ABSTRACT
Objective: Ossicular developmental anomalies related to the miscommunication between second branchial arch and otic capsule derivatives are relatively uncommon as a unilateral finding in patients without associated craniofacial developmental abnormalities. Specifically, the relative position of the developing tympanic facial nerve (fallopian canal) to that of the stapes anlage and oval window niche may lead to ossicular anomalies responsible for a conductive hearing loss.

Materials and Methods: This is a retrospective case series review. The clinical history, audiometric data, and intraoperative findings are discussed for the selected cases.

Results: The stapes was found to be mobile in each of five patients suspected of having a conductive or mixed hearing loss due to otosclerosis. A dehiscence, inferiorly malformed, tympanic segment of facial nerve was found to be impinging on the stapes crura in all cases and to be associated with a malformed stapes crura in one case. No further middle ear surgery was performed in any of the cases and conventional amplification was recommended postoperatively.

Conclusion: Although pre-operative imaging studies and the audiometric configuration of the mixed hearing loss in our patients suggested a possible diagnosis other than otosclerosis, exploratory tympanotomy was necessary in order to make a definitive diagnosis.

BACKGROUND
• Congenital dehiscence of the facial (Fallopian) canal is common. It has been described in 57% of temporal bone specimens in a large anatomic series. 1
• Dehiscence occurs predominantly along the tympanic segment (83%), adjacent to the oval window niche. The facial nerve may be found protruding outside the Fallopian canal in up to 23% of cases. 2
• Ten to 15 percent of these dehiscences were considered clinically significant in terms of the presence of the nerve in the surgical field during middle ear exploration. 1
• Various clinical series on stapes surgery estimate the incidence of facial canal dehiscence to be between 3% and 11.4% and for nerve prolapse to occur in 7% .2, 2,3
• Facial nerve anomalies are commonly associated with congenital anomalies of structures derived from the first and second branchial arch and otic capsule and may be expected in conditions such as aural atresia and Treacher-Collins syndrome, among others.
• The most consistent audiologic characteristic in patients with an anomalous facial nerve is congenital conductive hearing loss.
• The degree of hearing loss, however, is often associated with a concurrent ossicular malformation and not, necessarily, due to the nerve’s effect on sound conduction.
• We present five cases of unilateral conductive hearing loss thought to be due, primarily, to a dehiscent tympanic segment of the facial nerve causing impingement and/or abnormal development of the stapes crura.

PATIENTS AND METHODS
• Retrospective case review of clinical and operative records for patients who had middle ear exploration between 2002 and 2010 and found to have facial nerve dehiscence with impingement on the stapes crura.
• Average age at surgery was 30.6 years (range, 15-55 yrs.).
• Four patients had primary surgical procedures.
• All but one patient had a suspected diagnosis of otosclerosis.

RESULTS
• All but two of the patients (patients 3 and 5) complained of gradual worsening hearing loss of adult onset.
• None of the patients had obvious syndromic features of the head and neck.
• Patient 3 had a previous history of mesotympanic cholesteatoma and had a tympanoplasty procedure two years prior. Patient 5 had hearing loss recognized during childhood.
• Table 1 summarizes the audiometric and intraoperative findings.
• None of the patients who had primary middle ear exploration had temporal bone CT scan findings consistent with otosclerosis.
• High-resolution temporal bone CT for patient 5 showed a soft tissue density at the level of the oval window (Figure 1), suggesting a dehiscent mal-positioned facial nerve or a middle ear neoplasm.

Table 1. Summary of findings.

![Image](image1.png)

Table 1. Summary of findings.

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Hearing loss</th>
<th>audiometric findings</th>
<th>operative findings</th>
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<td>25</td>
<td>M</td>
<td>nerve source OSL type A</td>
<td>SS and STAI</td>
<td>removed dehiscent facial nerve impinging on stapes crura, 7 dB hearing loss for stapes crura</td>
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<td>30</td>
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<td>SS and STAI</td>
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<tr>
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<td>SS and STAI</td>
<td>removed dehiscent facial nerve impinging on stapes crura</td>
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</table>

DISCUSSION
• The presenting complaints, audiometric results, and intra-operative findings suggest that a dehiscent tympanic segment of the facial (Fallopian) canal with nerve prolapse was a significant factor in causing conductive hearing loss in this group of patients.
• Facial canal dehiscence with nerve prolapse is best understood in the context of the embryology of the facial canal.
• By 5-6 weeks of gestation, the horizontal and vertical segments of the facial nerve appear.
• It is not until 10 weeks of gestation that the facial canal begins to form around the facial nerve primordium.
• The facial canal develops as a result of the fusion between the cartilaginous primordium otic capsule and Reichert’s cartilage (second branchial arch derivative).
• Complete ossification of the canal is not complete until about 1 year postnatally. 4
• Congenital dehiscence of the facial canal may result from improper communication between the derivatives of the second branchial arch and the otic capsule.
• Failure or delayed union of the stapes crura (second branchial arch derivative) and the lamina stapediales (otic capsule derivative) can either contribute to, or be affected by, an abnormally coursing facial nerve. 5, 6
• Conductive hearing loss due to an abnormally coursing facial nerve, in the absence of a congenital ossicular malformation and/or inherent syndromic features, is a rare phenomenon, reported in less than 60 cases in the literature. 7, 8
• Clinical presentation can be similar to that of otosclerosis or congenital cholesteatoma.
• Patients may have no significant history of ear disease or trauma and demonstrate no evidence of abnormalities on clinical exam or imaging studies.

CONCLUSIONS
• Dehiscence of the tympanic segment of the facial canal is a common occurrence and should always be a consideration during safe middle ear surgery.
• The combination of facial canal dehiscence with facial nerve prolapse and impingement on the stapes crura is an, otherwise, rare finding during middle ear exploration for conductive hearing loss.
• Clinical examination, audiometry, and radiographic studies may suggest the possibility of this occurrence, however, surgical exploration is typically required to make the diagnosis.

REFERENCES