

## Abstract

**Introduction:** X-linked stapes gusher syndrome is a rare form of congenital sensorineural hearing loss (SNHL). Affected patients have abnormal configuration of the lamina cribrosa and internal auditory canal (IAC), leading to an increased risk of perilymph gusher with surgical manipulation. We report a case whereby intra-operative otoendoscopic visualization allowed real time visualization of the inner ear (IE) anatomy, which has the potential to impact on electrode choice in cochlear implants (CI).

**Case:** A 12-year-old boy has been followed up in our Pediatric Otolaryngology department with bilateral SNHL and X-linked stapes gusher syndrome for five years. He initially performed well with bilateral traditional hearing aids, but developed progressive mixed loss and worsened speech performance in the left ear, leading to the offer of cochlear implantation. The pre-operative computed tomography of the temporal bone showed the characteristic bulbous IAC with a dysplastic cochlea and no apparent modiolus. Intra-operative cochlear endoscopy however, showed an apparent modiolus without direct communication to the IAC.

**Discussion:** Electrode choice during CI in patients with IE malformation is based on the expected location of spiral ganglion cells. Cochlear endoscopy offers an additional tool for assessment of inner ear anatomy, which may assist in optimal electrode selection.

**Conclusion:** We advocate the use of intracochlear endoscopy in selected cases as it offers a better resolution than the high resolution CT and has the potential to allow customization of electrode choice based on membranous inner ear anatomy.

## Case

- A 12-year-old boy with X-linked stapes gusher syndrome initially performed well with bilateral traditional hearing aids, but developed progressive mixed loss and worsening performance of his left ear. His pre-operative audiogram is shown in figure 1.
- He was sent for CI candidacy assessment and was deemed to be suitable candidate for his left ear. The pre-operative CTTB showed a bulbous IAC with a dysplastic cochlea and no apparent modiolus characteristic of this condition (figure 2).
- A CI24RE (ST) implant was selected by the CI team due to uncertainty regarding the location of the spiral ganglion cells.
- The standard approach was utilized for his CI. A 1mm cochleostomy was made anteroinferior to the round window (RW). Upon entering the cochlea, a moderate perilymphatic gusher was encountered. A 1.3mm salivary endoscope was placed at the entrance of the cochlea to assess the intracochlear anatomy. There was a modiolus without direct communication to the IAC (figure 3).
- Full insertion was achieved on the first attempt without any difficulty. The gusher was controlled with packing of periosteal tissue at the cochleostomy site. An intra-operative plain X-ray confirmed the electrode placement.

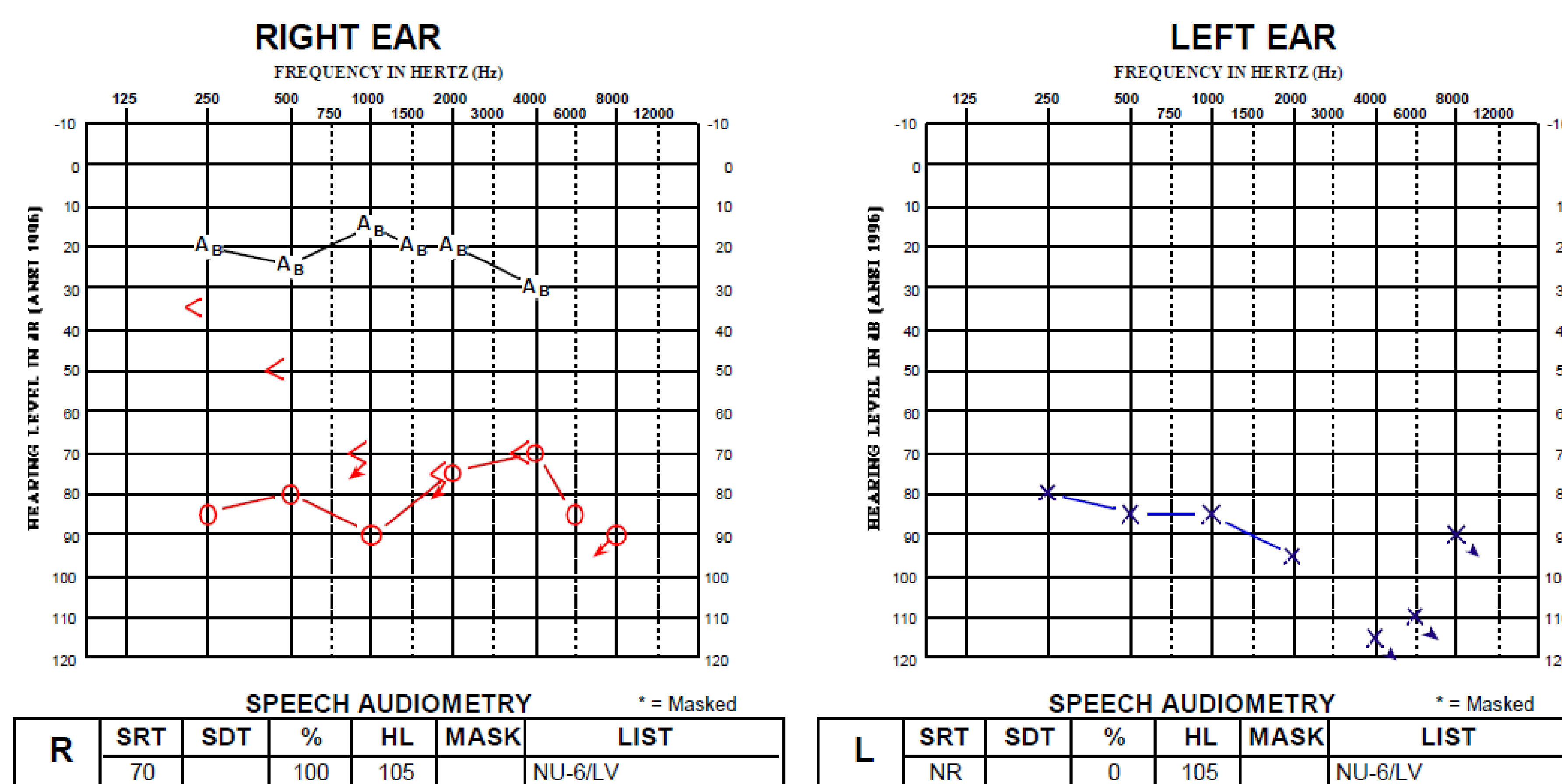


Figure 1: Pre-operative audiogram with speech testing



Figure 2: Dysplastic cochlea with no modiolus on CTTB as shown by the white arrow.

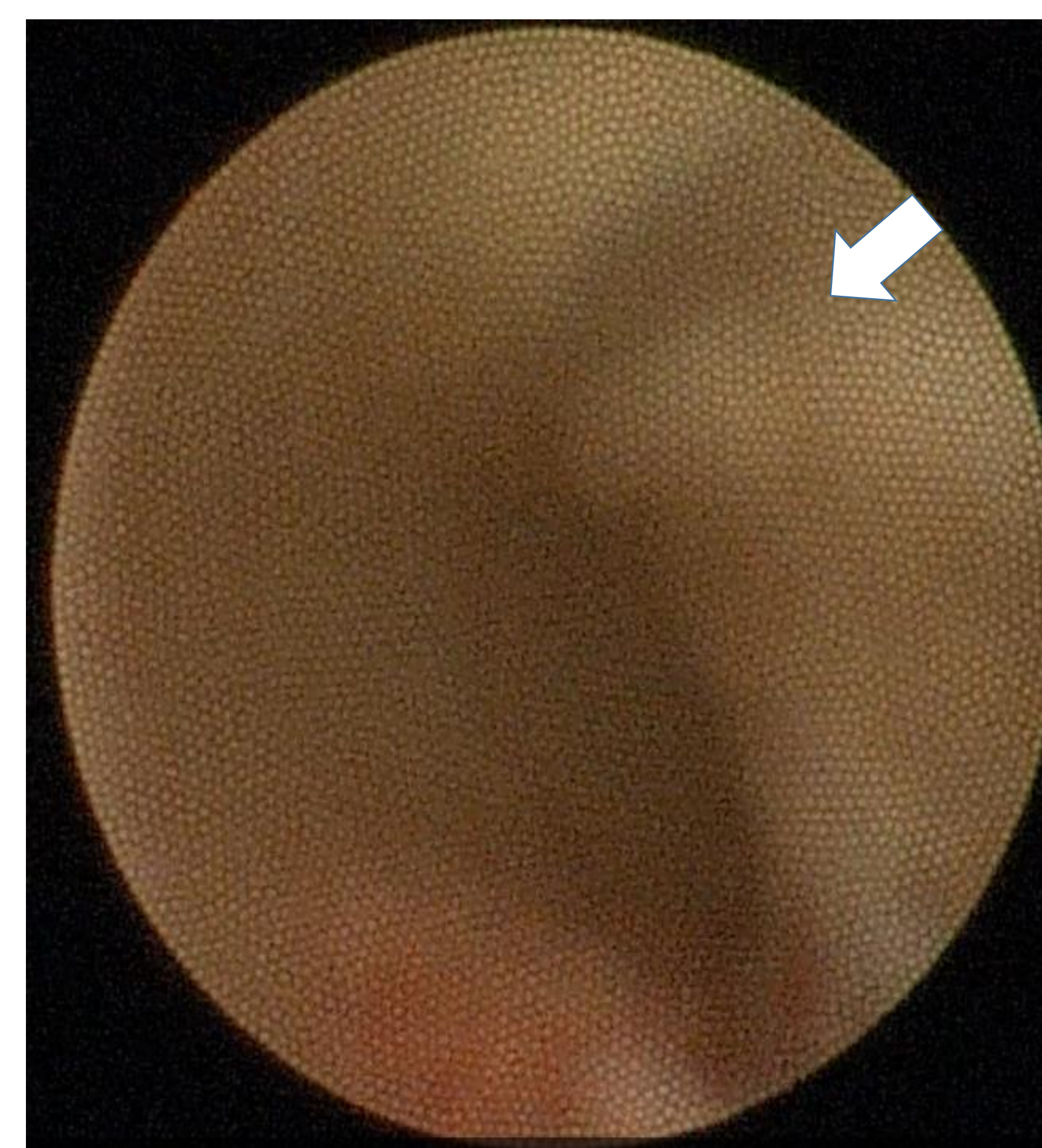


Figure 3: Intracochlear endoscopy of a left ear through a cochleostomy showing the presence of a modiolus (white arrow) with no obvious connection to IAC

## Discussion

- When the cochlear modiolus and osseous spiral lamina are deficient, this absent bony septum creates a common space seen on the imaging study [10] (figure 2). This abnormal communication between the IAC and vestibule is responsible for the perilymphatic gusher.
- Patients with IE anomalies may have both fewer and atypical positions of their spiral ganglion cells (SGC) [11]. This has functional implications because at least 10 000 functional SGNs are necessary for effective speech discrimination [12]. This number can be further reduced by surgical trauma [12].
- When there is uncertainty regarding the likely location of spiral ganglion cells, a fully banded CI electrode may be chosen to allow for full and multi-directional stimulation, whereas a directional electrode may be more appropriate if there is greater confidence in the location of spiral ganglion cells [13].
- The proper electrode choice may have important hearing outcome implications for patients with IE anomalies following CI due to challenges of reaching an optimal level of cochlear stimulation, decreased dynamic range, a wider pulse width, and weakened neural synchrony [9]. The functionality of CI is correlated to the number of SGCs and their distances from the stimulating electrode [14].
- While CTTB provides important information for surgical planning, the resolution is not currently sufficient to assess the fine detail of the structures of the inner ear.
- Our case demonstrates that intra-operative endoscopic findings may identify anatomic features not available on CT, indicating that cochlear endoscopy may be a useful tool to better delineate the intracochlear anatomy. The endoscopic intracochlear view gave us accurate and real-time information on the anatomy of the IE, which confirmed the presence of a modiolus.
- Although we did not alter electrode choice in this patient, the endoscopic findings suggest that a directional electrode may have been appropriate. This assumes that spiral ganglion cells are likely to be centrally located when membranous inner ear structures are preserved, as opposed to a peripheral location seen in common cavity malformations [10]. We suggest that cochlear endoscopy allows better delineation of the intracochlear anatomy, which may impact the electrode selection and potentially the hearing outcomes in patients with anomalous inner ear anatomy and uncertain location of spiral ganglion cells.

## Conclusions

- We advocate the use of intracochlear endoscopy in selected cases as it offers a better resolution of the intracochlear anatomy than high resolution CTTB.
- This has the potential to allow customization of electrode choice based on membranous inner ear anatomy, and potentially improve the hearing outcomes in patients with anomalous IE anatomy and uncertain location of spiral ganglion cells.

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