

A stepwise algorithm for the management of cerebrospinal fluid gusher during cochlear implantation

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Objectives

The aim of this study was to assess the incidence of CSF gusher during cochlear implantation in children with and without congenital inner-ear malformation and to establish a simple stepwise algorithm for managing CSF gusher at the time of cochleostomy.

Materials and methods

A total of 54 congenitally deaf children were included in a retrospective study between January 2011 and December 2012. All cases underwent classical cochlear implantation surgeries via mastoidectomy and posterior tympanostomy approach.

Results

Nine patients developed gusher at the time of the cochleostomy. Among the nine cases, only one child did not show any preoperative radiologic evidence of any bony cochleovestibular malformation, whereas the remaining eight cases had different congenital inner-ear malformations with known risk for intraoperative gusher during surgery.

Conclusion

We concluded that the CSF gusher is a surgical difficulty or an intraoperative challenge rather than a bad prognostic determinate for the postoperative audiologic performance, and in cases of congenital cochleovestibular malformation that develop gusher, a high degree of congenital anomaly of the cochlea, and not the degree or the amount of gusher, is correlated to the poor patient performance. Finally, we were able to achieve a simple stepwise algorithm for the management of gusher during cochlear implantation.

Keywords:

cerebrospinal fluid gusher, cochlear implantation, common cavity malformation, large vestibular aqueduct syndrome, wide internal auditory meatus

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Introduction

Cerebrospinal fluid (CSF) gusher is generally used to describe the profuse flow of CSF during accessing the basal turn of the cochlea during cochlear implantation whether using ordinary cochleostomy or round window insertion. Before the cochlear implant era, this condition was termed perilymphatic gusher and was expected to be encountered during stapedectomy in patients with an abnormally wide internal auditory meatus (IAM) or the large vestibular aqueduct syndrome (LVAS). In cases of gusher, the fluid coming from the inner ear is the CSF and not the perilymph owing to it being a few microliters [1].

The first report of a gusher during cochlear implant surgery in an inner-ear malformation was by Miyamoto *et al.* [2]. They experienced a gusher during cochleostomy in an ear with Mondini deformity.

Normally, CSF is not present in the cochlea; the CSF in the subarachnoid space extends laterally into the internal auditory canal (IAC) as far as the fundus, where it is separated from the perilymph by a bony plate termed the cribriform plate; however, it can access the cochlea

and may be encountered during cochlear implantation either through an abnormal developmental pathway of the otic capsule, usually a deficient cribriform plate, or after traumatic disruption of the temporal bone.

Gusher-prone developmental abnormalities of the cochlea-vestibular apparatus include an abnormally wide IAC (IAM), LVAS, incomplete partition type II (Mondini) malformation, and common cavity malformation [3]. The majority of these anomalies usually become uncovered with preoperative imaging; however, unexpected CSF leaks may occur at the time of accessing the basal turn of the cochlea for inserting the electrode array, whether through a round window or through the ordinary cochleostomy anteroinferior to the round window, without any preoperative radiologic evidence of any congenital inner-ear malformation.

The wide variability of the incidence of CSF gusher during surgery in different reports by different authors may be attributed to the inclusion of minor leaks, leading to an overestimation of the number of CSF leaks among different centers, which explains the huge range in the incidence of

leaks. Papsin [4] reported seven cases in a series of 103 patients with malformations; they also stated that the term gusher should be used only to describe the pulsatile egress of fluid that lasts for up to a minute and then subsides.

The known risk of a CSF gusher was previously considered as a relative contradiction to cochlear-implant surgery. The first case with LVAS was first implanted in 1995, and since then, patients with a variety of preoperatively identified cochlear and labyrinthine abnormalities have been implanted with minimal complications and a reasonable postimplantation audiologic profile gain [5].

Materials and methods

A retrospective analysis of 84 deaf children: 18 of them showed inner-ear malformation on preoperative radiologic assessment and no bony cochleovestibular malformation was found in the other 66 cases. All cases underwent cochlear implantation between January 2011 and December 2012 after taking the appropriate informed consents at the Alexandria University Hospital and the Alexandria Ear Hospital to assess the incidence of CSF gusher, different management strategies for CSF gusher at the time of surgery, and also the postoperative performance among these cases.

Preoperative evaluation included otologic examination, audiologic evaluation, and psychological assessment.

A high-resolution computed tomography (CT) scan of the temporal bone and MRI of the IAM was performed for all cases before surgery to detect the different types of congenital anomalies, which were classified according to Jackler *et al.* [6].

Surgeries were performed using a standard postauricular incision, mastoidectomy with posterior tympanotomy; cochleostomy was performed in all cases anteroinferior to the round window, to be followed by the insertion of the cochlear implant array and fixation of the receiver stimulator in the predrilled well to house the stimulator receiver of the implant electrode; meticulous wound closure in three layers was accompanied by a pressure dressing for 5 days. All cases with CSF gusher were detected at the time of surgery.

Results

Nine cochleostomy gushers were identified among the 84 studied patients, at an incidence of 10.7%. The mean age of these nine patients was 31 months (ranging from 12 to 46 months).

Table 1 shows that there was a significant difference regarding the occurrence of CSF gusher during cochleostomy among ears with congenital inner-ear malformation when compared with others with no anomalies (the *P* value using Fisher's exact test was 0.000005).

Among the nine cases with CSF gusher, we neither needed lumbar drain nor experienced postoperative meningitis; the gusher was totally controlled at the time of surgery, and no postoperative leak was encountered in any of them.

In our studied group, only one patient among the 66 cases did not show any evidence of congenital inner-ear malformation on preoperative radiologic evaluation, but developed CSF gusher at the time of the cochleostomy; her parents noticed a delayed speech development at the age of 24 months. Further investigations revealed bilateral profound sensorineural hearing loss. A CT scan of the temporal bone was normal, but she did not benefit from hearing aids; therefore, cochlear implant was planned and performed at the age of 32 months on her left ear.

During surgery, CSF gusher occurred, and after waiting for its flow to decrease, a regular straight electrode array (Pulsar; Medel, Innsbruck, Austria) was inserted into the cochleostomy. CSF continued to leak from the cochleostomy; however, it stopped by just sealing of the cochleostomy with the periosteum without packing of the whole basal turn. After 12 months, she had an average pure tone threshold at 32 dB, answers 100% to a closed set and 42% to an open set battery of speech perception test. She developed auditory abilities as expected from those without gusher.

Inner-ear anomalies that developed gusher during cochleostomy in this study included the following:

- (a) Three cases with Mondini's cochlear dysplasia: The cochlea appeared radiologically smaller than normal, and were partially lacking the interscalar bony septum, with confluence of the apical turn and the middle turn (Fig. 1).

In this study, we implanted three cases with Mondini malformation: two of them were twins and were implanted at the age of 36 month with the Medel Pulsar straight array, whereas the third was 14 months

Table 1 Distribution of the studied group

	CSF gusher at the time of cochleostomy		Total
	Yes [n (%)]	No	
Congenital inner-ear malformation	8 (44)	10	18
No congenital inner-ear malformation	1 (1.51)	65	66
Total	9 (10.7)	45	84

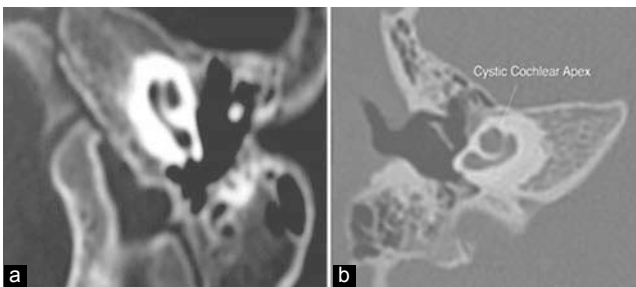
CSF, cerebrospinal fluid.

old at the time of implantation and was implanted with Medel Sonata.

In one of the brothers, the CSF gusher persisted despite immediate placement in the reverse Trendelenburg positioning and tight packing of the cochleostomy with periosteum after implant electrode insertion; further packing of the basal turn of the cochlea with small pieces of muscles also failed to stop the gusher. Packing of the whole middle ear and closure of the Eustachian tube with fascia and muscles harvested from the wound was needed after removal of the incus buttress and the incus for better visualization and better access to the Eustachian tube; no residual leak was noted after these measures. The CSF gusher that occurred in the other brother was controlled by sealing of the cochleostomy around the electrode array with the periosteum.

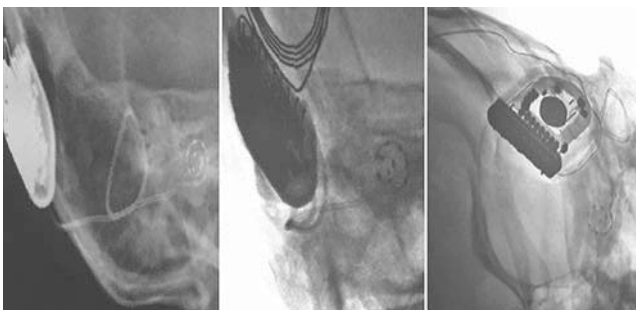
The control of the CSF gusher in the third case with Mondini syndrome was very simple when compared with the other two cases. CSF started to regress spontaneously by continuing suction in the anti-Trendelenburg position, and the insertion of the electrode array, which was easy, ensured complete arrest of the CSF gusher.

Figures 1



Axial computed tomography scan showing (a) right and (b) left temporal bones on which there is confluence of the apical and the middle cochlear turn (incomplete partition type II – Mondini malformation).

Figures 3

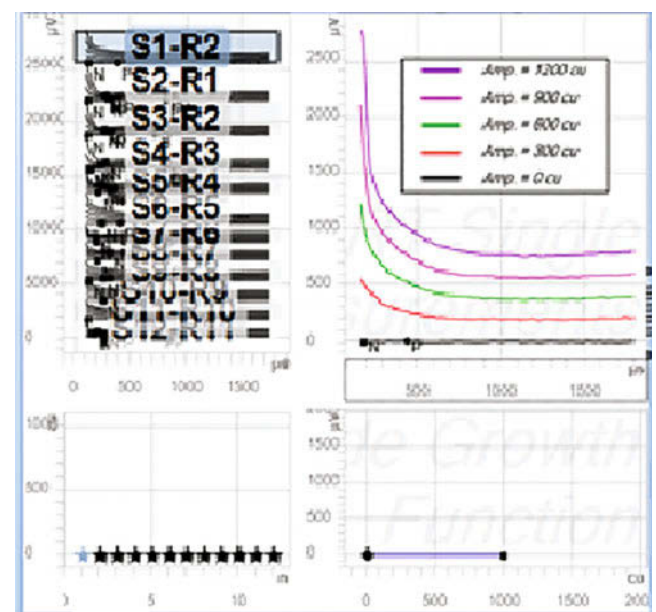


Mondini malformation: postoperative Stenver's view radiograph showing coiling of the electrode array in the confluent apical and middle cochlear turn.

None of the three cases showed evoked response telemetry (ERT) on intraoperative testing in the 12 channels of the implant array; however, on the initial stimulation of the implant 1 month after the surgery, ERT wave forms were present in the basal five to six channels of the implant, whereas the apical and the mid channels still did not show any wave form even with a high current level, which caused facial nerve stimulation in one case (Figs. 2–5).

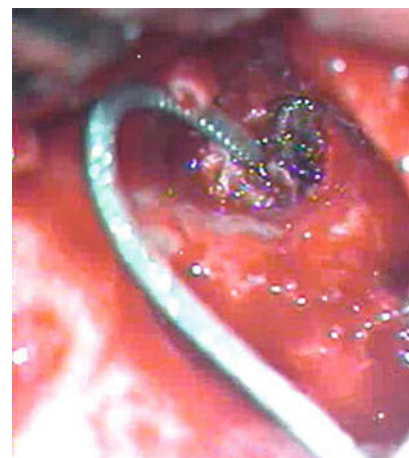
One year after the surgery, the average pure tone threshold in the first case was 38 dB, answered 80% to a closed set and 45% to an open set speech perception

Figures 2



Mondini malformation: no evoked response telemetry (ERT) wave forms could be detected in any channels on intraoperative testing.

Figures 4



Packing of the whole middle ear and the Eustachian tube with muscles after removal of the incus and its buttress with muscle.

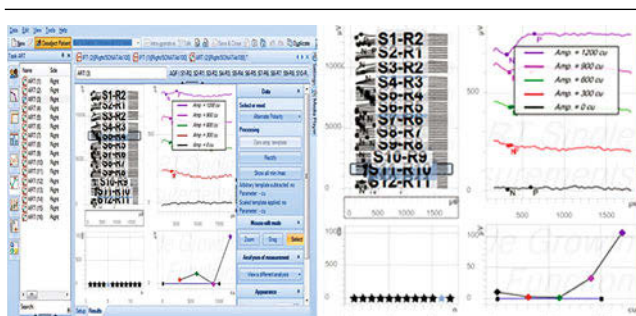
test. Despite the considerable difficulty during surgery and the severe degree of gusher necessitating obliteration of the middle ear and closure of the Eustachian tube, this child developed auditory abilities with little difference from that generally expect from those without any congenital anomalies or CSF gusher during surgery. The audiologic performance of the second patient (her twin) was slightly better as she showed a pure tone threshold of 35 dB, and answered 100% to a closed set and 60% to an open set speech perception test. In contrast, the performance of the third case was subsatisfactory, he had a pure tone threshold of 60 dB, scored 60% to a closed set, and 32% to an open set speech perception test, and these results were attributed to the very young age of the child at the time of implantation and also to the low socioeconomic background of his parents.

(b) Two cases with LVAS: In these patients, the vestibular aqueduct diameter was larger than 1.5 mm at the midpoint, and their diameter was more than double the posterior semicircular canal on axial CT scan.

One patient was implanted when she was 24 months old with a Nucleus freedom straight array, whereas the other patient was implanted at 36 months of age with Medel Sonata. The CSF gusher in the first case was removed by continual suction, elevation of the patient's head, decreasing the end-tidal CO₂ below 28, and hyperventilation of the patient until the egress stopped after a few minutes even before we started inserting the electrode array, whereas in the other case, CSF continued to leak during the insertion, but was stopped by sealing of the cochleostomy around the implant array with the periosteum.

One year after the implantation, the average pure tone threshold for both of them was 36 dB; the average response was 89% to the closed set speech perception threshold and 67% to open set tests (Fig. 6).

Figures 5



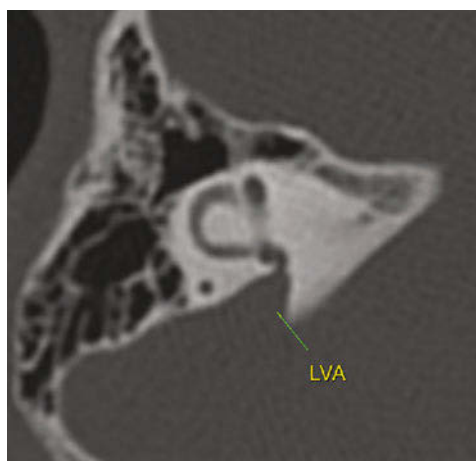
Mondini malformation: evoked response telemetry (ERT) 1 month after surgery. (a) No ERT wave forms could be detected in the apical and the middle channels; (b) good ERT wave forms on the basal channels.

(c) One case was diagnosed with Pendred syndrome: Being the second child of a family with a history of a deaf child, 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. The preoperative radiology showed Mondini cochlear dysplasia with large vestibular aqueduct; a genetic study confirmed the diagnosis as Pendred syndrome, and her mother gave a history of thyroid disease, which was first diagnosed during her first pregnancy (Fig. 7).

At the time of surgery, severe CSF gusher occurred immediately at the time of cochleostomy; the gusher in this case was the most aggressive gusher we encountered among the nine cases. The CSF continued to egress vigorously after the insertion of the electrode array (Medel Sonata); packing of the basal turn around the electrode array with sealing of the cochleostomy failed to control the continuous flow. Obliteration of the whole middle ear with muscles after removal of the incus and its buttress was carried out to allow proper visualization and closure of the Eustachian tube. No further leak was detected after all these measures during wound closure, and there was no need for lumbar drain insertion, despite the vigorous CSF leak. Tight bandage, a parenteral broad-spectrum antibiotic, and a carbonic anhydrase inhibitor (Cidamax tablets) were used for 1 week after the surgery.

No ERT was present on initial intraoperative testing of the implant; the impedance of the electrode array was within the range of compliance, except for channel 8, which showed an open circuit, indicating a broken channel; however, good ERT responses started to appear clearly for the first 6 channels on the second

Figures 6



Axial computed tomography (CT) scan of the left temporal bone showing a very wide vestibular aqueduct [large vestibular aqueduct syndrome (LVAS)] with normal developed cochlea.

visit for mapping; we were obliged to leave the last two channels switched off because of the high current level needed to induce auditory response, but with very distressing facial nerve stimulation.

One year after the implantation, she had an average threshold at 52 dB, responded 68% to a closed set and 63% to open set tests. At that time, all the electrodes were active, except electrode number 8. She has no problem in hearing any sounds except hearing crackling sensations, especially on stimulation of channels 11 and 12. Her ability to talk relatively progressed regarding her age and duration from implantation.

(d) One case with a wide IAC: This case was detected at the time of surgery after CSF gusher was encountered; on careful reviewing, the diameter of the IAC on an ultrathin-cut CT scan was more than 10 mm; the cochlea and the vestibule showed normal development radiologically, and no other congenital inner-ear malformation was noticed. The partition between the lateral end of the canal and the inner ear appeared deficient (deficient lamina cribrosa). This 46-month child was implanted with a Medel Pulsar straight array. During surgery, aggressive egression of the CSF was encountered from the cochleostomy site, which continued to leak for more than 15 min; the leak also persisted from around the electrode array after insertion, but stopped after complete sealing of the cochleostomy with a small piece of the periosteum. Immediate intraoperative testing of the electrode array showed that the impedance of the 12 channels was within the range of compliance, but no ERT was detected at any channel even at a high current level, which started to appear 3 weeks after

the surgery on initial stimulation of the cochlear implant at our audiology unit.

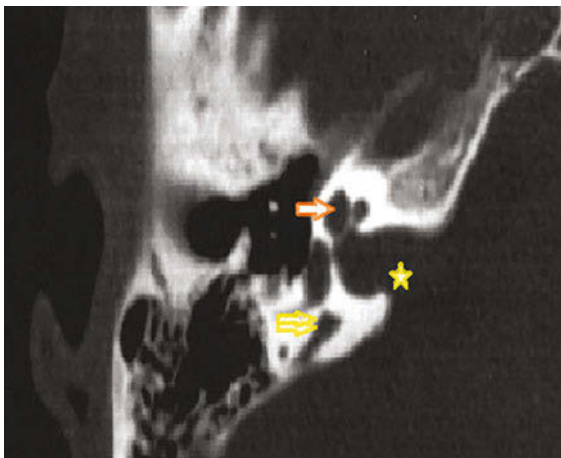
One year after surgery, the patient had a pure tone threshold hearing of 32 dB, responded 58% to a closed set speech perception threshold and 31% to an open set test (Fig. 8).

(e) One case with common cavity: In this patient, the whole inner ear was replaced radiologically by a cavity, which comprised both the vestibule and the cochlea, the common cavity was anterior to the IAM in the axial cuts, and the IAM opened in the centre of the cavity. Hearing loss was discovered at the age of 12 months; audiologic assessment showed bilateral profound hearing loss and a CT scan showed bilateral common cavity inner-ear deformity.

Cochlear implantation was performed at the age of 24 months. CSF gusher occurred, which did not stop spontaneously; full insertion of a straight electrode array (Nucleus 24K) was followed by packing of the whole cavity with small pieces of muscles till the CSF started to decrease, and this was followed by sealing of the cochleostomy periosteum.

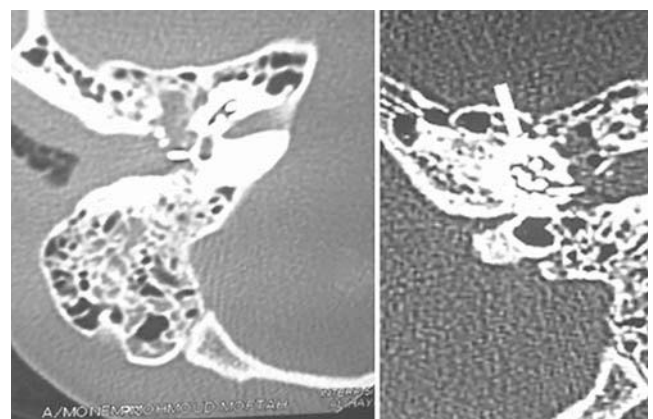
The postoperative period was subsatisfactory. Implant electric auditory brain stem response was performed; the patient responded to four electrodes at the time of activation and programming, but gradually, the number of active electrodes was increased to seven out of 22 electrodes at 6 months after surgery. The common ground artifact, which allows the assessment of the waveform reversal in phase around electrode 9 as the array takes the first turn around the cochlea, showed that the electrode array folded back on itself, and there was more than one point of reversal (Fig. 9).

Figures 7



Axial computed tomography scan of the left ear showing confluence of the apical-turn and the middle-turn Mondini malformation (red arrow) with a large vestibular aqueduct (yellow arrow) and a very wide internal auditory meatus (IAM) (yellow star).

Figures 8



Axial-cut computed tomography (CT) scan showing a wide internal auditory canal: the implant array appears inside the cochlear lumen; the cochlea shows normal development.

The 1-year postoperative audiologic evaluation was very subsatisfactory as the child, after 12 months, had a threshold hearing of 72 dB hearing level, responded to 32% to a closed set, but was not yet able to score in open set tests (Fig. 10; Table 2).

Discussion

Although the occurrence of CSF gusher during cochlear implantation has been described by many authors in the literature, the incidence and the management of this surgical difficulty during surgery remains very variable among different institutes.

Kempf *et al.* [7] reported an overall incidence of CSF gusher as it relates to the implantation itself of about 0.01%. Papsin reported that gusher was present in 6.7% of the 103 patients with malformations. They stressed on the importance to distinguish between the actual pulsatile egress of fluid and the simple ooze of CSF so as not to overestimate the number of CSF leaks, explaining the wide range of variability of the incidence of gusher [4].

Phelps and colleagues described two different types of CSF outflow during cochleostomy: gentle flow of clear fluid is called 'oozing' and a profuse flow is termed 'gusher'. Oozing is the result of a small defect between the malformed inner ear and the IAC. In the author's practice, oozing is an intermittent flow of CSF in small quantities, which usually stops after a few minutes. The defect between the IAC and the malformed ear is small and the CSF outflow is easily controlled with

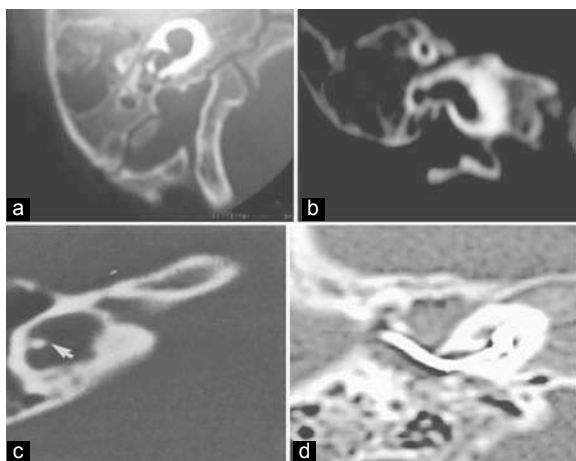
soft tissue packing around the electrode, whereas in cases of gusher, there is a wide communication between the subarachnoid space and the inner ear. In these patients, there is profuse CSF outflow on making the cochleostomy; this type of CSF flow is more common in type II [8].

Excessive CSF can usually access the cochlea through patent developmental pathways of the otic capsule, the most common of which are a wide IAC, common cavity malformation, and a large vestibular aqueduct (LVAS), and any anomaly that results in incomplete turns in the cochlea increases the risk of a CSF gusher as in cases of incomplete partition type II termed after Mondini [1,9]. In a review of the literature, most authors reported the incidence of gusher to be between 40 and 50% of their patients with inner-ear malformations; CSF leak from the cochlea can usually be anticipated as this occurs most often in cases of a congenitally abnormal inner ear [10–12].

Not all cases with inner-ear malformations have gusher during surgery. Sometimes, despite a wide defect at the end of the IAC, no gusher occurs upon entering the inner ear [13]. The majority of labyrinthine anomalies will be uncovered with preoperative imaging; even after careful preoperative evaluation, unexpected gusher may occur in the absence of any labyrinthine development malformation.

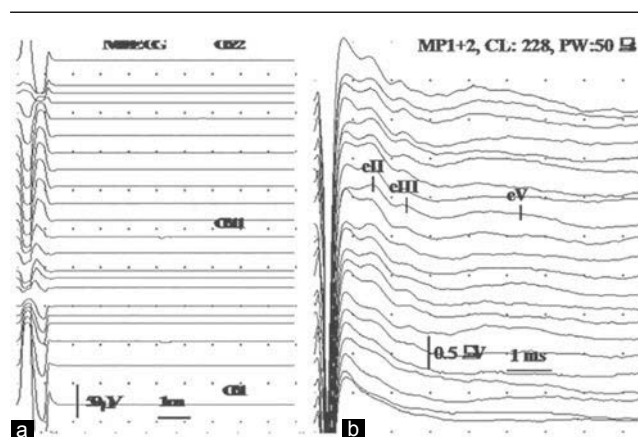
In the present study, eight out of the nine patients who developed gusher showed preoperative radiologic evidence of congenital inner-ear malformation; the incidence of gusher among the 18 cases with inner-ear malformation was 44% and was significantly high when compared with the other group, which did not show any developmental

Figures 9



(a, b) Axial and coronal computed tomography (CT) scan of the right temporal bone showing common cavity deformity. Note the confluence of the cavity, which comprises both the vestibule and the cochlea; the internal auditory meatus (IAM) opens in the centre of the cavity, with a deficient cribriform plate separating the cochlea from the IAM. (c, d) The electrode array within the common cavity.

Figures 10



(a) An implant electric auditory brain stem response common ground artifact showing more than the point of reversal, meaning that the electrode array folded back on itself; (b) a few of the electrodes yielded responses at the initial time of stimulation using the monopolar mode of stimulation.

Table 2 Patient information and intraoperative findings

Number	Inner-ear anomaly	Age at surgery	Type of device	How the gusher stopped	Average PTA	Closed SPT (%)	Open SPT (%)
1	Mondini malformation	12 months	Medel Sonata	Stopped after electrode insertion	60	60	32
2	Mondini malformation	36 months	Medel Pulsar	Sealing the cochleostomy around the electrode by pieces of fascia stopped the gusher	35	100	60
3	Mondini malformation	36 months	Medel Pulsar	Packing of the whole middle ear and Eustachian tube closure+packing of basal turn of the cochlea around the electrode with muscles+sealing cochleostomy	38	80	45
4	LVAS	24 months	Nucleus freedom straight array	Stopped spontaneously before electrode insertion	38	82	60
5	LVAS	36 months	Medel Sonata	Electrode insertion+sealing the cochleostomy	34	96	74
6	Mondini with LVAS Pendred syndrome	15 months	Medel Sonata	All measures+obliteration of the whole middle ear and closure of the Eustachian tube	52	68	63
7	Abnormal wide IAC	46 months	Medel Pulsar	Stopped after electrode insertion	32	58	31
8	Common cavity	42 months	Nucleus 24K	Packing of the whole cavity after electrode insertion also to insure alignment of the electrode to the wall of the cavity+cochleostomy sealing	72	32	0
9	No bony cochlear malformation	32 months	Medel Pulsar	Sealing the cochleostomy around the electrode by piece of fascia stopped the gusher	32	100	42

IAC, internal auditory canal; LVAS, large vestibular aqueduct; PTA, average pure-tone audiometry 12 months after surgery; SPT, speech perception threshold testing.

malformations (1.51%). All cases with modiolar defects in the 18 cases showed gusher. Three cases with a wide IAC, four cases with preoperative radiologic evidence of cochlear hypoplasia, two cases of hypoplastic semicircular canal, and lastly one case with a wide vestibular aqueduct did not develop gusher during surgery.

It is important that the CT scan of the inner ear be reviewed carefully before surgery. This serves a two-fold purpose. One is to assess the ability to insert the whole array and the second is to be prepared for a possible gusher. The family and the patient can be counseled appropriately. They should be warned of potential complications, the potential for prolonged hospital stays, and the incidence of meningitis, despite the meningococcal vaccine was given for all cases prior to surgery.

In our study, the incidence of gusher was 10.7%; this high incidence compared with other studies may be attributed to the small number of the studied group and also to the inclusion of cases with a simple CSF ooze, in which the gusher stops spontaneously by continuous suction. In the current study, we were able to achieve a scoring system for the gusher according to the measures needed to stop the gusher (Table 3) (Figs. 11–14).

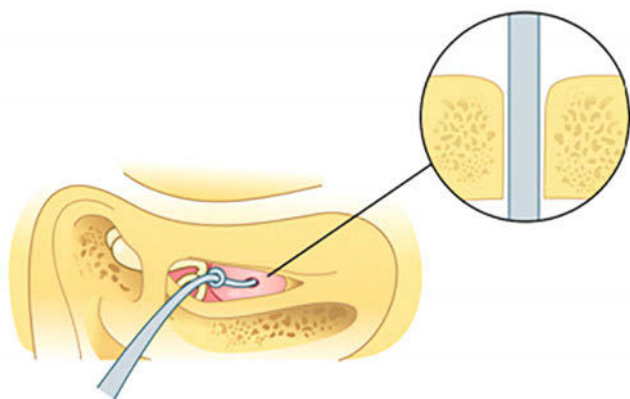
We classified CSF egress during cochlear implantation into simple CSF ooze, which stops spontaneously by continuous suction, positioning of the patient in the anti-Trendelenburg position, hyperventilation, and decreasing the end-tidal CO₂ below 28. Simple gusher included cases in which the gusher stopped with simple insertion of the electrode array, whereas when gusher stopped on sealing the cochleostomy with muscles harvested from the temporalis muscle, it was termed mild gusher. In some cases with moderate to severe gusher basal turn packing around the electrode array and packing of the whole middle ear and closure of the Eustachian tube consecutively was required. Among the nine cases, CSF gusher was severe in one case with Mondini malformation and another case of Mondini with a large vestibular aqueduct. In the case with common cavity malformation, the gusher stopped, obviously regressed after electrode insertion, and packing of the cavity was performed only to ensure the alignment of the electrode array to the cavity wall to ensure stimulation of the largest number of the spiral ganglion neuron, which is already sparse among this entity of congenital inner-ear malformations.

Table 3 Cerebrospinal fluid gusher staging

Stages	Measures needed	
0. No CSF gusher	No CSF leak	No CSF leak
1. Simple CSF ooze	CSF leak stops spontaneously or by simple medical measures	The egress of CSF stops either spontaneously by continuous suction, positioning of the patient in the anti-Trendelenburg position, hyperventilation, decreasing the end-tidal CO ₂ below 28
2. Simple gusher	Simple insertion of the electrode array stops the CSF gusher	Figure 11
3. Mild	Sealing of the cochleostomy site with the periosteum	Figure 12
4. Moderate	Packing of the basal turn around the electrode array	Figure 13
5. Severe	Packing of the whole middle ear and closure of the Eustachian tube	Figure 14

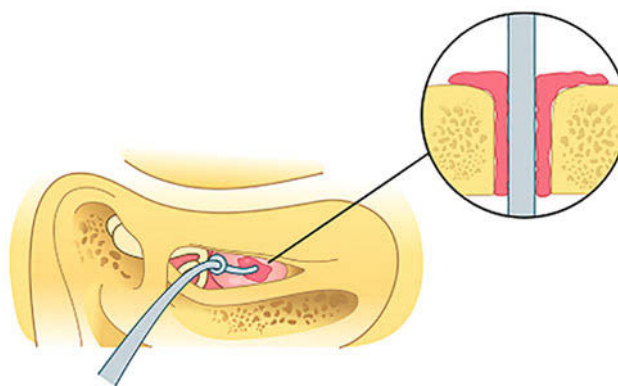
CSF, cerebrospinal fluid.

Figures 11



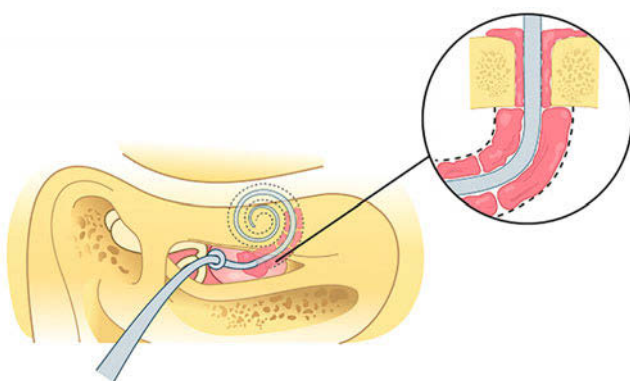
Simple insertion of the electrode array stopped the cerebrospinal fluid (CSF) gusher.

Figures 12



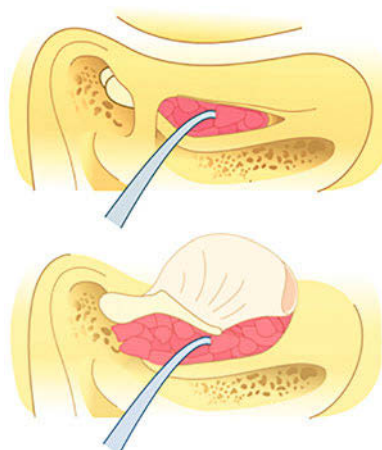
Sealing of the cochleostomy site with the periosteum, the fascia, or muscles

Figures 13



Packing of the basal turn around the electrode array.

Figures 14



Packing of the whole middle ear and closure of the Eustachian tube.

The stepwise algorithm for the management of cerebrospinal fluid gusher during cochlear implantation

In the current study, CSF gusher was expected in 17 cases with preoperative radiologic evidence of developmental labyrinthine anomalies; seven of them

developed CSF gusher during cochleostomy; two cases were detected at the time of surgery after CSF gusher was encountered; in one case, the preoperative radiologic evaluation showed normal labyrinthine

development radiologically, and no other congenital inner-ear malformation; on carefully reviewing, the diameter of the IAC on an ultrathin-cut CT scan was more than 10 mm; the cochlea and the vestibule were noticed, whereas in the other one, there was no malformation, with a normal IAC diameter and no obvious cause of gusher detected radiologically.

In gusher-prone cases, when the gusher was encountered, a special management algorithm to control this gusher was carried out as follows (Fig. 15).

Nonsurgical measures

Head elevation and positioning of the patient in the anti-Trendelenburg position, hyperventilation, and decreasing the end-tidal CO₂ below 28; the main aim of this measure is to decrease the cerebral blood flow with a subsequent decrease in the CSF pressure and thus decrease the CSF leak.

Positioning of the patient usually causes a transient decrease in the cerebral blood flow, usually lasting transiently for a few minutes, which is sufficient enough to insert the electrode array into the cochlear lumen until reflex increase in the cardiac output with subsequent increase in the intracranial tension.

Hyperventilation causes a decrease in the cardiac output and subsequently decreases the intracranial pressure and the CSF pressure.

Decreasing the end-tidal CO₂ below 28mmHg causes cerebral vasoconstriction.

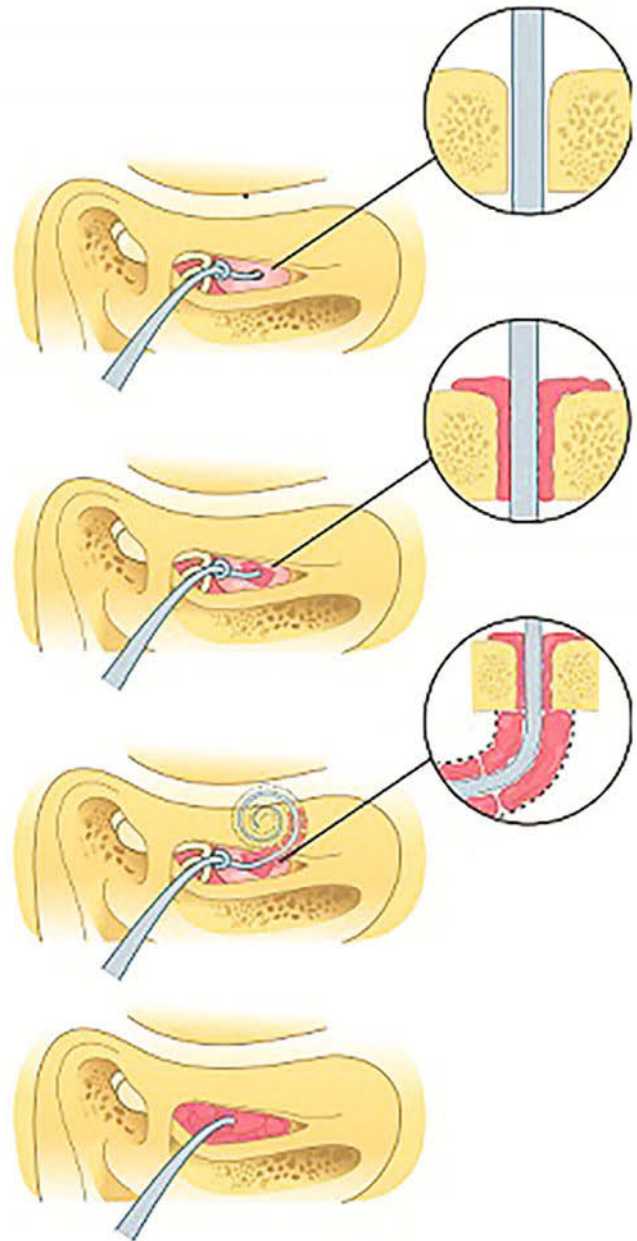
In two out of the nine cases with gusher, the CSF was removed with continual suction with previous measures until the gusher egress stopped or slowed down sufficient enough to introduce the electrode array, which ensured total arrest of gusher.

Electrode array insertion through the cochleostomy

CSF gusher increases the technical difficulty of the operation, because refraction through the clear fluid makes it less easy to visualize the electrode as it passes through the cochleostomy. At the same time, the surgeon may introduce the electrode array towards a bony structure, and the tip of the electrode may be damaged [13,14].

During cochlear implantation in gusher-prone cases, there is a debate about the size of cochleostomy; some authors claimed that a small cochleostomy is the one of choice, allowing the electrode cable to partly block the flow of CSF, whereas others reported the importance of a slightly larger cochleostomy to

Figures 15



The stepwise algorithm for the management of cerebrospinal fluid (CSF) gusher.

allow extra mobility with the instruments to pack the cochleostomy reinforced with connective tissue, muscle, and fibrin [4,7].

In this study, we used a 1-mm course diamond bur to create the cochleostomy anteroinferior to the round window. The tip of the suction was introduced at the edge of the cochleostomy, making a continual suction of CSF either totally or until sufficient visualization was obtained to introduce the electrode array into the cochlear lumen without traumatizing the tip of the electrode array.

In the present study, electrode insertion was sufficient to stop the egress of gusher in two

cases, with no need for using soft tissue around the cochleostomy site or to be enforced around the electrode in the basal turn. Sealing around the electrode array with fibrin glue was performed just to confirm no postoperative leak.

Packing the cochlea: either sealing the cochleostomy site or whole packing the basal turn

Firm packing around the electrode is important in patients with gusher. Continuous leak usually has bad sequels, including the risk of CSF rhinorrhea through the Eustachian tube or continuous leak through the incision site with subsequent meningitis, and the risk of electrode extrusion [15,16].

The application of soft tissue (muscle or fascia) is important in cases of gusher. The fascia or the muscle can be applied around the cochleostomy site or packing of the basal turn around the electrode array may be carried out.

In this study, sealing the cochleostomy site after electrode insertion was sufficient to stop CSF leak in three cases, and there was no need to obliterate the whole basal turn around the electrode array. Two cases needed further application of soft tissue into the cochlear lumen: one of these two cases was a common cavity and the soft tissue used was used not only to stop the gusher but also to align the electrode array against the wall of the common cavity so as to stimulate the largest possible number of spiral ganglion neurons, which is usually sparse in this type of deformity.

Middle-ear obliteration with subsequent closure of the Eustachian tube

This technique is usually left for cases with severe CSF gusher, which continue to egress vigorously after the insertion of the electrode array and packing of the basal turn around the electrode array with sealing of the cochleostomy fails to control the continuous flow.

In our study, we needed this technique in only one case with radiologic evidence of Mondini with LVA, in which obliteration of the whole middle ear with muscles after removal of the incus and its buttress to allow proper visualization and closure of the Eustachian tube to prevent escape of the CSF into the nasopharynx was performed.

No intraoperative or postoperative lumbar drain was performed in any of our cases; there was no need for the use of any special electrode in any case, and also, no postoperative CSF leak or meningitis was encountered.

Owing to the small number of cases in this study we were not able to perform a statistical analysis of the data or compare them with cases without gusher, but in general, the immediate intraoperative electrophysiologic study of the implant showed no responses at all channels in all cases, whereas initial (1 month after surgery) and late postoperative evoked responses of the implant were within normal ranges, except for the common cavity case, in which the responses was very subsatisfactory; also, except for this case, the postoperative audiologic performance in all cases was not evidently different from that generally expected from those without any congenital anomalies.

Conclusion and recommendation

- (1) CSF gusher is a well known and expected surgical difficulty rather than a complication of cochlear implantation. Proper preoperative evaluation will help the surgeon to sort out gusher-prone cases to be prepared for this difficulty.
- (2) In this study, we established a scoring system for cases of CSF gusher according to the different surgical techniques needed.
- (3) In this study, we were able to perform a simple logic stepwise algorithm for the management of CSF gusher if encountered during cochlear implantation.
- (4) CSF gusher should be considered as a surgical difficulty that a cochlear-implant surgeon may experience rather than as a complication or a predictor of the postoperative performance of cases that develop this difficulty during surgery.
- (5) Owing to the small number of cases in our study, we were not able to evaluate the use of different types of implant arrays with a stopper at the cochleostomy site provided by different companies or include other aggressive measures to stop the gusher, such as subtotal petrosectomy, because we did not need them.
- (6) Our study was carried out on a small number of cases; future study on a larger number of cases or a multicentric experience might be needed.

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Conflicts of interest

There are no conflicts of interest.

References

- 1 Janssens S, Govaerts PJ, Casselman J, Van Rompaey W, Van Langenhove A, Somers T, Offeciers FE. The LAURA multichannel cochlear implant in a true Mondini dysplasia. *Eur Arch Otorhinolaryngol* 1996; 253:301–304.
- 2 Miyamoto RT, Robbins AJ, Myres WA, Pope ML. Cochlear implantation in the Mondini inner ear malformation. *Am J Otol* 1986; 7:258–261.
- 3 Fahy CP, Carney AS, Nikolopoulos TP, Ludman CN, Gibbin KP. Cochlear implantation in children with large vestibular aqueduct syndrome and a review of the syndrome. *Int J Pediatr Otorhinolaryngol* 2001; 59:207–215.
- 4 Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. *Laryngoscope* 2005; 115(Suppl 106):1–26.
- 5 Slattery WH III, Luxford WM. Cochlear implantation in the congenital malformed cochlea. *Laryngoscope* 1995; 105:1184–1187.
- 6 Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope* 1987; 97(Suppl 40):2–14.
- 7 Kempf HG, Tempel S, Johann K, Lenarz T. Complications of cochlear implant surgery in children and adults. *Laryngorhinootologie* 1999; 78:529–537.
- 8 Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. *Am J Otol* 1994; 15:551–557.
- 9 Loundon N, Rouillon I, Munier N, Marlin S, Roger G, Garabedian EN. Cochlear implantation in children with internal ear malformations. *Otol Neurotol* 2005; 26:668–673.
- 10 Luntz M, Balkany T, Hodges NV, Telischi FF. Cochlear implant in children with congenital inner ear malformations. *Arch Otol Head Neck Surg* 1997; 123:974–977.
- 11 Juich I, *et al.* Surgical considerations regarding cochlear implantation's in congenitally malformed cochlea. *Otol Head Neck Surg* 1998; 121: 495–498.
- 12 Sennaroglu L, Sarac S, Ergin T. Surgical results of cochlear implantation in malformed cochlea. *Otol Neurotol* 2006; 27:615–623.
- 13 Beltrame MA, Frau GN, Shanks M, Robinson P, Anderson I. Double posterior labyrinthotomy technique: results in three Med-El patients with common cavity. *Otol Neurotol* 2005; 26:177–182.
- 14 Graham JM, Ashcroft P. Direct measurement of cerebrospinal fluid pressure through the cochlea in a congenitally deaf child with Mondini dysplasia undergoing cochlear implantation. *Am J Otol* 1999; 20: 205–208.
- 15 Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformations. *Arch Otolaryngol Head Neck Surg* 1997; 123:974–977.
- 16 Sennaroglu L, Aydin E. Anteroposterior approach with split ear canal for cochlear implantation in severe malformations. *Otol Neurotol* 2002; 23: 39–42 discussion 42–43.