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## Case Report

# A Case of Congenital Midline Fistula of the Upper Lip

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## Abstract

Congenital fistulas of the lip are commonly found in the lower lip and accompany cleft lip. They are seen as a symptom of Van der Woude syndrome, which is predominantly hereditary. In contrast, congenital fistulas of the upper lip are rare. A number of hypotheses have been proposed to explain the pathogenesis of fistulas of the upper lip, including fusion failure of facial prominences and absence of mesoblasts, suggesting a relationship between this condition and the development of cleft lip. The pathogenesis of this disorder has been attracting attention. We report the case of a 5-year-old girl with congenital fistula of the upper lip.

**Key words:** Congenital fistula—Midline fistula—Sinus—Upper lip—Lip pit

## Case

The patient was a 5-year-old girl who visited our hospital with the chief complaint of swelling of the upper lip at the midline accompanied by discharge of pus. A fistula had been noted in the upper lip at the midline at birth, and the patient had repeatedly experienced swelling and discharge of pus from the upper lip (Fig. 1). Prior to presenting at this hospital, the patient had fallen over and hit her upper lip, resulting in marked swelling and discharge of pus from the fistula. The patient visited a dental practitioner and was referred to our department after anti-inflammatory treatment. The medical histories of the patient and her

family showed nothing remarkable. Present illness: A 0.5 × 0.5-mm fistula was observed 2 mm below the base of the philtrum. Swelling and redness were observed in the upper labial frenulum.

**Treatment and prognosis:** The fistulous tract was removed under general anesthesia. A bougie was inserted into the lacrimal duct about 18 mm from the fistula (Fig. 2). The fistulous tract terminated in the upper labial frenulum. The skin surrounding the fistula was dissected in a spindle shape, and the fistulous tract was explored towards the anterior nasal spine with the lacrimal duct bougie. The tract and upper labial frenulum were resected and the fistula removed (Fig. 3). No postoperative



Fig. 1 The congenital midline fistula of the upper lip

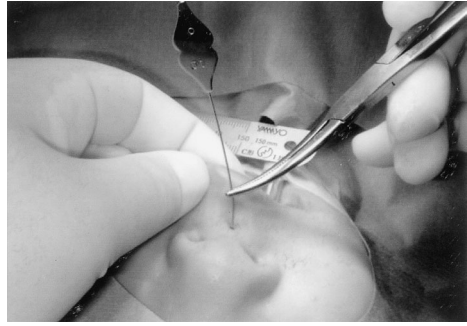


Fig. 2 A bougie was inserted into the lacrimal duct about 18mm from the fistula

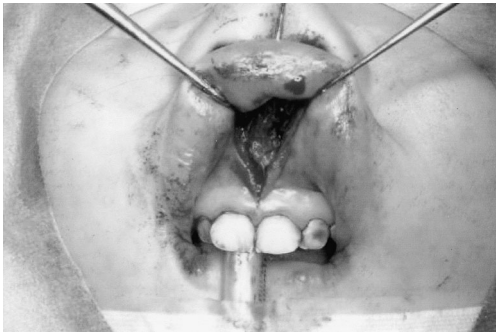


Fig. 3 The tract and upper labial frenulum were resected and the fistula removed



Fig. 4 No postoperative recurrence was noted at her one-year follow-up examination

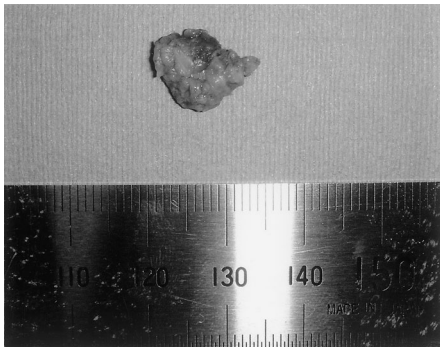


Fig. 5 Findings on resected section  $2 \times 5 \times 15$  mm

The resected sample contained tissues from the midline of the upper lip. A fistulous tract was observed in the center of the sample, consisting of fibrous connective tissue covered with cornified stratified squamous epithelium (Fig. 6). Dermal appendages such as sebaceous glands and hair follicles were found beneath the lining epithelium. An ulcer had formed where the epithelium was missing. Bleeding, capillary vessel growth and enlargement, and inflammatory cell infiltration including lymphocytes and plasma cells were observed.

recurrence was noted at her one-year follow-up examination (Fig. 4).

The resected sample ( $2 \times 5 \times 15$  mm) is shown in Fig. 5.

## Discussion

Lannelongue and Menard<sup>11)</sup> reported the first case of congenital midline fistula of the upper lip in 1891, and approximately 40 cases



Fig. 6 The fistulous tract itself, consisting of fibrous connective tissue covered with cornified stratified squamous epithelium, was observed in the center of the sample (HE)

have been reported outside Japan up to the present time. The incidence of congenital midline fistula of the lower lip is only 0.001%, and that of the upper lip is even lower. In Japan, the first case of congenital midline fistula of the upper lip was reported by Tange<sup>17)</sup> in 1965. So far, 20 more cases have been reported domestically, including two reports from our department (Koeda *et al.* in 1995<sup>9)</sup> and the present case).

The locations of the previously reported midline fistulas are shown in Fig. 7. All treatments performed included surgical resection of the fistula and removal of the fistulous tract. Postoperative prognoses were good and no esthetic problems occurred.

Complications included midline upper cleft lip<sup>4,11,15,18)</sup>, abnormalities of the maxillary labial frenum<sup>16,18)</sup>, midline nasal fistula<sup>18)</sup>, cleft alveolus<sup>4)</sup>, cleft uvula<sup>4)</sup>, posterior nasal spine defect<sup>4)</sup> and Pierre-Robin's syndrome<sup>18)</sup>.

A range of hypotheses have been put forward to explain the pathogenesis of congenital midline fistula of the upper lip, including the fusion of facial prominences theory<sup>10,19)</sup>, the merging of mesoblasts theory, and the invasion of abnormal epithelium theory<sup>1)</sup>. A strong association has been suggested between midline cleft lip<sup>4,11,15,18)</sup> and congenital midline fistula of the upper lip. One theory, in particular, suggests that midline fistula of the

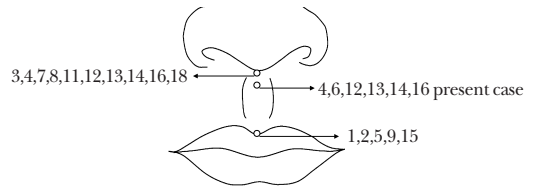


Fig. 7 The circles indicate the reported location of upper lip fistula in references  
The numbers indicate those of references.

upper lip is caused by planarization between the bilateral medial nasal processes due to a developmental problem of the front nasal prominence (medial nasal process) during the first 4–8 weeks of embryonic life, which would explain why fistulas are often formed at the midline and accompany midline cleft lip of the incomplete type due to problems occurring during the developmental process of the upper lip. No genetic tendency has been observed in this disorder, in contrast to with angulus or lower lip fistulas.

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