Mandibular Distraction to Relieve Airway Obstruction in Children With Cerebral Palsy

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Objective: To determine the effectiveness of distraction osteogenesis of the mandible for relief of airway obstruction in neurologically impaired children.

Design: Prospective pilot study.

Setting: Tertiary care children’s hospital.

Patients: Five children with upper airway obstruction secondary to hypotonia were identified by airway endoscopy, pulse oximetry, and polysomnography. Four children were tracheotomy-dependent because of upper airway obstruction, and 1 was being considered for tracheotomy because of progressive airway obstruction.

Interventions: All patients underwent distraction osteogenesis of the mandible for relief of their airway obstruction.

Main Outcome Measures: Treatment success was determined by endoscopy, continuous pulse oximetry, and polysomnography.

Results: Four of the 5 children underwent distraction osteogenesis of the mandible with successful resolution of airway obstruction or tracheotomy decannulation. One child did not show adequate improvement of upper airway obstruction and remained tracheotomy-dependent. Follow-up was 2 to 40 months.

Conclusions: Children with cerebral palsy and hypotonia of the upper airway may achieve relief of their chronic airway obstruction following distraction osteogenesis of the mandible. Appropriate selection criteria must be adhered to, including demonstration of tongue base obstruction on flexible laryngoscopy.


Children with cerebral palsy (CP) and neurogenic disorders with associated muscle hypotonia are at high risk for developing significant upper airway obstruction and obstructive sleep apnea. Furthermore, the frequent occurrence of seizure, gastroesophageal reflux, and hypersalivation disorders in these children can exacerbate the observed apnea. For these reasons, the airway management of these patients has been difficult. In most instances, these children become tracheotomy-dependent, leading to severe deleterious effects on the development of communicative skills and to significant financial, medical, and social burdens on the families. Long-term, tracheotomy-specific mortality rates of 0% to 3.4% have been reported in these children. Compared with children with nonneurogenic upper airway obstruction, children with neurogenic disorders have significantly longer periods of tracheotomy dependency, often permanent, with inability to achieve decannulation. Multiple studies have shown that aggressive surgical management in these children may avoid the long-term dependency and hazards of tracheotomy. Surgical management algorithms usually dictate tonsillectomy, adenoidectomy, and uvulopalatopharyngoplasty as initial procedures for treatment of the upper airway obstruction. Children with neurologic disorders often have obstruction that is refractory to these procedures. Different surgical techniques have been used to achieve decannulation or to avoid tracheotomy. These have included tongue reduction, tongue-hyoid suspension, tongue-lip adhesion, septoplasty with turbinectomy, salivary gland excision, epiglottoplasty, and mandibular advancement. Recently, distraction osteogenesis of the mandible has been established as a successful method for the treatment of tongue base airway obstruction in the pediatric population.
rologically induced hypotonia can lead to tongue base airway obstruction secondary to floppy, posteriorly displaced tongues and poor pharyngeal muscle tone. Theoretically, the generalized hypotonia may lead to micrognathia, as the forward thrust of the tongue and floor of mouth muscles are important for normal mandibular growth and development. Hypotonic, akinetic mice have been shown to develop micrognathia.8

This study was undertaken to determine the effectiveness of distraction osteogenesis of the mandible for relief of airway obstruction in neurologically impaired children. Specifically, the rate of avoidance of tracheotomy or ability to achieve decannulation was analyzed.

METHODS

PATIENTS

The study was conducted at a tertiary care children’s hospital from June 1, 1997, to July 31, 2001. The patients consisted of 5 children, 3 boys and 2 girls, with upper airway obstruction secondary to neurogenic disorder–related muscle hypotonia. The obstruction was identified by airway endoscopy, pulse oximetry, and polysomnography. Four children were tracheotomy-dependent because of upper airway obstruction, and 1 was being considered for tracheotomy because of progressive airway obstruction. All children had been followed up since birth and monitored before and after distraction osteogenesis of the mandible in a prospective fashion, with each child serving as his or her own control. In addition to the airway obstruction, all of the children had developmental delay, failure to thrive, seizure disorders, and gastroesophageal reflux disease, among other medical comorbidities. Associated problems in some included posthypoxic encephalopathy, microcephaly, dysphagia, and hypersalivation.

EVALUATION

All children were evaluated before surgery with flexible endoscopy, focusing on the tongue base airway obstruction. Dynamic endoscopic evaluation was accomplished in the operating room with the children sedated but spontaneously breathing (mimicking the condition of sleep, in which muscle relaxation seems to compound the airway collapse and obstruction). Jaw thrust maneuvers were performed while viewing the site of obstruction to visualize the effect of mandibular advancement on overall tongue base airway patency. The endoscopic evaluation was repeated once during the distraction process, under sedation in the operating room, and after decannulation. All children underwent preoperative 3-dimensional and routine computed tomographic imaging of the mandible to aid in surgical planning.

MANDIBULAR DISTRACTION

The surgical technique of mandibular distraction osteogenesis has been described in detail.7 In summary, under general anesthesia, the mandible is approached intraorally (Figure 1). Osteotomies are done anterior or posterior to the mandibular angle but not through it. These are accomplished by first creating periosteal pockets, exposing the lingual and buccal cortices. Outer corticotomy is then carried out with a side-biting burr (Figure 2). The site of the pins is then determined using a 25-gauge needle, planning placement of the nearest pins 5 to 8 mm from the osteotomy line and marking the corresponding skin sites. Small 5- to 10-mm incisions are made through the skin, and blunt dissection parallel to the facial nerve is carried down to the mandible. The pins are drilled into solid, inferior mandibular bone with a firmly positioned 2-hole drill guide (Figure 3) and a handheld drill (Figure 4). The pins are bicortical. Two pins are placed on either side of the osteotomy in as straight a line as possible. The distractor is then fitted over the pins until a point 5 to 10 mm from the skin. The distractor is conformed to precisely fit the pins at this point, and not after completion of the osteotomies (Figure 5). After determining what the position of the distractor will be, it is removed and the osteotomies are completed. The osteotomies are con-
tined from buccal to lingual along the superior and inferior aspects of the mandible but not through the middle. The final separation is accomplished with a chisel so as to not injure the inferior alveolar nerve (Figure 6). After ensuring that the osteotomy is indeed complete by passing a nerve hook all the way between the segments, the pins and distractor are positioned onto the previously determined sites without changing any angles. Easy linear distraction is then tested by performing 2 to 3 mm of linear mandibular distraction. If there are no impediments to distraction, the bone segments are placed 1 to 2 mm from each other in their original positions. Copious irrigation is done, and the wounds are closed in layers. After completion of one side, the head is turned and the contralateral osteotomies are done in an exact, sequential manner.

DISTRACTION PROTOCOL

Specific details of the distraction protocol have been previously described.7 In summary, the distraction process is begun 3 to 5 days after surgery. Each day, 1.0 to 1.5 mm of distraction is performed. Care is taken to log every distraction session and to ensure the direction of distraction turning is correct. Turning in the incorrect direction will lead to fraction compaction rather than distraction. Usually, the surgeon performs the initial few distraction sessions to ensure appropriate functioning of the device. The parents are then encouraged to continue the process. Most children are discharged 5 days after distraction placement. Fifteen millimeters of distraction is performed (usually during 10 days), which is adequate in most patients. The need for any further distraction is determined based on flexible endoscopy findings. The device allows a maximum of 3 cm of distraction. After discontinuing the distraction process, consolidation of the regenerate bone is allowed to occur for 4 to 8 weeks. Distractor device and pin removal can be done under general anesthesia or sedation without much difficulty.

EVALUATION FOR DECANNULATION

One month after device removal, the airway is assessed for patency by flexible endoscopy and bronchoscopy in the operating room under sedation with spontaneous ventilation. The airway is also evaluated for other airway anomalies, such as suprastomal granulations or collapse, which often occurs as a result of long-term tracheotomy. These must be addressed with laser excision or with laryngotracheal reconstruction (LTR) before the patient is decannulated.

RESULTS

The mean age at the time of mandibular distraction osteogenesis was 29 months. Four of 5 children were tracheotomy-dependent. Three of the tracheotomy-dependent children were subsequently able to achieve decannulation. One child did not show adequate improvement in upper airway obstruction and remained tracheotomy-dependent. Follow-up ranged from 2 to 40 months.

Two of the tracheotomy-dependent children underwent LTR for suprastomal tracheal collapse at the time of distraction. These 2 children had a tracheotomy for long periods (35 and 21 months). The obstruction conferred by the anterior tracheal wall collapse was severe in both patients. This was deemed a “second” airway lesion. Although the tongue base airway level had improved after distraction, it was believed that the suprastomal obstruction would impede their ability to be decannulated safely. These 2 children were unable to tolerate Passy-Muir valves. Laryngotracheal reconstruction for suprastomal airway expansion was then performed, as a single stage at the time of decannulation, resulting in marked improvement of the suprastomal airway in both patients. One child failed extubation after LTR. He required the tracheotomy to be replaced. Concomitant supraglottic collapse and laryngomalacia may have contributed to the decannulation failure. Table 1 summarizes our results in the 5 children.
One child was being evaluated for tracheotomy for chronic airway obstruction, obstructive sleep apnea, and multiple admissions with stridor and respiratory distress. He had been born with birth asphyxia and spastic quadriplegia. In addition to chronic airway obstruction, the child had microcephaly, seizure disorder, and gastroesophageal reflux disease. In the 2 months before distraction, the child had been admitted 3 times with respiratory distress. While orotracheally intubated, the child underwent extubation. After 10 mm of linear distraction had been accomplished, bedside flexible laryngoscopy was performed to determine the degree of tongue base airway patency improvement. He was extubated successfully at that time and was able to avoid tracheotomy.

The child who was unable to achieve decannulation after mandibular advancement was the second birth of quadruplets and was born prematurely at 24 weeks of gestation. He had bronchopulmonary dysplasia, static encephalopathy, developmental delay, gastroesophageal reflux disease, and laryngotraheomalacia. The child was tracheotomized 3 months after birth. Osteotomies and distractions were performed at 38 months of age. Laryngotraheal reconstruction was performed 3 months after the distraction procedure. The child did not show adequate improvement in his airway obstruction and failed extubation after LTR. He required tracheotomy, remaining dependent on the cannula.

The endoscopic assessment of tongue base airway patency revealed improvements in all children. The degree of improvement was mild in the child (patient 2) in whom decannulation was not successful. (Table 2).

Finally, there were no major complications related to the distraction procedure in the series. No blood transfusions were required. No instances of bony or soft tissue infection occurred. Pin sites remained mostly clean and healthy during the distraction sequences. No early removal of pins was required. Scarring was minimal and acceptable in all patients. No facial paralysis occurred.

Mandibular advancement appears to be an excellent technique to advance the tongue base and improve the airway of adult and pediatric populations in cases of moderate to severe oropharyngeal airway collapse secondary to glossoptosis and retrognathia (eg, Pierre Robin sequence).7,9,10 The improvement in airway dimensions achieved appears to be verifiable by 3-dimensional computed tomography.11 In adults, mandibular advancement is usually accomplished by sagittal split osteotomy procedures. Distraction osteogenesis was first described in 1969 by Ilizarov and Ledyaev12 in the Soviet Union as a means to lengthen the long bones of patients with limb asymmetries. The concept of distraction osteogenesis as a means to lengthen the mandible was subsequently proposed by multiple authors.13,14 Since then, other series have alluded to distraction osteogenesis elongation of the mandible as a means to treat airway obstruction in children with mandibular hypoplasia secondary to craniofacial deformities.15,16 Recently, distraction osteogenesis was shown to be a successful mechanism for upper airway patency restoration and a proven method to avoid decannulation in infants and children.7 To our knowledge, no series exist that describe mandibular lengthening by distraction as a means to improve the airway in children with compromise secondary to neurologically induced muscle hypotonicity, as commonly is the case in CP and spastic encephalopathy. Only isolated case reports of success in children with CP have been published.3,5 It is theorized that, by enlarging the upper airway by mandibular advancement, children with CP may reach a certain threshold at which pharyngeal hypotonia, floppy tongue muscles, and uncoordinated laryngeal movements do not result in significant respiratory compromise. The mechanism could be considered as similar to performing a jaw thrust maneuver in a person with alcohol or narcotic overdose–induced upper airway relaxation-respiratory compromise.

Tracheotomy is often required in children with CP and other neurologic disorders. It remains the only proven means of alleviating the tongue base level airway obstruction. Without tracheotomy, these patients are likely to have progressive upper airway obstruction, obstructive sleep apnea with pulmonary hypertension, cor pul-
monale, and probable death. Because of the difficulties that have existed in treating the neurogenically induced tongue base level airway obstruction, once tracheotomized, children with CP have significantly greater difficulty in achieving decannulation. Furthermore, in this population, tracheotomy can impair already rudimentary communication skills, increase the need for specialized and institutionalized care, and decrease quality of life. Long-term, tracheotomy-specific complications have decreased with improvements in technique, nursing care, and monitoring. Pediatric tracheotomy—specific mortality rates have decreased to 0% to 3.4%.\(^{1,2,16-20}\) Despite these improvements, the risks associated with tracheotomy, such as subglottic stenosis, airway obstruction, granulation tissue, bleeding, and death, must be considered and discussed with the families.\(^ {21}\) This pilot series introduces the use of mandibular distraction osteogenesis as a potential treatment modality that may provide an option to children with CP requiring a tracheotomy. It may also be used concomitantly with tracheotomy as a means to decrease the length of time that tracheotomy is required in this population. As mandibular distraction becomes more commonplace, its role in the airway management of these complex patients should become clearer.

In our series, 3 of 4 children with CP with tracheotomy were able to become decannulated. One child with progressive airway difficulty was able to avoid tracheotomy. Subjective airway rating percentage scores also improved in all children. The child who failed decannulation after mandibular distraction also had significant laryngomalacia with supraglottic airway obstruction. Although distraction helped improve his tongue base airway patency, the child was unable to compensate sufficiently, remaining tracheotomy-dependent. In this patient, congruent supraglottic airway obstruction represented another level of airway obstruction. It is possible that with time the supraglottic larynx will grow and become more rigid and that decannulation may be achieved in this child. Supraglottoplasty remains a treatment option.

It could be argued that objective evidence of upper airway improvement after distraction osteogenesis, such as predistraction and postdistraction respiratory distress index scores, is lacking in our cohort. Because it is not useful to obtain preoperative polysomnograms in children with a tracheotomy in place, these data could not be obtained. Plugging the tracheotomy to obtain polysomnogram results for research study purposes is unsafe. Even without predistraction quantitative apnea data, it was apparent that all children were having significant respiratory compromise, episodes of desaturations, and bradycardia. Furthermore, normative polysomnographic respiratory disturbance index data that can quantify severity of obstruction in children do not exist. Some authors would argue that adult polysomnogram criteria are not applicable to children.\(^ {22}\) Children with CP have much less tolerance to apneic events and are at greater risk of obstruction-related cardiopulmonary complications than otherwise healthy adults.

To date, this is the largest series of children with neurogenically induced muscle hypotonia in whom distraction osteogenesis was a successful means of relieving significant airway obstruction. Although our series offers encouraging results, follow-up is limited. Longer tracheotomy-free periods will be needed to prove the lasting effectiveness of distraction osteogenesis in these children.

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REFERENCES


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