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Outcome of cochlear implantation in children with enlarged vestibular aqueduct (EVA) and mondini dysplasia (incomplete partition type II)

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Outcome of Cochlear Implantation in Children with Enlarged Vestibular Aqueduct (EVA) and Mondini Dysplasia (incomplete partition type II)

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Abstract

Background

Congenital inner ear abnormality is a major cause of sensorineural hearing loss in children, about 20% of children with congenital sensorineural hearing loss (SNHL) have associated malformations of the temporal bone, and increased experience in cochlear implantation has led to more children with abnormal cochleo-vestibular anatomy submitted to this procedure.

Aim

To evaluate the outcome of cochlear implantation in cases with enlarged vestibular aqueduct (EVA) and Mondini deformity (incomplete partition type II) following cochlear implantation and comparing the results with cochlear implant cases without inner ear anomalies.

Patients and methods

69 children all less than 6 years of age, CI surgeries underwent from April 2014 to December 2015 at the National Hearing & Speech institute (HSI) Giza-Egypt. Eight patients were congenital prelingual severe to profound hearing loss with enlarged vestibular aqueduct (EVA) and Mondini deformity (incomplete partition type II). Isolated EVA was detected in four cases, two cases with bilateral incomplete partition type II (Mondini deformity) and two cases with bilateral incomplete partition II (Mondini deformity) associated with dilated vestibule & vestibular aqueducts bilaterally with short lateral, SCC (semicircular canal). Beside those 8 cases, a random sample (16 control cases). The ECAP thresholds determined with neural response telemetry (NRT) or Auditory response threshold (ART) software provide a good starting point for locating the behavioral T and C levels. Auditory skills were evaluated before start of rehabilitation and after 6 months of rehabilitation using LittleEARS Questionnaire and Auditory Checklists and compared to the auditory skills of other cochlear implantees with normal inner ear. Results: This study showed that the auditory skills and audiological performance of children with congenital ear anomalies developed over a period of 6 months after cochlear implantation, in a similar manner to those of young children with radiologically normal inner ears.

Conclusion

Cochlear implantation can be successfully performed in children with enlarged vestibular aqueduct (EVA) and Mondini dysplasia (incomplete partition type II). These children and their parents can expect significant auditory benefits from this intervention.

Keywords: Cochlear implant, Enlarged vestibular aqueduct, Mondini Dysplasia (Incomplete partition type II)

INTRODUCTION

Congenital inner ear abnormality is a significant cause of sensorineural hearing loss in children [1]. About 20% of children with congenital sensorineural hearing loss have associated malformations of the temporal bone [2]. Increased experience in cochlear implantation has led to more children with abnormal cochleovestibular anatomy submitted to this procedure.

Jakler *et al.* [3] proposed a classification of cochleovestibular malformations based on polytomography and related to

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embryological genesis. Sennaroglu and Saatci [4] suggested a rating based on computed tomography (CT) findings, which is an essential tool in the field of cochlear implantation. The authors described two types of incomplete partition (IP): IP type I and type II.

Recently, IP was recognized as type III [5]. It may occur in one ear (unilateral) or both ears (bilateral) and can cause varying degrees of sensorineural hearing loss [6]; the condition can also predispose a patient to recurrent meningitis [7].

IP type I is also known as cystic cochleovestibular malformation, where the cochlea has no interstellar septum and modiolus resulting in an empty cystic cochlea. A dilated cystic vestibule accompanies this with developmental arrest at the fifth week of gestation [8]. As regards IP II, the apical and middle cochlea turns are undifferentiated and form a cystic apex. The vestibule is mildly dilated, and the vestibular aqueduct is always enlarged, and developmental arrest occurs at the seventh week of gestation.

Enlarged vestibular aqueduct (EVA) is commonly defined as having a width larger than 1.5 mm, measured at the midpoint between the common crus. Valvassori and Clemis [9] first described the external aperture into the posterior fossa radiologically. Regarding the inner ear abnormality associated with sensorineural hearing loss, EVA has been reported to be the most common [10]. EVA accompanies malformation of the cochlea and semicircular canals [11].

Cochlear implantation is an essential method of auditory rehabilitation for children with profound sensorineural hearing impairment, who do not derive benefit from amplification [3]; however, children with congenital inner ear malformations show a critical test to even the most experienced clinicians. In fact, many cochlear implant (CI) centers deferred implantation in these children due to the uncertainty regarding surgical feasibility and performance expectations [12].

Early application of CI shows that it is of great importance for the development of adequate auditory performance and language skills [13]. Cochlear implantation efficiently increases both auditory perception and language development in children with congenital inner ear anomalies.

Tracking the auditory skill development of young children is quite essential. Some studies have elected to use auditory skills questionnaires completed by a therapist or a parent. Alternatively, rating scales of auditory development, such as the Category of Auditory Performance [14] or the Infant-Toddler Meaningful Auditory Integration Scale [15], the Auditory Skills Checklist (ASC) [16], and the LittleEARS Auditory Questionnaire (LEAQ) [17] [Appendix 1] have been used.

The ASC is an assessment test that uses a structured parent interview and a clinician observation to obtain information about auditory skills development. The ASC is administered to a child regularly to clinically observe the child's progress, monitor effectiveness of amplification, and to identify the

absence of auditory skills due to the hearing impairment. By combined information from the parent and the examiner, the examiner assigns a rating regarding the child's skill: 0, absence of skill; 1, inconsistent or emerging skill development; 2, consistently demonstrates the skill [16].

The LEAQ is a quick and useful tool for the assessment of auditory skills in a growing population of infants and toddlers. The LEAQ was created to assess the auditory skills of children who receive hearing devices either hearing aids or CIs. The LEAQ relies on a parent questionnaire, which has been shown to be a reliable way of assessing auditory skills development in children [17].

The electrode impedance is a method of measuring resistance encountered by electricity passing through wires, electrodes, and biological tissues [18] and the unit of impedance is $K\Omega$. While the neural response telemetry system renders possible the measurement of the compound action potential threshold [19], both are applied as intraoperative and postoperative verification methods for patients who undergo cochlear implantation.

Aim of the study

The aim of this study was to evaluate cases with EVA and Mondini deformity (IP type II), following cochlear implantation and comparing the results with CI cases without inner ear anomalies.

PARTICIPANTS AND METHODS

Sixty-nine children all less than 6 years of age who underwent CI surgeries from April 2014 to December 2015 at the National Hearing and Speech Institute (HSI) Giza, Egypt. Eight patients had congenital, prelingual severe to profound hearing loss with EVA and Mondini deformity (IP type II).

Study design

All young children underwent a thorough otorhinolaryngological examination and audiometric tests using behavioral audiometry, aided free-field audiometry, tympanometry, and electrophysiological tests including auditory brainstem response (ABR), and otoacoustic emission (OAE).

Preoperative evaluation

Each child with a diagnosis of prelingual severe to profound hearing loss received high-resolution computed tomography (HRCT) examination and inner ear and brain magnetic resonance imaging (MRI). All studies were performed using a standard temporal bone protocol with contiguous 0.5 mm scans of the temporal bone acquired in the axial and coronal planes.

Radiologists reviewed the CT images on a clinical picture archiving and communicating system and examined the temporal bone for the presence of cochlear, vestibular, or semicircular canal abnormalities. The diagnosis of Mondini dysplasia type II was confirmed by the presence of a shortened cochlea with only $1\frac{1}{2}$ turns, incomplete interscalar septum or osseous spiral lamina between the middle and apical turns, and a fully developed basal turn.

The width of the vestibular aqueduct (VA) was measured at the operculum and at the midpoint. EVA is diagnosed radiographically when its anteroposterior diameter exceeds 1.5 mm on computed tomography (CT) scan of the temporal bone, measured midway between its aperture and crus communes [9,20–22].

There was a negative history for maternal illnesses including TORCH [toxoplasmosis, other (syphilis, varicella-zoster, parvovirus B19), rubella, cytomegalovirus, and herpes infection], Rh-incompatibility, neonatal jaundice, or meningitis.

Their other developmental milestones were average. There was a negative history of visual disturbances, hypothyroidism, syncopal spells, or renal problems. General examination showed average intelligence quotient (IQ) with no neurological dysfunction.

Inner ear anomalies included isolated EVA detected in four cases, two cases with bilateral IP type II (Mondini deformity), and two cases with bilateral IP II (Mondini deformity) bilaterally associated with the short, lateral semicircular canal.

The data obtained from 24 patients in this study were divided into the following two groups according to the presence or absence of congenital anomalies:

Group A (8 cases)

Eight patients with congenital anomalies were implanted. One case was implanted with HiFocus 1 J electrode (12.5%) and three cases were implanted with HiFocus Mid-Scala Electrode (37.5%) (AB, Sylmar, CA, USA). Four cases were implanted with Sonata TII100+ titanium implant footprint with Standard Electrode (50%) (SA; Med-El, Innsbruck, Austria).

The mean age of patients was 4 years, 8 months (range: 3 years, 4 months to 5 years and 9 months). Of the eight cases, four were men (50%), and four were women (50%).

Approaches included a cochleostomy in five cases (62.5%) and round window approach in three cases (37.5%).

Group B (16 control)

Sixteen patients with normal inner ear anatomy were implanted. Two of them received HiFocus 1 J Electrode (12.5%), eight cases with HiFocus Mid-Scala Electrode (50%) (AB, Sylmar, CA, USA). Sonata TII100+ titanium implant footprint with Standard Electrode was used in five cases (31.25%) and Sonata TII100+ titanium implant footprint with Flex 28 electrode was used in one case (6.25%) (SA; Med-El, Innsbruck, Austria) (SA; Med-El, Innsbruck, Austria).

The mean age of patients was 4 years (range: 2 years, 1 month to 5 years and 9 months). Of the 16 cases, four were men (25%) and 12 were women (75%).

Approaches included a cochleostomy in seven cases (43.75%) and round window approach in nine cases (56.25%).

Overall, patient's right ear was operated in group B because they were right-handed and had similar inner ear anatomy and hearing level on both sides.

The Ethics Review Committee of the National Hearing and Speech Institute (HSI) and the parents approved the study, or legal guardians of each child signed a written informed consent before entry into the study.

All children received vaccinations for Pneumococcus and Haemophilus influenza (**PREVNAR 13**®vaccines)

Intraoperative protocol

Two senior surgeons performed CI surgeries. Cochleostomy was positioned inferior and anterior to the round window was done in 12 patients (5 in-group A and 7 in-group B) (total 50%) and round window approach in 12 patients (3 in-group A and 9 in-group B) (50%). The round window (RW) membrane was exposed via transmastoid facial recess approach. We accessed the scala tympani directly through the round window. To completely visualize the circumference of the round window membrane, we used a 1 mm micro drill to remove the superior lip of the round window niche.

CSF pulsatile ooze or leak started in five cases of group A. Then the head end of the table was raised and intravenous 20% mannitol drip (1.5 g/kg body weight for more than 20 min) was started. The leak was significantly reduced within 10 min and the electrode array of the implant inserted via the RW or cochleostomy. None of the cases in group A required lumbar drainage.

IO parenteral antibiotics were given. Complete insertion of the electrode array up to the mark was achieved. Temporalis muscle and fascia were used to seal tightly around the electrode array.

The IO telemetry showed satisfactory impedance and neural response in all the selected electrodes.

Postoperatively

Oral acetazolamide, twice daily, was used for 3 days. All patients were kept with the prophylactic intravenous antibiotics and analgesic for 5 days.

During follow-up, they did not have any vestibular symptoms or signs of CSF otorrhorrhea. They had no further problem before or after the implant was turned on and during 3 months of auditory habilitation.

A petrous bone CT, carried out 2 days after the surgery, showed a correct position and no kinking of the array electrode.

At 4 weeks after surgery the implant switch-on programming session was performed by an audiology physician. Regular programming was done to the cases, and all patients were referred postoperatively for auditory and language rehabilitation.

The children get used to their new hearing capacity by gradually increasing the stimulation levels over some sessions. The levels vary from soft to rather loud. If the level is too high, the stimulation could cause discomfort and thus induce rejection of the CI. The ECAP thresholds determined by neural response telemetry or the auditory response threshold (ART) software provide a good starting point for locating the behavioral T and C levels. This allows for a faster fitting procedure using higher stimulation levels than previously.

Auditory skills were evaluated before the start of rehabilitation and after 6 months of rehabilitation using LEAQ and ASC and compared with the auditory skills of other cochlear implanters with the normal inner ear.

The full administration of the ASC takes ~10 min to complete, with less time required for very young children. Reassessment at 6-month intervals was conducted to follow the progression of auditory skill development. At the time of administration, each item of the scale was rated as one of the following: 0 = does not have skill, 1 = inconsistent or emerging skill, and 2 = developed skill.

The 35 questions on the ASC comprise a total raw score of 70, with a range of the total score between 0 and 70 (detection = 18 points, discrimination = 14 points, identification = 14 points, comprehension = 24 points). Higher scores reflect higher or more-developed functional auditory skills.

The LEAQ is a parent questionnaire consisting of 35 ‘yes/no’ questions which can be evaluated by parents in less than 10 min. The LEAQ tests the receptive, semantic, and early expressive language skills of young children in response to the auditory input. The total score of ‘yes’ answers is compared with the normal curve of normal hearing children established by Coninx *et al.* [23]. Both tests were performed at 1 month and repeated at 6 months after starting rehabilitation.

RESULTS

In this study, IO impedance ART or NRI were measured for both groups, 1 month and 3 months follow-up for impedance measurements was done. Regular programming, assessment, and intensive habilitation were followed up for each child at specified times using the ASC and the LEAQ.

Data were described statistically regarding mean ± SD and by percentages when available. Examination of numerical factors between the study groups was finished utilizing the Mann–Whitney *U*-test for independent samples. Within each group, comparison of numerical variables was made using Wilcoxon signed-rank test for paired (matched) samples.

P values under 0.05 were considered statistically significant.

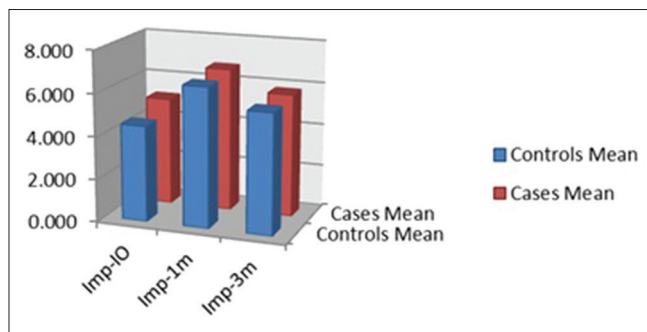


Figure 1: Comparison of mean between control and cases for measured intraoperative impedance (Imp-IO), 1 and 3 months postoperative impedances (Imp-1m and Imp-3m, respectively).

All statistical calculations were done using computer program SPSS (SPSS Inc., Chicago, Illinois, USA) release 15 for Microsoft Windows (2006). Table 1 and Fig. 1 show the mean and SD of ART and impedance (1 and 3 months, postoperatively) in both groups, there is no statistically significant difference between both groups at all measured parameters. Table 2 and Fig. 2 show mean and SD of IO impedance, LEAQ, and ASC (1 and 6 months, postoperatively). Moreover, there is no statistically significant difference between both groups at all measured parameters. Table 3 shows comparisons between ASC and LEAQ at 1 and 6 months, postoperatively, in the control and

Table 1: ART and intraoperative, 1, and 3 months postoperative impedances in both groups

Groups	ART	Imp-IO	Imp-1m	Imp-3m
Controls				
Mean	429.714	4.512	6.554	5.646
SD	347.1356	3.0387	0.8175	1.3072
Cases				
Mean	579.281	5.098	6.746	5.783
SD	405.0142	3.4383	2.8051	2.0775
<i>P</i>	0.374	0.646	0.582	0.624

ART, auditory response threshold; Imp, impedance; IO, intraoperative; 1m, 1 month; 6m, 6 months.

Table 2: IQ, LittleEARS Auditory Questionnaire and Auditory Skills Checklist (1 and 6 months postoperatively) in both groups

Groups	IQ	LEAQ-1m	LEAQ-6m	ASC-1m	ASC-6m
Controls					
Mean	90.94	10.5	19.38	12.31	22.38
SD	4.781	2.160	4.349	3.535	6.761
Cases					
Mean	94.38	12.25	22.38	12.00	21.63
SD	10.336	5.97	5.655	5.043	7.800
<i>P</i>	0.595	0.925	0.460	0.493	0.759

No statistically significant difference between both groups at all measured parameters. ASC, Auditory Skills Checklist; Imp, impedance; IQ, intelligence quotient; LEAQ, LittleEARS Auditory Questionnaire; 1m, 1 month; 6m, 6 months.

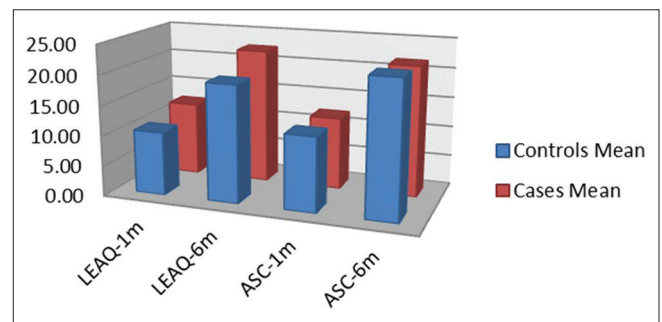


Figure 2: Comparison of mean between control and cases for measured LittleEARS Auditory Questionnaire (LEAQ) and Auditory Skills Checklist (ASC) (1 and 6 months postoperatively).

Table 3: Comparison between LittleEARS Auditory Questionnaire and Auditory Skills Checklist at 1 and 6 months, postoperatively, in control and study groups using Wilcoxon signed-rank test

	LEAQ-1m	LEAQ-6m	P	ASC-1m	ASC-6m	P
Control						
Mean	10.50	19.38	0.012*	12.31	22.38	0.011*
SD	2.160	4.349		3.535	6.761	
Cases						
Mean	12.25	22.38	0.011*	12	21.63	0.012*
SD	5.970	5.655		5.043	7.80	

Statistically significant difference in both groups at measured parameters between 1 and 6 months. ASC, Auditory Skills Checklist; Imp, impedance; LEAQ, LittleEARS Auditory Questionnaire; 1m, 1 month; 6m, 6 months. *P<0.05 was considered statistically significant.

Table 4: Comparison between EVAS and Mondini type II using Mann-Whitney U-test for ART, IO impedance, 1 and 3 months postoperative impedances

Anomaly	ART	Imp-IO	Imp-1m	Imp-3m
EVAS				
Mean	643.25	6.851	6.819	6.616
SD	388.349	4.233	0.392	1.608
Mondini				
Mean	515.313	3.344	6.672	4.951
SD	470.132	1.209	4.265	2.374
P	0.773	0.248	1.000	0.248

No statistically significant difference between EVAS and Mondini type II at all measured parameters. ART, auditory response threshold; EVA, enlarged vestibular aqueduct; Imp, impedance; IO, intraoperative; 1m, 1 month; 6m, 6 months.

Table 5: Comparison between EVAS and Mondini type II using Mann-Whitney U test for IQ, LittleEARS auditory questionnaire and auditory skills checklist (1 and 6 months) postoperatively

Anomaly	IQ	LEAQ-1m	LEAQ-6m	ASC-1m	ASC-6m
EVAS					
Mean	99.00	11.00	22.50	13.00	24.50
SD	12.193	2.944	4.796	5.292	6.403
Mondini					
Mean	89.75	13.5	22.25	11.00	18.75
SD	6.602	8.386	7.182	5.354	8.884
P	0.189	0.882	0.661	0.375	0.243

No statistically significant difference between EVAS and Mondini type II at all measured parameters. ASC, Auditory Skills Checklist; EVA, enlarged vestibular aqueduct; Imp, impedance; IQ, intelligence quotient; LEAQ, LittleEARS Auditory Questionnaire; 1m, 1 month; 6m, 6 months.

study groups. There is a statistically significant difference in both groups at measured parameters between 1 and 6 months with improvement at 6 months. Tables 4 and 5 and Figs. 3 and 4 show a comparison between EVA and Mondini type II; there is no statistically significant difference between EVA and Mondini at all measured parameters. Tables 6 and 7

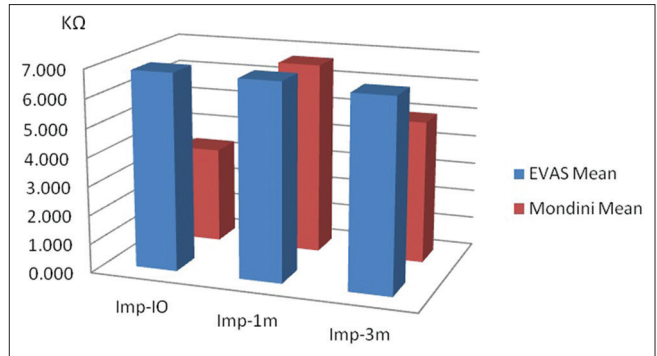


Figure 3: Comparison of mean between enlarged vestibular aqueduct (EVA) and Mondini types II measured in this study for intraoperative impedance (Imp-IO), 1 and 3 months postoperative impedances (Imp-1m and Imp-3m, respectively).

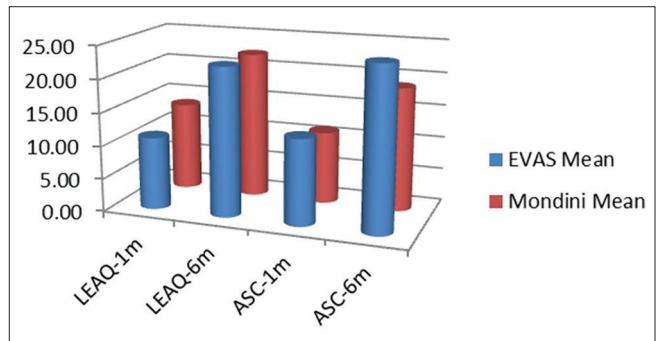


Figure 4: Comparison of mean between EVA and Mondini types II measured in this study for LittleEARS Auditory Questionnaire (LEAQ) and ASC (1 and 6 months postoperatively).

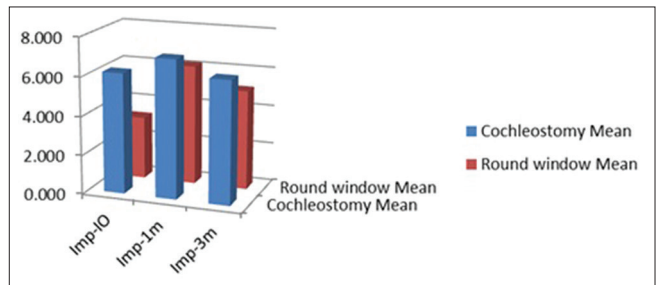


Figure 5: Comparison of mean between cochleostomy approach and round window approach for intraoperative impedance (Imp-IO), 1 and 3 months postoperative impedances (Imp-1m and Imp-3m, respectively).

and Figs. 5 and 6 show a comparison between cochleostomy and round window approach. There is the statistically significant difference between cochleostomy and round window approach at ART and IO impedance. The ART and IO impedance were at a lower level with round window than the cochleostomy approach.

DISCUSSION

Cochlear implantation is now an established means of rehabilitating severe-to-profound sensorineural hearing loss

Table 6: Comparison between cochleostomy and round window approach using Mann-Whitney U-test for ART, IO impedance, 1 and 3 months postoperative impedances

Approaches	ART	Imp-IO	Imp-1m	Imp-3m
Cochleostomy				
Mean	718.938	6.180	7.085	6.286
SD	358.8388	3.4933	1.8589	1.7653
Round window				
Mean	240.201	3.234	6.150	5.098
SD	160.557	1.828	1.416	1.095
P	0.004*	0.011*	0.166	0.05

*P 0.004 statistically significant. There is statistically significant difference between cochleostomy and round window approach at ART and intraoperative impedance. ART, auditory response threshold; Imp, impedance; IO, intraoperative; 1m, 1 month; 6m, 6 months.

Table 7: Comparison between cochleostomy and round window approach using Mann-Whitney U-test for IQ, LittleEARS Auditory Questionnaire and Auditory Skills Checklist (1 and 6 months, postoperatively)

Approaches	IQ	LEAQ-1m	LEAQ-6m	ASC-1m	ASC-6m
Cochleostomy					
Mean	94.58	12.50	22.08	13.33	24.33
SD	8.639	4.719	5.992	4.075	6.213
Round window					
Mean	89.58	9.67	18.67	11.08	19.92
SD	4.078	1.969	2.871	3.728	7.217
P	0.099	0.051	0.072	0.126	0.064

No statistically significant difference between cochleostomy and round window approach at all measured parameters. ART, auditory response threshold; ASC, Auditory Skills Checklist; Imp, impedance; IQ, intelligence quotient; LEAQ, LittleEARS Auditory Questionnaire; 1m, 1 month; 6m, 6 months.

in patients for whom traditional amplification provides limited benefit. Outcomes are overall highly successful with low complication rates [24].

This study demonstrated that the auditory skills of children with congenital ear anomalies developed over a period of 6 months after cochlear implantation, similar to those of young children with radiologically normal inner ears.

The result of this study is similar to the study done by Miyamoto *et al.* [25]. They published results of a retrospective case-control study on the outcome of cochlear implantation in 23 patients with EVA and 46 control patients and concluded that cochlear implantation was beneficial for the treatment of hearing loss in EVA as well as in control patients [25]. The study done by Chen *et al.* [26] compared 62 infants with LVAS for the development of auditory skills after CI and found results similar to those of infants with a normal inner ear.

In a research conducted by other authors, children with more severe malformations (common cavity, cochlear dysplasia, and hypoplastic cochlea) performed worse than those with more minor cochlear anomalies [27,28].

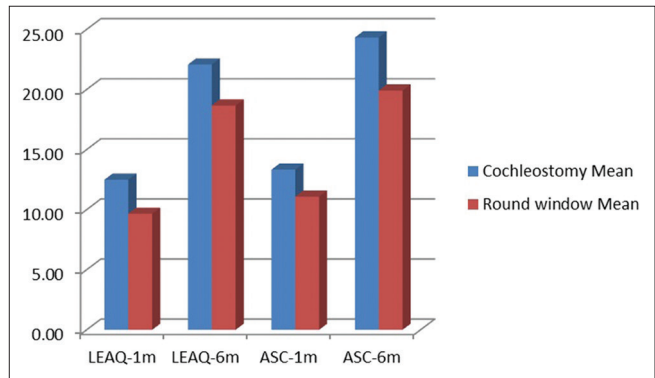


Figure 6: Comparison of mean between cochleostomy approach and round window approach for LittleEARS Auditory Questionnaire (LEAQ) and ASC (1 and 6 months, postoperatively).

In our study, five (62.5%) out of eight cases had IO cerebrospinal fluid (CSF) pulsatile ooze or leak that was controlled by the protocol mentioned before with no effect on the outcome of surgery and without the need to do lumbar puncture which matches the results of Aschendorff *et al.* [29]. Most of the literature reported CSF gusher or leak as a common problem encountered during and after cochlear implantation [30]. Although CSF leak management in the temporal bone is well described [31,32], the literature is less clear on the incidence and management of CSF leaks uncovered during cochleostomy. In his review of 298 children implanted at the Hospital for Sick Children in Toronto, Papsin [8] described 103 with anomalous bony labyrinthine anatomy. Cochleostomy gusher was noted in 6.7%, and a common cavity deformity was most commonly associated with a gusher.

IO management included reverse Trendelenburg positioning to allow the flow of CSF to subside and firmly packing with free temporalis muscle and fascia around the inserted electrode.

Hoffman *et al.* [33] identified 21 CSF leaks in 50 surgeries performed on children with anomalous cochleovestibular anatomy. Sixteen of these subsided with packing the cochleostomy alone, four required lumbar drainage, and one required revision surgery.

Luntz *et al.* [34] reviewed 10 CIs in which five perilymph/CSF leaks occurred. Four of these leaks stopped with packing the cochleostomy, whereas one required plugging of the middle ear and the eustachian tube with muscle and fascia.

In a meta-analysis, Fahy *et al.* [35] found that half of the patients with EVA [17 of 34 (50%)] experienced CSF gushers at cochleostomy, all controlled by muscle packing around the electrode and without lumbar drains.

We determined the results of audiological performance after cochlear implantation in the group of children with inner ear abnormalities compared with the group of congenitally deaf children without radiologically detectable malformations. The audiological performance in both groups was similar. Although the criteria for cochlear implantation had excluded children

with inner ear malformations, presuming malperformance after implantation, our results agreed with the recently published studies reporting satisfactory hearing results in these patients except those with a common cavity malformation [34,36,37].

A retrospective case–control study on the outcomes of cochlear implantation in 23 patients with EVA and 46 control patients concluded that cochlear implantation was beneficial for the treatment of hearing loss in EVA as well as in the control patients [25]. Chen and colleagues compared 62 infants with LVAS for the development of auditory skills after a CI. Moreover, he found results similar to those of infants with a normal inner ear [26].

The aftereffects of the present cases and the review of the literature agree that implantation in EVA and Mondini dysplasia (IP type II) can be performed without IO and postoperative complications. The outcome of hearing and speech development is being followed, but preliminary assessment showed no difference compared with implantees with normal cochlea at 6 months' follow-up. Therefore, cochlear implantation in deaf children with EVA and Mondini dysplasia (IP type II) is feasible and practical.

Overall results are encouraging. Several factors are crucial to the success of cochlear implantation. These factors consist of the age at implantation, the duration of deafness, the mode of communication, and the participation and support of the child's family during rehabilitation.

CONCLUSION

- (1) Cochlear implantation can be successfully performed in children with EVA and Mondini dysplasia (IP type II). These children and their parents can count on massive auditory improvement from this intervention
- (2) IP and enlarged vestibular aqueducts can be safely implanted; acceptable hearing results are expected.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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27. Have an auditory memory for number of items (e.g., being able to remember boat, apple, cup, and shoe would be for items)?
28. Have an auditory memory for phrases/sentences (e.g., 'The girl jumped over the fence to get the ball.')?
29. Auditory sequence a story with three events, 4 events, 4+ events (e.g., 1st event=Steve went to the store; 2nd event=He bought dog bones; 3rd event=Steve took the bones home to the dog)?
30. Understand the question forms What, Where, Who, Why, When (e.g., 'Where is the dog?'; 'Who broke the cup?')?
31. Understand concepts in phrases and sentences (e.g., in, under, between, in front)?
32. Understand the use of negatives in phrases and sentences (e.g., no, not, no more)?
33. Understand frequently heard phrases/sentences (e.g., 'Brush your teeth and get ready for bed.')?
34. Acquire information incidentally through audition alone?
35. Understand most of what is said through audition alone?