Case report

Congenital Bilateral Maxillomandibular Syngnathia with Delayed Presentation and Surgical Intervention – a Case Report 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 nasogastric tube prior to the surgery. 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 4 years post-operative review. Through 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 when they turn 16 years old.

KEY WORDS:

Maxillomandibular fusion /TMJ ankylosis/ Craniofacial surgery /Congenital facial defect/ Delayed presentation

Introduction

Congenital craniofacial defects account for approximately 20% of all birth defects, one of these is congenital fusion of the maxilla and mandible(Laster *et al*., 2001). 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 (synechiae) to bony fusion (synostosis)(Ugurlu *et al*., 2005). 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 hemiatrophy and Horner's syndrome(BURKET, 1936).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(Kumar *et al*., 2021).

The fusion of the bones canextend 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(Jackson *et al*., 1996). 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 (Kumar *et al*., 2021). 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*(Ugurlu et al., 2005, Konas et al., 2015, Al-Mahdi et al.*, 2016), 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(Rahaman *et al*., 2014).

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.

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 parents fed the patient through a nasogastric (NG) tube 5-6 times a day to source the nutrition. No records were

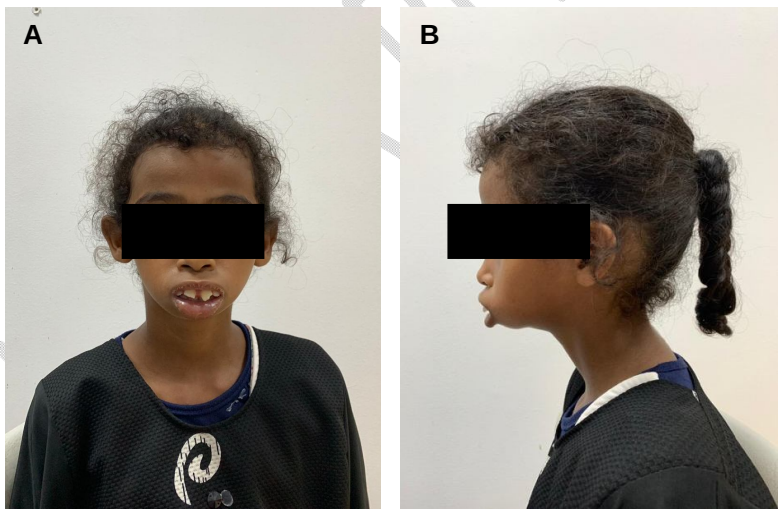

Figure 1Preoperative extraoral photographs A. Frontal B. Side profile view.

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 thethree-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**).

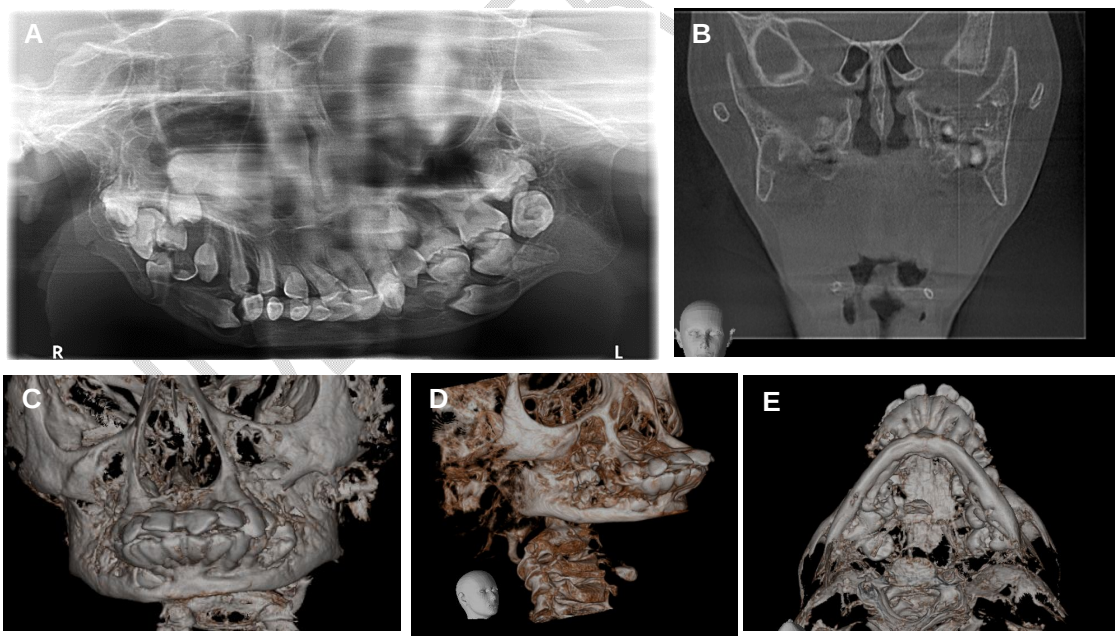

Figure 2 Radiological images: A. OPT, B. MRI, C-E CBCT

After the fibreoptic intubation, a planned tracheostomy was performed, followed by a bilateral trans- and extraoral preauricular incisions to expose the temporalis and the temporomandibular joints.Gap arthroplasty with coronoidectomyto 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 procedures.

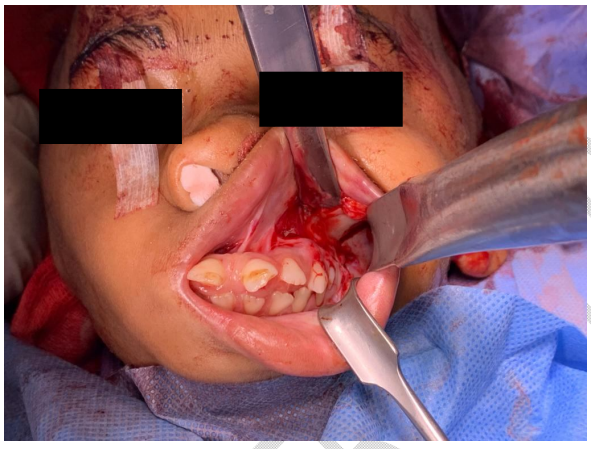

Figure 3 Perioperative photograph demonstrating the extent of the bony fusion

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 40cmwas achieved perioperatively, with a continuously stable mouth openingpost-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.

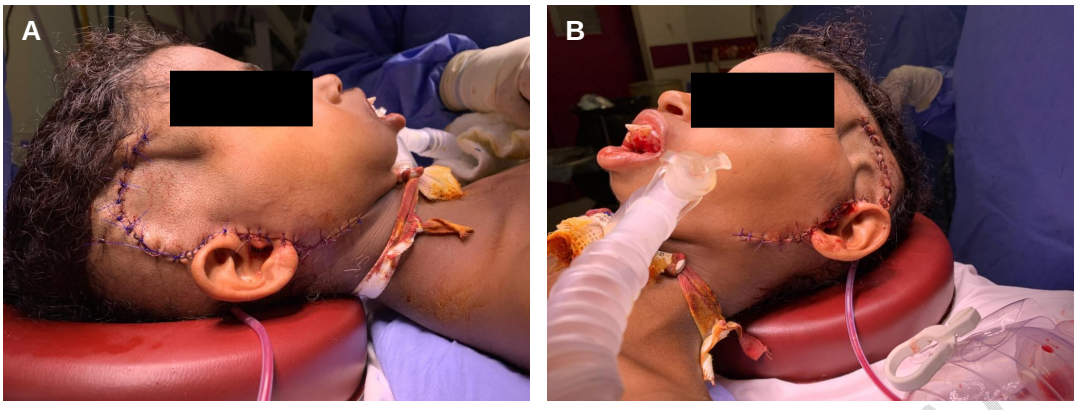

Figure 5 Immediate post-operative photos. A. Right, B. Left

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

Discussion

Most syngnathia 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(Daniels, 2004). Interestingly, the reported patient was able to survive until the age of 9-years without any appropriate surgical management. Despite the delayed presentation and severely malnourished, the patient was able to receive appropriate management and follow up.

Dawson *et al*. classified syngnathia into simple and complex syngnathia in 1997(Dawson *et al*., 1997). Simple syngnathia does not present with other abnormalities in the head and neck, whereas complex syngnathia is characterised by other co-extistent 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(Laster *et al*., 2001). 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(Shams *et al.*, 2006), which was the case in this patient. The lack of mouth opening led to the challenge in general anaesthetics, which was overcame by using the fibreoptic laryngoscopes in intubation. 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(Mortazavi and Motamedi, 2007). Pre-existing ankylosis of TMJ due to disuse was an added difficulty to the surgical considerations. Bilateral temporalis flap was used for the interpositioningto prevent TMJ re-ankylosis.

Based on previous case reports, the functional outcomes in isolated syngnathia cases are likely to be successful in simple fusions(Koeda *et al.*, 2010, Nuruddin bin Mohd, 1965). 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(Rao *et al*., 1997, Nwoku and Kekere-Ekun, 1986).

A careful surgical planning and regular follow-ups including long-term rehabilitation are considered fundamental to achieving a satisfactory 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 in the interpositioning of ankylosed TMJ. The reported case did not show any airway issues or feeding difficulties during the follow up period for 4 years.

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