

# Congenital Unilateral Zygomaticomaxillomandibular Fusion in a 4-year-old Girl

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**Summary:** Congenital zygomaticomaxillomandibular (ZMM) fusion is an extremely rare and debilitating condition. Fusion of the jaws results in a severe limitation of the mouth opening accompanied by breathing and feeding difficulties. Mandibular fusions to the maxilla, palate, zygoma, and temporal bone are described in literature. We present the case of 4-year-old girl diagnosed with congenital unilateral ZMM fusion. She was treated surgically at 1 year and at 4 and a half years to release the fusion and to improve mouth opening. Second surgery was indicated due to recurrence. Management of young patients with congenital maxillomandibular fusion is challenging. The scarcity of cases makes formulating practice guidelines very difficult. If surgery is performed, the importance of postoperative physical therapy cannot be overstated. The goal of this article is to emphasize the risk of recurrence in ZMM fusion after surgical treatment and importance of regular follow-up. (*Plast Reconstr Surg Glob Open* 2024; 12:e5677; doi: 10.1097/GOX.0000000000005677; Published online 23 April 2024.)

In congenital ZMM fusion, a distinction is made between bony ankylosis (synostosis or syngnathia) and fibrous adhesions (synechia). Kumar et al<sup>1</sup> provided an overview of congenital ZMM fusion cases available in the current literature. They reported 62 synostosis cases, 48 synechia cases, and eight combination cases at the time of publication in 2021.<sup>1</sup> Complete closure of the mouth is extremely rare, as reported by Raiteb et al.<sup>2</sup> The most important issue in small children with congenital syngnathia/synechia are feeding and respiration problems which may be life-threatening.

Etiology of ZMM remains unknown, but many hypotheses have been suggested in the literature.<sup>1</sup> Interestingly, as was stated by Burket<sup>3</sup> back in 1936, a true congenital fusion is exceptionally rare. Fusion resulting from bony healing after a traumatic forceps delivery is generally classified as congenital, although it is acquired at birth.<sup>3</sup> At present, multiple classification systems are available to subdivide maxillomandibular fusions, but none of them is comprehensive.<sup>4-6</sup> In the review by Kumar et al,<sup>1</sup> a

clinically oriented classification system is proposed which could be applied to improve treatment planning.

Surgical treatment of ZMM fusion is carried out at birth if breathing and feeding difficulties are life-threatening. It is advised to postpone surgical procedures if survival is likely without surgical intervention. The goal of surgical treatment is the release of fused tissues to improve mouth opening and thereby resolve breathing, feeding or eating, and speaking problems. Dental hygiene should be taken into account, which is often complicated due to the limited mouth opening. CT with preoperative three-dimensional planning is the current practice standard. Surgical approach may be intraoral or extraoral. Depending on the type of fused tissues, release of mucosal adhesions or bony osteotomies is performed. Most authors advocate interposition of grafts or material to prevent recurrence of fusion. The use of a buccal fat pads, abdominal fat grafts, and silicon sheets has been suggested in the literature.<sup>7,8</sup> After surgery, intensive follow-up and physical therapy are crucial in maintaining the surgical improvement in mouth opening.

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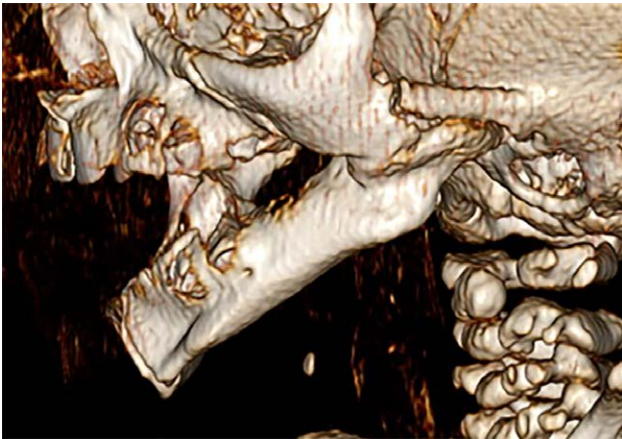
## CASE REPORT

The young patient was referred to our center 2 weeks after birth with difficulties of breast feeding and a noticed facial asymmetry of the lower jaw. Pregnancy and C-section at 40 weeks were complication-free. The infant had minimal asymmetry in the lower jaw, with the chin deviating toward the left and an asymmetrical position of the ears. No deformities in hands and feet were noticed. There were no congenital clefts, branchial deformities, or

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intraoral mucosal defects visualized. Mostly attributed to asymmetric mandibular growth with anterior open bite, mouth opening was limited to 9 mm, measured at the mid-line between upper and lower alveolar gingiva. The CT scan at birth showed unilateral ZMM fusion on the left side and incomplete bony fusion of the mandibular symphyseal region (Fig. 1).

Because of adequate food intake and growth, no surgical treatment was performed. At the age of 1 year, surgery was performed to release the unilateral ZMM fusion. A silicon sheet was interpositioned temporarily to prevent relapse. The sheet was removed after 8 weeks (Fig. 2).



**Fig. 1.** Three-dimensional reconstruction of CT images showing bony maxillomandibular fusion on the left side (CT scan taken at birth).



**Fig. 2.** Coronal section of CT scan showing silicon sheet interposition (arrow) after bony osteotomy of maxillomandibular fusion on left side (CT scan taken after surgery, on the age of 1 year old).

After surgery, early active physical therapy including the use of therabite (TheraBite Jaw Motion Rehabilitation System; Atos Medical) was started, but unfortunately, parents were not compliant. At the age of 4 years, the patient was referred back because of delayed speech development. The mouth opening was severely limited and fixed to 9 mm of opening with a persistent anterior open bite. Speech was completely incomprehensible. CT imaging showed no bony fusion (Fig. 3). At the age of 4 and a half years, a second surgery was performed. The inferior alveolar and lingual nerve were not encountered during exposure. Bony osteotomy was performed using an ultrasonic device (Piezotome M+; Acteon). After release of the fusion, a maximal mouth opening of 30 mm, measured between incisal edges of the central deciduous incisors was achieved with the use of Denhardt forceps. Next, a free fat graft from the inguinal region was interpositioned and suspended perizygomatically. A stab incision through the skin of the cheek was performed to make this possible.

Two days after surgery, the patient was pain free, and extensive physical therapy was started to aid in maintaining the established gain in maximal mouth opening. The active postoperative mouth opening was 15 mm, measured between incisal edges of the central deciduous incisors (Fig. 4).

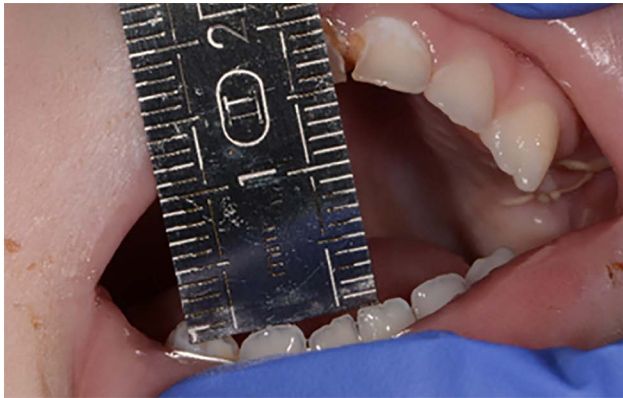
Unfortunately, the parents did not adhere well with postoperative instructions and physiotherapy was not followed strictly. At 6 months postoperatively, the patient presented with a maximal mouth opening of 6 mm.

## DISCUSSION

Management of young patients with congenital ZMM fusion is challenging. The scarcity of cases makes



**Fig. 3.** Three-dimensional reconstruction of CT scan showing no bony fusion of the left maxilla and mandible. Fibrous fusion could not be excluded on these images (CT scan taken at the age of 4 years old, before second surgery).



**Fig. 4.** Active postoperative mouth opening of 15 mm, measured between incisal edges of the central deciduous incisors (photograph taken at the age of 4 and a half years old, after second surgery).

formulating practice guidelines very difficult. Individual case by case evaluation will always be needed because presentation of clinical cases is extremely varied. It needs to be considered that not all patients effectively need surgical release of fusion, as some patients have survived without treatment.<sup>9</sup> Young patients who present with ZMM fusion require long-term follow-up. Additional surgery at a later age will probably be needed. Costochondral grafts are the current standard of care in replacing the temporomandibular joint in the growing child.<sup>10</sup>

If surgery is performed, however, the importance of postoperative physical therapy cannot be overstated. Physical therapy needs to start early after surgery, needs to be prolonged, and needs to comprise aggressive physical exercises to maintain the best possible result. Multiple cases in the literature demonstrated significant relapse of maximal mouth opening when patients were unable to perform jaw-opening exercises after surgery.<sup>1</sup> At our center, physiotherapy is started 24–48 hours after surgery. During hospital admission, daily mouth opening exercises are performed and explained to the patient. We advise extensive and regular physiotherapy to be continued for the first 2 years after treatment.

### CONCLUSIONS

We believe it is vital to emphasize the importance of regular active physical therapy. However, young patients

with craniofacial deformities tend to have multiple health issues leading to extensive medical care. Unfortunately, this often leads to bad compliance with time-consuming physiotherapy and increased susceptibility of relapse in mouth opening.

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### DISCLOSURE

*The authors have no financial interest to declare in relation to the content of this article.*

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