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Congenital Stapes Ankylosis in Children: Surgical Findings and Results in 35 Cases


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Objective: To evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital stapes ankylosis with or without other ossicular malformations (Teunissen and Cremers class I and class II malformations).

Study Design: A nonrandomized, nonblinded case series of prospectively collected data.

Setting: A tertiary referral center.

Patients: Twenty-eight consecutive pediatric patients who underwent 35 surgical procedures for congenital stapes ankylosis with or without other ossicular malformations and had available postoperative pure-tone audiometry.

Intervention: Primary stapedotomy with vein graft interposition and reconstruction with a Teflon piston, bucket handle prosthesis or total ossicular replacement prosthesis.

Main Outcome Measures: Pre- and postoperative audiometric evaluation using four-frequency (0.5, 1, 2, and 4kHz) audiometry. Air-conduction thresholds, bone-conduction thresholds, and air-bone gaps (ABGs) were measured. Postoperative audiometry was performed at 3, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter.

Results: Overall, a postoperative ABG closure of 10 dB or less was achieved in 73% of class I cases and in 50% of class II cases. A postoperative ABG closure of 20 dB or less was achieved in 77% of class I cases and 67% of class II cases. Postoperative sensorineural hearing loss occurred in one class I case (4%) and none of the class II cases.

Conclusion: Stapedotomy is a safe and feasible treatment option in children with congenital stapes ankylosis.

Key Words: Computed tomography imaging—Magnetic resonance imaging—Thin and dehiscent superior semicircular canals.


Conductive hearing loss is rarely caused by congenital middle ear malformations. Reported incidence varies between 0.5 and 3% in patients undergoing surgical treatment for conductive hearing loss (1,2). Congenital middle ear malformations are classified as major when they are accompanied by tympanic membrane or external ear malformations. Minor congenital middle ear malformations are limited to the middle ear and can be classified into four main groups according to the Teunissen and Cremers classification (3): (class I) isolated congenital stapes ankylosis, (class II) stapes ankylosis associated with other ossicular malformations, (class III) congenital malformations of the ossicular chain with a mobile stapes footplate, and (class IV) severe (IVA) aplasia or (IVb) dysplasia of the oval or round window. The majority of minor congenital middle ear malformations are classified as class I (31%) or class II (38%) malformations (3).

In the past, stapes surgery in case of congenital middle ear malformations has been thought to be associated with increased risks of perilymphatic gusher and sensorineural hearing loss (SNHL) (4,5). Meanwhile, numerous authors have published the results of case series of patients undergoing stapes surgery for congenital middle ear malformations with satisfactory results (2,3,6—10).

The objective of this study was to evaluate surgical findings and hearing results in children undergoing middle ear surgery for congenital stapes ankylosis with or without other ossicular malformations (class I and class II malformations).

METHODS

Between January 1991 and October 2014, 225 operations for congenital malformations of the middle ear were performed by the same surgeon (first author) in the same tertiary referral center. Of these 225 operations, 69 were performed in children and congenital stapes ankylosis was identified in 35 of these cases. The remaining 34 cases underwent surgery for class III malformations.
and class IV congenital malformations and were excluded from further analyses.

**Study Population**

This is a nonrandomized, nonblinded study of prospectively collected data of 28 children, younger than 18 years of age, who underwent 35 consecutive operations for class I and class II congenital middle ear malformations from 1991 to 2014. Whenever we refer to cases in this article, we refer to operations and not patients. Of the 28 included children, seven had sequential bilateral middle ear surgery. Postoperative pure-tone audiometry was available for all of these patients. No patients were excluded because they lacked postoperative pure-tone audiometric data. None of the included children were diagnosed with a concomitant syndrome. The presence of concomitant syndromes was evaluated using computed tomography (CT) imaging only. Children diagnosed with juvenile otosclerosis or osteogenesis imperfecta were excluded, because these disease entities have their own approach and outcome, the results of which have been published previously (11,12). Juvenile otosclerosis and osteogenesis imperfecta were diagnosed based on family history, a clinical history of progressive conductive hearing loss, intraoperative findings, and (otosclerotic focus) preoperative CT imaging. CT imaging was also used to exclude other causes of conductive hearing loss, such as enlarged vestibular syndrome.

**Audiometric Assessment**

A serial assessment of hearing status was conducted before and at 5, 6, 9, 12, 18, and 24 months after surgery and at a yearly interval thereafter. A low-frequency evaluation in the pre- and postoperative air-bone gaps (ABGs), air-conduction (AC) thresholds, and bone-conduction (BC) thresholds. Only AC and BC thresholds that were obtained at the same time postoperatively were used for calculation of the mean postoperative ABG. Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone averages for AC and BC thresholds, obtained at the last follow-up visit, were used in this study. Furthermore, closure of the ABG to within 10 dB or less was evaluated, as well as ABG closure to within 20 dB or less. Audiometry was reported according to American Academy of Otolaryngology-Head and Neck Surgery guidelines (13), except for thresholds at 3 kHz, which were substituted in all cases with those at 4 kHz. Audiometric data were collected prospectively using the Otology-Neurotology Database (ONDB) (AS Multimedia, Inc., Cassagne, France) (14).

**Statistical Analyses**

Means, standard deviations (SDs), and percentages were calculated. Differences in pre- and postoperative hearing outcomes were calculated as well as their corresponding 95% confidence intervals. Pre- and postoperative BC thresholds, AC thresholds, and ABGs were compared using a paired-samples t-test. All analyses were performed using SPSS version 20.0 (SPSS Inc., Chicago, IL).

**Surgery**

A transcanal procedure with facial nerve monitoring using the NIM 2.0 (Medtronic, Inc., Jacksonville, FL) was performed in all cases. The anatomy and mobility of the ossicular chain were carefully checked. The facial nerve was identified and its relationship with the ossicular chain and/or stapes remnant was assessed (15). The Teunissen and Cremers classification system (3) was used to define the type of malformation and ossicular chain status. The surgical technique for the repair of the ossicular chain was dictated by the surgical findings at the time of surgery and were described according to the Teunissen and Cremers classification. Surgical data were collected prospectively using the ONDB (AS Multimedia, Inc.) (14).

**Teunissen and Cremers Class I Cases**

Of the total series of 35 cases, 23 cases (66%) were class I cases, defined as stapes fixation only without any other simultaneous malformation (Supplementary Digital Table 1, http://links.lww.com/MAO/A383). Of these 23 cases, 22 were primary operations and one was a revision operation (case 14). Eleven procedures were performed on the right ear (48%) and 12 on the left ear (52%). Four patients were operated sequentially on both sides. Sex ratio was 44% women and 57% men. Mean age was 11.5 years (with a range of 7–17 yrs). Some authors have advocated waiting until age 8 to 10 years before surgical intervention and treating children with hearing aid amplification instead (3,5,8). In a few cases in this series, surgery was performed before the age of 8 years, because these cases suffered from severe hearing loss and a hearing aid was not well tolerated.

In two of 23 cases (9%) middle ear exploration revealed fixation of the stapes owing to a bony bar between the pyramidal process or the promontory and the stapes superstructure (cases 1 and 21) (Fig. 1A). The stapes footplate was normal in both cases. The bony bridge was carefully drilled out with the Skeeter drill using a 0.7-mm diameter diamond dust burr. The stapes remained mobile and the entire ossicular chain was preserved. In one case (case 1), no ossiculoplasty was attempted whereas in another case (case 21) a 0.4-mm diameter Teflon piston was positioned between the incus and the mobile stapes footplate without removing the superstructure (incus-to-footplate assembly). The choice between these two techniques was arbitrary.

An isolated congenital stapes fixation without the presence of a bony bridge was seen in 21 cases (91%). The technique was similar to our approach in primary stapedotomy: a regular 0.8-mm stapedotomy was performed using a combination of a laser and the Skeeter Microdrill (Medtronic Xomed, Inc., Jacksonville, FL), followed by vein graft interposition and incus-to-stapedotomy assembly (14). The KTP laser (Lumenis, Inc., Salt Lake City, UT) was used in 16 cases (70%), the CO2 laser (Omniguide, Inc., Cambridge, MA) in six cases (26%), and the Revolix Thulium laser (LISA Laser, Inc., Pleasanton, CA) in one case (4%). Ossiculoplasty was performed using a 0.4-mm-diameter Teflon piston of appropriate length when the incus was of normal length (20 cases [95%]). A bucket-handle (cup)–type prosthesis made of Teflon was used when the facial nerve was dehiscent (one case [5%]).

Of these 23 cases, a perilymphatic gusher occurred bilaterally in one patient (cases 2 and 3) (two cases [9%]). Both cases were treated with stapedotomy with vein graft interposition. Gelfoam was used to fill the oval window and keep the vein graft in its proper position.

**Teunissen and Cremers Class II Cases**

Of the entire series of 35 cases, 12 cases (34%) were class II cases, defined as stapes fixation with other ossicular malformation (Supplementary Digital Table 2, http://links.lww.com/MAO/A384). Of these 12 cases, eight were primary operations and four were revision operations (cases 2, 4, 10, and 11). One patient was operated bilaterally. Eight procedures were performed on the right ear (67%) and four on the left ear (33%).
Sex ratio was 75% women and 25% man. Mean age was 12.4 years (with a range of 6–18 yrs).

A simultaneous malleus ankylosis was identified in one case (case 7) (8%) and a total ossicular replacement prosthesis, composed of a hydroxyapatite head and a 0.4-mm-diameter Teflon shaft, was positioned between the malleus and the stapedotomy (malleus-to-stapedotomy procedure). The malleus relocation technique was used to increase the stability of the prosthesis (14,16). The malleus relocation technique was introduced by the first author in 1997. The malleus is dissected free from the tympanic membrane and the tensor tympani is sectioned close to its insertion into the malleus. The incus is then removed. The anterior malleal ligament is stretched by placing a hook anterior to the neck of the malleus, which is then pulled posteriorly, repositioning it so that it lies directly over the footplate. The position of the malleus is maintained by its superior ligament, which is then preserved.

The incus was malformed in 10 cases (83%). In eight cases, the incus appeared hypoplastic and short, but it was preserved and a Teflon piston was inserted (incus-to-stapedotomy procedure). A total ossicular replacement prosthesis was positioned between the malleus and the stapedotomy (malleus-to-stapedotomy procedure) in two cases. In one of these cases, the process was short, just reaching the area of the facial nerve. In the other case, the incus consisted of a fibrous band, which was connected to the malformed and fixed stapes. The malleus relocation technique was used in the two latter cases.

In one case (8%), stapedotomy was not performed at all, because the facial nerve coursed across the oval window, which itself was totally obstructed (case 12) (Fig. 1B). The surgery was aborted for the high risk of facial nerve injury. This was the only surgery that was aborted out of all class I and class II cases.

Furthermore, one case (8%) with a persistent stapedial artery (case 2) and one case (8%) with a perilymphatic gusher (case 1) were observed in this class II group.

RESULTS

Teunissen and Cremers Class I Cases

Overall Results (Table 1 and Supplementary Digital Table 1, http://links.lww.com/MAO/A385)

Postoperative data were available for all cases. The average follow-up duration was 21 months (with a range of 3–83 mo). A significant postoperative SNHL, defined as a change in the average BC threshold of 15 dB or more, was observed in one case (4%) in this series. This case experienced a dead ear immediately after surgery. This case was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n = 22 cases), but was included in the assessment of the postoperative AC and BC thresholds (n = 23 cases). Overall, the postoperative four-frequency ABG was closed to 10 dB or less in 16 of 22 cases (73%) and to 20 dB or less in 17 of 22 cases (77%). The postoperative four-frequency ABG was 12 dB (SD 15) compared with 29 dB (SD 8) preoperatively. The postoperative four-frequency average AC threshold was 32 dB (SD 22) compared with 46 dB (SD 10) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 17 dB (95% CI, 9–24, p = 0.000) and 14 dB (95% CI, 4–24, p = 0.008). Postoperatively, the four-frequency average BC threshold was 20 dB (SD 19) compared with 17 dB (SD 7) preoperatively, thus the BC threshold deteriorated with 3 dB (95% CI, −11 to 5, p = 0.437).

Hearing Results With at Least 1-year Follow-up (Supplementary Digital Table 1, http://links.lww.com/MAO/A383, and Supplementary Digital Table 3, http://links.lww.com/MAO/A385)

Postoperative data with at least 1-year follow-up were available for 14 of 23 cases (61%). The average follow-up duration was 28 months (with a range of 12–83 mo). The case presenting with a significant postoperative SNHL was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n = 13 cases), but was included in the assessment of the postoperative AC and BC thresholds (n = 14 cases). The postoperative four-frequency ABG was closed to 10 dB or less in 8 of 13 cases (62%) and to 20 dB or less in 9 of 13 cases (69%). The postoperative four-frequency average ABG was 16 dB (SD 18) compared with 29 dB (SD 9) preoperatively. The postoperative four-frequency average AC threshold was 39 dB (SD 24) compared with 48 dB (SD 10) preoperatively. Postoperatively, the four-frequency average BC threshold was 25 dB (SD 23) compared with 19 dB (SD 7) preoperatively. Only the
average ABG threshold improved significantly with a mean gain of 13 dB (95% CI, 1–25, \( p = 0.037 \)).

The audiometric results of all individual cases are visualized using the Amsterdam Hearing Evaluation Plots in Figures 2 and 3 (17). The gain in AC thresholds following surgery is dependent on preoperative ABG and both of these measures are plotted in Figure 2. Every dot below the solid diagonal line indicates a gain in AC threshold that is larger than the preoperative ABG. Every dot above the dotted diagonal line indicates an unsuccessful result, an ABG closure of more than 10 dB. The effect of surgery on BC thresholds is depicted in Figure 3 by plotting pre- and postoperative BC thresholds. The two dotted diagonal lines enclose those cases in which BC threshold did not change more than 10 dB. Every dot below the lower diagonal line indicates an improvement of the BC thresholds of more than 10 dB.

Teunissen and Cremers Class II Cases

Overall results are available in (Table 2 and Supplementary Digital Table 2, http://links.lww.com/MAO/A384).

Postoperative data were available for all cases. The average follow-up duration was 26 months (with a range of 3–119 mo). No significant postoperative SNHL was observed in this series of cases. Overall, the postoperative four-frequency ABG was closed to 10 dB or less in six of 12 cases (50%) and to 20 dB or less in eight of 12 cases (67%). The postoperative four-frequency average ABG was 13 dB (SD 14) compared with 31 dB (SD 12) preoperatively. The postoperative four-frequency AC threshold was 29 dB (SD 15) compared with 49 dB (SD 10) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 18 dB (95% CI, 6–31, \( p = 0.008 \)) and 20 dB (95% CI, 8–32, \( p = 0.004 \)). Postoperatively, the four-frequency average BC threshold was 17 dB (SD 3) compared with 18 dB (SD 6) preoperatively; thus, the BC threshold improved with 2 dB (95% CI, −1 to 4, \( p = 0.221 \)).

Hearing Results With at Least 1-year Follow-up

(Supplementary Digital Table 2, http://links.lww.com/MAO/A384, and Supplementary Digital Table 4, http://links.lww.com/MAO/A386)

Postoperative data with at least 1-year follow-up were available for eight of 12 cases (67%). The average follow-up duration was 37 months (with a range of

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**TABLE 1.** Overall pure-tone audiometric results for Teunissen and Cremers class I cases (number of procedures = 23)

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Difference (95% CI)</th>
<th>( p )</th>
</tr>
</thead>
<tbody>
<tr>
<td>AC (mean (SD) dB)</td>
<td>46 (10)</td>
<td>32 (22)</td>
<td>14 (4–24)</td>
<td>0.008</td>
</tr>
<tr>
<td>BC (mean (SD) dB)</td>
<td>17 (7)</td>
<td>20 (19)</td>
<td>−3 (−11 to 5)</td>
<td>0.436</td>
</tr>
<tr>
<td>ABG (mean (SD) dB)(^a)</td>
<td>29 (8)</td>
<td>12 (15)</td>
<td>17 (9–24)</td>
<td>0.000</td>
</tr>
<tr>
<td>ABG closure ≤10 dB (n (%))(^a)</td>
<td>—</td>
<td>16 (73)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>ABG closure ≤20 dB (n (%))(^a)</td>
<td>—</td>
<td>17 (77)</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>SNHL &gt;15 dB (n (%))</td>
<td>—</td>
<td>1 (4)</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

\(^a\) One case with SNHL was not included in the assessment of the postoperative ABG and ABG closure to within 10 and 20 dB (n = 22).

95% CI indicates 95% confidence interval; AC, air conduction; BC, bone conduction; ABG, air-bone gap; SD, standard deviation; SNHL, sensorineural hearing loss. Means were calculated using AC and BC thresholds at 0.5, 1, 2, and 4 kHz.

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**FIG. 2.** Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The solid diagonal line indicates total air-bone gap closure. The area between the diagonal lines indicates successful surgery with an air-bone gap of 10 dB or less. Audiometric data at last follow-up were used (23 class I cases and 12 class II cases).

**FIG. 3.** Amsterdam Hearing Evaluation Plot. Each black dot represents one case. The two diagonal lines enclose the cases in which bone conduction did not change more than 10 dB. Audiometric data at last follow-up were used (23 class I cases and 12 class II cases).
The postoperative four-frequency ABG was closed to 10 dB or less in five cases (63%) and to 20 dB or less in seven cases (88%). The postoperative four-frequency average ABG was 8 dB (SD 11) compared with 33 dB (SD 14) preoperatively. The postoperative four-frequency AC threshold was 25 dB (SD 14) compared with 52 dB (SD 12) preoperatively. Both ABG and AC threshold improved significantly with mean gains of 25 dB (95% CI, 9–41, \( p = 0.008 \)) and 27 dB (95% CI, 12–41, \( p = 0.003 \)). Postoperatively, the four-frequency average BC threshold was 17 dB (SD 3) compared with 19 dB (SD 7) preoperatively; thus, the BC threshold improved with 2 dB (95% CI, –3 to 6, \( p = 0.375 \)).

**Stability of Hearing Results**

Short-term follow-up (3 mo) was compared with long-term follow-up (1 year) for those cases with available pure-tone audiometry at these two follow-up moments (Supplementary Digital Table 5, http://links.lww.com/MAO/A387). The differences in postoperative four-frequency average AC thresholds, BC thresholds, and ABG between these follow-up moments were not statistically significant. Nor were the differences in ABG closure to within 10 dB or less and ABG closure to within 20 dB.

### DISCUSSION

In this study, we evaluated the hearing outcomes after stapedotomy for congenital stapes ankylosis in 23 pediatric class I cases and 12 pediatric class II cases. Overall, a postoperative ABG closure to within 10 dB was achieved in 73% of class I cases and in 50% of class II cases. A postoperative ABG closure to within 20 dB was achieved in 77% of class I cases and 67% of class II cases. Postoperative SNHL occurred in one class I case (4%) and none of the class II cases.

A rapid systematic literature search identified nine articles (2,3,6–10,18,19) reporting audiometric results following stapes surgery in children with class I and class II congenital stapes ankylosis. We only included studies that included at least 10 cases and performed a subanalysis for class I and class II congenital stapes ankylosis in five studies (Table 3). An ABG closure of 20 dB or less in class I cases was reported in six studies and was reached in 25 to 80%. Two studies reported the percentage of class II cases with ABG closure to within 10 dB: 39 and 23%. ABG closure to within 20 dB in class II cases was reported in three studies and was reached in 30 to 72%

Both juvenile otosclerosis and congenital stapes fixation are indications for middle ear surgery in children. Juvenile otosclerosis is more likely in cases with progressive conductive hearing loss and a positive family history for otosclerosis (20). Furthermore, intraoperatively established white otosclerotic foci in the anterior oval window with a lack of vascularization are characteristic for juvenile otosclerosis (20). Although the family history may contribute to a definitive diagnosis, it needs to be noted that both diseases can be hereditary and may simultaneously run in one family. CT imaging can be helpful in ruling out large vestibular aqueduct syndrome or other inner ear abnormalities that are associated with a higher risk of perilymph gusher during surgery and consequent SNHL (21). Furthermore, incudostapedial discontinuity and increased thickness of the oval window may be witnessed on CT. However, it is not uncommon for the CT scan to be entirely normal in congenital stapes ankylosis and these abnormalities are not pathognomonic for congenital middle ear malformations (6,21). Although CT scans were performed routinely in all cases included in this study, the findings were not routinely collected in the ONDB and therefore the results of CT scanning were not described in this paper. It is not unlikely that congenital stapes ankylosis is sometimes falsely considered to be juvenile otosclerosis and the other way around. Both are characterized by fixation of the stapes and both can be successfully treated with stapes surgery. Several studies were identified that compared the surgical results of congenital stapes ankylosis with juvenile otosclerosis. Children with juvenile otosclerosis generally showed better hearing results following stapes surgery than children with congenital stapes fixation. Risk differences were calculated. A risk difference is the difference between the observed risks in two treatment groups (22). Risk differences for ABG closure to within 10 dB differed between 4 and 38% in favor of juvenile otosclerosis in eight studies (23–30). Risk differences for ABG closure to within 20 dB differed between 0 and 30% in favor of juvenile otosclerosis in six studies (23–25,27,29,30) It needs to be noted that these studies included a variety of congenital stapes malformations and did not report them according to the
TABLE 3. Reported pure-tone audiometric results following surgical treatment of Teunissen and Cremers class I and class II cases in literature

<table>
<thead>
<tr>
<th>References</th>
<th>N</th>
<th>Class</th>
<th>Mean Follow-up Duration in Months (range)</th>
<th>% ABG Closure &lt;10 dB</th>
<th>% ABG Closure &lt;20 dB</th>
<th>Post ABG Closure (mean [SD])</th>
<th>Post AC Gain (mean [SD])</th>
<th>% SNHL &gt;10 dB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kisilevsky et al.</td>
<td>22</td>
<td>Class I</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>11</td>
<td>35</td>
<td>18</td>
</tr>
<tr>
<td>Park and Choung</td>
<td>46</td>
<td>Class I</td>
<td>12 (3–83)</td>
<td>NR</td>
<td>12</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Albert et al.</td>
<td>28</td>
<td>Class I</td>
<td>12 (6–82)</td>
<td>NR</td>
<td>12</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Welling et al.</td>
<td>21</td>
<td>Class I</td>
<td>&gt;4</td>
<td>48</td>
<td>20 (14)</td>
<td>31 (18)</td>
<td>18 (21)</td>
<td>7</td>
</tr>
<tr>
<td>Park and Choung</td>
<td>46</td>
<td>Class I</td>
<td>12 (3–119)</td>
<td>NR</td>
<td>13</td>
<td>29 (22)</td>
<td>14 (23)</td>
<td>4</td>
</tr>
<tr>
<td>This study</td>
<td>23</td>
<td>Class I</td>
<td>21 (3–83)</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Albert et al.</td>
<td>28</td>
<td>Class I</td>
<td>12 (6–82)</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Teunissen and</td>
<td>44</td>
<td>Class II</td>
<td>12</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Cremers</td>
<td>12</td>
<td>Class II</td>
<td>12</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

n indicates number of procedures; ABG, air-bone gap; AC, air conduction; SNHL, sensorineural hearing loss; SD, standard deviation; NR, not reported.

a0.5, 1, 2, and 3 kHz.
b0.5, 1, and 2 kHz.
c0.5, 1, 2, and 4 kHz.
dMedian with range instead of mean with SD.

Teunissen and Cremers classification or did not perform subanalyses according to this classification. Furthermore, all of the sample sizes were small and as a result none of these risk differences were statistically significant.

In the past, stapes surgery in case of congenital middle ear malformations has been thought to be associated with an increased risk of SNHL. In our series, only one class I case and none of the class II cases showed a postoperative SNHL exceeding 15 dB. In the included studies, SNHL was usually defined as a change in BC thresholds exceeding 10 dB. In these studies, SNHL was reported in less than 10% of surgically treated class I and class II cases (6–10).

Case series are vulnerable to selection bias. This case series was characterized by a clear study objective, explicit in- and exclusion criteria, consecutive patient enrollment, prospective outcome data collection, and a high follow-up rate. These strengths help to limit selection bias (22,31). Limitations of this case series include a relatively low sample size and the lack of a comparator group. Furthermore, follow-up duration was not standardized with a wide range of 3 to 119 months as a result.

CONCLUSION

Stapedotomy in children with congenital stapes ankylosis class I and class II was associated with satisfactory pure-tone audiometric results, with an ABG closure of 10 dB or less in 73% of class I cases and in 50% of class II cases. An ABG closure of 20 dB or less was achieved in 77% of class I cases and 67% of class II cases. Postoperative SNHL occurred in one class I case (4%) and none of the class II cases.

REFERENCES


