hearing threshold in 3 frequencies (500, 1000 & 2000 Hz) in the latest PTA.

†Speech perception tests in Persian language.

‡Because of very young age of the child and short postoperative period the speech perception tests could not be performed.

The postoperative period was uneventful. The electrode was activated at the regular 3 weeks time. Being the first case with a congenital anomaly in our center, she developed auditory abilities with little difference from what we generally expect from those without. After 60 months she now has a pure tone threshold at 38 dB, answers 100% to a closed set and 42% to an open set battery of speech perception test. Taking into account the poor socio-economic background she comes from, she now has an almost close to normal auditory and phonatory abilities. She attends speech therapy at our institute on a regular basis.

**Case 2.** As the second child of a family of which both parents are medical doctors, the complete bilateral deafness of the child was detected early at the age of 6 months. She was born full-term with a normal delivery. Further investigations revealed an incomplete partition or classic Mondini's dysplasia (10-12). Using hearing aid was not beneficial for her and so she was planned for a cochlear implant on her left ear which was done at the age of 28 months. Cochleostomy was performed, but maybe due to the small size of cochlea, the electrode insertion was unsuccessful. Postoperatively she had severe vertigo which subsided after a few days. Five months later she was referred to another center where a cochlear implant was done on the right ear with insertion of full length of the electrode array. After activation of the implant she returned to our institute for auditory rehabilitation. Two years after the implantation, she had an average threshold at 30 dB, responded 89% to a closed set and 67% to open set tests. At this time all the electrodes are active. She has no problem in hearing any sounds except "y" and "h". Her ability to talk has relatively progressed well according to her age and duration from implantation.

**Case 3.** (Fig. 2) Her parents noticed progressive decrease in hearing of their child since the age of 5 years. At the age of 11, severe hearing loss caused difficulty for the child to communicate even with using her hearing aid, thus she was referred to our center. On CT scan, a bilateral enlarged vestibular aqueduct was noticed and audiometric assessment showed bilateral profound hearing loss to total deafness. She underwent cochlear implantation at the age of 12 years. While approaching through the facial recess it was noted that the chorda tympani was placed much more medially than its usual place. CSF gusher occurred at the time of cochleostomy which was controlled with packing the site with muscle after full insertion of the electrode array of a multi-channel Nucleus-22 cochlear implant. Twenty-eight months after the surgery her average threshold is 30dB, answers 100% to a closed set and 95% to an open set speech perception test.

Her younger sister also has a progressive decrease in hearing with occasional attacks of vertigo. She also was diagnosed to have bilateral enlarged vestibular aqueduct on CT. At the age of 6, her pure tone audiometry stands at 30-40dB while it has been slowly decreasing over the last few years.

**Case 4.** (Fig. 3) When he was two years old his parents sought medical advice for delayed language development. Bilateral total deafness was found on audiologic examination and bilateral narrow internal auditory canal was noted on CT scan. He also suffered from repeated pulmonary infections and corneal ulcers which suggested the diagnosis of familial dysautonomia, otherwise known as Riley-Day syndrome.

Because of unfit medical conditions his implant was delayed up to the age of 4, when his vision had also diminished considerably. Therefore cochlear implant was the only possible way of giving the child a means of communication with the exterior. With a positive promontory test while expecting a relative poor audiologic result and explaining it to his parents, he was scheduled for implant. After the insertion of electrodes, anomalous stimulation of facial nerve was noted by reflex checking. During activation and programming, only 7 electrodes gave auditory responses. Although after 27 months he has not yet been able to score open set tests but his average threshold audiometry is 42 dB and answers 42% to closed set speech tests. He benefits from the implant by detecting the sounds of his environment and the procedure has been quite helpful to him.

**Case 5.** As a case of enlarged vestibular aqueduct diagnosed at the age of 2 years with bilateral profound sensorineural hearing loss who did not benefit using hearing aid for 6 months, she was scheduled for cochlear implant. During surgery CSF gusher occurred which was controlled with packing the site. Postoperative period was uneventful. After 16 months she has a threshold hearing of 32 dB, responds 58% to a closed set and 31% to an open set test.

**Case 6** (Fig. 1). Her mother (a nurse) realized her hearing loss relatively early and consulted our institute. Audiologic assessment showed bilateral profound hearing loss. On CT scan, bilateral common cavity inner ear deformity was noted with facial nerve placed anterior to the ossicles.

At the age of 10 months she became a candidate for cochlear implant. Trans-mastoid labyrinthotomy approach as first described by Mc Elveen *et al.* (13) was chosen. The prominence of lateral semicircular canal was absent and instead a single large bony capsule was protruding from the middle ear into the mastoid cavity and the opening into the inner ear was made in this capsule just behind the attic area in the mastoid antrum, posterior to the incus. The facial nerve was not seen in this approach. CSF gusher occurred which was again controlled with packing the opening after full insertion of electrodes. It was also noted that stapes crura were placed vertically to their normal position (up and down) and persistent stapedial artery was also present.

The postoperative period was uneventful. She responded to a few electrodes at the time of activation and programming but gradually the number of active electrodes was increased to 15 pairs at 6 months post-surgery, with an average threshold at 53 dB.

Because of the very young age of the child and short postoperative period, the speech perception tests could not be performed.

# DISCUSSION

While reviewing literature we found considerable variation in the findings and outcome of cochlear implants in patients with inner ear malformations (Tables 3 and 4).

Retrospective literature review has inherent limitations (14-19). The quality of the acquired data must be viewed in the light of extreme variability in diagnostic criteria, surgical technique and reporting methods. Nonetheless, such reports are of value when studying such an unusual problem. CSF gusher has been reported in 50% of patients, the incidence of which varies depending upon the type of malformation. The initial management of a CSF gusher has been to allow the flow of CSF to subside followed by firmly packing the cochleostomy site with soft tissue.

This was effective in 19 of the 20 gushers. Continuous lumbar cerebrospinal fluids drain (CLCFD) was needed in just one of 20 gushers which eventually needed a revision surgery to control the leak. No case of meningitis was reported in association with a CSF gusher.

**Table 3.** Literature review

| Author | No. of patients | Device | Anomaly | CSF leak | Finding and complications |
|---|---|---|---|---|---|
| Mangaberia [10] | 1(adult) | 3M/House | IP | 0 | |
| Miyamoto[11] | 1 | 3M/House | CC | 1 | None |
| Jackler[7] | 3* | 3M/House | IP | 0 | |
| | | | CC | 0 | Packed |
| | | | 2 Hypo | 0 | |
| Silverstien[12] | 1(adult) | Nucleus | Hypo | 0 | None |
| Molter[13] | 1 | Nucleus | CC | 0 | Abnormal VII entered through SCC |
| Tucci[14] | 6(3 adults) | Nucleus | CC | 1 | Packed, CWD |
| | | | 3 IP | 1 | Packed and drained |
| | | | 2Hypo | 1 | Abnormal VII, CWD to visualize electrodes into IAC |
| Slattery[15] | 10(3 adults) | 13M/House | 2 CC | 2 | Packed, 1 abnormal VII, incus removed |
| | | 9 Nucleus | 2VA | 0 | |
| | | | 2VA+ V | 1 | Packed |
| | | | Hypo | 0 | Abnormal VII, CWD, facial paralysis |
| | | | 3 IP | 1 | Packed, 1 abnormal VII, CWD |
| Hoffman[16] | 3 | 3 Nucleus | 2 IP | 2 | 2 Packed, 1 needed CLCFD, 1 device |
| | | | identical twin sisters | | failure, re-implanted. |
| | | | VA | 1 | Packed |
| Woolley[17] | 4 | Nucleus | 2 IP | 1 | Packed, 1 abnormal VII |
| | | | VA | 1 | Packed |
| | | | CC | 1 | Packed |
| Luntz[18] | 10 | Nucleus | 4 IP | 0 | 1 device failure, re-implanted |
| | | | 3 CC | 2 | Packed |
| | | | 3 VA | 3 | Packed, temporary VII paralysis |
| McElveen[9] | 4 | 2 Clarion | 4 CC | 0 | 4 abnormal VII, Transmastoid |
| | | 2 Nucleus | | | Labyrintotomy |
| Juichi[19] | 1 | Nucleus | CC | 1 | Abnormal VII, Transmastoid Labyrintotomy |

Abbreviations: IP, incomplete partition; CC, common cavity; Hypo, cochlear hypoplasia; VA, enlarged vestibular aqueduct; VA+V, enlarged vestibular aqueduct and vestibule; VII, facial nerve; IAC, internal auditory canal; CWD, canal wall down mastoidectomy; SCC, semicircular canal.
* 4 ears implanted in 3 children.

Facial nerve anomalies have been reported in 27% of patients with inner ear malformation undergoing cochlear implant. Facial paralysis as a complication of surgery has been reported in at least 2 cases in the literature that was reviewed, one of which was permanent.

## Surgical Findings

The facial nerve showed anatomic abnormalities in 2 of the patients. In one of them (case 3) chorda tympani nerve was more medial than it is normally placed and in the child with common cavity (case 6) the facial nerve was placed anterior to the ossicles. In the same child, the crura of the stapes were directed vertically and persistent stapedial artery was present. Facial nerve palsy occurred in none of the cases,but there was anomalous activation of the facial nerve and twitching in two children upon activation of the electrodes, which was managed by turning off the offending electrodes during the postoperative programming.

CSF gusher occurred in 4 patients, which was controlled with packing the cochleostomy site. There was no need for middle ear obliteration, continuous lumbar drain or revision surgery to handle the CSF leak in any of the cases.

**Table 4.** Literature review summary (12 reports, 40 patient)

| | |
|---|---|
| **Sex** | |
| Male | 24 |
| Female | 16 |
| **Ear implanted** | |
| Left | 22 |
| Right | 18 |
| **Inner ear malformation** | |
| Common cavity | 15 |
| IP (Mondini) | 15 |
| Hypoplastic | 6 |
| LVA | 10 |
| **Facial nerve anatomy** | |
| Normal | 29 |
| Abnormal | 11 |
| **Facial nerve paralysis** | |
| Permanent | 1 |
| Temporary | 1 |
| **CSF gusher** | 20 |
| Common cavity | 8 |
| IP (Mondini's) | 5 |
| Hypo plastic | 1 |
| LVA | 6 |

Abbreviations: IP, incomplete partition; LVA, enlarged vestibular aqueduct.

With the exception of the child with Mondini's dysplasia, in all the other 5 cases, full length of the electrode array was inserted.

No other surgical complications such as severe bleeding, infection, meningitis or device extrusion were observed.

## Electrode Activation and Programming

Details of post-implant auditory performance are presented in table 2. The number of electrodes in use varies from a minimum of 7 to a maximum of 22. For patient 4, low levels of stimulation of some of the electrodes produced discomfort with facial twitching, thus these electrodes were excluded from the program. For patient 6, stimulation of some of the electrodes produced severe vertigo, nystagmus and a feeling of discomfort, which were again excluded from the program. This result might have occurred due to the presence of vestibular nerve endings in the same common cavity. In the other 4 children, all 22 electrodes could be activated without a problem.

## Post-implant Auditory Performance

Average PTA threshold changed from 28 dB to 53 dB. All patients demonstrated scores above chance in the closed set speech perception test, with two of them reaching 100%. With the exception of patient 4, all the others who could perform the GASP test scored at or above 50%, with one scoring 24 out of 24. Four out of six children could develop open set speech recognition. Patient 6, being the youngest of the case series could not respond to speech perception tests.

All six patients described reportedly use their device a minimum of 12 hours each day. In conclusion cochlear implantation is feasible and relatively safe in the child with inner ear dysplasia. However, the presence of a malformation presents a challenge to the implant surgeon, most notably because of anomalous facial nerves and/or CSF gusher (16).

As described in literature, cochlear implantation is contraindicated in a narrow IAC which suggests an eighth nerve with severely decreased nerve fibers. However this is relative and in presence of positive promontory test, implantation can be performed knowing the limitations of performance. A cochlear aplasia or a narrow IAC with negative promontory test showing absent eighth nerve is an absolute contraindication to cochlear implantation. Such patients are candidates for brain stem auditory implantation. All other degrees of inner ear malformations ranging from common cavity to incomplete partition and enlarged vestibular aqueducts can be safely implanted, taking the necessary precautions with fairly acceptable hearing results expected. Further larger scale standardized studies are required to conclude about the results of cochlear implant in patients with inner ear malformations.

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