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Atlantoaxial Rotatory Fixation after Microtia Reconstruction Surgery

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Background: Nontraumatic atlantoaxial rotatory fixation after microtia reconstruction surgery is a rare complication. Intraoperative cervical hyperextension and/or excessive rotation and postoperative inflammation have been reported as causes of atlantoaxial rotatory fixation. We herein describe cases of atlantoaxial rotatory fixation after microtia reconstruction surgery.
Methods: This was a retrospective study of 80 patients (165 surgeries) who underwent microtia reconstruction surgery in Dokkyo Medical University Hospital between April 2006 and December 2012. The patient- and operation-related variables were obtained from medical charts. Neck radiographs and computed tomography scans of patients with atlantoaxial rotatory fixation were evaluated to check for cervical spine abnormalities.
Results: Five cases of atlantoaxial rotatory fixation after microtia reconstruction surgery were recorded. Three of these five cases were diagnosed with Klippel-Feil syndrome after the onset of atlantoaxial rotatory fixation. No significant difference was found in the operative duration and other variables between patients with atlantoaxial rotatory fixation and those without. All patients immediately underwent conservative treatment and showed complete recovery and no recurrences.
Conclusions: Although atlantoaxial rotatory fixation is a rare complication, surgeons should consider it in patients with neck problems following microtia reconstruction surgery. A patient with microtia may have unrecognized Klippel-Feil syndrome. Patients with Klippel-Feil syndrome are more likely to develop atlantoaxial rotatory fixation, which may have severe consequences. Thus, it is crucial to preoperatively identify Klippel-Feil syndrome with neck radiography and to detect atlantoaxial rotatory fixation at the earliest. *(Plast Reconstr Surg Glob Open 2021;9:e3760; doi: 10.1097/GOX.0000000000003760; Published online 23 August 2021.)*

INTRODUCTION

Grisel syndrome, a nontraumatic atlantoaxial rotatory fixation, is a rare condition in children.[1–3] Most atlantoaxial rotatory fixations occur following an upper respiratory tract infection or otolaryngological procedure.[4]

Atlantoaxial rotatory fixation after microtia reconstruction surgery is thought to occur following prolonged and/or excessive rotation and/or hyperextension of the head during the surgical procedure; however, the contribution of Klippel-Feil syndrome to atlantoaxial rotatory fixation after otoplasty surgery remains unknown. Herein, we described five patients with atlantoaxial rotatory fixation following microtia reconstruction surgery. Three patients were postoperatively diagnosed with Klippel-Feil syndrome, defined as a congenital cervical spine fusion. We proposed that not only intraoperative cervical hyperextension and/or excessive rotation but also unrecognized Klippel-Feil syndrome may cause atlantoaxial rotatory fixation after microtia reconstruction surgery.

MATERIALS AND METHODS

This study was approved by the ethics committee of Dokkyo Medical University Hospital (review no. R-38-7J). All patients and their parents were provided with verbal and written information about the study before obtaining their written consent for participation.

In our institution, we perform microtia reconstruction surgery in two stages. The first stage includes otoplasty with rib cartilage, and the second stage involves ear elevation with or without tympanoplasty.[5,6] A retrospective

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Patient	Status of Microtia	Other Medical Comorbidities	Age (year)	Gender	Stage of Surgery	Timing of AARF Diagnosis	Treatment for AARF	Klippel-Feil syndrome
1	Left	VSD Anal Atresia	11	Boy	1st Stage	POD 6	Cervical Collar, Traction for 2 weeks	+
2	Left	None	10	Boy	2nd Stage	POD 3	Cervical Collar	–
3	Bilateral	Moebius syndrome	11	Boy	2nd Stage (left)	POD 6	Cervical Collar	+
4	Right	First and second brachial arch syndrome	9	Girl	1st Stage	POD 7	Cervical Collar, Traction for 1 week	+
5	Right	None	10	Girl	2nd Stage	POD 3	Cervical Collar	–

Fig. 1. Patient characteristics. VSD, ventricular septal defect; AARF, nontraumatic atlantoaxial rotatory fixation.

chart review of data of the patients who underwent microtia reconstruction surgery (first or second stage) between April 2006 and December 2012 was completed. Because the susceptible age of atlantoaxial rotatory fixation is less than 12 years, patients aged 12 or more years at the first-stage surgery were excluded from this study. The preoperative patient demographic variables including sex, condition of microtia, medical comorbidities, surgery-related variables of the stage of surgery (first or second), and duration of the operation were obtained from the medical charts.

Symptomatic microtia and other congenital anomalies (eg, cardiac, gastrointestinal, and urological anomalies) were considered medical comorbidities, except for accessory ear. Neck radiographs and computed tomography scans of patients with atlantoaxial rotatory fixation were evaluated for cervical spine abnormalities.

For all statistical analyses, SPSS software (IBM Corporation, Tokyo, Japan) was used. Fisher exact test was used to assess the effect of sex, condition of microtia, medical comorbidities, and stage of surgery on the risk of atlantoaxial rotatory fixation. Mann-Whitney U test was used to analyze the relationship between the operative duration and the risk of atlantoaxial rotatory fixation.

RESULTS

In total, 80 patients, representing 165 surgeries, were analyzed. Five patients developed atlantoaxial rotatory fixation, indicating a 3% incidence rate. Details of five patients with atlantoaxial rotatory fixation are summarized in Figure 1. Three of five patients were postoperatively diagnosed with Klippel-Feil syndrome. The incidence of Klippel-Feil syndrome among patients with microtia and patients with microtia and medical comorbidities were 3.75% and 20%, respectively. No significant difference was

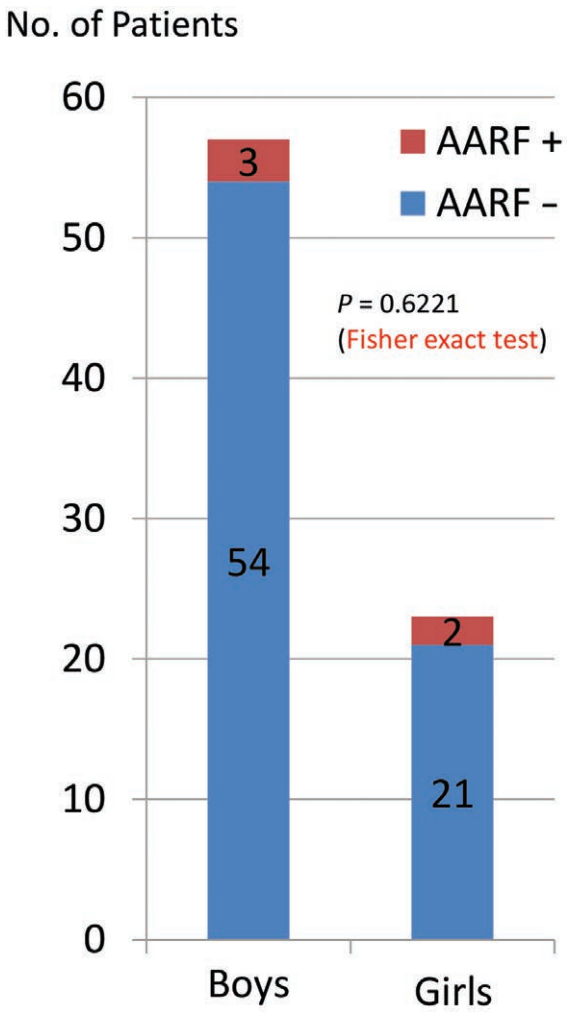

Fig. 2. Effect of sex on AARF.

found in the following variables: sex, condition of microtia, stage of surgery, and operative duration (Figs. 2–5). However, the presence of medical comorbidities showed a significant correlation with atlantoaxial rotatory fixation ($P = .0432$) (Fig. 6).

DISCUSSION

Nontraumatic atlantoaxial rotatory fixation, or Grisel syndrome, causes a painful torticollis (cock robin posture) and is predominantly seen in pediatric patients. Atlantoaxial rotatory fixation occurs after head and neck infection or following routine otolaryngological procedures in children. Although the mechanism of atlantoaxial rotatory fixation is unknown, anatomical features of the spine in children and inflammation play a major role in atlantoaxial rotatory fixation. The transverse ligament of the atlas mainly contributes to the stability of the C1–C2 joint.[7] The pharynx is anatomically adjacent to the C1–C2 joint and shares lymphatic drainage with it. Any inflammation in the pharynx may spread to the C1–C2 joint and result in laxity of the transverse ligament.[7]

Atlantoaxial rotatory fixation has been reported as a postoperative complication of surgery for congenital ear deformities; however, the contribution of Klippel-Feil syndrome to atlantoaxial rotatory fixation after otoplasty surgery has not been reported.[8–14] Intraoperative cervical hyperextension and/or excessive rotation and perioperative inflammation are stated as the cause of atlantoaxial rotatory fixation after otoplasty. In two of our five patients who did not have Klippel-Feil syndrome, the cause of atlantoaxial rotatory fixation appeared to be the same as that suggested in previous reports; however, in the other three who had Klippel-Feil syndrome, this syndrome appeared to be the cause of their developing atlantoaxial rotatory fixation. Patients with cervical spine abnormalities such as Klippel-Feil syndrome, Down syndrome, and Marfan syndrome are at a high risk of developing atlantoaxial rotatory fixation and suffering severe consequences.[15] None of the three patients with Klippel-Feil syndrome were recognized preoperatively as having this syndrome.

Klippel-Feil syndrome is a congenital bone disorder characterized by the abnormal fusion of two or more cervical vertebrae. However, less than 50% of patients with Klippel-Feil syndrome manifest all three of the classic signs: short neck, low hairline, and limited range of motion of the neck. Klippel-Feil syndrome is estimated to

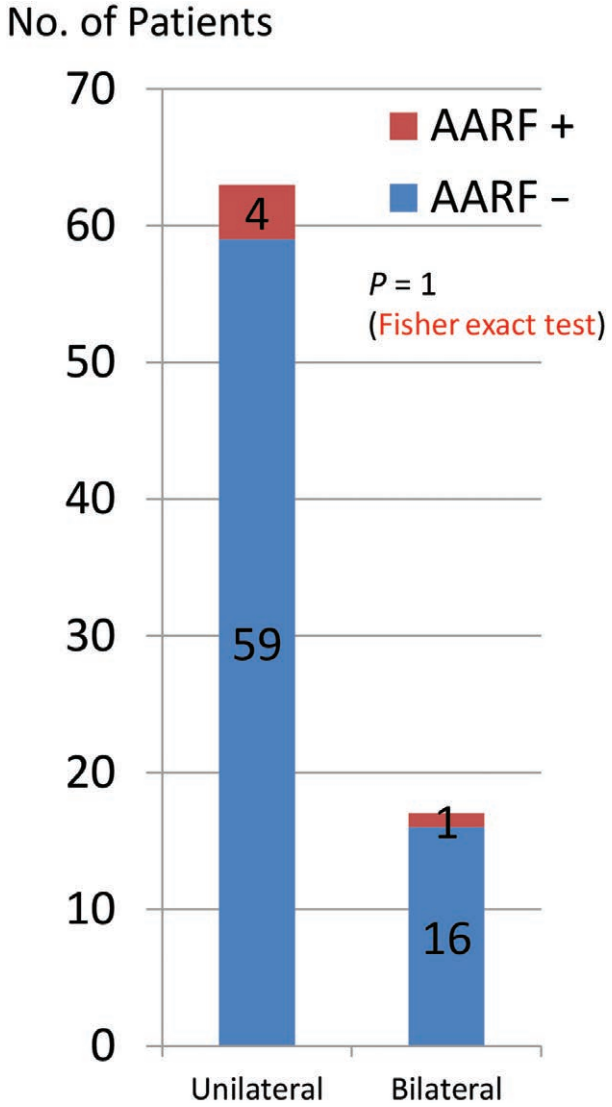

Fig. 3. Effect of the condition on AARF.

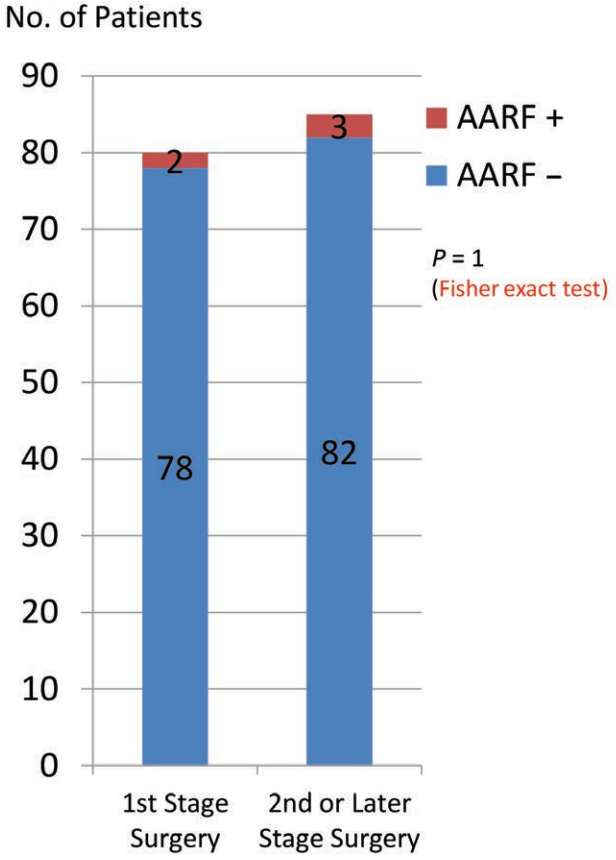

Fig. 4. Effect of the stage of surgery on AARF.

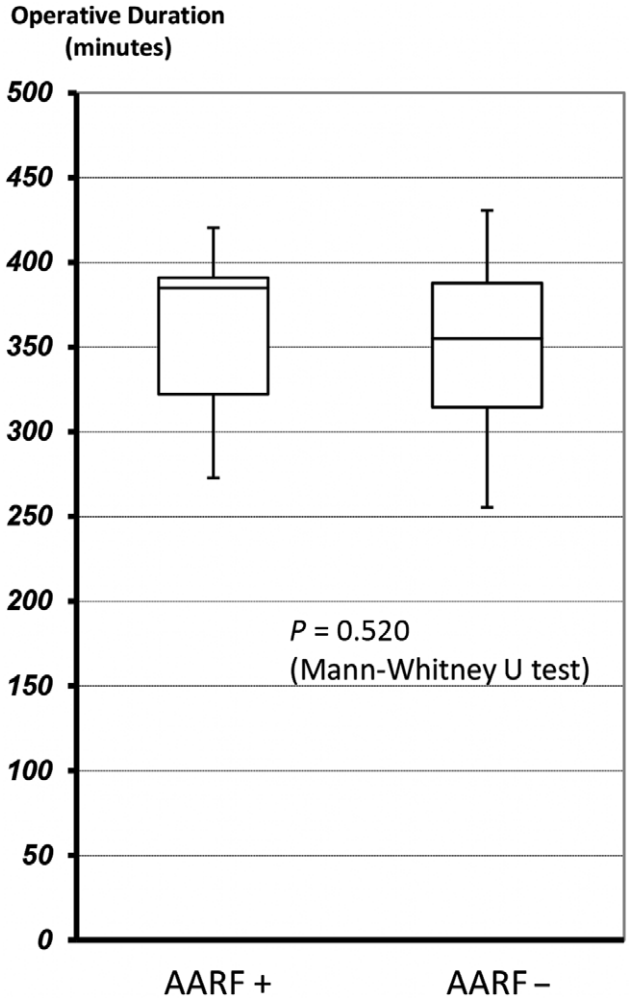

Fig. 5. Effect of operative duration on AARF.

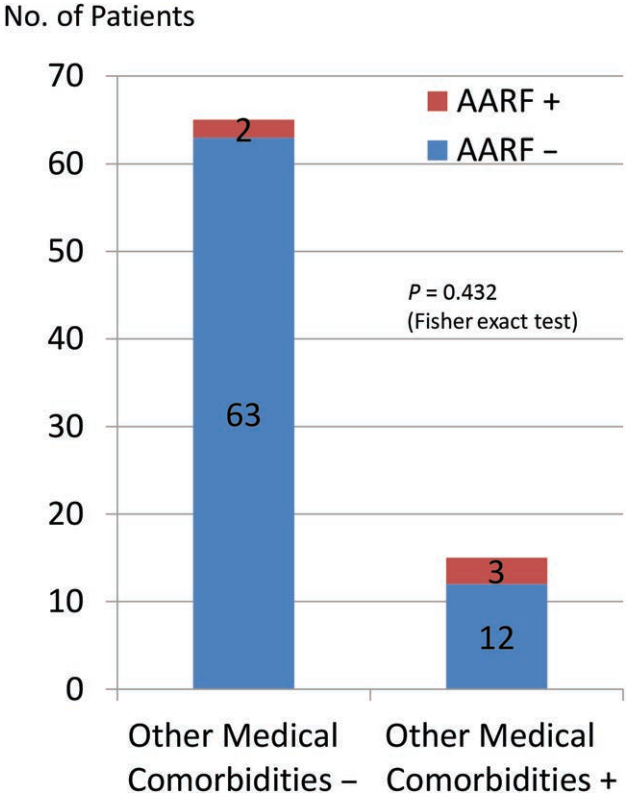

Fig. 6. Effect of other medical comorbidities on AARF.

occur in one in 42,000 births. Its common complications include skeletal abnormalities, renal and genitourinary abnormalities (25%–35%), central nervous system abnormalities (eg, deafness and psychomotor retardation) (12%–20%), cardiovascular abnormalities (3.5%–14%), and cleft lip and palate (10%).[16–20] Additionally, auricular deformities are a known comorbidity, although their prevalence has not been reported.

In this study, three (3.75%) of 80 patients were found to have Klippel-Feil syndrome, and the prevalence of Klippel-Feil syndrome in patients with microtia appeared to be higher than that in the general population. In addition, 20% (3 of 15) of the patients with medical comorbidities had Klippel-Feil syndrome. In some patients, microtia and congenital cervical spine fusion may be seen as part of a group of a few syndromes, such as Wildervanck, Duane, and Goldenhar syndromes. However, none of the three patients met the criteria for those syndromes. The prevalence of Klippel-Feil syndrome might be higher in patients with microtia, especially those with medical comorbidities, than in the general population. Because patients with Klippel-Feil syndrome have a high risk of developing atlantoaxial rotatory fixation and its severe consequences, we suggest that patients with microtia and medical comorbidities should be evaluated for any cervical spine abnormalities with neck radiography to preoperatively recognize whether they have Klippel-Feil syndrome. If we were aware of the presence of this syndrome, we could pay meticulous attention to the neck position during surgery, which could result in the early detection of atlantoaxial rotatory fixation. Conservative treatment (cervical collar and traction) should be started within 4 weeks of onset, because the delay in initiating treatment might result in recurrence or surgery.[11,21,22] All five patients with atlantoaxial rotatory fixation improved with conservative treatment and have shown no recurrence to date. Currently, in our institution, we routinely take preoperative cervical spine radiographs of patients with bilateral microtia, as well as those with microtia and other medical comorbidities. We recommend that surgeons take preoperative cervical spine radiographs of such patients and take utmost care of patients' cervical position during surgery to provide early detection and treatment for atlantoaxial rotatory fixation.

CONCLUSIONS

We reported five cases of postoperative atlantoaxial rotatory fixation in patients with microtia. From a retrospective review of medical charts, the prevalence of Klippel-Feil syndrome is higher in patients with microtia, especially those with microtia and other medical comorbidities, than in the general population. Further research is necessary to confirm this possibility.

Surgeons who perform microtia reconstructive surgery should keep in mind the possibility of Klippel-Feil syndrome and pay attention to whether the patients have atlantoaxial rotatory fixation to avoid its severe consequences.

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