Stapedotomy in Congenital Fixation with Cochlear Hypoplasia: A New Concept Among the Treatment Options for Cochlear Hypoplasia

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OBJECTIVES: To introduce the concept of stapedotomy as a new treatment alternative in cochlear hypoplasia (CH) and propose a new guideline for its management.

METHODS: Forty-two primary cases out of 355 presented with congenital stapes fixation between January 2003 and September 2015 were included in the study. Computed tomography scans of all cases with congenital stapes fixation were reviewed, and cases with inner ear anomalies were taken into account. Eleven cases had various inner ear anomalies, and 9 cases had various types of CH. In the present paper, only the CH cases with stapes fixation, all of whom underwent stapedotomy, are reviewed regarding preoperative audiological and radiological characteristics as well as surgical findings and postoperative audiological results.

RESULTS: The patients were aged between 4 and 22. There were 2 males (3 ears) and 4 females (6 ears). Three cases had bilateral stapedotomy. The remaining 3 cases had unilateral surgery. The average preoperative air–bone gap (ABG) was 50.3 dB. Postoperative hearing: preoperative ABG was 50.3 dB. Postoperative ABG was calculated as 20.1 dB hearing.

CONCLUSION: Hearing loss (HL) in hypoplastic cochlea demonstrates the full spectrum of HL types. CH is a unique inner ear anomaly that can be treated with all of the available rehabilitation modalities. As a result of current findings, a new treatment algorithm for CH is proposed.

KEYWORDS: Congenital stapes fixation, cochlear hypoplasia, congenital hearing loss, inner ear malformations, stapedotomy

INTRODUCTION

Stapes fixation can be due to congenital, otosclerotic, or tympanosclerotic etiology. If we exclude chronic otitis media and tympanosclerosis, congenital stapes fixation is very rarely encountered when compared to fixation due to otosclerosis. Vincent et al. reported that only 28 children among 2,525 patients were operated on for stapes fixation.4 Similarly, House et al. reported 13 children with stapes fixation in their large series.5

The most important symptom is nonprogressive hearing loss (HL) which is present since birth. The children present with a normal ear canal and tympanic membrane. Otitis media with effusion may be a coincidental finding, but conductive hearing loss (CHL) persists after the effusion is treated. In addition to stapes, other ossicles may also be fixed, the facial nerve may be anatomically misplaced to the oval window area, or the oval window may be atretic.

Treatment of CHL is very important at this age. Studies show that even in CHL, brain deprivation is unavoidable. For example, Webster and Webster (1979) found that experimentally produced CHL in mice at sensitive periods of development leads to insufficient maturation in most auditory brainstem neurons.6 Auditory abilities are also affected by these periods of non-stimulation, especially in the first year of life; otitis media in the first year of life was found to be associated with some of the higher-order
auditory processing abilities in the long term. Even single-sided uncorrected CHL may lead to deficits in auditory functions such as binaural processing.

Language development is another area that is affected by HL. Children at the language development stage who develop CHL are at risk of not learning important aspects of language. For this reason, sufficient hearing must be restored in these children as early as possible.

There are different treatment options in congenital ossicular fixation. Providing hearing aids is the earliest available option; however, in cases that present with severe ossicular pathology such as oval window atresia or ossicular discontinuity between the tympanic membrane and the oval 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 the use of bone-anchored hearing devices, but for children, the surgical placement of a bone-anchored device can be too premature. Bone conduction devices with head band appear to be the best option 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 addition, other ossicles may become fixed, necessitating surgical options such as manubrio-stapedioplasty or atticotomy.

Histologically, stapes fixation and oval window anomalies can be encountered in inner ear malformations. Recently, the first author investigated 41 temporal bone specimens with inner ear malformations in the Massachusetts Eye and Ear Infirmary (MEEI). Fourteen of the 41 cases with inner ear malformations presented either with oval window atresia or fixed stapes footplate. 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 CH, may cause CHL or mixed HL by stapes fixation.

Although stapedotomy is performed in middle ear anomalies, it is not defined in inner ear malformations. Therefore, the aim of this study is to report the clinical findings and outcomes of stapedotomy in cases with CH who had stapes fixation.

**MATERIALS AND METHODS**

We reviewed all cases of congenital CH managed between January 2003 and September 2015.

CH patients were defined by measuring the basal turn length; we included the cases with less than 7.5 mm length into the CH group (unpublished study). CH-IV is still an exception to this: they have a normal basal turn and hypoplastic middle and apical turns.

There were 9 ears with CH among 6 patients (Table 1). The CH patients were divided into 4 groups, as follows:

1. CH-I: cochlea with smaller external dimensions, with internal architecture (modiolus and IAC) absent, and with or without a thin bony partition between the cochlea and the internal auditory canal (IAC).
2. CH-II: external shape resembling a cochlea, but smaller and rounder than normal. Severely deformed modiolus resulting in a cystic cochlea.
3. CH-III: cochlea with smaller external dimensions and 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 smaller external dimensions, normal basal turn, and hypoplastic middle and apical turns.

**Table 1. Patient Characteristics, Operative Findings, and Audiological Findings of the Cases**

| Patient | Age | Sex | Side | Operation year | Operation findings | Preoperative Bone | Preoperative Air | Preoperative ABG | Postoperative Bone | Postoperative Air | Postoperative ABG | Basal Turn Length (mm) |
|---------|-----|-----|------|----------------|-------------------|------------------|------------------|------------------|------------------|------------------|------------------|------------------------|
| 1       | 16  | M   | Left | 2008 | FN at OW, drill to vestibulotomy, bone cement, Stapedotomy | 12 | 69 | 57 | 12 | 28 | 16 | 7.40 |
| 2       | 11  | F   | Left | 2012 | Stapedotomy, oozing | 10 | 54 | 44 | 10 | 45 | 35 | 6.72 |
| 3       | 8   | F   | Left | 2012 | OW atresia, drill to make vestibulotomy stapedotomy | 10 | 47 | 37 | 15 | 30 | 15 | 7.17 |
| 3       | 10  | F   | Left | 2012 | OW atresia, drill to make vestibulotomy stapedotomy | 6 | 47 | 41 | 17 | 40 | 23 | 7.07 |
| 4       | 4   | M   | Left | 2013 | FN inferior to OW, OW atresia, drill to vestibulotomy, gusher | 10 | 59 | 49 | 10 | 30 | 20 | 7.45 |
| 5       | 11  | F   | Left | 2013 | All ossicles fixed, atticotomy, stapedotomy | 50 | 115 | 65 | 44 | 61 | 23 | 6.23 |
| 6       | 22  | F   | Right| 2013 | Stapedotomy | 10 | 66 | 55 | 11 | 25 | 14 | 7.75 |
| 4       | 5   | M   | Right| 2014 | FN inferior to OW, OW atresia, drill to vestibulotomy, gusher | 10 | 55 | 45 | 10 | 31 | 21 | 7.42 |
| 5       | 13  | F   | Right| 2014 | All ossicles fixed, atticotomy, stapedotomy | 50 | 110 | 60 | 44 | 64 | 20 | 6.23 |
Preoperative audiological and radiological characteristics, surgical findings, and postoperative audiological results are reported.

RESULTS
In the present series, high resolution computerized tomography (HRCT) images of 43 primary congenital stapes fixation cases were reviewed, and cases with inner ear anomalies were taken into account. Eleven cases had various inner ear anomalies, and 9 cases had various types of CH. 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 in this paper). If the revision cases are excluded, 42 primary cases out of 355 presented with congenital fixation. Eleven ears of 6 patients had various inner ear pathologies. Nine had cochlear pathologies and 2 had only vestibular anomalies. The age of the patients ranged between 4 and 22 (Table 1). There were 2 males (3 ears) and 4 females (6 ears). There were 7 cases of CH-III and 2 cases of CH-IV. According to the description, in CH-III, basal, middle, and apical turns are smaller in size when compared to those of the normal cochlea (Figure 1). In CH-IV, the basal turn is normal, but the middle and apical turns are smaller than normal (Figure 2). Three cases had bilateral stapedotomy. The remaining 3 cases had unilateral surgery.

Preoperative Findings
Preoperative hearing level can be seen in Table I. The average preoperative ABG was 50.3 dB. Two cases were particularly interesting, one from the audiological point of view and the other from inheritance.

Patient 5 had severe mixed HL. Her air and conduction threshold average were 115 and 50 for the left and 110 and 50 for the right ear. The family was informed that the operation was planned to enable her to hear better with the hearing aid and was aimed at decreasing the air–bone gap. She was operated bilaterally. All of her ossicles were found to be fixed bilaterally. Because of the decrease in ABG postoperatively, she made better use of her hearing aid and her speech showed tremendous improvement.

Patient 6, with bilateral CH-IV, had pure CHL of 55 dB. Interestingly, her mother had exactly the same anomaly (CH-IV) but with profound sensorineural hearing loss (SNHL). Her mother had received cochlear implant (CI) surgery with severe facial nerve anomaly, necessitating complete removal of the ear canal and blind sac closure because the facial recess did not provide sufficient visibility of the round window and promontory. Her daughter had a unilateral successful stapedotomy.

Additional deformities: Patient 3 had severe heart anomaly and had undergone a heart operation in addition to cleft palate operation, prior to hearing evaluation. Patient 5 had facial anomalies and small oral orifice and auricles, in addition to a problem with eyesight.

Surgical Findings
Patient 1 had severe facial nerve abnormality at the tympanic segment. Normally, on coronal section, the facial nerve is located superior and lateral to the oval window (Figure 3). Patient 1 had facial nerve located at the oval window (Figure 4). Using a drill, vestibulotomy was performed inferior to the facial nerve. Using glass ionomer cement, the incus was extended toward the opening in the vestibule, and a Teflon piston was attached between this extension and the vestibulotomy.

Patient 2 had congenital cholesteatoma in the contralateral ear. She had undergone tympanoplasty with ear canal reconstruction 2.5 years prior to stapedotomy, with no recurrence of the disease.

The oval window was atretic bilaterally in Patient 3. It was not possible to make the fenestra using a perforator, necessitating the use of
Figure 4. Left coronal computed tomography showing the facial nerve over the oval window (White arrow).

Patient 5 had all ossicles fixed. In both operations, atticotomy and mobilization of the malleus and incus were performed before stapedotomy. A 0.6 mm Teflon piston of 4.5 mm length was used on both sides. On the left side, the facial nerve was found covering almost 75% of the footplate. No drilling was done as the footplate was easily perforated with a perforator pick.

Patient 4 had an anatomically misplaced facial nerve, which was located inferior to the oval window on both sides (Figure 5). His operation was the most difficult of the 9 cases. 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. By taking into account the position of the incus, a vestibulotomy was created using a 0.6 mm diamond burr. Cerebrospinal fluid (CSF) leakage occurred in both sides. A 0.4 mm Teflon 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. Both incuses were inclined medially, which made Teflon piston insertion difficult. A few drops of cement were used to fix the piston onto the angled incus. The patient had already been immunized by Pneumococcal vaccination. Ear examination at 3-month-intervals revealed no fluid in the middle ear that would suggest CSF. The patient regained near-normal hearing and excellent speech development without hearing aid (Video 1).

DISCUSSION

Until now, congenital stapes fixation has been regarded as part of the middle ear anomalies. Pathogenesis was believed to be related to developmental anomalies of the branchial arches. This theory is still valid, but the oval window is also part of the cochlea. The vestibular surface that forms the inner layer of the footplate is developed from the endosteum; therefore, it is not surprising to observe an oval window development anomaly because of inner ear malformation. This report is the first in the literature providing the link between CH and an oval window developmental anomaly.

Histopathological findings in IEM were published recently. Sennaroglu reported that footplate fixation or oval window atresia is seen in CH. In CH-I, the cochlea has a severe deformity like a bud, resulting in severe SNHL; therefore, stapes surgery is not done, but out of 3 CH-I cases in the MEEI temporal bone collection, 2 had a fixed stapes footplate and 1 had an aplastic oval window. In specimens with CH-II, the oval window was aplastic in 1, and the stapes footplate was fixed in 3. There were 5 specimens with CH-III: the oval window was normal in 1 specimen, fixed in 1, and aplastic in 3. This shows that oval window aplasia or stapes footplate fixation is common in CH. In cases of severe SNHL, usually CI or auditory brain stem implantation (ABI) (particularly in CH-I with cochlear nerve (CN) aplasia) is done. In CH-II, CH-III, and CH-IV, there may be mixed HL where stapedotomy can be successful. The patient may have to use a 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, 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. The arrest in the membranous labyrinth must happen between 10th and 20th weeks, after the basal turn reaches full size but before the middle and apical turns enlarge to their normal size.

This can be explained by embryology: the stapes footplate is part of the otic capsule, and according to Anson, 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 the invasion of the primitive cartilage by periosteal tissue. If there is an arrest of otic capsule development before the formation of the footplate, it is natural that the stapes become fixed to the oval window. However, it is still difficult to explain stapes fixation in CH-IV with normal basal turn.

Most probably, CH-I and CH-II cases also have a certain percentage of stapes fixation. However, due to the severe malformation and possibly due to the absence of the CN, they are candidates for CI or ABI.
The CH-II cases are accompanied by stapes fixation and a defective modiolus. The partial modiolar defect may not be diagnosed because of the resolution of present day HRCT, but histopathological examination shows a defective modiolus in all cases. These patients have SNHL because of the shorter cochlea, and the fixed footplate provides the conductive component. The author has performed stapedotomy in cases of CH with mixed HL. Postoperatively, these cases benefit more from HA. Patients with CH who have profound SNHL are candidates for CI. CH with cochlear aperture aplasia necessitates an ABI.

HRCT is very important in congenital conductive or mixed HL. Radiology may demonstrate ossicular fixation to the attic wall. Radiology also shows the type of pathology and the position of the facial nerve. In particular, a coronal section shows the position of the facial nerve in relation to the oval window. In the present series, there are cases with the facial nerve in normal position, at the oval window, and inferior to the oval window (Figures 3, 4, and 5). The surgery becomes very challenging if the coronal section demonstrates the facial nerve at or inferior to the oval window.

Another use of HRCT is to show the defect between the cochlea and the IAC, which may result in CSF leakage. Patient 4, who had CSF leakage, did not have a defect between the IAC and cochlea but presented with demineralization all around the cochlea, which may cause CSF from the subarachnoid space to reach the cochlea and hence result in CSF leakage at the time of fenestration into the vestibule.

Difficulties during Surgery
Surgical difficulties in these patients can be grouped under 5 headings:

Facial Nerve Anomaly
The facial nerve (FN) may take an unexpected route. In CH-IV, the labyrinthine segment of the FN is anterior to the cochlea. This is almost a pathognomonic finding. Therefore, the tympanic segment may not be in the usual location in these cases. The tympanic segment was located much superior to the oval window in Patient 6. In her mother’s case, the mastoid segment was anteriorly located, preventing visibility of the landmarks through the facial recess. The most difficult situations are where the FN is at (Patient 1) or inferior (Patient 4) to the oval window. Surgically, these are the most challenging cases, requiring FN monitoring and a stimulator.

Oval Window Atresia
Some cases do not have an oval window formation. Those cases can be classified as atretic and require drilling. Using a laser is not advisable because the bone is thick, and a laser may cause excessive heating, resulting in thermal damage to the FN. The correct location can be estimated by taking into account the position of the incus, and if present, the remnants of the stapes superstructure. During drilling, it is necessary to avoid making a tunnel. It is advisable to carefully lower the bony surface 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.

Incus Abnormality
In cases of otosclerosis, the incus is positioned more horizontally during surgical exploration and piston insertion is easier. In cases of oval window atresia, the incus is more obliquely positioned, possibly because of the absence of the stapes, making Teflon piston placement more difficult. As the incus is oblique, a few drops of cement can be applied to avoid piston movement postoperatively.

Involvement of other Ossicles
This finding is likely unrelated to the inner ear abnormality but makes the surgery more challenging nonetheless, involving the opening of the attic and the removal of all bony tissues mobilizing the ossicles. This was present in Patient 5.

CSF Leakage
This is the most serious surgical complication. If not managed properly, it may lead to meningitis. It is mandatory to have the children vaccinated before the procedure. Nowadays, Pneumococcal and Haemophilus vaccinations are routinely administered in many countries.

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

Outcome
In cases with pure CHL, it is possible to close the ABG and have near-normal hearing without a 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 in Patient 2 (Table 1), the results are not as good as stapedotomy in otosclerosis. This is due to the additional severe anatomical abnormalities.

In light of these findings, the treatment options for CH must be reevaluated. Until now, hearing aids, CI, and ABI were the only treatment modalities in CH. It is appropriate to also include stapedotomy and bone-anchored hearing aids (if there is an anomaly that cannot be corrected with the stapes procedure). The following algorithm is appropriate for children with hypoplastic cochlea (Figure 6): They should be evaluated with a thorough audiological examination and axial and coronal HRCT with MRI. In patients with mild to moderate SNHL, a hearing aid may be sufficient. In cases of CHL or mixed HL, stapedotomy may be performed to close the ABG as much as possible. In cases of mixed HL, stapedotomy should be performed to provide better use of hearing aids in the postoperative period. If severe SNHL is present and CN can be identified on MRI, CI with a short (around 20 mm) and thin electrode is appropriate. If CN is absent, ABI is the only treatment modality. If there are bilateral hypoplastic cochlear nerves, bimodal stimulation with CI and ABI may be appropriate.
Contraindications to Surgery in Congenital HL
For patients with CH with CHL, stapedotomy can be offered, but it is difficult to think of stapes footplate fixation in congenital mixed HL. The operation should never be done if HRCT demonstrates IP-II or IP-III.10

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

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REFERENCES
1. Kontorinis G, Goetz F, Lenarz T, Schwab B. Ear and further anatomic anomalies in children undergoing stapedotomy. Otol J Otorhinolaryngol Relat Spec. 2001;73(2):76-81.
2. Vincent R, Sperling NM, Oates J, Jindal M. Surgical findings and long-term hearing results in 3,050 stapedotomies for primary otosclerosis: A prospective study with the otology-neurotology database. Otol Neurotol. 2006;27(8)(suppl 2):S25-S47.
3. House JW, Sheehy JL, Antunez JC. Stapedectomy in children. Laryngoscope. 1980;90(11 Pt 1):1804-1809.
4. Webster DB, Webster M. Effects of neonatal conductive hearing loss on brain stem auditory nuclei. Ann Otol Rhinol Laryngol. 1979;88(5 Pt 1):684-688.
5. Wilmington D, Gray L, Jahrsdoerfer R. Binaural processing after corrected congenital unilateral conductive hearing loss. Hear Res. 1994;74(1-2):99-114.
6. Gravel JS, Wallace IF, Ruben RJ. Auditory consequences of early mild hearing loss associated with otitis media. Acta Otolaryngol. 1996;116(2):219-221.
7. Sennaroğlu L. Histopathology of inner ear malformations: Do we have enough evidence to explain pathophysiology? Cochlear Implants Int. 2016;17(1):3-20.
8. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balk Med J. 2017;34(5):397-411.
9. Sennaroğlu L. Cochlear implantation in inner ear malformations – a review article. Cochlear Implants Int. 2010;11(1):4-41.
10. Massey BL, Hillman TA, Shelton C. Stapedectomy in congenital stapes fixation: Are hearing outcomes poorer? Otolaryngol Head Neck Surg. 2006;134(5):816-818.
11. Anson BJ, Winch TR, Warpeha RL, Donaldson JA. The blood supply of the otic capsule of the human ear, with special reference to that of the cochlea. Ann Otol Rhinol Laryngol. 1966;75(4):921-944.