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Scala tympani drill-out technique for oval window atresia with malformed facial nerve: A report of three cases

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Abstract

Objective: To report a scala tympani drill-out technique for managing malformed facial nerve covering the entire oval window (OW).
Methods: Data from three cases with OW atresia, malformed stapes and abnormal facial nerve courses were reported, in which a scala tympani drill-out technique was employed with a TORP between the tympanic membrane and scala tympani fenestration for hearing reconstruction.
Results: Air conduction hearing improved in two of the three cases following surgery. In the third case, there was no improvement in air conduction hearing following a canal wall up mastoidectomy and tympanoplasty. There were no vertigo, tinnitus or sensorineural hearing loss in the three cases.
Conclusion: The scala tympani drill-out technique, which is basically fenestration at the initial part of the basal turn, provides a choice in hearing reconstruction when the OW is completely covered by abarrently coursed facial nerve.

Keywords: Oval window; Facial nerve; Malformation; Scala tympani; Fenestration

1. Introduction

In some rare cases of congenital aplasia of the OW, malformed stapes and abnormal facial nerve locations covering the OW present a challenge to reconstruction of the ossicular chain to improve hearing. Such malformations are typically acompanied by conductive hearing loss, normal external ear canal and normalsize middle ear cavity. The semicircular canal drill-out technique may be a treatment option, but the hearing outcomes are not always satisfactory (Su et al., 2014). Sterkers and Sterkers (1988) reported a vestibular drilling technique above the facial nerve at the usual location of the Fallopian canal. Plester (1971) reported the promontorial window technique and the prosthesis was inserted into the vestibular scala. Vibrant Soundbridge (VSB) implantation on the round window is a new technique and may be a good choice (Ahrsdoerfer, 1980; A1Dagna et al., 2014; Zernotti et al., 2013). In this paper, we report three cases in which the scala tympani drill-out technique was used. Details of this technique are introduced and discussed.

2. Materials and methods

Cases data

Table 1 lists the information of the three patients with bilatearl conductive hearing loss, normal external ear canals and tympanic membranes treated from July 2014 through August 2015. Pre- and post-operative air bone-gap (AB-gap) and AB-gap closure were recorded in accordance to the 1995

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Table 1
Patient profiles.

Case.	Sex	Age	Side	CT findings	Operative findings	Surgical treatment	AB-gap		AB-gap closure	Testing time
							Pre	Post		
1	M	11	L	Atresia of OW; Malformed stapes; Facial nerve located at the inferior border of the OW niche;	Osseous mass of stapes superstructure; horizontal facial nerve without bone canal cover the OW	Tympanoplasty with TORP implantation	45	15	30	3 months
2	M	8	R	Mastoiditis; Absence of stapes; Facial nerve located at the inferior border of the OW niche;	Absence of stapes superstructure; Horizontal facial nerve without bone canal cover the OW	Canal wall up mastoidectomy; tympanoplasty with TORP implantation	40	35	5	3 months
3	F	10	R	Atresia of OW; Malformed stapes; Facial nerve located at the inferior border of the OW niche;	Osseous mass of stapes superstructure; Horizontal facial nerve without bone canal cover the OW	Tympanoplasty With TORP implantation	55	20	25	4 months

guidelines by the Committee on Hearing and Equilibrium (1995).

Pre-operative coronal high resolution CT (HRCT) (Fig. 1A) showed the OW niche tapering to a"V"shaped central depression (yellow arrowhead) and the facial nerve (Fig. 1A & B, black arrowhead) located at the inferior border of OW atresia. The malformed stapes superstructure (Fig. 1C, red arrowhead) was seen over the exposed facial nerve. Scala tympani drill-out (Fig. 1C, white arrowhead) was located anterior-inferior to the round window membrane (Fig. 1B, blue arrowhead) and covered by a fascia graft (Fig. 1D, white dotted arrowhead), and a TORP (Fig. 1E: long white arrowhead) implanted between the tympanic membrane and the fenestration. Post-operative CT imaging (Fig. 1F) displayed the TORP in touch with the scala of tympani. Air conduction hearing was improved to 30 dBHL at 3 months after surgery (Fig. 4A) and has remained so at this writing. There was no vertigo, tinnitus or sensorineural hearing loss.

Case 2 presented with mastoiditis and no stapes. The facial nerve (Fig. 2A and B, black arrowhead) was completely exposed and entirely obliterated the oval window. Canal wall up mastoidectomy was completed, followed by fenestration of the scala tympani at a location anterior-superior to the round window membrane (Fig. 2A, white arrowhead). The TORP was implanted between the tympanic membrane and fenestration. Fig. 2D shows the TORP column base in touch with

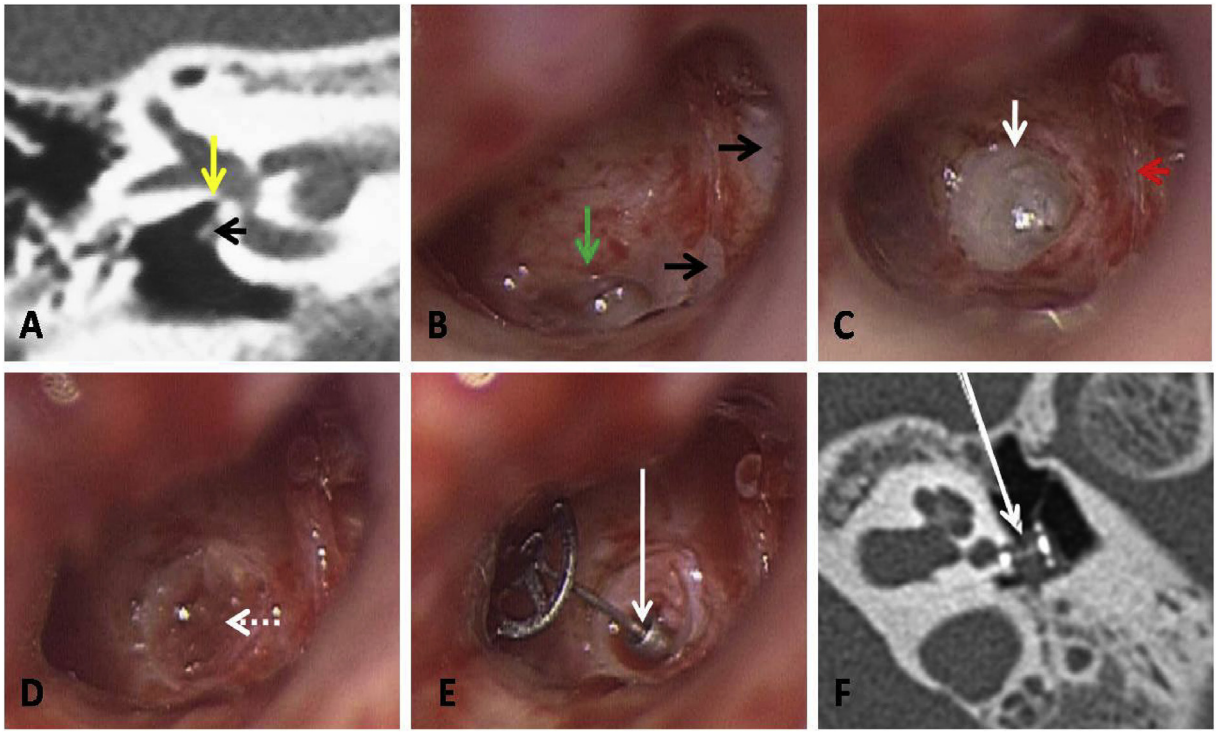

Fig. 1. Images from Case 1 A. yellow arrowhead:atresia of OW; black arrowhead: facial nerve; B. blue arrowhead: round window niche; black arrowhead: facia nerve; C. short white arrowhead: fenestra of scala tympani drilled-out; red arrowhead: malformed stapes superstructure; D. white dotted arrowhead: fascia on the bottom of fenestra; E&F. long white arrowhead: TORP.

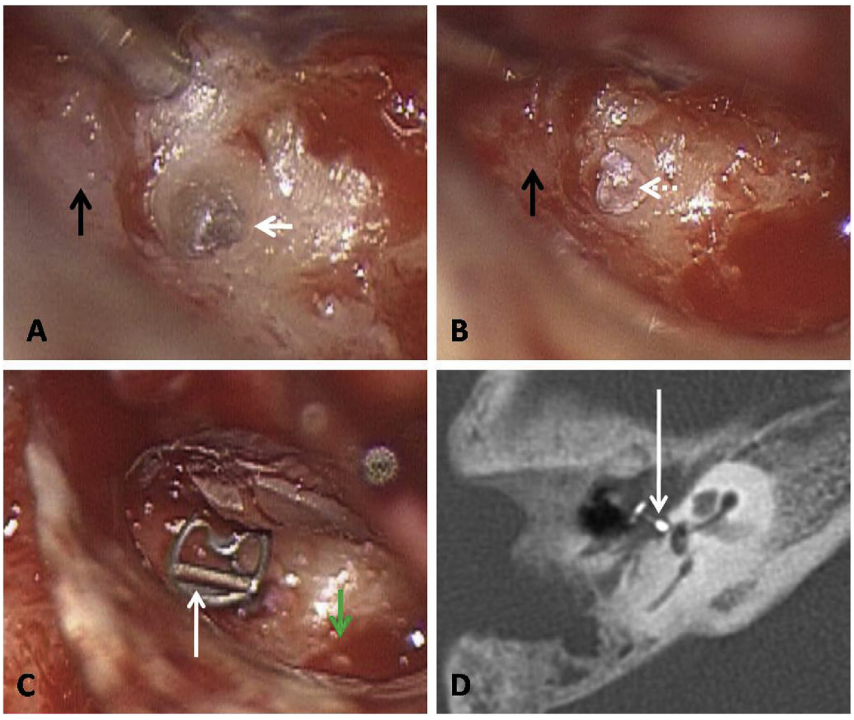

Fig. 2. Images from Case 2 A. black arrowhead: facial nerve; short white arrowhead: fenestra of scala tympani drilled-out; B. white dotted arrowhead: fascia on fenestration; C. blue arrowhead: round window niche; C and D. long white arrowhead:TORP.

the scala tympani. There was no vertigo, tinnitus or sensorineural hearing loss. There was no air conduction hearing improvement (Fig. 4B).

Case 3 had malformed stapes superstructure and incudostapedial joint (red arrowhead), which was pushed toward the promontory by the exposed facial nerve (black arrowhead). The oval window was completely covered by the facial nerve. Fig. 3B shows the position of scala tympani drill-out (white arrowhead) at a location anterior-inferior to the round window membrane. Fig. 3C and D show the implanted TORP (long white arrowhead) between the tympanic membrane and fenestration. The white dotted arrowhead indicates the

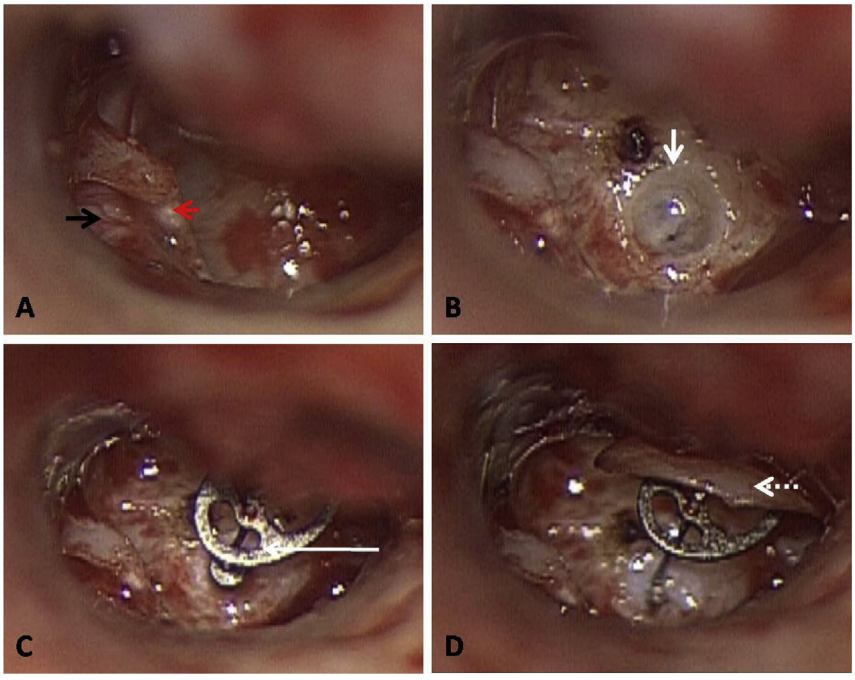

Fig. 3. Images from Case 3 A. black arrowhead: facial nerve; red arrowhead: malformed stapes superstructure; B. short white arrowhead: fenestra of scala tympani drilled-out; C. long white arrowhead: TORP; D. white dotted arrowhead: cartilage piece.

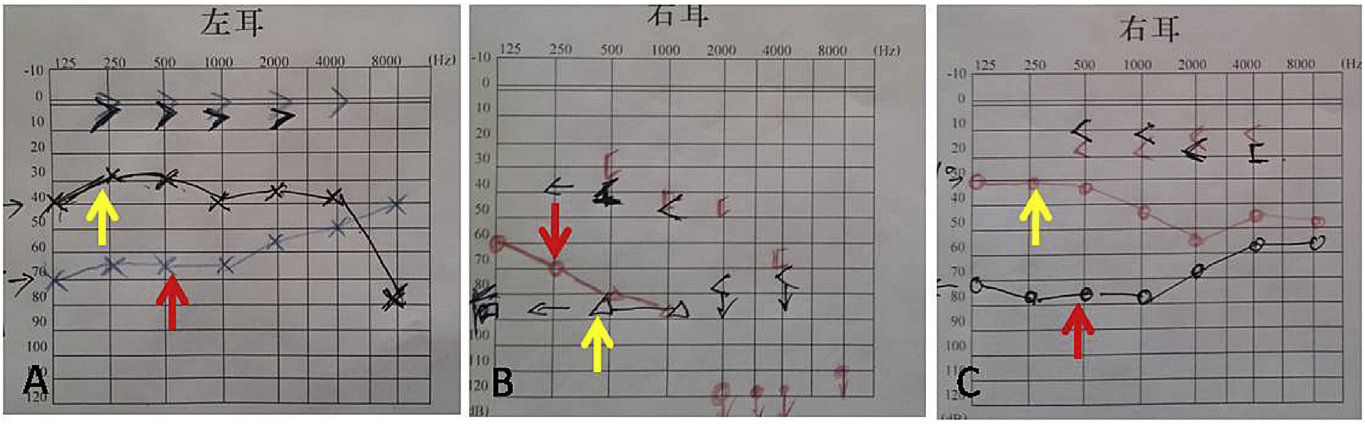

Fig. 4. Pre- and post-operative audiograms. A:case 1; B: case 2; and C: case 3. red arrowhead: pre-operative air conduction thresholds; yellow arrowhead: post-operative air conduction thresholds.

cartilage graft between the TORP and tympanic membrane. Air conduction hearing in this case improved to 25 dBHL at 4 months after surgery (Fig. 4C). There was no vertigo, tinnitus or sensorineural hearing loss.

3. Discussion

3.1. Diagnosis

Pre-operative diagnosis of congenital aplasia of the OW, malformed stapes and abnormal facial nerve courses can be difficult. Literature and our experiences indicate that HRCT and multi-planar reconstruction (MPR) play an important role in the pre-operative diagnosis of malformed middle ear structures. According to the reports by Zeifer et al. (2000), Veillon et al. (2001) and Yang et al. (2015), malformations of this area can be summarized as follows:1) tapering of the oval window niche to a"V"shaped central depression on coronal HRCT (layer thickness = 0.628 mm) (Fig. 1A, yellow arrowhead, Fig. 5B); 2) a footplate showing bone but no cartilage densities on coronal or horizontal HRCT (thickness = 0.7 mm or greater, Fig. 5A); and 3) the horizontal

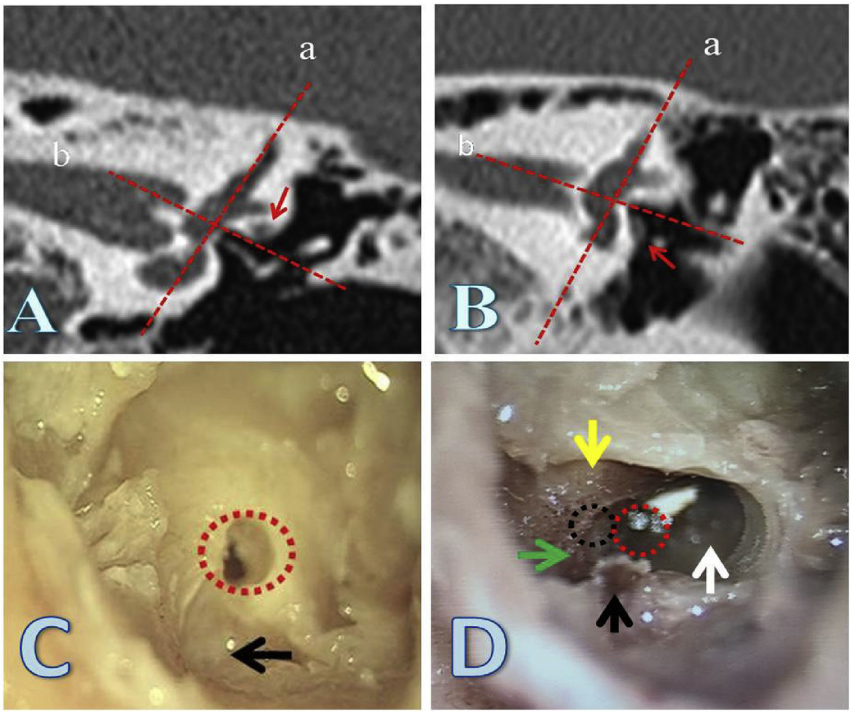

Fig. 5. Important landmarks. A and B. CT coronal imaging. line a: parallel to the plane of superior semicircular canal; line b:crossing "line a" through the foot of horizontal semicircular canal; red arrowhead: facial nerve (A: normal position; B: abnormal position). C and D: landmarks on cadevar specimen C. black arrowhead: round window membrane; red dotted circle: location of scala tympani drilled-out. D: black arrowhead: round window membrane; white arrowhead: scala tympani; yellow arrowhead: scala vestibuli; blue arrowhead: osseous spiral lamina; red dotted circle: desired location of scala tympani drilled-out; black dotted circle: less than optimal location of scala tympani drilled-out.

facial nerve located by a line parallel to the plane of superior semicircular canal (Fig. 5A and B, line a) and a line crossing "line a" at the foot of horizontal semicircular canal (Fig. 5A and B, line b) on coronal HRCT. Normally, the facial nerve is located in the upper outer quadrant of the red cross lines and below the horizontal semicircular canal (Fig. 5A, red arrowhead). If located in the lower outer quadrant and below the inferior border of the OW, it is abnormal (Fig. 5B, red arrowhead).

3.2. Location of fenestration

The position of fenestration must be considered firstly. The optimal location is the initial part of scala tympani, corresponding to the promontory wall anterior-inferior to the round window membrane (not the niche, Fig. 5C, red dotted circle). At this location, vibration through the fenestration can be best conducted to the perilymph in the scala tympani. If anterior-superior to the round window membrane, part of the fenestration may face the scala media and scala vestibuli (Fig. 5D, black dotted circle), and vibration can be conducted to the perilymph and endolymph at the same time, potentially damaging the basilar membrane, spiral ligament and stria vascular. In his cochlear implant research, Briggs et al. (2005) reported that, to avoid damaging the basilar membrane and spiral ligament, the position of cochleostomy should be located inferior, rather than superior, to the round window to ensure scala tympani exploration. In our cases, lack of hearing improvement in Case 2 maybe due to the fenestration that was anterior-superior to the round window membrane (Fig. 5D, black dotted circle). In case 2, infection in the middle ear cavity or cholesteatoma may also affect hearing results. Caution must be taken agaist fenestration in such case to prevent inner earinfection.

3.3. Fenestration techniques

The bone at the fenestration site is removed to the level of endosteum using a 1.0 mm diamond burr at low-speed until the blue shadow is seen. Care should be taken not to disturb the endosteum and the membranous labyrinth. Once the fenestra is created, a thin piece of fascia should be placed over the fenestration to protect and support the endosteum. After measuring the distance from the tympanic membrane to the fascia, a titanium TORP of appropriate size is placed between the tympanic membrane and the fenestration. Lastly, the base of the TORP should be sealed with fat graft to prevent perilymph leak, and the top of the TORP covered with a cartilage graft underneath the tympanic membrane to prevent prosthesis extrusion.

4. Conclusion

Congenital aplasia or atresia of the oval window (OW), with malformed facial nerve and stapes is a rare embryologic defect. The scala tympani drilled-out technique of fenestration in the initial part of scala tympani, provides us with a new option in hearing reconstruction when the area of OW is completely covered by the malformed facial nerve.

Conflict of interest

All of the authors declare that they have no conflicts of interest concerning this article.

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