Published in final edited form as:

Otol Neurotol. 2014 June ; 35(5): e199–e201. doi:10.1097/MAO.0000000000000274.

Bifid facial nerve in congenital stapes footplate fixation

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Keywords

Congenital stapes; footplate fixation; stapedectomy; facial nerve; bifid facial nerve; congenital otosclerosis

A 7-year-old male with a history of submucous cleft palate, congenital heart disease, and torticollis presented with since-birth right-sided hearing loss. The patient had no previous otologic history other than pressure equalization tube placement at 9 months of age. Audiometric evaluation revealed normal hearing thresholds on the left with a maximal conductive hearing loss on the right side without a distinct Carhart notch (Fig. 1A). Fine cut computed tomography (CT) of the temporal bones was obtained demonstrating bilateral well-aerated mastoid and middle ear cavities with normal inner ear morphology; specifically, the right cochlea was not dysplastic and the vestibular aqueduct was normal in size. The right ear demonstrated a malformed long process of the incus, a thinned stapes suprastructure and a thickened footplate. The labyrinthine and mastoid segments of the facial nerve appeared within normal limits, while the tympanic portion was poorly defined but appeared to straddle the oval window (Fig. 2 & 3). The contralateral middle ear was radiographically unremarkable.

After informed consent, the patient and family elected to proceed with middle ear exploration and possible stapedotomy. The tympanomeatal flap was elevated and palpation of the malleus and incus revealed normal mobility, however the incus and stapes were severely malformed. The long process of the incus was foreshortened and slightly medially rotated with a malformed lenticular process and the stapes crurae were merely fibrous bands connected to a thick, immobile footplate. The tympanic segment of the facial nerve was noted to be distinctly bifid, coursing around the footplate superiorly and inferiorly (Fig 4). Facial nerve stimulation confirmed an anomalous split facial nerve configuration. A stapedotomy was carefully performed using a microdrill until the footplate was “blue lined” and a CO2 laser with micropick was used to open the vestibule. A 4-mm self-crimping Nitinol stapes prosthesis was placed without difficulty and the patient awoke from

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CONFLICT(S) OF INTEREST TO DECLARE: There are no relevant conflicts of interest to disclose

The following study has not been presented or published previously and is not currently under consideration elsewhere.

anesthesia with normal facial nerve function and Weber examination lateralized to the operative ear. Three months following surgery the patient was noted to have persistent bilateral serous effusions and progressive anterior pars tensa retraction requiring tympanostomy tube placement. Follow-up audiometry was performed approximately 4 months following surgery and demonstrated a 52 dB improvement in pure-tone average with a residual 17dB air-bone gap; bone conduction thresholds remained unchanged (Fig 1B).

Stapes footplate fixation is known to be the most prevalent congenital ossicular abnormality.³ The common embryologic origin, shared by the facial nerve, long process of the incus, stapes suprastructure and outer table of the footplate helps explain the constellation of findings within the present case. While rare, an aberrant facial nerve course is more common among patients with external auditory canal atresia or dysplastic inner ears. While even less common, an anomalous tympanic course can also be seen in patients with congenital footplate fixation, having normal external and inner ears.^{3,4}

In contrast to juvenile otosclerosis, congenital footplate fixation presents with a life-long, non-progressive conductive hearing loss. While audiometric findings are somewhat variable, most patients are diagnosed with advanced conductive hearing loss, often without a distinct Carhart notch. All pediatric patients undergoing stapedectomy should be evaluated with preoperative CT to rule out inner ear malformations and the associated risk of gusher. The above case demonstrates the importance of understanding and applying embryology during otologic surgery. When a surgeon encounters an anatomic anomaly intraoperatively, they must carefully inspect other structures with similar embryologic origins to avoid misadventure.

Acknowledgments

FINANCIAL MATERIAL & SUPPORT: No funding or other support was required for this study

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Figure 1.

A, Pre-operative audiogram revealing a maximal conductive loss without a Cahart notch. B, Post-operative audiogram demonstrating a 52 dB pure-tone average gain with a residual 17 dB air-bone gap and unchanged bone conduction thresholds.

Figure 2.

High-resolution axial and coronal CT of a right temporal bone with congenital footplate fixation and a bifid tympanic facial nerve segment. A, Axial CT demonstrating a fibrous malformed stapes suprastructure (white arrow) and a thickened non-distinct footplate. B, Coronal CT demonstrating the inferior limb of the bifid facial nerve next to the stapedial tendon (white arrow).

Figure 3.

For illustration purposes, a preoperative CT of a normal temporal bone and the CT of the index patient were segmented using automated software in order to create 3-dimensional renderings of the fallopian canal (magenta), ossicles (blue) and inner ear (tan). Figure A & B demonstrate the expected course of the facial nerve, superior to the oval window niche. In contrast, Figures C & D reveal the anomalous course of the facial nerve, with the stapes suprastructure and footplate situated between the tympanic facial nerve segments (black arrows). The preoperative CT rendering does not clearly distinguish the bifurcation, but rather demonstrates the footplate resting in the middle of the tympanic facial nerve segment.

Figure 4.

A, Endoscopic view of the anomalous facial nerve prior to removing the stapes suprastructure. B–D With the stapes suprastructure removed, the bifid facial nerve can be clearly seen coursing superior and inferior to the stapes footplate. (FN=facial nerve; In=incus; ST=stapedial tendon; RWN=round window niche; FP=footplate; CT=chorda tympani)