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Evaluation and Management of Children with Obstructive Sleep Apnea Syndrome

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Abstract

Obstructive sleep apnea syndrome (OSAS) is a common pediatric disorder characterized by recurrent events of partial or complete upper airway obstruction during sleep which result in abnormal ventilation and sleep pattern. OSAS in children is associated with neurobehavioral deficits and cardiovascular morbidity which highlights the need for prompt recognition, diagnosis, and treatment. The purpose of this state-of-the-art review is to provide an update on the evaluation and management of children with OSAS with emphasis on children with complex medical comorbidities and those with residual OSAS following first-line treatment. Proposed treatment strategies reflecting recommendations from a variety of professional societies are presented. All children should be screened for OSAS and those with typical symptoms (e.g. snoring, restless sleep, and daytime hyperactivity) or risk factors (e.g. neurologic, genetic, and craniofacial disorders) should undergo further evaluation including referral to a sleep specialist or pediatric otolaryngologist and overnight polysomnography, which provides a definitive diagnosis. A cardiology and/or endocrinology evaluation should be considered in high-risk children. For the majority of children, first-line treatment is tonsillectomy with or without adenoidectomy; however, some children exhibit multiple levels of airway obstruction and may require additional evaluation and management. Anti-inflammatory medications, weight loss, and oral appliances may be appropriate in select cases, particularly for mild OSAS. Following initial treatment, all children should be monitored for residual symptoms and polysomnography may be repeated to identify persistent disease, which can be managed with positive airway pressure ventilation and additional surgical approaches if required.

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Keywords

Adenotonsillectomy (AT); diagnosis; obstructive sleep apnea syndrome (OSAS); pediatrics; polysomnography; sleep disordered breathing (SDB)

INTRODUCTION

Sleep disordered breathing (SDB) in children refers to a spectrum of disorders which occur during sleep including central apnea, hypoventilation, and obstructive hypoventilation. The most severe manifestation of obstructive hypoventilation, obstructive sleep apnea syndrome (OSAS), is characterized by recurrent events of partial or complete upper airway obstruction during sleep which leads to abnormal ventilation and sleep pattern [1, 2]. The prevalence in children is 2–4% [3] and is increasing with the rising trend of childhood obesity [4]. OSAS is associated with neurobehavioral deficits [5], cardiovascular morbidity [6], poor quality of life [7], and increased health care utilization [8] which underscores the importance of detection and treatment. Consequently, the American Academy of Pediatrics (AAP) recommends screening for OSAS at routine medical visits. The diagnosis should be considered in children with typical symptoms (e.g. snoring, restless sleep, or daytime hyperactivity) or risk factors (e.g. craniofacial, neurologic, or genetic disorders) [9] and is confirmed with overnight polysomnography.

While the pathophysiology of pediatric OSAS is multifactorial, the most common cause is overgrowth of the adenoid and tonsils leading to upper airway restriction during sleep. Accordingly, first-line treatment is tonsillectomy with or without adenoidectomy, collectively referred to as adenotonsillectomy (AT) [1, 10]. Respiratory parameters improve following AT in the majority of children, but 40–75% have some level of persistent disease [11–13]. When AT is not indicated, if there is residual OSAS following surgery, or when medical comorbidities are present, additional evaluation and a modified management plan is often required. This may involve positive airway pressure (PAP) ventilation, weight management, dental procedures, secondary airway surgery, or medical therapy. Choice of treatment is tailored to the individual child depending on their comorbidities, preferences, and disease severity and is planned in consultation with a specialist in pediatric otolaryngology or sleep medicine.

EVALUATION

A proposed strategy for the evaluation of children with suspected OSAS is presented in Table 1.

I. History and physical examination

Clinical history—The most common complaints reported by parents of children with OSAS are snoring and difficulty breathing during sleep. Although almost all children with OSAS snore, snoring has low specificity for OSAS and clinical symptoms alone cannot reliably distinguish OSAS from primary snoring [14, 15]. Other symptoms during sleep include paradoxical breathing (in which the thoracic wall moves inward with respiration),

or obstructive hypopnea) per hour of sleep or (2) obstructive hypoventilation, manifested by $P_{ETCO_2} > 50$ mm Hg for $> 25\%$ of sleep time and/or arterial oxygen desaturation coupled with snoring, paradoxical thoracoabdominal movement, or flattening of the nasal airway pressure waveform [26]. Patients up to age 18 may be scored using the diagnostic criteria for children, but sleep specialists can choose to score children ≥ 13 years using adult criteria [25]. Particularly in very young children, central apnea may co-exist with OSAS. Careful scoring of the polysomnogram is needed to distinguish between central and obstructive events as the etiology and management of central apnea differs from OSAS. Several polysomnographic respiratory parameters of importance and their definitions are presented in Table 4. For further detail the reader is directed to the *AASM Manual for the Scoring of Sleep and Associated Events* [25].

There is no universally accepted classification for OSAS severity in children, although many studies use the apnea hypopnea index (AHI) to categorize OSAS as mild (AHI 1–4.9), moderate (AHI 5–9.9), or severe (AHI > 10) [10]. A cutoff of AHI > 30 to denote very severe OSAS has also been proposed [27]. Other objective findings of disease severity include presence and length of oxygen desaturations, degree of hypoventilation, sleep fragmentation, and decreased total sleep time. It is important to note that the severity of polysomnogram findings does not always correlate with the degree of morbidity and thus treatment decisions should take into account the child's symptoms and relevant comorbidities in addition to the objective polysomnogram findings.

It should be mentioned that there is controversy regarding indications for polysomnography in children. Guidelines published by the AASM, AAP, and European Respiratory Society (ERS) recommend polysomnography for all children when the clinical assessment is suggestive of OSAS (Table 1). In contrast, the American Academy of Otolaryngology—Head & Neck Surgery favors a more limited use of polysomnography and recommends routine use only in high risk populations (children with age < 2 , obesity, Down syndrome, craniofacial abnormalities, neuromuscular disorders, sickle cell disease, or mucopolysaccharidoses) or if the need for surgery is uncertain or there is discordance between the history of SDB and the tonsillar size on physical exam [28].

III. Screening tools

Due to the scarcity of accredited sleep laboratories and significant resources required to perform polysomnography, there has been considerable effort invested into the development of alternative screening or diagnostic methods which could be more widely available.

Questionnaires—Owing to their convenience, a reliable screening questionnaire to identify pediatric OSAS would have significant clinical utility. A number of clinical scoring instruments have been developed including the OSA score [29], pediatric sleep questionnaire (PSQ) [30], and OSA-18 [31], among others [32]. The diagnostic accuracy varies depending on the cutoff score and study population but is generally too low to be considered an alternative diagnostic method. While they cannot accurately identify OSAS, questionnaires do have clinical utility in assessing the impact of OSAS on quality of life. For example, the PSQ showed stronger correlation to behavior, sleepiness, and quality of life metrics than

polysomnogram parameters, leading the authors to suggest that PSQ score is a useful adjunct to polysomnography [33]. Other groups have developed predictive models using a combination of symptoms-based questions and physical exam findings or results from home sleep apnea testing (HSAT) [34, 35], but their utility in clinical practice has not been determined.

Home sleep apnea testing and pulse oximetry—HSAT is performed in the patient's home and monitors fewer physiologic variables than full polysomnography. In theory, HSAT could have significant benefits over laboratory-based polysomnography in terms of cost, resources, and disruption to family life. HSAT is an acceptable method for the diagnosis of OSAS in uncomplicated adult patients [36]; however, an AASM task force found insufficient evidence to support the use of HSAT in children [37]. Further validation studies are required to assess the feasibility and validity of these studies and determine their optimal role in pediatric OSAS.

Continuous pulse oximetry has also been studied as a screening or diagnostic tool when polysomnography is not available. Although intermittent oxygen desaturations on overnight continuous pulse oximetry is highly suggestive of OSAS [38], not all children with OSAS exhibit nocturnal hypoxemia [39] and therefore children with negative studies require follow-up polysomnography to rule out OSAS. A large prospective study recently evaluated the diagnostic utility of nocturnal oximetry to estimate the AHI of habitually snoring children [40]. An automatic analysis algorithm achieved an accuracy of 75.2, 81.7, and 90.2% in diagnosing children with AHI 1, 5, and 10 respectively, raising the possibility that overnight oximetry could play an important role in the diagnosis of pediatric OSAS especially in places where polysomnography is not available [40].

IV. Upper airway evaluation

Radiographic evaluation—Several radiological techniques are available to evaluate the upper airway including lateral neck radiographs, cephalometrics, fluoroscopy, computerized tomography (CT), and magnetic resonance imaging (MRI) [41–44]. These modalities are most often used in the research setting to study the pathophysiology of OSAS, though a small number of studies have explored their diagnostic potential with varying results [45]. In clinical practice, radiographic techniques are typically reserved for upper airway evaluation in children with comorbid conditions such as craniofacial anomalies or neurological conditions and may assist in identification of level(s) of obstruction and treatment selection. Conventional techniques for upper airway evaluation such as fluoroscopy, lateral neck cephalometry, and CT have substantial limitations as they evaluate neck anatomy in the awake state (and thus may not demonstrate dynamic airway collapse occurring in sleep) and expose children to radiation. In light of these limitations, dynamic MRI and drug-induced sleep endoscopy (DISE) have emerged as alternative methods of assessment.

Dynamic MRI—MRI is particularly powerful since it may be used to reconstruct the upper airway (including soft tissues and skeletal structure) [43, 46] in three dimensions and to evaluate upper airway dynamics [47, 48]. Dynamic MRI captures dynamic changes of the upper airway throughout the respiratory cycle and is typically used for children with

persistent OSAS following AT or for high risk children with multiple sites of upper airway obstruction [49]. Identified sites of obstruction may then be targeted surgically. Reports regarding MRI-directed surgical outcomes are sparse; however, a meta-analysis including 68 patients found that the most common procedure performed after dynamic MRI was lingual tonsillectomy (57%) and the mean AHI reduction was 7.4 events/hour [50]. At present dynamic MRI is not widely used and is only performed at a small number of specialized sleep centers.

Drug-induced sleep endoscopy (DISE)—In DISE, the upper airway is evaluated with a flexible endoscope while the child is in a pharmacologically-induced sleep-like state [51]. This method enables dynamic evaluation of the airway and identification of areas of obstruction which may become surgical targets. DISE-directed procedures include lingual tonsillectomy, supraglottoplasty, nasal surgery, revision adenoidectomy, uvulopalatopharyngoplasty, and partial midline glossectomy [52]. Despite its increasingly widespread use, studies evaluating outcomes after DISE-directed therapy are sparse. A small retrospective study found that approximately half of children undergoing DISE-directed surgery experienced resolution of OSAS; however, in the subgroup of children who underwent DISE for residual OSAS following AT, there was no improvement in obstructive AHI [52]. A small prospective study of 20 children reported a decrease in mean AHI from 6.1 to 1.9 following DISE-directed surgery [53]. Although use of DISE continues to grow, larger and more rigorous studies should be performed in order to evaluate outcomes associated with DISE-directed surgical management and identify subpopulations who benefit most from this approach.

MANAGEMENT

A proposed strategy for the management of children with OSAS is presented in Table 5.

I. Expert evaluation

The link between OSAS and cardiometabolic morbidity in adults is well established [54], and current evidence also supports this relationship in the pediatric population [6]. Accordingly, children with severe OSAS and those with pre-existing risk factors for cardiometabolic disease may require evaluation by a specialist. Children with severe OSAS are often referred to cardiology prior to surgery, as are children with overt cardiac manifestations (such as elevated blood pressure, or signs of pulmonary hypertension). Children who are obese may also benefit from an endocrinology evaluation.

II. Surgical management

Adenotonsillectomy—In healthy children over two years of age with adenotonsillar hypertrophy, first-line treatment of OSAS is AT [1, 10]. This recommendation is supported by the results of the Childhood Adenotonsillectomy Trial (CHAT), a large randomized controlled trial which found that surgical intervention with adenotonsillectomy was associated with significant improvements in behavior, quality of life, and polysomnographic findings as compared to watchful waiting. At seven months follow up, polysomnogram findings normalized in 79% of surgical patients compared to 49% in the watchful waiting

arm [7]. In a meta-analysis of three studies (including CHAT), Chinnadurai and colleagues reported a 4.8-point improvement in AHI for patients undergoing surgical intervention compared to watchful waiting [55].

Classically, tonsillectomy was performed using “cold” techniques to dissect the tonsillar capsule off the pharyngeal constrictor muscles. Alternatives to conventional cold dissection include electro-surgery (mono- or bi-polar) and plasma-mediated ablation (coblation) [56]. Partial tonsillectomy, a modified procedure in which the majority of tonsillar tissue is removed but the tonsillar capsule remains in place, is increasingly being utilized to treat OSAS in children [57]. Complication rates of partial tonsillectomy (also known as tonsillotomy and intracapsular tonsillectomy,) are lower compared to standard tonsillectomy; however, few studies have examined its therapeutic effect [57]. The subset of children with both obesity and asthma are more likely to experience resolution of OSAS following traditional tonsillectomy compared to partial tonsillectomy [58], but these findings should be evaluated in prospective trials. Immediately following tonsillectomy, children should be monitored for respiratory complications which range in severity from mild oxygen desaturation to postoperative pulmonary edema. Other complications include throat pain, bleeding, nausea/vomiting, dehydration, delayed feeding, velopharyngeal insufficiency, and rarely death [10]. Most tonsillectomies are performed as outpatient procedures although children with age < 3 or severe OSAS (AHI > 10 or oxygen saturation nadir < 80%) are at increased risk for complications and should be observed overnight [10].

AT is generally successful in improving respiratory parameters but is not curative in all cases. In a multicenter study of almost 600 children undergoing AT for OSAS, AHI decreased from 18.2 to 4.1, but complete resolution of OSAS was observed in only 27.2% of children [12]. Risk factors for persistence include age > 7, severe disease, chronic asthma, and obesity [12, 59].

Other upper airway surgeries—Surgical procedures beyond AT are typically reserved for children with comorbidities leading to airway obstruction at multiple levels or for children with persistent OSAS following AT. Potential causes of airway obstruction include turbinate hypertrophy, nasal septal deviation, redundant palatal tissue, large tongue base, retrognathia, regrowth of the adenoids, laryngomalacia, or lingual tonsil hypertrophy [60]. Obstruction at the level of the tongue base can be addressed using a number of procedures including lingual tonsillectomy and posterior midline glossectomy. Obstruction may also be addressed at the level of the palate (uvulopalatopharyngoplasty [61]), lateral pharyngeal walls (expansion sphincter pharyngoplasty [62]), nasal cavity (turbinate reduction [63]), or adenoids (revision adenoidectomy [64]).

When airway obstruction is severe due to craniofacial disorders restricting the upper airway, more extensive surgical approaches are used. In children with mandibular insufficiency as seen in Pierre-Robin Sequence, Treacher-Collins Syndrome, and other causes of congenital retrognathia, mandibular advancement or mandibular distraction osteogenesis can effectively alleviate upper airway obstruction and avoid tracheostomy dependence [65, 66]. Genioglossus advancement and hyoid suspension have also been used to treat OSAS, though the body of evidence is small and most studies involve adults [67, 68].

Tracheotomy—Tracheotomy is the definitive surgical treatment for OSAS. The majority of patients who require tracheotomy have either severe craniofacial abnormalities or neuromuscular conditions causing hypotonia [69]. Although many children remain tracheostomy-dependent, the procedure can also be used as a temporary measure to manage severe OSAS while awaiting other surgical treatment.

III. Non-surgical management

Positive airway pressure (PAP)—Children with small tonsils/adenoids and those who are not surgical candidates or whose parents decline surgery should be managed with PAP therapy during sleep. PAP therapy maintains airway patency by serving as a pneumatic splint for the soft tissues of the upper airway leading to increased cross-sectional airway area throughout the respiratory cycle. Initiation of PAP therapy begins with an overnight sleep study with PAP titration, ideally in an accredited sleep laboratory or in a hospital where an expert technician or respiratory therapist can monitor respiratory and sleep parameters. Pressure adjustments are performed with the goal of eliminating respiratory events and correcting gas exchange abnormalities while also maintaining patient tolerability. Technical details regarding PAP titration may be found elsewhere [70]. Continuous PAP (CPAP) and bilevel PAP (BPAP) are equally effective, although CPAP is used most frequently. PAP therapy in children improves both respiratory parameters and neurobehavioral sequelae of OSAS such as attention span, hyperactivity, daytime sleepiness, and school performance [71, 72]. Studies in adults have demonstrated improvement in blood pressure and cognitive function [73, 74], but data regarding the corollary outcomes in children is limited.

Although PAP therapy is an effective treatment modality for children of all ages and with a range of underlying pathophysiological mechanisms, effectiveness is limited by poor adherence in some studies. In a prospective study on PAP compliance in children, Marcus and colleagues noted a high dropout rate (35%), low average nightly use (5.3 hours/night), and frequent overreporting of adherence by parents as compared to objective compliance data [71]. Discomfort associated with the face mask is frequently reported and likely contributes to poor adherence. Compliance can be significantly improved by close clinical follow up in addition to intensive education [75] and gradual introduction of the PAP mask [76]. Side effects include nasal congestion, oronasal dryness, epistaxis, eye irritation from air leak, facial pain, and skin abrasion [77]. Though rare, skin necrosis and ulceration have been reported [78]. Long-term CPAP use has also been linked to midface hypoplasia [79] and midfacial retrusion over time [80]. These skeletal changes appear to be common, as global facial flattening and maxillary retrusion can be observed in 68% and 37% of patients, respectively [81]. Accordingly, routine follow up with special attention to midface growth is essential in children when long term PAP therapy is anticipated.

Medical therapy—Medical therapies have been proposed as an alternative to surgery, especially in children with mild OSAS. A six-week course of once-daily intranasal budesonide has been shown to improve quality of life and respiratory parameters measured on polysomnography with a sustained effect two months after discontinuation [82, 83], and a recent meta-analysis found that montelukast improved the AHI by 55% when used alone and by 70% when used in conjunction with intranasal corticosteroids in children with mild

OSAS [84]. These therapies hold promise as potential treatments for mild uncomplicated OSAS in children. Of note, obese children and those with age > 7 are less likely to benefit from anti-inflammatory therapy [85].

Dental procedures—Rapid maxillary expansion is an orthodontic procedure which expands the airway by increasing the width of the hard palate using a dental device secured over the maxillary teeth and an accompanying expansion screw. Two studies have shown normalization of polysomnogram parameters and symptoms using this technique in children with maxillary restriction and dental malocclusion [86, 87]. Oral appliances including tongue devices and mandibular advancement devices move the tongue and mandible forward and away from posterior pharynx to improve upper airway patency. In a small group of children with mild OSAS and dysgnathia, oral jaw-positioning appliances were shown to reduce AHI, improve sleep, and alleviate daytime sleepiness [88]. A recent Cochrane review of randomized studies for oral appliances in pediatric OSAS identified only this one study and concluded there is insufficient evidence to support or refute their utility [89].

SPECIAL CONSIDERATIONS

Certain children with OSAS present unique challenges to treatment and their management warrants further discussion.

I. Residual OSAS after adenotonsillectomy (AT)

Not all children experience resolution of OSAS following AT and some (including those with chromosomal, craniofacial, or neuromuscular disorders) are at increased risk of persistence [12, 59, 90]. Residual OSAS may be identified by repeat polysomnogram (which is recommended for those with risk factors) or by recurrence of clinical symptoms. In the latter case, the diagnosis should be confirmed with polysomnography. Options for the management of persistent OSAS include airway evaluation with possible surgical intervention, PAP, and medical therapy. DISE or dynamic MRI can be used to evaluate the upper airway and identify sites of persistent obstruction which can be targeted surgically, as discussed above. PAP is effective in this setting, although difficulties with tolerability and poor adherence still apply [72]. A combination of montelukast and intranasal budesonide was also shown to improve AHI and oxygen nadir in children with mild OSAS following AT [83]. Lastly, weight management should be recommended for children who are overweight or obese (see next section).

II. Children with complex comorbidities

Obesity—Obesity is reaching epidemic proportions across all age groups, including children, and increases the risk for OSAS with an odds ratio of 4.5 [91]. Some studies have reported the prevalence of OSAS to be as high as 45% in obese children [92]. The cause of this association is not fully elucidated, but adenotonsillar hypertrophy, increased airway collapsibility, alterations in body fat composition, and abnormal ventilatory response likely each play a role [4]. If adenotonsillar hypertrophy is present, AT is first-line treatment and significantly improves OSAS symptoms and polysomnogram parameters; however, persistent OSAS is identified in 33–76% of obese children compared to 15–37% of non-

obese children [93]. While the reason for high failure rates is unclear, residual adenoid tissue and increased soft palate volume have been observed in obese children following AT and may contribute to persistent airway obstruction [94]. PAP is a good treatment alternative for children without adenotonsillar hypertrophy or those with residual OSAS following AT. A small number of studies have explored the hypothesis that weight loss may improve OSAS in obese children. Andersen and colleagues prospectively enrolled 62 overweight or obese children with AHI > 2 in an individualized, family-centered obesity treatment protocol. After one year, the AHI normalized in 44% of children and the change in BMI standard deviation score was associated with the decrease in AHI; however, most of the children (71%) had mild OSAS at baseline [95]. Although the effect of weight loss on OSAS outcomes is not well studied, weight management is routinely recommended. A decrease in BMI is associated with improved metabolic outcomes in obese children and should be routinely encouraged notwithstanding its potential benefits regarding OSAS [96]. Lastly, further surgical management may be considered although studies reporting outcomes of secondary surgery such as uvulopalatopharyngoplasty or supraglottoplasty in this population are sparse and have so far demonstrated poor efficacy [97].

Down syndrome—The prevalence of OSAS among children with Down syndrome may be as high as 70% and almost half have severe disease [98]. Accordingly, the AAP recommends routine screening for SDB/OSAS in children with Down syndrome [99]. The pathophysiology of OSAS in this population is multifactorial and includes hypoplasia of the midface/mandible, relative macroglossia, lingual tonsil hypertrophy, comorbid laryngomalacia, obesity, decreased airway tone, and hypopharyngeal collapse [100–102]. Recognition and treatment of OSAS is particularly important as the consequences of untreated OSAS may exacerbate underlying cardiac disease, behavioral problems, and cognitive challenges [103–105]. The pathophysiology, morbidity, and treatment of OSAS in patients with Down syndrome was reviewed recently and highlights the need for further research regarding treatment options and outcomes in this high-risk population [106]. AT is first-line treatment but has a low success rate of approximately 10–20% [24, 107]. Residual OSAS may be treated with PAP or further surgery. DISE-directed therapy resulted in improvements in polysomnogram parameters in a small cohort of children with Down syndrome, but the study was not powered to detect statistically significant differences [108].

Craniofacial disorders—Children with disorders associated with midfacial hypoplasia and/or mandibular deficiency deserve special attention in their evaluation and management. We discuss here Pierre-Robin sequence; however, a similar approach may be applied to similar disorders (see Tables 2 and 5). Pierre-Robin sequence is characterized by a triad of micrognathia, glossoptosis, and upper airway obstruction with or without cleft palate and can occur as an isolated finding or in the context of other congenital abnormalities or syndromes. The presence of micrognathia with glossoptosis significantly increases the risk of developing OSAS and studies have reported a high prevalence (46–83%) of OSAS [109]. The severity of upper airway obstruction and presentation of SDB varies widely; some children experience respiratory distress requiring intubation at birth, while others present later in life with poor weight gain [110]. Initial management with lateral or prone positioning and placement of a nasopharyngeal airway relieves the obstruction in some

children [111]. A trial of PAP therapy is warranted in those requiring further respiratory support. If surgical management is required, tongue-lip adhesion or mandibular distraction osteogenesis have been shown to improve the AHI and relieve upper airway obstruction [111]. Because upper airway dimensions can improve over time [112], PAP therapy is a good alternative to consider prior to mandibular surgeries.

Neurologic Disorders—Children with neurologic disorders can be affected by a number of sleep abnormalities including OSAS, nocturnal obstructive hypoventilation, and diurnal hypoventilation. Predisposing factors to OSAS include reduced ventilatory responses, reduced activity of respiratory muscles during sleep, reduced upper airway dilator tone, and poor lung mechanics due to the underlying neuromuscular disorder [113]. Depending on the severity of disease, PAP therapy can be accomplished with CPAP, BPAP, or invasive ventilation and tracheotomy may be required in severe cases. For further discussion of sleep in children with neurologic disorders the reader is directed elsewhere [113].

CONCLUSION

In the coming years, pediatric OSAS will continue to burden our society. Improved methodologies to better diagnose and manage children with this disorder will continue to be developed as we improve our knowledge of its pathophysiology and the mechanisms leading to its adverse outcomes.

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TABLE 1:**Evaluation of the Child with Suspected OSAS or Residual OSAS following AT**

<p>Otherwise healthy children</p> <hr/> <p>Clinical evaluation: history and physical exam (recommended by AAP [1], AASM [114], AAO-HNS [10], and ERS [115]) Diagnostic polysomnogram (recommended by AAP [1], AASM [26, 114], and ERS [115])</p> <ul style="list-style-type: none"> • AAO-HNS recommends polysomnography if the need for surgery is unclear or if there is discordance between the clinical history and the tonsillar size on physical exam [28] <p>Alternative testing when polysomnography is not available</p> <ul style="list-style-type: none"> • AAP: options include nocturnal video recording, nocturnal oximetry, daytime nap polysomnography, or ambulatory polysomnography [1] • ERS: options include ambulatory polysomnography or polygraphy, nocturnal oximetry, Pediatric Sleep Questionnaire, or Sleep Clinical Record [115] • AASM: does not recommend home sleep apnea testing for OSAS diagnosis in children [37] • AAO-HNS: home-based studies may be considered but are not recommended for routine use [28] <p>Consider endocrinology/weight management referral for obese children Consider cardiology referral for children with severe sleep apnea or cardiometabolic risk factors</p> <hr/> <p>Children with complex comorbidities</p> <hr/> <p><i>Includes children with obesity, neuromuscular disorders, craniofacial abnormalities, genetic syndromes, sickle cell disease, etc.</i> Clinical evaluation: history & physical exam (recommended by AAP [1], AASM [114], AAO-HNS [10], and ERS [115]) Diagnostic polysomnogram (recommended by AAP [1], AASM [114], AAO-HNS [10], and ERS [115]) Expert evaluation (e.g. pediatric pulmonology, pediatric otolaryngology, sleep medicine, craniofacial team) Consider endocrinology/weight management referral for obese children Consider cardiology referral for children with severe sleep apnea or cardiometabolic risk factors</p> <hr/> <p>Children with residual OSAS following AT</p> <hr/> <p>Post-treatment reevaluation of high-risk patients</p> <ul style="list-style-type: none"> • AAP: High-risk children (defined as those with significantly abnormal baseline polysomnogram, sequelae of OSAS, obesity, or continued symptoms following treatment) should be reevaluated with objective testing or referred to a sleep specialist [1] • AASM: Polysomnography should be repeated following adenotonsillectomy in children at risk of recurrence: mild preoperative OSAS with residual symptoms following surgery, pre-operative evidence of moderate-severe OSAS, obesity, craniofacial abnormalities, or neurologic disorders [114] • ERS: Patients at risk of persistent OSAS or with persistent symptoms should be reevaluated with polysomnography or polygraphy [115] <p>Expert evaluation (pediatric pulmonology, sleep medicine, pediatric otolaryngology, endocrinology) Polysomnography titration for positive airway pressure therapy (AAP [1]) Nasopharyngoscopy or drug-induced sleep endoscopy Imaging (computed tomography, dynamic magnetic resonance imaging) Cardiology referral for persistent, untreated OSAS</p> <hr/>
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AAO-HNS American Academy of Otolaryngology—Head & Neck Surgery, *AAP* American Academy of Pediatrics, *AASM* American Academy of Sleep Medicine, *ERS* European Respiratory Society, *OSAS* obstructive sleep apnea syndrome

TABLE 2:**Medical Conditions Associated with OSAS****Craniofacial Anomalies:**

Achondroplasia
 Apert syndrome
 Beckwith-Wiedemann
 Choanal stenosis
 Crouzon syndrome
 Down syndrome
 Goldenhar syndrome
 Hallerman-Streiff syndrome
 Klippel-Feil syndrome
 Laryngomalacia
 Marfan syndrome
 Micrognathia
 Midface hypoplasia
 Nager syndrome
 Palate formation (e.g. high and narrow hard palate)
 Pfeiffer syndrome
 Retrognathia
 Robin sequence
 Rubinstein-Taybi
 Stickler syndrome
 Subglottic stenosis
 Treacher-Collins syndrome

Neurological disorders:

Arnold-Chiari malformation
 Cerebral palsy
 Möbius syndrome
 Muscular dystrophy
 Myasthenia gravis
 Myelomeningocele
 Myotonic dystrophy
 Poliomyelitis
 Spinal muscular atrophy
 Syringobulbia
 Syringomyelia

Miscellaneous disorders:

Airway papillomatosis
 Congenital hypothyroidism
 Face and neck burns
 Family history of obstructive sleep apnea syndrome

History of prematurity

Mucopolysaccharidoses (e.g. Hurler, Hunter)

Obesity

Polycystic ovary syndrome

Prader Willi syndrome

Sickle cell disease

Postoperative Disorders:

Post adenotonsillectomy leading to naso- and/or oro-pharyngeal stenosis

Post pharyngeal flap: leading to naso- and/or oro-pharyngeal stenosis

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TABLE 3:

Diagnostic Criteria for Pediatric OSAS Based on AASM *International Classification of Sleep Disorders*, 3rd ed.[26] (Diagnosis Requires A + B)

A. The presence of one or more of the following:

1. Snoring
2. Labored, paradoxical, or obstructed breathing during the child's sleep
3. Sleepiness, hyperactivity, behavioral problems, or learning problems

B. Polysomnography demonstrates one or more of the following:

1. One or more obstructive or mixed apneas, or obstructive hypopneas, per hour of sleep
 2. A pattern of obstructive hypoventilation, defined as at least 25% of total sleep time with hypercapnia ($P_{ETCO_2}^* > 50$ mm Hg) and/or arterial oxygen desaturation in association with one or more of the following: snoring, flattening of the inspiratory nasal pressure waveform, and paradoxical thoracoabdominal motion.
-

* P_{ETCO_2} , end-tidal CO_2 .

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TABLE 4:

Pediatric Respiratory Events Derived from Polysomnography (modified from The *AASM Manual for the Scoring of Sleep and Associated Events*) [25]

-
- **Obstructive apnea:** At least a 90% drop in peak signal excursion of the oronasal thermal sensor in the presence of continued respiratory effort lasting longer than 2 respiratory cycle times.
 - **Central apnea:** At least a 90% drop in peak signal excursion of the oronasal thermal sensor in the absence of respiratory effort and at least one of the following: (1) the event lasts 20 seconds; (2) the event lasts at least 2 respiratory cycles and is associated with 3% oxygen desaturation; or (3) the central event lasts at least 2 respiratory cycle times and is associated with heart deceleration.
 - **Mixed apnea:** Criteria for apnea need to be met for at least 2 respiratory cycles and associated with absent respiratory effort during one portion of the event and inspiratory effort in another portion.
 - **Hypopnea:** All the following three criteria need to be met: (1) at least 30% decrease in the amplitude of the nasal pressure signal; (2) duration 2 respiratory cycles times; and (3) 3% oxygen desaturation or the event is associated with arousal.
 - **Obstructive hypopnea:** To score hypopnea as obstructive, the following need to be met: (1) snoring during the event; (2) increased inspiratory flattening of the nasal pressure flow signal compared with baseline breathing; and (3) associated thoracoabdominal paradox occurs during the event but not during pre-event breathing.
 - **Hypoventilation:** Criteria are met when end-tidal CO₂ (P_{ETCO₂}) >50 mmHg persists for more than 25% of total sleep time.
 - **Apnea index (AI):** Number of obstructive and/or central apneic events per hour of sleep.
 - **Obstructive apnea index (oAI):** Number of obstructive apneic events per hour of sleep.
 - **Hypopnea index (HI):** Number of hypopneas per hour of sleep.
 - **Apnea/hypopnea index (AHI):** The summation of apnea index and hypopnea index.
 - **Obstructive apnea/hypopnea index (oAHI):** The summation of obstructive apneic events and hypopneic events per hour of sleep.
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TABLE 5:

Management of the Child with OSAS

Otherwise healthy children		
Mild OSAS	Moderate OSAS	Severe OSAS
<ul style="list-style-type: none"> • AT vs watchful waiting for 6 months • Intranasal corticosteroid • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • Specialist referral (e.g. sleep medicine, otolaryngology) • AT • PAP therapy • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • Specialist referral (e.g. sleep medicine, otolaryngology) • Cardiology evaluation • AT with overnight observation • PAP therapy • Weight management, if overweight/obese

Children with complex comorbidities		
Mild OSAS	Moderate OSAS	Severe OSAS
<ul style="list-style-type: none"> • Specialist referral (e.g. sleep medicine, otolaryngology) • AT • Intranasal corticosteroid • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • Specialist referral (e.g. sleep medicine, otolaryngology) • AT with overnight observation • Other airway and/or craniofacial surgery • PAP therapy • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • Specialist referral (e.g. sleep medicine, otolaryngology, craniofacial team) • Cardiology evaluation • AT with overnight observation • Other airway and/or craniofacial surgery • PAP therapy • Weight management, if overweight/obese

Children with residual OSAS following AT		
Mild OSAS	Moderate OSAS	Severe OSAS
<ul style="list-style-type: none"> • Intranasal corticosteroid • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • PAP therapy • Weight management, if overweight/obese 	<ul style="list-style-type: none"> • PAP therapy • Weight management, if overweight/obese • Secondary surgical intervention • Tracheostomy

AT, adenotonsillectomy

OSAS, obstructive sleep apnea syndrome

PAP, positive airway pressure

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