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.)

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0194-4998/98/$5.00 + 0 **23/1/87915**

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

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

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

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-