

Case Report

Endoscopy-Assisted Transmeatal Cochlear Implantation in Multiple Ear Deformities

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To study congenitally deaf children with inner ear malformations that usually have comorbid anomalous facial nerves and middle ear deformities. To determine the feasibility of endoscopy-assisted transmeatal cochlear implantation with the purpose of reducing the risks of iatrogenic facial nerve injury. This report presents a unique technique in a pediatric case with multiple ear anomalies: microtia, cochlear hypoplasia with an aberrant facial nerve, a sigmoid sinus deformity leading to a narrow mastoid cavity, and a flat promontory wall without round window. A cochlear implant electrode array was successfully inserted endoscopically using the transmeatal approach in the present case. It caused no postoperative surgical complications, and the patient was then able to hear binaurally and functionally. For patients with comorbid multiple ear deformities, using endoscopy for cochlear implantation transmeatally is an alternative method providing a better visualization of the middle ear anatomy, an avoidance of injury of facial nerve, and an assurance of precise insertion of the electrode when the posterior tympanotomy approach is not applicable.

KEYWORDS: Cochlear implant, inner ear malformation, congenital deafness, facial nerve, sigmoid sinus

INTRODUCTION

The facial recess approach, with electrode arrays inserted into the scala tympani after a posterior tympanotomy, is well established as the standard technique of cochlear implantation (CI). For patients with anatomic malformations, particularly a narrow mastoid cavity, alternative methods without mastoidectomy have been reported.^{1–4} Cochleovestibular anomalies like a common cavity, an incomplete partition (IP), cochlear hypoplasia (CH), an enlarged vestibular aqueduct, and hypoplasia of the internal auditory canal (IAC), are surgically challenging and yield uncertain auditory outcomes.⁵ We report a case of inner ear malformation (IEM) with an anomalous facial nerve and a narrow mastoid cavity for which we used endoscopy-assisted transmeatal approach as an alternative method of CI.

CASE PRESENTATION

The patient in this case was 16-year-old congenitally deaf boy with a 46xy, inv (9) (P13q21.21) karyotype and comorbid with multiple anomalies (cleft palate, right microtia, and right facial asymmetry). His genetic inversion conformed with none of the established syndromes. He visited our hospital first in 2002, when an auditory assessment indicated bilateral profound sensorineural hearing loss. High-resolution computed tomography (HRCT) of the temporal bone showed that the patient's left cochlea had less than 2 turns (CH-III) and his right cochlea had hypoplastic middle and apical turns (CH-IV) (Figure 1). He had a left facial nerve anteriorly and inferiorly displaced and a right facial nerve anterior to the cochlea. Because his hearing aids had limited value, he had received a cochlear implant (Nucleus CI24R) in his left ear using the retrofacial approach in 2004 when he was 3 years old, as previously reported.⁶

Postoperative rehabilitation led to gradual significant improvements in the patient's hearing and speech. Although he kept using the right hearing aid, the speech detection threshold was 75 dB HL (decibels of hearing level) and the word discrimination score was 0% in his right ear. A second CI was requested to improve his binaural hearing. A follow-up temporal bone HRCT showed the electrodes of the left implant well within the left cochlea, and his right sigmoid sinus had developed an engorged emissary vein leading

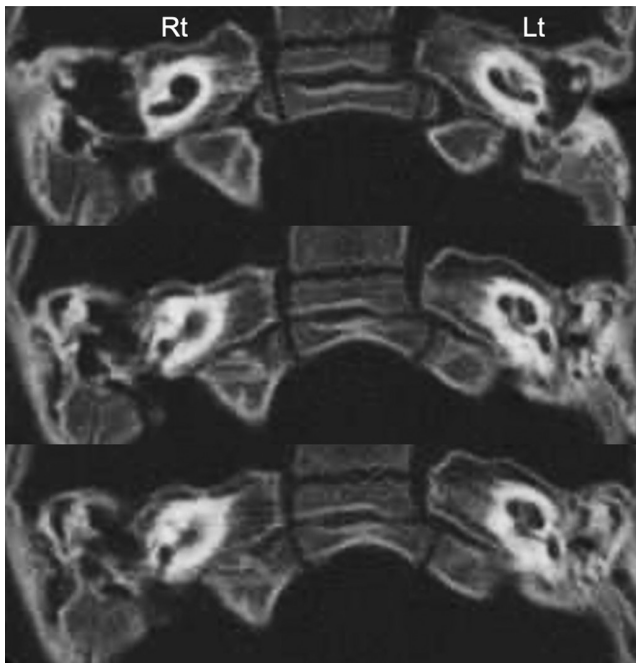


Figure 1. Axial sections of first high-resolution computed tomography (HRCT) in 2004 show bilateral cochlea hypoplasia with the right side more severe. Rt, right; Lt, left.

to an extremely narrow mastoid cavity (Figure 2). Considering the deformed cochlea and mastoid cavity, we designed a “endoscopy-assisted transmeatal CI” for this case in March 2018.

We first made a standard post-auricular incision and then created an anteriorly based periosteal tympanomeatal flap elevated to the annulus. The external auditory canal (EAC) was widened using cutting burrs, and a groove was drilled along the posteroinferior aspect of the EAC and turned into a tunnel when accessing the annulus. This tract was prepared as a route for the electrode array. A simple mastoidectomy and a well for a receiver-stimulator were prepared as usual. After elevating the tympanomeatal flap, a transmeatal exploratory tympanotomy was done using a rigid 0-degree endoscope (3 mm diameter; 14 cm long) (Karl Storz Co., Tuttlingen, Germany) and a connected HD camera system (Karl Storz). We saw a flat promontory without a visible round window niche. No facial canal could be grossly identified, but a bony prominence on the cochlea showed electrical signals on the intraoperative facial nerve monitor, which indicated an inferiorly displaced tympanic segment of a facial nerve (Figure 3A).

The round window is normally about 2 mm (range: 1.39–3.57 mm) inferior to the oval window.⁷ In this case, however, the expected round window was obscured by the displaced facial nerve (Figure 3B [white circle]). A cochleostomy was performed inferior to the facial nerve (Figure 3B [black circle]), and the scala tympani was exposed with a minimal gusher (Figure 3C). A slim, straight electrode array of the device (Nucleus CI422) was fully inserted into the scala tympani (Figure 3D). The rest of the wire was placed in the groove and covered with bone wax and chopped concha cartilage. The impedance level of all electrodes was within normal limits, and all channels showed a moderate current unit in the neural response telemetry immediately after operation. There were no remarkable

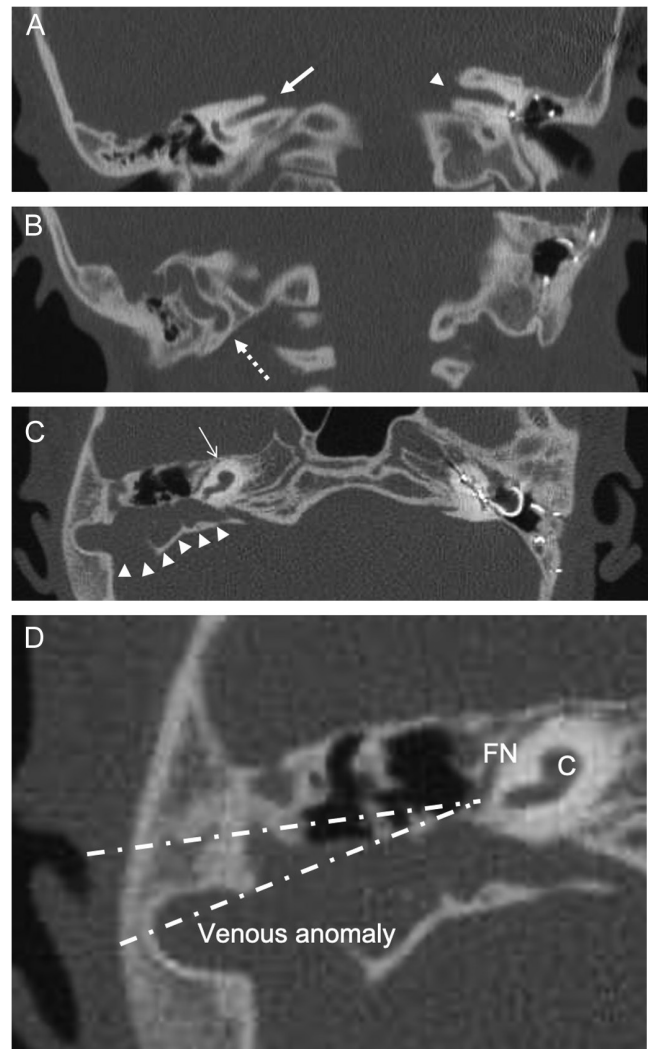


Figure 2. HRCT in 2018 before the second cochlear implantation. (A) The coronal view shows the relatively narrow right IAC (1.79 mm in diameter, arrow) compared with left side (2.41 mm in diameter, arrow head); (B) The coronal view shows the engorged emissary vein within the right mastoid bone (dotted arrow); (C) The axial view shows the right hypoplastic cochlea (long arrow) and deformed sigmoid sinus (line by arrowhead); (D) The axial view shows the small mastoid cavity with a limited retrofacial recess (angles between dotted lines). C, cochlea; FN, facial nerve.

intraoperative or postoperative complications, and the electrode was well placed in the cochlea as shown in postoperative HRCT (Figure 4). After 12 months of follow-up, his second implanted ear had certain auditory perception with a score of 28% in the Lexical Neighborhood test, and it performed well even in a noisy environment with an only larger signal-to-noise ratio than that of the left ear (+15 dB vs. +1 dB, respectively, in the Hearing in Noise test).

DISCUSSION

About 20% of patients with congenital sensorineural hearing loss have IEM, which is generally categorized as CH and IP.^{5,8–11} Probably 14–16% of IEMs—most often cases of CH with an unknown etiology—are comorbid with an anomalous facial nerve.¹² Sennaroglu et al.¹³ suggested that surgeons should be prepared to make necessary technical modifications. In our case, the tympanic segment of the right facial nerve was displaced anteriorly and inferiorly, which blocked the round

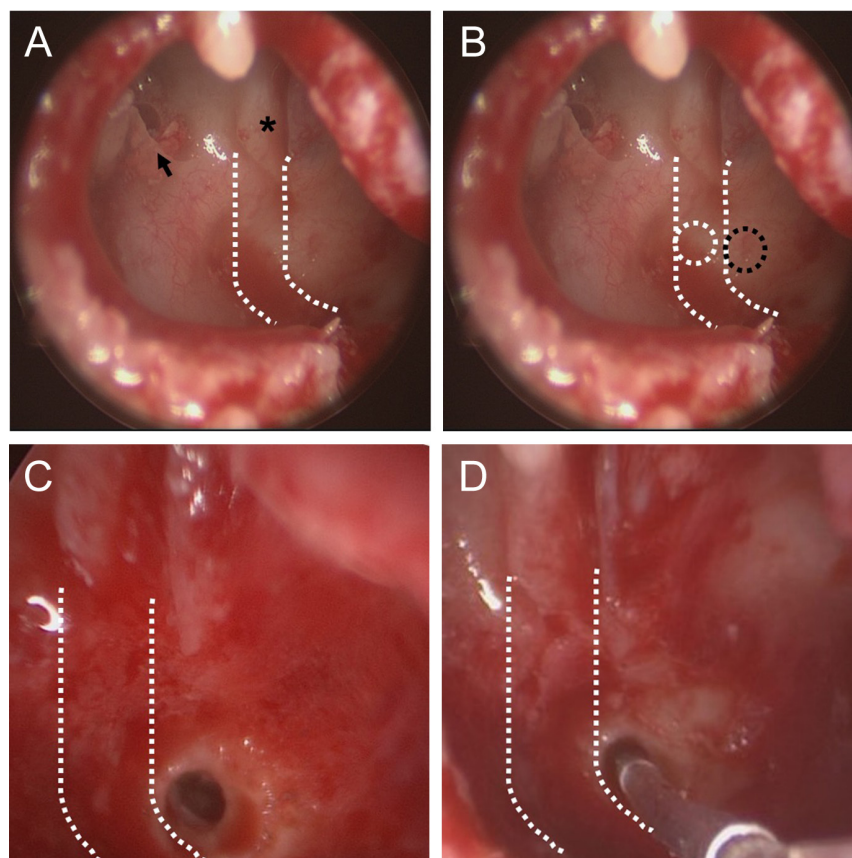


Figure 3. Intraoperative endoscopic view. (A) shows the oval window (arrow), a flat promontory wall with a facial nerve prominence (asterisk) on it, and the assumed course of the facial nerve (dotted lines); (B) shows the suspicious round window (white circle) 2 mm from the oval window and the cochleostomy site (black circle); (C) cochleostomy was done; (D) fully inserting the electrode array.

window. Moreover, the patient's deformed sigmoid sinus obscured the facial recess such that a posterior tympanotomy or retrofacial pathway was impossible. Consequently, we needed a safe and feasible alternative approach to prevent unwanted facial palsy and massive bleeding.

Apart from radical middle fossa approach, there are 3 non-mastoidectomy procedures: the suprameatal approach and its modifications by conducting the electrode through a tunnel drilled in the suprameatal region superior to Henle's spine¹; the transcanal approach, or Veria technique,² performed by drilling a subperiosteal tunnel parallel to the long process of the incus, from 11 o'clock laterally to 9 o'clock medially; and the pericanal approach,³ which places an electrode directly through a vertical groove made at the posterosuperior region of the bony EAC. These procedures have the advantages of eliminating facial nerve or chorda tympani injury and of shortening operation time, without significantly higher complication rates compared to the conventional mastoidectomy approach.¹⁴ However, the electrode must be obliquely (posterosuperiorly) inserted and then turned to a near-vertical direction relative to the cochleostomy and redirected in an anteroinferior direction into the scala tympani. Heightened caution and special skills are required to prevent kinking or misplacing the electrode.

We modified some alternative procedures. First, we used endoscopy to visualize the hidden area of the middle ear obscured by the posterior canal wall, which does not require drilling or curetting the bony

structure, and is useful for pediatric patients with IEM.¹⁵ Although a flat promontory was identified in this case, the location of the scala tympani could be predicted by recognizing the facial nerve prominence and oval window, using endoscopy. Second and unique to this case, we created a groove for the inducing electrode, in order to insert the electrode without kinking the wire and also to avoid injuring the aberrant facial nerve. With no need to change the direction, it ensured the insertion of electrode array into the cochlea through a natural course with a low resistance.

After 1 postoperative year, our patient could verbally communicate using the second implant. In a large-scale study, there was no significant difference in speech reception scores between children with and without an anomalous cochlea.⁵ Accordingly, anomalous cochleovestibular anatomy should not play a significant role in candidacy assessment.

CONCLUSION

Our transmeatal approach assisted by an endoscope might be a feasible alternative method for complicated cases. This approach should be used only if the standard transmastoid route with posterior tympanotomy approach is not possible.

Informed Consent: Informed consent was obtained from the patient who participated in this study.

Peer Review: Externally peer-reviewed.

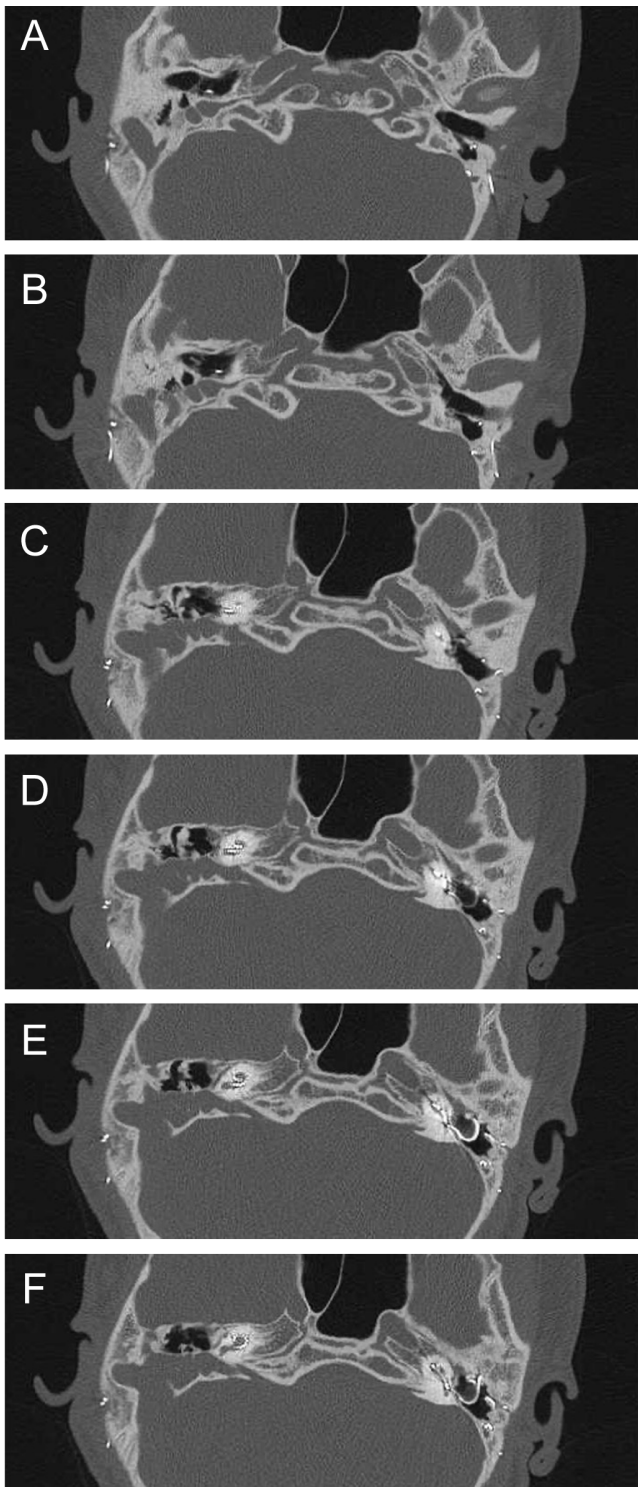


Figure 4. (A-F) Axial views of the postoperative HRCT show the course of electrodes from the external auditory canal to the right cochlea.

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