Positive Airway Pressure Therapy in Children

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KEYWORDS

- Child • Obstructive sleep apnea • Positive airway pressure
- Noninvasive positive pressure ventilation
- Neuromuscular disorders

Children with obstructive sleep apnea syndrome (OSAS) have recurrent episodes of partial or complete airway obstruction during sleep, resulting in hypoxemia, hypercapnia, and sleep disruption. The earliest and best description of pediatric obstructive apnea can be found in Sir William Osler's 1892 textbook, "The Principals and Practice of Medicine" in a chapter entitled "Chronic Tonsillitis"[1]:

> *"The direct effect of chronic tonsillar hypertrophy is the establishment of mouth-breathing. The indirect effects are deformation of the thorax, changes in the facial expression, and sometimes marked alteration in the mental condition… At night the child's sleep is greatly disturbed; the respirations are loud and snorting, and there are sometimes prolonged pauses, followed by deep, noisy inspirations. The child may wake up in a paroxysm of shortness of breath…"*

Childhood OSAS is a common cause of morbidity in approximately 2% of children.[2] Because of the documented association of childhood OSAS with adverse neurocognitive, cardiovascular, metabolic, growth, and inflammatory sequelae, more children are being referred to sleep laboratories for polysomnography, based on the recommendations of the American Academy of Pediatrics (AAP) guidelines for management of OSAS.[3]

In the past, OSAS was primarily associated with adenotonsillar hypertrophy in children. However, because of the rising incidence of childhood obesity, this diagnosis increasingly affects the treatment and incidence of childhood OSAS. Obese children are more likely to have residual obstructive sleep apnea (OSA) after adenotonsillectomy. Tauman and colleagues[4] have shown that the polysomnogram may not always normalize after surgical resection. The use of noninvasive positive pressure ventilatory support using continuous positive airway pressure (CPAP) and nocturnal noninvasive positive pressure ventilation (NIPPV, also referred to as *BiPAP* or *bilevel PAP*) in children of all ages has increased in the past decade.

This article describes the use of pediatric positive airway pressure in the treatment of sleep-disordered breathing (SDB), highlighting pathophysiology and clinical practical considerations in the nonsurgical management of childhood OSAS and chronic respiratory failure.

PATHOPHYSIOLOGY OF SDB

OSAS

The essential feature of OSAS is increased upper airway resistance during sleep. In contrast to

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adults, in whom OSAS occurs predominantly in men, prevalence of OSAS does not differ between the sexes during childhood.[5] Anatomic narrowing of the upper airway and neuromuscular tone of upper airway musculature determine the propensity for upper airway collapse during sleep. Narrowing of the nasopharynx caused by allergic rhinitis, turbinate hypertrophy, deviated nasal septum, and maxillary constriction can all increase resistance in the nasal compartment. Palatal factors, tongue-base collapse, macroglossia, and posterior displacement of the mandible in craniofacial syndromes may also contribute to narrowing of the oropharynx or hypopharynx and result in OSAS.

Adenotonsillar hypertrophy is the most common cause of OSA in children between 2 and 8 years of age.[6] Children with other comorbidities, such as obesity, Trisomy 21, and cerebral palsy, may concurrently have abnormal central arousal thresholds, disordered neural control or airway caliber, and decreased pharyngeal tone. Dynamic airway collapse may occur at multiple sites in children with OSA. Hence, this is a multifactorial disorder with overlapping influences that predispose patients to obstructed breathing. The combination of genetic influences, neuromuscular tone, and structural anatomic factors variably and additively affect the presentation and severity of OSA.[7]

Obesity is an increasingly important and common risk factor for OSA in children.[8] Upper airway narrowing results from deposition of adipose tissue around the neck and jaw with external compression. Respiratory mechanics are altered with decreased chest wall compliance and upward displacement of the diaphragm by the obese abdomen, especially in the supine position. Lung volumes are reduced during sleep, with increased risk of desaturation associated with obstructive events. Gas exchange abnormalities with hypercapnia and obesity-related hypoventilation can occur in children[9]; hence, monitoring for hypoventilation during sleep is mandated in pediatric polysomnography using end-tidal carbon dioxide (CO_2) or transcutaneous CO_2 monitoring devices according to the guidelines for testing published by the American Academy of Sleep Medicine (AASM) in 2007.[10]

Chronic obstructed breathing with chronic hypoxemia, hypercarbia, or both may eventually lead to pulmonary hypertension in severe OSA.[11] Cardiovascular morbidity, right heart strain, and cor pulmonale may then ensue. Children with OSAS show signs of increased sympathetic activity, autonomic dysfunction, endothelial dysfunction, and a trend toward higher arterial blood pressure.[12,13]

Impaired growth has been seen in some children with OSAS[14] and has been thought to be related to increased work of breathing during sleep. These children have been shown to have impaired secretion of nocturnal growth hormone. Secondary nocturnal enuresis can also be a consequence of OSA, which may be caused by brain natriuretic peptide or atrial natriuretic peptide secretion.[15] Neurocognitive consequences include a higher incidence of attention deficit-hyperactivity disorder[16] and impaired learning[17] and memory,[18] affecting daytime behaviors and school performance in vulnerable children. Chronic OSA can therefore have reversible and irreversible consequences for children. Many of the these morbidities have improved after treatment with adenotonsillectomy[19] and efficacious use of NIPPV in children.[20]

CHRONIC RESPIRATORY FAILURE

Children who are at risk for chronic respiratory failure as a result of neuromuscular weakness, scoliosis, cerebral palsy, or myopathic disorders may ventilate adequately when awake but may decompensate with hypoventilation during sleep (**Box 1**, **Table 1**). Normally, during sleep, a decrease in tidal volume occurs, especially during rapid-eye-movement (REM) sleep, with a reduction in the respiratory rate and central respiratory drive. The P_{CO_2} rises by 3 to 7 mm of Hg, and a corresponding drop in PaO_2 of 3 to 9 mm of Hg may occur, compared with the waking state. Hence, gas exchange abnormalities and relative hypercapnia become apparent in vulnerable individuals during sleep, leading to sleep disruption.[21,22]

CENTRAL SLEEP APNEA AND CENTRAL HYPOVENTILATION

Causes in children include prematurity (central sleep apnea), Arnold-Chiari malformation,[23] and congenital central hypoventilation syndrome. Children with congenital central hypoventilation syndrome (a genetic disorder caused by a mutation in the *PHOX2B* gene) may have decreased tidal volume or respiratory rate during sleep, but may not have frank central apnea. These disorders tend to present during infancy, although late diagnoses have been reported.[24]

DIAGNOSIS

A history of snoring is a sensitive, though not specific, symptom of OSAS. Primary snorers are those who snore but have no obstructive apnea, gas exchange abnormalities, or sleep