



SURGICAL MANGEMENT OF MANDIBULAR HYPOPLASIA USING MANDIBULAR DISTRACTOR

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Abstract:

Management of Pierre robin syndrome nonsurgically or surgically is still controversial as mandibular hypoplasia is one of the critical feature seen in Pierre robin sequence. It could cause respiratory distress. Mandibular distraction is a innovative and current technique in managing such hypoplastic mandible .Mandibular distraction involves use of extraoral or intraoral mandibular distractor to increase the ,mandibular length , increase pharyngeal and airway space

Introduction:

Mandibular hypoplasia is one of the feature seen in pierre robin sequence . If associated with glossoptosis and u shaped incomplete cleft palate it is known as Pierre robin syndrome(PBS) Risk of death exists in the first moments of life for a infant wih pierre robin's..Micrognathia could be one of the causes for respiratory distress after delivery . Glossoptosis and posterior displacement of tongue attributes to this This trifecta shows a deleterious effect on varied stages of infancy.As oral cavity is a conduit for gastrointestinal tract ,feeding problems are also common .Depending to the severity ,this can lead to issues like choking , less weight gain and also associated gastroesophageal reflux in children with PBS.Other manifestations include heart murmurs, pulmonary hypertension and pulmonary stenosis .

Management of Pierre robin syndrome nonsurgically or surgically is still controversial . Management involves non –surgical management involves positioning in prone or lateral position ,tongue lip adhesion ,use of nasopharyngealcannula .Other surgical methods involves mandibular distraction and tracheostomy . Tracheostomy, which bypasses the site of obstruction is highly effective but the number of complications associated are so numerous that it is used in extreme situations.

Mandibular distraction is a innovative and current technique in managing such hypoplastic mandible .Mandibular distraction involves use of extraoral or intraoral mandibular distractor to increase the ,mandibular length , increase pharyngeal and airway space .

CASE REPORT :

A 10 month old male infant diagnosed with Pierre robin Sequence was referred to the head and neck opd with severe gastric regurgitation and mild hypoventitation . The parents gave a history of airway complications at birth but was managed non surgically . Patient presented to the department with nasogastric tube since birth with severe gastric refluxpatient was obese with delayed normal milestones He presented with typical “bird face deformity “ with ,retrognathic mandible , negligible neck throat angle ,inability to open mouth . Ct scan showed retrognathic mandible with elongated coronoid process ,absence of definite gonial angle bilateral ,ill-defined sigmoid notch..



Pre- distraction

Based on the CT scan and age of the patient bilateral mandibular distraction osteogenesis (MDO) with single vector system (intraoral mini distractor for mandible ,orthomax baroda-no affiliation ,no conflict of interest)was planned.

Surgical technique :

Bilateral mandibulotomies and placement of distraction devices :

Extraoral approach 2-3 cm incision was placed along the relaxed skin tension line of the neck , at least one finger breadth below the mandible. A “vertical mandibulotomy” is performed bilateral using a piezoelectric device just anterior to the angle of the mandible . The mandibulotomy is kept incomplete to avoid minimal damage to the inferior alveolar nerve .The distractor is placed perpendicular to the placed vertical cut bilaterally . The mandibulotomy is then completed using a combination of straight osteome and piezoelectric device . A trial of linear distraction is performed to rule out green sticking .A punchhole was made bilateral to create a portal for the key of the distractor and closure was done



Skin incision placed



Osteotomy cut with distraction

The patient was kept on 72 hour latency period .Activation is started on the 4th day twice daily with 12 hours apart.The patient was kept on intravenous antibiotic and analgesic and oral thereafter .throughout the entire distraction period , nasogastric feeding is continued in order to avoid microleakage of food debri .Activation was done for 15 days or till the distraction key stops turning .Therafter consolidation phase begins where in patient is regularly monitored for weight gains and post operative pin care . The consolidation period is kept for 45 days



Post consolidation period

The distractor is then removed via the same scar extraorally under a light anesthesia and scar is revised



6 month post distraction

Discussion:

PRS patients develop respiratory distress caused by glossoptosis and micrognathia ,but beyond this problem can also cause difficulty in eating and talking to a varied degree.It may or maynot be associated with syndromes like sticklers(commonly seen associated with PRS) with associated 22q 11 DS genetic sequencing.

In patients with severe micrognathia ,the tongue takes up proportionately more volume in the oropharynx ,resulting in glossoptosis in the setting of small mandible Due to this abnormal positioning of the tongue and size of the mandible occlusion of the nasal and oral pharynx occurs creating not just a problem for breathing but also for oral intake and increase chances of aspiration when food is taken orally.

Feeding difficulties are common as infants struggle to breathe during eating. Gastroesophageal reflux and aspiration are common sequelae of this process.. Airway obstruction and resulting intrathoracic pressures have been identified as causes for increased gastroesophageal reflux.⁹ This when presented along with cleft palate ,causes poor calorific intake and are unable to gain weight during early postnatal period ..

Conservative management like positioning in prone or lateral position ,tongue lip adhesion ,use of nasopharyngeal cannula , But use of nasopharyngeal cannula requires constant review and can cause obstruction of tube for mucosal secretions or aspiration of gastric contents .¹⁵

Surgical advances involves Mandibular Distraction osteogenesis (MDO) .It is most stable and an elective procedure which allows the advancement and elongation of jaw as a result of intial osteotomy and progression of tongue in order to reduce supraglottic obstruction.It involves 3 stages :latency , activation and consolidation phase .Latency period according to literature suggests 0-2 days.. During activation phase, mandibular distraction occurs at a specific speed, generally 1 mm per day (0.5 mm in the morning and 0.5 mm in the evening). The length of distraction is strictly dependent on the severity of the defect in order to obtain optimal functionality or to correct some types of malocclusion such as open bite and asymmetry. The osteotomy is preferred just adjunct to the angle of mandible as least number of tooth buds as present causing minimal damage in the future .This also reduces changes of infection of the neo-callus formed .A unidirectional vector distractor is preferred as the growth of the callus can be easily moulded in a growing child .

As a highly invasive surgical technique, the possibility of postoperative complications is inevitable: temporary paresthesia or minor alveolar nerve damage, facial nerve or mandibular

marginal nerve injures, altered or failed consolidation of the mandibular segments, wound infections, dental damage and ankylosis of the temporomandibular joint. The consolidation period is 6- 3 weeks .As infants have a faster osteogenesis potential , a maximum of 3 weeks gives enough time interval to consolidate the new callus formed .

A 3-5 mm of relapse is noted post 1 month of distraction to allows tissue to settle and bone remodeling to occur . It is necessary to emphasize that tracheostomy aims to treat immediately clinical symptoms but has no influence on pathophysiology of Pierre Robin syndromeThe only impediment is to detect the unpredictable growth of the neo- bone formed in a growing child and requires long term follow up till growth spurt is complete

Conclusion :

Any mandibular hypoplasia which may or maynot be associated with PRS should be corrected with mandibular distractor immediately at birth . This technique opens airway immediately at birth and provides better clinical results as the child grows .

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