Endoscopic Release of Intraoral Synechiae in Popliteal Pterygium Syndrome

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An infant with typical popliteal pterygium syndrome had intraoral fibrous bands binding the maxillary and mandibular alveolar ridges. This resulted in dramatically restricted mouth opening. These bands were divided surgically with endoscopic assistance.

Schonauer F, La Rusca I, Sordino D, Settimi A, Molea G. Endoscopic release of intraoral synechiae in popliteal pterygium syndrome. Ann Plast Surg 2002;49:XXX–XXX

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Received Jan 24, 2002, and in revised form Apr 11, 2002. Accepted for publication Apr 11, 2002.

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Popliteal pterygium syndrome (or facial genitopopliteal syndrome) is characterized by any combination of deformities, including popliteal webs and craniofacial, genitourinary, and extremity anomalies. Oral membranes or fibrous bands may be present in addition to the typical webs between the ischium and the calcaneum. We describe a new case in which intraoral bands were divided endoscopically soon after birth.

Patient Report

A 3,150-g white female infant was transferred from a district hospital on day 10 after birth. The main findings in this infant were bilateral popliteal webs extending from the ischium to the calcaneum that limited severely the extension of the knee joints, and bilateral cleft lip and left unilateral cleft palate. Both active and passive mouth opening was impossible. A large oval lip pit was observed in the center of the lower lip (Fig 1). Other anomalies such as incomplete bilateral cutaneous syndactylies between the fourth and fifth toes and genitourinary anomalies (hyperpigmented hypertrophic clitoris and hypoplastic labia majora) were present. Feeding was possible exclusively via nasoenteral tube. Radiographic examination demonstrated a bilateral curved tibia. Temporomandibular joint views, voiding cystourethrography, and chromosome studies were normal. AQ: 1

Nasotracheal intubation was achieved and surgery was performed on day 22. Intraoral anatomy was investigated with a 3-mm endoscope (Storz). Bilateral fibrous bands (1.7 cm long with a base of 0.5 cm) were seen binding the maxillary and mandibular alveolar ridges posteriorly (Fig 2). F2 These pillars were divided sharply under direct endoscopic vision, and bipolar hemostasis was performed (Fig 3). Immediate release of the temporomandibular joint was achieved (Fig 4). F3 F4

During the same surgical session, the left leg was operated. The popliteal web was released with a double-opposing Z-plasty, and the sciatic nerve was divided and elongated with a graft of homolateral sural nerve divided into three cables. The right leg was operated at 2 months in the same way. The cleft lip was corrected at 3 months and the cleft palate was corrected at 6 months.

Discussion

Our case is a typical example of popliteal pterygium syndrome with facial anomalies (a large sinus of the lower lip, bilateral cleft lip and unilateral cleft palate, intraoral bands).[1] A review of the literature suggests that this malformation may represent an autosomal dominant condition with variable expression. The patient's family history did not reveal such a finding. However, we cannot rule out this possibility entirely be-

DOI: 10.1097/01.SAP.0000020050.55576.9C

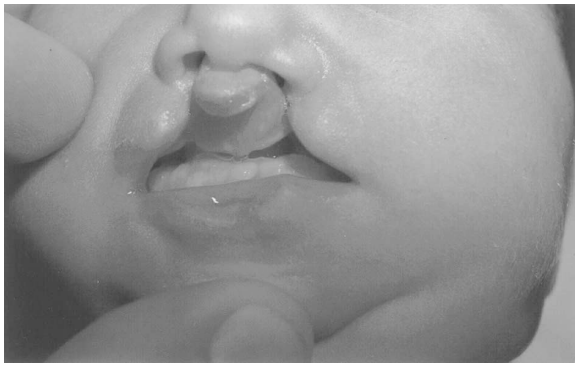

Fig 1. Facial abnormalities and restricted mouth opening.

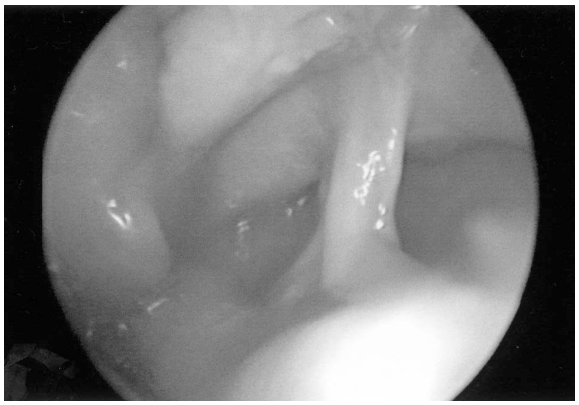

Fig 2. Endoscopic view of the intraoral synechiae.

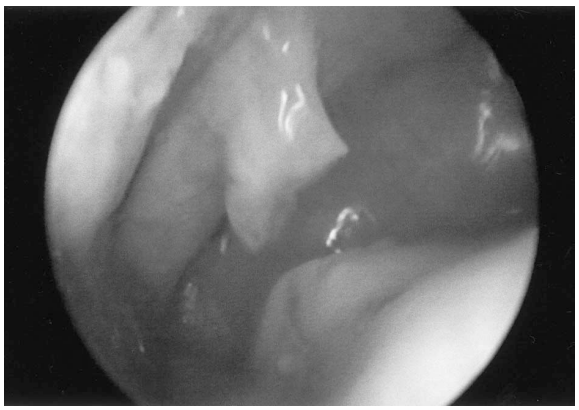

Fig 3. Division of the synechiae.

cause not all family members were available for direct questioning.

Hetch and Jarvinen[2] first described a patient in who laterally based oral webs extended from the maxillary tuberosity to the mandible. Wynne and

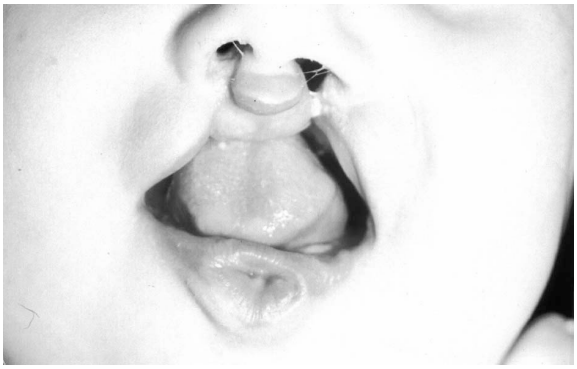

Fig 4. Postoperative mouth opening.

colleagues,[3] in 1982, described a patient with a sail-like membrane that extended from the premaxilla to the floor of the mouth, which was divided surgically soon after birth.

Congenital intraoral bands may consist of epithelium supported by varying amounts of connective tissue. If they just include soft tissues they are called *synechiae*; if bone is present the condition is defined as *syngnathia*.[4] Proposed embryological explanations for synechia formation are either failure of division (persistent oropharyngeal membrane) or abnormal epithelial behavior.[5]

These bands have been corrected without anesthetic in the nursery using surgical instruments[6] or silk ligatures,[5] under local anesthesia with a nasopharyngeal airway by division with scissors,[7] divided surgically under mask ventilation and oxygenation on the second day of life after failed attempts at nasotracheal intubation,[8] and again with oxygen by mask under direct vision with insulated needle tip cautery.[9] In another patient a tracheostomy was performed at 1 week of age, followed by division of the bands with a scalpel.[10]

The first problem with this condition is placing an endotracheal tube. We recommend a setting with optimal anesthesia equipment, including a fiberoptic endoscope. When general anesthesia is achieved, division of the intraoral bands can be performed easily. AQ: 2

We report the first patient in whom posterior oral fibrous bands, neither visible nor approachable via an open view, were diagnosed and then divided surgically under endoscopic direct vision. We recommend this simple, safe, and direct

approach to detect and correct intraoral anomalies that are limiting suction or oral feeding.

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