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Obstructive sleep apnea syndrome in the pediatric age: the role of the pneumologist

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Abstract. – OBJECTIVE: This review paper aims to summarize the current state of knowledge on the role of the pneumologist in the diagnosis and respiratory treatment of children affected by obstructive Sleep Disordered Breathing (SDB).

MATERIALS AND METHODS: A literature review has been performed on the following topics: obstructive SDB and its clinical entities, indications for respiratory treatment of pediatric SDB, and Continuous Positive Airway Pressure (CPAP) and Noninvasive Positive Pressure Ventilation (NIPPV) treatment approach to obstructive SDB.

RESULTS: OSDB is related to obesity, craniofacial pathologies, neuromuscular disorders and, most commonly, adenotonsillar hypertrophy. Adenotonsillectomy is the first-choice treatment in children with obstructive apnea secondary to adenotonsillar hypertrophy. CPAP and NIPPV are recommended in cases where Obstructive Sleep Apnea (OSA) persists after surgery or when surgery is contraindicated. Treatment interventions are usually implemented gradually by separately addressing each abnormality that would predispose to obstructive SDB, then reevaluating after each intervention to detect any residual disease and to assess the need for additional treatment.

CONCLUSIONS: Many pediatric patients continue to experience problems and symptoms such as hypersomnia and apnea after adenotonsillectomy and need CPAP/NIPPV treatment. Current knowledge is still incomplete, especially with regard to the mechanisms of pathogenesis of pediatric OSA, the factors affecting pediatric OSA, and the phenotypic variability of the disease. A better understanding of these aspects would contribute to the development of new therapies.

Key Words
Continuous Positive Airway Pressure, Noninvasive Positive Pressure Ventilation, Tonsillectomy, Adenotonsillectomy, Obstructive sleep disordered breathing.

List of Abbreviations

SDB: sleep disordered breathing; OSA: obstructive sleep apnea; PSG: polysomnography; AHI: apnea-hypopnea index; TST: total sleep time; HST: Home sleep testing; CPAP: continuous positive airway pressure; NIPPV: Noninvasive positive pressure ventilation; BiPAP: Biphasic positive airway pressure.

Introduction

Pediatric obstructive sleep disordered breathing (SDB) comprises a range of obstructive respiratory disorders during sleep; characteristic clinical indications include snoring and/or increased respiratory effort secondary to increased risk of upper airway resistance and pharyngeal collapse[1]. These phenomena may cause hypoxia, hypercarbia, increased respiratory efforts, considerable intrathoracic pressure changes, and sleep fragmentation[2]. Obstructive sleep apnea (OSA) is one of the most common causes of SDB in children. It affects neurocognitive development and the child's general state of health. Moreover, SDB has a multifactorial etiopathogenesis and its correct diagnosis and therapy require a multidisciplinary approach that involves otorhinolaryngologists, neurologists, pneumologists, maxillofacial surgeons, dentists, cardiologists and pediatricians[3].

The term "obstructive SDB" is used whenever intermittent obstruction symptoms in the upper airways are observed during sleep; however, the severity of the disease is determined by studying daily drowsiness, along with objective tests like polysomnography (PSG). In a recent meta-analysis, the prevalence of regular snoring in children was 7.45%, while the presence of OSA syndrome showed a

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frequency ranging from 1% to 4%. The apnea-hypopnea index (AHI) is the polysomnographic parameter that measures SDB severity by the number of mixed, obstructive, and central apneas and hypopneas per hour during a complete sleep[4,5].

The aim of this review article is to evaluate the role of the pneumologist in the diagnosis and respiratory treatment of children affected by obstructive SDB.

Materials and Methods

A literature review has been performed on articles retrieved from PubMed and Scopus from the last 30 years on the following topics: obstructive sleep disordered breathing and its clinical entities, indications for respiratory treatment of pediatric SDB, and Continuous Positive Airway Pressure (CPAP) and Noninvasive Positive Pressure Ventilation (NIPPV) treatment approach to obstructive SDB.

Results

Obstructive Sleep Disordered Breathing and Its Clinical Entities

The spectrum of pediatric obstructive SDB has four degrees of severity [6]:

Primary snoring: the mildest and most prevalent manifestation. Habitual snoring is defined as snoring that occurs more than 3 nights per week without apnea, hypopnea, frequent arousal, or gas exchange abnormalities.

Upper Airway Resistance Syndrome (UARS): involves snoring, increased respiratory effort, and frequent arousal, without recognizable presence of obstructive events or gas exchange abnormalities.

Obstructive hypoventilation: characterized by nocturnal snoring with a high increase of carbon dioxide in the blood in the absence of recognizable obstructive events.

OSA syndrome: manifests itself with frequent events of partial or complete obstruction of the upper airways (hypopnea, obstructive or mixed apnea) with normal oxygenation, ventilation and sleep interruption.

Indications for Respiratory Treatment of Pediatric SDB

The primary standard test to diagnose obstructive SDB and assess its severity is PSG [1,7]. PSG allows for objective diagnosis, classification and severity of the pathologic condition. AHI total sleep time (TST), the number of mixed, obstructive and central apneas and hypopnea per hour of total sleep time, is the PSG parameter used to diagnose and quantify the severity of SDB. AHI TST tracks obstructive, mixed apnea or obstructive hypopnea events per hour of sleep, hypoventilation during sleep (when PaCO2 > 50 mmHg for > 25% of TST), isolated snoring, paradoxical thoracoabdominal motion, and/or flow limitation. Most sleep centers consider an obstructive AHI $\leq$ 1 episode per hour TST to be normal, 1 < AHI $\leq$ 5 as mild OSA, 5 < AHI $\leq$ 10 as moderate OSA and AHI >10 as severe OSA.

PSG is indicated in children with symptoms of obstructive SDB who have not yet undergone adenotonsillectomy, especially in the presence of obesity. PSG has also been recommended after adenotonsillectomy in patients with persistent symptoms of OSA syndrome despite surgery[7].

When PSG is not available, possible alternatives include nocturnal oximetry studies, nocturnal cardiorespiratory PSG recording, and home sleep testing (HST) and ambulatory PSG recording[8].

Nocturnal oximetry studies consist in continuous recording with trend analysis and an average time of 3 seconds or less. Oximetry studies have high specificity but low sensitivity in the diagnosis of pediatric OSA. According to the McGill criteria, the diagnostic elements of oximetry studies can be summarized as three or more clusters of desaturation $\geq$ 4%, with at least three desaturations at < 90% being considered pathological. However, the rate of false negative or inconclusive results is high. In a recent study on 589 patients suspected of OSA, Hornero et al[9] have demonstrated that the automated analysis based on the neural network of oximetry saturation recordings provides a comprehensive identification of the severity of OSA among children with a high probability of OSA pre-testing. These results strongly suggest that nocturnal oximetry can provide a simple and effective diagnostic alternative to nocturnal PSG, leading to prompter interventions and potentially better results.

Nocturnal cardiorespiratory PSG recording is a regular PSG exam without the electroencephalography, electromyography and electrooculography sensors. It is easier than PSG to perform and to set. Some authors have reported good results with in-lab PSG[10,11].

HST and ambulatory PSG recording are also alternative studies to PSG. Outpatient cardiorespiratory nocturnal PSG has proven to be effective in guiding the diagnosis of obstructive SBD. Cardiorespiratory nocturnal PSG tests are considered as a

less expensive alternative to in-lab PSG; moreover, the results may be more representative of the child's typical home sleep. Comparison data with in-lab PSG are still limited, but results from a recent study by Alonso-Alvarez et al[12] comparing outpatient HST with in-lab PSG in 50 children clinically suspected of OSA, has shown promising diagnostic potential.

In addition to PSG, self-administered questionnaires that investigate the presence of characteristic symptoms, risk factors and predictors of OSA can be used; however, they are mainly used in the adult population[13-15] although some have been applied to pediatric patients as well[16].

Ventilatory Treatment for Obstructive SDB

Treatment for children with obstructive SDB may be surgical or non-surgical; the choice depends on the underlying etiology. The European Respiratory Society Taskforce [1] recommends a gradual approach to treatment, up to full resolution of OSA. The taskforce recommends treatment in children with AHI > 5; in children with AHI between 1 and 5, ventilatory treatment may be initiated in the presence of comorbidity. In the common case of significant adenotonsillar hypertrophy, the first step in treating pediatric SDB is adenotonsillectomy. In OSA pediatric patients without significant adenotonsillar hypertrophy, positive airways pressure therapy is recommended to keep the upper airways open throughout the respiratory cycle, improve lung function and reduce breathing. Starting CPAP treatment in children may be difficult; time must be taken to educate the parents on the importance of the treatment and to make sure they are involved [17].

Indications for CPAP and NIPPV are residual OSA after adenotonsillectomy (AHI > 5) and OSA syndrome related to obesity, craniofacial abnormalities, or neuromuscular disorders. If nocturnal hypoventilation occurs (end-tidal carbon dioxide tension PCO2 > 50 mmHg for > 25% of total sleep time or peak end-tidal PCO2 ≥ 55 mmHg), NIPPV is preferred [1]. Indications are summarized in Table I. NIPPV of the airway has been found to produce improved gas exchange, along with improvements in attention deficit, sleepiness, behavior, and quality of life. Treatment with CPAP/NIPPV should be initiated in the hospital. Several nasal/oronasal masks should be tried to choose the one that the child accepts best. Treatment should be started in daytime to accommodate incremental pressures. In some cases, in-hospital application needs to be for several nights, depending on the child's tolerance of CPAP/NIPPV.

Discussion

The pneumologist plays a central role in the respiratory treatment of children affected by obstructive SDB when the surgical approach is contraindicated or, more commonly, OSA persists after surgery. In fact, many pediatric patients continue to experience problems and symptoms such as hypersomnia and apnea after adenotonsillectomy and need CPAP/NIPPV treatment. Furthermore, positive airway pressure therapy is recommended in all cases of SDB secondary to obesity, craniofacial pathologies, and neuromuscular disorders.

Treatment with CPAP/NIPPV can be started in a team approach to make the process fun and reduce anxiety. When night and day PaCO2 is elevated, which occurs in children with other comorbidities such as neuromuscular disease, craniofacial syndromes or obesity hypoventilation, NIPPV treatment may be preferentially indicated. In NIPPV, the machine offers greater inspiratory/expiratory pressure comfort, so is recommended when the child does not tolerate CPAP. Biphasic positive airway pressure (BiPAP) treatment may also be better tolerated in children who do not tolerate CPAP because of better positive exhalation pressure levels. CPAP and NIPPV complications include nasal congestion, rhinorrhea, epistaxis, skin facial erythema due to the mask, and discomfort due to air build-up in the abdomen. It is important to monitor adherence to CPAP or NIPPV and to manage complications to optimize patient adherence. Regular

Table I. Indications for ventilatory treatment of obstructive Sleep Disordered Breathing (SDB).

– AHI > 5 episodes (CPAP/NIPPV).
– CPAP or NIPPV (for nocturnal hypoventilation) may be beneficial in cases of AHI 1–5 episodes in the presence of: morbidity from the cardiovascular system; morbidity from the central nervous system; enuresis; somatic growth delay or growth failure; decreased quality of life; risk factors for SDB persistence.
– SDB respiratory treatment (CPAP or NIPPV) is a priority in the presence of: serious craniofacial abnormalities; neuromuscular disorders; achondroplasia; Chiari malformation; Down syndrome; mucopolysaccharidoses; Prader-Willi syndrome.

long-term control should be performed with respect to respiratory work pressure and device interface in order to prevent cutaneous decubitus. Generally, PSG recording is recommended 6 weeks after adenotonsillectomy. Children treated by CPAP or NIPPV should be reviewed at least every 12 months after initial treatment.

Conclusions

CPAP is recommended in cases where OSA persists after surgery, when surgery is contraindicated or in patients with SDB secondary to obesity, craniofacial pathologies, and neuromuscular disorders. CPAP/NIPPV treatment is complex and requires many multidisciplinary specialists, as many children do not tolerate CPAP. Our knowledge is still incomplete, especially with regard to the mechanisms of pathogenesis of pediatric OSA, the factors affecting pediatric OSA, and the phenotypic variability of the disease. A better understanding of these aspects would contribute to the development of new therapies.

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Conflict of Interests

The authors declare that they have no conflict of interest.

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