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Hernan Goldsztein and Joseph B. Roberson, Jr

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Anatomical Facial Nerve Findings in 209 Consecutive Atresia Cases

Hernan Goldsztein, MD[1], and Joseph B. Roberson Jr, MD[1]

Abstract

Objective. Describe intraoperative facial nerve findings in 209 consecutive atresia cases. Identify preoperative and intraoperative anatomical variants that should alert the surgeon to potential high-risk facial nerve anatomy.

Study Design. Case series with chart review.

Setting. Tertiary care subspecialty private practice.

Methods. Retrospective review of 209 consecutive atresia cases treated between 2007 and 2011. Descriptive analysis of intraoperative findings. Logistical regression models with generalized estimating equations were used to examine the effect of preoperative variables over the operative findings.

Results. Two hundred and nine consecutive patients (ages 2-48) underwent atresia repair between 2007 and 2011. Preoperative Jahrsdoerfer Scale was 9 (23%), 8 (42%), 7 (19%), 6 (2%), 5 or less (2%). The facial nerve was found to have an abnormal course in 39% of the cases and not identified in 1%. It was congenitally dehiscent in 53% of cases and was surgically exposed in 10%. The most common site of congenital dehiscence was in the tympanic segment (57%). Facial-stapes contact was found in 11% of cases. The stapedius tendon was absent in 30% of cases. A single patient had a mild transient postoperative paresis (House-Brackmann 2).

Conclusion. Atresia repair remains one of the most challenging procedures in otology. In spite of modern preoperative imaging, the facial nerve remains at risk. When performing surgery on patients with preoperative facial nerve paresis and/or lower Jahrsdoerfer scores, the surgeon should be aware of a higher incidence of facial nerve abnormalities. Thorough knowledge of anatomical variations and meticulous surgical technique are mandatory to safely perform these surgeries.

Congenital aural atresia (CAA) and microtia occur in approximately 1:10,000-20,000 births as a result of the aberrant development of the first and second branchial arches. As a consequence, there is hypoplasia or aplasia of the external ear (pinna and external auditory canal) and the middle ear.[1] The development of the facial nerve (FN) is intimately related with the development of the middle ear and the mastoid process. The FN canal initially appears as a sulcus in the cartilaginous otic capsule. Ossification starts from 2 distinct centers (anterior near the apex of the cochlea and posterior at the pyramidal eminence) at 20 and 25 weeks of gestation, respectively.[2] The bone progressively encases the facial nerve and the process is usually completed 3 months after birth. As the mastoid process and tympanic ring grow postnatally, they displace the nerve medially.

In cases of congenital atresia, the tympanic portion of the FN tends to be dehiscent in a greater proportion of patients, possibly due to lack of merging of the ossification centers. As a consequence of the impaired development of the mastoid process and tympanic ring, the vertical segment of the FN tends to lie more anterior and lateral and adopt a more horizontal disposition in its way to the stylomastoid foramen. The second genu of the FN tends to be located more lateral and anterior, potentially obscuring access to the middle ear. Additionally, there is a higher incidence of overlap of the oval window and contact with the stapes superstructure. These anatomical variations place the FN at risk for injury during atresia repair, particularly in the inferoposterior portion of the atretic bone just lateral to the middle ear.[3]

With the advent of modern high-resolution CT scans, the surgeon can evaluate the position of the FN preoperatively and plan the procedure appropriately. However, CT scan evaluation does not always correspond with the intraoperative findings.[4] In this work we intend to describe our intraoperative findings in 209 consecutive atresia cases and attempt to identify preoperative factors that can alert the otologic surgeon for FN abnormalities to be encountered.

[1]California Ear Institute, Palo Alto, California, USA

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Corresponding Author:
Joseph B. Roberson Jr, MD, California Ear Institute, 1900 University Ave. Suite 101, E. Palo Alto, CA 94303, USA
Email: jbr@calear.com

Materials and Methods

Institutional Review Board exemption status was obtained for this project from the Western Institutional Review Board (Olympia, Washington). All patients were seen and treated at the senior author's (JBR) practice consecutively between the years of 2007 and 2011. Standard microsurgical techniques were used as described in a prior publication.[5] Continued intraoperative nerve monitoring was performed for all cases using an NIM 2.0 nerve monitoring system (Medtronic Inc, Minneapolis, Minnesota). Paired subdermal electrodes were placed in the orbicularis oculi and oris. Ground and stimulation return electrodes were placed subdermally in the suprasternal notch as indicated by the manufacturer. Demographic, medical, and intraoperative data were prospectively accumulated in a password-protected Bento database (FileMaker, Inc, Santa Clara, California) since 2007. This information was de-identified and exported into Excel (Microsoft Corporation, Redmond, Washington) for retrospective review. Inclusion criteria were patients with congenital atresia operated on primarily; revision cases were excluded. Minor atresia cases (presence of external auditory canal and a tympanic membrane) were also excluded. Descriptive analysis was performed with Excel. Logistical regression models with generalized estimating equations were used to examine the effect of preoperative variables over the operative findings. All analyses were done with SPSS version 20.0 (IBM Corporation, Armonk, New York).

Results

Descriptive Analysis

The patient population consisted of 209 consecutive patients undergoing atresia repair between 2007 and 2011 by the senior author (JBR). Preoperative demographics are summarized in **Table 1**.

The stapes superstructure was found to be abnormal in approximately a third of patients (37%). We most commonly noticed a concave suprastructure with unusually thin crura. We often identified an associated abnormal oval window and footplate (**Figure 1**). The stapedius tendon was absent in 30% of patients. The FN was found to have an abnormal course (lying more lateral and anterior than usual atresia cases) in 39% of the cases and not identified in 1%. Facial nerve–stapes contact was found in 11% of cases (**Figure 2**). The nerve was congenitally dehiscent in 53% of cases and was surgically exposed in 10%. The most common site of congenital dehiscence was in the tympanic segment (57%), and the most common site of surgical exposure was in the descending segment just inferior to the ossicles (**Figure 3**). Intraoperative FN potentials were recorded in 14 patients; however, a single patient had a mild transient postoperative paresis (House-Brackmann 2) as a consequence of surgery. In all instances the potentials were single events, noncontinuous, and self-limited recorded when operating close to the FN, such as lysing adhesions, drilling along the course of the

**Table 1.** Summary of the Demographic Characteristics of the Patient Population

|	
Age	
Median	5
Range	(2-48)
Gender	
Male	135 (65%)
Female	66 (32%)
Side	
Right	112 (54%)
Left	51 (24%)
Bilateral	46 (22%)
Preoperative Jahrsdoerfer score	
3	1 (0%)
4	2 (1%)
5	3 (1%)
6	5 (2%)
7	40 (19%)
8	88 (42%)
9	49 (23%)
Not available	19 (9%)
Preoperative Paresis	
Yes	9 (4%)
No	200 (96%)
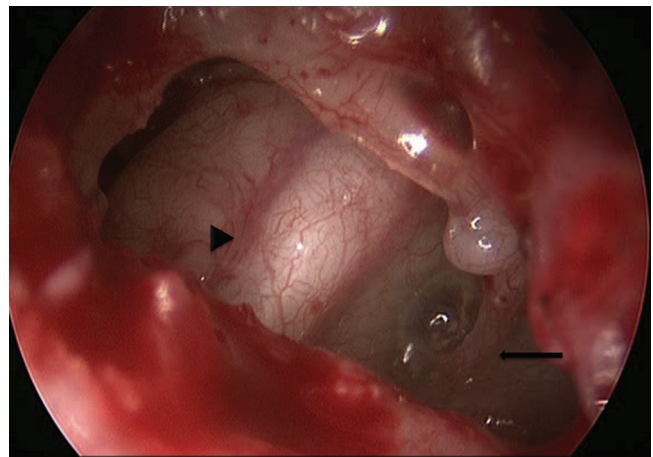
Figure 1. Intraoperative view of a right ear with congenital atresia. Arrowhead points toward the dehiscent horizontal segment of the facial nerve. Black arrow points toward the stapes suprastructure.

nerve, or when mobilizing the ossicles that were in contact with the nerve.

Logistic regression was used to analyze preoperative variables and see how well they could predict postoperative FN findings. Preoperative variables included age at time of surgery, gender, side (right, left, right of bilateral, or left of bilateral), preoperative Jahrsdoerfer Scale, and presence
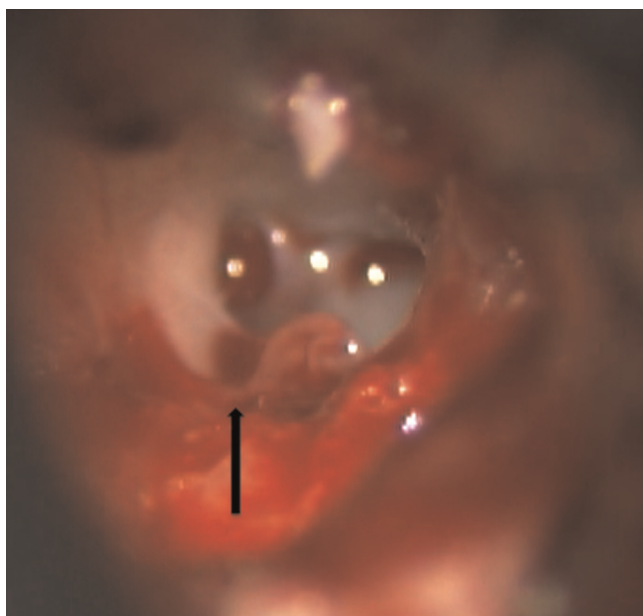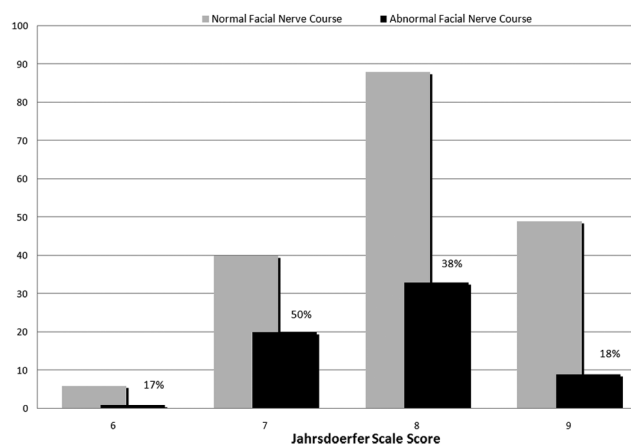
Figure 2. Intraoperative view of a right ear with congenital atresia. The arrow points at the site of contact between the stapes suprastructure and the dehisced horizontal segment of the facial nerve.
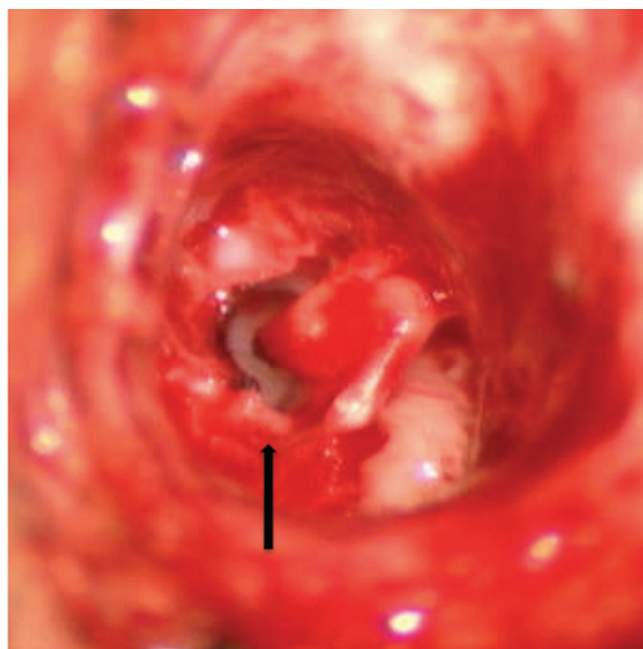
Figure 3. Intraoperative view of a left ear. The arrow points to the surgically exposed descending segment of the facial nerve. Potentials were recorded on the nerve monitor but there was no facial nerve injury.

Figure 4. Patients with abnormal facial course versus patients with normal facial nerve distributed by Jahrsdoerfer Scale score. A significant trend toward normal facial nerve anatomy was observed with increasing Jahrsdoerfer score (P = 0.01).

of FN paresis. No preoperative variables predicted facial-stapes contact or recorded FN potentials by intraoperative nerve monitoring. As mentioned previously, an abnormal course of the FN was found in 39% of patients; preoperative Jahrsdoerfer Scale score (P = .01) (**Figure 4**) and preoperative FN paresis (P = .04) predicted a significant abnormal course. Left atretic ears were more likely to have abnormal FN course, but this relationship did not reach statistical significance (P = .077).

Discussion

Atresia and microtia are the result of first and second arch developmental abnormalities, which have significant effect on structures of the ear, including the FN course. External ear deformities are commonly accompanied by malformations of the middle ear cleft.[1] Most patients present with a maximal conductive hearing loss. Treatment options for this condition include observation, bone conduction devices (both implantable and nonimplantable), middle ear implantable hearing aids, and surgical correction. Not all patients are amenable for surgical correction or middle ear implants, however, and determining candidacy is critical for success. Facial nerve anatomical evaluation and preservation both preoperatively and intraoperatively are paramount to successful outcome.

Several classification systems have been developed to describe the extent of the malformations and determine candidacy for surgical repair (Altman, De la Cruz, Jahrsdoerfer, Schuknecht, and Chiossone).[1] Perhaps the most important tool in the evaluation of patients with CAA has been the high-resolution CT scan. CT scans are critical for many of these classifications, including the most commonly used Jahrsdoerfer grading system of candidacy for surgery of CAA. This grading system relies almost exclusively on the CT and gives points based on the presence of the stapes, mastoid and middle ear pneumatization, presence of the oval and round windows, FN course, status of the ossicles, and external appearance.[6]

Dedhia et al felt that the Jahrsdoerfer system did not completely describe the anatomy and potential intraoperative challenges. They propose a modified grading system adding the position of the tegmen and the position of the

malleus-incus complex with respect to the stapes, and they divide the FN grade into 2 distinct points based on the relationship of the nerve to the middle ear and to the oval window.[7] In their series the FN obstructed access to the oval window in 41% of the cases and obstructed access to the middle ear in 21%. However, in their pilot study they warned that CT findings did not always correlate with intraoperative findings.[8]

Yu et al correlated preoperative CT scans with intraoperative findings in the identification of the FN.[4] In this work CT proved to be an invaluable tool for the preoperative evaluation of the FN course. The authors pointed out that in some cases CT was unable to identify the nerve, especially when there was opacification of the middle ear. When looking at the relationship between the FN and the oval and round windows, as well as bony dehiscence, they found the predictive power of CT less reliable.

Facial nerve injury has been reported in 0% to 11% of cases[3,4] and is perhaps the most feared risk of these surgeries. Therefore in this article we intended to analyze anatomical FN findings in a large cohort of patients with CAA. Our series is in concordance with previous reports of a male and right-sided predominance. There is a logical bias toward higher Jahrsdoerfer scores given that the patient population mostly consists of atresiaplasty candidates. Our findings of an abnormal course and dehiscence of the FN (39% and 53%, respectively) is in line with prior reports.[1,3] Although the assignment of an abnormal course was partially arbitrary, it was based on the senior author's extensive experience with atresiaplasty. Given the inherent developmental anomalies in patients with CAA, all of them present certain FN anatomical variation. Unusual lateral and anterior displacement of the nerve for the normative CAA patient at or after the second genu was assigned an abnormal course. We routinely utilize facial nerve monitoring for our cases. The FN was surgically exposed in 10% of patients, and intraoperative FN potentials were detected in only 14 patients. In spite of these findings, a single patient had a delayed transient paresis with no permanent injury.

Due to the extremely low incidence of FN sequelae in our patient group we were not able to identify any preoperative variables that correlated with increased operative risk to the FN. The single patient with a very mild (House Brackmann 2/6) transient facial weakness had no intraoperative potentials and a 270-degree circumferential dehiscent horizontal portion of the facial nerve from the geniculate to the second genu. This facial nerve was the most dehiscent in this segment of all cases in this series. The paresis started 10 days after surgery and lasted for a total of 3 weeks. Facial nerve monitoring is believed by the authors to be mandatory in this group of patients. Anatomic abnormalities undoubtedly put the FN at greater risk of injury and should be sought diligently preoperatively. Indeed, abnormal position of the FN may be the sole factor to prevent a surgeon from performing atresia repair in some cases.

Obstruction of the oval window and the middle ear by the FN has been reported in the past,[4,7] but we have not seen a report specifically addressing facial-stapes contact. Contact of the ossicular chain and the FN may have implications for postoperative hearing results. We found 11% of patients having soft tissue synechae between these structures (**Figure 2**). When possible and safe we have lysed these connections to release the stapes. A future publication will analyze this patient population with respect to hearing outcomes.

Although not reaching statistical significance ($P = .077$), it was interesting that left ears had a higher incidence of abnormal FN configuration. Further analysis with larger numbers may shed light on this observation. Another interesting finding is the absence of the stapedius tendon in 30% of patients given its intimate embryological development with the FN and stapes.

Preoperative FN paralysis should alert the surgeon to anatomical abnormalities at the time of surgery. CT scanning preoperatively is indispensable in determining FN position; however, the facial nerve can be expected to have more abnormalities than high-resolution CT scans will detail preoperatively.[4,7,8] Extreme intraoperative care and diligence is needed for successful surgical outcome with regards to the facial nerve. Finally, in spite of attempts by many authors to replace, modify, or improve Dr Jahrsdoerfer's scale for congenital atresia, it remains one of the most used tools, as the composite score shows a correlation with abnormal FN anatomy at the time of surgery. It is our opinion that more data than the Jahrsdoerfer score relays are needed to make a fully informed decision regarding successful CAA surgery with respect to the facial nerve.

Limitations of this study include inherent limitations of retrospective studies. The assignment of an abnormal course of the FN or abnormal structure for the stapes is to a certain degree arbitrary but based on the senior author's extensive experience in atresiaplasty. As this study is purely descriptive in nature, it does not detail functional outcomes. Future projects will be directed toward correlating these findings with hearing outcomes and revision rates.

Conclusions

Atresia repair remains one of the most challenging procedures in otology. In spite of modern preoperative imaging, the facial nerve remains at risk given the variable development of the first and second branchial arches. We strongly believe in continuous intraoperative facial nerve monitoring for these cases. Most importantly, thorough knowledge of anatomical variations and meticulous surgical technique are mandatory to safely perform these surgeries. Facial nerve–stapes contact is present in 11% of patients and the stapedius tendon was absent in 30% of the population. Patients with preoperative facial nerve paresis and lower Jahrsdoerfer scores have a higher incidence of abnormal facial nerve anatomy.

Author Contributions

Hernan Goldsztein, MD, designed study, collected data, analyzed data, wrote article; **Joseph B. Roberson Jr**, MD, designed study, collected data, revised article.

Disclosures

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