Cochlear Implantation in a Child With Recurrent Meningitis Due to a Congenital Stapedial Footplate Defect

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A 9-month-old male with bilateral profound sensorineural hearing loss was referred to otolaryngology clinic for cochlear implantation (CI) evaluation. He had been using hearing aids since 5 months of age. There was no other known family history of hearing loss. The patient had a normal head and neck examination without syndromic features. Temporal bone computed tomography scan showed bilateral cochlear incomplete partition type I and enlarged vestibules (Figure 1). Magnetic resonance imaging (MRI) of the lateral internal auditory canals demonstrated 3 nerves on the right side and 4 nerves on the left side. Risks and benefits of a left CI were reviewed with the parents, and a higher risk of facial nerve aberration and cerebrospinal fluid (CSF) leak was reviewed. While undergoing CI evaluation, the patient had 2 hospitalizations for bacterial meningitis. After discharge, a repeat MRI was obtained which showed no postmeningitis cochlear ossification. Given the new history of recurrent meningitis in addition to the abnormal inner ear anatomy, bilateral external auditory canal (EAC) closures were planned along with the left CI to protect the patient from a higher risk of CSF leak.

The patient underwent the planned procedures. The right EAC was transected at the bony cartilaginous junction, and the skin cuff was everted and closed in multiple layers. The right middle ear was dry. The left EAC was closed in a similar fashion. When the middle ear was entered, significant...
inflammatory tissue was noted along with a large gush of CSF originating from a congenital defect in the left stapedial footplate (Figure 2). The stapes was removed and the oval window was packed with muscle. The eustachian tube was also packed with muscle. A cortical mastoidectomy was then performed and the facial recess was opened for CI placement. A straight complete banded electrode was placed into the round window and packed with muscle. Intraoperative testing of the CI showed responses from all electrodes. The patient had no additional episodes of meningitis at a 1-year follow-up.

Recurrent meningitis in children is a potentially life-threatening infection requiring a thorough search for the underlying mechanism.\(^1\) Over 30\% of the patients with meningitis and 90\% of those with recurrent meningitis were found to have otolaryngological etiologies.\(^2\) Congenital inner ear malformation is an uncommon yet known etiology of recurrent meningitis in children. There have been a few case reports describing CSF fistulas caused by various inner ear malformations including bony defects in the stapedial footplate, margin of the oval window, tegmen tympani, and roof of the eustachian tube.\(^3\)\(^-\)\(^5\) Our report further details the management option of performing EAC closure, CSF fistula repair, and CI placement concurrently.

The stapes footplate is embryologically derived from the otic capsule, and its defect is likely a congenital abnormality developed along with the other inner ear malformations. Increased CSF pressure to the medial surface of the oval window common among patients with inner ear malformations may also have contributed to the development of the CSF fistula in the stapedial footplate.\(^6\) Small defects in the stapes are likely to be missed with available imaging modalities. Exploratory tympanotomy is often necessary to diagnose of CSF leak in patients with a high suspicion for inner ear malformation causing recurrent meningitis. If a CSF leak is found at the oval window, the stapes should be removed and the vestibule needs to be packed firmly with temporalis fascia or muscles. A CI can safely be implanted simultaneously to restore hearing.

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