CASE REPORT
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Simultaneous Bilateral Cochlear Implantation in an Adult with Bilateral Ossification of the Scala Tympani: Image-Guided Planning of Scala Vestibuli Insertion and Postoperative Hearing Outcomes

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ABSTRACT

The scala tympani is the preferred position for cochlear implant electrode array insertion to ensure optimal outcomes. Here, we report the case of an adult with bilateral rapidly progressive profound SNHL and bilateral ossification of the scala tympani. Image-guided surgical planning was used to assist bilateral simultaneous cochlear implantation, gain direct access to the scala vestibuli, and determine the electrode array length. The short-term postoperative auditory performances are presented and highlight that meticulous planning and management by an experienced cochlear implant team enable optimal auditory performances even in anatomically difficult cases.

Keywords: Bilateral, sensorineural hearing loss, cochlear implantation, cochlear fibrosis, scala vestibuli

Introduction

Cochlear implantation (CI) is routinely used in patients with severe-to-profound sensorineural hearing loss (SNHL) for whom hearing aids do not provide adequate speech understanding. Cochlear implantation involves the placement of an electrode array to provide electrical stimulation to the remaining auditory ganglion cells. The preferred position of the electrode array is the scala tympani, accessed through the round window membrane, to ensure optimal hearing outcomes.[1-3] However, in some cases, the scala tympani is not patent due to either partial or complete ossification. This "labyrinthitis ossificans" can be caused by meningitis, otosclerosis, Cogan syndrome, autoimmune inner ear diseases etc.[2] It presents the CI team with a challenging situation with respect to the surgical approach, electrode selection, and insertion.

Several studies have compared the auditory performance of the scala tympani electrode position with that of the scala vestibuli electrode position, and the results are variable. O'Connell et al, for example showed negative results, while Gu et al and Trudel et al demonstrated at least comparable results.[1,2,4] It is unclear why these results vary; however, the level of ossification may have an effect on unexpected drill-out procedures that are more invasive towards the osseous spiral lamina and basilar membrane. On the other hand, electrode array translation can also explain this variability.[5,6]

We present the case of an adult patient with bilateral rapidly progressive profound SNHL and progressive bilateral ossification of the scala tympani who underwent bilateral simultaneous CI with image-guided surgical planning of scala vestibuli insertion.

Case Presentation

A 20-year-old male presented at the Dept. of Otorhinolaryngology and Head & Neck Surgery at the hospital with rapidly progressive bilateral severe-to-profound SNHL. At the first presentation a bilateral moderate SNHL and bilateral

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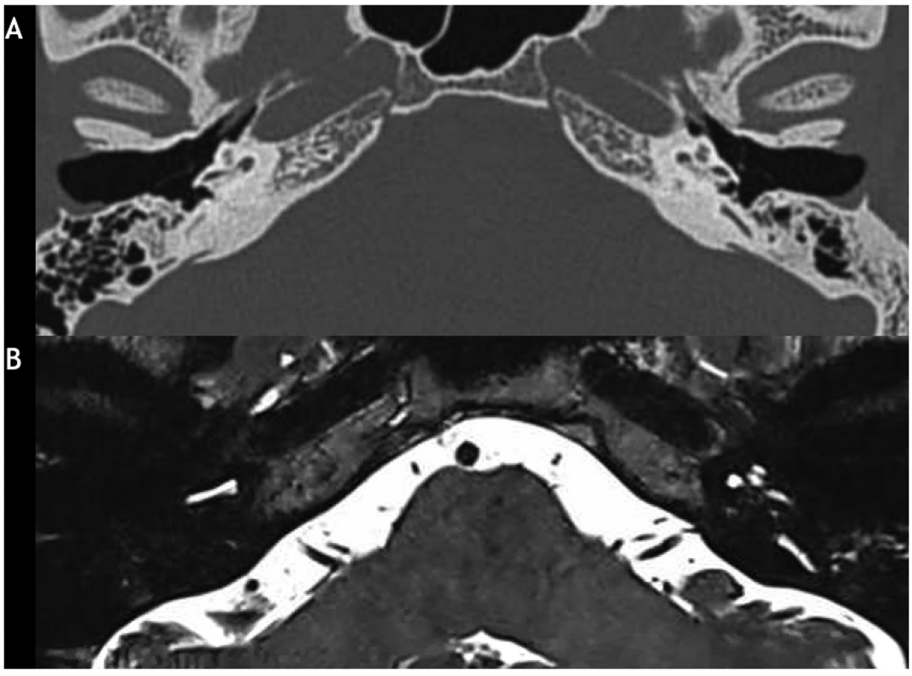

Figure 1. Preoperative temporal bone imaging. A. High-resolution temporal bone CT scan identifying bilateral ossification of the scala tympani. B. Magnetic Resonance Imaging (MRI) of the temporal bone and cerebellopontine angle confirming the loss of T2-signal but patent scala vestibuli without fibrosis.

absence of caloric response on electronystagmography were identified. High-resolution temporal bone CT and MRI were unable to identify anatomical abnormalities. In the next few years, the hearing level evolved towards profound deafness three years after the first presentation. The patient has a medical history of ulcerative colitis treated with methylprednisolone and azathioprine. There was no known family history of hearing loss, and a hearing loss gene panel testing analysis in 2019 was unable to identify any abnormalities.

Microscopic evaluation of the external auditory canal and tympanic membrane was normal. Tympanometry demonstrated a normal type A curve bilaterally, confirming normal aeration of the middle ears. At final presentation, no response could be identified using tonal liminal audiometry and speech audiometry. No reproducible Auditory Brainstem Recording (ABR) traces could be detected. A repeat high-resolution temporal bone CT scan meanwhile demonstrated bilateral ossification of the scala tympani in the basal turn, while the absence of T2-signal was confirmed on MRI (Figure 1A-B).

Main Points

- The scala tympani is the preferred position for the cochlear implant electrode array to ensure optimal auditory performance.
- Ossification of the scala tympani can be readily diagnosed with repeat CT imaging.
- Image-based surgical planning enables the exact determination of the scala vestibuli cochleostomy site.
- Cochlear duct length measurements can also be performed in cases of anticipated scala vestibuli position to determine optimal electrode length.

Whole-exome sequencing was performed to ensure a more in-depth genetic analysis. A heterozygous c.2719G>A p.(Gly907Arg) variant was detected in the *COL4A3* gene, previously unreported and absent in the gnomAD control population. This variant may potentially be pathogenic but does not provide a definitive etiology for the hearing loss. *COL4A3* mutations are known causes of Alport syndrome type 2 (OMIM #203 780) and autosomal dominant benign familial hematuria (OMIM #141 200).

The patient opted for bilateral simultaneous CI. Image-guided planning software (Otoplan, Cascination Med-EL) was used to analyze the preoperative high-resolution temporal bone CT scan and determine cochleostomy site and cochlear duct length (Figure 2A-D). The anatomy of the facial recess, ossicles, and cochlea was reconstructed three-dimensionally in order to determine the exact cochleostomy site to enable the opening of the scala vestibuli with a structure preservation technique (Figure 1). The calculated cochlear duct length (organ of Corti from 0 degree) on the right side was 33.20 mm and 34.57 mm on the left side. Because of the intended scala vestibuli cochleostomy, a 24-mm flexible lateral wall electrode (Med-EL Synchrony 2 S-vector Flex 24) was chosen to ensure full insertion.

The surgical procedure, bilateral simultaneous CI, was performed by the senior author. A standard facial recess approach was used, and cochleostomy in the scala vestibuli was performed on both sides. Full insertion of the electrode array was achieved and confirmed on a postoperative high-resolution CT scan. The postoperative period was uneventful with no pain, no vertigo, no facial weakness, and discharge after an overnight stay. The speech processors (RONDO 3, MED-EL GmbH, Innsbruck, Austria) were activated three weeks after surgery. The 3-month auditory performances are presented in Table 1

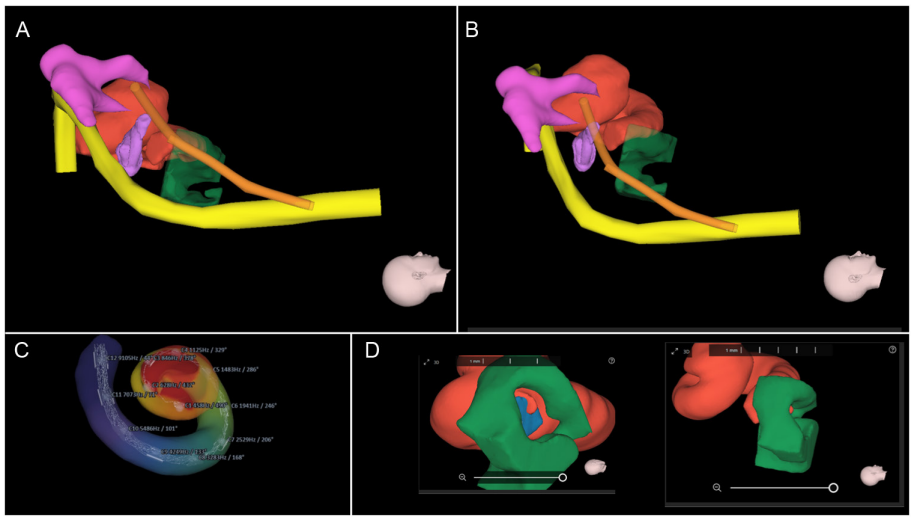

Figure 2. Preoperative image-guided surgical planning (Otoplan, Cascination). A-B. Three-dimensional reconstruction of the right ear with slightly different head angles to demonstrate the absence of the scala tympani but the presence of the scala vestibuli (red). C. Three-dimensional planning with a 24-mm flexible lateral wall electrode to determine optimal cochlear coverage. D. Reference of normal surface anatomy of a right ear with patent scala tympani and scala vestibuli. Yellow, facial nerve; orange, chorda tympani; purple, ossicles; red, cochlear turns; green, round window niche; blue, round window membrane.

Table 1. Evolution of Auditory Performance After CI Activation

	Baseline		3-Month Post CI Activation		
	Unaided Right	Unaided Left	CI Only (Right)	CI Only (Left)	Best-Aided (Both CIs)
Pure tone average (PTA)[a]	>120 dB HL	>120 dB HL	25 dB HL	28 dB HL	/
Speech in quiet (SPIQ)[b]	0%	0%	88%	97%	94%
Speech in noise (SPIN)[c]	not possible	not possible	+2.00 dB SNR	+0.67 dB SNR	−1.33 dB SNR
A§E® phoneme discrimination test	not possible	not possible	95%	100%	/
Sound localization	not possible	not possible	/	/	−30.0°

Hz,[a]The PTA value is determined by the measurements at 500 Hz, 1000 Hz, 2000 HzHz, and 4000 Hz. [b]SPIQ was measured using the Nederlandse Vereniging voor Audiologie (NVA) list at 65sixty-five dB SPL in the sound field. [c]SPIN was measured with the Leuven Intelligibility Sentence Test (LIST) using the adaptive procedure (speech adaptive and noise fixed at 65 dB SPL) inin the sound field.[5,6] CI, Cochlear implantation.

demonstrating an excellent short-term outcome with respect to speech perception in quiet and in noise, phoneme discrimination, and sound localization.

Patient consent for publication was obtained.

Discussion

The present case report discusses how a similar philosophy of structure preservation technique combined with image-based surgical planning can be used in the case of preferred scala vestibuli electrode position.

Many studies have been conducted on the comparability of these two methods, showing contradictory results. The postoperative consonant-nucleus-consonant (CNC) scores in the publication of O'Connell et al suggested that the usage of the scala tympani has better outcomes than the scala vestibule.[4] However, in 2018, Trudel et al described at least comparable auditory outcomes when comparing the scala vestibuli technique to the scala tympani technique. Their study assessed the largest cohort of patients available at that moment.[2] More recent, a retrospective study from Gu et al showed that there was no significant difference in auditory performance between the two groups 1 year after the procedure.[1]

In conclusion, high-resolution temporal bone CT can not only be used to determine the patency of the scala tympani, but also to assist in surgical planning with respect to the exact location of a minimally invasive cochleostomy to enter the scala vestibuli, and determine cochlear duct length. Based on literature and the present case report, equivalent auditory performance can be achieved with careful planning and management by an experienced CI team.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Informed consent was obtained from the patient who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

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Declaration of Interests: Oliver M. Vanderveken is the Editor in Chief at B-ENT, and Vincent Van Rompaey is an Associate Editor at B-ENT, however, their involvement in the peer-review process was solely as authors. The other authors have no conflict of interest to declare.

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