

Skeletal expansion combined with soft-tissue reduction in the treatment of obstructive sleep apnea in children: Physiologic results

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Twenty consecutive children, ranging in age from 6 days to 18 years, were treated with skeletal expansion, in addition to soft-tissue reduction, for medically refractory obstructive sleep apnea. The underlying diagnoses were craniofacial microsomia ($n = 6$), Down syndrome ($n = 3$), Pierre Robin syndrome ($n = 3$), cerebral palsy ($n = 3$), Nager's syndrome ($n = 1$), Treacher Collins syndrome ($n = 1$), cri du chat syndrome ($n = 1$), juvenile rheumatoid arthritis ($n = 1$), and temporomandibular joint ankylosis ($n = 1$). Fourteen children had severe medically refractory sleep apnea and were tracheostomy candidates; in the remaining six, tracheostomies were placed shortly after birth and could not be decannulated. Overnight, 12-channel polysomnography was obtained before and after surgery. The mean apnea index improved from 7.42 to 1.26, the mean respiratory disturbance index improved from 25.24 to 1.72, and the mean lowest apnea-related oxygen saturation improved from 68% to 88%. Of the 14 children with medically refractory obstructive sleep apnea, two required tracheostomies. Of the six patients with tracheostomies, five have been decannulated at the time of this writing. Skeletal expansion in conjunction with soft-tissue reduction in the pediatric population permits substantial increases in the volume of both the nasopharynx and oropharynx. Creative use of conventional osteotomies and the application of distraction osteogenesis have enabled surgeons to apply maxillofacial and craniofacial techniques in treating children with obstructive sleep apnea. (*Otolaryngol Head Neck Surg* 1998;119:476-85.)

Obstructive sleep apnea (OSA) in children can cause cardiopulmonary compromise and even death. Children with craniofacial and neurologic disorders are at particularly high risk for upper-airway obstruction. Regardless of the underlying cause of OSA, it is the resulting disproportion in skeletal and soft-tissue dimensions that anatomically compromises the upper airway. Sleep apnea in children is often insidious; changes in behavior, developmental delays, and deterioration in school performance may herald its onset. Noisy breathing during sleep, waking episodes, pauses in respiration, and daytime somnolence are frequently reported by the families of children with OSA.

Sleep apnea may have both a central and an obstructive component.¹⁻³ Central apnea may result from a variety of central nervous system abnormalities, such as brain stem compression and increased intracranial pressure. The peripheral obstructive component is always the result of one or more anatomic obstructions between the nares and bronchi. Once the diagnosis of peripheral OSA is confirmed on polysomnography, treatment is tailored to the severity and location of the problem. Medical therapy includes the use of weight reduction, supplemental oxygen, tooth-borne anterior repositioning devices, oropharyngeal and nasopharyngeal airways, positive-pressure mask ventilation, and, ultimately, intubation. Standard surgical approaches include relief of nasal obstruction, tonsillectomy, adenoidectomy, and uvulopalatoplasty. In adults in whom these measures fail, extensive skeletal expansion procedures—such as maxillary and mandibular advancement with tongue hyoid suspension—have provided relief of OSA.^{4,5} In children, however, few alternatives exist, and tracheostomy must be performed.

To stabilize the airway in children and prevent the placement of permanent tracheostomy with its cumulative morbidity and cost, we have employed an aggressive surgical approach using a combination of soft-tissue reduction and skeletal expansion. We previously reported findings from a group of 28 children with medically refractory sleep apnea who were treated primarily with the use of soft-tissue techniques and tongue hyoid suspension. At that time, we had not accumulated

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sufficient experience with skeletal expansion in the pediatric population. In children, procedures to expand the craniofacial skeleton often must be modified, either because of the patient's age or to address specific anatomic abnormalities of the upper airway. The purpose of this study was to present in detail the types of skeletal osteotomies we have used, focusing on a variety of modifications that have enabled their application in children with OSA.

METHODS AND MATERIAL

Over the past 6 years, 70 children under the age of 18 years at our institution have undergone surgical treatment of medically refractory OSA. Since January 1993, skeletal expansion and soft-tissue reduction have been routinely applied, when indicated, in children of all age groups with OSA. During this time, 20 consecutive children have undergone skeletal osteotomy, along with a soft-tissue procedure to correct OSA (Table 1). Our study group consisted of six girls and 14 boys, ranging in age from 6 days to 17 years. The underlying diagnoses were craniofacial microsomia ($n = 6$), Down syndrome ($n = 3$), Pierre Robin syndrome ($n = 3$), cerebral palsy ($n = 3$), Nager's syndrome ($n = 1$), Treacher Collins syndrome ($n = 1$), juvenile rheumatoid arthritis ($n = 1$), cri du chat syndrome ($n = 1$), and temporomandibular joint ankylosis ($n = 1$).

Of these 20 children, 14 had severe medically refractory sleep apnea and were tracheostomy candidates. In the remaining six, permanent tracheostomies for OSA were placed shortly after birth and the patients could not be decannulated in accordance with conventional criteria; instead they underwent skeletal expansion for tracheostomy removal.

Each patient underwent a diagnostic workup by a multidisciplinary team composed of a craniofacial surgeon, a pediatric otolaryngologist, and a pediatric pulmonologist. Medical evaluation included history and physical examination, chest and cervical spine radiographs, testing to rule out gastroesophageal reflux, 12-lead ECGs, flexible upper-airway endoscopy during spontaneous ventilation, and lateral fluoroscopy under sedation. When necessary, CT of the head and neck and cardiac ultrasonography were performed. Lateral cephalograms were obtained whenever possible.

Overnight, 12-channel polysomnograms were obtained in the 12 patients who were not intubated before surgery and who did not undergo tracheostomy. OSA is defined as complete cessation of ventilation. The apnea index (AI)—defined as the total number of apneic events divided by the total sleep time, multiplied by 60—was calculated from the polysomnographic data.⁶ The respiratory disturbance index (RDI)—defined as the number of apneas and hypopneas divided by the total sleep time, multiplied by 60—was also calculated.⁶ Last, the lowest oxygen saturation during an apneic or hypopneic event was recorded. Polysomnograms were obtained before surgery, after

Table 1. Study population ($n = 20$)

Diagnosis	No. of patients
Craniofacial microsomia	6
Pierre Robin syndrome	3
Down syndrome	3
Cerebral palsy	3
Treacher Collins syndrome	1
Cri du chat syndrome	1
Nager's syndrome	1
Juvenile rheumatoid arthritis	1
Temporomandibular joint ankylosis	1

Table 2. Soft-tissue procedures

Procedure	No. of patients
Tonsillectomy and adenoidectomy	4
Septoplasty/turbinectomy	5
Tongue reduction	6
Uvulopalatoplasty	10
Tongue hyoid suspension	12
TOTAL	37

Table 3. Skeletal procedures

Procedure	No. of procedures
Mandibular advancement	
Sagittal split	5
Inverted L	2
Costochondral graft	2
Mandibular distraction	10
Temporomandibular joint arthroplasty	1
LeFort I procedure	1
LeFort III procedure	3
Bipartition	1
TOTAL	25

surgery but before hospital discharge, and during follow-up if indicated by clinical history and findings.

Medically refractory OSA is considered to be present if clinical signs and symptoms or repeat polysomnographic studies fail to demonstrate improvement, even after weight reduction or the use of supplemental oxygen, tooth-borne anterior repositioning devices, or positive-pressure mask ventilation. In those children with adenoidal or tonsillar hypertrophy and nasal obstruction, conventional surgical treatments—such as turbinectomy, septoplasty, and tonsillectomy and adenoidectomy—are recommended. Children in whom these conventional therapies fail are then referred for more aggressive skeletal expansion combined with soft-tissue reduction procedures. In children with coexisting skeletal deformities such as severe retrognathia or maxillary hypoplasia, a combined approach is often played out: Tonsillectomy and ade-

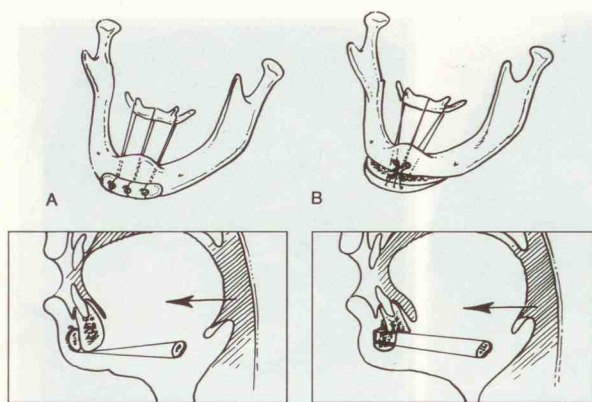


Fig. 1. Tongue hyoid suspension. **A**, In the younger child with unerupted dentition, a rib graft is onlaid over the inferior chin. Three permanent sutures are placed around the hyoid, and an inferior myotomy is made, releasing the central hyoid. The lingual surface of the mandible is stripped and the hyoid suspended to the rib graft, pulling it and its attachment to the tongue into a more anterior position. This pulls the tongue away from the retropharynx. **B**, In the older child whose permanent dentition has erupted, an osseous genioplasty is performed. The sutures are passed around the stabilizing metal plate, and the central lingual aspect of the mandible is not stripped. For further details of tongue hyoid suspension, see Burstein et al.⁶

noideotomy are performed at the same time as the other procedures. On the basis of our experience, we believe we can select those patients who would benefit from such a combined approach.

The soft-tissue and skeletal procedures used to increase the caliber of the airway are shown in Tables 2 and 3. We have previously discussed our indications and techniques for soft-tissue reduction in children with sleep apnea.⁶ We adhere to an airway zone concept to define functional and anatomic airway zones.⁶ Zone I extends from the nares to the velum. Zone II extends from the lips to the hypopharynx, excluding all laryngeal structures. Zone III extends from the epiglottis to the trachea, including the larynx. Last, Zone IV extends from the subglottis to the bronchi. Zones I and II are the focus of our investigation.

In tongue hyoid advancement (Fig. 1)⁶ in infants and children with primary or mixed dentition, onlay rib grafts are used in place of an osseous genioplasty to increase the anteroposterior length of the mandible, and the hyoid complex is suspended from the most anterior rib graft. The tongue hyoid suspension is carried out through a small submental incision. The hyoid bone is easily palpated and dissected by following the superior strap muscles to the central hyoid. The inferior strap muscles are released from the central hyoid, allowing the hyoid to be pulled upward and forward. Care must be taken not to stray from the central hyoid, avoiding injury to the superior branch of the recurrent laryngeal nerve. Once the

hyoid has been freed, two or three 2.0 permanent sutures are passed around it. The lingual aspect of the mandible is then dissected in the subperiosteal plane until all the muscle attachments have been freed. If a genioplasty is to be performed, the genioglossus is not detached. Two or three permanent sutures are then passed through the periosteum adjacent to the genioglossus. The hyoid sutures are then passed through holes in the anterior mandible around the stabilizing plate or into the rib graft through holes or around the screws. This pulls the hyoid forward and upward. The submental wound is closed in layers.

All patients with Goldenhar's syndrome and those lacking mandibular rami and condyles underwent unilateral or bilateral costochondral grafting, which produced or are producing sagittal advancement of the mandible, anteriorly repositioning of the tongue, and clearing of the retropharynx.

Mandibular osteotomies were performed in one of two ways, depending on the age of the patient and the status of the permanent dentition. If both the first and second permanent molars had erupted, bilateral sagittal split osteotomies of the mandible with rigid fixation were used for advancement (Fig. 2). In younger children whose permanent molars had not yet erupted, either an inverted L-osteotomy or distraction osteogenesis (Fig. 3) was performed. When mandibular shape and size were conducive, an inverted L-osteotomy with rigid fixation from either an intraoral or extraoral approach was used. In general, children younger than 6 or 7 years of age underwent distraction osteogenesis with an external lengthening device. This procedure involved external exposure of the ramus, angle and body of the mandible through the Risdon approach, and application of two distraction pins caudal and cranial to the proposed osteotomy of the mandible with care taken to preserve the inferior alveolar nerve.⁷ To obtain an immediate 1- to 1.5-cm advancement, however, an interposition corticocancellous bone graft was placed in the osteotomy gap and fixed to one side of the mandible (Fig. 3E and F). Distraction was then begun at a rate of 1 mm/day, beginning on postoperative day 5. The rationale behind immediate osseous expansion and bone grafting was to create an initial sagittal advancement of the base of the tongue, with the object of decreasing the postoperative ventilation time these children would require.

In children with midfacial retrusion or in whom anterior midface repositioning was warranted to sagittally advance or lower the nasal floor and soft palate, a LeFort I or III procedure was performed (Fig. 4). When narrowing of the width of the nasopharynx was present, transverse expansion was carried out with either a subcranial facial bipartition (if aesthetically tolerated) or a segmental LeFort I osteotomy (Fig. 5).

RESULTS

Over the preceding 2.5 years, 20 consecutive children with OSA were treated with the use of a combina-

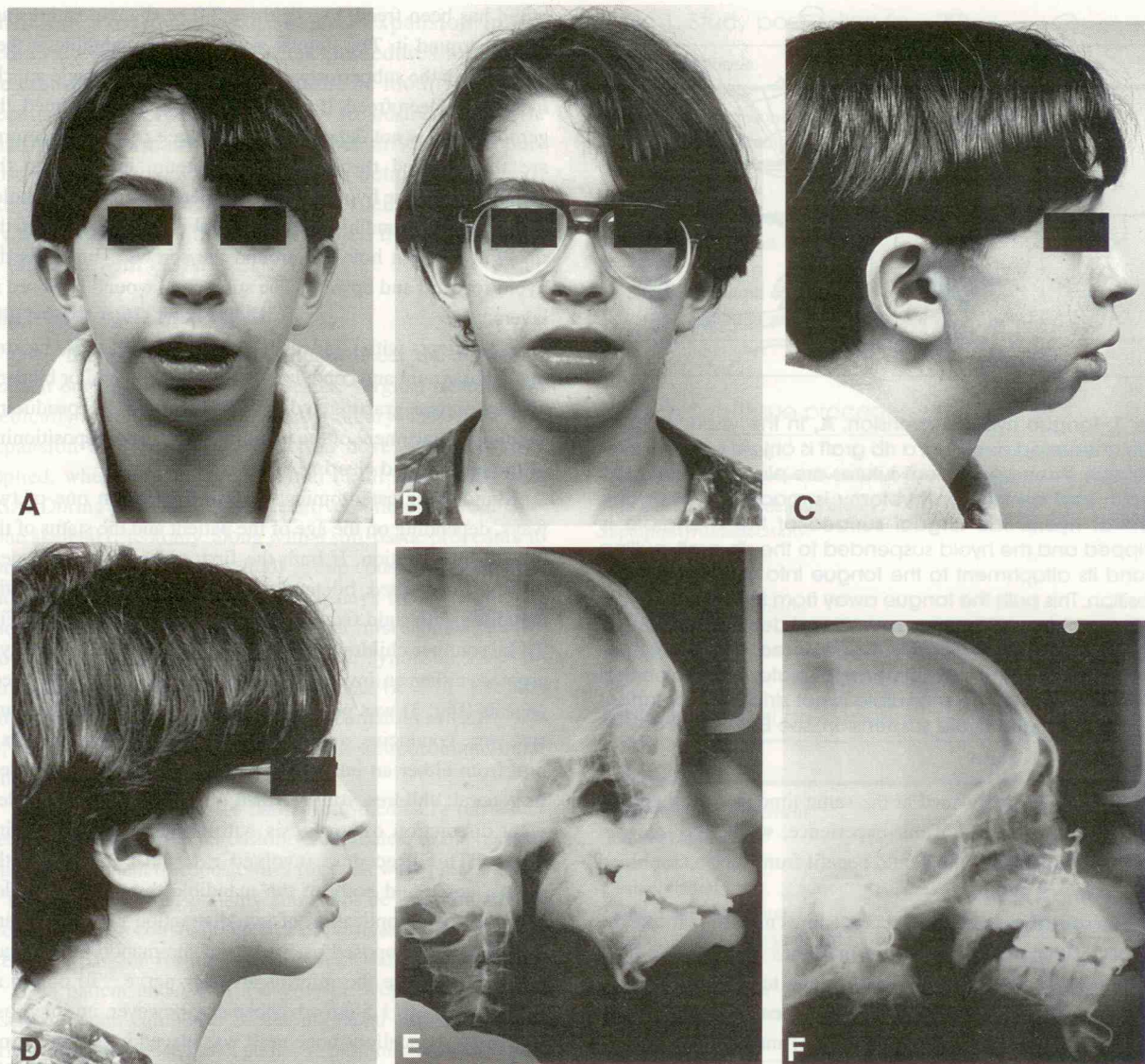


Fig. 2 **A**, Preoperative frontal photograph of 16-year-old boy with juvenile rheumatoid arthritis and juvenile diabetes who presented with severe retrognathia, vertical maxillary excess, transverse maxillary collapse, relative macroglossia, and obstructive sleep apnea. The patient underwent LeFort I clockwise rotation with transverse maxillary expansion, mandibular advancement, genioplasty, tongue reduction, uvulopalatoplasty, and tongue hyoid suspension. The patient's preoperative RDI was 15, his postoperative RDI 2.9; the lowest preoperative oxygen saturation was 76% and the lowest postoperative saturation 91%. **B**, Postoperative frontal photograph, 6 months after surgery. **C**, Preoperative lateral photograph. **D**, Six-month postoperative lateral photograph. **E**, Preoperative lateral cephalogram. Note thread-sized airway in front of soft palate. **F**, Three-month postoperative lateral cephalogram. Note the enlargement of airway dimensions.

tion of skeletal expansion and soft-tissue reduction. Each patient who was not intubated or did not have a tracheostomy was prospectively evaluated by means of polysomnography. Serial preoperative and postoperative polysomnographic results were available in 12 children; these are shown in Table 4. Postoperative sleep studies were conducted before the patient's discharge

from the hospital and repeated if clinical signs or symptoms of OSA recurred during follow-up. Two of the 20 children were intubated before surgery, and polysomnography could not be obtained. One of these two children had retrognathia, an elongated soft palate, and bilateral bony temporomandibular joint ankylosis. At 5 days of age he underwent tongue hyoid suspension, uvu-

