Bilateral Pediatric Mandibular Distraction for Micrognathia with Temporomandibular Joint Ankylosis and Sleep Apnea

Abstract
Mandibular retrognathism is one of the important contributing anatomical factors to the obstructive sleep apnea (OSA). Such patients suffer from number of apneic or hypopneic events during sleep such as snoring, daytime sleepiness, fatigue, inability to concentrate, and irritability. Distraction osteogenesis is a less invasive surgical technique in the management of OSA by correcting the reduced airway space. Apart from correcting functional disturbances due to OSA, it also corrects the facial profile resulting in the substantial improvement in cosmetic appearance. We report a case of a 3-year-old boy who was struggling with severe retrognathic chin and OSA causing hypopneic episodes and snoring. He was successfully treated by bilateral mandibular distraction which resulted in significant improvement of respiratory distress and feeding as well as evidential advancement of the mandible was achieved.

Keywords: Bilateral mandibular distraction, distraction osteogenesis, obstructive sleep apnea, pediatric distraction, temporomandibular joint ankylosis

Introduction
Retrognathism or retrpositioning of the lower jaw is reported to be a significant risk factor for the obstructive sleep apnea (OSA). OSA is believed to be a disorder of repeated upper airway collapse during sleep subsequently leading to oxygen desaturation that causes disrupted sleep pattern. Affected individual may present with snoring, witnessed apneas, daytime somnolence (excessive sleepiness), fatigue, behavioral, cognitive, and growth impairment. Increase of the airway space is the suggested rationale of treatment. Maxillomandibular advancement (MMA) is the preferred surgical approach to improve the volume of the airway space. It is reported that improvement of the airway dimensions by skeletal advancement of jaws remains the most effective surgical therapy for OSA. These jaw advancement surgeries when performed for larger dimension are often accompanied by a high rate of relapse. When performed at young age, the rate of relapse is higher as the growth centers continue to grow in spite of its newer position. The presence of the temporomandibular joint (TMJ) pathologies such as ankylosis complicates the issue.

Distraction osteogenesis (DO) is a relatively less invasive surgical technique which stimulates the native bone to expand in its dimension by vector control and has been reported to be a successful treatment modality in the management of OSA. The procedure also eliminates the need for bone grafting, less surgical dissection, and high degree relapse rates that are often associated with jaw advancement surgeries.

Case Report
A 3-year-old boy reported to our hospital with chief complaint of small lower jaw with fall back of the tongue and thereby causing obstruction in the airway and cessation of his breath intermittently. History revealed that the patient had difficulty in opening his mouth and swallowing, and heavy snoring was observed. History revealed by parents suggested that the boy had complications at birth and that there was no history of trauma related to the facial skeleton. The initial clinical examination revealed a hypoplastic mandible with chin receded and class II dental relationship. Mouth opening was minimal, and there were no TMJ movements palpable. The oral cavity appeared to be small leading to the diagnosis of micrognathia. On examination of the three-dimensional computed...
tomography, the patient had hypoplastic mandible with bilateral TMJ ankylosis [Figure 1].

This case featured a stage 2 ankylosis, where the extension of the lesion had involved the sigmoid notch. It was a Type IV classification of ankylosis where the joint is totally obliterated by an expanded bony block between the ramus and the skull (Sawhney 1986).

On intraoral examination, dentition was present in both the jaws, and a small submucosal cleft in the palate was noticed. The relatively reduced tongue space due to micrognathia was what had caused the OSA. No evidence of abnormality in the brain and other intracranial structures was found. No related records were available to identify the cause of TMJ ankylosis or micrognathia or its association. Diagnostic sleep apnea study indicated a poor compromise in oxygenation during sleep and reduced CPAP (Continuous Positive Airway Pressure) with intubation.

The goal of the therapy was to initially relieve the patient from OSA symptoms by increasing the airway volume. The TMJ ankylosis posed an additional challenge, as it would prevent the growth or movement of the lower jaw. The age of the patient was not in favor of treating the TMJ ankylosis as the desired growth was not yet achieved nor it was advisable to postpone the surgery until patient attains the desired age for relieving the TMJ ankylosis. Hence, DO was chosen as the treatment of choice. The treatment plan was to widen the TMJ using a horizontal mandibular ramus distractor. The distraction movement and vector placement were planned in such a way that the desired growth could be achieved at the end of activation period.

**Surgical procedure**

A fiberoptic-guided nasal intubation was performed initially for the administration of general anesthesia (GA) as the patient had reduced mouth opening due to bilateral TMJ ankylosis. While the patient was under anesthesia, patient’s neck was prepared and infiltrated with local anesthetic, and tracheostomy was performed to prevent the fall back of the tongue postoperatively. As the tracheal ring was incised and retracted, the nasoendotracheal tube was visualized which was removed, and a tracheostomy tube was inserted through which the GA gases were administered for further surgical procedure [Figure 2].

The surgical site was prepared using povidone-iodine disinfectant and patient was draped in sterile towels. Intraorally, buccal vestibular incisions were placed approximately 5 mm below the attached gingiva on either sides of the mandible. The mucoperiosteal flap was raised and as per the presurgical planning, an osteotomy cut was placed through the outer cortex of the mandibular body where there were no tooth buds. Intraoral horizontal ramus distractors (KLS Martin, Germany) were used on both sides, and bur holes were drilled for the adaptation of the distractor plates. Care was taken not to injure the inferior
alveolar nerve path and all developing tooth buds. After placement of the distractors, the function, position, and occlusion were checked. Once the position and movements of the distractors were confirmed, the screws were fixed after which the lingual cortex was split using an osteotome. Similarly, the opposite side was also split. Once again, a thorough final checkup of the function of both the internal distractors was performed, and the intraoral incisions were sutured [Figure 3].

After 4\textsuperscript{th} day, the distractors were activated to achieve about 25 mm of mandibular length. Initially, 1 turn of 0.5 mm distraction (0.5 mm/day) was performed until 5 mm was achieved followed by 2 turns of 1 mm distraction (2 mm/day), and finally, again, 1 turn of 0.5 mm/day distraction regimen was performed. Thus, completing 25 mm, sutures and tracheostomy tube were removed. Distractors were left in situ till the completion of consolidation (mineralization) period. After a period of 3 months, distractors were removed without any complications, and desired results were achieved. Postoperative sleep apnea studies too revealed better oxygenation and positive favourable response in CPAP. Later, surgery was planned for correction of TMJ ankylosis [Figure 4].

**Discussion**

The abnormal anatomical component of micrognathia with retrognathic chin restricts the airspaces compromising the airflow. Such restricted airflow leads to myriads of health consequences, some with disastrous consequences including hypoxia, hypercapnia, pulmonary heart disease, and pulmonary hypertension on a longer term. Till the last decade of 20\textsuperscript{th} Century, the “gold standard” of treatment for OSA was tracheostomy. This invasive procedure was associated with a high morbidity such as tracheomalacia, chronic bronchitis, laryngeal stenosis, and risk of death due to mucus plugs or extrusion/dislocation of the cannula.\textsuperscript{[6,7]} Long term dependence on tracheostomy has its own limitations leading to diminished quality of life. The other alternative procedures included nasal reconstruction, uvulopalatopharyngoplasty, advancement genioplasty, bilateral sagittal split ramus osteotomy with advancement genioplasty, and inferior mandibular osteotomy with hyoid myotomy. In most severe cases, MMA with advancement genioplasty was indicated. All these procedures are extensively invasive which have their own anatomical and physiological complications, especially in a growing child.\textsuperscript{[8]}

Since 20\textsuperscript{th} century, DO has been emerging as an alternate treatment modality for patient suffering from OSA. DO offers several advantages over the other conventional techniques by eliminating the need for bone grafting and involving less surgical dissection because the lengthening is the result of natural bone healing in a gap created by a simple osteotomy. The incremental skeletal movement permits accommodation of the soft tissues as well as promotes its growth. The procedure also could help to lengthen or widen the bone by a determined vector in a controlled fashion. A larger dimension of skeletal movement was realized with minimum risks, complications, and morbidity. The associated soft tissue structures such as muscles, blood vessels, and nerves are also newly formed along with the neoosteogenesis. This accounts to the increased better soft tissue adaptation, a factor that was not achieved by conventional techniques. Although less surgical dissection is necessary for DO, the procedure is highly technique sensitive, especially in achieving the
proper alignment of the distraction devices as well as vector control.[9]

The presence of TMJ ankylosis complicated the OSA scenario. We could not decipher whether the micrognathia and genial retrusion are the cause or the effect of the TMJ ankylosis. The net result was that TMJ ankylosis and mandibular retrusion restricted the airspace. Correction of the TMJ ankylosis at the age of 3 years was not preferred as there were other local issues – retruded mandible. Correction of TMJ ankylosis in children is a matter of debate. While some suggest deferring the surgery till oral functional movements are achieved, some are in favor of early intervention.[10] In the present case, it was decided to defer the surgery till the growth of midface stabilizes.

To correct the severe OSA that the child suffered from, we had no other better option to perform than the DO of the mandible. In a growing child, mandibular elongation by progressive distraction is gradual and thus better tolerated by our patient, both functionally and morphologically. The procedure was less time consuming, and placement of the distractors causes no particular problem apart from the care required in achieving the proper vectors of distraction. The development of miniaturized intraoral distractors and reduction in their cost should result in more widespread use of the procedure.[11]

There are recent reports such as that by Hu et al.[12] that has performed such a procedure on adults and achieved success while there are very few reports of the same in children as old as 7 years.[13] In the present case, the procedure was successfully performed in a 3-year-old patient.

Procedurally, to facilitate administration of anesthesia and sustain the space till sufficient airspace was created, the tracheostomy was performed. During the procedure and in immediate postoperative situation, the child had no issues with this approach. The desired airspace was achieved.

The case is being reported for its unique challenges, a bilateral TMJ ankylosis in a 3-year-old patient, probably affecting the jaw growth centers while reduced airspace volume, causing severe reduction in quality of life, and was successfully rehabilitated with DO [Figure 5].

**Conclusion**

DO is a powerful tool for surgical reconstruction of complex jaw deformities. The goal of treatment in infants with severe micrognathia is to focus on breathing and feeding and to optimize growth and nutrition. It appears to be the superior method of reconstruction technique when performed correctly and effectively lengthens the jaws without the use of grafts.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient’s parents have given consent for images and other clinical information to be reported in the journal. The patient’s parents understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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