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# Airway-Compromising Mandibular Hypoplasia in Neonates

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The use of distraction osteogenesis (DO) for neonates (less than 3 months old) with mandibular hypoplasia has been helpful in relieving airway and feeding difficulty. Surgical approaches to managing upper airways in Pierre Robin sequence (PRS) infants have included tracheostomy, glossopexy, hyomandibulopexy, tongue-lip adhesion, circummandibular wire, and subperiosteal release of the floor of the mouth. Tracheostomy has been considered by many to be the gold standard of treatment in airway difficulties, but it has not been without its detractions. There are potential complications, such as granulation tissue formation, innominate artery hemorrhage, pneumothorax, tracheal tube obstruction, cricoid cartilage injury, and accidental decannulation. Long-term problems include delayed development of speech/language skills, pulmonary infections, behavioral problems, and problems with parent-child social interactions.

Neonates with airway-compromising retrognathia usually have a nonspecific working diagnosis of PRS. PRS has been described with both syndromic and isolated, nonsyndromic variations. In the isolated, nonsyndromic variation, the clinical triad of cleft palate, micrognathia, and glossoptosis and the associated feeding difficulties and airway disturbances are the only malformations noted. In the syndromic variation, the clinical triad and associated feeding and airway maladies are only part of a greater number of malformations associated with a genetic or developmental syndrome. The syndromic associations include Stickler syndrome, velocardiofacial syndrome, fetal alcohol syndrome, Treacher Collins syndrome, distal arthrogryposis, and chromosome 6q deletion.

Upper airway obstruction in infants has been associated with failure to thrive, gastroesophageal reflux, hypoxia, hypercapnia, cor pulmonale, neurologic impairment, and death. Mortality rates reported have varied widely, from 5 to 65%, and death has been thought to be a consequence of the combined effects of malnutrition, exhaustion, pulmonary sepsis, and/or sudden cerebral anoxia. The mortality rate has been reported as 22.8% in PRS associated with a syndrome but only 5.9% in nonsyndromic PRS patients.[1]

## Indications

The indications for DO in the neonate should be relatively rigid and straightforward. Severe airway compromise is the main indication for the use of DO in neonates. The neonate should be airway compromised to the point that the consideration of a tracheostomy has been made. The threshold for making the decision may be lowered during the winter months as the potential for upper respiratory infection increases. An example would be a neonate who is born in the month of November and on initial evaluation is able to deal with airway compromise by positional control of lateral and prone sleeping positions. This same infant who has an upper respiratory tract infection 3 months later may have severe airway compromise in any position. This infant could now be in mortal danger if certain care is not taken. This is especially true in a family situated in a rural area without immediate specialized care.

The second indication is feeding difficulty in the infant (greater than 3 months old). This indication may be controversial but has come about through the observation of the feeding ability of multiple airway-compromised infants. In virtually all of the DO cases performed by this author, the feeding in distracted neonates was essentially normalized. The neonates who had cleft palates still had to use some form of cleft feeder. The timing of mandibular advancement in this situation may be variable owing to the possible placement of gastric tubes.

## Evaluation

The nature and mechanisms in airway obstruction in PRS patients are multifactorial, and the treatments proposed reflect this. There is great variability in the severity of airway compromise and in the treatments designed to manage the airway compromise.

The evaluation should take place as rapidly as possible, and early surgical intervention should be accomplished if possible. The age range of neonatal patients when DO was performed at our institution was 10 to 68 days, with a mean age of 26.4 days ($n$ = 10). Additionally, DO was performed on two infants over 6 months of age, with an average age of 245 days.

The neonate should be stabilized and airway temporization accomplished prior to beginning any studies. Initial studies include a polysomnographic (PSG) study if possible. This can usually be done at the bedside with or without oxygen, and the neonate may be awake or asleep. If the neonate requires intubation or a laryngeal mask airway, a sleep study cannot be obtained. The PSG study, if done, will give information as to the severity of the obstruction and desaturation levels in an objective manner.

Thorough evaluation of the airway aimed at determining the mechanism of the obstruction should be performed prior to initiating long-term management. Sher used flexible fiber-optic nasopharyngoscopy to identify the nature of airway obstruction in patients with craniofacial anomalies, including PRS.[2] The author reported on 53 cases of PRS and categorized the type of obstruction found in relation to the primary diagnosis (syndromic vs nonsyndromic). In this study, 53.5% of the patients had type I obstruction, 20.8% type II, 9.4% type III, 9.4% type IV, and 1.9% other. The four types of obstructions were described as follows:

- Type I: Posterior movement of the dorsum of the tongue to the posterior pharyngeal wall. This is primarily an anteroposterior-positioning problem.
- Type II: Posterior movement of the tongue, but the contact is with a long soft palate or cleft palatal tags, which then impinge on the pharyngeal wall.
- Type III: The lateral pharyngeal walls move medially, causing them to approximate.
- Type IV: Circular or sphincteric constriction of the pharynx, with movements occurring in all directions.

This information is helpful in predicting if the distraction surgery will be helpful in the resolution of the airway compromise. It is of the utmost importance that an airway evaluation is performed, not only for the above reasons but also to evaluate the subglottic airway. This evaluation has cancelled the impending distraction surgery in two cases when subglottic stenosis was found to be the cause of the airway compromise. If DO would have proceeded, certain failure to resolve the airway obstruction would have resulted.

High-quality three-dimensional computed tomography (CT) and the fabrication of a stereolithographic model are recommended. The use of stereolithographic models for presurgical evaluation and planning has been shown to decrease the operative time.[3] ClearView Stereolithographic Models from Medical Modeling Corporation, Golden, CO were used; vital structures, such as the inferior alveolar nerve and the developing tooth buds, are visible in these models, and the location of the corticotomy was planned with knowledge of these structures. The distraction appliance can be prebent to fit the mandibular contours present in each case, which significantly reduces operating time and improves accuracy. In most instances, the turnaround time for the fabrication of these models was 24 to 48 hours so as not to delay surgery.

Along with a thorough systemic evaluation, additional consultations would include genetics, cardiology, pulmonology, neurosurgery, and other members of the craniofacial team as needed.

## Biologic Foundation

The use of DO in neonates for mandibular advancement is a sound treatment in that the majority of cases that have had endoscopic evaluation show a type I or II airway. In these situations, the advancement of the mandible will, in most instances, resolve the compromising position of the tongue base. A video of the tongue position prior to and after mandibular DO advancement shows tongue repositioning from the dorsal side of the soft palate to a normal floor of the mouth resting position. This change in position normalizes the airway and feeding. The likely failures of DO would include neonates with Treacher Collins syndrome and other craniofacial anomalies, including malformation of the ramus, condyle, and glenoid fossa. These types of cases need additional bone stock that may not be satisfied by DO alone.

Several authors have observed the relief of upper airway obstruction by the use of mandibular DO. Long-term results describe stabilization of the airway obstruction and better feeding by the infant.[4,5]

## Advantages and Disadvantages

The advantages of DO for these patients would include the ability to avoid a tracheostomy or progression to decannulation if the patient already trached. Normalization of oral feeding is also an advantage. The benefits of these changes would be physical and psychosocial. The physical benefits are evident, but long-term follow-up will be needed to determine the longitudinal effects on dentition and mandibular growth. The longest follow-up for neonates treated with DO by our institution is 5 years, and clinical and radiographic evaluations show that there is no growth disturbance in the mandible and all primary teeth are developed and erupted normally. This patient does not have any airway or feeding difficulty as recorded by subjective parental observation and objective PSG evaluation postsurgically. The interaction and socialization of toddlers may progress more readily without the tracheostomy. The reluctance of some medical professionals and certainly the insurance companies to partake in quality of life issues probably underestimates this psychosocial benefit.

## Technique

Once the stereolithographic models have been obtained, the surgery is planned. The location of the corticotomy is planned to avoid vital structures and to allow distraction in the desired direction (Figure 37-1). The distraction appliance is adapted and cut to fit the stereolithographic model at this time and then is sterilized prior to placement at the time of surgery. The vector of the appliance is parallel to the inferior border of the mandible.

We are currently using the KLS Martin L.P. (Jacksonville, FL) micromandibular internal distraction appliance (Figure 37-2), which is available in 10, 15, and 20 mm distraction lengths. The appliances are contoured on the stereolithographic models so that they are as parallel as possible. Access to the surgical site is via a submandibular approach. It is not advised to try the intraoral route to place the device. This will compromise paralleling the devices with each other and the inferior border. Additionally, many other surgeons have called our institution after failed attempts via the intraoral approach. The appliance is positioned but not secured. The corticotomy is marked on the mandible as was predicted on the stereolithographic model, the appliance is removed, and an incomplete corticotomy is performed with rotary instrumentation using a small-diameter taper fissure bur (701). The corticotomy is extended to all aspects of the mandible that can be readily visualized. The distraction appliance is then anchored to the mandible using 1.0 mm bone screws, avoiding vital structures, as seen on the stereolithographic model. Do not use self-drilling screws as the neonate mandible will splinter. Always drill the hole for screws using the appropriate drill. The appliance is activated, and then the corticotomy is completed using osteotomes if needed. Often activation of the appliance alone will complete the corticotomy (Figure 37-3). The

**FIGURE 37-1** Stereolithographic model fabricated from computed tomography for preoperative surgical planning.

**FIGURE 37-2** A 20 mm micromandibular distraction device produced by KLS Martin.

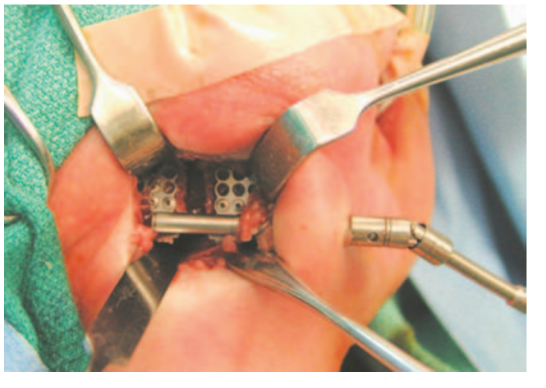

**FIGURE 37-3** Mandible is exposed from a submandibular approach. The prebent distraction device is placed appropriately and the mandible is marked, the appliance is removed, and superior, lateral, and inferior border corticotomy is performed. The corticotomy is carried on the medial side of the mandible as far as can be visualized. The appliance is now returned and secured to the mandible. Do not use self-drilling screws. The appliance is then activated, and the mandibular corticotomy is now completed with osteotomes. The appliance is now returned to its original position.

drive mechanism is a simple screw in an antirotation slot drive and exits the skin via a small submental incision. The drive shafts are available in various lengths to accommodate transcutaneous placement and are available in rigid and flexible designs. One-dimensional DO is obtained with this appliance. The submandibular incision is closed in an aesthetic manner (Figure 37-4).

Owing to the rapid healing responses of neonates, our center does not use a latency period. The distraction process is initiated immediately and proceeds at a faster rate than for adults. Our treatment protocol is as follows:

1. No latency period
2. 1.5 mm DO per day
3. Three activations per day (0.5 mm per activation)
4. Minimum of 6 weeks' consolidation after DO is completed
5. Removal of the device through the original incision with aesthetic closure

## Morbidity

In our experience, three complications have occurred. The first was a minor wound infection that was readily treated with antibiotics and wound care. Scarring was seen on all cases but was not significant owing to its submandibular placement. The third complication occurred when the endotracheal tube was inadvertently dislodged during the course of the surgery. This occurred after general anesthesia had been initiated and just as the surgery was about to begin. During efforts to reintubate the patient, the patient suffered barotrauma to both lungs when being ventilated using a face mask. The surgery was stopped, and tracheostomy and chest tubes were placed to treat the bilateral pneumothorax. The patients' lungs were allowed to heal, and, subsequently, the distraction appliance was placed uneventfully; the infant ultimately was decannulated, had resolution of apnea, and resumed normal feeding.

**FIGURE 37-4** Postoperative view of a patient showing paralleling of the right and left appliances as closely as possible.

Facial scarring, sensory and motor nerve damage, major hemorrhage, and loss of fixation should be considered the highest occurring risks. There has not been one case of a lack of consolidation at our institution. In fact, most patients show exuberant bone growth to the extent of bone covering the mesh panel of the distractor.

The dentition has been disrupted in very few of the patients, with malpositioned impacted teeth being the most common problem (Figure 37-5).

## Cost

The cost of treatment is approximately $10,000 in undiscounted fees (prior to insurance write-offs). This includes surgical fees and appliances. Billing should include mandibular osteotomies and a code for an unlisted procedure to account for the time to care for the DO and care of the consolidating mandible. At this time, there are no current procedural terminology (CPT) codes for DO. The hospital costs for these types of cases are high owing to intensive neonatal care, CT scans, and the number of specialists involved during the treatment of a neonate.

The comparison of alternative surgical intervention (DO) with tracheostomy shows that the costs of tracheostomy care, supplies, nursing, and doctor visits quickly outpace the cost of the alternative surgery if successful. Cohen and colleagues reported that the breakeven point occurred at 6 months.[6]

## Case Report

A below-normal birth weight neonate with no diagnosed genetic abnormalities underwent consultation by the cleft craniofacial team for acute airway obstruction. The airway was stabilized with an oral airway. The clinical examination was consistent with a working diagnosis of PRS. The mandibular hypoplasia and glossoptosis caused airway collapse, and a tracheostomy was being considered (Figure 37-6). A polysomnogram showed an apnea index of 8.3 with supplemental oxygen. Preoperative CT scans were made, and a

**FIGURE 37-6** A 25-day-old neonate lateral profile view prior to distraction osteogenesis. A surgical airway was elected owing to the inability to fiber-optically intubate the neonate.


**FIGURE 37-5** Changes in dentition include malpositioned teeth.

stereolithographic model was obtained. The neonate was taken to the operating room for DO at 25 days. A surgical airway was established after noninvasive attempts made to secure the airway failed.

Bilateral DO devices were placed through submandibular incisions, and after 1 cm DO had occurred, the bone was allowed to consolidate for 6 weeks. During this consolidation time, the neonate had started normal feeding with a Haberman nipple (Figure 37-7). The DO devices were removed, and the neonate was decannulated while in the operating room at 6 weeks post-DO and was again discharged without respiratory difficulty and normal feeding. A postoperative polysomnogram at 9 months showed an AI of 0.0. The 4-year follow-up showed good continued growth (Figure 37-8) and no interference with tooth formation and eruption. The occlusion is slightly Class III.

**FIGURE 37-7** Post–distraction osteogenesis lateral profile view 10 days after surgical placement of distractors.

**FIGURE 37-8** A 48-month postoperative lateral profile view shows a slightly concave profile that is consistent with Class III malocclusion. No airway or swallowing difficulty was noted at the examination.

## Conclusion

The use of DO for the airway-compromised neonate is a valuable tool in the resolution of breathing problems. It may also be an option for correction of eating or swallowing difficulty in the retrognathic neonate. Thorough evaluation needs to take place prior to the surgical decision to distract to ensure that noninvasive methods will not yield positive airway changes. Meticulous attention to the placement of the devices and the resultant vectors need to take place to prevent open bite–type malocclusions after DO is completed.

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