

# Synechia of the Oral Cavity with Cleft Palate: A Rare Congenital Anomaly

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## ABSTRACT

**Aim:** Fusion defects of maxilla and mandible are a rare congenital anomaly that affect the growth and development of patients in varying extents. Authors intend to present a case of incomplete synechia of the oral cavity.

**Background:** Congenital maxillo-mandibular fusions present with varying degree of involvement of mucosa, soft tissues and bone and can cause aerodigestive problems in the neonatal period.

**Case description:** A 5-day-old male child was referred from pediatrics for evaluation for complaints of inability to feed. On examination, a mucosal band was noted connecting the floor of the mouth with the hard palate, with free margins on both sides. After proper consent, the mucosal band was completely excised under local anesthesia and hemostasis was achieved. The rest of the oral cavity and oropharynx was normal except cleft palate.

**Conclusion:** These types of anomalies have to be diagnosed as early as possible in the early neonatal life. Early diagnosis and appropriate management should be instituted to prevent risks of asphyxia, aspiration pneumonitis, growth retardation, malnutrition, facial growth abnormalities, and improper eruption of teeth.

**Clinical significance:** As there are various types of presentations, the treatment needs to be individualized.

**Keywords:** Maxillo-mandibular fusion, Oral synechia, Orofacial anomalies, Syngnathia.

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## BACKGROUND

Fusion of maxilla and mandible can involve only the soft tissues or both hard and soft tissues of the oral cavity. It may present with respiratory or feeding difficulty and facial growth abnormalities due to temporomandibular joint fixation at a later age.<sup>1</sup> Fusion of the maxilla and mandible mostly occurs in association with other oral and orofacial anomalies such as cleft lip and cleft palate.<sup>1-3</sup> Decision to discuss this case was taken, first, to highlight this rare birth defect to the readers; secondly, our case did not find any place in the most accepted classifications of this condition.

## CASE DESCRIPTION

A 5-day-old neonate weighing 2 kg was referred to us from the Department of Pediatrics for evaluation and management of restricted mouth opening and inability to feed. The baby was a full-term born with normal vaginal delivery in a hospital without any adverse consequences. According to the mother since the time of birth, the neonate has not been able to open his mouth and feed. The child has been kept under observation with nasogastric tube feeding. There was no similar history in other siblings of the child. The patient was simply diagnosed as a case of tongue tie from the Department of Pediatrics and they have sent the child for its release. However, on examination of the oral cavity, it was observed that there was an incomplete fibrous band between the floor of the mouth and hard palate anteriorly (Fig. 1). He rest of the structures of the oral cavity could not be visualized.

After obtaining proper informed consent from the parents, the child was transferred to the ENT ward. Since it was not possible to intubate the patient, excision of fibrous attachment was planned under intravenous sedation. Once under sedation, the band was

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excised and hemostasis achieved. After complete excision of the band, complete visualization of oral cavity was done. Remarkably, the child's tongue was normal (Fig. 2). On further inspection, presence of cleft palate involving both hard and soft palate areas could be noted (Fig. 3).

The child fared well in the recovery period. He was sent to the pediatrics and plastic surgery department for further management regarding cleft palate and low birth weight.

## DISCUSSION

Incidence of congenital birth defects in neonates is about 2.5%. Congenital fusion defects of the maxilla and mandible with other anatomical oral and orofacial abnormalities are usually rare.<sup>3</sup> Though congenital fibrous bands commonly involve either between alveolar ridges of the maxilla and mandible or between the tongue and palate, it can involve any part of the oral cavity.<sup>1,2</sup> The term



Fig. 1: Front view of the oral cavity showing incomplete congenital mucosal band (synechiae) between floor of mouth and palate



Fig. 2: Front view of the oral cavity after removal of mucosal band showing normal tongue

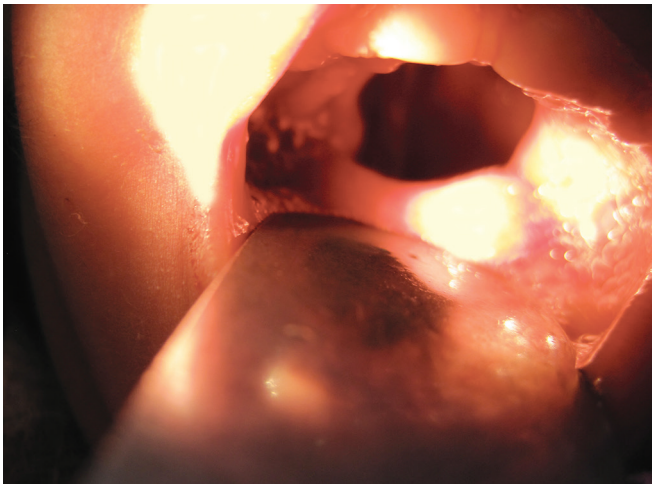


Fig. 3: Front view of the oral cavity after removal of mucosal band showing cleft palate

syngnathia denotes congenital fusion bands between maxilla and mandible and epithelium, connective tissue, muscle and bone all may be involved in the process.<sup>3</sup> It is known as synostosis when it comprises of bones and soft tissues or synechiae when there is only involvement of soft tissues. Bony maxilla-mandibular fusion is an extremely rare anomaly.

Two classification systems of such fusion have been proposed by Dawson and colleagues<sup>4</sup> and Laster and coworkers.<sup>5</sup> Dawson divided the cases of syngnathia into:

- Type I: Simple—with no other birth defects in head and neck;
- Type II: Complex—with two subtypes:
  - Type IIa: Syngnathia with aglossia;
  - Type IIb: Syngnathia with agenesis or hypoplasia of the proximal mandible.

Laster modified Dawson's classification system as:

- Type Ia: Simple anterior syngnathia characterized by bony fusion of the alveolar ridges and without other congenital deformities in head and neck
- Type Ib: Complex anterior syngnathia characterized by bony fusion of the alveolar ridges only and associated with other congenital deformities in head and neck

- Type IIa: Simple zygomaticomandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex, causing only mandibular micrognathia
- Type IIb: Complex zygomaticomandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex and associated with clefts or temporomandibular joint ankyloses.

However, both classification systems do not classify simple anterior incomplete synechiae with cleft palate into any type. A new system of classification which takes into consideration the fusion of soft tissues and bones separately along with the presence of other congenital orofacial anomalies should be devised.

Various causes have been considered as a reason for the syndrome; however, none has been proved. Some described the cause to include persistence of the buccopharyngeal membrane,<sup>6</sup> amniotic constriction bands in the region of the developing branchial arches, environmental insults, drugs such as large doses of vitamin A.<sup>7,8</sup>

A proper radiological investigation including high resolution CT scan can support the clinical recognition of this condition and its nature. Such congenital bands interfere with feeding, breathing, general health of the patient, growth and development, intubation for anesthesia and difficulty in breathing, aspiration pneumonitis and other problems.<sup>8</sup> Early intervention is needed to secure airway followed by management of feeding problems as delayed intervention in the case of maxillo-mandibular fusion can predispose the neonate to asphyxia, aspiration pneumonitis, malnutrition, growth retardation, poor growth of the facial skeleton, and poorly aligned dental eruption. However, treatment varies as per the extent and nature of anomalies.

## CONCLUSION

These type of anomalies have to be diagnosed as early as possible in the early neonatal life. Early diagnosis helps in institution of appropriate management plan for the child. Early intervention is needed to secure airway followed by management of feeding problems as delayed intervention in the case of maxillo-mandibular fusion can predispose the neonate to asphyxia, aspiration pneumonitis, malnutrition, growth retardation, poor growth of the

facial skeleton, and poorly aligned dental eruption. Management plan varies as per the need of the individual case.

### CLINICAL SIGNIFICANCE

Congenital fusion of maxilla and mandible is a rare anomaly. It may present with aerodigestive problems and facial deformities. As there are various types of presentations, treatment needs to be individualized.

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