Cochlear Implantation in Syndromic Patients with Aural Atresia: Case Series

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Background

- Hearing loss (HL) in congenital aural atresia results from external and middle ear anomalies including canal atresia, ossicular fixation, and hypoplastic mastoid air cells[1].
- Though conductive loss predominates, progressive or late-onset mixed HL has been documented, requiring evaluation for possible cochlear implantation.
- Anatomic challenges include hypoplastic mastoid, inner ear malformations, and facial nerve anomalies (aberrant course, dehiscence, bifurcation) which drive surgical candidacy
- Few reports describe cochlear implantation in atresia and outcomes remain under-characterized.
- We present two cases of congenital aural atresia in patients with Treacher-Collins syndrome and Crouzon syndrome who underwent cochlear implantation.

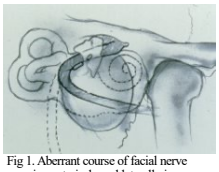

Fig 1. Aberrant course of facial nerve coursing anteriorly and laterally in congenital aural atresia obstructing access to round window for cochlear implantation

Case Description 1.

Case 1. 72 year old female with Treacher-Collins syndrome

HPI: A 72 year old female with Treacher-Collins syndrome presented to our institution with a decrease in speech understanding in both ears over the preceding several months. She has a history of bilateral external auditory canal (EAC) atresia and had been using a Cochlear™ 5 SP Bone conduction hearing aid (BAHA) headband attachment for several years.

She has a history of left meatoplasty performed at age 15. She denies otorrhea, facial weakness, or autophony. Her only other symptom is bilateral tinnitus which she has had at baseline and is unchanged.

Physical Exam:
AD: Involuted helix, EAC atresia
AS: Small cavity ending in a blind pouch without a tympanic membrane, a well-formed helix.

Pre-operative Audiogram:
Profound mixed hearing loss AU with maximal conductive components. Speech recognition in quiet on AzBio is 55% (Fig 3.)

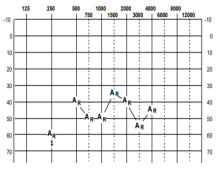

Fig 2. Baseline audiogram prior to development of mixed HL

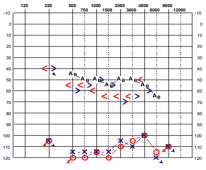

Fig 3. Audiogram with mixed HL

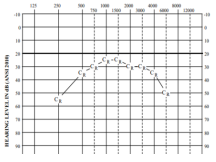

Fig 3. Audiogram following CI and programming

Case Description 1. Continued

Case 1. Continued

Imaging: CT Temporal Bone (Right)
The tympanic cavity and mastoid air cells are hypoplastic and there is non-specific soft tissue density within the epitympanum, hypotympanum, and extending along the ossicles into the oval window. The malleus is absent. The incus and stapes are present but malformed. The facial nerve runs in a more anterior and lateral course with no vertical segment, crossing deep to the TMJ. The IAC, cochlea, vestibular aqueduct, and semicircular canals are normal.

Operative Notes: Right cochlear implantation
- There was no mastoid pneumatization, EAC, or middle ear space.
- The lateral semicircular canal was identified and blue-lined.
- Facial nerve was found to be taking a far lateral and anterior course into the space just below glenoid fossa, obstructing access to round window.
- To avoid injury to the facial nerve, a retrofacial approach was taken

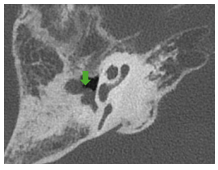

Fig 6. Pre-op CT right temporal bone

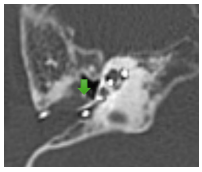

Fig 7. Post-op CT right temporal bone

Case Description 2.

Case 2. 18 year old M with Crouzon syndrome s/p bilateral CI

HPI: An 18 year old male with Crouzon syndrome presented to our institution with sudden onset decrease in hearing following a trip. He has a history of bilateral conductive hearing loss (CHL), bilateral EAC atresia, and bilateral microtia. He had been using a BAHA on the left for several years prior to this event and initially believed that the device had malfunctioned. He was treated with an oral steroid taper and sent in his BAHA to be fixed with no improvement in hearing.

History of bilateral partial pinna reconstruction at 7 and 9 years old.

Physical exam: Bilateral microtia and EAC atresia. Right > left partial pinna reconstruction.

Pre-operative Audiogram:
Rapidly progressive SNHL AU, stable CHL AU, resulting in profound mixed HL AU; prior testing revealed NI BC; 0% speech understanding at 55dB

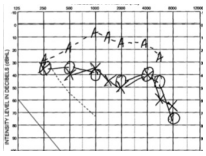

Fig 4. Baseline audiogram prior to development of mixed HL

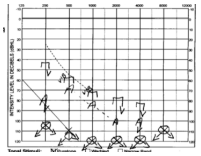

Fig 5. Audiogram with rapidly progressive mixed HL

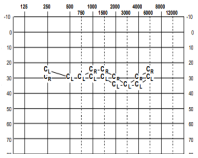

Fig 6. Audiogram following CI and programming

Case Description 2. Continued

Case 2. Continued

Imaging: CT Temporal Bone
The stapes and long process of the incus are normal bilaterally. The head of the malleus is dysmorphic bilaterally. The apical turn of the cochlea is somewhat globular and there is deficiency of the modiolus. The vestibular aqueducts are normal bilaterally. The superior and posterior semicircular canals are normally formed. The horizontal semicircular canals are shortened and widened bilaterally. There is improved aeration of the mastoid air cells bilaterally. The facial nerve demonstrates a typical intratemporal course.

Operative Notes 1. Right cochlear implantation
- A large horseshoe arcuate post-auricular incision with extension into the temporalis area and occipital was created.
- There was no ear canal and no normal landmarks.
- The tegmen, sigmoid sinus, and lateral semicircular canals were identified.
- The atretic plate was removed to gain access to the middle ear space.
- The facial nerve ran in a more typical course and the round window was approached from a more anterior-lateral direction.

Operative Notes 2. Left cochlear implantation
- An arcuate postauricular incision was made extending to temporalis area.
- The location of the mastoid space was identified using stereotactic image guidance.
- There was no sigmoid sinus. Instead, there was a large anomalous vein running superiorly along the mastoid.
- The lateral semicircular canal, facial recess, chorda tympani, stapes, and round window were identified.
- The facial nerve ran in a more typical course and the round window was approached from a more anterior-lateral direction following removal of atretic plate

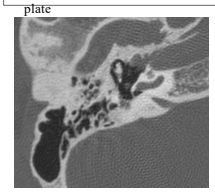

Fig 8. Pre-op CT right temporal bone

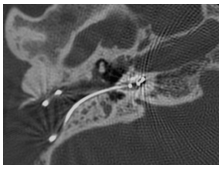

Figure 9. Post-op CT right temporal bone

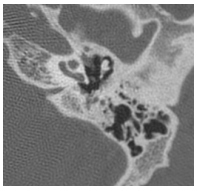

Fig 10. Pre-op CT left temporal bone

Discussion

- Cochlear implantation in syndromic aural atresia is feasible though requires flexible surgical planning due to variable anatomy.
- Because the facial nerve is often anteriorly displaced limiting a standard facial recess approach, surgeons should be prepared to perform a retrofacial approach to access the round window safely.
- Stereotactic image guidance or intraoperative navigation can improve safety when anatomic landmarks are obscured.
- These cases expand the scope of CI candidacy and underscore the importance of recognizing SNHL in syndromic aural atresia.
- Beyond intra-operative considerations, this study highlights the under recognition of the development of mixed hearing loss in syndromic aural atresia.
- Thus far, there have been six cases of isolated SNHL in Crouzon syndrome and 10% prevalence of mixed hearing loss in Treacher-Collins[2].
- No inner ear abnormalities have been described in either syndrome.

References
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Acknowledgements
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