

Clinical assessment of a delayed Congenital Bilateral Maxillomandibular Syngnathia– a comprehensive management and Literature Review

ABSTRACT

Syngnathia refers to maxillomandibular fusion and may present with other defects, the airway and feeding being the primary considerations. Congenital syngnathia is an extremely rare condition only described predominantly in Asia and with some cases in Africa. To date, only 21 cases of true bony bilateral syngnathia have been reported. Surgical interventions were usually instituted rather early, most cases received surgery within the first 15 days of birth. The oldest age reported to receive a reconstructive surgery was 4 years old to date.

Here we describe the surgical management of abilateral complete extraoral maxillomandibular fusion as well as alveolar fusion intra-orally with temporomandibular joint (TMJ) involvement, who had managed to survive to the age of 9 years without any surgical intervention. The family managed to feed the patient through a Ryles nasogastric prior to the surgical intervention. At presentation, the patient was severely underweight due to malnourishment.

This is also the first report of use of bilateral temporalis flap as an interposition flap to prevent TMJ re-ankylosis. Peri-operatively, a mouth opening of 40mm was achieved. General improvements were confirmed by a stable mouth-opening of 20mm as well as an increased weight of 2 kgs at the 2years post-operative review.& at 4 year review continued to be stable at 20mm with additional weight of 2 kg.

The minimal post-operative follow up and care including physiotherapy for jaw movements and speech, the quality of feeding and phonation improved significantly. Additional rehabilitation measures including the distraction of TMJ replacement are being carefully planned to commence when the patient turns 16 year old if the bone quality has improved such that this procedure can be attempted.

KEYWORDS:

Maxillomandibular fusion, TMJ ankyloses, Craniofacial surgery ,Congenital facial defect,Delayed Diagnosis

Introduction

Congenital craniofacial defects account for approximately 20% of all birth defects, one of these is congenital fusion of the maxilla and mandible(1). Syngnathia is the maxillomandibular fusion with or without ankylosis of the temporomandibular joint (TMJ). The condition presents with a broad spectrum of severity, ranging from single mucosal bands of fibrous tissues (synechia) to bony fusion (synostosis)(2).

Congenital syngnathia is a very rare anomaly. The first case was reported in 1936 by Burket, where the patient presented with TMJ ankylosis as well as gingival fusion related to facial hemi atrophy and Horner's syndrome(3). Since then, there has been limited cases that has been reported to date. A total of 118 cases have been reported to date for congenital syngnathia, of which only 21 cases had bilateral bony fusion(4).

The fusion of the bones can extend to any part of the jaw to any part of the head and neck including the fusion of the mandible to zygomatic complex, temporal bone

extraorally, to the alveolus, tuberosity and hard palate intraorally(5). Syngnathia can severely limit the mouth opening, causing difficulty in respiration and feeding, thus the condition can make the normal day to day life activities and general anaesthesia very challenging. Particularly for young children, syngnathia also interferes with the normal craniofacial and dental development, which adversely affects the physical and mental health of the patients and their families.

The age of the presentation was mostly very early in reported cases, with most cases within the range of at birth up to a year, with a very few surviving cases after the period without any surgical treatments (4). Most of the reported cases to date included unilateral or incomplete fusions with only a few bilateral fusions presented at the first few months. The extensiveness of the bony fusions in most of these cases were less severe with only 3 cases with bilateral bony fusion with TMJ involvement out of 62(2, 6, 7), of which cases they had earlier surgical interventions to enable feeding. The oldest case reported to date to receive a surgery was 4 years old, reported by Rahman et al(8).

In this case report, we present a 9-year-old Yemeni girl that presented with bilateral complete extraoral maxillomandibular fusion as well as alveolar fusion intra-orally with TMJ involvement. The authors would also like to highlight that this is the first case report to discuss a patient who survived to the age of 9 without any surgical interventions& also the first to receive a interpositional temporalis flap to prevent reankylosis and maintain the mouth opening.

OBJECTIVES:

1. To give a outline of presentation of Syngnathia

2. Discuss the work up and investigations required
3. Discuss the surgical interventions and long term sequelae following surgery
4. Explore the role of interpositional flaps in Syngnathia surgery to minimise reankylosis and improve long term results

Case Report

A 9-year-old girl presented to the department of oral and maxillofacial surgery department at King Fahad Central Hospital in Jazan, Saudi Arabia, with a severe facial deformity with inability to open her mouth, which had been continuous since birth. At the presentation, she was unable to articulate her jaws or produce meaningful speech. Clinically, the lack of articulation was due to the lack of space for the tongue to move, which was completely boxed in due to the bony fusions.

The patient comes from an underprivileged rural area of Yemen, where consanguineous marriage is common culturally. In their family, only one of their older siblings is affected by almost the same condition. There was no previous history of similar surgical intervention in the family. The patient would self intubate a Ryles naso gastric tube via her nostril & the parents would pour long life milk 5 to 6 times a



Figure 1 Preoperative extraoral photographs A. Frontal B. Side profile view.

day, this was her only means of nutrition. . No records were available from the mother's obstetric history or the birth.

Clinical examination revealed a very malnourished child with a hypoplastic maxilla and mandible extraorally, Intraorally there was complete fusion of the alveolus with no gap anywhere across the arch precluding any possibility of oral feeding.

An orthopantomogram (OPT) showed the collision of deciduous and permanent dentitions due to the lack of intraoral space. It also showed the evidence of underdeveloped ramus bilaterally, as well as bony fusion across the maxilla mandibular region, zygoma, temporomandibular joints.

A Computerised Tomography (CT) scan was performed to confirm the three-dimensional (3D) structure and discern the type and extent of the bony fusion. The CT scan results also showed the fusion being close to the carotid and jugular veins at the base of skull (See **Fig 2A-E**).

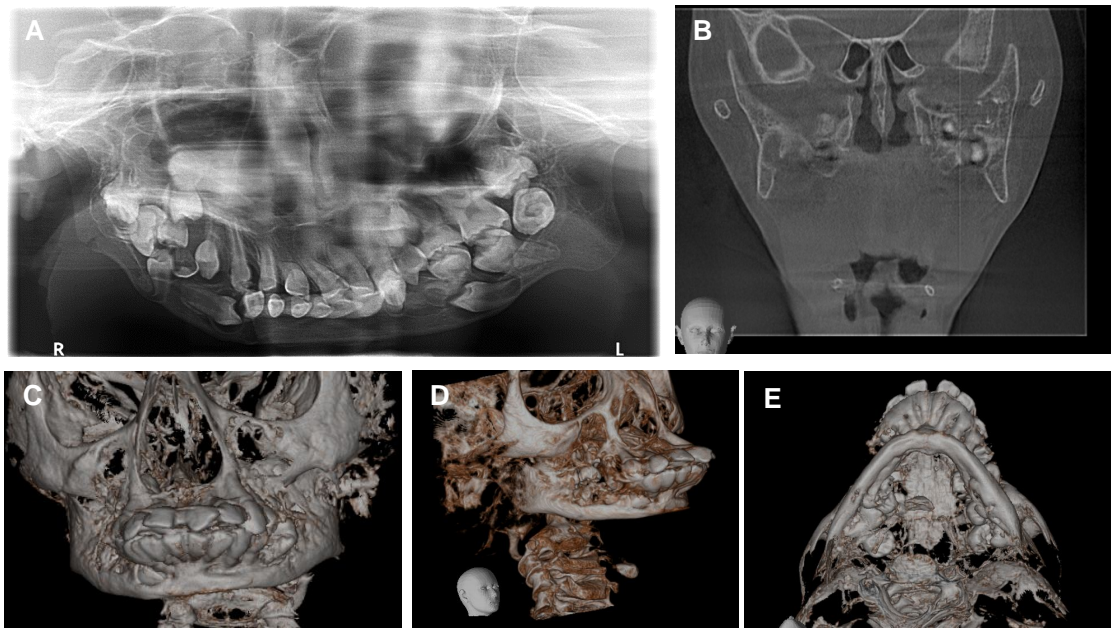


Figure 2 Radiological images: A. OPT, B. MRI, C-E CBCT

After the fibreoptic intubation, a planned tracheostomy was performed, followed by a bilateral intraoral I mandibular incisions , extraoral preauricular incisions to expose the temporalis and the temporomandibular joints. Gap arthroplasty with coronoidectomy to create a surgical space was performed. Before the intraoral procedure, the extent of the bony fusion was assessed under general anaesthesia (See **Fig 3**). Intraoral bony adhesions were released using a fine drill and saw blade. Dental extractions were performed to remove any teeth interfering with the surgical

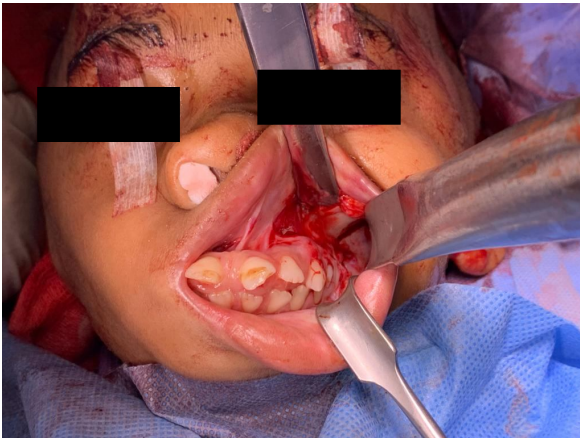


Figure 3 Perioperative photograph demonstrating the extent of the bony fusion procedures.

The general quality of the bone was suboptimal with very thin and fragile areas, possibly due to the lack of nutrition, which could affect the chance of successful osseo-distraction. A mouth opening of 40cm was achieved perioperatively, with a continuously stable mouth opening post-operatively (see **Fig. 4A and B**).



Figure 4A. Perioperative mouth opening at 40mm, B. Postoperative outhopening

Bilateral temporalis flaps were raised and interpositioned into the gap arthroplasty to prevent re-ankylosis and improve the long-term management of the condition.

Following the reconstruction, the surgical sites were closed and dressed accordingly (See **Fig 5**).

Following the operation, she was rehabilitated in the intensive care unit (ICU), and tracheostomy decannulation was achieved in 3 days. The patient was discharged 5 days post operatively.

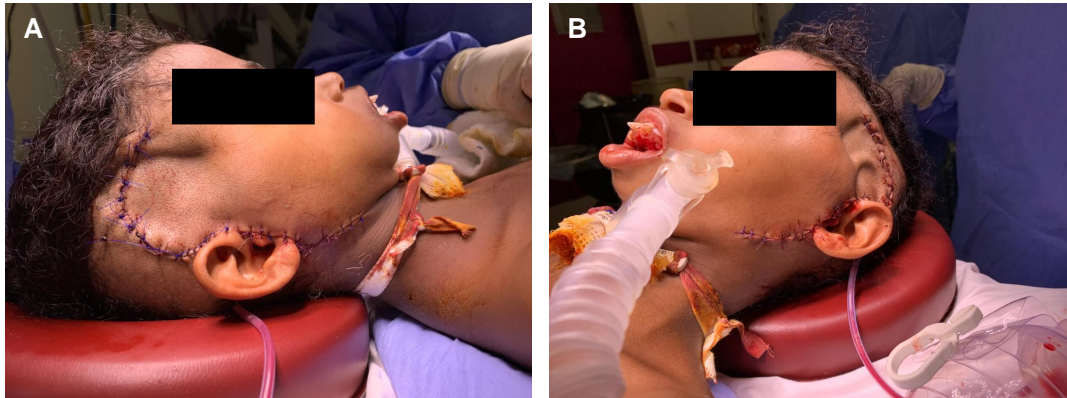


Figure 5 Immediate post-operative photos. A. Right, B. Left

At the review at 2 years later the mouth opening was stable at 20mm with improved speech and articulation. General nourishment has improved significantly demonstrated by regular oral intake of the food as well as an increase of the weight by 2kg. However, in-depth follow up have been challenging for the patient and their family due to their limited access to care and transport, therefore most reviews were carried out via telephone. At the 3 year review the mouth opening was still stable at 20mm.

Further rehabilitation in the form of distraction and TMJ replacement is being planned when the patient is 16 years old.

Discussion

Most synnathia cases are detected promptly following the birth because the baby is not able to open the mouth for normal feeding. The affected patients are usually presented to a relevant specialist within hours of birth(9). Interestingly, the reported patient was able to survive until the age of 9-years without any appropriate surgical

management. Despite the delayed presentation and severe malnourishment, the patient was able to receive appropriate management and follow up.

Dawson et al. classified syngnathia into simple and complex syngnathia in 1997(10). Simple syngnathia does not present with other abnormalities in the head and neck, whereas complex syngnathia is characterised by other co-existent anomalies in the area. Laster et al. suggested a modified version of the syngnathia classification, where they added more categories for the rare cases of bony fusions(1). The patient reported here, had bilateral maxillomandibular fusion alongside the zygoma and TMJ involvement, which could be classed as Type 2b according to the modified classification.

Surgical sectioning of the fusion is essential for normal functions such as everyday feeding, airway securement, jaw functions and speech, so it should be performed at the earliest opportunity. Delayed surgical management is associated with an elevated risk of TMJ ankylosis which may result in the lack of mandibular development and facial deformities(11), which was the case in this patient. The lack of mouth opening led to the challenge in rendering a general anaesthesia, which was overcome by using the fiberoptic laryngoscope for intubation purposes.

Insufficient intraoral space resulted in surgical challenges in creating sufficient space for the jaw articulation, making the reconstruction very difficult following the jaw division. The patient's bone was fragile due to the malnourishment, and excessive force may easily result in jaw fracture(12). Pre-existing ankylosis of TMJ due to disuse was an added difficulty to the surgical considerations along with fusion of the zygoma and the skull base.

. Bilateral temporalis flap was used as an interpositional flap to prevent/minimise TMJ re-ankylosis.

Based on previous case reports, the functional outcomes in isolated syngnathia cases are likely to be successful in simple fusions(13, 14). However, it is difficult to standardise the management due to the extreme rarity of the conditions, so each case should be considered individually considering the location and severity. The functional outcome for jaw fusion cases are generally worse in complex cases involving several fusion sites, leading to reankylosis(15, 16).

A careful surgical planning and regular follow-ups including long-term rehabilitation are considered fundamental to achieving a satisfactory long term outcome. Optimal surgical division is essential for all the areas of concern with a sufficient separation of bone fusions to prevent any recurrence. To the authors' knowledge, this case is the first to report the usage of bilateral temporalis flaps as an interpositional graft/flap to prevent reankylosis of the TMJ and has been shown to be successful with a 4 year follow up & the first case report of first treatment aged 9 and having survived to this age without surgical intervention.

Furthermore the temporal/zygomatic branches of facial nerve bilaterally showed normal function despite the extensive facial /temporal approaches.

The reported case did not show any airway issues or feeding difficulties during the follow up period for 4 years.

We would recommend aggressive gap arthroplasty and an interpositional temporalis flap in chronic TMJ ankylosis to prevent early relapse .

Conclusion:

We have successfully surgically treated a delayed presentation of a complex rare case of Syngnathia with a follow up of 4 years.

Workup with 3 D scans and knowing the points of ankylosis is important. Early recognition and treatment is obviously the best way forward, however given the delayed presentation and the extensive ankylosis, we have used a interpositional flap which has been successful in preventing reankylosis and maintaining a functional mouth opening . We recommend interpositional flaps/grafts in such cases.

The use of a crepe bandage or a sports kinesiology tape extraorally will help with reducing the extraoral swelling and keeping patients comfortable . (17)

Consent

As per international standards, parental written consent has been collected and preserved by the author(s).

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References

1. Laster Z, Temkin D, Zarfin Y, Kushnir A. Complete bony fusion of the mandible to the zygomatic complex and maxillary tuberosity: case report and review. *International Journal of Oral and Maxillofacial Surgery*. 2001;30(1):75-9.
2. Ugurlu K, Karsidag S, Huthut I, Yildiz K, Bas L. Congenital Fusion of the Maxilla and Mandible. *Journal of Craniofacial Surgery*. 2005;16(2):287-91.
3. BURKET LW. CONGENITAL BONY TEMPOROMANDIBULAR ANKYLOSIS AND FACIAL HEMIATROPHY: REVIEW OF THE LITERATURE AND REPORT OF A CASE. *Journal of the American Medical Association*. 1936;106(20):1719-22.
4. Kumar V, Rattan V, Rai S. Congenital Maxillomandibular Syngnathia: Review of Literature and Proposed New Classification System. *J Maxillofac Oral Surg*. 2021;20(1):19-36.
5. Jackson I, Agrawal K, Bush K. Congenital bilateral maxillo-mandibulo zygomatic fusion with bilateral oblique facial clefts. *European Journal of Plastic Surgery*. 1996;19(5):262-4.
6. Konas E, Aliyev A, Tunçbilek G. Congenital Maxillomandibular Syngnathia: A New Management Technique Using Distraction Techniques. *Journal of Craniofacial Surgery*. 2015;26(1):e68-e70.
7. Al-Mahdi AH, Koppel DA, Al-jumaily HA, Mohammed AAH, Boyd D. Congenital Bilateral Zygomatico-Maxillo-Mandibular Fusion Associated With Gum Fusion. *Journal of Craniofacial Surgery*. 2016;27(1):e20-e3.
8. Rahaman MM, Kabir MI, Rabby MAI, Rahman MM, Uddin MN, Rashid MHU, et al. Congenital syngnathia in bangladesh: a very rare case report. *Medicine Today*. 2014;26(2):114-7.
9. Daniels JSM. Congenital maxillomandibular fusion:: a case report and review of the literature. *Journal of Cranio-Maxillofacial Surgery*. 2004;32(3):135-9.
10. Dawson KH, Gruss JS, Myall RWT. Congenital Bony Syngnathia: A Proposed Classification. *The Cleft Palate Craniofacial Journal*. 1997;34(2):141-6.
11. Shams MG, Motamedi MHK, Abad HLD. Congenital fusion of the maxilla and mandible: brief case report. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*. 2006;102(2):e1-e3.
12. Mortazavi SH, Motamedi MHK. Congenital fusion of the jaws. *The Indian Journal of Pediatrics*. 2007;74(4):416-8.
13. Koeda S, Suzuki T, Nei H, Inahara H, Takata Y, Goto S, et al. A case of congenital unilateral maxillo-mandibular bony fusion in an 8-year-old girl. *Asian Journal of Oral and Maxillofacial Surgery*. 2010;22(4):220-4.

14. Nuruddin bin Mohd S. Congenital partial fusion of the mandible and maxilla: Report of a case. *Oral Surgery, Oral Medicine, Oral Pathology*. 1965;20(1):74-6.
15. Rao S, Oak S, Wagh M, Kulkarni B. Congenital midline palatomandibular bony fusion with a mandibular cleft and a bifid tongue. *British journal of plastic surgery*. 1997;50(2):139-41.
16. Nwoku AL, Kekere-Ekun TA. Congenital ankylosis of the mandible: Report of a case noted at birth. *Journal of maxillofacial surgery*. 1986;14:150-2.

17. Patil S, Rajanikanth K. Comparative Assessment of Post-Operative Sequelae Following Surgical Removal of Mandibular Third Molar with and Without Extra-Oral Application of Kinesiologic Tape. J. Pharm. Res. Int. [Internet]. 2021 Dec. 27 [cited 2025 Jan. 6];33(60B):2824-9. Available from: <https://journaljpri.com/index.php/JPRI/article/view/6718>