Surgical Therapy for Severe Refractory Sleep Apnea in Infants and Children: Application of the Airway Zone Concept

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Obstructive sleep apnea in children may result in hypoxia, right-sided heart failure, and sudden death. Children with craniofacial deformities and/or cerebral palsy are at high risk for the development of obstructive sleep apnea. Prompted by the excellent results obtained in adults when sleep apnea was managed by an aggressive surgical approach, we undertook a similar treatment philosophy in children. Twenty-eight patients representing four diagnostic groups were evaluated and operated on for severe upper airway obstruction: Down syndrome (n = 5), cerebral palsy (n = 12), Goldenhar syndrome (n = 4), and a mixed apnea group (n = 7). Tracheostomy was avoided in 25 of 28 patients (89 percent), with a marked decrease in apnea (median 90 percent) and hypopnea (median 87 percent) episodes. Tongue hypertrophy, suspension and skeletal expansion procedures, which were the mainstay of treatment, were applied for the first time in children and adolescents with obstructive sleep apnea. (Plast. Reconstr. Surg. 96: 54, 1995.)

Accepted treatments for obstructive sleep apnea include medical modalities such as positive-pressure mask ventilation, supplemental oxygen, temporary nasopharyngeal airways, and oropharyngeal airway placement.1-3 Standard surgical approaches include tonsillectomy, adenoidectomy, uvulopalatopharyngoplasty, and relief of nasal septal obstruction.3-5 Unfortunately, severely affected children may be refractory to standard medical and surgical treatments. These children may have no alternative other than permanent tracheostomy. While effective, this entails a significant increase in the level of home or institutional care required for the child. There also may be deleterious psychological, communicative, and social effects in the affected child and his or her family due to permanent tracheostomy.10-12

The focus of the present investigation is to provide a logical, sequential treatment strategy for children with severe obstructive sleep apnea refractory to standard medical and surgical treatments. Our goal is to avoid permanent tracheostomy whenever possible. In order to better define the anatomic problems, we have developed the concept of functional and anatomic airway zones (Fig. 1). 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. Finally, zone IV extends from the subglottis to the bronchi. We report on the application of the airway zone concept and multidisciplinary
team approach in the management of complex obstructive sleep apnea in children.

**Patients and Methods**

Twenty-eight patients ranging in age from 18 months to 17 years (mean 5 years and 3 months) were treated surgically for severe refractory sleep apnea. All had anatomic sites of obstruction in zones I and II. There were 17 males and 11 females. Follow-up ranged from 6 to 48 months (mean 16 months.) All our patients had multiple problems ranging from cerebral palsy to Down syndrome. For purposes of discussion, patients are divided into four diagnostic groups: group I (mixed), group II (Down syndrome), group III (cerebral palsy), and group IV (Goldenhar syndrome) (Table I). All prospective surgical candidates were evaluated by the pediatric pulmonology service (Scott, Teague, Montgomery) and the craniofacial surgery team (Burstein, Cohen). Medical evaluation included physical examination, chest and cervical spine radiographs, testing to rule out gastroesophageal reflux, 12-lead electrocardiogram, cardiac ultrasonography, flexible upper airway endoscopy during spontaneous ventilation, computed axial tomography (CAT scan) of the head and neck, and whenever possible, lateral head cephalograms. Overnight 12-channel polysomnograms were obtained in all patients. Obstructive apnea is defined as complete cessation of ventilation. The apnea index, defined as the total number of apneic events divided by the total sleep time and multiplied by 60, was calculated from the sleep data. The respiratory disturbance index, defined as the number of apneas and hypopneas divided by the total sleep time and multiplied by 60, also was calculated. All patients had undergone maximal medical treatment without significant improvement before being entered in our treatment protocol. Statistical analysis of the preoperative and postoperative apnea index and respiratory disturbance index values was carried out.

**Surgical Procedures**

All patients underwent a combination of procedures selected according to their sites of obstruction as defined by physical examination, endoscopy, and radiographic evaluation (Table II). Eight patients had previously undergone tonsillectomy and adenoidectomy for pre-
sumed adenotonsillar hypertrophy but still had significant airway obstruction. Procedures are listed by anatomic site in the following paragraphs.

**Zone 1 (Nasal/Nasopharynx).** Adenoidectomy, if not previously done, septoplasty, and inferior turbinectomy were performed in various combinations according to anatomic findings.

**Zone 2 (Oropharynx/Hypopharynx).** Tonsillectomy, if not previously done, and uvulopalatopharyngoplasty were carried out. Patients with Down syndrome and macroglossia underwent simultaneous central tongue reduction. The tongue was reduced through a central elliptical excision extending from the circumvallate papillae to the tongue tip. The width of the ellipse is determined by the amount and location of the greatest bulk. Care must be taken to stay at least 2 cm from the lateral border of the tongue to prevent damage to the hypoglossal nerve, lingual nerve, and lingual artery. In patients with retromicrognathia or lack of base of tongue support for neurologic reasons, a combination of bony and soft-tissue procedures were done. In children and adolescents with permanent dentition, sagittal split osteotomy of the mandible in combination with horizontal mandibular osteotomy and hyoid-tongue suspension was carried out (Figs. 2 and 3). In infants and children with primary or mixed dentition, onlay rib grafts are used in place of horizontal mandibular osteotomy 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 can easily be palpated and dissected by following the superior strap muscles to the central hyoid. The inferior strap muscles are then transected from the central hyoid segment to allow the hyoid to be pulled upward and forward. Care must be taken to stay on the central hyoid segment to avoid injury to the superior branch of the recurrent laryngeal nerve. Once the hyoid has been freed, two 2-0 permanent sutures are passed around it. The lingual aspect of the mandible is then dissected in the subperiosteal plane until all the muscular attachments have been freed; if a ge-
nioplasty is to be done, the genioglossus is not detached. Two 2–0 permanent sutures are then passed through the periosteum adjacent to the genioglossus muscle. The hvoid sutures are then passed through holes in the anterior mandibular segment or the most distal rib graft, and the hvoid is pulled forward and upward. In a similar fashion, the tongue sutures are passed through the same holes and brought forward and downward (Figs. 2 and 3). The submental wound is closed in multiple layers with absorbable sutures; no drains are used. In selected children with mixed dentition, mandibular lengthening was carried out using costochondral grafts for ramal reconstruction and/or distraction osteogenesis with mandibular corticotomies.

**POSTOPERATIVE MANAGEMENT**

All patients were kept intubated in the intensive care unit for 5 to 7 days with mechanical ventilatory support to allow the oropharyngeal and floor of mouth edema to decrease. Twelve hours prior to planned extubation, intravenous steroids were started along with vasoconstrictive nasal drops. These were used for the first 72 hours after extubation and then were stopped without tapering. The patients were monitored for 24 to 48 hours in the intensive care unit after extubation and then were placed on the floor with continuous pulse oximetry. An immediate postoperative sleep study was obtained prior to discharge from the hospital and was repeated 3 to 6 months postoperatively. Intravenous antibiotic prophylaxis was given for the first 7 days postoperatively.

**RESULTS**

The patients were divided into four groups according to pathologic diagnosis (Table I). Statistical analysis for intergroup and intra-group variations in preoperative and postoperative apnea index and respiratory disturbance index were carried out using nonparametric statistical methods. Wilcoxon signed-rank tests were used to produce statistical significance tests for the average change within subject groups. Kruskal-Wallis tests were used to compare median changes across diagnostic groups, and Spearman rank correlation coefficients were used to assess the degree of dependence of the changes in apnea index and respiratory disturbance index on the preoperative values.\(^{15}\)

Twenty-five of the 28 patients treated had improvement in their apnea index or respiratory disturbance index measurements. The median change overall was −3.0 for the apnea index \((p = 0.0009)\) and −12.1 for the respiratory disturbance index \((p < 0.0001)\). The median percentage change for the apnea index was −9.0 percent, and it was −87 percent for the respiratory disturbance index. Figures 4 and 5 compare the preoperative and postoperative median apnea index and respiratory disturbance index for the study subjects by diagnosis group. Two patients, one in group II and one in group III, required emergency intubation for airway obstruction before preoperative sleep studies could be obtained. Postoperative studies demonstrated an apnea index of 2.66 and a respiratory disturbance index of 3.00 for the group II patient and an apnea index of 4.00 with

![Graph](image_url)

**Fig. 4.** Comparison of median preoperative and postoperative apnea index in the four diagnostic treatment groups.
a respiratory disturbance index of 9.20 for the group III patient. Generally accepted measurements for normal children are an apnea index of less than 4 and a respiratory disturbance index of 8 or less.\textsuperscript{14-16}

Three patients in group III were considered complete failures. All were afflicted with severe cerebral palsy with flaccidity. Repeated attempts at decannulation failed; two of these individuals were salvaged with tracheostomies. The third had aspiration pneumonia and died of septic complications. Four patients had good initial results but had gradual deterioration, with moderate degrees of obstructive apnea occurring between 4 and 6 months after surgery. Two patients, noted to have redundant lateral pharyngeal walls, had good response to repeated uvulopalatopharyngoplasty and lateral pharyngeal tightening with maintenance of airway support. One patient had massive regrowth of adenoidal tissue and was salvaged with repeated adenoidectomy. Finally, one patient who had an excellent initial response had acute deterioration and was found to have broken the sutures suspending the base of the tongue–hyoid complex. Resuspension with surgical wire was curative.

Two patients in group III exhibited a marked increase in apneic events 2 to 6 months after initially successful surgery. Both individuals had severe cerebral palsy with low functional levels. Polysomnography coupled with electroencephalography revealed that the apneic events were related to seizure activity that was not clinically evident. Treatment with anticonvulsants was effective in both cases.

\textbf{DISCUSSION}

The application of the anatomic zone concept of upper airway obstruction to the diagnosis and treatment of obstructive apnea was successful in avoiding tracheostomy in 25 of 28 patients, with significant improvement in both apnea index and respiratory disturbance index measurements (see Figs. 4 and 5). We attribute our success to being able to thoroughly evaluate and subsequently select those patients with anatomic obstruction in zones I and II who were amenable to surgical therapy. Patients with extreme flaccidity and cerebral palsy were generally not good candidates for upper airway surgery. Patients ultimately selected for operative therapy had definite structural craniofacial findings that were treatable by alteration of abnormal morphologic findings.

There is no general agreement as to what represents a normal apnea index or respiratory disturbance index in children, with ranges of 3 to 5 for apnea index and 5 to 10 for respiratory disturbance index being given.\textsuperscript{14-16} Compromised patients, such as those with cerebral palsy (group III), will have far less tolerance for apneic events than a child without neurologic compromise and with normal pulmonary reserve. Although the initial median values for the apnea index and respiratory disturbance index of the group III patients were relatively low (see Figs. 4 and 5), all these individuals were hospi-
talized because of severe oxygen desaturation episodes often accompanied by bradycardia.

There were significant intergroup differences in the severity of the preoperative apnea index and respiratory disturbance index scores (see Figs. 4 and 5). The patients in groups II and IV had more severe apnea than those in groups I and III preoperatively, with greater improvement in apnea index and respiratory disturbance index postoperatively. These differences reached statistical significance \( p < 0.05 \). The most notable feature in groups II (Down syndrome) and IV (Goldenhar syndrome) was the relative imbalance between soft tissues and their skeletal supporting framework. In group II, maxillary hypoplasia magnified the macroglossia found in all the Down syndrome patients. In group IV, the absence of one or, in one case, both condyles and mandibular rami gave the soft tissues of the oral and pharyngeal cavities little rigid support. We postulate that this lack of skeletal framework predisposes these patients to prolapse of the oropharyngeal soft tissues into the airway during inspiration. These patients responded well to skeletal expansion.

Macroglossia was present in all of the Down syndrome patients, and central tongue retraction was used as an adjuvant measure to prevent protrusion of the tongue beyond the oral cavity as the hyoid-tongue complex was advanced. We and others have observed that macroglossia alone is seldom sufficient to precipitate significant obstructive apnea.\(^{17,18}\) We contrast our positive results with those of Mixter et al.\(^ {19} \) and Lauritzen et al.\(^ {20} \) who performed heroic midface advancements at an early age to improve the airway with relatively little success. Perhaps their patients were more severely affected than our group. Alternatively, they probably did not address the multifactorial etiology of the airway problem. We speculate that in their patients midface advancement improved the nasopharyngeal airway dimensions but did not correct the narrowed nasal airways or lack of base of tongue support that is often found in this group of syndromic patients. Their approach simply moves the obstruction forward, leaving the nasal airway resistance unchanged. It does not address possible obstructive sites in zone II.

Jones\(^ {21} \) has presented encouraging early results combining monobloc advancement with facial bipartition in Apert’s patients with severe obstructive apnea. The facial bipartition procedure as described by Tessier\(^ {22} \) in the treatment of orbital telorbitism increases the width of the maxilla and entire nasal airway while decreasing the interorbital distance.

Neuromuscular factors are an important part of the pathophysiology of obstructive sleep apnea. Sauerland and Harper\(^ {23} \) and others\(^ {24,25} \) have demonstrated that the resting tone of the tongue musculature during sleep is decreased in individuals with obstructive apnea. Flexible nasopharyngeal endoscopy performed during sleep in patients with obstructive sleep apnea demonstrates collapse of the lateral pharyngeal walls with decrease in resting muscle tone.\(^ {23-25} \) These studies support our observation that patients with neuromuscular flaccidity such as is seen with cerebral palsy are at risk for obstructive apnea. Fujita\(^ {26} \) has described a simple relationship among airway resistance, flow, and pressure (Fig. 6). Note that the airflow will be decreased by increases in airway resistance or decreases in the pressure gradient between the upper and lower respiratory tracts. During rapid eye movement (REM) sleep, the diaphragmatic and accessory respiratory muscles continue to generate relatively constant negative pressure gradients, while there is a decrease in tonic pharyngeal and tongue muscle activity resulting in a greater tendency toward luminal collapse and thereby airway obstruction. In normal individuals, slight increases in arterial carbon dioxide and decreases in oxygen trigger the increase in tonic activity in the pharyngeal and tongue musculature that counteracts this tendency.\(^ {23,24} \) In individuals who have chronic neuromuscular conditions such as cerebral palsy, which is often accompanied by thoracic scoliosis, the ability to generate a negative pressure gradient may be severely compromised. We can appreciate from Figure 6 that in individuals who have a low pressure gradient between the upper and lower respiratory tracts, any changes in airway resistance will have a profound effect on airflow.

Adenotonsillar hypertrophy is a common cause of obstruction and may result in cor pulmonale in severe cases.\(^ {5} \) Several of our patients

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V = \frac{PD(\text{PA-PI})}{R}
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**Fig. 6.** Mathematical expression of upper airway dynamics in respiration: \( V \) = airflow, \( PD \) = pressure difference, \( \text{PA} \) = atmospheric pressure, \( \text{PI} \) = inspiratory pressure, and \( R \) = respiratory resistance.
had previous adenotonsillectomy in an attempt to relieve the airway obstruction. Although temporary improvement was reported in a few patients, all eventually deteriorated, requiring further treatment. This reflects the severe nature and multifactorial etiology of obstruction in these compromised patients. The role of the soft palate and lateral pharyngeal walls in snoring and sleep apnea has become the subject of intense interest. Uvulopalatopharyngoplasty was initially advocated as treatment for both snoring and sleep apnea. There is general agreement that uvulopalatopharyngoplasty is effective in snoring but unreliable in treatment of severe obstructive apnea. In our series we performed an extended uvulopalatopharyngoplasty in order to remove redundant tissue from the lateral pharyngeal walls as an adjuvant measure.

Abnormalities in size, position, or morphology of the skeletal tissues will have an impact on the soft tissues of the upper airway zones and therefore airflow. In the group III patients, contributing skeletal abnormalities included turbinate hypertrophy, sepal deviation, choanal narrowing, and relative retromicrognathia caused by vertical maxillary overgrowth. The effect of poor upper pharyngeal muscle tone combined with retromicrognathia and a narrow transverse maxillary dimension seen in patients with cerebral palsy was particularly challenging. The base of tongue position was retrodisplaced by poor genioglossus tone and the retrodisplaced mandible. The tongue-hyoid suspension combined with segmental or total mandibular advancement was quite effective in maintaining oral pharyngeal patency by increasing soft-tissue support through skeletal expansion. The mechanical effect of suspending the hyoid from the mandible is to pull the base of the tongue forward and downward away from the posterior oropharyngeal wall, thereby increasing the airway diameter (see Figs. 2 and 3). Riley et al. have described a similar approach in adults with obstructive sleep apnea, but ours is the first reported series in children. Unlike the Riley procedure, our approach advances not only the base of tongue and genioglossus but also the rest of the intrinsic tongue muscles forward by moving their periosteal attachments anteriorly. This tends to tighten the anterior tongue musculature, preventing glossophtosis.

The three complete failures in our series occurred in patients with cerebral palsy and extreme flaccidity, with essentially absent head control, intrinsic pulmonary disease, as well as compromised diaphragmatic and accessory respiratory musculature function. Despite resolving the skeletal to soft-tissue disproportion, as well as positioning the tongue forward, these patients could not overcome even minimal upper airway resistance. Although we have not added standard spirometry to our preoperative evaluation protocol at this time, it may be prudent to do so in patients with intrinsic pulmonary disease or with poor thoracic muscular function. Partial failures were due to overly conservative initial uvulopalatopharyngoplasty adenoidectomy in three patients and to mechanical failure of the hyoid suspension in one patient. All were salvaged without tracheostomy.

Seizure disorders often coexist with obstructive sleep apnea in patients with craniofacial anomalies, neurologic compromise, or cerebral palsy and may be aggravated by short hypoxic episodes. It is crucial to separate apneic events caused by seizures from pure central or obstructive events, since therapy is quite different depending on etiology. We recommend a combined overnight sleep and EEG study with video monitoring in those patients with a poorly controlled seizure disorder. We were able to salvage two patients who appeared to be initial surgical failures with improved seizure control.

The potential short- and long-term medical and surgical complications of tracheostomy in children with craniofacial conditions or neurologic compromise have been well described. Mixter et al. described granulation tissue causing complete tracheal obstruction and death in their report. In patients who attend special education classes, such as those with Down syndrome, avoidance of tracheostomy has a tremendous psychological advantage. In patients who are homebound or institutionalized, tracheostomy care can be time-consuming and costly. Parents or other nonmedical caregivers may find tracheostomy care overwhelming, making noninstitutional care impossible. In an era of cost restraints, an economic justification for new procedures such as the tongue-hyoid suspension is of great importance. We surveyed our own institution for the cost of tracheostomy versus the procedures described in this report over 3 years in order to provide an economic comparison. Although the initial cost of surgical therapy was higher than that of tracheostomy, this relationship was reversed after the first 6 months because of the
ongoing cost of tracheostomy supplies, otolaryngology follow-up, and home, institutional, or school nursing help.

Our results suggest that a multidisciplinary team approach, thorough preoperative evaluation, and the application of the zone concept of airway management can successfully treat severe obstructive sleep apnea in medically compromised infants and children, avoiding the necessity for tracheostomy in most cases.

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REFERENCES


