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REVIEW ARTICLE

Pediatric Obstructive Sleep Apnea: Surgical Techniques beyond Tonsillectomy and Adenoidectomy

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ABSTRACT

Sleep disordered breathing (SDB) affects 1 in 10 children in the United States and poses a growing threat to childhood health. Although tonsillectomy and adenoidectomy is considered the standard of care for treatment of pediatric SDB, up to 25% of children present with persistent symptoms after surgery. Success of treatment modalities, such as continuous positive airway pressure (CPAP) is affected by compliance. Management of residual SDB is a complex, and often controversial topic. Here we discuss options for managing childhood SDB that persists after initial management with tonsillectomy.

Keywords: Adentonsillar hypertrophy, Multilevel sleep surgery, Obstructive sleep apnea, Pediatric sleep disordered breathing.

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INTRODUCTION

Pediatric sleep-disordered breathing (SDB) is a spectrum of sleep disorders that include in increasing severity – primary snoring, upper airway resistance syndrome, and obstructive sleep apnea (OSA). While all three conditions are associated with varying degrees of partial or complete airway obstruction during sleep, OSA leads to a cycle of hypoxemia, respiratory compensation, heart rate variability, and awakening. Sleep-disordered breathing most commonly occurs as a result of hypertrophy of palatine (*tonsil*) and nasopharyngeal (*adenoid*) lymphoid tissues. Tonsillectomy and adenoidectomy (T&A) is the first line of treatment and leads to improvement or resolution of SDB in the majority of children.[1]

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The growing awareness of the negative consequences of SDB has made it the principal indication for T&A in the last decade. Tonsillectomy and adenoidectomy for SDB leads to improvements in sleep, behavior, and quality of life. However, only 80% of the low-risk children undergoing the procedure will normalize the sleep disorder as measured by polysomnography (PSG). A proportion of children will need reevaluation for recidivism that may hinder normal growth and development. Surgical outcomes are also suboptimal in children with obesity, craniofacial and neuromuscular disorders. Although treatment strategies, such as nasal continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BiPAP) have been proposed in children in whom T&A provided only partial benefit, compliance is poor.[2]

With half a million tonsillectomies performed in children in the United States every year, there is a critical need to determine whether additional surgical procedures may provide durable benefit when CPAP/BiPAP fail. The purpose of this review is to examine the role of surgical procedures for persistent OSA after T&A.

CLINICAL EVALUATION OF CHILDREN WITH PERSISTENT OSA AFTER T&A

Less than 5% of children undergo PSG after T&A.[3] Although T&A for OSA results in significant improvement in respiratory parameters in the majority of healthy children, there is a lack of correlation between improvements in PSG and quality of life.[4] In addition, children with severe OSA are more likely to have persistent OSA after T&A with an odds ratio of 1.5 to 2.5, compared with those with mild or moderate OSA.[5] In these children, it is important to follow a structured history and physical examination and direct attention to critical symptoms such as snoring and witnessed apneic events as they may reflect persistence of OSA.

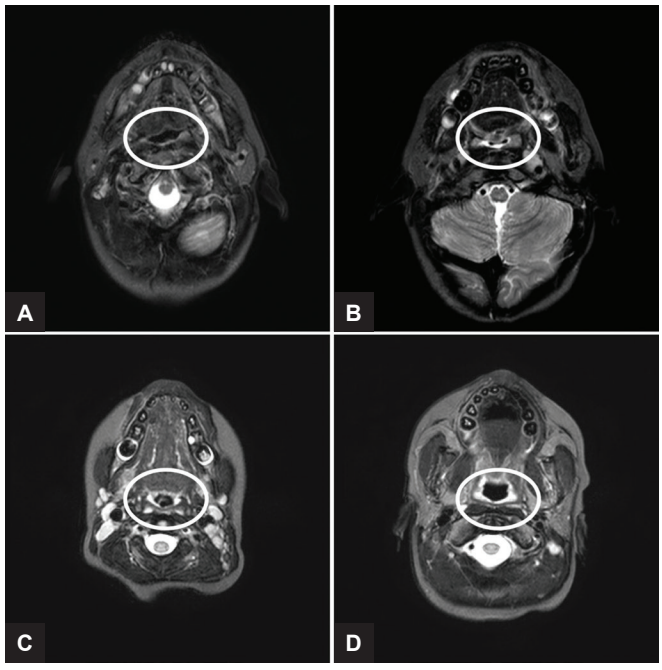
The growing incidence of obesity as well as postoperative weight gain observed in children undergoing T&A could make a strong case for more frequent and routine PSG to monitor progression of OSA. At this time, there is paucity of data supporting routine postoperative PSG in all children (American Academy of Otolaryngology—Head and Neck Surgery guidelines) considering the cost of such diagnostic testing. However, rebound weight

gain warrants repeat PSG in symptomatic children. Furthermore, follow-up PSG is unequivocally necessary in children with craniofacial malformations, Down syndrome, or neuromuscular disorders. Once a diagnosis of persistent OSA is made, it is important to explore the sites of residual upper airway obstruction. Classification systems used in management of adult OSA, such as the Müller maneuver, Fujita and Mallampati scores have been unsuccessful in children. In lieu of these, imaging protocols may be necessary, and the following section examines the various options.

IMAGING UPPER AIRWAY OBSTRUCTION

Imaging can provide critical information regarding upper airway dimensions and sites of obstruction in children with persistent OSA. The contribution of nonlymphoid soft tissue in the upper airway of children with a diagnosis of OSA cannot be overstated especially when obesity is present. These areas, including the tongue, soft palate, lateral pharyngeal walls, parapharyngeal fat pads, and mandible, may be studied with nasopharyngoscopy, cephalometry, fluoroscopy, computed tomography (CT) and magnetic resonance imaging (MRI). Awake techniques are limited by lack of cooperation in some children. Among these methods, the least invasive is upper airway flexible endoscopy under sedation. Termed drug-induced sleep endoscopy (DISE), fiber-optic examination of the upper airway under optimal sedation (e.g., propofol) has been shown to have high interrater reliability as well as validity when compared with PSG. Durr et al[6] have formulated a standardized protocol to examine the upper airway for OSA. In addition to the VOTE strategy for functional assessment of velum, the oropharyngeal lateral walls, the tongue base, and epiglottis, additional sites of potential obstruction include the inferior turbinates, the adenoid pad, and the larynx (laryngomalacia).

Both cephalometry and CT scanning are of limited value due to exposure to ionizing radiation and reduced soft tissue resolution. As MRI offsets these issues, it has generated considerable interest. Refinements such as respiratory gating, three-dimensional (3D) reconstruction, and phase-contrast imaging have collectively contributed to reducing motion artifacts and improving scan times. At the apex of MRI technology for localization of obstruction is cine MRI, a relatively new method that encompasses advances in dynamic imaging. First used for cerebrospinal fluid flow studies, cine MRI provides simultaneous visualization of multiple images of the airway from various projections without overlap.[7] The reliability of cine MRI is affected by the type and depth of sedation administered. Consequently, standardized protocols are necessary to ensure that the depth of sedation closely approximates stages of natural sleep. Representative images from our institution are shown in Figures 1A to D.



Figs 1A to D: Site of obstruction imaging using cine MRI; (A) Example of axial short tau inversion recovery image outlining (white circle) a relatively normal airway caliber at the level of the hypopharynx; (B) at the base of the tongue, the airway is considerably effaced; (C) and (D) similar sequences from a separate patient with airway narrowing at the level of the hypopharynx, yet a relatively normal caliber at the level of the tongue

NONSURGICAL MANAGEMENT OF RESIDUAL PEDIATRIC OSA

Medical management of persistent OSA after T&A spans pharmacologic therapy, the use of CPAP, and oral appliances. Mild persistent OSA (apnea-hypopnea index, AHI < 5) after T&A may be successfully treated with nasal corticosteroids and oral anti-inflammatory therapy such as leukotriene inhibitors, e.g., montelukast. Some studies have described clinical improvement and complete normalization of sleep by PSG using these therapies.[8] This line of management takes into consideration the likely surge in inflammatory mediators as a consequence of untreated OSA. That said, the routine use of anti-inflammatory medications, regardless of the route of administration, has been shown to be ineffective in the management of moderate to severe OSA (AHI > 5).

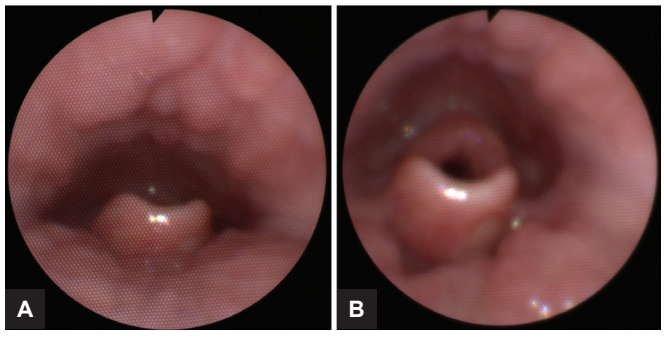
Oral appliances, designed to improve oropharyngeal airflow by mechanical effects on subsites within the upper airway, such as the tongue and palate, have limited benefit in children and their success is closely related to compliance. Some degree of success may be observed when occlusion problems are identified and custom-made orthodontic appliances are used.

Both CPAP and BiPAP are highly effective in children with OSA, but in general, their use is limited by disappointingly high dropout rate and parental assessment of either therapy notoriously overestimating actual usage.[9]

SURGERY OF THE OROPHARYNX

One of the first described oropharyngeal surgeries for management of OSA is uvulopalatopharyngoplasty (UPPP), which comprises expansion of the oropharyngeal airway by removal of tonsillar tissue and the anterior surface of the soft palate, as well as the uvula. Uvulopalatopharyngoplasty is one of the commonest surgeries performed for management of adult OSA. It is usually performed by electrocautery, cold steel, or cold radiofrequency ablation. Uvulopalatopharyngoplasty in combination with T&A was first described as an alternative to tracheostomy in children with significant neurological impairment and moderate to severe OSA. Kerschner et al[10] studied a small group of children who underwent UPPP and T&A, and noted that the short-term clinical improvements may not be durable enough for application to a wider population of children in whom neuromuscular tone may be a principal contributor to OSA. The procedure is rarely performed in children though modifications of UPPP may have a role in children especially as an adjunct to CPAP therapy.

Tongue base surgery for pediatric OSA has gained popularity in the last decade. The tongue may be a surgical target when it is (i) absolutely enlarged due to overgrowth of lymphoid tissue at its base, or relatively enlarged as a consequence of craniofacial morphology—specifically Down syndrome, (ii) malpositioned either due to resting or sleep-related neuromuscular tone abnormalities, or (iii) due to a combination of (i) and (ii). Relatively straightforward techniques to approach and reduce the volume of tongue tissue have been described recently[11] with more widespread availability of cold radiofrequency ablation (Figs 2A and B). This technique may be used to minimize lingual tonsillar tissue via either a small incision anteriorly (SMILE, submucosal minimally invasive lingual excision) or following endoscopic exposure with laryngoscopic assistance. Tongue base reduction is facilitated by submucosal dissection, akin to a partial glossectomy. On the other hand, lingual tonsillectomy removes lymphoid tissue without surgically addressing intrinsic tongue musculature. The success rates (defined by postoperative reduction of AHI by ≥ 50%) of both procedures approach 50%, but submucosal dissection may be associated with more complications. Use of endoscopic and ultrasonographic guidance reduces the risk of injury to the lingual neurovascular bundle. Successful surgical outcomes are related to patient and procedure selection, as well as the experience of the surgeon and the center performing the procedure.



Figs 2A and B: Tongue base reduction to improve oropharyngeal airway: (A) Preoperative flexible endoscopic examination showing significant base of tongue obstruction that appears to retroflex the epiglottis toward the airway; and (B) same patient following cold radiofrequency ablation of tissue from the base of tongue improving airway luminal diameter

Altering the tension applied to the genioglossus muscle can provide secondary benefits on the position and tone of the tongue, by reducing upper airway collapsibility. This may be traditionally achieved by advancing the muscle itself, in combination with a trapdoor drilled out of the mandible, or by using the Repose™ system that relies on passing a suture through the substance of the tongue. Although principally approved for adults with OSA, there are few reports detailing their use in a pediatric population with limited success. Synchronous additional suspension sutures around the hyoid prevent collapse and maintain an improved oropharyngeal airway. Overall success rate when performed in combination with ablation of the base of tongue is reported to be in excess of 60%.[12]

SURGERY OF THE FACIAL SKELETON

Facial dysmorphisms and craniofacial syndromes account for the majority of facial skeletal defects that lead to upper airway obstruction. Prone positioning resolves upper airway obstruction in many infants with Pierre–Robin sequence by alleviating airway obstruction caused by a retrognathic and a malpositioned and obstructive tongue. A smaller proportion of children require a temporary tracheostomy for relief of upper airway obstruction. The gold standard to provide a long-lasting upper airway free of obstruction in children with Pierre–Robin sequence has become distraction osteogenesis. The procedure exploits the ability of the pediatric facial skeleton to heal rapidly by osteosynthesis. Prior to surgery, all infants undergo 3D CT scan of the craniofacial skeleton, followed by image reconstruction. In many series, distraction osteogenesis facilitated either avoidance or early decannulation of tracheostomy in about 80% of the patients undergoing the procedure.[13]

Rapid maxillary expansion (RME) is an orthodontic procedure used to treat contracted maxillary arches. The principal goal of RME in children with a reduced nasopharyngeal and oropharyngeal airway is to address the posterior cross-bite and widen the maxilla, with secondary benefits in management of OSA. Villa et al[14] have shown that RME causes substantial reduction of AHI when performed in children with existing dentofacial anomalies. Rapid maxillary expansion has an innate tendency to alter facial growth patterns, hence this procedure should be chosen carefully.

SURGERY OF THE HYPOPHARYNX

Although lateral pharyngeal collapse is widely described in both adults and children with OSA, methods to address hypopharyngeal airway caliber and tone have not been studied extensively. The most comprehensive of these procedures, the expansion sphincter pharyngoplasty (ESP), involves a combination of tonsillectomy, expansion pharyngoplasty, rotation of the palatopharyngeus muscle, a partial uvulectomy, and closure of the anterior and posterior tonsillar pillars.[15] The procedure is performed following an initial endoscopic evaluation under sedation to document the site of maximal obstruction. When controlled for demographic variables, associated conditions, and preoperative AHI, ESP resulted in more resilient reduction of severity of OSA, projecting the feasibility of the procedure as both an adjunct and an alternative to T&A in patients with documented hypopharyngeal collapse.

TRACHEOSTOMY FOR PEDIATRIC OSA

A tracheostomy offers complete relief of upper airway obstruction and facilitates access to the lower airway for pulmonary toilet as well as mechanical ventilation. A tracheostomy may be performed (i) as a temporizing measure for relief of upper airway obstruction in young children unrelated to OSA (e.g., Pierre–Robin sequence), (ii) in primary pulmonary pathology as a consequence of prematurity, or (iii) in untreated severe OSA to avoid the severe secondary detrimental effects on cardiopulmonary physiology (e.g., right heart failure). Unfortunately, there is significant paucity of data on the incidence of pediatric tracheostomies for the sole indication of severe OSA, as it may be more common to associate OSA as a secondary diagnosis attached to craniofacial syndromes or pathology related to neuromuscular tone. In one such series, Robison et al[16] reported that the incidence of tracheostomy in a cohort of young children (age range, 3–24 months) was less than 2%. The authors described that the maximum objective resolution of OSA was seen in this group, followed by children who were treated by CPAP/BiPAP. Pediatric compared with adult tracheostomies, although performed at moderate frequency in tertiary care institutions, have an elevated risk profile, with greater incidence of accidental decannulation and acute airway obstruction. Further research is needed to determine the physiologic and anatomic profiles of children who may be better candidates for a tracheostomy compared with other surgical procedures.

NOVEL THERAPIES

Current trends in research within neuromuscular control mechanisms of upper airway patency have spurred development of novel therapies utilizing implanted stimulators. Initial research examining genioglossal activation patterns demonstrated the clear and steep decline in neuromuscular tone during sleep from electromyographic recordings from genioglossus in adult patients with OSA. This is particularly seen during rapid eye movement sleep. Conceptual application of this finding has led to the invention of a hypoglossal nerve stimulator (Inspire™ system, Inspire Medical, Maple Grove, MN, USA), which has provided promising results in its first clinical trial, with a greater than 67% response rate.[17] Although not currently approved in children, this technology may have application in children in whom the primary mechanism for upper airway obstruction is related to neuromuscular tone problems rather than obesity.

A STEPWISE APPROACH TOWARD MANAGEMENT OF RESIDUAL OSA AFTER T&A

At our institution, we have formulated a standardized approach toward management of residual OSA after an initial diagnosis of severe OSA. When a PSG confirms OSA in a child with symptoms lasting more than 3 months, we recommend T&A as the first line of treatment, regardless of medical comorbidities or the severity of OSA. Children who are symptomatic more than 3 months after surgery undergo reevaluation with flexible endoscopy in the office. The nasal cavity, postnasal space, tongue base, and larynx are evaluated and an allergy evaluation and/or repeat adenoidectomy initiated. A postoperative PSG is obtained 3 to 6 months after surgery in symptomatic children and all children with an AHI > 10. Symptomatic children with an AHI > 5 are evaluated in the multidisciplinary sleep medicine clinic and started on PAP therapy (CPAP or BiPAP) and continued for a minimum of 6 months. During this time, compliance reports from the on-board computer attached to the PAP machine are regularly monitored. Treatment for comorbidities such as nutritional intervention for obesity is also initiated. If compliance is poor, or if there is concern regarding rising pressures, a cine MRI is performed under anesthesia to

localize the site of obstruction. Drug-induced sleep endoscopy is rarely performed as a sole procedure in children and is usually performed in the operating room prior to surgical intervention. The surgical procedure performed is based on the findings of the cine MRI and DISE. The most common patient seen has Down syndrome and undergoes a tongue base procedure (lingual tonsillectomy and/or tongue base reduction using cold radiofrequency ablation). The most common palatal procedure is an ESP at the time of the tongue base procedure or as a sole procedure. Children are routinely observed overnight on a regular floor and few need admission to the intensive care unit. A PSG is obtained 3 months after surgery and PAP is reinitiated after repeat titration. Surgical success is defined as reduction in AHI by 50% or more.

POSTOPERATIVE MANAGEMENT FOLLOWING PEDIATRIC OSA SURGERY

The severity of preoperative OSA is a predictor of postoperative respiratory adverse events (PRAEs) in children undergoing OSA surgery. The spectrum of PRAEs includes incidents such as transient laryngospasm that responds to conservative management—to catastrophic upper airway obstruction and postoperative pulmonary edema that needs reintubation and mechanical ventilation. The physiology of upper airway neuromuscular control is modulated by chronic hypoxia at the level of the brain stem, which in turn accentuates the loss of tone and protective reflexes subsequent to administration of opioids and benzodiazepines. Consequently, there is a need for close coordination between the otolaryngologist, anesthesiologist, and the critical care team to ensure safety of the child during the postoperative period.

Unless medically contraindicated, all children undergoing OSA surgery should receive intraoperative corticosteroids (e.g., dexamethasone), which may also need to be continued in the immediate postoperative period. Pain control is achieved using acetaminophen, nonsteroidal anti-inflammatory drugs, and judicious use of opioids. The child may also benefit from nasal CPAP and humidified oxygen therapy. Intensive care unit stay is not needed in all children with OSA and rarely needed beyond the 24 hours following surgery.

CONCLUSION

The rising prevalence of pediatric OSA poses clinical and scientific challenges in diagnosis and management. Although it is clear that T&A is an effective procedure that leads to clinically significant reduction of the burden of upper airway obstruction, nonresponders and partial responders constitute a population of about 100,000 children every year. Current and historic studies have largely focused on the effectiveness of T&A, leaving much room to formulate strategies for the management of residual OSA. Summarized in Table 1, procedures to address residual OSA should be part of the armamentarium of a clinician with expertise in management of upper airway obstruction. Unfortunately, attempts to compare the pathophysiology of residual OSA in children to adults have not been fruitful. Areas that require further research include imaging upper airway obstruction, which can be targeted by surgical procedures, and the role of multilevel airway surgery.

Table 1: Summary of surgical procedures other than T&A, grouped by surgical site

Site of obstruction	*Procedure*
Nasal	Adenoidectomy
	Septoplasty
	Inferior turbinate reduction
Tongue, including base of tongue	Lingual tonsillectomy
	Tongue base reduction
	SMILE
	Wedge excision procedures
Uvula and palate	UPPP
	Uvulectomy
	Palatal expansion
Pharyngeal and hypopharyngeal	Expansion pharyngoplasty
Maxilla and mandible	RME
	Maxillomandibular advancement
	Distraction osteogenesis
Multilevel	Multilevel surgery
	Tracheostomy

T&A: Tonsillectomy and adenoidectomy; SMILE: Submucosal minimally invasive lingual excision; UPPP: Uvulopalatopharyngoplasty; RME: Rapid maxillary expansion

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