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Clinical Practice Guideline: Diagnosis and Management of Childhood Obstructive Sleep Apnea Syndrome

Section on Pediatric Pulmonology and Subcommittee on Obstructive Sleep Apnea Syndrome

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Section on Pediatric Pulmonology, Subcommittee on Obstructive Sleep Apnea Syndrome

Clinical Practice Guideline: Diagnosis and Management of Childhood Obstructive Sleep Apnea Syndrome

ABSTRACT. This clinical practice guideline, intended for use by primary care clinicians, provides recommendations for the diagnosis and management of obstructive sleep apnea syndrome (OSAS).

The Section on Pediatric Pulmonology of the American Academy of Pediatrics selected a subcommittee composed of pediatricians and other experts in the fields of pulmonology and otolaryngology as well as experts from epidemiology and pediatric practice to develop an evidence base of literature on this topic. The resulting evidence report was used to formulate recommendations for the diagnosis and management of childhood OSAS.

The guideline contains the following recommendations for the diagnosis of OSAS: 1) all children should be screened for snoring; 2) complex high-risk patients should be referred to a specialist; 3) patients with cardiorespiratory failure cannot await elective evaluation; 4) diagnostic evaluation is useful in discriminating between primary snoring and OSAS, the gold standard being polysomnography; 5) adenotonsillectomy is the first line of treatment for most children, and continuous positive airway pressure is an option for those who are not candidates for surgery or do not respond to surgery; 6) high-risk patients should be monitored as inpatients postoperatively; 7) patients should be reevaluated postoperatively to determine whether additional treatment is required.

This clinical practice guideline is not intended as a sole source of guidance in the evaluation of children with OSAS. Rather, it is designed to assist primary care clinicians by providing a framework for diagnostic decision-making. It is not intended to replace clinical judgment or to establish a protocol for all children with this condition and may not provide the only appropriate approach to this problem. Pediatrics 2002;109:704–712; obstructive sleep apnea, infant, child, adenoidectomy, tonsillectomy, meta-analysis, polysomnography, sleep disorders, snoring.

ABBREVIATIONS. OSAS, obstructive sleep apnea syndrome; PS, primary snoring; REM, rapid eye movement; CPAP, continuous positive airway pressure; PPV, positive predictive value; NPV, negative predictive value.

INTRODUCTION

bstructive sleep apnea syndrome (OSAS) is a common condition in childhood and can result in severe complications if left untreated. Nevertheless, there is no consensus on the best methods of evaluation and management of this syndrome in children. Therefore, the American Academy of

The recommendations in this statement do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

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Pediatrics has supported the development of a practice guideline for the diagnosis and management of childhood OSAS.

The purpose of this clinical practice guideline is to 1) increase the recognition of OSAS by pediatricians to decrease diagnostic delay and avoid serious sequelae of OSAS; 2) evaluate diagnostic techniques; 3) describe treatment options; 4) provide guidelines for follow-up; and 5) discuss areas requiring additional research.

This practice guideline focuses on uncomplicated childhood OSAS, that is, the otherwise healthy child with OSAS associated with adenotonsillar hypertrophy and/or obesity who is being treated in the primary care setting. This guideline specifically excludes infants younger than 1 year, patients with central apnea or hypoventilation syndromes, and patients with OSAS associated with other medical disorders, including but not limited to Down syndrome, craniofacial anomalies, neuromuscular disease (including cerebral palsy), chronic lung disease, sickle cell disease, metabolic disease, or laryngomalacia. These important patient populations are too complex to discuss within the scope of this paper and require specialist consultation. In addition, patients with lifethreatening OSAS who present in cardiorespiratory failure will not be covered here, because these patients require urgent treatment.

METHODS OF GUIDELINE DEVELOPMENT

Details of the methods of guideline development are included in the accompanying technical report published online.1 Committee members signed forms confirming that they did not have a conflict of interest. The guidelines were based on data available from the medical literature. A computerized search of the National Library of Medicine's PubMed database (http://www.ncbi.nlm.nih.gov/entrez/query. fcgi?db=PubMed) from 1966-1999 (later updated to include 2000) was performed using the following keywords: sleep apnea syndrome, apnea, sleep disorders, snoring, polysomnography, airway obstruction, adenoidectomy, tonsillectomy (adverse effects, mortality), and sleep-disordered breathing. The search was limited to articles involving children. Studies involving infants, animal studies, and articles written in languages other than English were excluded. Reviews, case reports, letters to the editor, and abstracts were not included. A total of 2110 articles were found. Committee members then screened the articles, first by title and then by abstract, to obtain articles relevant to the guideline. After screening, a total of 278 articles were reviewed in full by committee members. An additional 6 articles, primarily from foreign publications, could not be obtained from local libraries. None of these were considered particularly germane to the guideline. In addition to the literature search, committee members supplemented the articles with additional publications thought to be relevant and with those published after 1999. Details of the literature grading system are available in the accompanying technical report published online. Review of the literature revealed that there were very few randomized controlled studies. When the evidence was poor or lacking, there was extensive discussion among committee members to achieve consensus. The guideline notes whether a decision was based on objective evidence or on consensus decision.

DEFINITION

OSAS in children is a "disorder of breathing during sleep characterized by prolonged partial upper airway obstruction and/or intermittent complete obstruction (obstructive apnea) that disrupts normal ventilation during sleep and normal sleep patterns."2 Symptoms include habitual (nightly) snoring (often with intermittent pauses, snorts, or gasps), disturbed sleep, and daytime neurobehavioral problems. Daytime sleepiness may occur but is uncommon in young children. Complications include neurocognitive impairment, behavioral problems, failure to thrive, and cor pulmonale, particularly in severe cases. Risk factors include adenotonsillar hypertrophy, obesity, craniofacial anomalies, and neuromuscular disorders. Only the first 2 risk factors are discussed in this guideline.

OSAS needs to be distinguished from primary snoring (PS), which is defined as snoring without obstructive apnea, frequent arousals from sleep, or gas exchange abnormalities.³ Although PS is usually considered benign, this has not been well evaluated, because most studies of snoring children did not discriminate between PS and OSAS.

PREVALENCE

OSAS occurs in children of all ages, from neonates to adolescents. It is thought to be most common in preschool-aged children, which is the age when the tonsils and adenoids are the largest in relation to the underlying airway size.4 Three studies have evaluated the prevalence of childhood OSAS. These studies did not use conventional polysomnography, used adult rather than pediatric polysomnographic criteria, or studied only a selected high-risk sample of the population; thus, a definitive epidemiologic study has not yet been performed. Despite these limitations, the 3 studies showed similar prevalence rates of approximately 2%.5-7 In contrast, PS is more common; habitual snoring occurs in 3% to 12% of preschool-aged children. 5,6,8-10 Thus, the clinician needs a method to distinguish OSAS from PS. OSAS occurs equally among boys and girls.7 One study indicated that the prevalence is higher among African American individuals than among white individuals.⁷

SEQUELAE OF OSAS

Untreated OSAS can result in serious morbidity. Early reports documented such complications as failure to thrive, cor pulmonale, and mental retardation.¹¹ These severe sequelae appear to be less common now, probably because of earlier diagnosis and treatment. Although failure to thrive is the exception these days, children with OSAS still tend to have a growth spurt after adenotonsillectomy. $^{12-14}$ In the past, cor pulmonale with heart failure was not an uncommon mode of presentation for OSAS in children, but it is now rare. Although overt right heart failure now occurs less often, asymptomatic degrees of pulmonary hypertension may be common. 15 Systemic hypertension can occur. 16-18 Many reports have suggested that children with OSAS are at risk of neurocognitive deficits, such as poor learning, behavioral problems, and attention-deficit/hyperactivity disorder. 19-22 However, many of these studies were case series based on histories obtained from parents of snoring children without objective evaluation, control groups, or sleep studies to distinguish PS from OSAS. One recent study showed that children with sleep-disordered breathing were more likely to do poorly at school, and many improved after adenotonsillectomy.²³ If untreated, OSAS may result in death. Early OSAS literature described children who presented with cardiorespiratory failure or coma, some of whom died.24-26

METHODS OF DIAGNOSIS

Diagnostic methods that have been scientifically evaluated include history and physical examination, audiotaping or videotaping, pulse oximetry, abbreviated polysomnography, and full polysomnography. The goals of diagnosis are to 1) identify patients who are at risk for adverse outcomes; 2) avoid unnecessary intervention in patients who are not at risk for adverse outcomes; and 3) evaluate which patients are at increased risk of complications resulting from adenotonsillectomy so that appropriate precautions can be taken.

History and Physical Examination

A sleep history screening for snoring should be part of routine health care visits. In children, OSAS is very unlikely in the absence of habitual snoring. If a history of nightly snoring is elicited, a more detailed history regarding labored breathing during sleep, observed apnea, restless sleep, diaphoresis, enuresis, cyanosis, excessive daytime sleepiness, and behavior or learning problems (including attention-deficit/hyperactivity disorder) should be obtained. Findings on physical examination during wakefulness are often normal. There may be nonspecific findings related to adenotonsillar hypertrophy, such as mouth breathing, nasal obstruction during wakefulness, adenoidal facies, and hyponasal speech. Evidence of complications of OSAS may be present. These include systemic hypertension, an increased pulmonic component of the second heart sound indicating pulmonary hypertension, and poor growth (although conversely, some children with OSAS are obese).

Although history and physical examination are useful to screen patients and determine which patients need additional investigation for OSAS, there is controversy about their roles in determining which patients require treatment. A number of studies have shown that there is no relation between the size of the tonsils and adenoids and presence of OSAS.^{27–30} This is because OSAS is thought to be attributable to a combination of adenotonsillar hypertrophy and the neuromuscular tone of the upper airway during sleep rather than to structural abnormalities alone. Thus, the presence of large tonsils and adenoids does not necessarily indicate that the patient has OSAS.

An accurate diagnosis is required not only to ensure that appropriate treatment is provided and to avoid unnecessary treatment, but also to determine which children are at risk of complications resulting from treatment. Several studies have objectively evaluated the utility of a standardized history alone; history and physical examination; or history, physical examination, and audiotaping or videotaping to diagnose OSAS. In 1984, a study evaluated the efficacy of a questionnaire-derived OSAS score.31 The questionnaire was administered first to patients with polysomnographically proven OSAS and controls without OSAS and then prospectively to snoring patients being evaluated for suspected OSAS. The score was able to distinguish between patients with known OSAS and controls. However, three quarters of subjects had an indeterminate score. A more recent study by the same authors with a much larger sample found that the score had a sensitivity of 35% and specificity of 39%.³² A number of other studies have shown that this score has limited utility when applied to snoring children being evaluated for OSAS^{33,34} or when applied to obese patients.^{35,36} Thus, this questionnaire has minimal usefulness in the evaluation of OSAS.

Other studies have evaluated the utility of history and physical examination in distinguishing children with PS from those with OSAS.^{33,34,37–41} None of these studies were able to reliably discriminate between OSAS and PS.

There are a number of reasons why the history can be misleading. The loudness of snoring does not necessarily correlate with the degree of obstructive apnea. Thus, children may have very noticeable snoring without apnea. Children with OSAS experience obstruction primarily during rapid eye movement (REM) sleep, which occurs predominantly in the early morning hours when their parents are not observing them,⁴² thus leading to an underestimation of apnea. Some children have a pattern of persistent partial upper airway obstruction associated with gas exchange abnormalities, rather than discrete, cyclic apneas ("obstructive hypoventilation"²). These children will not manifest pauses and gasps in their snoring, and therefore, the condition may be misdiagnosed as PS.

Nocturnal Polysomnography

Nocturnal polysomnography (sleep study) is the only diagnostic technique shown to quantitate the ventilatory and sleep abnormalities associated with sleep-disordered breathing and is currently the gold standard. Polysomnography can be performed satisfactorily in children of any age, providing that appropriate equipment and trained staff are used. Furthermore, pediatric studies should be scored and interpreted using age-appropriate criteria as outlined in the American Thoracic Society consensus statement on pediatric polysomnography.² Polysomnography, by definition, can distinguish PS from OSAS. It can objectively determine the severity of OSAS and related gas exchange and sleep disturbances. As such, it can help determine the risk of postoperative complications (Table 1). However, although it is generally believed that children with severely abnormal results of sleep studies are at increased risk for complications of OSAS, formal studies have not been performed to evaluate the correlation between polysomnographic parameters and adverse outcomes in children with OSAS.43 Thus, although we know which polysomnographic parameters are statistically abnormal,44 studies have not definitively evaluated which polysomnographic criteria predict morbidity. In addition, there is currently a shortage of facilities that perform pediatric polysomnography. The availability of pediatric polysomnography is expected to improve, especially with the computerized equipment currently available.

Audiotaping or Videotaping

Two studies have examined the use of audiotaping, ^{33,41} and 1 study has examined the use of videotaping, ⁴⁵ alone or combined with clinical findings, in establishing a diagnosis. In these studies, sensitivity ranged from 71% to 94%, and specificity ranged from 29% to 80%. Positive predictive values (PPVs) were 50% ⁴¹ and 75% ³³ for audiotaping and 83% for videotaping. ⁴⁵ Sounds of struggle on audiotapes were found to be more predictive of OSAS than were pauses. ³³ The negative predictive value (NPV) ranged from 73% to 88%. Although these techniques may have promise, the discrepancies in results from different centers indicate that additional study is necessary.

Abbreviated Polysomnography

Several studies have evaluated abbreviated polysomnographic techniques. Overnight oximetry can be useful if it shows a pattern of cyclic desaturation. Brouillette et al³² performed oximetry in a group of

TABLE 1. Risk Factors for Postoperative Respiratory Complications in Children With OSAS Undergoing Adenotonsillectomy

Age younger than 3 years
Severe OSAS on polysomnography
Cardiac complications of OSAS (eg, right ventricular hypertrophy)
Failure to thrive
Obesity
Prematurity
Recent respiratory infection
Craniofacial anomalies*
Neuromuscular disorders*

^{*} Not discussed in these guidelines.

children with suspected OSAS and compared it with simultaneous full polysomnography. Patients with complex medical conditions were excluded. Compared with polysomnography, they found a PPV of 97% and an NPV of 47%, indicating that oximetry was useful when results were positive. However, patients with negative results of oximetry required full polysomnography for definitive diagnosis. False-positive results were found in patients with mild coexistent medical problems, such as obesity and asthma, suggesting that this technique is useful only in otherwise healthy children.

Nap polysomnography is appealing, because it can be performed in the daytime and is, therefore, more convenient for patients and laboratory staff. Studies have shown a PPV of 77% to 100% and an NPV of 17% to 49%. 46,47 In children with OSAS, overnight polysomnograms demonstrate more severe abnormalities than do nap studies. Thus, nap polysomnography may be useful if results are positive, although it may underestimate the severity of OSAS. An overnight study should be performed if the results of the nap study are negative. The difference in predictive value between nap and overnight studies is probably attributable to the decreased amount of REM sleep during nap studies as well as the decreased total sleep time.

Unattended home polysomnography in children has been evaluated by only 1 center. Home polysomnography yielded similar results to laboratory studies. However, it should be noted that the equipment used in this study was relatively sophisticated and included respiratory inductive plethysmography (a method for determining ventilation without using oronasal sensors), oximeter pulse wave form, and videotaping. The utility of unattended home studies in children using commercially available 4- to 6-channel recording equipment has not been studied.

Summary of Diagnostic Techniques

In summary, history and physical examination are poor at predicting OSAS. Most studies have shown that abbreviated or screening techniques, such as videotaping, nocturnal pulse oximetry, and daytime nap polysomnography tend to be helpful if results are positive but have a poor predictive value if results are negative. Thus, children with negative study results should undergo a more comprehensive evaluation. The cost efficacy of these screening techniques is unclear and would depend, in part, on how many patients eventually required full polysomnography. In addition, the use of these techniques in evaluating the severity of OSAS (which is important in determining management, such as whether outpatient surgery should be performed) has not been evaluated.

TREATMENT OPTIONS

Tonsillectomy and Adenoidectomy

Adenotonsillectomy is the most common treatment for children with OSAS. Adenoidectomy alone may not be sufficient.^{38,49} In otherwise healthy chil-

dren with adenotonsillar hypertrophy, polysomnographic resolution occurs in 75% to $100\%^{37,49-51}$ after adenotonsillectomy; this is associated with symptom resolution. Although obese children may have less satisfactory results, many will be adequately treated with adenotonsillectomy, and it is generally the first-line therapy for these patients.

Potential complications of adenotonsillectomy include anesthetic complications; immediate postoperative problems, such as pain and poor oral intake; and hemorrhage. In addition, patients with OSAS may develop respiratory complications, such as worsening of OSAS or pulmonary edema, in the immediate postoperative period. Death attributable to respiratory complications in the immediate postoperative period has been reported in patients with severe OSAS. Identified risk factors are shown in Table 1.^{53–58} High-risk patients should be hospitalized overnight after surgery and monitored continuously with pulse oximetry.

Continuous Positive Airway Pressure (CPAP)

For patients with specific surgical contraindications, minimal adenotonsillar tissue, or persistent OSAS after adenotonsillectomy or for those who prefer nonsurgical alternatives, CPAP therapy is an option.^{59–61} However, unlike adenotonsillectomy, which is a 1-time procedure that is usually curative, CPAP will need to be used indefinitely. CPAP is delivered using an electronic device that delivers constant air pressure via a nasal mask, leading to mechanical stenting of the airway and improved functional residual capacity in the lungs. The pressure requirement varies among individuals; thus, CPAP must be titrated in the sleep laboratory before prescribing the device and periodically readjusted thereafter. 59 CPAP is a long-term therapy and requires frequent clinician assessment of adherence and efficacy. It is generally tolerated in older children. 59,60 Young children or older children with learning or behavioral problems may require behavioral or desensitization techniques to accept this form of therapy.⁶² Attention to compliance with this therapy is crucial.

Other Treatment Modalities

Most adjunctive measures in the treatment of childhood OSAS have not been prospectively evaluated. Avoidance of environmental tobacco smoke and other indoor pollutants, avoidance of indoor allergens, and treatment of accompanying rhinitis may be helpful. In obese patients, weight loss strategies should be used. However, implementation of adjunctive therapies should not delay specific treatment of OSAS.

Oxygen therapy is sometimes prescribed in special cases to alleviate nocturnal hypoxemia in children with OSAS. However, there are few, if any, indications for its use in the otherwise healthy child with OSAS. Oxygen therapy does not prevent sleep-related upper airway obstruction and resultant problems, such as sleep fragmentation and increased work of breathing. Furthermore, it may worsen hy-

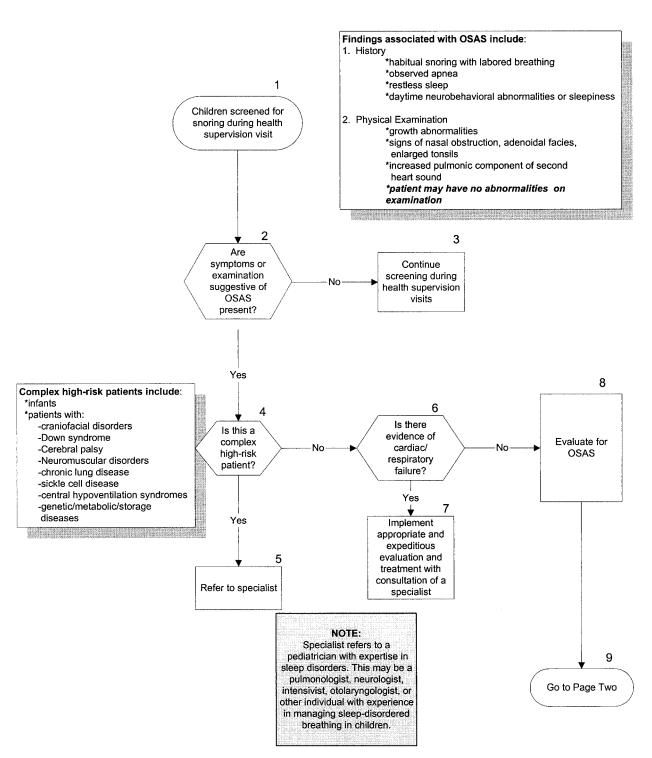


Fig 1. Diagnosis and management of uncomplicated childhood OSAS.

poventilation.⁶³ If oxygen therapy is to be used in children with OSAS, it should be evaluated during continuous Pco₂ monitoring to assess its effect on hypoventilation.

Other surgical options are available for patients not responding to usual treatment. These patients require care from pediatric surgical specialists. Surgical treatment options include uvulopharyngopalatoplasty, craniofacial surgery, and in severe cases, tracheostomy.

Follow-up of Patients Undergoing Surgical Treatment for OSAS

All patients should have clinical follow-up for reassessment of symptoms and signs associated with OSAS after initial treatment. Patients with mild to moderate OSAS who have complete resolution of symptoms and signs do not require objective testing to document resolution. Patients who have continued symptoms or signs, who have severe OSAS,³⁷ or

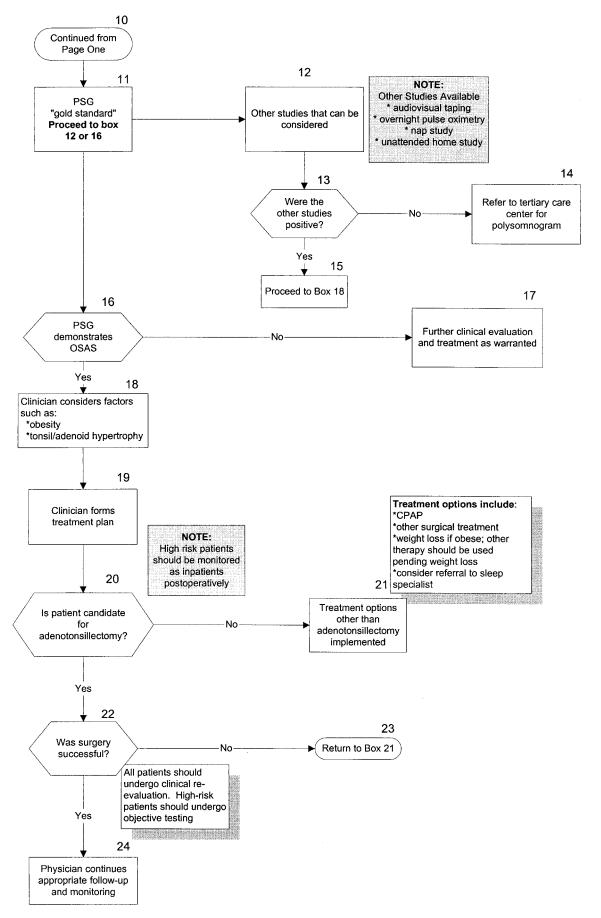


Fig 1. (continued)

who are obese require objective reevaluation to determine whether additional therapy, such as CPAP, is required. Objective data regarding timing of post-operative evaluation are not available. Most clinicians recommend waiting 6 to 8 weeks before reevaluation to ensure that upper airway, cardiac, and central nervous system remodeling is complete.

SUMMARY OF RECOMMENDATIONS FOR THE DIAGNOSIS AND MANAGEMENT OF UNCOMPLICATED CHILDHOOD OSAS

The following recommendations accompany an algorithm (Fig 1). As previously noted, these recommendations relate to otherwise healthy children older than 1 year with OSAS secondary to adenoton-sillar hypertrophy and/or obesity and who are not in cardiorespiratory failure.

- 1. All children should be screened for snoring. As part of routine health care maintenance for all children, pediatricians should ask whether the patient snores. An affirmative answer should be followed by a more detailed evaluation. (Evidence for this recommendation is good, and the strength of the recommendation is strong.)
- 2. Complex, high-risk patients (Fig 1) should be referred to a specialist. (Evidence is good that these children are at increased surgical risk and require more complex management; the strength of the recommendation is strong.)
- 3. Patients with cardiorespiratory failure cannot await elective evaluation. It is expected that these patients will be in an intensive care setting and will be treated by a specialist; thus, these patients are not covered in this practice guideline.
- 4. Thorough diagnostic evaluation should be performed. History and physical examination have been shown to be poor at discriminating between PS and OSAS (evidence is strong). Polysomnography is the only method that quantifies ventilatory and sleep abnormalities and is recommended as the diagnostic test of choice. Other diagnostic techniques, such as videotaping, nocturnal pulse oximetry, and daytime nap studies, may be useful in discriminating between PS and OSAS if results of polysomnography are positive. However, they do not assess the severity of OSAS, which is useful for determining treatment and follow-up. In any case, because of their high rate of false-negative results, polysomnography should be performed in the event of negative results of the other diagnostic techniques. Additional study of audiotaping is necessary. (Evidence for and strength of the recommendation are strong.)
- 5. Adenotonsillectomy is the first line of treatment for most children. CPAP is an option for those who are not candidates for surgery or do not respond to surgery. (Evidence for and strength of the recommendation are strong.)
- High-risk patients should be monitored as inpatients postoperatively. (Evidence that these patients are at high risk of postoperative complications is strong. Strength of the recommendation is strong.)

7. Patients should be reevaluated postoperatively to determine whether additional treatment is required. All patients should undergo clinical reevaluation. High-risk patients should undergo objective testing. (Evidence is good, strength of the recommendation is strong.)

RESEARCH RECOMMENDATIONS

Specific research questions have been delineated by an American Thoracic Society workshop.⁴³ Areas requiring additional investigation include:

- 1. Accurate prevalence data.
- 2. Identification of risk factors of complications resulting from OSAS, including the relationship of OSAS severity to specific outcomes.
- Development and evaluation of low-cost, highsensitivity, and high-specificity screening methods for OSAS.
- 4. Delineation of the natural history of treated and untreated PS and OSAS.
- 5. Assessment of long-term efficacy of adenotonsillectomy, CPAP, and other OSAS treatments.

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Section on Pediatric Pulmonology and Subcommittee on Obstructive Sleep Apnea Syndrome

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