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 microtia reconstruction surgery remains unknown. Herein, we described five patients with atlantoaxial rotatory fixation after microtia reconstruction surgery. Three patients were postoperatively 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. 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.)
A 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 (e.g., 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 observed.

![Patient characteristics.](image1)

### No. of Patients

| 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.

![Effect of sex on AARF.](image2)

Fig. 2. Effect of sex on AARF.

\[ P = 0.6221 \] (Fisher exact test)
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. 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

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**Fig. 3.** Effect of the condition on AARF.

**Fig. 4.** Effect of the stage of surgery 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%). 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.

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.

**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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