

# Syngnathia Without Any Other Associated Anomaly: A Very Rare Case Report

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## Abstract

Congenital bony fusion of the mandible and maxilla (syngnathia) without any other oral anomalies is a very rare condition. Numerous cases with other anomalies like cleft lip, cleft palate, aglossia have been described. Syngnathia can also occur with Vander Woude syndrome and popliteal pterygium syndrome. This report presents a very rare case of syngnathia with true bony fusion between mandible and maxilla, unusually with no other intra-oral or systemic anomalies, of which only a few cases exist in the world literature.

## INTRODUCTION

Syngnathia is a rare anomaly involving fusion between the maxilla and the mandible. The fusion may be due to soft tissue adhesions between the two or a true bony fusion between maxilla and mandible<sup>1,2</sup>. The latter is a very rare anomaly. These children present in the immediate neonatal period with difficulties in the airway protection and maintenance as well as feeding problems<sup>3</sup>. Most have other associated anomalies like popliteal pterygium syndrome, Vander Woude syndrome requiring concurrent management. Surgical management involves division of the bony fusion or break down of the adhesions in the first few days of life. Depending upon the severity, these children present formidable anaesthetic challenges. The prognosis is generally favourable.

We present a very rare case report of bony fusion between the maxillary and mandibular alveoli without any other associated anomaly.

## CASE REPORT

An 1800 gms male infant was born to a 30 year old Woman (G<sub>4</sub>P<sub>4</sub>A). It was the product of a consanguineous marriage with a full term unremarkable pregnancy with poor antenatal follow up. It was a normal vaginal delivery conducted at home. Shortly following delivery, the baby was noted to have difficulty in opening his mouth with no anterior jaw opening. The parents got alarmed and immediately consulted a local physician who examined the baby and confirmed the fusion between maxilla and mandible.

There was no other associated local or systemic anomaly. The maternal and paternal history was negative for any facial cleft. All other siblings were normal. There was no record of TORCH (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes), VDRL and alpha-fetoproteins during the pregnancy. The baby was referred to Department of Plastic and Reconstructive Surgery, Sher-i-Kashmir Institute of Medical Sciences, Srinagar on 2<sup>nd</sup> day of his life because of inability to feed the child. The baby was admitted in Neonatal ICU (Intensive care unit). A nasogastric tube was put in for feeding and the patient was maintained on humidified oxygen by mask. Clinical examination revealed complete bony fusion between the maxilla and the mandible involving the entire alveolar margin with a small gap about 12 mm on left side in the canine region (Figure 1).

**Figure 1**

Figure 1: Photograph showing bony fusion between Maxilla and Mandible with a small gap (12mm) in the left canine region.



Very feeble motion was palpable over each Temporomandibular (TM) joint. A Computed Tomography (CT) scan was done which showed bony fusion between the two alveolar ridges with a small gap on the left side (Figure 2).

**Figure 2**

Figure 2: Computed Tomography Scan showing the bony fusion between the two alveolar ridges with a small gap on the left side.



The patient was kept in neonatal ICU for optimization before surgery. All base line investigations were done including Haemogram, Kidney function tests, Liver function tests, X-ray chest. The patient was assessed by the anaesthesia team in anticipation for surgery and preparations were made for

awake blind nasotracheal intubation. On 12<sup>th</sup> day of his life, patient was taken to the operating room. He was successfully anaesthetized by awake blind nasotracheal intubation. The bony fusion was broken using a micro-osteotome and hammer from right molar to left molar area. After breaking down the fusion, the jaws were gently separated to achieve a mouth opening of 15mm (Figure 3).

**Figure 3**

Figure 3: Postoperative photograph showing separation of the Maxilla and mandible with adequate mouth opening.



This gap was maintained by putting two small silastic sheets in the raw molar areas on either side which was secured by few stitches with the surrounding tissues. He tolerated the entire procedure well and returned to the recovery room, breathing spontaneously, well oxygenated. He was shifted back to neonatal ICU and on first postoperative day bottle feeding was started. He could not suck well. Probably because of fusion of the mandible and maxilla he had not developed an active sucking reflex in utero. So a pacifier was used to enhance the development of active sucking reflex (Figure 4).

### Figure 4

Figure 4: Separated alveoli with a pacifier in situ to enhance sucking reflex.



Within a few days, the baby started feeding from a bottle and was thriving well. His postoperative period was uneventful and was discharged from the hospital on 10<sup>th</sup> postoperative day. At that time, spontaneous mouth opening was around 16mm. At 2 weeks postoperatively, the silastic sheets were removed and the patient was well on follow-up. At the age of about 3 months, the patient developed loose motions, he was again admitted and I.V. fluids were given. The loose motions got controlled, but unfortunately, the baby was found dead in the morning once her mother got up. The exact cause could not be known as no consent for autopsy was given.

### DISCUSSION

Maxillomandibular fusion is a rare group of anomalies varying in severity from simple mucosal adhesions (synechia) to extensive bony fusion (syngnathia). This depends upon the amount of mesodermal penetration. True bony fusion is a very rare anomaly and only a handful of case reports exist in the literature<sup>4,5,6</sup>. The soft tissue fusion (synechia) have been extensively reviewed by Gartlan et al, and were classified as bucopharyngeal membrane remnants or as ectopic membranes depending upon their presumed

origin.

Bony fusion (syngnathia) without any associated systemic or intraoral anomaly, as in our case report, is an extremely rare entity and only a few cases reports exist in the literature<sup>8</sup>. Most of these reports are inadequate in description, inconsistent and confusing in nomenclature and with limited useful conventional imaging<sup>9,10,11,12</sup>.

The cause of congenital bony fusion is not certain. Some of the postulated causes including Goodacre and Wallace's experimental studies include persistence of bucopharyngeal membrane, amniotic constriction bands in the region of the developing first branchial arch, environmental insults, drugs such as meclozine and large doses of Vitamin A. A review of 5 cases presented by Dawson et al<sup>9</sup>, provides no evidence of any familial tendency, history of drug and toxin exposure or consanguinity. In our case report, although there was a history of consanguinity, but there was no family history or any exposure to drugs or toxins as reported by some authors<sup>10,13,14,15</sup>.

The congenital bony fusion can be clinically recognized and diagnosed at or soon after birth of the affected neonate as they have airway and feeding difficulties<sup>5,9</sup>. The diagnosis can be confirmed by conventional radiography using high resolution or spiral CT scan (Figure 2). It also tells about the condition of TM joints, any hypoplasia of the other facial bones.

The problems associated with syngnathia include maintenance and protection of airway, feeding difficulties and induction of anaesthesia. On the basis of reports from the literature, functional results especially in isolated cases of syngnathia are likely to be good<sup>9,10,11,12</sup>. Since this condition is very rare, so it is difficult to standardize the treatment. Therefore, treatment is individualized. In terms of jaw function and its outcome, it is more problematic in complex cases.

Dawson et al, have proposed a system of classification and elaborated on treatment recommendations. Their proposed classification is aimed at treatment and likely functional outcome, rather than on etiology or pathogenesis of the malformation. The airway is the first priority, followed by feeding problems to be overcome by placing nasogastric tube or gastrostomy tube.

Since the occurrence of bony fusion of maxilla and mandible is extremely rare, and there is high rate of association

between bony syngnathia and other regional and systemic malformations,<sup>4,7,11,12,14,17,18</sup> the patient should be kept under the supervision of a team of clinicians skilled in the diagnosis and appropriate treatment of the congenital oral and maxillofacial anomalies<sup>9</sup>.

The optimal treatment of bony syngnathia is the surgical division of the fusion under anaesthesia. Awake blind nasal intubation as we did in our case report, is the ideal way of introducing anaesthesia. In cases of failed awake blind nasal intubation, tracheostomy may be required<sup>6</sup>. Proper physical therapy and feeding should be resumed as soon as possible after the surgery as we did.

The significant points about our case report include: it was an isolated pure bony fusion without any associated local (cleft lip/palate) or systemic anomalies. To our knowledge, only few cases exist in the world literature so far. Secondly there is a history of consanguinity among the parents which suggests the possibility of autosomal recessive inheritance.

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