Temporalmandibular Joint Ankylosis After Early Mandibular Distraction Osteogenesis: A New Syndrome?

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Abstract: Distraction osteogenesis (DO) has been one of the most innovative concepts in crano-maxillofacial syndromology and surgery over the last 25 years. Early mandibular distraction in severe micrognathia has recently been recognized as an effective treatment option to safely relieve upper airway obstruction associated with mandibular deficiency. An increased incidence in temporomandibular joint complications during DO in neonates has recently been reported, especially in syndromic patients.

The authors report 2 children affected by severe micrognathia and severe respiratory distress at birth. Early DO was performed during the first 2 months of the life in another institution with the aim of increasing mandibular length and upper airway size.

Both the patients had severe restricted jaw opening after DO and mandibular abnormalities.

Temporomandibular joint ankylosis after early mandibular distraction could be a considered a new pathological entity.

Key Words: Airway management, cerebro-costo-mandibular syndrome, mandibular osteogenetic distraction, pediatric surgery, Pierre Robin syndrome, TMJ ankylosis

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Temporomandibular joint (TMJ) ankylosis is a complex craniofacial disorder that involves fusion of the mandibular condyle to the glenoid fossa of the temporal bone leading to obliteration of the TMJ. Ankylosis causes significant mandibular growth retardation in both horizontal and vertical planes. The extent of growth retardation depends on onset, severity, and duration of insult to the TMJ. Unilateral TMJ ankylosis patients present with jaw asymmetry with or without retrognathia, while bilateral patients present with moderate to severe retrognathia leading to obstructive sleep apnea. Restoring normal anatomy and facial features is challenging in such patients. The main cause of ankylosis is trauma followed by local or systemic infection, such as ankylosing spondylitis and rheumatoid arthritis. Ankylosis can also be a result of inexact mandibular distraction surgery.

Pierre Robin syndrome (PRS) is characterized by microgenia with retrognathia cleft palate glossoptosis, and frequently is associated with airway obstruction and difficulty in swallowing. Respiratory obstruction may be severe and require immediate treatment at birth. Numerous surgical and nonsurgical techniques have been proposed for the treatment of severe respiratory obstruction and feeding difficulties.

Cerebro-costo-mandibular syndrome (CCMS) is a rare disorder with only a few reports in the scientific literature. It is a familial syndrome with an autosomal recessive pattern of inheritance.

The syndrome bears a resemblance to PRS as the affected infants demonstrate mandibular hypoplasi and glossoptosis with resultant feeding and respiratory distress. However, it differs in the associated rib-gap defects. In the algorithm of the treatment of micrognathia, distraction osteogenesis (DO) with micro-distractors has recently been considered a surgical option for syndromic patients during the neonatal age.

The treatment of airway obstruction in the newborn with PRS is complex; sometimes, it may constitute a real surgical emergency. The evolution of devices for osteo-distraction and the use of hybrid techniques have allowed surgical treatment in the first few months of life.

The complications associated with the use of these devices are, however, well documented and should be taken into account.

The choice of appropriate distraction vectors is of paramount importance to avoid TMJ excessive loading and to prevent TMJ ankylosis.

Reankylosis should be taken into account and aggressive physiotherapy is critical in its prevention.

The authors describe 2 patients of TMJ ankylosis after early mandibular distraction and the treatment protocol for recovery of morphology and function.

METHODS

The study involved 2 patients who had had obstructive sleep apnea syndrome since birth due to micrognathia.

Aggressive early mandibular distraction had been performed during the first few months of life in both the patients at another institution.

The first patient, with PRS, had undergone 2 previous mandibular distractions over the years.

At the age of 10 he was admitted to our institution with severe restricted jaw opening and coronoid hyperplasia (Figs. 1–3).

The second patient with micrognathia and bilateral TMJ ankylosis due to CCMS was admitted to our institute at 2 years of age after a previous mandibular distraction.

At the age of 3 months, following increasing difficulty in feeding and breathing, she underwent tracheostomy, percutaneous endoscopic gastrostomy position, and early mandibular distraction. After this surgery, the patient developed coronoid hypertrophy and severe TMJ ankylosis (Fig. 4).
The patient was admitted to our institution at the age of 2 with severe airway obstruction, tracheostomy, and reduction in jaw opening.

Both the patients were evaluated by a multidisciplinary team and underwent fibroscopic study.

**Treatment Plan**

The treatment plan for the PRS patient was as follows:

1st step
- Mandibular lengthening by DO (Distractor device Martin Zurich II).
- After 5 days of latency the activation rate was 1.00 mm per day based on a 2 per-day-rhythm. The activation period was 16 days with 60 days of consolidation period. The mandibular osteotomies were performed using an ultrasound bony device (Fig. 5). Removal of distraction devices occurred after 6 months.
- Jumping genioplasty.

2nd step
- Removal of the bilateral ankyotic TMJ block via a coronal approach (gap arthroplasty with interpositional silastic block).
- Coronoidectomy.
- Myofasciotomy.
- Forced opening oral cavity.
- Early mandibular rehabilitation.

**FIGURE 1.** Pierre Robin syndrome. Severe restricted jaw open less than 1 cm.

**FIGURE 2.** Pierre Robin syndrome. Lateral view: severe micrognathia and tracheostomy.

**FIGURE 3.** Pierre Robin syndrome. Computed tomography scan right temporomandibular joint ankylosis and coronoid hyperplasia.

**FIGURE 4.** Cerebro-costo-mandibular syndrome. Severe ankylosis and trachostomy.
The planning treatment of the patient with CCMS was as follows:

1st step
- Coronal approach, coronoidectomy, and removal of the ankylotic block.
- Fasciotomy, myotomy, and stretching.
- Forced mouth opening.
- Early aggressive physiotherapy with bite openers and stretching exercises.

2nd step
- Mandibular DO (Distractor device Martin Zurich II).
- After 5 days of latency the activation rate was 1.00 mm per day based on a 2 per-day-rhythm. The activation period was 16 days with 60 days of consolidation period. The mandibular osteotomies were performed using an ultrasound bony device. Removal of distraction devices was after 6 months.

3rd step
- Removal of mandibular distraction device and removal of TMJ reankylotic block (gap arthroplasty with interpositional silastic block).
- Myofaciotomy stretching of muscle.
- Forced mouth opening.
- Rehabilitation with physiotherapy in the immediate post-operative period with bite opening and stretching exercises.

RESULTS

Regarding the PRS case, after 1st-step distraction surgery the elongation was 18 mm (including 2 mm intraoperative elongation). Comparison of predistraction and postdistraction CT scans confirmed a significant improvement in airway space (Figs. 6 and 7).

Direct visualization with flexible endoscopy demonstrated the improvement of the airways space with elevation of the tongue, and epiglottis off the posterior pharyngeal wall. At this stage, no change in ankylosis was obtained.

After the 2nd-step surgery (TMJ ankylosis release) we obtained a 3.2 cm intraoperative mouth opening.

The patient was treated by aggressive postoperative physiotherapy and as a result a maximum mouth opening of 3.3 cm was achieved (Fig. 8). After 4 months of follow-up and after ensuring the airway was clear, tracheotomy was removed (Fig. 9).

In summary, after the surgical procedure the patient had a significant recovery of morphology, function, and psychosocial habits.

As regards the CCMS case, the first step involving ankylosis surgery with the removal of the coronoid process led to a temporary increase of the mouth opening (2.5 cm).
Even if an aggressive physiotherapy protocol was established with bite openers and stretching of muscle, an immediate relapse of the ankylosis occurred.

After the second step involving mandibular distraction surgery, we obtained a 24 mm opening (4 mm intraoperative) with a significant change in the face profile (Fig. 10).

Prior to the removal of the distractor device, X-ray and ultrasound echography showed the callous formation and the degree of ossification of the mandible.

Due to the persistence of the ankylosis and mandible hypomobility with feeding problems, tracheostomy was still kept in place for safety reasons and for preventing aspiration and dyspnea.

The third surgical step was the removal of the distractor devices and of the bilateral reankylositic TMJ block with the interpositional silastic. The intraoperative mouth opening was 3.00 cm. An intensive mandibular rehabilitation protocol began in the immediate postoperative period, but after 6 months of follow-up progressive closing and limitation of mandibular movement occurred (Fig. 11).

**DISCUSSION**

Distraction osteogenesis has been one of the most innovative concepts in craniomaxillofacial surgery over the last 25 years.

In 1987, Guerrero et al. first performed a mandibular widening by DO and McCarthy et al. in 1992 published an extensive paper on the treatment of hemifacial microsomia.
In 1994, Wangerin and Gropp\(^1\) and in 1996 Diner et al\(^4\) published on the use of intraoral devices for mandibular distraction. After a quarter of a century of extensive use DO has today specific indications for congenital craniofacial and cleft deformities.

Mandibular distraction has been reported to improve upper airway obstruction in PRS.\(^2\)

The mechanism of improvement of the airways is mainly based on the advancement of the mandible and subsequent anterior displacement of the tongue base and epiglottis away from the posterior pharyngeal wall.\(^5\)

Ortiz-Monasterio\(^6\) reported that 83% of patients with PRS showed gastroesophageal reflux disease with all patients having resolution of the reflux after airway control.

Treatment options in children with PRS include positional therapy, intraoral devices, nasopharyngeal and endotracheal intubation, glossopexy-tongue-lip adhesion, tracheostomy, and mandibular traction.\(^7\)

Recently, it has been reported that the wire traction of the lower jaw could be considered the first choice for severe breathing problems in newborns affected by PRS.\(^9\)

Tracheostomy is associated with frequent morbidity, high cost, and occasionally mortality. Decannulation may take several years and there is a significant psychological impact on the caregivers and family members.\(^15\)

Distraction osteogenesis of the mandible is a relatively new treatment in infants with PRS.\(^7\) Initially, reports of mandibular advancement during the first months of life were limited but many more studies can be found in the recent literature.\(^2\)

Temporomandibular joint ankylosis in children is uncommon and is one of the most difficult and complex problems managed by oral and maxillofacial surgeons.

Operations for TMJ ankylosis are known to be complicated based on recurrence. Over a period of 100 years various treatments have been proposed including gap arthroplasty, interposition arthroplasty, reconstruction of the ramal condylar unit with a costochondral graft, and total reconstruction of the joint.\(^8,12\) In addition to recurrence, any intervention used to treat ankylosis is complicated due to the presence of facial nerves, the maxillary artery, the pterygoid plexus, and the deranged anatomy of the joint itself. It is therefore crucial to select a technique that allows completion of the osteotomy safely with minimal risk to the surrounding structures.

In syndromic micrognathia early mandibular distraction seems to be an appropriate indication. Benefits are functional, morphological, and aesthetic, with consistent psychological and familiar effects. An increased incidence in TMJ complications during DO in neonates has recently been reported, especially in syndromic patients.\(^10\)

Fan et al\(^1\) demonstrated an increased incidence of condylar pathology preoperatively in syndromic micrognathia compared with nonsyndromic patients. Another mandibular anomaly that may contribute to poor functionally and decreased mouth opening includes excessive height and more cephalad positioning of the coronoid process.\(^4\)

In neonatal micrognathia distortion, Andrews et al\(^18\) reported complications including dental injuries (6%), neck scarring (8%), wound infections (5%), and TMJ ankylosis (10%). The last complication, TMJ ankylosis, was seen only in syndromic distraction patients.\(^18\)

Surgical planning for early mandibular distraction should take into consideration the appropriate distractor vector.\(^19\)

The distraction vector should be as horizontal as possible according to the mandibular morphology.

For the determination of the particular type of ankylosis present, CT views alongside panoramic views are important; among CT views, the coronal view of the TMJ gives the best identification of type III ankylosis. Three-dimensional documentation of the joint provided by a CT scan, especially the coronal view, helps in evaluating the nature and severity of the ankylosis in great detail.\(^20\)

A century of exil procedures have been proposed for TMJ release and reconstruction, namely, gap arthroplasty, interpositional arthroplasty, and joint reconstruction. The advantages of gap arthroplasty are its simplicity and short operating time, but the disadvantages include shortening of the mandibular ramus, open bite, and increased risk of recurrence of ankylosis.\(^2,21,22\)

According to Vasconcelos et al\(^23\) there is no consensus regarding the ideal interpositional graft. When the TMJ is anatomically and functionally impaired, joint reconstruction is suggested; a clavicular bone graft, coronoid process, iliac crest, and metatarsus can be used. Because of its growth, biocompatibility, workability, great adaptability to the glenoid fossa, and anatomical similarity to the condyle, a costochondral graft is considered the best option for TMJ reconstruction.\(^24\) In using alloplastic materials, the advantages are as follows: analog anatomy to real joints, absence of donor-site morbidity, possibility of restoring the vertical dimension, reduction in surgery time, and a lower risk of recurrent ankyloses.\(^25\)

According to Givens,\(^26\) surgical treatment should be chosen according to the extent and type of ankylosis, patient age, onset and time of surgery, and whether the ankylosis is unilateral or bilateral. Nogueira and Vasconcelos\(^27\) reported that the rate of facial nerve injury (31%) is related to the complexity of surgery.\(^27\)

In the reported patient, the initial distraction surgical plan considered a vertical vector and during the early distraction process caused the dislocation of the mandibular condyle and coronoid process toward the medial cranial fossa with bone fusion and hypertrophy of the coronoid process.

The PRS patient after 2 previous mandibular distractions in another institution reported severe restricted jaw opening, hyperplasia, and tracheostomy. We planned mandibular distraction with a horizontal vector, coronoidectomy with releasing of ankylosis, recovery of mandibular movements, and removal of tracheostomy.

The CCMS patient after mandibular DO at the age of 3 months with vertical vector had coronoid hyperplasia and complete fusion of the TMJ and coronoid process to the middle cranial fossa, with severe restricted mouth opening. After ankylosis surgery with bone removal and aggressive early physiotherapy and intensive follow-up, the child had severe reankylosis.

We planned a new horizontal mandibular distraction and surgery of ankylosis at the time of distractor removal. Even if the elongation of the mandible was quite satisfactory as well as the correction of the profile, recovery of function was not obtained due to reankylosis. The surgical treatment of ankylosis now needs to be delayed until the child is old enough to be cooperative and both child and parents are willing to have the operation.

Temporomandibular joint prosthesis reconstruction should be considered the treatment plan according to the age of the patient.

**CONCLUSIONS**

Mandibular deficiency with ankylosis represents one of the most challenging cases in craniofacial surgery. The critical question is what is the first surgical procedure should be? Management of TMJ ankylosis is difficult as no standardized surgical protocols have been suggested. Failure and reankylosis are common. In pediatric age, the aim of surgery is to recover morphology and function with normal growth, appropriate speech development, dental occlusion, and normal development of the facial skeleton.

Planning and selection of an appropriate distraction vector are of paramount importance in early DO to avoid severe complications.
REFERENCES