**ABSTRACT**

Anomalies of the course of the facial nerve have been reported in association with middle and inner ear malformations. A rare facial nerve anomaly was incidentally discovered during the workup of a congenital conductive hearing loss in a pediatric patient associated with unilateral agenesis of the oval window. The bilateral labyrinthine segments of the facial nerve were noted to bifurcate just proximal to the geniculate ganglion bilaterally.

Bifurcation of the intratemporal portion is a rare malformation in which focal splitting of one or more facial nerve segments occurs. We describe the CT appearance of this anomaly and discuss its possible embryology.

This case highlights the importance of vigilance regarding facial nerve anatomical variations encountered during middle-ear surgery thus avoiding inadvertent damage.

**CONCLUSIONS:**

Congenital hearing loss may be associated with facial nerve anomalies. Facial nerve bifurcation is important to recognize in patients undergoing evaluation for congenital hearing loss and other congenital ear malformations.

Otolaryngologists should be cautious when exploring patients with conductive congenital hearing loss.

---

**CASE REPORT**

The patient was an eight year old boy who presented to the tertiary referral otologic practice for evaluation of a nonprogressive left conductive hearing loss present since birth.

He was noted to have other skeletal deformities including hypoplasia of the right hand, skeletal abnormality of the right foot, chronic dysphagia, and congenital torticollis.

A complete audiometric and otolaryngologic examination was performed. Otolologic examination was noted to be within normal limits. Tymanograms were normal. Hearing in the right ear was normal. The left ear revealed a maximal conductive loss.

A temporal bone CT was obtained and was interpreted as normal by the radiologist.

Independent review of the temporal bone CT was performed. The left ear was noted to have agenesia of the oval window and displacement of the tympanic segment of the facial nerve into the location of the absent oval window. In addition, a bifid labyrinthine segment of the facial nerve was noted. Two labyrinthine facial nerve canals of identical size were noted as well as a normal internal auditory canal on the left.

CT examination of the right ear was notable for a bifid course of the labyrinthine segment of the facial nerve.

After discussion of findings, recommendation for amplification or bone anchored hearing aid placement was made.

---

**DISCUSSION**

The exquisite detail afforded by a dedicated CT scan of the temporal bone allows evaluation of osseous abnormalities that may have previously gone unnoticed.

Many abnormalities of the facial nerve canal in the petrous temporal bone have been documented.

Anomalies of the facial nerve canal are infrequently found in normal temporal bones and are usually seen in association with middle and inner ear dysplasias.

Bifurcation and trifurcation facial nerve anomalies have been previously described in the otolaryngology literature. These have been reported to involve all portions of the nerve from the intracanalicular segment to the mastoid segment, with the most common anomalies occurring along the tympanic segment.

Bifurcation of the labyrinthine facial nerve segment appears to be the most rare of the intratemporal anomalies.

Although the dual labyrinthine canals possibly represented one for the facial nerve and one for the nerve intermedius, the latter nerve is significantly smaller than the facial nerve, yet the canals were of symmetric diameter.

The embryologic origin of facial nerve bifurcation anomalies is uncertain, because the facial nerve never exists as separate bundles during its development.

Formation of the nerve commences early in gestation with the facioacoustic primordium (derived from the neural crest and otic vesicle) separating into facial and acoustic components at the end of the 4th week. By the end of the 5th gestational week, the chorda tympani has differentiated from the distal facial nerve. By the 8th week, the orientation of the facial nerve within the temporal bone has been established, with the nerve’s ultimate position and bony covering determined by development of the stapes and membranous labyrinth.

The basic configuration of the facial nerve is completed at around 6 weeks, whereas the mesenchymal tissue through which it passes and the structures it innervates are still poorly formed. Abnormal separation of the facial nerve much earlier in embryogenesis may be necessary for a bifurcation or trifurcation anomaly to result. Early division of the nerve at around 4–6 weeks may allow the displacement of one segment by the developing temporal bone structures, such as pulling it anteriorly or laterally.

Facial canal ossification commences near the end of the 5th month in utero from second branchial arch cartilage (Reichert’s cartilage), but the mastoid segment canal largely forms postnatally with growth of the mastoid bone.

Although the case presented did not have surgical or pathologic confirmation, the images are highly suggestive of the bifurcation anomalies described in the surgical literature. The presence of an anomalous facial nerve component over the window may limit or preclude surgical access for stapedectomy, and an anomalous facial nerve segment through the mastoid bone may be at risk with mastoidectomy or cochlear implant placement.

---

**REFERENCES**