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Neonatal mandibular distraction osteogenesis

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Mandibular distraction osteogenesis is a surgical technique used in the management of tongue base obstruction in patients with micrognathia. Judicious and early application of this procedure has been shown to negate the need for tracheostomy and gastrostomy tube placement in select neonates with micrognathia and glossoptosis with and without cleft palate (Pierre Robin sequence). This article describes the operative technique using an external distraction system, with an emphasis on the importance of preoperative assessment and patient selection, as well as the expected postoperative course. Current data on short- and long-term outcomes and new technologies within this growing field are also highlighted.

Introduction

McCarthy first described mandibular distraction osteogenesis (MDO) in 1992. Since 1998, this technique has been reliably used in neonates with Pierre Robin sequence (PRS) to lengthen the mandible and alleviate tongue base obstruction.[1,2] An osteotomy is made, and after a latency period of several days, a distraction appliance is used to incrementally separate the bone segments at a slow and steady rate. The goal is to elongate the mandible to a desired length. Once the goal length is achieved, the hardware is left in place to stabilize the callus of regenerated bone as it is left to mature during the consolidation phase. In certain patients, early MDO can obviate the need for a tracheotomy and allow the infant to feed orally much sooner than in the past.[3] This article describes the use of MDO as an early method of managing airway obstruction in infants with PRS. Preoperative planning, surgical technique, postoperative care, outcomes, and new technologies within this growing field are reviewed.

Preoperative evaluation

The timeline for evaluation and management of a neonate with PRS is shown in Figure 1. In the initial evaluation of a PRS patient, the most pressing issue is the airway. If an infant exhibits signs or symptoms of acute airway obstruction, then the airway must be emergently secured via either endotracheal intubation or a tracheotomy. In emergent cases of airway obstruction, the laryngeal mask airway (LMA) may be used as an effective bridge to more definitive management.[4] For those neonates who are not in

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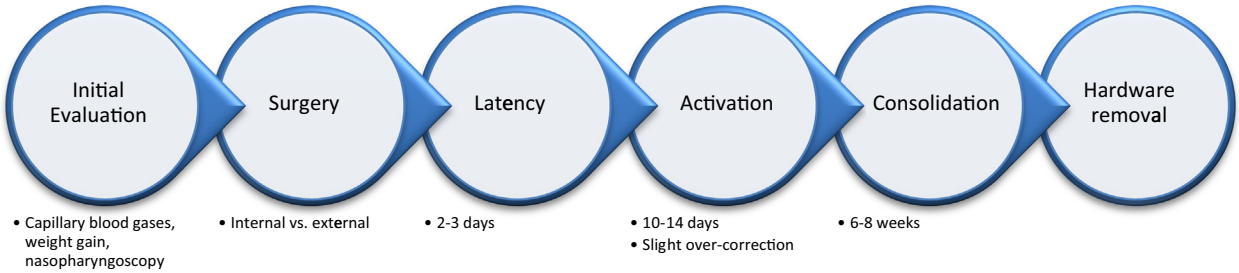

Figure 1 Timeline of neonatal mandibular distraction osteogenesis used by the senior author. (Color version of figure is available online.)

acute airway distress, evaluation should begin with a comprehensive history and physical examination to assess for any other abnormalities. Additional dysmorphic features, neurologic impairment, and cardiopulmonary dysfunction should be noted, as PRS can be associated with several conditions, including stickler and velocardiofacial syndrome. A flexible nasopharyngoscopy should be performed to evaluate the degree of tongue base obstruction and assess for any synchronous airway lesions such as choanal atresia, laryngomalacia, or glottic or subglottic stenosis, as this may influence the decision to perform a tracheotomy to secure the airway before any further surgical interventions.[5]

Most commonly, patients have mild to moderate intermittent airway obstruction. For this subset of patients, nonsurgical airway interventions should always be tried first. These may include repositioning in the prone or lateral positions, the use of a nasopharyngeal airway, or a customized oral appliance.[6] During this period, capillary blood gases may be obtained on a regular basis (daily or every other day) to objectively monitor the degree of chronic airway obstruction. Normal neonatal values for pCO_2 range from 35-48 mm Hg, and 22-27 mEq/L for HCO_3. Persistently elevated pCO_2 values in the age group of 50-59 years in conjunction with an increase in HCO_3 level are reliable markers for chronic respiratory acidosis with compensatory metabolic alkalosis, which may signal the necessity for surgical intervention. The senior author feels that polysomnography in the neonatal population is not a reliable means of assessing chronic airway obstruction, as normative data for neonates are lacking and these studies tend to overestimate the degree of sleep apnea.

The other major issue facing neonates with PRS is difficulty with feeding, which is closely intertwined with the problem of persistent airway obstruction. Traditional feeding devices for infants with cleft palate can be used with the assistance of trained feeding specialists.[7] However, if the child fails to gain weight appropriately in spite of these strategies or dietary supplementation, then surgical intervention can be considered. Similarly, if an infant has increased work of breathing or prolonged feeding times, then more support may be lent toward a surgical solution.[8]

Patient and hardware selection

Once the decision is made to proceed with a surgical airway intervention, one must assess whether the patient is a good candidate for MDO. Surgery is usually deferred until the patient reaches 2.5 kg, given the low blood volume of neonates as well as the need for appropriately sized endotracheal tubes (3.0-3.5 ETT) and distraction hardware. The senior author's contraindications to neonatal MDO include the absence of mandibular condyles, poorly defined glenoid fossae, and neurologic compromise such as seizures, hypotonia, chronic aspiration, or poor coordination. Children with Pruzansky III deformities are at risk for inadequate engagement of the condyle and the skull base, allowing posterior displacement of the ramus into the mastoid region and ineffective advancement of the anterior segment.[9] Children with neurologic impairment have been shown to have worse airway and feeding outcomes following MDO due to their propensity for airway compromise and dysphagia related to factors independent of tongue base obstruction.[10]

Both external and internal (or "buried") distraction devices are available. There are advantages and disadvantages to each approach, and these must be considered in conjunction with the patient's anatomy as well as the parents' wishes. External hardware allows for multivector distraction and is easy to troubleshoot and adjust during the activation process. In most cases, it does not require preoperative imaging and is easily removed after the consolidation phase. However, there is increased risk of injury to tooth buds from placement of bicortical K-wires, which secure the distractors to the bone (Figure 2). Addi-

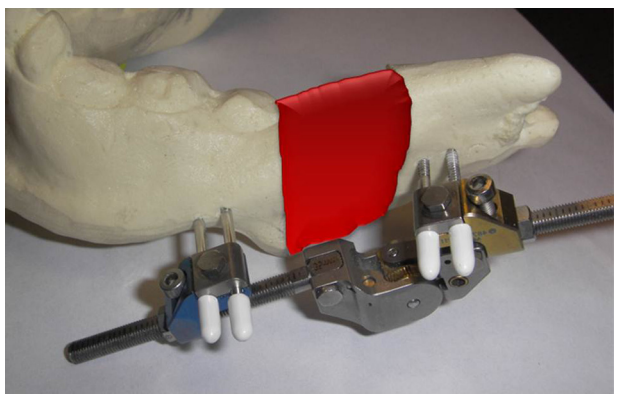

Figure 2 The concept behind mandibular distraction osteogenesis. Medical model demonstration of how lengthening regenerate bone (red) results in improved mandibular projection over time. A multivector external distractor is being used in this case. (Color version of figure is available online.)