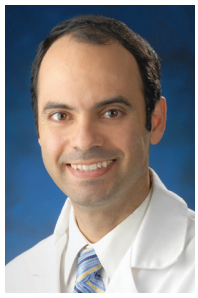

CLINICAL CONSULTATION

# Symptom: Conductive Hearing Loss

By Hamid R. Djalilian, MD

A 28-year-old male, who has been a lifelong hearing aid user, comes in for a hearing evaluation. He states that he was born with the hearing loss but is unsure of the cause. His birth history is normal and he denies any problems during his mother's pregnancy. There is no family history of hearing loss. Examination of the ears reveals a somewhat narrow ear canal bilaterally. On the right side, the middle ear contains an effusion. On the left side, the middle ear appears well aerated. The patient wants to explore surgical options for treatment of his hearing loss. On the right is his audiogram.

**What is your diagnosis? See page 44.**

Read past Clinical Consultation columns in a special collection on the *HJ* website: **bit.ly/HJClinicalConsultation**.

**Dr. Djalilian** is director of neurotology and skull base surgery and professor of otolaryngology and biomedical engineering at the University of California, Irvine.

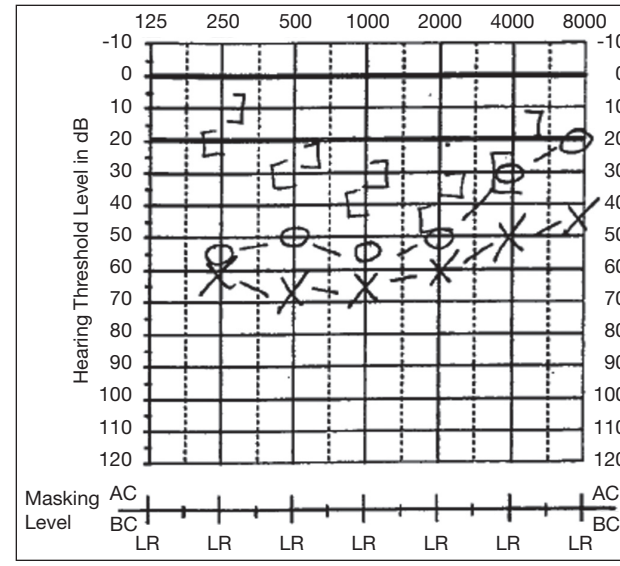


**Figure 1. Image of the patient's audiogram.**

# Diagnosis: First Branchial Cleft Anomaly (Type 1)

By Hamid R. Djalilian, MD

Continued from p. 42

Congenital conductive hearing loss is most commonly due to middle ear effusion. However, we generally would not expect the effusion to last for many years. It would be uncommon for middle ear effusion to last beyond the age of 6 months. In some cases, the middle ear mesenchyme, which surrounds the ossicles during the embryonic stage, may not fully resolve by the time of birth. This especially happens in children who are born prematurely. However, the middle ear mesenchyme is generally fully resolved by the time the infant is 6 months of age. Therefore, patients with congenital conductive hearing loss that has been non-fluctuating for many years may suffer from ossicular fixation (Maducdoc. *Int J Pediatr Otorhinolaryngol* 2015;79[12]:2277).

In the presence of congenital conductive hearing loss, a CT scan of the temporal bones is obtained when surgical treatment is contemplative. This patient's CT scan can be seen in figures 2 and 3. There appears to be traces of a previous mastoidectomy. However, this patient denies having had previous surgery. This led us to consider a congenital cholesteatoma of the mastoid as the potential cause of the mastoid defect (Thakkar. *Otol Neurotol* 2006;27[2]:282). Because of the unclear cause of the defect, an MRI was obtained to better evaluate the soft tissue contained within the area. The MRI shows the soft tissue detail, whereas the CT of the temporal bone shows the bony detail better. The MRI did not reveal cholesteatoma to be the cause of the mastoid defect. Surgical intervention was undertaken in order to better evaluate the cause of the hearing loss and abnormalities seen on CT scan.

**Patients with congenital conductive hearing loss that has been non-fluctuating for many years may suffer from ossicular fixation.**

In surgery, we found the area of the mastoid defect to be a duplication of the cartilaginous and bony external auditory canal. An instrument could pass through the duplicated canal into the middle ear, which coursed behind the normal bony ear canal that was connected to the auricle. The patient was clearly suffering from a type 1 first branchial cleft anomaly.

First branchial cleft anomalies are rare congenital malformations of the head and neck, comprising less than 10 percent of all branchial anomalies. A first branchial cleft anomaly commonly presents with a cyst or draining fistula in front or behind the auricle below the level of the ear canal.

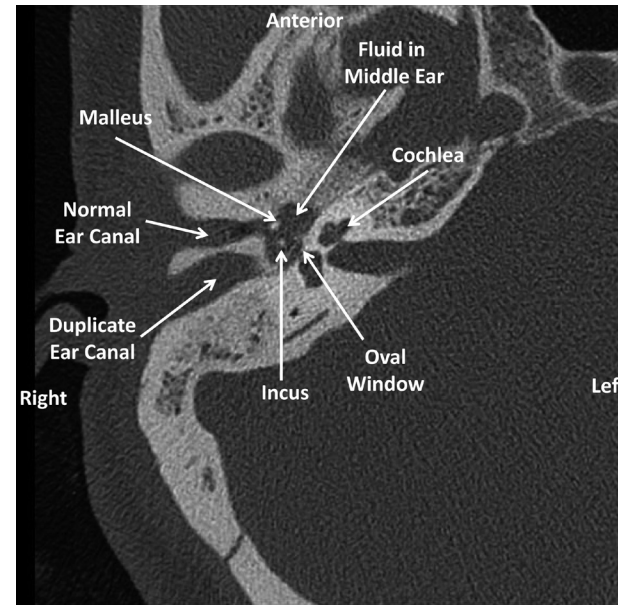


**Figure 2. Axial CT image of the right temporal bones at the level of the cochlea demonstrating duplicate ear canal with the bony wall separating the two canals.**

First branchial cleft anomalies are divided into two types. Type I anomalies most commonly present as a cystic mass that is skin lined and may or may not contain accessory skin structures such as hair, sweat glands, etc. Type I lesions occur medial to the auricle and can extend into the postauricular crease. These typically end at the bony plate at the level of the middle ear. Type II anomalies present as a cyst or fistula (an opening in the skin that intermittently drains). Type II lesions contain skin, accessory skin structures, and often cartilage. These may pass through the parotid salivary gland (located in front and below the ear) and can have a variable relationship to the facial nerve. These lesions may travel laterally, medially, or between the branches of the facial nerve, which makes their removal challenging.

Physical examination of the external auditory canal may show an opening into the ear canal on the floor of the canal. Though uncommon, a membranous attachment between the floor of the external ear canal and the tympanic membrane may be found as well. These webs can help with the diagnosis of the type II first branchial abnormality. When an anomaly is suspected, MRI of the area can help delineate the type of anomaly and the relationship to the parotid gland. The facial

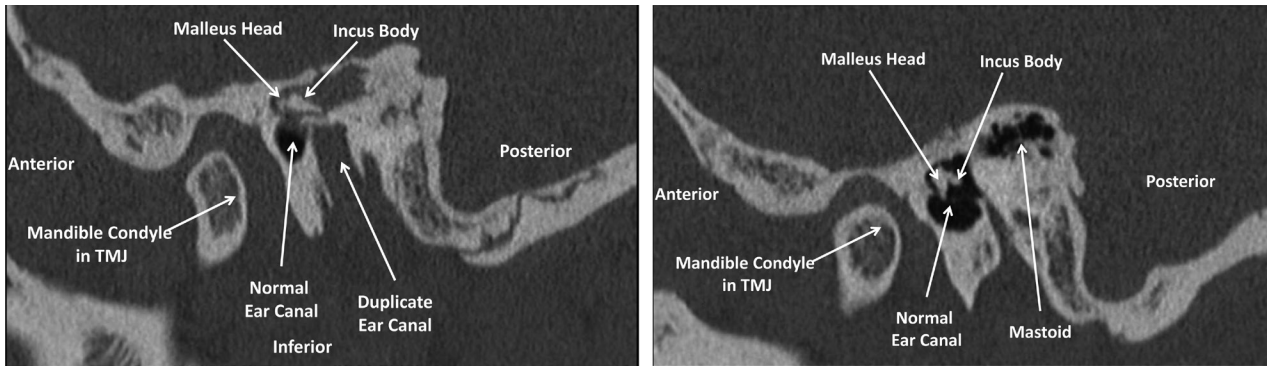


**Figure 3. (Left) Sagittal CT image of the right temporal bone showing the separation between the two canals at the level of the annulus showing the ossicles. (Right) Sagittal image of the left temporal bone shows the normal anatomy.**

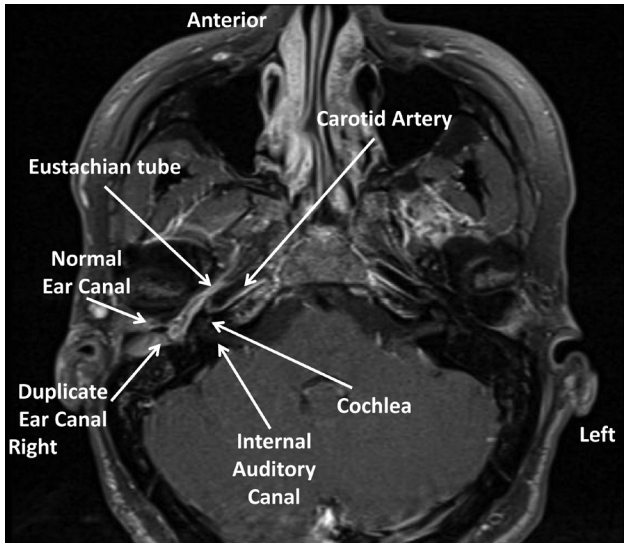


**Figure 4. Axial T1 post gadolinium image at the level of the cochlea showing the inflamed mucosa of the middle ear and Eustachian tube.**

nerve in the parotid gland is difficult to see on an MRI but an approximate location can be surmised. A CT of the parotid area is unable to show the level of soft tissue detail needed to visualize these abnormalities.

In this patient, evaluation of the middle ear revealed that there were no abnormalities of the ossicles. The presence of mucoid effusion and inflamed middle ear mucosa were the likely cause of hearing loss. Examination of the CT before surgery revealed that the mastoid was not well developed, which indicates poor eustachian tube function since childhood. The placement of a pressure equalization tube allowed the aeration of the middle ear and the normalization of hearing.

We did not find a connection between the normal ear canal and the duplicated one in this patient. Instead of excising the entire duplicated canal that was lined by skin, we connected the two canals and took down the wall between the two. This allowed for enlargement of the ear canal as the narrow canal caused problems with recurrent cerumen impaction. Dissection of the duplicated canal into the parotid gland was unnecessary, as this was a type I anomaly, which is generally a duplication of the canal.