Clinical Brief

Congenital Fusion of the Jaws

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ABSTRACT

Congenital fusion of the jaws is quite rare. It may be unilateral or bilateral, and involves only the soft tissues or both the hard and soft tissues. This anomaly may be seen separately or in association with syndromes. Maxillomandibular fusion restricts mouth opening causing problems in feeding, swallowing, and respiration. This condition can be easily treated. However, in long-standing cases, growth anomalies from TMJ ankylosis may occur.
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Congenital maxillomandibular fusion is a rare deformity seen in infants which often presents as part of a syndrome. Non-syndromic cases of jaw fusion are very rare. Fusion often involves the oral soft tissues but may involve both hard and soft tissues preventing mouth opening and feeding resulting in inability to thrive. Early treatment is indicated as long-standing cases are complicated by ankylosis of the temporomandibular joint (TMJ). Isolated cases of jaw fusion are sparse. The authors found only 10 cases in the literature. Here is reported our third case of congenital maxillomandibular fusion in an infant girl with description of the clinical features and treatment adding to the existing amount of literature on the subject.

CASE REPORT

A 2-month-old baby girl suffering from inability to open her mouth since birth was brought to our clinic for treatment because of inability to thrive. She had been fed *via* a nasogastric feeding tube since birth. Upon examination it was evident that the patient's jaws were fused in the posterior regions bilaterally. In the anterior region there was an 8 mm openbite. In the posterior regions intermaxillary fibrous adhesions were present

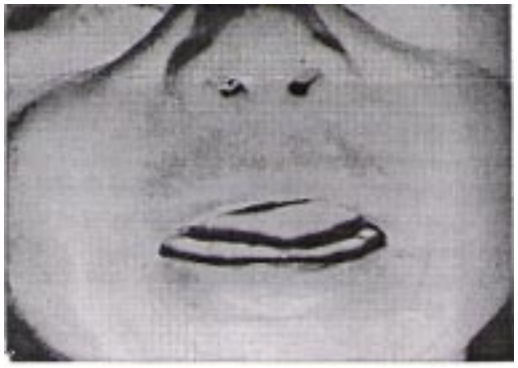

Fig. 1. Bilateral intermaxillary fibrous adhesions between the alveolar ridges of the maxilla and mandible in the molar area which extend posteriorly preventing mouth opening (note anterior open bite).

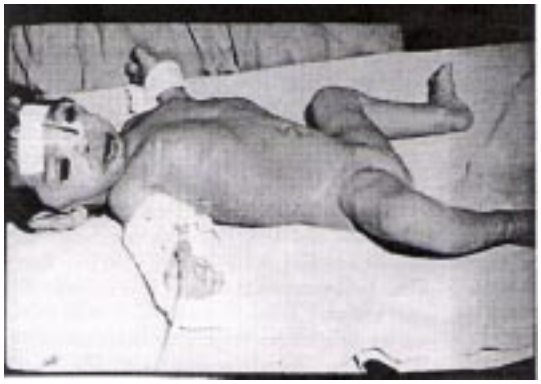

Fig. 2. Underweight infant upon admission with nasogastric feeding tube.

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between the alveolar ridges bilaterally (Figs. 1, 2). The medical history edical history of both parents was insignificant except that there was consanguinity in the parents (they were cousins). The mother had had a healthy gestation period and there was no account of illness, trauma, or drug use. The baby was the family's 2nd child, and there was no history of similar anomalies in close relatives except that the grandfather of the baby had syndactyly, The Ist child of the parents was normal but had died of meningitis. Routine laboratory exam values were normal.

TREATMENT

Following general anesthesia, the dense fibrous adhesions on both sides of the jaws were excised using surgical scissors. Manual manipulation and manipulation with an expander was gently used until an inter-alveolar mouth opening of 30 mm was obtained (Fig. 3).

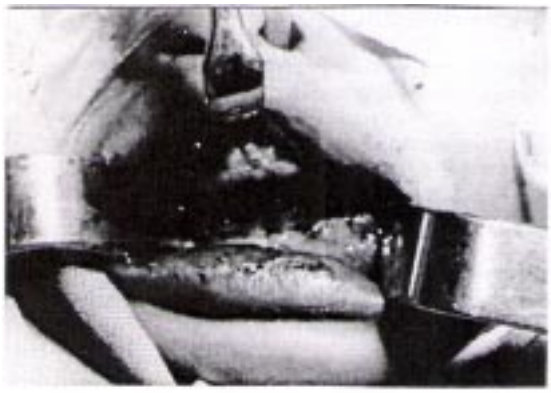

Fig. 3. Intraoperative view after lysis of adhesions between the alveolar ridges.

DISCUSSION

Congenital adhesions (synechiae) of the oral cavity are rare.[1,2] They arise between the upper and lower alveolar ridges (syngnathism) or between the tongue and palatal 'margins (glossopalatal ankylosis). These adhesions consist of membranes bands of epithelium supported by various amounts of connective tissue, muscle or bone.[2] Alveolar adhesions are rarely seen in isolation and often accompanied by additional congenital defects, such as cleft lip or palate, microglossia, micrognathia, or TMJ anomalies. Cleft palate is frequently seen with alveolar fusion.[1-8] In a review of 50 cases of alveolar synechiae between 1990 and 1993, Gartlan[6] found only 7 cases of isolated alveolar fusion; the remainder were associated with syndromes such as Van der Woude, cleft palate alveolar synechiae and oromandibular limb hypogenesis syndromes.

The etiology of synechiae is unknown. It is postulaled that during the 7th to 8th week of embryological alveolar ridge development when the tongue and palatal shelves are in close contact, the ensuing palatal closure depends on downward and forward contraction of the tongue; failure of the tongue to protrude predisposes to alveolar fusion. Genetic, teratogenic, or physical insults during this period also may lead to prolonged contact between oral structures, thereby, leading to abnormal fusion.[8-14] Others feel that adhesions may be remnants of the buccopharyngeal membrane.[15-16] Fhurmann[17] found a hereditary link in 5 family members with cleft palates and synechiae.

This was the authors' third encounter of congenital maxillomandibular fusion in infants. Our other case of congenital fusion of the mandible and maxilla was in a 1 year-old boy suffering from inability to open the mouth due to bilateral fusion of the jaws since birth. In this case there was no consanguinity in the parents or history of birth defects in the family. He was treated similarly by excising the fibrous adhesions on the left side and bony-fibrocartilage adhesions on the right side using electrocautery. Another case seen at our previous center was congenital unilateral fusion of the mandibular and maxillary alveolar ridges wel as the temporomandibular joint, and coronoid process in a 5-month-old boy which was reported by our collegues in 1996.[5] This case required use of an osteotome to treat the bony fusion and the temporomandibuar fusion required arthroplasty until the jaw could be mobilized. In this case, although the parents had no consanguinity, this patient presented other anomalies such as colobomas of the eyelids, facial hypoplasia, ear tags and pectus excavatum.

In all cases surgical division of adhesions was necessary for normal feeding, preventing airway obstruction, and allowing normal mandibular function and growth. The sooner treatment is rendered the less the possibility of TMJ ankylosis; ankylosis can lead to lack of mandibular growth and facial deformities. It should be noted that although surgery is simple, general anesthesia is often difficult. Fine fiberoptic laryngoscopes are required for nasoendotracheal intubation. Another point to be of mentioned is that normal-range mouth opening often cannot be obtained immediately after incising the adhesions and forceful manipulation. The TMJs are stiffened due to disuse, the alveolar ridges are soft and infantile bone is fragile and thus, excessive force may easily lead to jaw fracture and should be avoided. Normal mouth opening will automatically be regained within 1-2 weeks postoperatively.

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