Congenital Absence of the Incudostapedial Joint

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Abstract

Educational Objective:
Discuss features of the congenitally absent incudostapedial joint, its surgical management, and its embryology.

Objectives:
1) Describe features of the congenitally absent incudostapedial joint
2) Describe the surgical management of the congenitally absent incudostapedial joint with ossicular chain reconstruction and audiologic outcomes
3) Describe possible embryological etiology for the congenitally absent incudostapedial joint

Study Design Case series.

Methods: Retrospective review of 4 patients with isolated congenital absence of the incudostapedial joint treated between 2007 and 2012 by a single neurotologist. Temporal bone computed tomography images were reviewed. Preoperative and postoperative audiograms were compared.

Results: Patients underwent partial or total ossicular chain reconstruction and demonstrated significant improvement in hearing. No complications were observed postoperatively.

Conclusions: To date, few cases with this condition have been reported. This study may indicate a common embryologic origin for the incus long process and stapes capitulum. Based on extent of defect, partial or total ossicular chain reconstruction may significantly improve hearing.

Introduction

• Major congenital otologic malformations have been associated with an external ear abnormality
• Minor congenital otologic malformations are limited to the middle ear only (i.e. stapes, incus, malleus, oval window, round window)
• Non-progressive, conductive hearing loss in the setting of a normal tympanic membrane, without a history of trauma or infection, is highly suspicious for congenital ossicular anomaly
• Conductive hearing loss due to congenital ossicular malformation is rare, occurring in less than 1 in 15,000 births
• The incudostapedial joint (ISJ) is a diarthrotic, sphenoidal joint comprised of the long and lenticular processes of the incus, capitulum and neck of the stapes, and stapedius muscle & tendon
• There are a limited number of cases identifying ISJ anomalies
• In this case series we report 4 cases of congenital ISJ anomalies

Method

• Retrospective chart review at a university medical center from January 2007 to March 2012
• Preoperative and postoperative audiograms were assessed based on the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head & Neck Surgery guidelines for the evaluation of results of treatment of conductive hearing loss
• Successful treatment was defined when both of the following criteria were satisfied:
  o Air-bone gap reduced to 20 dB or less
  o Postoperative hearing gain of at least 15dB

Discussion

• The incus, stapes superstructure, and the ISJ are derivatives of the second branchial arch
• The stapes foot plate is a derivative of the lamina stapedialis, an otic capsule precursor. The stapes requires the longest duration of development, and therefore, has the greatest likelihood for malformation
• Isolated congenital anomalies of the stapes represent approximately 40% of congenital ossicular lesions
• Due to the common embryologic origin of the stapes superstructure and incus, it is not surprising that approximately one third of stapes anomalies are accompanied by incus anomalies
• Teunissen and Cremers (1993) classified congenital anomalies between four classes. Class I with stapes ankylosis and Class II with stapes ankylosis with an associated ossicular anomaly demonstrated favorable surgical outcomes with a 73% rate of post-operative air-bone gap less than 20 dB
• Conversely, Class III anomalies with mobile foot plate and ossicular discontinuity, and Class IV with dysplasia of the round or oval window have less favorable surgical outcomes.
• All patients in this study cohort demonstrated ossicular discontinuity (Class III ossicular anomaly)
• 2 patients (JJ and MM) who underwent TORP and PORP respectively had successful results with an Air-bone gap was reduced to less than 20 dB
• Patient SC who underwent an Applebaum ISJ prosthesis first followed by revision incus interposition had sub-optimal outcome. A PORP would have been more suitable

Bibliography


Figure 1: 4 patients with ISJ anomalies with audiologic findings, surgical findings, and treatments (ABG, air-bone gap)

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Baseline ABG</th>
<th>Post-Op ABG</th>
<th>ABG difference</th>
<th>Surgical Findings</th>
<th>Treatment</th>
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<tr>
<td>JJ</td>
<td>F</td>
<td>10</td>
<td>39 dB</td>
<td>14 dB</td>
<td>25 dB (64%)</td>
<td>Short Incus long process &amp; No Stapes Capitulum</td>
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<tr>
<td>MM</td>
<td>F</td>
<td>17</td>
<td>49 dB</td>
<td>18 dB</td>
<td>31 dB (63%)</td>
<td>Incus lenticular process &amp; Stapes capitulum suspended by fibrous adhesions</td>
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<tr>
<td>SC</td>
<td>F</td>
<td>14</td>
<td>63 dB</td>
<td>35 dB</td>
<td>28 dB (44%)</td>
<td>Short Incus long process &amp; No Stapes Capitulum</td>
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<tr>
<td>CA</td>
<td>M</td>
<td>6</td>
<td>41 dB</td>
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Figure 2: Air bone gap data obtained at 0.5,1,3, and 3kHz.

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<th>SD</th>
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<tr>
<td>Post-Operative</td>
<td>22 dB</td>
<td>18 dB</td>
<td>31 dB</td>
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<tr>
<td>Change dB</td>
<td>28 dB</td>
<td>3 dB</td>
<td>25-31 dB</td>
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Figure 3: Operative findings of MM: Incus lenticular process and stapes capitulum suspended by fibrous adhesions with no incus or stapes foot plate

Figure 4: Operative findings in JJ and SC: Short Incus long process and no stapes capitulum.

Figure 5: Patient CA CT imaging demonstrating abnormality of the ISJ with deficient incus long process and no stapes capitulum.