Received: 12 September 2024 | Revised: 7 November 2024 | Accepted: 7 December 2024

DOI: 10.1002/lio2.70055

ORIGINAL RESEARCH

Laryngoscope Investigative Otolaryngology

Short-process incudo-stapedioplasty in congenital ear malformation

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Funding information
Japan Society for the Promotion of Science, Grant/Award Number: JP24K12696

Abstract

Objectives: Although various stapedotomy and stapedectomy techniques exist, anchoring the piston can be challenging. We present a novel surgical approach for treating congenital stapes malformations with an atypical facial nerve trajectory.

Methods: This is a case of a 7-year-old boy presenting with bilateral conductive hearing loss. Prior attempts at tympanoplasty had proven unsuccessful in improving his hearing. Presurgical imaging studies revealed an unusual anatomical configuration, with the facial nerve positioned inferior to the oval window. This anatomical variation precluded the use of conventional prosthesis-anchoring techniques typically employed in stapedotomies. Thus, we devised an innovative approach, opting to anchor the prosthesis to the short process of the incus.

Results: This novel technique circumvented the atypical course of the facial nerve, allowing for successful reconstruction of the ossicular chain. The patient demonstrated an acceptable improvement (30 dB gain) in hearing 1-year post-surgery, with no reported complications.

Conclusion: This case underscores the critical importance of adapting surgical techniques to address the unique anatomical challenges that may arise in the context of congenital ear malformations. It also highlights the potential of the short process of the incus as a viable alternative anchoring site for stapes prostheses, thereby improving the outcomes of such complex cases. This technique not only restored the patient's hearing but also contributed valuable insights into the management of similar cases, potentially improving the quality of life for individuals with rare and challenging anatomical variations.

Level of evidence: 5.

KEYWORDS
congenital ear malformation, incus, prosthesis, stapedectomy, stapedotomy

1 | INTRODUCTION

Conductive hearing loss due to congenital stapes malformations is very rare, but the diagnosis is typically established at 3rd month of age.[1]

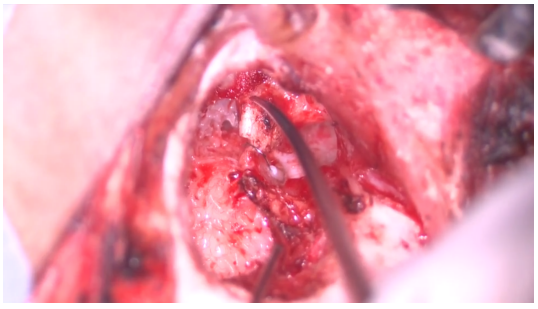
Because of the difficulties associated with anatomical variations, various stapedotomy and stapedectomy techniques have been developed. Stapedotomy is well known as an effective and safe procedure for treating stapes malformations. Incus stapedotomy and malleostapedotomy with microscopy have been widely used for stapes surgery. Additionally, the use of endoscopic stapedial surgery is also increasing. In these techniques, the prosthesis is usually anchored to the long process of the incus or handle of the malleus. However, in some patients, it is challenging to anchor the piston. Cases of congenital stapes fixation with facial nerve malposition are very rare.[2] Here, we report a case of a tympanic anomaly that was resolved using a novel prosthesis anchoring technique with the video of this surgery (Video 1).

2 | METHODS

This study was approved by the Ethics Committee of Okayama University Hospital (Approval #2010-026). To our knowledge, this is the first reported case using this surgical method.

A 7-year-old boy presented to our clinic with bilateral conductive hearing loss. The patient's pronunciation was indistinct. He had previously undergone ossiculoplasty at the age of 5 years for conductive hearing loss; however, the results were not satisfactory. During the previous surgery, he was diagnosed with a single stapes crus, a type of stapes malformation; the superstructure of the stapes resembled a short stick.[3] A cartilage columella was located between the incus and foot plate of stapes.

Pure-tone audiometric data before the second surgery showed conductive hearing loss and a Carhart's notch on both sides (Figure 1). This indicated that the previous reconstruction likely had unsatisfactory results. Computed tomography revealed that the facial nerve coursed inferiorly to the oval window on the right ear (Figure 2). Similar findings were also observed in the left ear.



VIDEO 1 The movie of the novel surgical method anchoring a prosthesis to the short process of the incus.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/lio2.70055

Informed consent was obtained from the patient and his parents. The surgery was performed under general anesthesia and via a retroauricular approach. The skin of the external ear canal was injected with 1% lidocaine with 1:100,000 epinephrine and elevated from behind the root of the right pinna to the tympanic annulus. After completing flap elevation, the short process of the incus and columella under the long process of the incus were identified. The scutum was already partially removed due to the previous surgery, and regrowth had occurred. An atticotomy was performed to secure working space. The lateral semicircular canal was clearly visible. Removal of the columella revealed that the facial nerve ran inferior to the suspected stapes because of an anatomical anomaly (Figure 3A). The facial nerve was not dehiscent. A hole was created in the suspected footplate using a perforator. The long process of the incus was too short for anchoring. The course of the facial nerve prohibited traditional methods of prosthesis anchoring. When we adjusted the degree of the prosthesis, it reached the short process of the incus without bending. Therefore, we anchored the prosthesis to the short process of the incus. The distance between the footplate and medial surface of the short process of the incus was measured to determine the required prosthesis size. A malleus-attaching piston prosthesis made of wire and Teflon (Schuknecht Wire Malleus Attachment Piston, 0.6 mm diameter, 6.0 mm length, Gyrus ACMI, MN, USA) was used. We drilled the upper area of the short process of the incus using a 1-mm skeeter drill; however, the degree of movement of the incus and piston was not sufficient, revealed by checking the stapes fixation with gentle testing of ossicular chain mobility. We drilled the lower area of the short incus process, and the efficiency reached a sufficient level (Figure 3B,C). Finally, the connective tissue was seated to seal the oval window and crimping area (Figure 3D). The posterior incudal ligament was not excised to avoid necrosis of the incus and to anchor the piston securely. There was no incidence of perilymphatic leakage during the surgery.

3 | RESULTS

One year later, hearing gains were observed on pure-tone audiometry as follows; 45 dB at 250 Hz (80–35), 35 dB at 500 Hz (70–35), 35 dB at 1000 Hz (65–30), 15 dB at 2000 Hz (55–40), 10 dB at 4000 Hz (50–40), and 25 dB at 8000 Hz (65–40). No bone conduction threshold shift was observed, and no facial palsy or dizziness occurred (Figure 1). The patient was satisfied with the results.

4 | DISCUSSION

The most common causes of congenital conductive hearing loss are stapes ankylosis and incudostapedial discontinuity.[1] Congenital stapes ankylosis represents 20%–35% of ossicular malformations and arises from the fixation between the peripheral lamina stapedialis and annular ligament.[1]

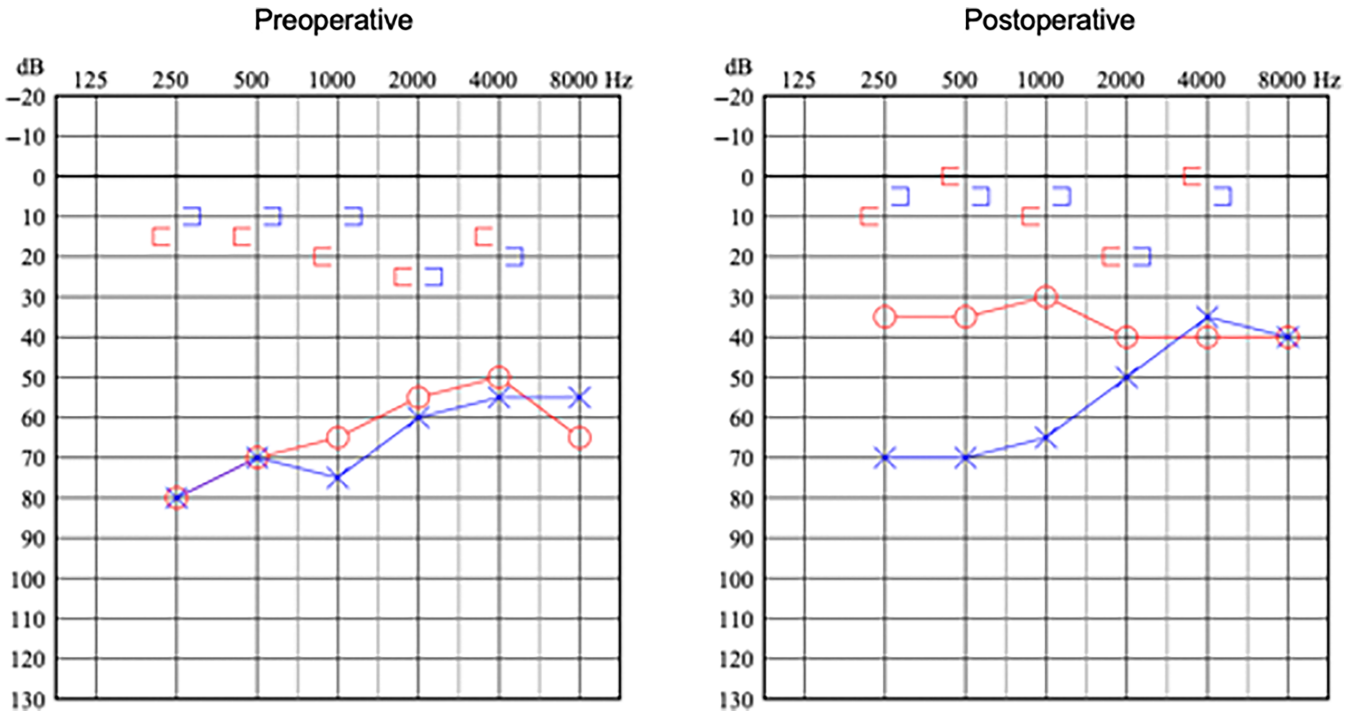


FIGURE 1 Pure tone audiometry before and after surgery.

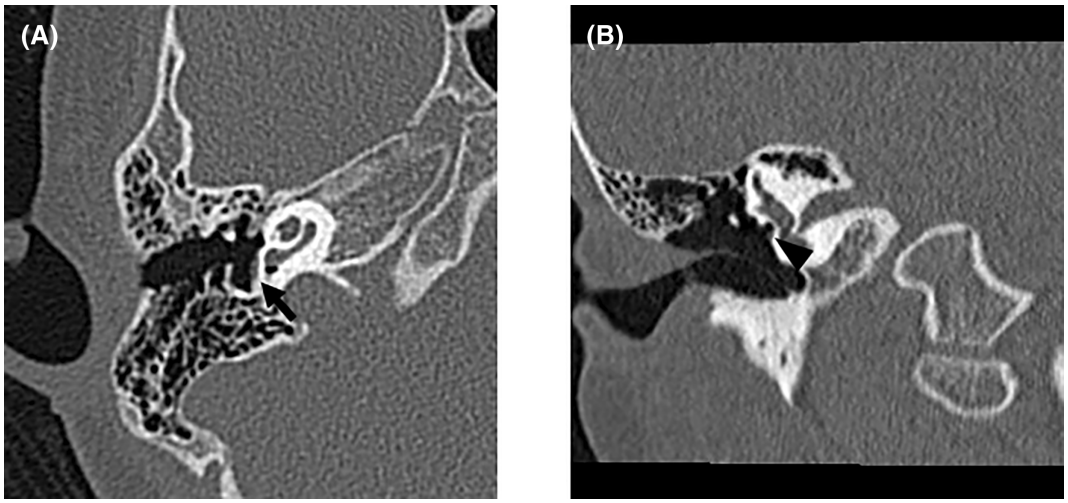


FIGURE 2 Computed tomography scan of the right ear. The facial nerve runs inferior to the oval window (A: Black arrow in axial view. B: Black arrowhead in coronal view).

Analyzing patients' anatomical features using high-resolution computed tomography before surgery provide valuable information. As observed in the present case, it is necessary for surgeons to consider that all patients with congenital stapes disease may have other congenital anomalies of the cochlea or facial nerve course to avoid iatrogenic injury. In our case, because of an anomaly of the temporal bone, the facial nerve ran on the opposite side of the oval window. An et al.[2] reported that an anomalous facial nerve in the middle ear cavity was found in 7 of 62 ears that underwent stapes surgery. Massey et al.[4] reported that 1 in 25 cases could not be completed because the facial nerve was positioned over the oval window, which precluded reconstruction.

In this case, the facial nerve was in a different position such that the piston could not be anchored to the long process of the incus. This technique requires the following: (1) Teflon wire pistons of suitable length and (2) a feasible way to establish a straight-line connection to the stapes.

Additional indications for our surgical technique are explained in the Methods section. Some reports have described that fenestration of the inner ear, including the vestibule and scala vestibuli, improves

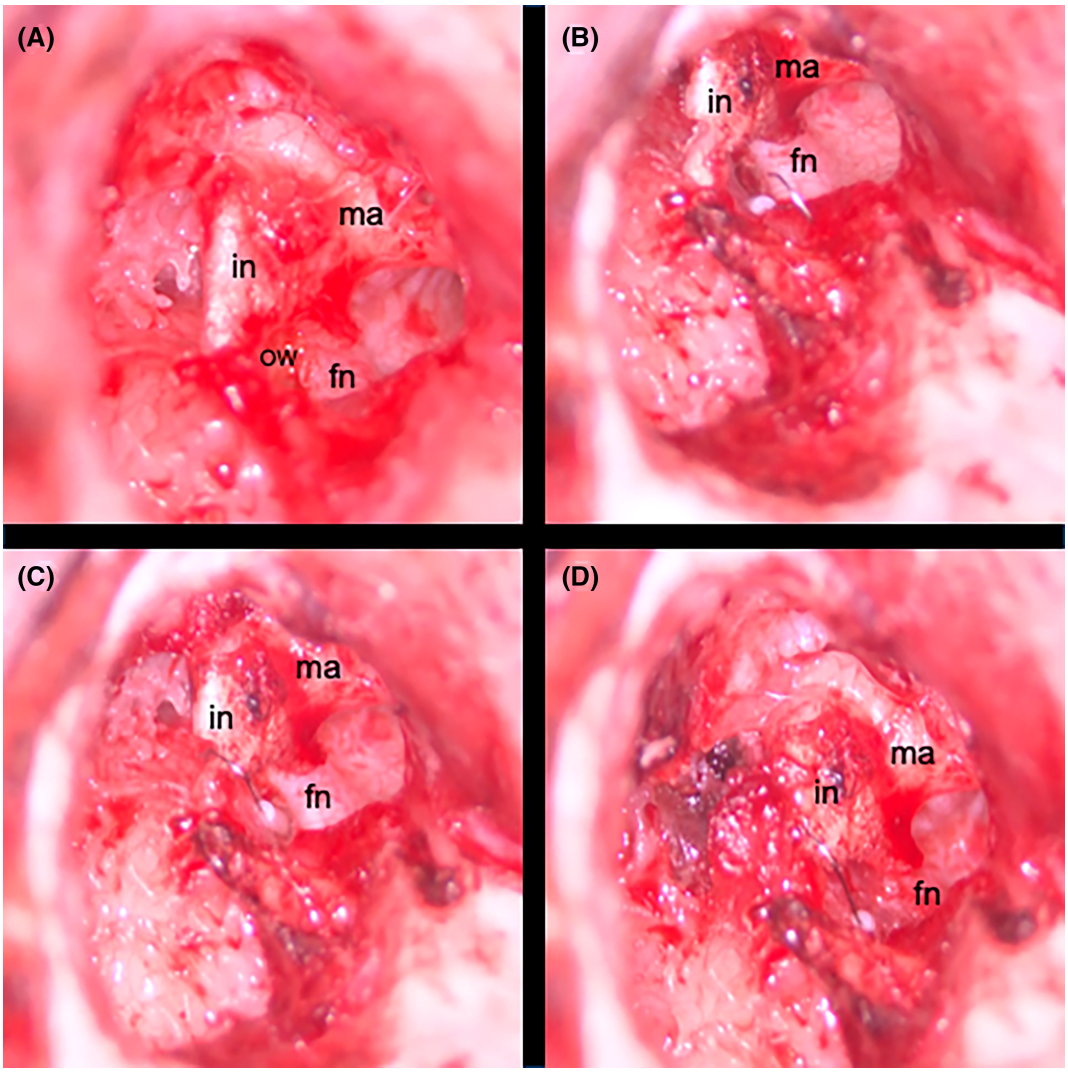


FIGURE 3 Surgical view of the right ear. (A) After atticotomy and removing the columella, the facial nerve can be seen running inferior to the oval window. (B) After arranging the gap on the short process of incus, the root is on the oval window. (C) After crimping the prosthesis on the short process of incus. (D) Sealing off the crimped area and oval window with connective tissue. fn, facial nerve; in, incus; ma, malleus; ow, oval window.

hearing without complications.[2,5] Considering the unclear visualization of the footplate, the fenestration of the vestibule may be performed. An et al. reported an interesting technique involving the attachment of the piston wire with soft tissue to the tympanic membrane.[2] While surgery through the ear canal may be considered an alternative approach for some patients, we deemed this method unsuitable for this patient due to his curved and narrow ear canal. Malleo-vestibulopexy could be considered an alternative procedure, but we determined that the angle was not appropriate for this approach. To the best of our knowledge, the only positions to anchor the piston are the long processes of the incus and malleus, excluding the case reported by An et al. Our method is the first in which the prosthesis was anchored to the short process of the incus. Although the air/bone gap was 20 dB, the hearing threshold improved from 65 to 30 dB at 1000 Hz. Even though longstanding closure of the air/bone gap to within 10 dB is expected in 78%–92% of the patients with otosclerosis, we believe this result is acceptable because of the unique anatomy.[4] The optimal location for the groove in this surgical technique is debatable. Although placing the wire distal to the axis of rotation may enhance the efficiency of stapes transmission, a more conservative approach is recommended. To ensure the safety and stability of the prosthesis, some margin should be maintained from the outside edge of the short process of the incus. This may necessitate placing the groove slightly more proximally, even if it potentially reduces the efficiency of energy transfer. The lower groove was also suitable for the stability of the prosthesis, but if we curved the groove more on the upper portion of the short process of incus, the air/bone gap may have decreased.

While our initial assessment suggested that a more distal location would optimize transmission within safety constraints, further discussion and investigation are warranted to determine the ideal groove placement for maximizing both stability and functional outcomes.

This surgical technique presents two points for debate: an intact posterior incudal ligament and functional incudomalleolar (MI) joint. The posterior incudal ligament plays an important role in incus stability; if it is compromised, the incus loses its secure fixation, predisposing the piston prosthesis to dislocation. Furthermore, disruption of the posterior incudal ligament can compromise the blood supply to the incus, especially if the ramus nutricia incudomallei is severed, potentially leading to incus necrosis. On the other hand, there is a possibility to reduce the air/bone gap if the ligament is compromised. Additionally, this technique is not feasible if the MI joint is disrupted, as it relies on the integrity of this articulation. In cases of a missing posterior incudal ligament and/or MI joint, the use of a columella or piston prosthesis may be considered. However, the choice of prosthesis is contingent upon the position of the facial nerve. If the facial nerve encroaches upon the surgical field, as in the present case, a bent malleus attachment may be used. If the surgeon deems ossiculoplasty unfeasible, bone-anchored hearing aids may offer a viable alternative.

The limitations of this report are as follows: (1) the lack of evaluation of long-term postoperative results; (2) age-appropriate word and sentence scores cannot be displayed; and (3) in some countries, bone conducting or bone-anchored hearing devices are options, but these could not be used in our patient due to the indication.

5 | CONCLUSION

Short-process incudo-stapedioplasty in congenital ear malformation results in good hearing improvement, without facial nerve injury or severe hearing loss. This method may be an option for stapes surgery in cases of conductive hearing loss.

FUNDING INFORMATION

This work was supported by JSPS KAKENHI Grant Number JP24K12696.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest.

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How to cite this article: Omichi R, Kariya S, Sugaya A, Ando M. Short-process incudo-stapedioplasty in congenital ear malformation. *Laryngoscope Investigative Otolaryngology*. 2025;10(1):e70055. doi:10.1002/lio2.70055