Relief of Upper Airway Obstruction With Mandibular Distraction Surgery
Long-term Quantitative Results in Young Children

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Objective: To evaluate the long-term benefits of mandibular distraction on sleep-related upper airway obstruction in young children with mandibular hypoplasia.

Design: Cross-sectional study. Subjects were examined for sleep-disordered breathing using medical history, physical examination results, and a written questionnaire. Subjects underwent standard overnight polysomnography, during which measures of sleep-disordered breathing were collected.

Setting: Tertiary care hospital.

Patients: Five children with upper airway obstruction from craniofacial anomalies treated with mandibular distraction, with a minimum follow-up of 12 months.

Main Outcome Measures: Apnea-hypopnea index, oxygen saturation nadir, and peak end-tidal carbon dioxide value.

Results: Of the 5 children, 3 were cured of upper airway obstruction as documented by polysomnography, with an apnea-hypopnea index of less than 1.5 and no snoring. The fourth child had primary snoring without apnea. The fifth child had severe obstructive sleep apnea, with an apnea-hypopnea index of 20.2.

Conclusions: Most children who undergo mandibular distraction for upper airway obstruction associated with mandibular hypoplasia demonstrate significant clinical improvement of obstructive sleep apnea. However, those children who continue to have symptoms of sleep-disordered breathing after surgery should undergo polysomnography for evaluation of persistent obstructive sleep apnea.

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MANDIBULAR DISTRACTION has been used in children with upper airway obstruction from mandibular hypoplasia since the first reported series was published in 1992.1 Most children treated with this technique have either unilateral or bilateral mandibular hypoplasia due to hemifacial microsomia, other syndromic disorders, or the Pierre Robin syndrome. Mandibular distraction may relieve sleep-related upper airway obstruction and allow some children to avoid tracheotomy. It can also be used to facilitate earlier decannulation in those children who have already undergone tracheotomy. These same children with craniofacial abnormalities are at high risk for persistent obstructive sleep apnea (OSA). Symptoms of OSA include snoring, gasping, respiratory pauses, and restless sleep. A clinical examination is sensitive but not specific for OSA.2 Polysomnography is the gold standard for the evaluation and diagnosis of OSA.

Published series3-12 have demonstrated the short-term efficacy of mandibular distraction in the treatment of OSA in patients with craniofacial disorders. However, the postoperative sleep study data in these series were typically obtained shortly after surgery. Other studies4 show improvement of airway obstruction with this surgery without complete cure of OSA. There is a paucity of literature regarding the long-term effectiveness of mandibular distraction in the prevention and treatment of OSA in children with craniofacial anomalies. Our objective was to examine the patients with mandibular hypoplasia who have had mandibular distraction at our institution to assess the long-term benefit in relieving upper airway obstruction and OSA.

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After obtaining approval from The Johns Hopkins School of Medicine institutional review board, eligible patients were identified by the craniofacial surgeon (C.V.) from his clinical patient database. Patients were recruited from the pediatric craniofacial clinic and otolaryngology clinic. Subjects eligible for this study included children aged 3 to 14 years with craniofacial anomalies who had undergone mandibular distraction for airway obstruction, and had undergone surgery a minimum of 12 months before recruitment. Exclusion criteria included non-English-speaking patients and patients who are tracheotomy dependent.

The study subjects were interviewed and examined by the investigators for signs and symptoms of sleep-disordered breathing, by medical history, a physical examination, and a written questionnaire. History of pharyngeal surgery or other treatment for OSA was recorded. A physical examination was performed with particular attention to the upper airway characteristics, including mandibular position and size and tonsillar size. The patient’s parent or caretaker completed a written questionnaire (the pediatric OSA symptom questionnaire) regarding symptoms of OSA. The questionnaire addressed 12 potential symptoms of OSA and the frequency of occurrence (Figure). The questionnaire was numerically scored as follows: 0 indicates never; 1, sometimes; 2, almost always; and 3, always. The possible total score range on the questionnaire was from 0 to 36. While this questionnaire has not been formally validated, it was used to facilitate a standard history and review of signs and symptoms.

Patients then underwent standard overnight polysomnography in a pediatric facility accredited by the American Board of Sleep Medicine. The study was performed by a trained pediatric polysomnographic technician, using standard techniques. A parent remained in the same room as the child throughout testing. During the sleep study, surface electrodes and monitoring devices (acquired digitally by Alice 3 and Alice 4, Atlanta, Ga) measured signals from the electroencephalogram, the right and left electrooculograms, the surface electromyogram, and an electrocardiogram; chest and abdominal wall motion; pulse oximetry; and end-tidal PCO2 (Novametrix, Wallingford, Conn). Airflow was measured by oronasal thermistor in all children. In addition, airflow was monitored using nasal pressure to obtain a quantitative signal (Protech, Mukilteo, Wash). All studies were monitored with real-time videotape for motion analysis and snoring recording. The medical history was obtained and physical examination, questionnaire, and polysomnography were performed in these patients for research purposes only.

Although no standard pediatric criteria exist for the diagnosis of pediatric OSA, pediatric criteria, as suggested by the American Thoracic Society,13 were used for evaluating sleep structure and respiratory patterns observed in the sleep studies. Sleep architecture was recorded according to standard criteria.14 Respiratory analysis consisted of defining apneas as a cessation of airflow for 2 or more breath cycles. Hypopnea was defined as a visual reduction in airflow over 2 or more respiratory cycles, accompanied by a 3% or greater decrease in oxygen saturation or terminated by an arousal.15 Hypopneas were measured preferentially with the nasal pressure monitor, a quantitative measure of respiratory flow that more reliably demonstrates airflow limitation compared with the thermistor.16 Obstructive apneas are indicated by the presence of paradoxical breathing movements and/or snoring in association with decreased airflow signals for 2 or more respiratory cycles. Central apneas are without evidence of respiratory effort, indicated by paradoxical thoracoabdominal movements, and are not recorded in the apnea-hypopnea index (AHI). Mixed apneas are a combination of central and obstructive events. The AHI is defined as the number of obstructive apneas, mixed apneas, and hypopneas occurring per hour of sleep. Although standard pediatric sleep scoring criteria have not been established, to our knowledge, healthy nonsnoring children without craniofacial anomalies are described as having an AHI much less than 1 event per hour.17,18 Patients were considered to have no OSA if the AHI was less than 1.5 and there was no snoring; primary snoring, if the AHI was less than 1.5 and snoring was present; mild OSA, if the AHI was 1.5 to 5.0; and significant OSA, if the AHI was greater than 5.0.

A pulse oximeter (Masimo Corporation, Irvine, Calif) was used, which has a short averaging time and, therefore, detects oxyhemoglobin changes sensitively with less movement artifact. Oxygen saturation measurements obscured by artifact (detected by lack of synchrony between the pulse wave and electrocardiogram) were deleted from analysis. The oxygen saturation nadir is the lowest saturation tabulated. Studies17,18 of normal polysomnographic respiratory values in children indicate that oxygen saturation rarely decreases to less than 93% except during central apneas.

<table>
<thead>
<tr>
<th>Circle One:</th>
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<tr>
<td>Always</td>
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<tr>
<td>1. My child snores while sleeping:</td>
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<tr>
<td>2. My child stops breathing or has pauses in breathing at night:</td>
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<tr>
<td>3. My child gasps while sleeping:</td>
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<td>4. My child chokes while sleeping:</td>
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<td>5. My child has restless sleep:</td>
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<tr>
<td>6. My child wakes frequently when sleeping:</td>
</tr>
<tr>
<td>7. Answer this question only if your child is toilet trained: My child has problems with bed-wetting:</td>
</tr>
<tr>
<td>8. My child sweats while sleeping:</td>
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<tr>
<td>9. My child extends his neck while sleeping or sleeps in strange positions/position:</td>
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<tr>
<td>10. My child is sleepy/tired during the day:</td>
</tr>
<tr>
<td>11. My child breathes through his mouth (is a mouth breather):</td>
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<tr>
<td>12. My child has a runny nose:</td>
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Figure. Pediatric obstructive sleep apnea symptom questionnaire.
RESULTS

Eight patients identified by the craniofacial surgeon (C.V.) from his clinical database were eligible to participate in the study. Examination of the clinical database also revealed that no patients who had undergone mandibular distraction longer than 12 months ago were still tracheotomy dependent. Five patients agreed to participate and completed the study (Table). One patient who declined the study cited inconvenience; the other 2 agreed to the study but did not complete their sleep studies despite several attempts at scheduling. The median age at the time of surgery was 49 months. Four patients were born with Pierre Robin syndrome, and the other was born with severe micrognathia without cleft palate. All patients underwent mandibular distraction at our institution, performed by one of us (C.V.), with the indication of respiratory distress from upper airway obstruction. The median follow-up from surgery was 47.5 months. One patient had undergone tracheotomy, but then underwent decannulation 34 months after mandibular distraction. Two patients clinically continued to have severe retrusion after distraction, and the remaining 3 had mild retrusion. One patient underwent a prior tonsillectomy, and the other patients had +1 or +2 tonsils. Dental occlusion was abnormal in 3 patients, and 4 patients reported abnormal dentition. No patients complained of lip numbness. The median size of overbite was 4 mm (range, 3-12 mm). None of the children were obese or had neurologic problems.

The median score on the pediatric OSA symptom questionnaire was 19 (range, 7-24) of a maximum possible score of 36.

The patients underwent standard overnight polysomnography. One patient had no snoring or OSA (AHI <1.5 and no snoring), and 3 patients had primary snoring (AHI <1.5 with snoring).

One patient had severe OSA, with an AHI of 20.2. The patient had undergone tracheotomy and had the highest score (score, 24) on the OSA symptom questionnaire. The caregiver for this patient answered always for items regarding snoring, gasping, stopping breathing, restless sleep, and mouth breathing on the questionnaire. This child had undergone polysomnography with her tracheotomy tube plugged 2 years before our study and had a respiratory disturbance index of 3.7 with an oxygen desaturation nadir of 88%. After full airway endoscopy and adenotonsillectomy, the patient was monitored in the hospital with her tracheotomy tube plugged and had no signs of sleep-disordered breathing. She subsequently underwent decannulation and remained symptom free for more than 1 year. Approximately 6 to 12 months before our study, the patient began developing symptoms of nocturnal airway obstruction. The results of the polysomnogram, done as part of this study, were communicated to her family and the treating physicians; her condition is successfully maintained using bilevel (biphasic) positive airway pressure.

The median oxygen saturation for the sleep studies was 87%. The median peak end-tidal carbon dioxide level was 34 mm Hg.

COMMENT

Children with craniofacial anomalies and mandibular hypoplasia are at high risk for upper airway obstruction in the neonatal period. Symptoms range in severity from increased work of breathing and OSA to continuous upper airway obstruction. Before mandibular distraction, children with craniofacial anomalies and severe airway obstruction required endotracheal intubation, long-term nasal airways, craniofacial surgery, and often tracheotomy.

In 1992, McCarthy et al described the first series of patients to undergo mandibular distraction, which can help avoid tracheotomy in some children with mandibular hypoplasia. Mandibular distraction involves the surgical creation of osteotomies in the mandible. The mandible is then serially distracted in small amounts with a rigid fixator device. Bone forms between the separated bone surfaces, with distraction rates of approximately 1 mm/d. The lengthening of the mandible corrects the hypoplasia of the mandible, with improved supraglottic and pharyngeal airway support. The original article by McCarthy et al described the use of mandibular distraction to improve aesthetic appearance in children with craniofacial anomalies. Several subsequent articles have documented the use of mandibular distraction to improve the airway in children with mandibular hypoplasia.

Carls and Sailer described a series of 7 children with severely hypoplastic mandibles and associated respira-
tory distress who underwent distraction. All 7 had relief of their airway symptoms after surgery. The distraction allowed for the decannulation of the 2 patients in this series who underwent prior tracheotomy. Sidman et al described a series of 11 micrognathic children with tracheotomies who underwent successful decannulation after mandibular distraction. In their report, patients were considered to have a successful outcome if they were able to undergo decannulation, but polysomnography was not routinely done postoperatively.

The successful use of mandibular distraction to treat OSA in children with craniofacial anomalies has been documented in the literature.5-12 Morovic and Monasterio described 7 children younger than 18 months who were treated with mandibular distraction. Postdistraction polysomnograms showed an improvement in the apnea indexes for all children, but these children continued to have obstructive events, ranging from 8 to 18 events per hour, indicating persistent OSA after surgery. Bell and Turvey described 8 patients with severe micrognathia who were treated with distraction. Four were successfully treated, as 2 of 3 patients with tracheotomies underwent decannulation and another 2 children were weaned from supplemental oxygen. However, no sleep study data were reported for this study. Monasterio et al described 15 patients with severe OSA with the Pierre Robin syndrome who had improvement of their apnea as documented by polysomnography that was performed a mean of 3.2 months after surgery. Cohen et al described a series of 20 patients, 12 who were studied with preoperative and postoperative polysomnograms. They reported an improvement in the mean AHI from 25.24 to 1.72; their postoperative sleep studies were performed before discharge from the hospital. None of these studies report long-term results of mandibular distraction as documented by polysomnography.

To our knowledge, the present study is the first report of long-term polysomnographic data after mandibular distraction. At our institution, neonates presenting with mandibular hypoplasia and respiratory distress without apparent neuromotor disease or complex craniofacial anomalies are candidates for mandibular distraction. All patients who have undergone distraction at our institution have either avoided tracheotomy or, if they did require tracheotomy, subsequently underwent decannulation. We have not done routine postoperative polysomnography, but have used clinical relief of airway obstruction, extubation, and close intensive care monitoring with normal oxygen saturations as primary outcomes for such surgery. We have performed sleep studies for symptomatic patients after surgery. For the 5 patients in our study, 2 underwent sleep studies before their participation in our study, 1 was being observed at another institution, 1 had been scheduled twice for a sleep study but did not follow through before he was recruited for our study, and 1 had no documented airway or sleep complaints in the medical record. We routinely admit patients for monitored observation with the tracheotomy tube cuffed before decannulation, and we also obtain sleep study results with the tracheotomy tube capped.

Our study has demonstrated the long-term improvement of OSA in 4 of 5 children with a median follow-up of 47.5 months. Of these 5 patients, 1 had severe OSA documented, with an AHI of 20.2. This patient was the only patient who completed the study who underwent a previous tracheotomy; this patient also had the highest score on the pediatric OSA symptom questionnaire. In our small sample of patients who underwent mandibular distraction, it is not possible to draw conclusions on the sensitivity and specificity of either clinical assessment or polysomnography for the diagnosis of OSA after this procedure. However, previous studies of children who have not undergone mandibular distraction have shown that medical history and physical examination alone are not reliable in assessing for OSA. Polysomnography is the gold standard for evaluation of OSA and, therefore, should be offered to children with obstructive symptoms after distraction.

Our study is limited by the small sample size. Additional long-term prospective evaluation of more children undergoing mandibular distraction is necessary to fully understand the benefits and limitations of this surgery.

In conclusion, most young children with upper airway obstruction from mandibular hypoplasia have long-term relief of OSA after mandibular distraction. In children with continued or recurrent symptoms of sleep-disordered breathing, clinical evaluation and polysomnography are recommended.

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REFERENCES


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**Announcement**

**Trial Registration Required**

In concert with the International Committee of Medical Journal Editors (ICMJE), *Archives of Otolaryngology–Head & Neck Surgery* will require, as a condition of consideration for publication, registration of all trials in a public trials registry (such as http://ClinicalTrials.gov or http://controlled-trials.com). Trials must be registered at or before the onset of patient enrollment. This policy applies to any clinical trial starting enrollment after July 1, 2005. For trials that began enrollment before this date, registration will be required by September 13, 2005, before considering the trial for publication. The trial registration number should be supplied at the time of submission.

For details about this new policy, and for information on how the ICMJE defines a clinical trial, see the editorial by DeAngelis et al in the June issue of *Archives of Otolaryngology–Head & Neck Surgery* (2005;131:479-480). Also see the Instructions for Authors on our Web site: http://www.archoto.com.