The Jahrsdoerfer Grading Scale in Surgery to Repair Congenital Aural Atresia

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Objective: To determine the predictive ability of the Jahrsdoerfer grading scale score in congenital aural atresia surgery.

Design: Retrospective review of medical records.

Setting: Tertiary referral center.

Patients: One hundred eight patients with aural atresia.

Main Outcome Measures: Demographic data, preoperative Jahrsdoerfer score, and postoperative audiometric outcomes were reviewed. One month postoperative, 4-tone pure-tone averages and speech reception thresholds were compared between ears scoring 6 or lower, 7, and 8 or higher on the Jahrsdoerfer grading scale. The percentage of ears with a speech reception threshold of 30 dB hearing level or lower for each group was calculated and compared between groups. Individual anatomical structures on the Jahrsdoerfer grading scale were evaluated for their ability to predict postoperative audiometric success.

Results: Of 116 ears evaluated, postoperative 4-tone pure-tone averages and speech reception thresholds were significantly poorer in ears scoring 6 or less on the Jahrsdoerfer grading scale compared with ears scoring 7 or higher ($P < .02$, t test). Ears scoring 6 or less had a 45% chance of achieving a postoperative speech reception threshold of 30 dB hearing level or lower, while ears scoring 7 or higher had an 89% chance ($P < .01$, $\chi^2$ test). Lack of middle ear aeration was the only anatomical factor predictive of poor audiometric outcome.

Conclusions: Compared with patients with a Jahrsdoerfer score of 6 or lower, patients with a score of 7 or higher had significantly better hearing postoperatively. Middle ear aeration may be the most important predictor of postoperative hearing outcome. The Jahrsdoerfer grading scale is an invaluable tool in the preoperative evaluation of patients with congenital aural atresia.


Surgery to repair congenital aural atresia is performed in an attempt to restore the normal sound-conducting mechanism of the ear, thereby improving and in many cases normalizing hearing. The operation involves drilling the bone of the tympanic ring to identify the middle ear space and the ossicular chain, freeing the ossicular chain from the surrounding bone, constructing an eardrum using a lateral surface temporalis fascia graft, lining the bony canal with a split-thickness skin graft, and delivering the skin graft through a meatus created in the reconstructed or native auricle.1-3

Not all patients with aural atresia, however, are candidates for surgery. A patient’s candidacy for surgery is based on audiometric findings and the anatomy of the temporal bone. High-resolution computed tomography remains the best method for evaluating the development and anatomy of the temporal bone in these patients.4 Important anatomical considerations when evaluating a child for surgery include the position and course of the facial nerve, location of the tegmen, presence of the stapes bone, and status of the oval window. The Jahrsdoerfer grading scale, proposed in 1992, assigns an anatomical score (1-10 [the higher the score, the better]) for the atretic ear based on the presence or absence of 9 structures (Table 1).5,6 The scale not only evaluates a patient’s candidacy for surgery but also, as some have proposed, predicts audiometric outcome. The higher the Jahrsdoerfer grading scale score, the better the chance for normal or near-normal postoperative hearing.5,6

Given the inherent difficulty of surgery to repair aural atresia, a tool that can accurately predict audiometric outcomes is invaluable in the assessment of atresia, especially unilateral atresia. The purpose of this study was to test the hypothesis that the preoperative Jahrsdoerfer score accurately predicts postoperative hearing results by comparing postoperative audiometric outcomes between groups of patients.
having different Jahrsdoerfer scores. The percentage of patients in each group that achieved “normal or near-normal hearing,” defined as a speech reception threshold (SRT) of 30 dB hearing level (HL) or lower, was also calculated to determine the ability of the Jahrsdoerfer score to predict normal or near-normal postoperative hearing. The specific anatomical structures analyzed by the Jahrsdoerfer grading scale were compared between groups with good and poor audiometric outcomes to determine the predictive ability of each structure.

METHODS

Medical records were reviewed retrospectively for all patients undergoing surgery to repair congenital aural atresia at the University of Virginia, Charlottesville, between January 2, 1996, and December 31, 2006. The study was approved by the University of Virginia Institutional Review Board. Patients were identified by the International Classification of Diseases, Ninth Revision diagnosis code for aural atresia or Current Procedural Terminology codes indicating surgery to repair congenital ear malformations. Patients undergoing revision aural atresia surgery or surgery to repair minor congenital middle ear malformations were excluded.

Patient demographic information was collected including age, sex, unilateral vs bilateral atresia, side of operation, preoperative Jahrsdoerfer score, and the specific point value (0, 1, or 2) assigned to each anatomical structure based on the Jahrsdoerfer grading scale. Audiometric data collected included preoperative and 1-month postoperative air conduction 4-tone pure-tone average (PTA; 500, 1000, 2000, and 3000 Hz) and SRTs. Also excluded were patients without complete audiometric data, patients in whom the operation was aborted without an attempt at reconstruction, and patients with substantial preoperative sensorineural hearing loss (ie, bone conduction 4-tone PTA >30 dB HL). Patients included in the study were classified into 3 groups on the basis of their preoperative Jahrsdoerfer score as follows: 6 or lower, 7, or 8 or higher. Scores with half points were rounded down to the nearest whole number (eg, 7.5 was rounded down to 7).

Statistical analysis was performed using single-factor analysis of variance to compare the preoperative and 1-month postoperative audiometric results (PTA and SRT) across the 3 groups of patients. The t test was then used to compare the 1-month postoperative PTA and SRT between selected groups. The percentage of patients achieving normal or near-normal hearing (ie, postoperative SRT ≤30 dB HL) was calculated for each group. The χ² test was then used to compare the probability of achieving normal or near-normal hearing between patients with different preoperative Jahrsdoerfer scores.

Regardless of the preoperative Jahrsdoerfer score, ears were classified into 2 groups: those with a postoperative 4-tone PTA either higher than 35 dB HL or 35 dB HL or lower. We used the PTA of 35 dB HL as the cutoff value because it marks the break point between mild and moderate hearing loss. The specific anatomical structures that lost points on the Jahrsdoerfer grading scale were identified for each ear. These structures were compared between the 2 hearing outcome groups (χ² analysis) to identify associations between abnormal structure and hearing outcome.

RESULTS

One hundred eight patients (127 ears) were identified. Sixty-nine patients (64%) were male, and 39 (36%) were female. Their median age at surgery was 8 years (age range, 4-62 years). Bilateral atresia was present in 39 patients (38%), unilateral atresia in the right ear in 47 patients (44%), and unilateral atresia in the left ear in 22 patients (20%). Eleven ears were excluded from the final analysis: 4 because of missing data, 2 because of preoperative sensorineural hearing loss, and 5 because the surgery was aborted without attempt at reconstruction. One of the 5 operations was aborted because a previously unidentified polymeric silicone (Silastic) implant was discovered during surgery; 1 was aborted because of a low-hanging dura that prevented adequate visualization for reconstruction; and 3 were aborted because of the position of the facial nerve. These ears had preoperative scores of 5, 6, and 7, and although each lost points on the preoperative evaluation for facial nerve position, it was thought preoperatively that the position of the nerve would not prevent a successful operation. Preoperative Jahrsdoerfer scores for the 116 included ears were as follows: 1 to 4 points, no ears; 5 points, 1 ear (0.9%); 6 points, 10 ears (8.6%); 7 points, 49 ears (42.2%); 8 points, 53 ears (45.7%); 9 points, 3 ears (2.6%); and 10 points, no ears. Surgery was not attempted in ears with scores lower than 5. The mean Jahrsdoerfer score for the 116 ears enrolled in the study was 7.46.

For statistical analysis, the 1 ear with a score of 5 was included in the group with a score of 6. Three ears with a score of 9 were added to the group with a score of 8. This left 3 groups for statistical comparison: 11 ears with a score of 6 or lower, 49 ears with a score of 7, and 56 ears with a score of 8 or higher. No significant difference was noted in preoperative air conduction 4-tone PTA and SRT across the 3 groups (Figure 1 and Figure 2, P= .19 [PTA] and P< .58 [SRT]), single-factor analysis of variance.

The mean postoperative air conduction 4-tone PTA and SRT for each group is shown in Figure 3 and Figure 4. There was a significant difference in both the postoperative air conduction 4-tone PTA and SRT across the 3 groups (P< .004, single-factor analysis of variance). When paired comparisons between groups were performed, the postoperative air conduction 4-tone PTA was significantly poorer for ears having a score of 6 or lower compared with ears having a score of 7 or of 8 or higher (Figure 3, P< .03 for both comparisons, t test). Likewise, the postoperative

### Table 1. Jahrsdoerfer Grading Scale Score for Congenital Aural Atresia

<table>
<thead>
<tr>
<th>Anatomical Structure</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stapes bone</td>
<td>2</td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
</tr>
<tr>
<td>Middle ear space</td>
<td>1</td>
</tr>
<tr>
<td>Facial nerve</td>
<td>1</td>
</tr>
<tr>
<td>Malleus-incus complex</td>
<td>1</td>
</tr>
<tr>
<td>Mastoid pneumatization</td>
<td>1</td>
</tr>
<tr>
<td>Incus-stapes connection</td>
<td>1</td>
</tr>
<tr>
<td>Round window</td>
<td>1</td>
</tr>
<tr>
<td>External ear</td>
<td>1</td>
</tr>
<tr>
<td>Total Possible Score</td>
<td>10</td>
</tr>
</tbody>
</table>

*Adapted from Jahrsdoerfer et al.*

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air conduction SRT was significantly poorer for ears having a score of 6 compared with ears having a score of 7 or of 8 or higher (Figure 4; \( P < .01 \) for both comparisons, \( t \) test). There was no significant difference in either the postoperative 4-tone PTA or the SRT between ears having a score of 7 and those having a score of 8 or higher (Figure 3 and Figure 4; \( P < .3 \), \( t \) test).

Normal or near-normal hearing (ie, postoperative SRT \( \leq 30 \) dB HL) was achieved in 98 of 116 ears (84%). Compared with 105 ears with a score of 7 or higher, 11 ears with a score of 6 or lower had a significantly lower chance of achieving normal or near-normal hearing (45% vs 89%; \( P < .01 \), \( \chi^2 \) test). No significant difference was noted in the likelihood of achieving normal or near-normal hearing between 49 ears having a score of 7 and 56 ears having a score of 8 or higher (\( P < 1 \); \( \chi^2 \) test).

One hundred two ears were identified with documented scores for each of the structures in the Jahrsdoerfer grading scale. Of the 102 ears, 28 (27%) had a postoperative 4-tone PTA greater than 35 dB HL and 74 (73%) had a postoperative 4-tone PTA of 35 dB HL or less. Only poor middle ear aeration was significantly more likely in the 28 ears with poorer postoperative PTA (\( P < .05 \), \( \chi^2 \) test) (Table 2). When the 3 ears in which surgery was aborted because of intraoperative facial nerve location were included in the statistical analysis, abnormal facial nerve position approached statistical significance (\( P = .08 \), \( \chi^2 \) test).

Repair of congenital aural atresia remains one of the more challenging operations in otology. Because of the difficulty of atresia surgery, its risks, and the often inconsistent postoperative hearing results, not all patients with congenital aural atresia are good surgical candidates, and the literature suggests careful selection of patients for repair of atresia.\(^5\)\(^6\)

By comparing postoperative PTA and SRT among 3 groups of ears with different preoperative Jahrsdoerfer scores, this study supports the use of the Jahrsdoerfer grading scale in the selection of patients for surgery. Our results demonstrate that ears scoring 6 or lower have a worse postoperative hearing outcome than ears scoring 7 or higher. In addition, ears scoring 8 or higher did no bet-
ter than those scoring 7, with 80% to 90% of ears in both groups achieving normal or near-normal hearing. When individual anatomical features were evaluated, lack of middle ear aeration was significantly more likely in ears with poor hearing outcome (Table 2).

The goals of atresia repair are to obtain the best possible hearing outcome and to construct a clean, well-epithelialized external ear canal and tympanic membrane. Previous studies have reported postoperative SRTs in the 10- to 25-dB HL range in certain patients.1 The risks of atresia repair include injury to the facial nerve, sensorineural hearing loss, failure to close the air-bone gap, and meatal or canal stenosis.7,9 Jahrsdoerfer10 discussed the difficulty of reconstructing an ear in which the facial nerve is displaced. In 3 of the excluded ears in the present study, the operation was aborted because of the position of the facial nerve. Facial nerve position approached significance as a predictor of poor audiometric outcome when the 3 aborted operations were included in the analysis.

Despite the deleterious effects of unilateral hearing loss,11 repair of unilateral aural atresia remains controversial, and it has been suggested that surgery be performed only in patients who have reliable preoperative indicators of a high likelihood of achieving normal hearing postoperatively. Based on the evaluation of anatomical structures on high-resolution computed tomography of the temporal bone, the Jahrsdoerfer grading system was developed to determine surgical candidacy and to predict which patients would have more favorable hearing outcomes.9

The patients evaluated in this study were an appropriate sampling of the population with congenital aural atresia. The demographic data, including the incidence of bilateral atresia and the male preponderance observed in this study, corresponds with published demographic data.12 No statistical difference was noted in the preoperative 4-tone PTA or SRT regardless of the assigned preoperative Jahrsdoerfer score. The postoperative audiometric data were obtained no earlier than 1 month postoperatively and no later than 3 months postoperatively. The current study does not purport to evaluate long-term hearing outcomes. One-month postoperative audiometric outcomes may decline, although de la Cruz and Teufert13 reported relatively stable hearing with no significant change in the air-bone gap from short- to long-term (>6 months) follow-up.13 A study by Lambert14 demonstrated some degradation in hearing after 1 year postoperatively.

Although there was no hearing difference between patients preoperatively, the postoperative audiometric data demonstrated the predictive ability of the Jahrsdoerfer grading system. Patients with a preoperative Jahrsdoerfer score of 6 or lower had a significantly worse audiometric outcome for both PTA and SRT compared with patients scoring 7 or higher. Compared with patients with a Jahrsdoerfer score of 7 or higher, patients with a score of 6 or lower had a significantly lower chance of achieving normal hearing. These patients had a 45% chance of achieving normal or near-normal hearing based on postoperative SRT. This was less than the 60% result reported in the literature.9

In the current study, ears with a preoperative Jahrsdoerfer score of 7 had a better audiometric outcome than predicted by the literature.4 No significant difference was noted in the postoperative 4-tone PTA and SRT between these patients and those scoring 8 or higher; all patients with a preoperative Jahrsdoerfer score of 7 or higher had an 88% to 90% chance of achieving normal or near-normal hearing on the basis of postoperative SRT. This highly favorable outcome for patients scoring 7 may be artificial, a result of rounding error. Scores with half points were rounded down to the nearest whole number; thus, a score of 7.5 was rounded down to 7. Therefore, rounding error may have skewed the data to more favorable outcomes for patients with a score of 7. We did not statistically analyze subgroups by half points. A score of 7 seems to be the break point between patients who did well audiometrically and those who did not.

Several studies have attempted to correlate surgical anatomy and audiometric outcome with preoperative factors. Age, severity of microtia, and revision vs primary surgery have been linked to hearing outcomes.15,16 The specific anatomical structures evaluated by the Jahrsdoerfer grading scale were compared between ears with a very good postoperative hearing outcome (4-tone PTA ≤35 dB HL) and ears with a suboptimal hearing outcome (4-tone PTA >35 dB HL). Lack of middle ear aeration was the only variable evaluated by the Jahrsdoerfer grading scale that was significantly more likely to occur in ears with a poorer hearing outcome. No statistically significant difference was noted in the presence or absence of the other 8 anatomical structures evaluated using the Jahrsdoerfer scale between the 2 groups, although the facial nerve approached significance when ears in which surgery was aborted were included in the analysis. Middle ear aeration may be the most important predictor of surgical success. Lack of aeration may result in difficulty identifying a middle space and ossicular chain during drilling, relifixation of the ossicular chain postoperatively, and postoperative stenosis with a constricted middle ear space and smaller tympanic membrane.

<table>
<thead>
<tr>
<th>Abnormal Structure</th>
<th>Ears With Abnormality, No. (%)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Postoperative 4-Tone PTA &lt; 35 (n=74)</td>
<td>Postoperative 4-Tone PTA &gt; 35 (n=28)</td>
</tr>
<tr>
<td>Stapes bone</td>
<td>64 (86)</td>
<td>24 (86)</td>
</tr>
<tr>
<td>Oval window open</td>
<td>4 (5)</td>
<td>0</td>
</tr>
<tr>
<td>Middle ear space</td>
<td>7 (9)</td>
<td>7 (25)</td>
</tr>
<tr>
<td>Facial nerve</td>
<td>13 (18)</td>
<td>8 (29)</td>
</tr>
<tr>
<td>Malleus-incus complex</td>
<td>10 (14)</td>
<td>6 (21)</td>
</tr>
<tr>
<td>Mastoid</td>
<td>2 (3)</td>
<td>3 (11)</td>
</tr>
<tr>
<td>Pneumatization</td>
<td>15 (20)</td>
<td>6 (21)</td>
</tr>
<tr>
<td>Incus-stapes</td>
<td>1 (1)</td>
<td>0</td>
</tr>
<tr>
<td>Round window</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>External ear</td>
<td>66 (89)</td>
<td>23 (82)</td>
</tr>
</tbody>
</table>

Abbreviation: PTA, pure-tone average.

**Table 2. Anatomical Structure as Predictor of Postoperative Hearing Outcome**
The significance of this study is 2-fold. First, the results can be useful in determining surgical candidacy and in counseling patients preoperatively. A patient with favorable anatomy scoring 7 or higher on the Jahrsdoerfer grading scale can be informed that there is an approximately 85% to 90% chance of achieving normal or near-normal hearing as measured by SRT postoperatively. Similarly, a patient who is contemplating surgery and has unilateral atresia with a score of 6 on the Jahrsdoerfer grading scale may not be a good surgical candidate because there is only a 40% to 50% chance of achieving normal or near-normal hearing postoperatively. Nevertheless, conventional hearing amplification in the newly constructed ear canal may offer a more acceptable means of hearing rehabilitation than a bone-oscillating or bone-anchored hearing device. In general, we do not recommend surgery in patients with unilateral atresia and a score lower than 6. For patients with bilateral atresia, we do not offer surgical reconstruction for patients with a score lower than 5.

Second, while this article demonstrates the effectiveness of the grading scale in predicting hearing outcome, the grading scale may be more detailed than necessary for determining surgical candidacy. The evidence that lack of middle ear aeration correlates significantly with poorer postoperative hearing results may enable simplification of the Jahrsdoerfer grading scale, or a modified Jahrsdoerfer grading scale. To determine a patient’s candidacy for surgery, computed tomography can be used to assess primary structures that would immediately preclude surgery: the position of the tegmen, the location and position of the facial nerve, the presence of middle ear aeration, and the appearance of the stapes bone or oval window. If the location of the facial nerve would prevent reconstruction or place it at risk during drilling, if the tegmen is too low to enable visualization, or if the middle ear space is not aerated, the patient is deemed not a candidate for surgery. After applying these criteria, if a patient is determined to undergo the surgery, the more detailed Jahrsdoerfer grading scale can then be used to make predictions about hearing outcome postoperatively.

The primary limitation of this study is the relatively small number of patients in the group with a preoperative Jahrsdoerfer score of 6 or lower. Although statistical significance was demonstrated, it is possible this smaller sampling of patients skewed the data in favor of poorer outcome. Nevertheless, the results support the continued use of the Jahrsdoerfer grading scale, and possibly a more simplified version, in the preoperative evaluation of patients with congenital aural atresia.

CONCLUSIONS

Given the inherent difficulty of surgery to repair aural atresia, the documented risks, and the often inconsistent postoperative hearing results, careful selection of candidates for surgery is paramount, particularly in patients with unilateral atresia. The results of the current study show that the Jahrsdoerfer score, based on high-resolution computed tomography of the temporal bone anatomy, can accurately predict postoperative hearing outcome and assist in determining surgical candidacy. It is a powerful tool in the preoperative evaluation and counseling of patients considering surgery to repair congenital aural atresia.

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Author Contributions: Dr Kesser had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Shonka and Kesser. Acquisition of data: Shonka and Livingston. Analysis and interpretation of data: Shonka and Kesser. Drafting of the manuscript: Shonka and Kesser. Critical revision of the manuscript for important intellectual content: Shonka, Livingston, and Kesser. Statistical analysis: Shonka and Kesser. Administrative, technical, and material support: Kesser. Study supervision: Kesser.

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Additional Contributions: Mark Conaway, PhD, assisted with the statistical analysis, and Elizabeth Aitcheson assisted with data gathering.

REFERENCES