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Mandibular Condylar Hypoplasia in Children With Isolated Unilateral Congenital Aural Atresia

Tyler R. Halle, MD; N. Wendell Todd, MD, MPH; Bruno P. Soares, MD

Objectives/Hypothesis: We hypothesized that children with isolated nonsyndromic unilateral congenital aural atresia have subclinical mandibular condylar hypoplasia ipsilateral to the atretic ear, and that the Jahrsdoerfer score is associated with the degree of condylar hypoplasia.

Study Design: Retrospective self-controlled case series.

Methods: We reviewed high-resolution computed tomography scans of the temporal bones of 68 children with isolated nonsyndromic unilateral congenital aural atresia. Images were viewed in the transverse (axial) plane perpendicular to the axis of the mandibular ramus and scanned from the mandibular notch to the condylar top. The slice where the condyle had the largest cross-sectional area was measured and checked for correlation with atresia status, age, and sex. The Jahrsdoerfer score of the atretic ear was calculated and correlated with condyle cross-sectional area, age, and sex.

Results: Cross-sectional area of the condyle ipsilateral to the atretic ear was, on average, 8.41 mm² smaller than the contralateral condyle (P < .0001). The Jahrsdoerfer score was not associated with the condylar cross-sectional area, age, or sex.

Conclusion: Isolated nonsyndromic unilateral congenital aural atresia is associated with mild hypoplasia of the mandibular condyle ipsilateral to the atretic ear. This is consistent with the hypothesis that congenital aural atresia is a variant of craniofacial (hemifacial) microsomia.

Key Words: Aural atresia, mandibular condyle hypoplasia, craniofacial macrosomia.

Level of Evidence: 4.

INTRODUCTION

Children with congenital aural atresia (CAA) frequently have concomitant abnormalities of the temporal bone, middle ear, and associated structures.1-6 CAA is a component of numerous syndromes and also is seen in association with other developmental abnormalities of craniofacial structures, both unilaterally and bilaterally. However, little is known about how the mandible may be affected in children with isolated nonsyndromic CAA.

The mandible, external ear, and middle ear structures are all embryologic derivatives of the first and second pharyngeal arches.7 Given these shared embryologic origins, it is not surprising that syndromic unilateral CAA has been associated with other abnormalities of the ear and temporal bone, including microtia and ossicular malformations,8 facial nerve dysfunction, and sensorineural hearing loss2; posterior displacement of the temporomandibular joint9; and inferior displacement of the tegmen mastoideum.4 Although anomalies of the mandible coexist with CAA in children with craniofacial (hemifacial) microsomia,8 little is known about the nature and extent of mandibular involvement in children diagnosed with isolated nonsyndromic unilateral CAA.

We hypothesized that children with isolated nonsyndromic unilateral CAA have subclinical hypoplasia of the mandibular condyle on the side of the atresia, and that the degree of condylar hypoplasia is positively correlated with severity of middle ear malformation. The specific aims of this study were to 1) assess the cross-sectional area (CSA) of the mandibular condyle ipsilateral and contralateral to the atretic ear; 2) correlate the Jahrsdoerfer score of middle ear malformation with condylar area, expecting to find lower scores in condyles with smaller areas; 3) check for age and gender correlates with Jahrsdoerfer score; and 4) check age correlates of condylar area ipsilateral and contralateral to the atretic ear.

MATERIALS AND METHODS

The university institutional review board approved this retrospective self-controlled case series study of previously acquired computed tomography (CT) scans of the temporal bone in children with unilateral CAA.

Subjects

Study subjects were selected by identifying patients, age birth through 18 years of age, who underwent CT of the temporal bones for congenital aural atresia. The patients were identified through database query search of radiology reports utilizing

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108 children in the database who underwent computed tomography of the temporal bones for assessment of congenital aural atresia during the years 2002-2012.

Excluded:
- 20 subjects with bilateral aural atresia or stenosis
- 18 subjects with syndromic causes of aural atresia and/or other structural abnormalities or malformations of the head and neck
  - Treacher-Collins syndrome (n=4)
  - Craniofacial microsomia (n=4)
  - Goldenhar syndrome (n=3)
  - Nager syndrome (n=1)
  - CHARGE syndrome (n=1)
  - Crouzon syndrome (n=1)
  - Trisomy 13 (n=1)
  - Trisomy 18 (n=1)
  - Choanal atresia (n=1)
  - Skull base dysplasia (n=1)

70 children with images that could be studied.

Excluded:
- 1 subject without the regions of interest on images
- 1 subject with cholesteatoma

68 children included in the study:
- 22 males with right-sided atresia
- 14 males with left-sided atresia
- 25 females with right-sided atresia
- 7 females with left-sided atresia

Fig. 1. Subject selection and enrollment.

ISYS: desktop 6 (ISYS Search Software, Englewood, CO) at Children’s Healthcare of Atlanta for the years 2002 through 2012. Children were excluded from the study if: 1) aural atresia was bilateral; 2) diagnosed with a syndrome associated with aural atresia prior to the reference CT scan; or 3) another confounding structural abnormality of the head and neck was diagnosed prior to the reference CT scan or visible on the reference CT scan. Patients also were excluded if the imaging did not adequately show the regions of interest, or if only postsurgical images were available.

In total, 108 CT scans of children with CAA were identified. After exclusions, 68 patients with isolated unilateral CAA were included in our study (Fig. 1). The sample population was 53% males and the atretic ear was on the right side in 69% of patients. The mean age at the time of the CT scan was 62.0 ± 43.4 months, and ages ranged from under 1 month to 206 months.

**Technique for Mandibular Condyle Measurement**

We used the transverse (axial) plane perpendicular to the axis of the mandibular ramus and selected the slice where the condyle had the largest cross-sectional area (CSA). This was a consistent and easily reproducible method that eliminated the individual variability of external ear anatomy between patients. The condyle CSA served as a surrogate marker for the size of the mandibular condyle.

High-resolution CT images of the temporal bones were viewed at a display field of view of 6.5 cm using Centricity™ Universal Viewer software (GE Healthcare, Barrington, Illinois, U.S.A.), which also was used to measure the CSA. The CT slice thickness of the imaging series was 0.625 mm in 37 patients and 1.25 mm in the remaining 28. First, the right condyle was measured and then the left condyle. Each was scanned from the mandibular notch to the condylar top. The slice in which the condyle appeared largest was selected, and its CSA was measured from the outermost margin of the visible bony cortex (Fig. 2). In cases for which the largest slice was not immediately apparent, multiple slices were measured and the largest value recorded. The mandibular condyle on the side of the nonatretic ear was used as a comparison for each patient.

**Jahrsdoerfer Score Calculation**

The Jahrsdoerfer score (Table I) is a 10-point scale largely based on the interpretation of high-resolution CT images of the temporal bone. Higher scores correlate with an increased rate of success that atresiaplasty surgery will restore hearing to normal or near-normal levels. The score was used as a marker for the severity of middle ear malformation. Its calculation was performed by an attending pediatric neuroradiologist (B.P.S.) at an academic children’s hospital. The scorer was blinded to the size of the mandibular condyles, and the score was calculated for the atretic ear using the CT images only.

During evaluation of the facial nerve, the point was deducted only when the course of the nerve was deemed
sufficiently abnormal to increase the risk of injury at surgery (i.e., significant lateral and anterior displacement). The point was not deducted for very minor deviations in the course of the nerve. In this manner, we accounted for mild anatomic variations of the facial nerve course that almost universally are seen among patients with CAA. The score point for the external ear appearance was subtracted only if microtia or severe dysplasticity was obvious on imaging (i.e., no correlation with physical examination was involved).

**Data Analysis**

Continuous variables were summarized as mean ± standard deviation. CSA of the mandibular condyle ipsilateral to the aural atresia was compared to the contralateral normal condyle using the paired *t* test. Multiple linear regression with standard least square estimates was used to assess the impact of 1) age, sex, and Jahrsdoerfer score on the condyle CSA; and 2) age and sex on the Jahrsdoerfer score. Univariate tests were used to further assess relationships that were statistically significant. All tests used a *P* value of .05 as the threshold for significance. Analysis was performed using SAS JMP 12 software (2015; SAS Institute, Cary, NC).

**RESULTS**

Mandibular condyle CSA was highly correlated between the two sides. On average, the CSA of the condyle ipsilateral to the aural atresia was 8.41 mm$^2$ smaller than the antimeric condyle ($P < .001$, $R^2 = .797$) (Fig. 3). The average ratio of the condyle CSA of the affected side to the unaffected side was 0.91 ± 0.11 and was not impacted by age or sex ($P > .05$).

The mean Jahrsdoerfer score for atretic ears was 7.53 ± 2.85. The median score was 9 and ranged from 0 to 10. When controlling for age and sex, the Jahrsdoerfer score did not impact the condyle CSA ipsilateral to the atresia ($P = .97$). Because aural atresia rarely occurs in the absence of some degree of microtia, this analysis was repeated after deducting the point for external ear appearance. Mandibular condyle CSA was still significantly smaller on the affected side ($P < .001$, $R^2 = .797$).

**TABLE I. Jahrsdoerfer Criteria as Used in the Study.**

<table>
<thead>
<tr>
<th>Points</th>
<th>Parameter</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>Stapes present</td>
</tr>
<tr>
<td>1</td>
<td>Oval window open</td>
</tr>
<tr>
<td>1</td>
<td>Middle ear space large and favorable</td>
</tr>
<tr>
<td>1</td>
<td>Malleus-incus complex well formed</td>
</tr>
<tr>
<td>1</td>
<td>Mastoid well pneumatized</td>
</tr>
<tr>
<td>1</td>
<td>Incus and stapes connected</td>
</tr>
<tr>
<td>1</td>
<td>Round window present and open</td>
</tr>
<tr>
<td>1</td>
<td>Appearance of external ear is normal (by CT)</td>
</tr>
<tr>
<td>1</td>
<td>Facial nerve position</td>
</tr>
<tr>
<td>10</td>
<td>Total available points</td>
</tr>
</tbody>
</table>

Adapted with permission from Yeakley and Jahrsdoerfer.$^9$  
CT = computed tomography.
appearance from all patients. This reduced Jahrsdoerfer score also did not impact condyle CSA ($P = .91$). Consequently, we reduced the multiple linear regression model to assess the impact of age and sex on condyle CSA bilaterally.

Controlling for sex, condyle CSA ipsilateral to the aural atresia increased by 0.36 mm$^2$ with each additional month of age ($P < .001$) and 0.42 mm$^2$ per month on the contralateral normal side ($P < .001$). Age also was positively correlated with condyle CSA bilaterally on univariate analysis ($P < .001$) (Fig. 4). Controlling for age, condyle CSA in males was larger than females by 3.60 mm$^2$ ipsilateral to the aural atresia ($P = .04$) and 4.84 mm$^2$ on the contralateral normal side ($P = .01$). On univariate analysis using the unpaired two-sample $t$ test, the correlation between sex and condyle CSA was not statistically significant ipsilateral to the aural atresia ($t_{65} = 1.49, P = .14$) but trended toward significance in the contralateral normal condyle ($t_{65} = 1.84, P = .07$).

Neither age nor sex impacted the Jahrsdoerfer score when controlling for the other ($P = .62$ and $P = .35$, respectively).

**DISCUSSION**

Congenital aural atresia, in isolation or in association with other congenital craniofacial anomalies, has numerous consequences for affected individuals and presents a challenge for otologists. This study shows that CAA is associated with decreased CSA of the mandibular condyle on the side of the atresia, but that a lower Jahrsdoerfer score does not correlate with smaller condylar area. This suggests that CAA is associated with mild hypoplasia of the mandibular condyle, but that the degree of condyle hypoplasia is not associated with the severity of middle ear malformation.

The ear and mandible share embryologic origins. The first pharyngeal arch (Meckel’s) cartilage gives rise to the incus and malleus, whereas the surrounding mesenchymal tissue develops into the mandible. The stapes is a derivative of the second pharyngeal arch (Reichert’s) cartilage. Both the first and second pharyngeal arches contribute to the development of the auricle. The external auditory canal is a product of the first pharyngeal groove and forms by obliteration of tissue in a lateral-to-medial direction, ultimately terminating at the lateral surface of the tympanic membrane. Thus, embryologic development of the ear and mandible are intimately linked processes.

It then is perhaps surprising that no correlation was identified between the Jahrsdoerfer score and the size of the atretic condyle. Takano et al. found that several components of the Jahrsdoerfer score correlated with the degree of mandibular hypoplasia observed in a group of children with congenital microtia. However, this study included children with clinically obvious mandibular deformities in whom these differences were best appreciated. Therefore, the Jahrsdoerfer score likely correlates with the degree of mandibular hypoplasia across a broader range of mandibular deformities in the setting of congenital microtia or aural atresia, while being insensitive to subtle differences in condyle size like those presented here.

The condylar hypoplasia was seen across all ages, and the ratio of the CSA of the affected/unaffected sides...
was not correlated with age. These findings suggest that the mandibular hypoplasia was congenital and the relative severity of hypoplasia did not change over time. This is consistent with the idea that isolated unilateral CAA represents a mild variant of craniofacial microsomia in which mandibular hypoplasia is subclinical. The absence of a correlation between age and Jahrsdoerfer score implies that the severity of middle ear malformation is determined prior to birth. Although CAA is more prevalent in males, sex is not related to the severity of the malformation.

Congenital hypoplasia of the mandibular condyle, whether unilateral or bilateral, usually co-occurs with other malformations of first and second pharyngeal arch derivatives as part of a syndrome. Craniofacial microsomia is one such syndrome encompassing a wide range of malformations of the structural derivatives of the first and second pharyngeal arches, particularly the ear and mandible. Similarities between the epidemiologic characteristics and ear malformations of isolated microtia, CAA, and craniofacial microsomia—in addition to the shared embryologic origins of affected structures—have led to the suggestion that these may represent different manifestations of the same underlying disorder. Our findings further support this assertion.

We acknowledge several limitations to our study. First, the CSA of the mandibular condyle was measured under the assumption that it correlates with the condylar volume, but this was not directly demonstrated. It is not established whether this measure correlates with other morphologic or dynamic differences in the temporomandibular joint. Also, the Jahrsdoerfer score is inherently limited by its reliance on subjective assessments of particular anatomic structures. In order to minimize the variability that this may introduce, we applied the Jahrsdoerfer score in a strict and systematic fashion across all study subjects. No CT features have been shown to accurately predict increased risk of facial nerve injury during atresiaplasty surgery, and this issue was not specifically addressed in our study. However, we applied the Jahrsdoerfer score, deducting a point for “facial nerve position” when its course was significantly deviated and potentially resulting in a difficult surgical approach, as we explained in the Methods section. Our study did not address the important question of how much room there is between the condyle and the facial nerve, where the surgeon has to create the ear canal.

Whether these findings have clinical orthodontic implications remains undetermined. It is possible that over time even very mild unilateral mandibular hypoplasia could increase the risk for temporomandibular joint disorder via the slight biomechanical imbalance created by asymmetric condyles. In addition, given the disproportionate increase in airway resistance from small decreases in airway diameter, subclinical bilateral mandibular hypoplasia could increase the risk of sleep-disordered breathing and obstructive sleep apnea. The data presented here do not attempt to address these questions.

CONCLUSION
Our results show that patients with isolated nonsyndromic unilateral CAA have mild hypoplasia of the mandibular condyle ipsilateral to the atretic ear. These data support the assertion that CAA represents a manifestation of craniofacial microsomia in which mandibular hypoplasia is subclinical.

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BIBLIOGRAPHY