Stapedotomy

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14.1 Introduction

Congenital ossicular chain abnormalities behind normal ear canal and tympanic membrane are much rarer when compared to conductive hearing loss (CHL) caused by acquired causes. They usually fail neonatal hearing screening tests. It can be diagnosed early if air and bone conduction ABR is done. They usually present as non-progressive hearing loss since birth. If there is additional sensorineural hearing loss component, they usually present with poor language development.

In congenital CHL stapes may be fixed alone or three ossicles may be involved in different degrees. They may all be fixed. Stapes fixation is the most important of all, because stapedotomy may improve hearing loss significantly. However, stapedotomy in pediatric population is controversial. In this group of patients there may be higher risk of sensorineural hearing loss (SNHL), facial nerve (FN) injury, and meningitis when compared to normal population. These are the main reasons for stapedotomy to be controversial in pediatric population.

In 2014, first author investigated the specimens with inner ear malformations in Massachusetts Eye and Ear Infirmary (MEEI) [1]. Interestingly 14 of 41 cases had oval window atresia or stapes footplate fixation. This brings the concept of performing stapedotomy in cases with IEMs. Here we present 11 cases of IEMs who underwent stapedotomy between 2008 and 2014.

14.2 Histopathology

Histologically stapes fixation and oval window anomalies can be encountered in IEMs. Sennaroglu L investigated 41 temporal bone specimens with IEMs in the MEEI [1]. Fourteen of the 41 cases with IEMs presented either with fixed stapes footplate (Fig. 14.1a) or oval window atresia (Fig. 14.1b). Twelve of these cases presented with cochlear hypoplasia (CH). As the oval window is part of the cochlea, fixation of the footplate or the oval window atresia can be expected in a hypoplastic cochlea. It was interesting to note that cochlear abnormalities, particularly cochlear hypoplasia, may cause CHL or mixed hearing loss by stapes fixation.

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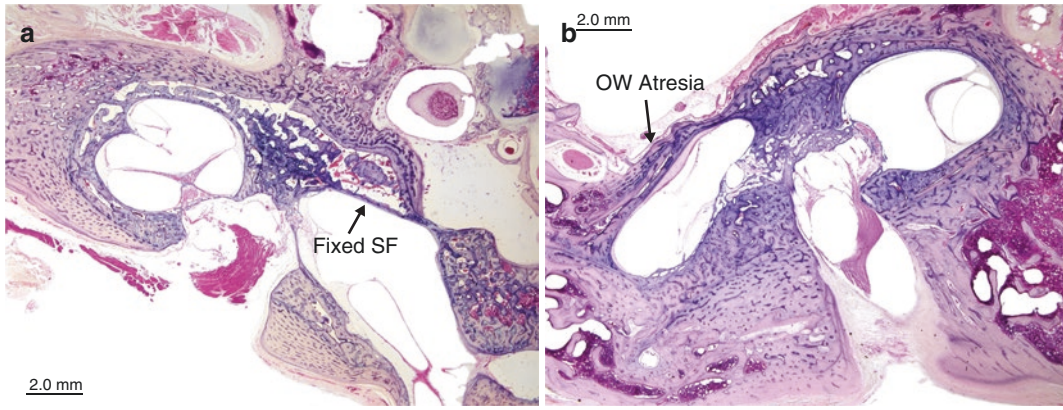

Fig. 14.1 Histopathological findings of oval window in cochlear hypoplasia: (**a**) stapes footplate (SF) fixation, (**b**) oval window (OW) atresia. (With permission of Department of Otolaryngology of Massachusetts Eye and Ear Infirmary)

14.3 Literature Review

In his report on histopathology of IEMs, Sennaroglu L. mentioned that majority of stapes footplate fixation or oval window atresia are found in cochlear hypoplasia (CH) [1]. Out of three CH-I cochleae in MEEI, two had a fixed stapes footplate and one had an atretic oval window. In specimens with CH-II, oval window was atretic in one specimen, and the stapes footplate was fixed in three. There were five specimens with CH-III: oval window was normal in one specimen, fixed in one, and atretic in three. There was no specimen with CH-IV. These cases show that it is common to find stapes footplate fixation or oval window atresia in cochlear hypoplasia. It also implies that if there is conductive or mixed type hearing loss in CH stapedotomy may result in better hearing. The patient may have to use hearing aid after surgery depending on the bone conduction level and air-bone gap. If there is pure conductive loss, stapes surgery may result in near normal hearing.

According to Sennaroglu [1], in CH-III, the developmental arrest in the membranous labyrinth most probably occurs between 6 and 8 weeks, resulting in a cochlea whose dimensions are smaller than normal, with normal internal architecture. In CH-IV there is a normal basal turn but small middle and apical turns. Arrest in the membranous labyrinth must be between 10th and 20th weeks, after the basal turn reaches full size but before the middle and apical turns enlarge to their normal size.

Congenital stapes fixation can be explained by embryology: the stapes footplate is part of the otic capsule, and according to Donaldson et al. [2], the base of the stapes is originally continuous with the otic capsule. Then it is segregated through a retrogressive process in the cartilage. The reorganized tissue becomes the annular ligament. A transcapsular channel (fissula ante fenestram) is formed as a result of invasion of the primitive cartilage by periotic tissue. If there is an arrest of the otic capsule development before the formation of the footplate, it is natural that the stapes becomes fixed to the oval window; however, it is still difficult to explain stapes fixation in CH-IV with normal basal turn.

CH-I and CH-II cases also have stapes fixation in some cases. But due to the severe malformation and profound SNHL, they are candidates for implantation. CH-II cases are accompanied by a defective modiolus. Because of the resolution of present day HRCT, the partial modiolar defect may not be diagnosed, but histopathological examination shows the defective modiolus in all cases. Because of the shorter cochlea they have SNHL and the fixed footplate provides the conductive component. The author has performed stapedotomy in cases of CH with mixed hearing loss. Postoperatively, these cases benefit more from HA. Patients with CH who have profound sensorineural hearing loss (SNHL) are candidates for CI. CH with cochlear aperture aplasia necessitates an ABI.

14.4 Clinical Findings

Between 2008 and 2014, nine cases of cochlear hypoplasia underwent stapedotomy (two cases of vestibular dilatation are not included). They all applied with the complaint of hearing loss, which was non-progressive and present since birth. They present with a normal ear canal and tympanic membrane. Otitis media with effusion may be a coincidental finding but conductive hearing loss persists after the effusion is treated. In addition to stapes, other ossicles may also be fixed, the facial nerve may be misplaced at the oval window area, or the oval window may be atretic.

14.5 Radiological Findings

Radiology is very important in congenital conductive or mixed hearing loss. Radiology may demonstrate ossicular fixation to the attic wall.

Radiology also shows the type of IEMs. There are four groups of cochlear hypoplasia where external dimensions are smaller than normal cochlea with various internal architecture deformities (Chaps. 1 and 26 for more details):

1. CH-I: Cochlea with absent internal architecture (modiolus and ISS) with/without a thin bony partition between the cochlea and the IAC.
2. CH-II: External shape resembled a cochlea, but it was smaller and rounder than normal. Modiolus is defective resulting in a cystic cochlea.
3. CH-III: Small cochlea with normal internal architecture. The only difference from a normal cochlea was that the CH-III cochlea consisted of approximately one and a half turns.
4. CH-IV: Cochlea with normal basal turn, hypoplastic middle, and apical turn.

CH-I is like a bud without any internal architecture and outcome with stapes surgery is not expected to be good. In CH-II there may be a risk of gusher. CH-III (Fig. 14.2a, b) and CH-IV (Fig. 14.2c, d) are the best candidates for stapedotomy.

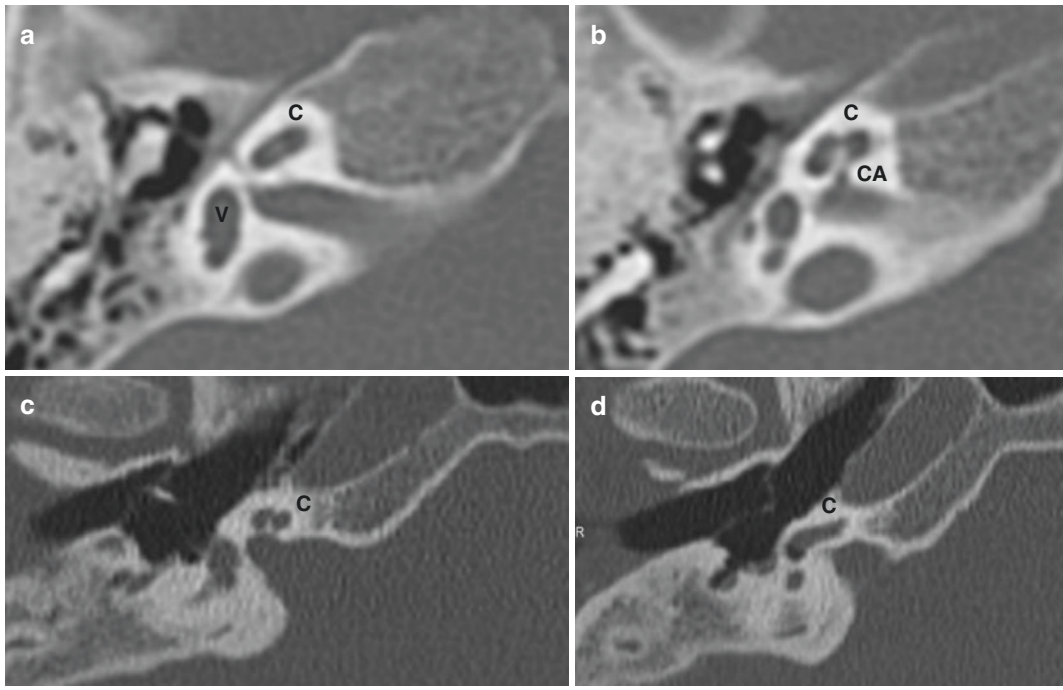

Fig. 14.2 (**a**, **b**) Cochlear hypoplasia type III. Cochlea (C) consists of hypoplastic basal and middle turns with hypoplastic vestibule (V). Note stenotic cochlear aperture (CA). (**c**, **d**) Cochlear hypoplasia type IV. Cochlea (C) consists of normal basal turn and hypoplastic middle and apical turns

Temporal CT shows the position of the facial nerve. Coronal sections in particular show the position of the facial nerve in relation to oval window. In the present series there are cases with facial nerve in more lateral position (Fig. 14.3a), at the OW (Fig. 14.3b) and inferior to the OW (Fig. 14.3c). The surgery becomes very challenging if the coronal section demonstrates the facial nerve at or inferior to the oval window on coronal section.

Another use of HRCT is to show the defect between the cochlea and internal auditory canal (IAC) which may cause CSF leakage. Case 14.1 who had CSF leakage did not have a defect between IAC and cochlea but presented with demineralization all around cochlea, which may cause CSF from subarachnoid space to reach cochlea and hence result in CSF leakage at the time of fenestration into the vestibule.

14.6 Audiological Findings

Audiological findings of these patients are given in Table 14.1. It is possible to diagnose pure conductive hearing loss (see Case 14.1) and mixed type hearing loss (see Case 14.2). Out of these nine ears, seven were pure conductive and two were mixed. Although both benefit from stapedotomy, there is a possibility for near normal hearing in cases of conductive hearing loss (see Case 14.1). In case of mixed hearing loss, aim of the surgery is to make the patient benefit more from hearing aids in the postoperative period (see Case 14.2).

All patients had cochlear hypoplasia (CH). More common presentation for CH is SNHL. Depending on thresholds, hearing aids or CI may be the method for habilitation. If there is no cochlear nerve ABI may be indicated as well.

14.7 Management

There are different treatment options in congenital ossicular fixation. Providing hearing aids is the earliest and most appropriate option, particularly in bilateral cases; however, in cases that present with severe ossicular pathology such as oval window atresia or ossicular discontinuity between the tympanic membrane and the oval

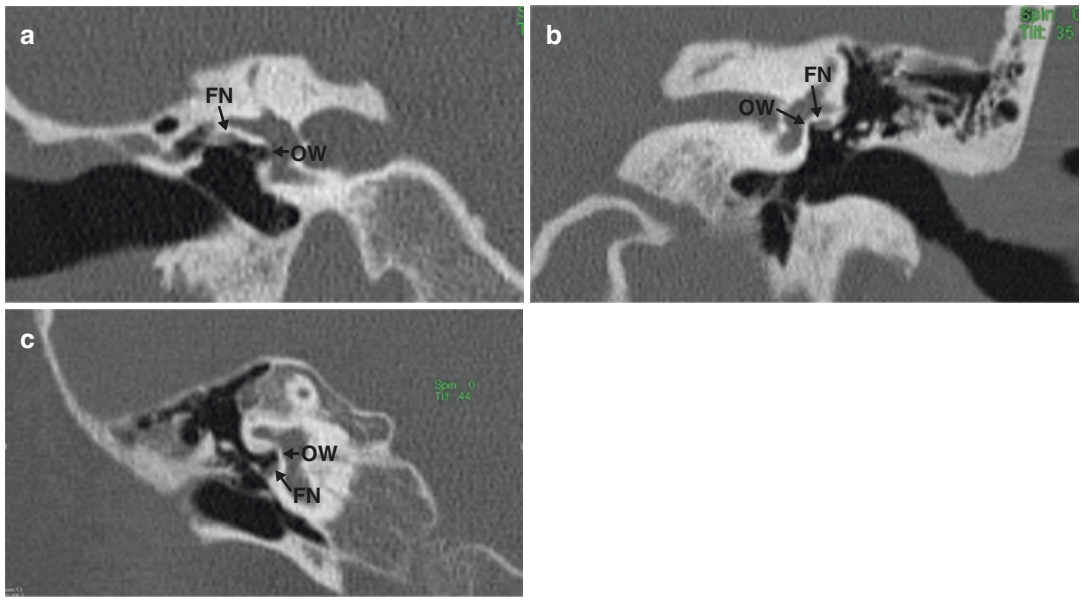

Fig. 14.3 Abnormalities of tympanic segment of facial nerve (FN) (coronal section) (**a**) FN in a more lateral position in relation to oval window (OW) and stapes and inferior to lateral semicircular canal (cochlear hypoplasia type IV), (**b**) FN at the OW, (**c**) FN inferior to oval window

window, hearing aids may not be sufficient or well tolerated because of the large air-bone gap (ABG). Stapedotomy is another option that is not universally accepted for children, because the procedure involves exposing the child's inner ear. Yet another option is bone anchored hearing devices, but for children, the surgical placement of a bone anchored device can be too early. Bone conduction devices with head band may be appropriate if the patient does not benefit from a hearing aid. As this is a treatment option for ear canal atresia cases, where the surgical reconstruction of the ear canal is usually not satisfactory, the families usually look for other treatment possibilities in ossicular pathologies. In such a situation, stapedotomy is one of the treatment options. In addition, other ossicles may become fixed, necessitating surgical options such as manubriostapedioplasty [3] or atticotomy.

14.8 Surgery

There are certain difficulties during the surgery of CHL in IEMs. These can be grouped under five headings:

1. **Facial Nerve Anomaly:** Facial nerve (FN) may have an abnormal location in congenital cases. In CH-IV the labyrinthine segment of the facial nerve (FN) is anterior-superior to the cochlea (patient #6 in Table 14.1). This is almost a pathognomonic finding. Therefore, tympanic segment may not be in the usual location. Two cases with CH-IV had stapedotomy and their tympanic segment was located more superior to its normal location relative to oval window (Fig. 14.3a). This did not cause any difficulty during stapedotomy. Her mother with identical abnormality underwent cochlear implantation where she needed subtotal petrosectomy to visualize round window because of mastoid segment anterior dislocation (see Case 26.3 Chap. 26). Most difficult situation is when tympanic segment of FN is located at the oval window (patient #1). Preoperatively, this situation can be seen on coronal sections of temporal bone CT passing through the oval window. This needs special technique for drilling into vestibule while the burr is very close to the dehiscent FN and incus has to be extended for piston placement [4]. FN can also be seen inferior to the OW

Table 14.1 Audiological findings of the operated patients

Patient	Age	Sex	Side	IEM	Op date	Operation findings	Preoperative			Postoperative		
							Bone	Air	ABG	Bone	Air	ABG
1-TU	16	M	Left	CH-III	2008	FN at OW, drill to make vestibulotomy, bone cement, stapedotomy	22	72	50	12	28	16
2-SA	11	F	Left	CH-IV	2012	Stapedotomy, oozing	10	54	44	10	45	35
3-HG	8	F	Left	CH-III	2012	OW atresia, drill to make vestibulotomy & stapedotomy	13	47	34	13	25	12
3-HG	10	F	Right	CH-III	2013	OW atresia, drill used to make vestibulotomy & stapedotomy	10	48	38	13	32	19
4-IH	4	M	Left	CH-III	2013	FN inferior to OW, OW atresia, drill to make vestibulotomy, gusher	10	59	49	13	28	15
5-HS	11	F	Left	CH-III	2013	All ossicles fixed, atticotomy, stapedotomy	37.5	85	47.5	30	61.25	31.25
6-DK	22	F	Right	CH-IV	2013	Stapedotomy	10	66	55	11	25	14
4-IH	5	M	Right	CH-III	2014	FN inferior to OW, OW atresia, drill used to make vestibulotomy, gusher	10	55	45	12	22	10
5-HS	13	F	Right	CH-III	2014	All ossicles fixed, atticotomy, stapedotomy	37.5	89	51.50	33.75	71.25	37.5

(patient #4 in Table 14.1). Surgically, these are the most challenging cases, requiring facial nerve monitoring and a stimulator.

2. **Oval Window Atresia:** Some cases do not have an oval window formation. Those cases can be classified as atretic and require drilling for making fenestra into vestibule. Using a laser is not advisable because the bone is thick, and a laser may cause excessive heating, resulting in thermal damage to FN. Correct location can be estimated by taking into account the position of the incus and if present, the remnants of stapes suprastructure. During drilling, it is necessary to avoid a tunnel, but carefully lower the thickness of the bone to open the vestibule at the final moment by taking into consideration the direction of the prosthesis from the incus to the fenestra. If this is not planned correctly, drilling after opening the fenestra may cause severe SNHL.
3. **Incus Abnormality**: In cases of otosclerosis incus is positioned more horizontally during surgical exploration. In cases of oval window atresia, incus is more obliquely positioned possibly because of the absence of the stapes, making stapes piston placement more difficult. There is a possibility of the piston sliding off the incus which is positioned more oblique than normal. It is advisable to use a few drops of cement to stabilize the piston and avoid its slipping off incus towards vestibule.
4. **Involvement of Other Ossicles**: This finding is likely to be unrelated to the IEMs but makes the surgery more challenging in this situation. An atticotomy is necessary removing all the bone immobilizing the ossicles. This was present in patient #5 in Table 14.1. It is advisable to use endaural approach in all these cases so that more manipulation can be done around malleus and incus if necessary.
5. **CSF Leakage:** This is the most serious complication of the surgery. If the leak is not controlled properly, it may lead to meningitis. It is mandatory to have the children vaccinated before the procedure. Nowadays, pneumococcal and haemophilus vaccination is routinely done in many countries.

CSF leakage can be expected in these cases. If a piston tightly fitting into the fenestra is used, inserting the fascia around the piston into the vestibule becomes difficult. Based on our experience, using a 0.6 mm drill and a 0.4 mm stapes piston is suggested so that fascia can be inserted sufficiently into the vestibule. Passing the piston shaft through the fascia and inserting them together allows the fascia to surround the piston all around (see Case 14.1).

Contraindications to surgery in congenital hearing loss: In patients with CH with CHL, stapedotomy can be suggested to the families, but it is difficult to think stapes footplate fixation in congenital mixed HL. The operation should never be done if HRCT demonstrates IP-II or IP-III (Fig. 14.4a–c).

14.9 Complications

This procedure carries the risk of injury to the facial nerve, sensorineural hearing loss, and meningitis. Patients without vaccination against pneumococcus and haemophilus influenza should not be operated.

14.10 Clinical Experience

Between January 2003 and September 2015, the first author performed stapedotomy in 355 cases that presented with normal tympanic membrane and ossicular pathologies. Fifty-one of these had congenital ossicular anomalies with a normal ear canal and tympanic membrane (ear canal atresia is not included). If the revision cases are excluded, 42 primary cases out of 355 presented with congenital fixation. Eleven of these cases had various inner ear malformations. Nine had cochlear pathologies and two had only vestibular anomalies.

Nine cases presented with cochlear hypoplasia (CH) (Table 14.1). As stapes is part of the cochlea, the surgical findings are investigated in particular from this perspective. They have been classified according to the recent classification system [5, 6].

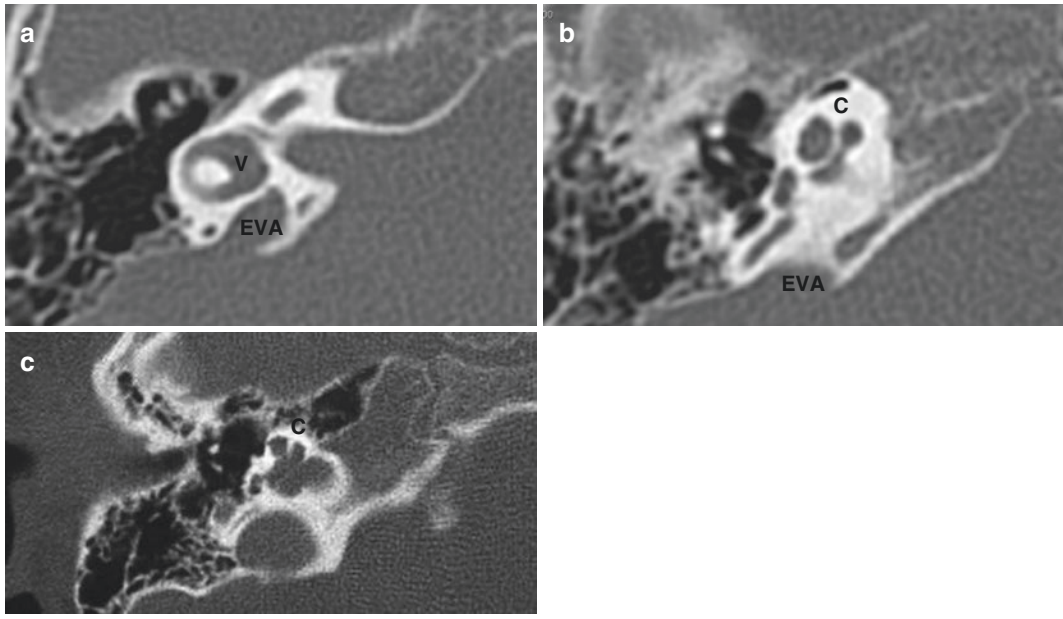

Fig. 14.4 Pathologies with mixed hearing loss where stapedotomy is contraindicated: (**a**, **b**) IP-II cochlea (C) with cystic apex, enlarged vestibular aqueduct (EVA), and minimally dilated vestibule (v). (**c**) IP-III cochlea (C) with interscalar septa and absent modiolus

The age of the patients was between 4 and 22 (Table 14.1). There were two males (three ears) and four females (six ears). There were seven cases of CH-III and two cases of CH-IV. Three cases had bilateral stapedotomy. Remaining three cases had unilateral surgery.

14.11 Cases

Case 14.1: IH (Patient #4) 4-Year-Old Male Patient

He applied with bilateral hearing loss since birth. He failed hearing screening on both sides. On HRCT of the temporal bone he had bilateral CH-III (Fig. 14.5a, b). Coronal sections showed that FN was located inferior to oval window on both sides (Fig. 14.5c, d). His preoperative audiological evaluation revealed bilateral moderately severe conductive hearing loss (Fig. 14.5e). His operation was the most difficult of the nine cases. Left side was operated on 20 September 2013 and right side on 25 November 2014. On both sides, the oval window was completely atretic without any footplate or annular ligament formation; however, round window was present bilaterally. Incus and malleus were present (Video 14.1). By taking into account the position of the incus, a vestibulotomy was created using 0.6 mm diamond burr. Cerebrospinal fluid (CSF) leakage occurred on both sides. A 0.4 mm stapes piston was passed through a piece of fascia (2 × 2 mm) and then placed between the incus and the fenestra. The fascia was inserted all around the piston into the vestibule in a dumb-bell fashion. CSF leak stopped completely. Incus was inclined medially on both sides, which made piston insertion difficult. A few drops of cement were used to fix the piston onto angled incus. The patient had already been immunized with Pneumococcal and Haemophilus influenza vaccination. Ear examination at 3 months' intervals revealed no fluid in the middle ear that would suggest CSF leakage.

After operation, his air conduction hearing thresholds showed improvement and ABG decreased to 20 dB from 50 dB. He had mild con-

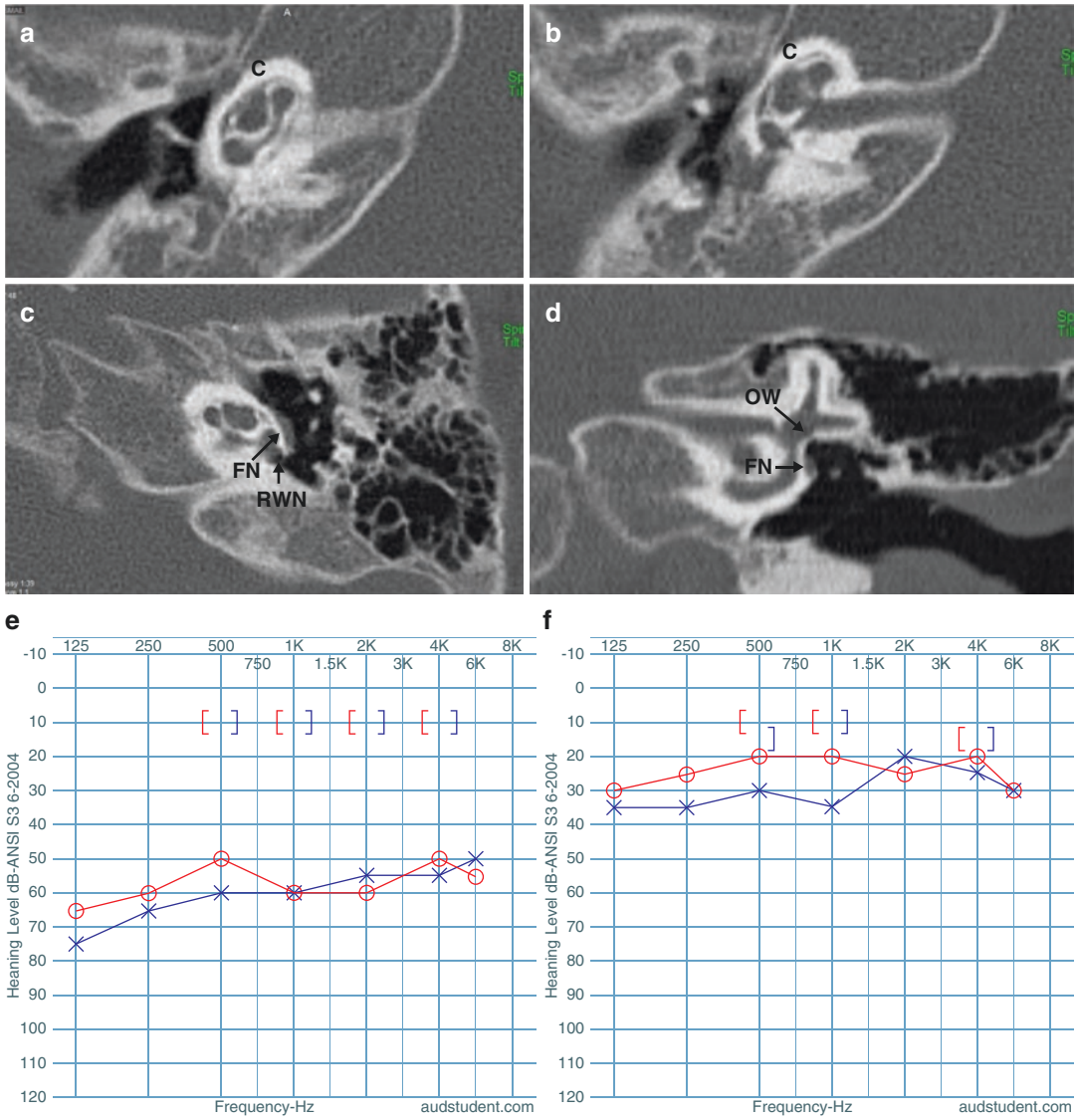

Fig. 14.5 Case 14.1. (**a**, **b**) Axial sections showing cochlear hypoplasia type III (C); note demineralization around cochlea. Modiolus appears to be normal. (**c**) Axial section showing tympanic segment of the facial nerve (FN) over the round window niche (RWN) on the promontory (left ear). (**d**) Coronal section showing FN below the oval window (OW). (**e**, **f**) Preoperative (**e**) and postoperative (**f**) hearing thresholds

ductive hearing loss and was able to hear without hearing aids. He developed excellent speech without hearing aid.

CSF leakage most probably came from bone demineralization around cochlea. Coronal section is very important to visualize FN in relation to oval window. This patient shows that it is possible to obtain hearing without hearing aids in pure conductive hearing loss.

Case 14.2: HS (Patient #5) 11-Year-Old Female Patient

She applied with bilateral severe mixed type hearing loss since birth. She had congenital cleft palate and developmental delay. She had been using hearing aids with very poor language development. CT revealed bilateral CH-III with extremely small middle and apical turns (Fig. 14.6a, b). Cochlear aperture was stenotic.

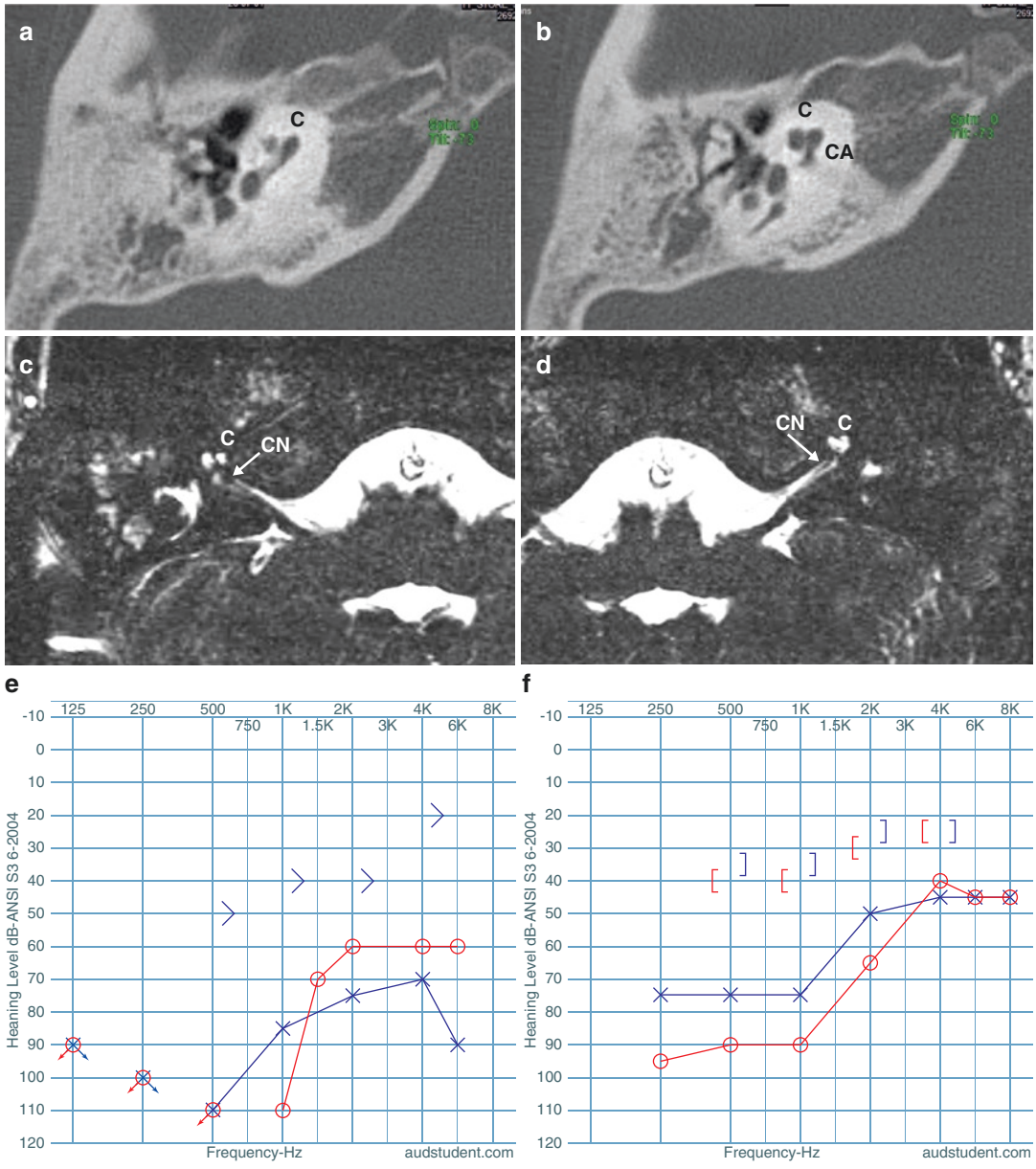

Fig. 14.6 Case 14.2. (**a**, **b**) Cochlear hypoplasia type III (C) consisting of hypoplastic basal turn and extremely small apical turn. Please note cochlear aperture stenosis. Axial sections showed cochlear nerve (CN) on right (**c**) and left (**d**) sides. (**e**, **f**) Preoperative (**e**) and postoperative (**f**) hearing thresholds

Cochlear nerve was present bilaterally (Fig. 14.6c, d). Due to developmental delay, testing was very difficult. Therefore, ears were tested at different sessions through insert earphones. First preoperative audiogram was done when she was 4 years old. She had rising audiogram and average air conduction thresholds (500–1000–2000–4000 Hz) were 85 dB for left and 89 dB for right ear. Average bone conduction threshold was 37.5 dB (masked bone conduction thresholds did not get). ABG was 47.5 dB for left and 51.50 for right ear (Fig. 14.6e).

On both sides she had all ossicles fixed. Left side was operated on 29 January 2011 and right

side was operated on 29 April 2014. In both operations, atticotomy and mobilization of the malleus and incus were performed before stapedotomy. A 0.6 mm stapes piston with 4.5 mm length was used on both sides. On the left side, the facial nerve was covering almost 75% of the footplate.

In her latest postoperative audiogram, air conduction hearing thresholds showed improvement. She had moderately severe mixed HL on the left ear and severe mixed HL on the right side with rising configuration. ABG also decreased from 47.5 to 31.25 for left ear and from 51.50 to 37.5 for right ear (Fig. 14.6f). She uses her hearing aids regularly and thresholds with hearing aids were between 35 and 20 dB. She made much better use of hearing aid after bilateral stapedotomy. Both her receptive and expressive language were significantly poorer than peers. Although she had better performance at closed-set tests (pattern perception and word identification), she performed poorly at open-set test (sentence recognition).

Case 14.3: (Patient #1) TU, 16-Year-Old Male Patient

His left ear was explored in 2008. Facial nerve was located at the oval window (Fig. 14.3b). Using a drill, a vestibulotomy was performed inferior to facial nerve. Using glass ionomer cement, incus was extended towards the opening in the vestibule, and a piston was attached between this extension and the vestibulotomy.

Surgery of Case 14.3 was difficult. We have to use a drill to make a fenestra. This may cause SNHL. Position of incus was over the FN. Therefore, cement was used to extend incus for attachment of the piston towards the vestibulotomy.

Preoperatively he had severe mixed type HL on the left and moderately severe mixed type HL on the right side with ABG of 50 and 41 dB (Fig. 14.7a). On the left side, there was a notch around 2000 Hz. He was operated on left side and his hearing thresholds were improved and ABG was decreased (Fig. 14.7b).

Case 14.4: HG 8-Year-Old Female Patient

She applied with bilateral moderate CHL. She had been using hearing aids until that time. In both ears, stapes was absent, and incus position was more oblique when compared to normal otosclerosis cases. Oval window was atretic bilaterally. It was not possible to make the fenestra using a perforator, necessitating the use of a 0.6 mm diamond drill to make the fenestra. Insertion of the hook of the stapes piston onto incus was more difficult when compared to nor-

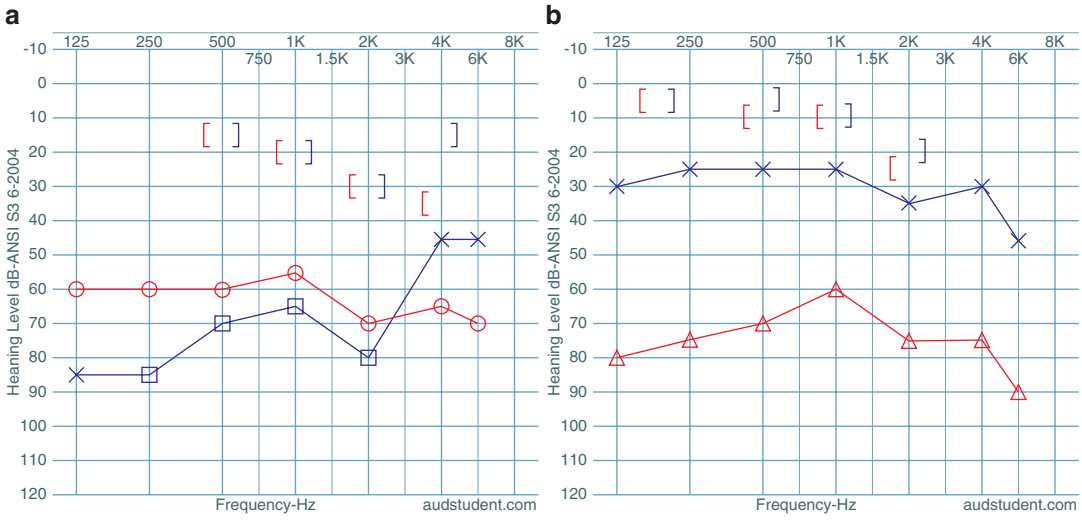

Fig. 14.7 Case 14.3 (**a**, **b**) Preoperative (**a**) and postoperative (**b**) hearing thresholds

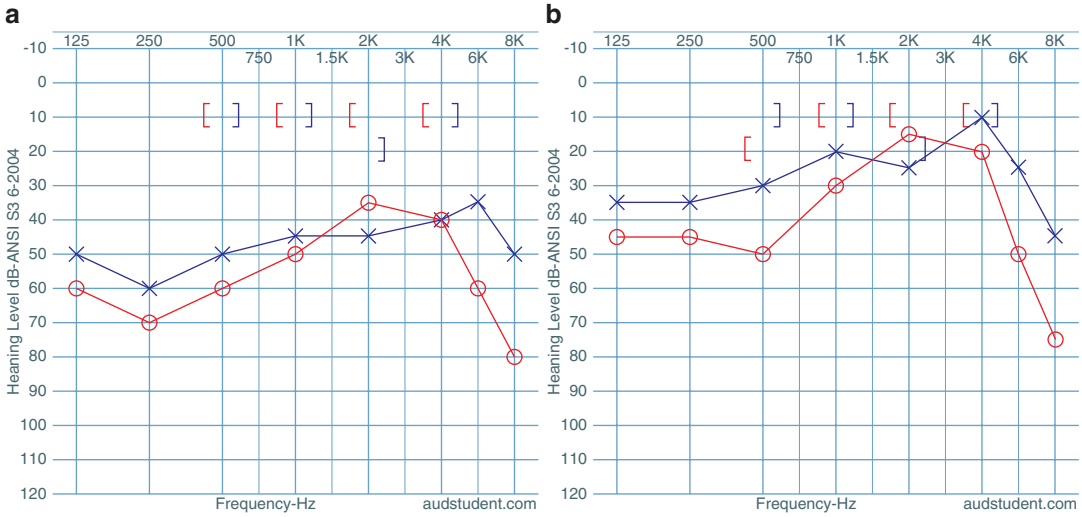

Fig. 14.8 Case 14.4 (**a**, **b**) Preoperative (**a**) and postoperative (**b**) hearing thresholds

mal stapedotomy, where the incus is more horizontal. Two drops of glass ionomer cement were used to fix the prosthesis to avoid sliding inferiorly.

Her preoperative audiogram showed bilateral moderate CHL with ABG of 34 dB on left ear and 38 dB on right ear (Fig. 14.8a). Although conductive component remains, postoperative audiogram showed bilateral improvement on air conduction thresholds. On left ear there was slight CHL with 12 dB ABG and on the right ear there was mild CHL with 19 dB ABG (Fig. 14.8b).

14.12 Outcome

Preoperative and postoperative hearing levels can be seen in Table 14.1. The average preoperative ABG was 46 dB and postoperative ABG was 21 dB.

In cases with pure CHL, it is possible to close the ABG and have near normal hearing without hearing aid. In children with mixed HL, the goal is to close the ABG as much as possible and allow the child to benefit more from the hearing aid. The latter should be made clear to the family when obtaining informed consent. In spite of tremendous improvement in all cases except patient #2 (Table 14.1), the results are not as good as stapedotomy in otosclerosis. This is due to the additional severe anatomical abnormalities.

14.13 Conclusion

Congenital stapes fixation and oval window abnormalities can be seen in inner ear malformations particularly accompanying cochlear hypoplasia. Stapedotomy is an acceptable treatment option as it provides sufficient hearing gain postoperatively. In mixed hearing loss the aim of the operation is to provide better benefit from hearing aid. The surgery may be complicated as a result of fixation of other ossicles, facial nerve abnormality, and CSF leakage. In the light of the findings of the present study stapedotomy has to be an option in conductive or mixed hearing loss in cochlear hypoplasia among other treatment alternatives.

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