Mandibular Distraction for Micrognathia and Severe Upper Airway Obstruction

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Objective: To determine whether the use of mandibular distraction osteogenesis (DOG) can help to avoid tracheotomy or achieve decannulation in patients with mandibular hypoplasia and severe upper airway obstruction.

Design: Retrospective medical record review (spanning a 27-month period).

Setting: Tertiary care children’s hospital.

Subjects: Group A (n=8) was composed of infants with Pierre Robin sequence and no tracheotomy (mean age, 2.5 months); group B (n=6), older nontracheotomized micrognathic children with obstructive sleep apnea (OSA) (mean age, 69 months); and group C (n=12), tracheotomized children with complex congenital syndromes (mean age, 33 months).

Intervention: Bilateral mandibular DOG with endoscopic (n=24) and/or radiographic (n=17) airway evaluation (mean follow-up, 16 months [range, 2-42 months]).

Outcome Measures: Group A, tracheotomy avoidance; group B, resolution of OSA (clinically or on polysomnography); and group C, decannulation.

Results: Group A, 7 patients (88%) successfully avoided tracheotomy; group B, 5 patients (83%) had resolution of OSA; and group C, 2 patients (17%) underwent decannulation.

Conclusions: Mandibular DOG (1) allows tracheotomy avoidance in infants with isolated Pierre Robin sequence and (2) relieves OSA in older micrognathic children without tracheotomy. However, mandibular DOG does not frequently lead to decannulation in tracheotomized patients with complex congenital syndromes.

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Congenital micrognathia and glossoptosis are most commonly seen in patients with Pierre Robin sequence (PRS), but may also be associated with disorders such as Treacher Collins syndrome, Nager syndrome, and hemifacial microsomia. Although the majority of children born with micrognathia are either asymptomatic or can be treated conservatively (eg, prone positioning and nasopharyngeal airways), up to 23% of children with PRS may have major respiratory obstruction, necessitating more aggressive intervention. Some, but not all, patients may outgrow their micrognathia.

Tracheotomy is traditionally the safest and most effective treatment option in patients with PRS and severe upper airway obstruction and is performed in as many as 12% of cases. Tracheotomy, however, is associated with high cost, frequent morbidity, and occasional mortality, with an average age of 3.1 years at decannulation. Therefore, the development of other effective methods of airway management is desirable. A new advance in the treatment of children with micrognathia and severe upper airway obstruction is mandibular distraction osteogenesis (DOG), in which an attempt is made to advance the tongue base anteriorly via its muscular attachments to the mandible, relieving pharyngeal airway obstruction. The purpose of the present study was to review a tertiary care pediatric hospital’s experience with mandibular DOG in 26 consecutive children with micrognathia and upper airway obstruction over a 27-month period.

METHODS

Twenty-six consecutive patients with micrognathia who underwent mandibular DOG in an attempt to relieve severe upper airway obstruction between March 2000 and May 2002 were identified using a patient database maintained by the Pediatric Plastic and Reconstructive Surgery Division at Children’s Hospital of Pittsburgh, Pittsburgh, Pa. This study was determined to be exempt from hospital Human Rights Committee review.
Group A consists of infants (aged <1 year) with micromaxillary and respiratory obstruction severe enough to require admission to the neonatal intensive care unit. Patients were evaluated by a multidisciplinary team involving the following specialties: neonatology, otolaryngology, plastic surgery, anesthesiology, genetics, pulmonology, and gastroenterology. The diagnosis of airway obstruction was based on a variety of factors, including weight gain, oral intake, and oxygen saturation, as well as results from sleep studies, laryngoscopy (flexible and direct), and bronchoscopy (rigid or flexible), when performed. Patients whose airway obstruction could not be controlled with conservative measures (ie, positioning and nasal airway), and for whom tracheotomy was recommended based on a consensus decision by the multidisciplinary team, underwent mandibular distraction in an attempt to avoid tracheotomy. Patients were excluded from consideration for mandibular distraction if any of the following conditions were identified: a second airway lesion, central sleep apnea, and severe untreated gastroesophageal reflux disease (GERD).

Group B consisted of micrognathic children without tracheotomy and without adenotonsillar hypertrophy, but with documented obstructive sleep apnea (OSA) on polysomnography (1-48 episodes of OSA lasting 13-30 seconds, with oxygen desaturations of 61%-91% on room air). In 2 patients with just 1 obstructive event during polysomnography, the relative lack of obstruction during sleep was deceptive, as these patients had frequent hypopneas (up to 24 events) with poor sleep efficiency, and even demonstrated stertor while awake, which could be relieved with a jaw thrust. After preoperative endoscopic and/or radiographic airway evaluations confirmed tongue base collapse as the primary site of airway obstruction in all group B patients, these patients underwent mandibular distraction to relieve the signs and symptoms of OSA.

Group C consisted of children with micrognathia who had already undergone tracheotomy and in whom decannulation had failed or who were thought to be unsuitable for decannulation. After preoperative endoscopic and/or radiographic airway evaluation confirmed tongue base collapse as the primary site of airway obstruction, these patients underwent mandibular distraction with the intention of relieving tongue base obstruction and allowing decannulation.

Two of us (J.P.B. and C.B.G.) performed all mandibular distraction procedures at Children's Hospital of Pittsburgh. All patients underwent bilateral distraction. Distraction devices were applied externally in 20 patients and internally in 6 patients. Although internal distractors are technically more challenging to apply (owing to the smaller working space), this method was preferred, when feasible, because of the following advantages: less chance of device dislodgment during consolidation and no visible external hardware, thereby allowing earlier return to school for older children, reduced parenteral and nursing anxiety, and less visible facial scarring.

**INTERNAL MANDIBULAR DOG**

For internal mandibular distraction procedures in neonates and infants, a combined intraoral and extraoral approach (2-cm Risdan incision) is used. Either a vertical or an inverted ‘L’ mandibular ostectomy is performed with a malleable retractor positioned lingually for protection of underlying structures. A microdistraction (KLS, Jacksonville, Fla) is fixed with 2 or 3 screws (1.0 mm) on either side of the ostectomy. The microdistraction rod is placed at an oblique vector to gain both ramal height and body length. The distractor turning arm is brought out intraorally with a 12F catheter for soft tissue protection. For older children, the placement of internal mandibular distraction is performed without an external incision. The osteotomy is performed through an intraoral dissection from the angle of the mandible to above the entrance of the inferior alveolar nerve just proximal to the lingula. A pediatric mandibular ramus distractor (Zurich; KLS) is percutaneously attached with 1.5-mm screws on either side of the ostectomy with an oblique vector.

**EXTERNAL MANDIBULAR DOG**

An intraoral approach is again used for placement of an external device, and a similar osteotomy is made from the angle of the mandible to just proximal to the inferior alveolar nerve entrance. In each proximal mandibular segment, 2 K-wires (0.062 mm) are percutaneously placed in parallel fashion. The distal K-wire is driven through both outer and inner mandibular body cortices below the inferior alveolar canal so that the external device is stabilized to the contralateral device. (In some cases, this transmandibular wire was also used for the proximal segment.) A multiplanar external distractor device (Synthes, Paoli, Pa) is positioned and used for unidirectional lengthening, angulation, and varus/valgus manipulation. For both external and internal distraction devices, a latency period of 1 to 2 days was allowed, following which distraction began at a rate of 2 turns (0.6 mm) 3 times a day (1.8 mm/d) in infants and 1 turn (0.5 mm) 2 times a day (1.0 mm/d) in children older than 1 year. In newborns and infants, active distraction was completed once airway obstruction was relieved (confirmed with bedside flexible laryngoscopy or conversion of tongue orientation from a vertical to a horizontal position). In older children, distraction continued until a slight class III occlusal relationship was established.

Postdistraction cephalograms were obtained to confirm bone consolidation (6 weeks in infants; 10-12 weeks in older children), after which the distraction hardware was removed. Follow-up studies included a cephalogram and a 3-dimensional computed tomographic scan immediately after hardware removal, with further cephalograms obtained at 6 months and 1 year.

### RESULTS

**GROUP A**

Eight infants (7 boys and 1 girl) with micrognathia, glossoptosis, and severe upper airway obstruction underwent mandibular distraction in an attempt to avoid tracheotomy. The mean age at DOG was 2.5 months (age range, 8 days to 8 months). Seven patients had PRS (6 isolated cases and 1 with associated degenerative encephalopathy), and 1 patient had Beal syndrome. The diagnosis of airway obstruction was based on oxygen desaturations (n=4), dinkiness and apnea on feeding (n=4), inability to tolerate extubation (n=1), and results from sleep studies (n=6). Tongue base collapse was confirmed as the primary site of obstruction by flexible nasolaryngoscopy and direct laryngoscopy (n=7) and computed tomographic scans with 3-dimensional reconstruction (n=3), while rigid (n=6) and flexible (n=1) bronchoscopy ruled out other airway lesions.
The mean length of distraction was 23 mm (range, 15-35 mm). After distraction, the patients remained intubated an average of 5.6 days (range, 1-8 days), and active distraction was performed for an average of 12 days (range, 8-18 days).

Seven patients (88%) were relieved of their upper airway obstruction and were discharged home an average of 7 days after DOG (range, 2-62 days). At the most recent follow-up visit (average length of follow-up, 13 months; range, 7-16 months), none was requiring regular home monitoring. Prolonged postoperative gavage feedings were required in 4 patients during hospitalization, but at the most recent follow-up visit, 6 of the 7 patients were tolerating a regular oral diet, while 1 patient was gastrostomy tube dependent.

One patient with PRS and degenerative encephalopathy underwent extubation 6 days after DOG and then underwent reintubation 2 days later owing to central apnea. Although a preoperative sleep study had shown 103 oxygen desaturations that were lower than 90%, all but 4 of which were thought to be due to obstructive and hypopneic events, the central apnea component of the patient’s condition either had been underappreciated or had rapidly worsened in the weeks after the study was performed. Tracheostomy and gastrostomy were strongly being considered before the family elected to withdraw life support 2 months after DOG.

Cleft palate repair (n=3) and adenoidectomy (n=1) were performed at later dates. Four patients underwent postoperative endoscopic airway evaluations, which confirmed anterior positioning of the base of tongue with a patent airway.

**GROUP B**

Six older nontracheotomized children (4 boys and 2 girls) with micrognathia, glossoptosis, and upper airway obstruction without adenotonsillar hypertrophy were seen as outpatients and diagnosed as having OSA by polysomnography. Mandibular DOG was performed in an attempt to relieve OSA. The mean age at DOG was 69 months (range, 13-161 months). Three patients had isolated PRS, and 1 patient each had Goldenhar syndrome, Cornelia de Lange syndrome, and trisomy 2 with craniosynostosis. Tongue base collapse was confirmed as the primary site of airway obstruction with flexible and direct laryngoscopy and rigid bronchoscopy (n=4). Preoperative computed tomographic scans with 3-dimensional reconstruction (n=8) and cephalograms (n=3) were obtained. The mean length of distraction was 31 mm (range, 25-38 mm).

Five patients (83%) no longer had any evidence of upper airway obstruction after surgery; 1 patient still required home monitoring for central apneas. Three of these 6 patients underwent postoperative sleep studies that documented resolution of OSA. The average length of follow-up was 17 months (range, 10-22 months).

**GROUP C**

Twelve tracheotomized patients (5 boys and 7 girls) with micrognathia, glossoptosis, and upper airway obstruction underwent mandibular DOG in an attempt to achieve decannulation. The mean age at DOG was 33 months (age range, 38 days to 93 months). Four patients had PRS (1 of whom also had arthrogryposis and a 14:X balanced translocation), 3 had Nager syndrome, 2 had Goldenhar syndrome, and 1 each had Treacher Collins, Lowe, and Marden-Walker syndromes. Previously, 2 had unsuccessfully undergone mandibular bone grafting. Tongue base collapse was confirmed as the primary site of airway obstruction by flexible laryngoscopy (n=12). Rigid bronchoscopy confirmed the absence of other airway lesions (n=12). Preoperative computed tomographic scans with 3-dimensional reconstruction (n=8) and cephalograms (n=3) were obtained. The mean length of distraction was 29.5 mm (range, 15-41 mm).

At the most recent follow-up visit (average length of follow-up, 21 months; range, 4-42 months), only 2 (17%) of the 12 patients (17%) had undergone decannulation (at 3 and 7 months after surgery). Neither was requiring home monitoring or gavage feedings, and both had patent pharyngeal airways confirmed by postoperative endoscopy. Of the 10 patients who remained with tracheostomy, 2 had preexisting ankylosis of the temporomandibular joint (TMJ), 1 developed TMJ ankylosis after DOG, 7 had chronic swallowing dysfunction and gastrostomy tube dependence, and 4 had previously undergone Nissen fundoplication for severe GERD. Decannulation was formally attempted in just 2 of the 10 patients, both of whom required another tracheotomy for persistent obstruction. Postoperative endoscopy revealed relief of tongue base obstruction but persistent epiglottic collapse against the posterior pharyngeal wall in 6 of the 10 patients.

In the present study, 7 reversible complications occurred in 6 patients (23%): premature callus consolidation requiring another DOG procedure (n=3), cheek abscess requiring incision and drainage (n=2), minor lip erosion from pin contact (n=1), and facial cellulitis (n=1). Major complications occurred in 3 patients (12%): unilateral total facial paralysis as a result of facial nerve injury (n=1), and 2 cases of TMJ ankylosis, one of which contributed to decannulation failure, and one of which led to tracheotomy in a patient with OSA and postdistraction mandibular osteomyelitis.

**COMMENT**

Because the tongue is held forward by its anterior muscular attachments to the mandible and hyoid bone, a lack of mandibular projection in patients with micrognathia may allow the tongue base to compress the epiglottis, leading to supraglottic obstruction. Traditional techniques for increasing the anterior projection of the mandible in adults, such as sagittal split osteotomies with bone grafts and rigid fixation, are not thought to be stable and reliable in small children. Other surgical interventions, such as tongue-lip adhesion, have fallen into disfavor owing to ineffectiveness and frequent complications.

Ilizarov and Ledyaev first popularized DOG for correction of leg length discrepancies in adults in 1969. Since McCarthy et al used DOG for the reconstruction of the deficient mandible in children in 1992, the technique has
become increasingly accepted as a management option for micrognathia, as evidenced by recent series reported by Denny et al.\textsuperscript{2,6} and Sidman et al.\textsuperscript{3} The primary objective of mandibular DOG is to advance the tongue base anteriorly via its muscular attachments to the distracted mandible, thus pulling the tongue out of the hypopharynx and relieving upper airway obstruction.\textsuperscript{8}

Reported complications from the use of mandibular DOG in children have included penetration of the floor of the mouth with a pin or loosening of a pin after a fall,\textsuperscript{2,9} development of an abscess at the pin site,\textsuperscript{10} inadequate distraction requiring a second DOG procedure,\textsuperscript{3} and facial scarring requiring revision.\textsuperscript{9} In patients old enough to be tested, inferior alveolar nerve function has remained intact.\textsuperscript{9,19}

The results from the present study indicate that 7 (88\%) of 8 patients younger than 1 year with micrognathia and severe upper airway obstruction (group A) can avoid tracheotomy and be discharged home after undergoing mandibular DOG. Other series have attained similar results. In 2002, Ortiz-Monasterio et al.\textsuperscript{10} (n = 4) and Denny and Kalantarian\textsuperscript{2} (n = 5) reported on neonates (aged <1 month) with PRS and severe upper airway obstruction, all of whom were able to avoid tracheotomy after mandibular DOG.

In the present series, it is unknown what the outcome would have been if patients identified as needing a tracheotomy by the multidisciplinary team had received neither tracheotomy nor mandibular DOG. Determination of the need for tracheotomy was based on clinical judgment and the assessment of a variety of factors, including intubation, oxygenation, and feeding status; polysomnography; response to positioning and nasopharyngeal airway; failure to thrive; GERD assessment; endoscopy; and imaging. However, because a strict protocol was not in place to precisely determine the need for tracheotomy, there is a potential for bias, both in our study and in other reports. This inherent weakness in study design may be overcome by establishing rigid indications for tracheotomy and mandibular DOG in the future.

The natural history of mandibular growth in patients with isolated PRS is typically one of continued development, although results may be neither consistent nor sufficient.\textsuperscript{4} The micrognathia associated with more complex conditions, such as Treacher Collins and Nager syndromes, is more likely to persist over time.\textsuperscript{3} Little is known about the long-term outcomes of untreated pediatric glossoptosis with respect to failure to thrive and life-threatening airway obstruction, since withholding definitive therapy is unethical unless the parents choose to withdraw support.\textsuperscript{3}

In the present study, 5 (83\%) of 6 older children with micrognathia and documented OSA, with no evidence of adenotonsillar hypertrophy and no concomitant airway lesions (group B), experienced resolution of OSA after mandibular DOG. Although a report by Denny et al.\textsuperscript{2} has demonstrated similar results, overall there is a lack of literature describing this subset of patients. It seems reasonable to address micrognathia and glossoptosis in older children as a potentially reversible etiologic factor in OSA.

In the present study, mandibular DOG procedures led to decannulation in only 2 (17\%) of 12 tracheotomized, micrognathic patients with complex congenital syndromes and severe upper airway obstruction (group C). Higher decannulation rates of 86\% (6 of 7 patients) and 88\% (7 of 8 patients) have been reported in the literature for such patients.\textsuperscript{3,11} Our relative lack of success with this patient population is most likely multifactorial in nature. Among the 10 patients who remained tracheostomy dependent, 7 had chronic swallowing dysfunction necessitating gastrostomy feedings, 4 had a history of severe GERD requiring surgery, and 3 had TMJ ankylosis. These conditions were either not present or not specifically addressed in other studies.\textsuperscript{3,11}

Persistence of tracheostomy does not necessarily reflect failure of mandibular DOG to enhance the airway, but rather may reflect caregiver bias. The fact that only 2 of 10 patients who still had a tracheostomy underwent formal decannulation attempts may reflect hesitancy to attempt decannulation in a child with a long history of airway obstruction, psychomotor delay, chronic speech and swallowing difficulties, hypotonia, and possible need for other nonairway surgical procedures requiring general anesthesia and unimpeded airway access. On the other hand, it is difficult to recommend a decannulation trial if post-DOG endoscopy fails to document an adequate airway.

Anatomically, persistent epiglottic collapse against the posterior hypopharyngeal wall was noted in 6 of our 10 patients who still had a tracheostomy after undergoing mandibular DOG. In one study reporting a decannulation rate of 88\% after mandibular DOG, many other adjunct procedures, such as tongue reduction, uvulopalatopharyngoplasty, and tongue-kyoid and epiglottal suspension, were performed.\textsuperscript{17} To achieve the best results in tracheotomized, micrognathic patients with complex airways, adjunct procedures need to be performed with mandibular DOG, more aggressive post-DOG decannulation protocols need to be followed, or entrance criteria for mandibular DOG may need to be limited to patients without (1) a history of severe GERD, (2) a history of chronic swallowing dysfunction and gastrostomy tube dependence, (3) hypotonia, and (4) preexisting TMJ ankylosis.

The natural decannulation rate for patients who have undergone tracheotomy for micrognathia and concomitant airway abnormalities and/or neurologic deficit is not easily determined, as there are few historical controls in the literature. In one study of 11 such complex airway patients, decannulation was achieved in all at an average age of 3.1 years (range, 14 months to 5 years), although it is not specifically mentioned whether adjunct surgical procedures were required to achieve this result.\textsuperscript{4}

In conclusion, mandibular DOG was highly successful for relief of upper airway obstruction in neonates with PRS and in older children with micrognathia and OSA. In contrast, DOG was much less successful in allowing decannulation of tracheotomized patients with complex syndromes, owing to a variety of potential factors, including underlying neurologic dysfunction, persistent supraglottic obstruction, and TMJ ankylosis.

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