CSF Fistula and Meningitis

12

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Special Features

1. Oval window is usually the site for spontaneous CSF fistula.
2. Cochleostomy can also be the source of CSF leakage.
3. Mostly seen in IP-I, to a lesser extent in CH-I and CH-II.
4. Stapes footplate defect is most probably due to endosteum developmental anomaly as a result of defective vascular supply from IAC.
5. Recurrent meningitis is common.
6. Vaccination against pneumococcus is very important but not sufficient.
7. Proper sealing of the leakage area with fascia in a dumbbell fashion is mandatory.
8. Subtotal petrosectomy may be necessary in some cases.
9. Stapes footplate must be examined for fistula during CI surgery in every IP-I and CH-II cases.
10. Surgeon should not leave operation theater without fully stopping the leakage.

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12.1 Introduction

Today, inner ear malformations (IEM) are among the most important causes of recurrent meningitis with otogenic origin. In addition, cochlear implantation (CI) in this particular group of patients can also cause recurrent meningitis. Cerebrospinal fluid fistula (CSF) in IEMs is associated with considerable morbidity and mortality, usually presenting a diagnostic challenge to the otologist. It may easily lead to recurrent meningitis unless, there is a surgical intervention to repair the fistula. Majority of the CSF fistulae are located at the stapes footplate [1–6]. Rarely one may encounter CSF fistula at the cochleostomy site [7]. **As a surgical principle, surgeon should not leave operation theater without completely stopping the leakage.** Therefore, diagnosis and treatment of CSF leaks are very important to prevent meningitis in this special patient group.

There are two groups of patients: spontaneous CSF leakage in IEMs and CSF leakage after CI surgery. Trauma can also produce CSF leaks in this special group of patients easier than in subjects with normal anatomy. Both groups present with recurrent meningitis. It is more difficult to diagnose the pathology in spontaneous CSF leaks. High suspicion is the key to diagnosis. Therefore, **in cases of recurrent meningitis it is mandatory to evaluate inner ear radiologically to rule out an IEM**.

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12.2 Histopathology and Pathophysiology

In 2016 Sennaroglu [8] reported the histopathology and possible pathophysiology in inner ear malformations. The findings provided understanding of the etiology and changed the concept of "CSF pressure causing footplate defect." Findings suggested that defective endosteum is most probably the cause of IP-I. All five patients with IP-I pathology in Massachusetts Eye and Ear Infirmary (MEEI) had a very thin and defective endosteum (innermost layer of the otic capsule) all around the cochleovestibular space, while the middle enchondral and outer periosteal layers were normal.

Three of the five cases with IP-I had a defect involving the stapes footplate. The defect was covered with a thin membrane. Embryologically, the vestibular part of the stapes footplate is derived from the endosteum, and a defective endosteum may be responsible for this defect.

If the embryology of the inner ear is investigated, it is noted that three layers of otic capsule have different vascular supply [9]. While inner endosteum (inner periosteal layer) has a vascular supply coming from the IAC, middle enchondral and outer periosteal layers have their vascular supply from the middle ear mucosa. According to Donaldson [9], blood vessels from the IAC supply the developing modiolus, the walls of the scala, the osseous spiral lamina, and the partition between cochlear turns. Therefore, it looks quite possible that histopathological changes in IP-I may be a result of defective vascular supply from the IAC blood vessels. As the vestibular surface of the stapes footplate is also derived from the endosteum, defective footplate development may be the result of abnormal endosteal development due to vascular arrest coming from the IAC. During development of the fetus, vascular channels are being formed circumferentially on the footplate, through the growth of the endosteal bone around blood vessels that are already present. Therefore, reduced vascular supply may cause a defective footplate.

In MEEI temporal bone collection, there were three IP-I specimens with a thin, but intact modiolar base, and a stapes defect at the same time (Fig. 12.1). Therefore, it is possible to have stapes footplate defect in cases with no CSF filling the cochlea. This is a very important finding demonstrating that high CSF pressure cannot be held responsible for the defective development at the stapes footplate. The author has operated on 11 cases with spontaneous CSF fistula and found that majority of the cases had a cystic structure present at the stapes footplate. Once it was punctured, CSF gushed out from this defect. This was repaired by introducing a piece of fascia through the defect into the vestibule, in a dumbbell fashion. Sometimes a bony defect with CSF leakage was also encountered. It is possible that in IP-I cochlea, where the modiolus is completely absent, high CSF pulsations acting on the thin membrane at the footplate may easily produce an oval window fistula. If there is a bony separation between the IAC and the cochlea, the footplate defect may not be noticed at all during the patient's lifetime. Stapes footplate fistula can only be noticed if there is a middle ear infection which may cause meningitis when the infection passes through the thin membrane at the oval window into CSF filling the cochlea.

None of the 11 spontaneous CSF fistulas due to IEMs operated on by the first author had a fis-

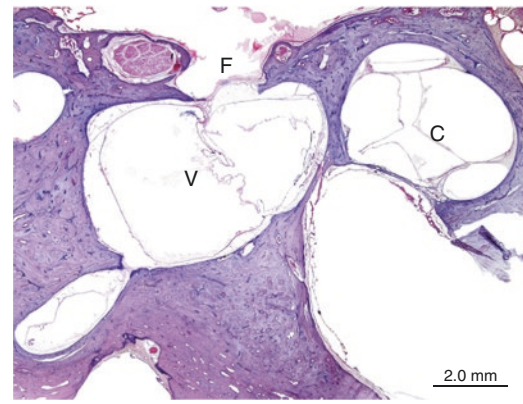

Fig. 12.1 Histopathological specimen showing stapes footplate where the normal bony footplate is replaced completely by thin fibrotic membrane (F) with intact modiolar base in a patient with IP-I specimen. As the cochlea is completely separated from internal auditory canal high cerebrospinal fluid pressure cannot be held responsible for the defective footplate (with permission of Massachusetts Eye and Ear Infirmary)

tula at the round window. All of the reported spontaneous CSF fistula cases in IP-I in the literature are located at the oval window [3, 5, 6, 10]. This also shows that this is observed in cases with a defective footplate.

In IP-III, which is observed in X-linked deafness, there is a larger defect in the fundus, a high pulsating CSF pressure in the cochlea. All 11 IP-III cases operated in our department had a severe gusher upon cochleostomy, but a spontaneous CSF fistula has never been encountered. The reason may be that the endosteum, which is deficient in IP-I (probably due to defective vascular supply coming from the IAC), is properly formed in IP-III. In IP-III, it looks possible that the endosteum is well developed. There is no histopathological specimen with IP-III but from HRCT it looks as if there is a thick endosteal layer in IP-III, while outer two layers of the otic capsule are missing. This causes stapes fixation without any fistula at the stapes footplate. Therefore, spontaneous CSF fistula formation has not been observed by our group or reported in the literature in IP-III. In other words, high CSF pressure cannot be held responsible for spontaneous footplate fistulas.

Another question is whether the footplate defect is present at birth or develops in time. The defect is most probably present at birth. High CSF pressure can cause a fistula through the defective footplate, or otitis media during childhood may result in recurrent meningitis. All cases that have been operated on in our department are children. No adult patient has been operated on so far: it appears that it is not a progressive disease, and that it must be present during childhood. If it had been the result of high pressure only, it would have been possible to see this clinical entity at all ages. This shows us that high pressure is not necessary all the time for the development of the oval window fistula. The defective development is most probably a result of a deficient periosteum present at birth, but high CSF pressure may produce a fistula in this already defective area. The prevalence of otitis media decreases considerably in adults. Therefore, adult patients with IP-I have very little chance to develop otitis media and meningitis.

All these features are valid for CH-I and CH-II as well. The only difference is the size of the cochlea; dimensions of the cochlea are less in CH-I and CH-II compared to IP-I. The occurrence of a gusher, a spontaneous CSF fistula at the footplate, and all other clinical findings can be observed in CH-II as well. The author has seen a spontaneous CSF fistula in two and a gusher during CI surgery in four CH-II cases and one spontaneous CSF fistula in CH-I.

12.3 Literature Review

In 1997 Hoffman et al. [11] reviewed the literature, along with the results of 200 institutions performing cochlear implants (which were queried by questionnaire) about IEMs. Out of 50 cases none had meningitis. In spite of this report, at present, IEMs are one of the most important causes of recurrent meningitis. This is the reason for devoting a chapter on this topic.

Phelps et al. [3] reported that meningitis occurred in 40% of the patients with severe cochlear dysplasia. They observed that patients with wider basal turn, possibly incomplete partition type I (IP-I), are more prone to risk of CSF fistula, whereas none of the patients with normal basal turn and enlarged vestibular aqueduct (possibly IP-II cases) had meningitis. Most probably their patients with "wider basal turn" are IP-I cases, where absence of interscalar septa and modiolus caused their basal turn look enlarged. This was an important observation indicating that stapes footplate defect was more often encountered in IP-I cases. However, the reason for recurrent meningitis is the absence of cribriform plate and modiolus resulting in a wide defect between the IAC and the cystic cochlea.

In 2010 Sennaroglu [1] reported in his review of IEMs that a wide defect in the cribriform plate and modiolus may cause the CSF to come adjacent to the medial surface of the oval and round window. Continuous CSF pressure may cause erosion, bony defect, and fistula at the stapes footplate. At that time CSF pressure was held responsible for bony erosion at the footplate. Recently, In 2016 Sennaroglu [8] reported the

histopathology and possible pathophysiology in IEMs and came to the conclusion that footplate defect is the result of endosteal developmental anomaly seen in IP-I and CH-II. If there is CSF filling cochlea, a middle ear infection causes meningitis when pathogens pass through the cystic or membranous footplate into inner ear.

In the literature, majority of the spontaneous CSF fistulas are reported to be located in the oval window. Phelps et al. [3] reported one patient with meningitis whose exploration of the middle ear revealed ballooning of the mucosa over stapes footplate and removal of the stapes produced a CSF gusher. Histopathological examination of another patient who had died of meningitis revealed a defect in the stapes footplate. Similarly Syal et al. [10] reported four cases where the site of leak was in and around oval window in all cases.

Shetty et al. [6] described two patients who had Mondini malformation in whom CT cisternography showed a CSF fistula at the lamina cribrosa (lateral wall of the internal auditory canal). They had CSF rhinorrhea in the presence of an intact tympanic membrane. Contrast material was seen in the middle ear cavity, having leaked through a defect in the lamina cribrosa and from there to an enlarged vestibule through the oval window. The cochlea was represented by a sac like diverticulum from the vestibule without apical turns. During surgery, they discovered a defect at the posterior aspect of the oval window, where the CSF was leaking.

It is possible to have multiple leak sides. Da Cruz et al. [5] reported a case with recurrent meningitis due to oval window fistula. After repairing the fistula with vein and fat graft, rhinorrhea continued and meningitis recurred. CT images in their report resembled a common cavity deformity. In their investigation, CT cisternography demonstrated two fistula sites: one at the oval window and the other in the tegmen. During revision surgery, two fistula were discovered: one fistula at the oval window and a second fistula at the tegmen. This necessitated a combined middle fossa and transmastoid approach. They made an important contribution that it is very important to look for another site of CSF fistula in recurrent cases.

CSF fistula may be seen in common cavity malformation as well. Mylanus et al. [4] reported recurrent meningitis in a patient with common cavity. Similar to other cases middle ear exploration revealed a fistula at the oval window which was sealed with temporalis fascia.

Although rarer than oval window, sometimes the leak site can be the cochleostomy. Page and Eby [7] reported a case of meningitis after minor head trauma developing 2 years after cochlear implantation in a child with Mondini malformation. This is the first case of meningitis after CI surgery in IEMs. The images in their report resemble IP-I malformation. CSF leakage was located at the cochleostomy around the electrode, and this was sealed with a temporalis fascia and muscle plug.

Contralateral side may also be the source of CSF leak and cause for meningitis in an implanted patient. Bluestone [12] stated that sometimes the implant may not be associated with the pathogenesis of post-implantation meningitis. Suzuki et al. [13] reported the temporal bone histopathological findings of a 6-year-old boy who had a CI surgery at another institution and was admitted with acute, fulminating pneumococcal meningitis, and died. His temporal bones were removed and the histopathology revealed bilateral Mondini malformations that had been implanted in his left ear. Interestingly, he had otitis media and labyrinthitis in the contralateral, right, non-operated ear that had spread to his meninges. The cochlear implant was not involved in the pathogenesis of the meningitis but the underlying inner ear anomaly and otitis media were. The left side with the cochlear implant was completely normal with no infection in the middle ear and no disruption in the footplate, annular ligament and the round window membrane. However, the non-operated right side had normal footplate and annular ligament but he had inflammatory necrosis of the round window membrane with many leucocytes in the scala tympani adjacent to the tympanic membrane. The suppurative labyrinthitis was attributable to the spread of middle ear infection through the round window membrane. The further spread of infection into intracranial CSF space was through the defective modiolus.

Findings in this patient demonstrated that round window may rarely be involved in transmission of infection as well.

12.4 Radiology

Radiology is the most important tool to diagnose preoperatively the CSF leak and the predisposing anatomic factors. HRCT is superior to MRI in demonstrating the bony defects or fractures. While MRI demonstrates the fluid characteristics better than HRCT. Shetty et al. [6] stressed the importance of CT in the evaluation of patients with CSF fistula. The presence of IEMs, a defect in the lamina cribrosa, or a bone fracture on HRCT may be the etiology of CSF gusher. CT cisternography can show CSF fistula in patients with CSF otorhinorrhea and unilateral hearing loss. A noninvasive method for confirming this finding is a fast spin-echo T2-weighted MR sequence through the region [14]. Retrospectively, Shetty et al. [6] concluded that plain high-resolution CT study of the temporal bones coupled with coronal MR cisternography of this region with the use of a fast spin-echo T2-weighted sequence would have noninvasively shown the site of the CSF fistula in their patients CSF fistula. Syal et al. [10] also found MRI (using 3D FSE T2WI and 3D FIESTA sequences) a useful technique in the assessment of patients with CSF fistulae; it is noninvasive, offers excellent anatomical detail, and has no radiation risk. Da Cruz et al. [5] also recommended a plain high-resolution CT coronal temporal bone study with MR cisternography to show the defect and the leak noninvasively, particularly in patients with bilateral CSF otorhinorrhea associated with unilateral hearing loss.

Radiology is the most important tool to diagnose CSF fistula. **All cases of recurrent meningitis, particularly with sensorineural hearing loss should have HRCT and MRI of the temporal bone to rule out a possible IEM**. Radiology may demonstrate:

1. Presence and type of IEM: In our department stapes footplate fistulas were observed in IP-I (Fig. 12.2a), CH-I (Fig. 12.2b, c), and CH-II (Fig. 12.2d).

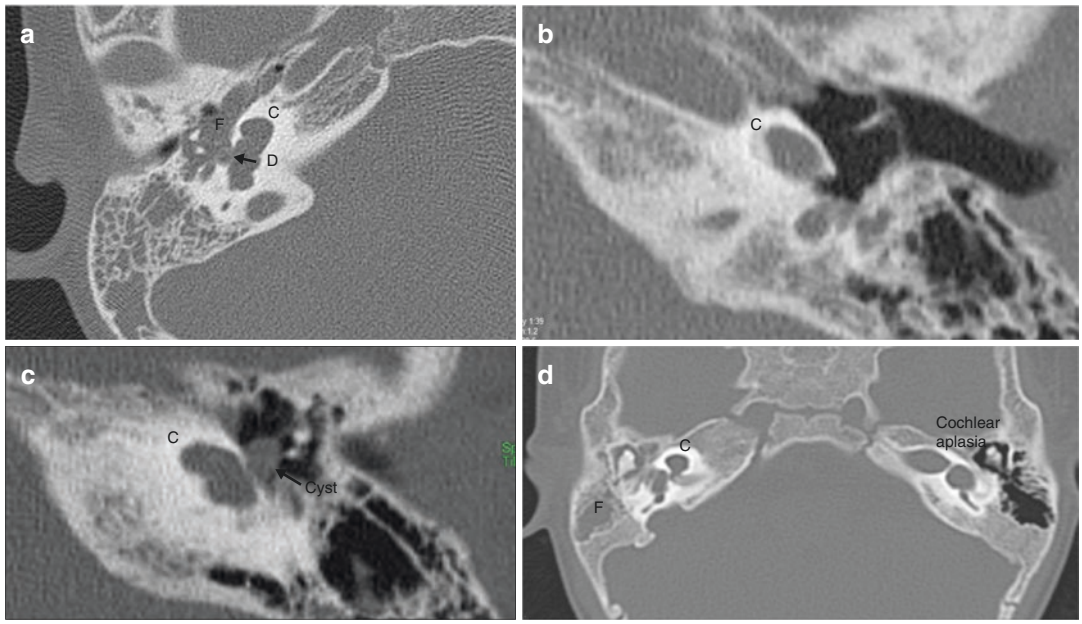

Fig. 12.2 Cases with stapes footplate fistula: (**a**) Incomplete partition type one cochlea (C), with defective footplate (D) and fluid (F) filling the middle ear, (**b** and **c**) cochlear hypoplasia type one (C) with cyst over the oval window, (**d**) cochlear hypoplasia type two (C) with fluid (F) filling middle ear and mastoid on the right side. Please note cochlear aplasia on the left side

2. Signs of oval window cyst: Opacity in the oval window area (Figs. 12.2c and 12.3).
3. Signs of CSF leak: Fluid filling middle ear and mastoid (Fig. 12.4a) and sometimes extending to nasopharynx via Eustachian tube (Fig. 12.4b). If this fluid has similar characteristic to the fluid filling the inner ear and IAC, it may be the sign of a CSF leak. This latter finding, if present, distinguishes OME from CSF leakage where both fluids demonstrate different characteristics on MRI.

To diagnose CSF leak in case of recurrent meningitis after CI surgery is more difficult. MRI cannot be performed. If there is no fluid in the middle ear and mastoid in the preoperative HRCT, and during operation, postoperative fluid signal on HRCT is an important sign indicating CSF leak (particularly in IP-I) (see **Case 3** below). One of our patients with oval window fistula presented like this. It may be necessary to have a CT cisternography in certain cases.

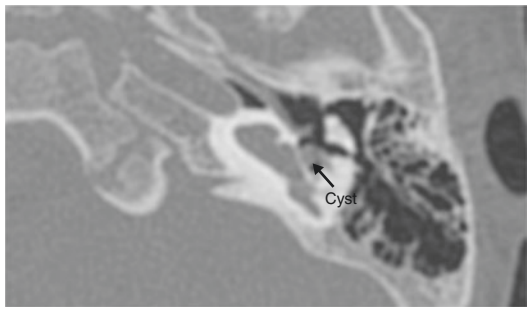

Fig. 12.3 Cystic opacity over the oval window area

12.5 Indications for Surgery

None of the patients who have been operated had a clear diagnosis of fistula before exploration. History of the patient is extremely important in the diagnosis and management of the patient with recurrent meningitis (see **Case 1** below). Therefore, suspicion of fistula in recurrent meningitis if there is one of the above pathologies is very important. If there is a suspicion of CSF leak it is advisable to go ahead with surgery rather than wait and see.

If one of the findings below is present there is possibility of a stapes footplate fistula necessitating middle ear exploration:

1. An opacity at the oval window area on HRCT in a patient with recurrent meningitis: oval window area has to be examined surgically.
2. Middle ear effusion in the above IEM types with recurrent meningitis.
3. Fluid in the middle ear which has similar characteristics on intensity with the inner ear fluids on MRI extending through Eustachian tube into nasopharynx.
4. Posttraumatic rhinorrhea or otorrhea in the above mentioned subtypes of IEM.

12.6 Treatment

SURGEON SHOULD ALWAYS KEEP IN MIND NOT TO LEAVE OPERATION THEATER WITHOUT FULLY CONTROLLING THE CSF LEAKAGE.

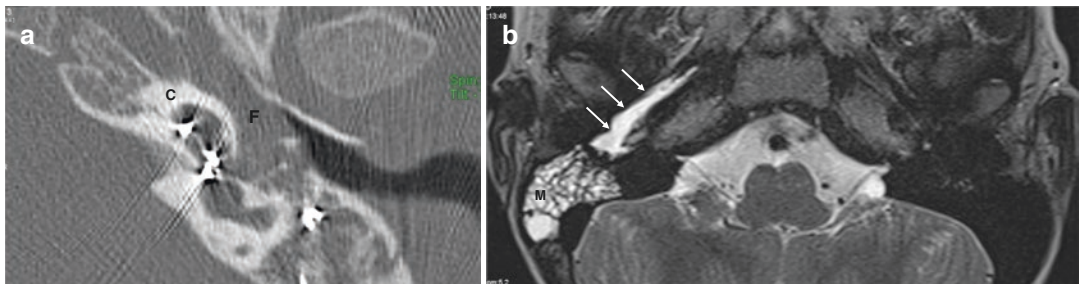

Fig. 12.4 (**a**) Incomplete partition type one cochlea (C) with implant and fluid (F) filling middle ear, (**b**) Fluid in the mastoid (M) and Eustachian tube (white arrows)

12.6.1 Vaccination

SNHL due to meningitis showed dramatic decrease after routine use of pneumococcal vaccination. However, IEMs still remain an important cause of recurrent meningitis. Although vaccination is more important in IEMs than normal population, appropriate management of the fistula site is priority. Vaccination only does not prevent meningitis in a case of IEM.

12.6.2 Middle Ear Exploration

Ear canal is narrower in children; exploration should be performed via endaural approach which allows enlargement of the ear canal. Atticotomy may be necessary to evaluate the ossicles. In general, more manipulation is necessary at the oval window than stapedotomy. Therefore, endaural approach provides larger exposure and is preferred over endomeatal approach in children. The latter approach has been used to manage footplate without damaging the electrode in an already implanted patient (see **Case 4** below).

There have been important changes in the management of these patients. In the past removal of the ossicles and obliteration of the vestibule was suggested in the management of these cases [1]. It has been observed that stapes is an important structure keeping the fascia in place against high pulsating CSF pressure. Now we believe that **every effort must be shown to keep the stapes in place**.

The fistula site is explored without removing any ossicles. Usually there is a cystic structure coming from the footplate (Fig. 12.5a). Without removing this cyst, it is impossible to evaluate the fistula. There is usually little serous fluid around. The fluid is removed with suction and facial nerve (FN) canal is evaluated. It is important to note any bony dehiscence at the tympanic segment at this stage because once the cyst is removed, there will be a gusher and during continuous CSF leakage it is not possible to evaluate the FN. During gusher it is possible to cause FN injury with surgical instruments if there is a bony dehiscence.

The cystic structure is then removed with cup forceps. As soon as it is removed there will be a CSF gusher from the footplate. It is important to avoid trauma to the facial nerve because clear fluid causes refraction and FN location may be misinterpreted. At this stage it is advisable to gently remove the fluid with suction and wait for 10–15 min until it decreases considerably and starts to pulsate.

At this stage the fistula site is inspected (Fig. 12.5b). Aim of surgery is to introduce a piece of fascia in a dumbbell fashion; one half of the fascia in the vestibule and the other half in the middle ear. If the defect is less than 0.7–0.8 mm, it is difficult to insert fascia through the opening and efficiently block the fistula. In that case it is advisable to use a 0.6 mm diamond burr to make a circular opening at the footplate (Fig. 12.5c). It is very important to keep the stapes intact at this stage. Then a fascia about 1 cm long and 2–3 mm wide is gently inserted into the defect (Fig. 12.5d). Ideally the fascia has to be placed in a dumbbell fashion in the defect (Fig. 12.5e).

There may be irregular defects or a defect at the anterior crus. It is advisable to enlarge the defect with a 0.6-mm diamond burr and apply the fascia as explained before.

The most difficult patients are the ones who will have a CI surgery at the same session where the stapes has a fistula. In two of them stapes dislocated during manipulation. Vestibule had to be obliterated. This carries the risk of filling up the space for electrode. It is advisable to insert the electrode and gently obliterate the vestibule (See **Case 5** below). However, the best treatment method is to keep the stapes in place and enlarge the fistula.

IF CI has to be applied as well, we start with transmastoid approach and open the facial recess. The facial recess is enlarged as much as possible because there will be prolonged surgical manipulation around the cochleostomy area. This approach is sufficient for electrode insertion. But it is insufficient for stapes footplate exploration. Therefore, additional transcanal exploration is necessary. The ear canal skin is elevated and then the footplate is managed as mentioned before.

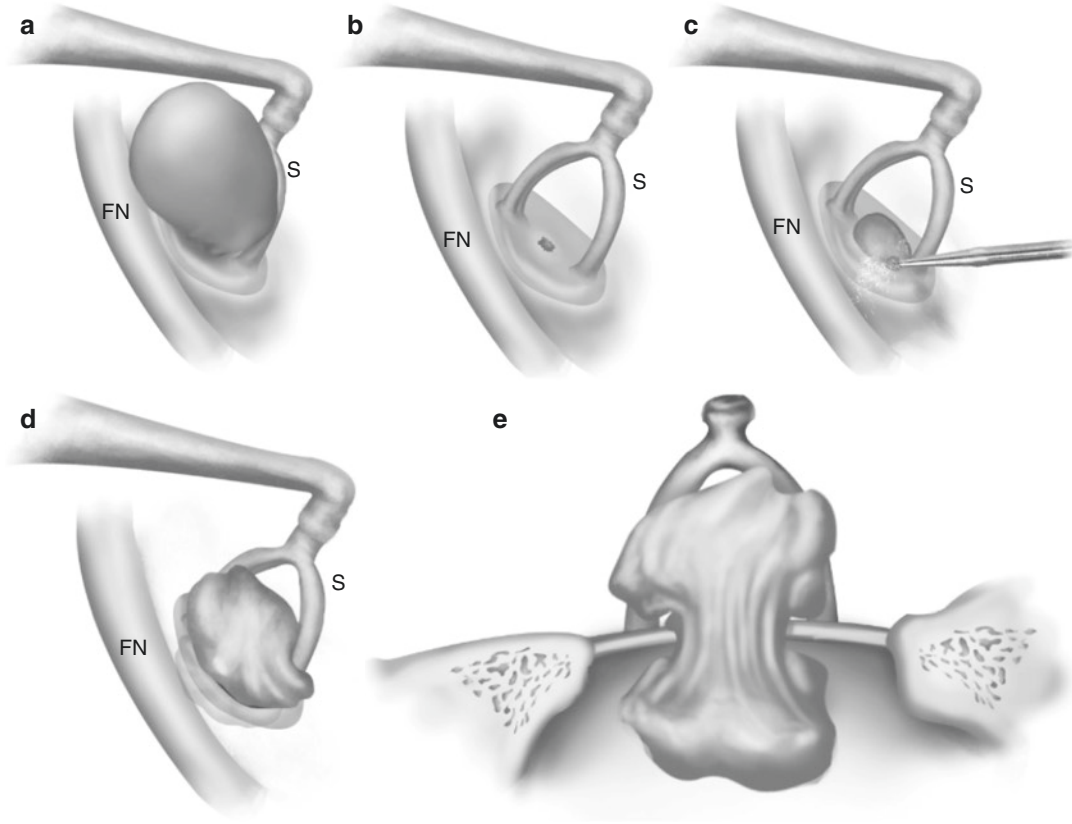

Fig. 12.5 Management of the stapes footplate fistula with a cyst: (**a**) A cystic structure coming from the footplate, (**b**) Footplate defect after cyst removal, (**c**) Enlargement of the fistula with diamond burr, (**d**) Placement of the fascia through the defect, while keeping the stapes in place to provide support to the fascia, (**e**) Fascia placed in dumbbell fashion through the defect

In situations where the stapes was removed during manipulation the vestibule is packed with fascia. It is advisable to block oval window by incus. Long process is removed by drilling and short process and body is inserted into the window with fascia between incus and the bony opening (Fig. 12.6a–d).

At times it is necessary to make prolonged manipulation at both windows. Keeping the fascia pieces in place against high pulsating CSF pressure is the most important part of the procedure. Therefore, postoperative CONTINUOUS LUMBAR DRAINAGE IS ALWAYS PART OF THE PROCEDURE.

In the literature, there are methods suggesting to remove the ossicles and to pack vestibule through the oval window with a layer of muscle or fascia followed by injection of fibrin glue [3, 4, 6, 10]. Another layer of muscle or fascia may be used on top. Syal et al. [10] placed intraoperative continuous lumbar drainage and lumbar drainage was continued postoperatively for 7 days in all four cases. Da Cruz [5] repaired the oval window fistula with vein and fat graft.

If a leakage is discovered at the cochleostomy site, this should be sealed as reported by Page and Eby [7]. Temporalis fascia was packed around the electrode array and into the vestibule until the leakage stopped. A piece of temporalis muscle was then placed in the middle ear space on top of the fascia. The wound was then closed, and a lumbar drain was inserted.

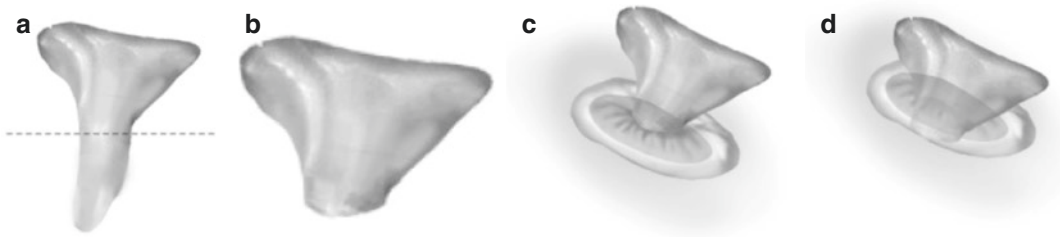

Fig. 12.6 (**a**–**d**) Stapes was removed during manipulation. Vestibule is obliterated with fascia and incus is placed into oval window defect after long process is removed

12.6.3 Subtotal Petrosectomy

Weber et al. [15] recommended subtotal petrosectomy with removal of the middle ear mucosa, closure of the Eustachian tube and ear canal in patients with recurrent meningitis.

Mylanus et al. [4] reported a patient with common cavity who had recurrent meningitis. Exploration of the middle ear revealed a fistula in the oval window which was sealed with temporal fascia. Upon recurrence of meningitis she had subtotal petrosectomy and a cochlear implantation through a labyrinthotomy, followed by a total obliteration with abdominal fat and closure of the external auditory canal. No CSF gusher was found and complete insertion was accomplished with an uncoiled, straight electrode array.

In their patient with multiple leakage sites Da Cruz et al. [5] used combined middle fossa and transmastoid approach. They repaired the oval window fistula with vein and fat graft. A soft tissue seal of temporalis muscle and fascia lata was inserted from above into the tegmen defect like a cork. Eustachian tube orifice was blocked and ear canal was closed as a blind sac.

We have performed this procedure in one patient who had recurrence of the stapes footplate fistula twice.

12.6.4 Continous Lumbar Drainage

This is a very important part of the management of the fistula. There is a strong pulsating CSF pressure acting on the reconstruction area. It tries to dislodge the fascia around the opening. In one case of footplate fistula, leakage was managed properly and no CLD was applied, there has been a recurrence of rhinorrhea 2 days after the surgery. Fistula site was re-explored and repaired once again and CLD was applied a week. No further leakage was noted.

Another patient with difficult surgery at the oval window and cochleostomy was managed properly in OR. At night CLD was removed accidentally and unfortunately rhinorrhea started the day after. He had 10 days of CLD and finally discharged without any leakage.

Our current management protocol is to employ CLD in all cases of gusher at the footplate or during cochleostomy even though the fistula is closed completely. As mentioned before, this diverts CSF to another area and lowers the CSF pressure acting on the fascia decreasing the chance of dislodging the fascia.

12.7 Clinical Experience

Eleven patients, who had spontaneous footplate fistula without any cochlear implant surgery, had the following subtypes of IEMs:

IP-I eight cases (Fig. 12.2a),
CH-I one case (Fig. 12.2b, c),
CH-II two cases (Fig. 12.2d).

Two of these patients had CI surgery in their contralateral ears and CI was not related to the fistula at all. Two of these cases received CI in the same setting during fistula closure (see **Case 5** below).

CSF leakage was seen after CI surgery in six cases: Two had been operated in our department: one had a leakage around electrode, other had stapes footplate fistula without any leakage around electrode (see **Case 3** below). Remaining four patients had been operated in other centers: two had CSF leakage around electrode, two had oval window fistula.

12.8 Site of Fistula

All 11 cases with spontaneous CSF fistula had their defect at the oval window. None of them had a spontaneous fistula at the round window. Literature findings support these findings. One case with accidental removal of CLD led to rhinorrhea. One case without CLD needed revision. One case with two recurrences had subtotal petrosectomy.

Six cases of recurrent meningitis were after CI surgery: three of these had fistula at the oval window and the remaining three had CSF leakage at the cochleostomy site. In cases with oval window fistula the footplate defect was repaired with fascia. In cases where the leak was from the cochleostomy site, area was closed with fascia. We had to change the modiolar hugging electrode in one patient with the FORM electrode.

In our department we had a patient who presented with recurrent meningitis due to spontaneous fistula on the contralateral side to CI surgery. She had bilateral IP-I defect and had been implanted 2 years prior to meningitis on the right side. She had two attacks of meningitis. Her CT demonstrated fluid on the left, contralateral middle ear, and mastoid. Exploration of the nonimplanted left ear revealed mucosal cyst coming from the stapes footplate. Perforation of the mucosa produced a CSF gusher which was controlled by packing fascia into the vestibule. She had two recurrences of CSF leak and the surgeon performed a subtotal petrosectomy in the end. This case demonstrates us that CI in IEMs may not always be the source of recurrent meningitis. This presents a diagnostic dilemma to the implant surgeon.

12.9 Cases

Case 1: EP 2y Old Male Patient, Operated July 2013

His sister was operated for ABI because of severe IEM. Her mother said she had a brother who had two attacks of meningitis. I asked her about his hearing and she said he was deaf on one side only. I told her that I want to examine him immediately. Next day he was brought from Istanbul to Ankara. His HRCT revealed an IP-I with fluid filling middle ear and mastoid on the right side. Left side had normal cochlea and well aerated middle ear and mastoid (Fig. 12.7). Next day he was operated via endaural approach and the footplate cyst was removed and defect was sealed with fascia. He had no further attack of rhinorrhea or meningitis (Video 12.1).

Case 2: NCB 3-Year-Old Female Patient Operated on 8.10.2013

She applied with recurrent meningitis. She had bilateral CH-II with fluid filling left middle ear and mastoid (Fig. 12.8). Left side was explored via endaural approach. There was a fistula at the stapes footplate with continuous CSF leakage. There was no cystic structure at the footplate.

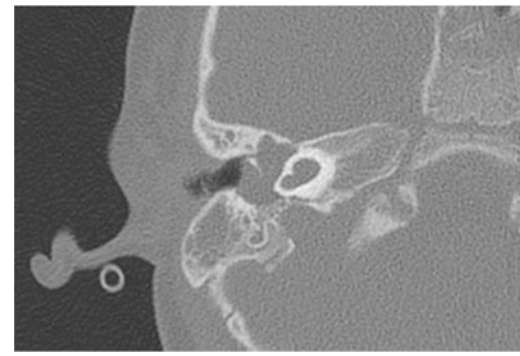

Fig. 12.7 Case 1. HRCT showing an IP-I with fluid filling middle ear and mastoid on the right side

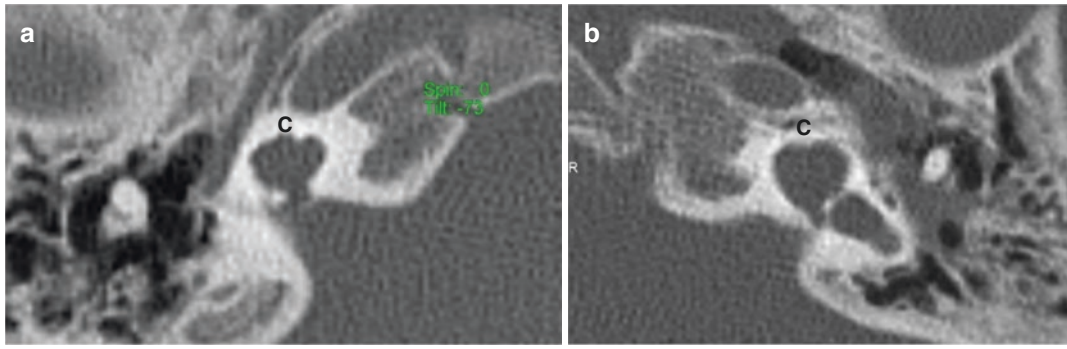

Fig. 12.8 Case 2. Bilateral CH-II with normal middle ear and mastoid on the right (**a**) and fluid filling middle ear and mastoid on the left side (**b**)

The ossicles were kept in place and the millimetric defect at the footplate was enlarged with a 1-mm diamond bur so that a piece of fascia can be passed through to stop the leakage. After the leakage was controlled tissue glue was used and a second layer of fascia was applied (Video 12.2).

Case 3: CK 6-Year Old Female Patient, Operated on 5.9.2005

In 2008 we were faced with recurrent meningitis in a child with bilateral IP-I who had received CI 4 years prior to meningitis attacks. She had bilateral IP-I deformity (Fig. 12.9a, b) with no fluid in the middle ear and mastoid. Right side was implanted in 2005. During cochleostomy there was gusher which was controlled completely and the electrode was fully inserted. Four years later she had meningitis. HRCT demonstrated normal electrode placement without any fluid on the right side (Fig. 12.9c). On the left side middle ear and mastoid were filled with fluid (Fig. 12.9d). Left side with fluid in the middle ear was explored via endaural approach. There was a cyst coming from the stapes fistula. The cyst was removed and this revealed a CSF leakage. Incus and stapes were kept in place and the defect was repaired with fascia. She had one more attack of meningitis and her surgeon performed subtotal petrosectomy on the left side.

Her HRCT enabled us to choose the correct side with CSF fistula. Her initial preoperative HRCT 4 years prior to meningitis attacks had normal ventilation in the middle ear. After meningitis CT demonstrated fluid in the middle ear and mastoid on the contralateral side to CI. Retrospective examination of the original CT demonstrates a soft tissue mass (cyst) on the left side (Fig. 12.9b).

Case 4: MP 1-Year-Old Female Patient, Operated on 14.6.2010

She had bilateral IP-I with defective fundus between cochlea and IAC (Fig. 12.10a). Right side was implanted in 2010. During surgery there was severe gusher. She was implanted with the initial prototype of FORM electrode. At the end of the surgery, there was no CSF leakage around the electrode. Six months later she applied with meningitis. HRCT demonstrated fluid filling middle ear and mastoid on the right side with cochlear implantation (Fig. 12.10b). Middle ear exploration was planned on the right side. Endomeatal approach was used. Electrode was functioning and the risk for damage to the electrode is avoided by using endomeatal approach. Middle ear was filled with CSF coming from a defect in the stapes footplate. No leakage was present at the cochleostomy area. Oval window fistula was repaired with a piece of fascia keeping the stapes in place (Video 12.3). She has not experienced any leakage or meningitis afterwards.

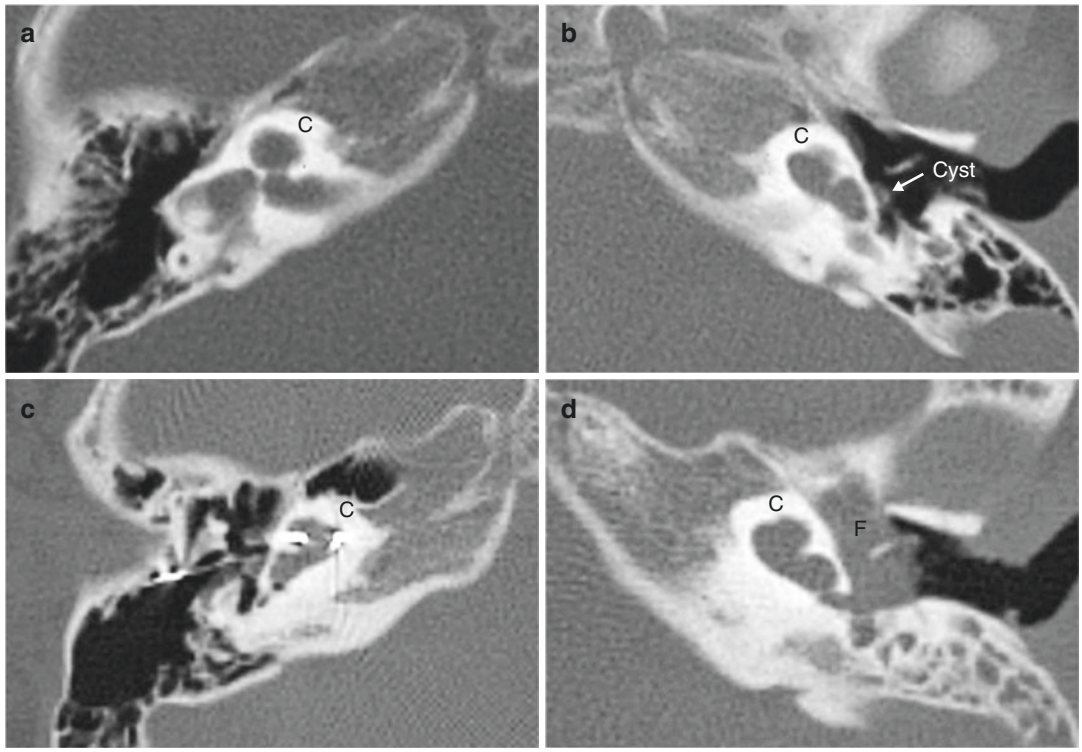

Fig. 12.9 Case 3 bilateral IP-I deformity (**a** = Right and **b** = Left) with no fluid in the middle ear and mastoid. Please note the soft tissue mass (cyst) on the left side which was noticed postoperatively. (**c**) HRCT showing normal electrode placement without any fluid on the right side, (**d**) Contralateral ear filled with fluid (*C* cochlea, *F* fluid)

Case 5: MD 1.5-Year-Old Male Patient Operated on 31.12.2012

He had posttraumatic rhinorrhea for 8 months prior to his visit. He had two times neurosurgical exploration of the anterior cranial fossa without any leakage point in another center. Main reason for application was deafness since birth. Unfortunately, his ears had not been evaluated before. Child was deaf on both sides since birth. HRCT demonstrated left side cochlear aplasia and right side CH-II with hypoplastic cochlear nerve. On the right side middle ear was filled with fluid.

CSF fistula reparation and cochlear implantation were planned at the same time. Postauricular approach was used. During mastoidectomy there was continuous CSF leakage coming from the antrum. Facial recess was opened. Leakage was coming from the stapes footplate. Initially footplate defect was explored via facial recess but it was impossible to evaluate and manage the fistula properly. Transmastoid approach was combined with transcanal approach where the footplate was controlled much better. Stapes had to be removed. Electrode was inserted via facial recess through cochleostomy. Defect was then closed with fascia and incus (Video 12.4).

He then received an ABI on the contralateral side with cochlear aplasia (Fig. 12.11).

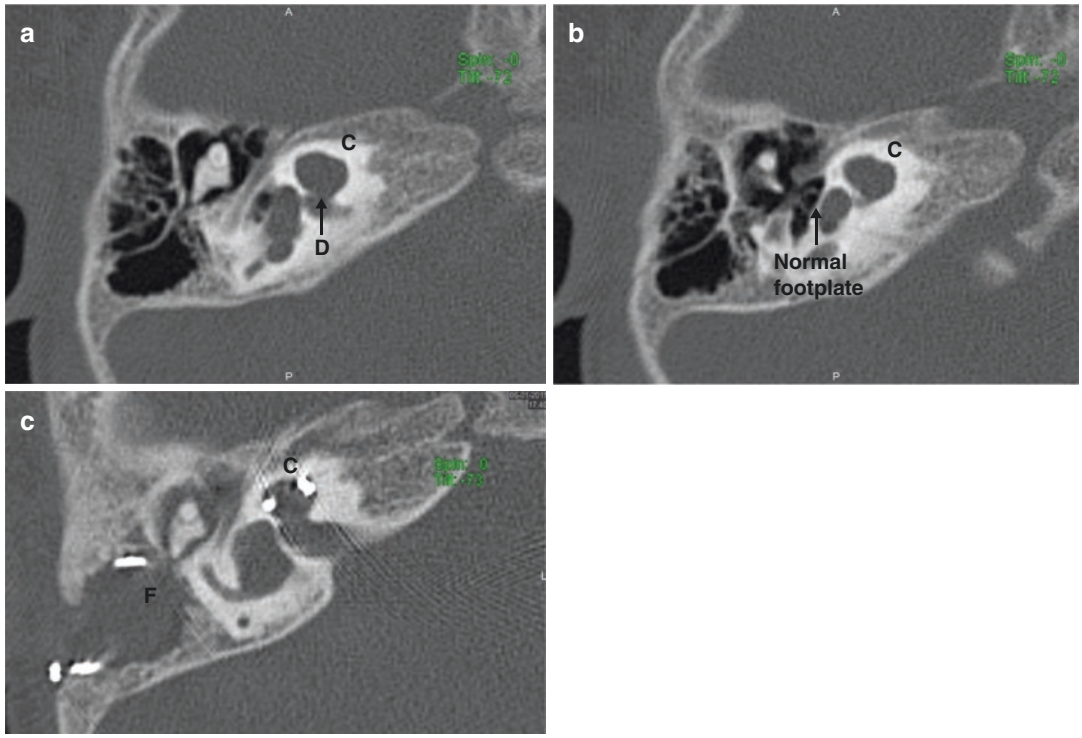

Fig. 12.10 Bilateral IP-I with defective fundus between cochlea and IAC. (**a**) Preoperative HRCT showing normal IP-I defect without footplate fistula or cyst, and normal ventilation, (**b**) Normal footplate, (**c**) HRCT showing normal electrode placement with fluid filling middle ear and mastoid (*C* cochlea, *F* Fluid)

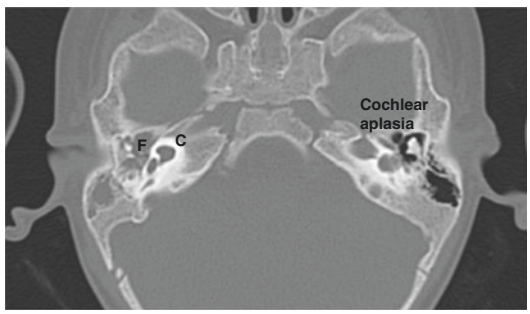

Fig. 12.11 HRCT showing left side cochlear aplasia and right side CH-II with hypoplastic cochlear nerve. On the right side, the middle ear was filled with fluid

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