Correlation Between Microtia and Temporal Bone Malformation Evaluated Using Grading Systems

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Objective: To evaluate the relationships between temporal bone abnormalities and the severity of microtia in Japanese patients using objective grading systems.


Setting: Academic, tertiary care, referral medical center.

Patients: One hundred forty-two ears of 109 Japanese patients (85 male and 24 female patients; mean age, 12.8 years [range, 2-36 years]) with microtia.

Main Outcome Measures: The severity of microtia was classified according to Marx classification. Developmental abnormalities of the temporal bone were evaluated by a computed tomographic (CT) scoring system modified after the system used by Jahrsdoerfer and colleagues, using high-resolution CT scans of the temporal bone. Correlations between the scores obtained from these 2 grading systems were evaluated using a nonparametric statistical method.

Results: Male preponderance and incidence of bilateral cases of approximately 30% were observed in our Japanese patients with microtia. There was no significant difference in the severity of microtia between unilateral and bilateral cases. The mean ± SEM total points in the CT scoring system (full marks, 10) was 7.9 ± 0.4 for grade I microtia, 6.6 ± 0.6 for grade II, and 6.4 ± 0.3 for grade III; the total points correlated inversely with the microtia grade. Development of the auricle correlated significantly with aeration in the middle ear spaces but not with ossicular development or formation of the oval/round windows. Proportion of acceptable surgical candidates according to the CT scoring system (>5 points) was 79% for grade I microtia, 52% for grade II microtia, and 65% for grade III microtia.

Conclusion: The principle “the better developed the auricle, the better developed middle ear” was confirmed in Japanese patients with microtia; however, even with grade II/III microtia, more than half of the patients were considered suitable for atresia surgery.


The incidence of congenital aural atresia ranges from 1 in 10000 to 1 in 15000 births. Congenital aural atresia is a serious birth malformation in which the auricle, the external auditory canal (EAC), middle ear structures, and the inner ear may fail to develop. The degree of microtia ranges from smaller pinna with a normal shape to almost complete absence of pinna with rudimentary soft tissue. The state of the auricle can be classified into 3 types using the classification described by Marx. The EAC anomaly varies from slightly narrow canal to complete atresia. Abnormalities of the middle ear structure include stapes deformity, absence of oval and/or round windows, aberrant course of facial nerve, poor pneumatization of the middle ear space, and fusion of malleus and incus. The incidence of inner ear abnormalities associated with microtia is estimated between 10% and 47%.

Surgery for congenital aural atresia is one of the most challenging and difficult procedures in otology because this condition is often accompanied by various temporal bone anomalies, such as aberrant facial nerve, deformity of ossicles, defect of oval window, and lack of mastoid pneumatization. Preoperative high-resolution computed tomography (HRCT) is indispensable for the surgical planning because it provides important anatomical information. For determining good surgical candidacy, Jahrsdoerfer et al developed a grading system based on preoperative temporal bone HRCT and appearance of the auricle. This computed tomographic (CT) scoring system consists of 9 parameters related to temporal bone anatomy. The presence of a well-defined stapes scores 2 points, whereas all other parameters are assigned 1 point.
The subjects included 142 ears of 109 patients with microtia (85 male and 24 female patients; mean age, 12.8 years [range, 2-36 years]), who were seen between August 1992 and October 2003 and underwent HRCT examination of the temporal bone at the University of Tokyo Hospital, Tokyo, Japan. Those who had other anomalies associated with systemic syndromes, such as Treacher Collins and Goldenharr syndromes, were excluded from this study.

The severity of microtia was classified into grades I, II, or III according to the classification used by Marx.3 In brief, grade I microtia exhibits only mild deformity, with the auricle being only slightly smaller than normal, each part of which can be clearly distinguished. In grade II microtia, the size of the auricle is one half to two thirds of the normal size and its structure is only partially retained. In grade III microtia, the auricle is severely malformed and usually exhibits a peanut shape.

The HRCT images of the temporal bone were obtained using the Aquilion Multi CT system (Toshiba, Tokyo, Japan). Continuous slices of 1.0-mm thickness were obtained in both axial and coronal planes at 120 kV (peak) and 160 mA. The anomalies of the temporal bone were graded according to the CT scoring system used by Jahrsdoerfer and colleagues, with slight modification, in which the parameter “appearance of external ear” in the original grading system was replaced with the parameter “external ear canal present.” The original grading system used by Jahrsdoerfer and colleagues is useful for selection of candidates for atresia surgery. The relationship between the development of the auricle and the structures of the temporal bone was evaluated in the present study. The replacement of the parameter of the “external ear canal present” was more appropriate for this study because the parameter of “appearance of external ear,” which is one of the most important elements, was included in the Marx classification. If the external auditory ear was absent or not more than a small mound of skin and cartilage on the face (ie, grade III according to Marx classification) no point was assigned in the classification used by Jahrsdoerfer and colleagues). No other changes were made to the other parameters including their assigned scores.

The distribution of parameters of the CT scoring system was examined in relation to the grade of microtia. We also evaluated the correlation between the CT scoring and Marx classification. For correlational analyses, Spearman nonparametric rank correlation coefficient was calculated for Marx classification and total scores of the CT scoring (full mark, 10 points) and between Marx classification and the following 3 subtotal scores reflecting specific development: (1) subtotal of parameters related to ossicular development (sum of “stapes present,” “malleus/incus complex,” and “incudostapedial connection”; full mark, 4 points); (2) subtotal of parameters related to windows connected to the cochlea (sum of “oval window open” and “round window”; full mark, 2 points); and (3) subtotal of parameters related to aeration of the middle ear cavity (sum of “middle ear space” and “mastoid pneumatization”; full mark, 2 points).

Table 1 gives the distribution of microtic ears according to the side and severity as classified using the Marx classification.3 The distribution according to the side and sex is given in Table 2. There was no significant difference in the severity of microtia between unilateral and bilateral cases (t test, 2-tailed). Male patients were predominant in our sample. Moreover, there was no significant difference in the severity of microtia between male and female patients. The inner ear anomalies such as agenesis of the cochlea and the enlarged vestibular aqueduct were not found in these patients.

Table 3 demonstrates the distribution of parameters of the CT scoring system in relation to the grade of microtia. Correlations with Marx classification were evalu-
ated in total points and 3 subtotal points related to ossicles, windows open to the inner ear, and aeration of middle ear (Table 4).

Total points correlated significantly with the microtia grade, indicating that better developed auricles have better developed middle ear spaces that are more suitable for surgery. For comparison between microtia grades, 1-way analysis of variance showed a significant difference between mean values by grades \( (P = .02) \), and the Sheffe test, a post hoc test suitable for multiple comparison, showed a significant difference between grades I and III \( (P = .02) \). The differences between the other pairs (grades I and II, \( P = .23 \); grades II and III, \( P = .91 \)) failed to reach statistical significance.

With regard to the 3 subtotal points, only the sum points of aeration of the middle ear correlated significantly with the microtia grade. One-way analysis of variance for intergrade comparison revealed significant difference only with sum points regarding aeration \( (P = .02) \) and not with sum points regarding ossicles \( (P = .29) \) or windows \( (P = .34) \). In the intergrade comparison for sum points regarding aeration, the Sheffe test showed significant difference only between grades I and III \( (P = .03) \) but not between the other pairs (grades I and II, \( P = .22 \); grades II and III, \( P = .96 \)). These results indicate that the difficulty of ossicular reconstructive surgery, which is closely related to the existence of oval/round windows and deformity of ossicles, does not necessarily depend on the microtia grade.

The proportion of candidates suitable for surgery according to the CT scoring system \((>5 \text{ points})\) was 79% for grade I microtia, 52% for grade II microtia, and 65% for grade III microtia.

**COMMENT**

Microtia has been reported to occur predominantly in male individuals (male-female ratio, 2:1). Furthermore, the incidence of bilateral microtia is reported at 10% to 30%, with right ear involvement in 55% to 65% of unilateral cases.\(^1\)\(^6\)\(^16\)\(^17\) The findings of the present study in Japanese individuals were similar to the aforementioned data (male-female ratio, 4:1; incidence of bilateral case, 30%; and incidence of right-sidedness in unilateral cases, 51%).

The severity of microtia was not different between unilateral and bilateral cases.

A previous study reported that the average atresia score according to CT grading system correlates with the severity of microtia,\(^10\) in which the average atresia score was 8.5 in grade I microtia, 7.2 in grade II microtia, and 5.9 in grade III microtia. Similarly, in our patients, total points of the CT scoring system correlated inversely with the severity of microtia. These results support the principle

### Table 3. Distribution of Each Parameter of the Computed Tomographic Scoring System*\(^a\)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Assigned</th>
<th>Marx Classification, %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Point</td>
<td>I</td>
</tr>
<tr>
<td>Related to ossicles</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stapes present</td>
<td>2</td>
<td>70</td>
</tr>
<tr>
<td>Malleus/incus complex</td>
<td>1</td>
<td>94</td>
</tr>
<tr>
<td>Incudostapedial connection</td>
<td>1</td>
<td>67</td>
</tr>
<tr>
<td>Related to the window’s connection to the cochlea</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
<td>85</td>
</tr>
<tr>
<td>Round window</td>
<td>1</td>
<td>94</td>
</tr>
<tr>
<td>Related to aeration development of middle ear</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Middle ear space</td>
<td>1</td>
<td>100</td>
</tr>
<tr>
<td>Mastoid pneumatization</td>
<td>1</td>
<td>85</td>
</tr>
<tr>
<td>Facial nerve</td>
<td>1</td>
<td>64</td>
</tr>
<tr>
<td>External ear canal present</td>
<td>1</td>
<td>58</td>
</tr>
</tbody>
</table>

*Modified after the system used by Jahrsdoerfer and colleagues\(^8\) in relation to the Marx classification.\(^3\)

†\(P < .001\).

### Table 4. Correlation Between Total/Subtotal Points and the Marx\(^3\) Classification*

<table>
<thead>
<tr>
<th>Variable</th>
<th>Points, Full Mark</th>
<th>Marx Classification</th>
<th>(r)</th>
<th>(P) Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total points</td>
<td>10</td>
<td>7.85</td>
<td>6.62</td>
<td>6.35</td>
</tr>
<tr>
<td>Subtotal points related to development of ossicles, windows, and aeration</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ossicles</td>
<td>4</td>
<td>3.00</td>
<td>2.52</td>
<td>2.53</td>
</tr>
<tr>
<td>Windows</td>
<td>2</td>
<td>1.79</td>
<td>1.67</td>
<td>1.61</td>
</tr>
<tr>
<td>Aeration</td>
<td>2</td>
<td>1.85</td>
<td>1.52</td>
<td>1.48</td>
</tr>
</tbody>
</table>

*Data are given as average points and Spearman rank correlation coefficient \((r)\) with \(P\) values.
“the better developed the auricle, the better developed middle ear.”

To our knowledge, the correlation between microtia grades and CT parameters (subtotals) regarding development of specific components of the middle ear has not been reported previously. Our study indicates that auricular development correlated significantly with the aeration in the middle ear spaces but not with the ossicular development or formation of the oval/round window. The auricle, middle ear epithelium lining the air cells, ossicles, and stapes footplate tapping the oval window are thought to arise from neural crest cells of the first branchial arch, endodermal cells of the first branchial arch, neural crest cells of the first and second branchial arches, and mesodermal cells of the otic capsule, respectively. Differences in correlation among specific components may reflect differences in their origin during development.

Takegoshi and colleagues performed a similar study on patients with mandibulofacial dysostosis and found a positive correlation between attic formation and microtia severity. The antrum and the mastoid air cells were absent in the patients with mandibulofacial dysostosis. In our study, the mastoid air cells were not necessarily absent. The mastoid pneumatization were 85% in grade I, 62% in grade II, and 63% in grade III in these patients with microtia, except for those with systemic syndromes.

Microtia is usually accompanied by atresia or stenosis of the EAC. Abnormal EACs can be classified into 3 types: almost normal; narrowing of the fibrocartilaginous canal; and narrowing and tortuosity of both the fibrocartilaginous and bony parts of the canal. Surgical reconstruction of the EAC and ossicular chain is much easier when the EAC is present, even when it is very narrow and tortuous, because this acts as a landmark to help the surgeon to reach the tympanic cavity more safely. In addition, the presence of the EAC was very closely related to the formation of the manubrium. The presence of the manubrium helps to reconstruct the tympanic membrane in its original position. These were the reasons why we replaced the parameter of “appearance of the auricle” in the original scoring system used by Jahrsdoerfer and colleagues with “existence of EAC.” As is evident from the distribution shown in Table 3, the parameter “existence of EAC” correlated significantly with the microtia grade (P<.001), confirming that replacement of the parameter in the overall scoring system would be beneficial in determining the candidacy for atresia surgery.

In conclusion, the principle “the better developed the auricle, the better developed middle ear” was confirmed in Japanese cases of microtia; however, even with grade III microtia, more than half of the patients were acceptable candidates for atresia surgery. Auricular development correlates significantly with aeration in the middle ear spaces.

Submitted for Publication: August 10, 2004; accepted December 22, 2004.

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REFERENCES


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