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1. Introduction

Obstructive Sleep Apnea Syndrome (OSAS) is a condition characterized by intermittent partial or total obstruction of the upper airways during sleep. The events of upper airway obstruction are associated with repetitive episodes of hypoxemia and microarousals, usually followed by autonomic activation. As a consequence, OSAS is related to sleep fragmentation, excessive daytime sleepiness and its consequences, cognitive and behavioural changes, and an increased risk of cardiovascular and cerebrovascular diseases.[1]

In childhood, OSAS is characterized by both intermittent obstruction and by prolonged periods of partial resistance/obstruction of the airways.[1] Methodological differences in diagnosing this disease have led to variable reports of prevalence, with the strongest evidence indicating a prevalence of 1 to 5%.2 The disease occurs in all childhood age ranges from the neonatal period to adolescence, being more common among preschoolers.

In the childhood age range, OSAS is more frequently associated with tonsil and adenoid hypertrophy and with other conditions including obesity, allergic rhinitis, craniofacial malformations, neuromuscular diseases, and genetic and metabolic syndromes.

Important clinical outcomes of the condition such as delayed growth and hyperactive behavior have been well established. [1]

2. Clinical aspects

Clinical Signs and Symptoms: The clinical signs and symptoms are mainly characterized by snoring, difficult breathing during sleep, nighttime breathing pauses, agitated sleep, and hyperactive behavior.
Snoring is present in the great majority of children, but may not be observed in infants or children with muscle weakness. Paradoxical breathing is frequently present due to a more complying thoracic cage in childhood.1

Different ventilatory patterns may characterize Sleep Disordered Breathing (SDB) in childhood, with the predominance or exclusive presence of each one in each child: [1]

1. Cyclic apneas, as observed in adults, with snoring associated with intermittent breathing pauses followed by noisy inspiraton and movements/microarousals.

2. Obstructive hypoventilation, with continuous snoring, without frequent pauses or microarousals. This pattern occurs in younger children and consists of prolonged periods of partial airway obstruction associated with hypercapnia or hypoxemia, or both.

3. A pattern similar to that known in adults as Upper Airway Resistance Syndrome, with snoring and intermittent periods of greater ventilatory effort associated with microarousals, with no changes in flow compatible with apnea or hypopnea.

In childhood, respiratory events may occur without being associated with microarousals, especially in younger children, due to the high arousal threshold. In addition, these events occur more during REM sleep, when the child is especially predisposed not to wake, and are rare during slow-wave sleep. [1] Typically, there is greater preservation of sleep architecture than in adults. For this purpose, the main pattern of sleep architecture change is the increase of slow-wave sleep and a reduction of REM sleep duration. [3, 4]

Snoring is usually reported by the caregivers, whereas breathing pauses may not be perceived. Or, conversely, the parents may report the observation of nighttime breathing pauses in children, with these events being of the central type – or even obstructive – but of an insufficient number to characterize OSAS. Thus, anamnesis alone is insufficient to exclude or diagnose sleep apnea in children who snore. [5, 6, 7, 8, 9]

In addition to snoring and breathing pauses, other signs observed are agitated sleep, night sweats, preferential decubitus with cervical hyperextension, and enuresis. Episodes of parasomnia and sleep bruxism may be more frequent. Morning headache, difficulty in getting up in the morning and excessive daytime sleepiness may occur, especially among older children. Excessive sleepiness is usually absent in younger children, who more commonly show daytime agitation.

Since tonsil and adenoid hypertrophy is the main cause underlying OSAS in the childhood age range, related clinical aspects may be present, such as mouth breathing syndrome and its orthodontic and craniofacial complications such as crossbite, high-arched palate, and long face syndrome with practically constant open mouth (figure 1); dysphagia and odynophagia; repeated upper airway infections; hearing loss; gastroesophageal reflux disease. So far, the degree of tonsil and adenoid hypertrophy has not been documented to predict the presence of OSAS in children who snore and are mouth breathers. [10, 11] Methodological aspects may be involved, in addition to the fact that other factors contribute to the presence or absence of OSAS, such as particularities of the neural control of ventilation in each child.
In younger children, OSAS may be related to difficulty in gaining weight, particularly when associated with genetic syndromes. In older children, obesity may be present. Regardless of their weight status, children may develop weight gain after treatment of OSAS, not infrequently increasing their food intake after improvement of olfaction, of dysphagia and of odynophagia induced by adenotonsillectomy.

Some of the clinical outcomes of this syndrome in childhood have been well established, whereas others are still under investigation. The complaint of school difficulty is relatively frequent and most studies have established an association between childhood OSAS and cognitive deficit. However, the correlation between the severity of the breathing disorder and the degree of neuropsychological impairment is still controversial due to methodological differences and to the lack of control of other relevant clinical variables such as obesity, in addition to environmental and social variables. More studies are still needed to determine the cognitive subdomains that are more affected, their relationship with hypoxemia and sleep fragmentation, and their evolution after the treatment of OSAS.

From a behavioral viewpoint, hyperactive behavior is the most common abnormality. Children tend to show a worse performance in tests of sustained attention and executive functions and may or may not fulfill the formal criteria of Attention Deficit Hyperactivity Disorder (ADHD). In addition, children with ADHD have a higher prevalence of SDB than controls. Aggressiveness, difficulty in social relations, and mood changes are other behaviors reported.

Excessive daytime sleepiness may be present in some children, although few studies have correlated polysomnography (PSG) parameters with objective sleepiness parameters. Some studies have demonstrated behavioral and cognitive improvement after the treatment of apnea in children, whereas others have detected persistence of the previous impairment. Well-designed studies are still needed, with the control of confounding variables such as...
family and social environment, educational level, time of disease evolution, and the presence of other sleep disorders.

Regarding the cardiovascular outcomes, despite the scarcity of well-designed studies, there is evidence indicating increased arterial pressure and repercussions on both the right and left ventricles. Arterial hypertension, pulmonary hypertension and cor pulmonale may occur in children with more severe disorder. There is a lack of well-controlled studies also regarding inflammatory markers, with C-reactive protein apparently increasing in more serious cases. [2]

3. Diagnosis

Ideally, the presence of OSAS should be investigated in all children with complaints of snoring and agitated sleep. However, the predictive value of the clinical history alone is low, with PSG being considered to be the gold standard for diagnosis. [17] Alternative methods of diagnostic complementation such as oximetry, evaluation of cardiovascular parameters, ambulatory evaluation of ventilatory parameters, and daytime PSG have not been recommended to define the diagnosis thus far, as they may not be sufficient when negative. Ideally, children with negative results should be referred to whole night PSG study. [2]

<table>
<thead>
<tr>
<th>A</th>
<th>Report of snoring or of increased breathing effort, or both during sleep</th>
</tr>
</thead>
<tbody>
<tr>
<td>B</td>
<td>The caregiver or the child reports at least ONE of the signs/symptoms below:</td>
</tr>
<tr>
<td></td>
<td>1. presence of a paradoxical breathing pattern during inspiration</td>
</tr>
<tr>
<td></td>
<td>2. arousal associated with movements</td>
</tr>
<tr>
<td></td>
<td>3. diaphoresis</td>
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<td></td>
<td>4. cervical hyperextension during sleep</td>
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<td></td>
<td>5. excessive daytime sleepiness, hyperactivity or aggressive behavior</td>
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<td></td>
<td>6. reduced growth rate</td>
</tr>
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<td></td>
<td>7. morning headache</td>
</tr>
<tr>
<td></td>
<td>8. secondary nighttime enuresis</td>
</tr>
<tr>
<td>C</td>
<td>PSG presents obstructive AHI ≥ 1/hour</td>
</tr>
<tr>
<td>D</td>
<td>PSG presents items 1 or 2 below:</td>
</tr>
<tr>
<td></td>
<td>1. At least ONE of the events below:</td>
</tr>
<tr>
<td></td>
<td>1.1. increased arousals related to increased breathing effort;</td>
</tr>
<tr>
<td></td>
<td>1.2. fall in oxygen saturation associated with obstructive breathing events;</td>
</tr>
<tr>
<td></td>
<td>1.3. hypercapnia during sleep;</td>
</tr>
<tr>
<td></td>
<td>1.4. marked negative oscillation in esophageal pressure</td>
</tr>
<tr>
<td></td>
<td>2. Periods of hypercapnia or desaturation, or both, during sleep, associated with snoring, paradoxical breathing and at least one of the events below:</td>
</tr>
<tr>
<td></td>
<td>2.1. frequent arousals;</td>
</tr>
<tr>
<td></td>
<td>2.2. marked negative oscillation of esophageal pressure</td>
</tr>
</tbody>
</table>

Table 1. Diagnosis of OSAS in Childhood and Adolescence
According to the criteria of the International Classification of Sleep Disorders [1], the diagnosis is based on clinical and PSG criteria (Table 1). From a clinical viewpoint, there must be the complaint of snoring and/or difficult breathing during the night, associated with at least one of the following signs and symptoms: paradoxical breathing, agitated sleep, nocturnal sudoresis, cervical hyperextension, excessive daytime sleepiness, hyperactivity or aggressive behavior, morning headache, and secondary enuresis.

From a polysomnographic viewpoint, an Apnea + Hypopnea Index (AHI) ≥ 1/hour should be present in association with sleep fragmentation, desaturation episodes, hypercapnia, or negative oscillations of esophageal pressure.

4. Clinical and complementary evaluation

4.1. Clinical evaluation and physical examination: Anterior rhinoscopy and endoscopy

The systematized measurement of cervical circumference routinely used for adults has not been standardized for children and therefore it is not routinely used in most services.

The otorhinolaryngology exam is always focused on the search of obstructive causes in the airways, from the nasal fossae to the regions of the hypopharynx and larynx. Bone changes such as micrognathia and deformity of the skull base (present, for example in individuals with Down Syndrome) should always be remembered. Complementary flexible nasofibroscopy is desirable, as it permits a precise evaluation up to the larynx region. The main causes of respiratory obstruction are:

Choanal Atresia: this is a congenital malformation that leads to nasal obstruction, nasal secretion and, when bilateral, respiratory stress breathing at birth. The diagnosis can be made by CT scan and nasal endoscopy (Figure 2).

Figure 2. Choanal Atresia. (A) Endoscopic view: posterior nasal fossa, left side, with impermeable choana; (B) Axial CT scan, showing the choanal atresia at the left side.
Adenotonsillar hypertrophy: the complaints reported by the mother usually starts when the child is already older than two years, although they may also start earlier. Depending on the severity of the case, the child has nighttime apnea which considerably frightens the parents, who are unable to sleep. Diagnosis of palatine tonsils hypertrophy is clinical (Figure 3), while adenoid hypertrophy, in most cases, the diagnosis is confirmed by simple lateral radiography or nasofibroscopy (Figure 4). It should be pointed out that Valera et al.[18], in 2005, in a retrospective study based on the analysis of clinical data in the medical records of 267 children, did not observe a correlation between the degree of adenotonsillary hypertrophy and the severity of OSAS. These data were later confirmed by Nolan and Brietzke (2011) [10], who concluded that the association between tonsil grading and OSAS severity should be considered at best weak.

Figure 3. Grade IV palatine tonsils

Figure 4. Adenoid hypertrophy. (A) Lateral X-Ray; (B) Endoscopic view, with important obstruction in nasopharynx due to adenoid hypertrophy.

Allergic rhinitis: children with allergic rhinitis who are not properly treated may present severe nasal obstruction because the hypertrophic nasal turbinates prevent the airflow.
Anatomical variations of the nasal turbinates: the most common of them is the Bollosa turbinate, when the middle turbinate is pneumatized. This variation may be only a endoscopic/ radiographic finding, but it may be also related to nasal obstruction and repeated rhinosinusitis. The diagnostic suspicion based on nasofibroscopy is confirmed by CT scan (Figure 5).

Figure 5. Coronal CT scan, showing bilateral concha bollosa

Septal deformities: important deviations of the septal wall can also induce mouth breathing and OSAS in children (Figure 6).

Nasal tumors: benign or malignant tumors in the nasal fossae of children may provoke unilateral or bilateral nasal obstruction and should be promptly diagnosed (Figures 7 to 9).

Figure 6. Anterior septal deviation.
Flexible nasal fibroscopy should reach the region of the larynx, also for the evaluation of changes in soft tissues such as macroglossia and of laryngeal diseases such as laryngomalacia (Figure 10). The procedure permits the diagnosis of hypotonia of the dilators of the lower airways present in children with neuromuscular abnormalities (hypotonic muscular dystrophies and cerebral palsy causing lack of coordination).

Some authors[19] recommend the use of anterior rhinomanometry, which measures nasal resistance in order to diagnose severe apnea in children. According to them, the nasal resistance of children is significantly reduced with age and increases in the presence of edema of the nasal fossae induced by adenoid enlargement. However, this exam is not routinely performed in these children.
4.2. Cephalometry

In view of the interaction between craniofacial changes and SDB in children, cephalometry is considered to be a useful exam for these patients. [20] The exam consists of radiography of the face in a systematic manner, so that the data for one patient can be compared to a data bank of normal values.

However, its routine use for the evaluation of patients with OSAS is still questioned by the major consensuses. [21] According to these consensuses, clinical evaluation can identify the main craniofacial changes when they are more exuberant and this should determine whether the patient needs cephalometry as an additional exam. Cephalometry, however, is essential for the indication of surgery in patients with craniofacial anomalies.
All professionals who deal with children should be aware of these as the main causes of OSAS in children and refer these patients to a specialist who will detect them and treat them correctly as soon as possible. Permitting the child to breathe through the nose before five years of age prevents the installation of changes of bone development and of facial muscles and will favor growth with the desired orofacial harmony.

4.3. Polysomnography

Nocturnal polysomnography (PSG) in a sleep laboratory is considered to be the gold standard for the evaluation of SDB since it provides an objective and quantitative evaluation of the respiratory and sleep architecture parameters [17].

Despite the scarcity of sleep laboratories with experience in treating children, diagnostic PSG recording in childhood can be acquired with few technical variations compared to adult examination, with the most important differential probably being the incorporation of capnography. The interpretation of the recording should be adapted to the childhood age range and the recommendation is to acquire and analyze the data according to the pediatric criteria of the sleep staging manual of the American Academy of Sleep Medicine (AASM) [22]. These criteria should be applied for children and adolescents up to 18 years of age, although, in selected cases, adult criteria can be applied to individuals older than 13 years.

Apneas should be identified by recording oronasal airflow with a thermistor, and hypopneas should be identified with pressure transducers using a nasal pressure tube [22].

The main events identified are:

1. Obstructive apnea: A reduction of basal air flow of 90% or more for at least two breathing cycles, accompanied by breathing effort.

2. Mixed apnea: A reduction of basal air flow of 90% or more for at least two breathing cycles, with breathing effort present only during one period of absence of airflow.

3. Central apnea: A reduction of basal air flow of 90% or more in the absence of breathing effort. Central apneas lasting more than 20 seconds and central apneas with a duration of two respiratory cycles accompanied by desaturation ≥ 3% or arousal are computed. For children younger than 1 year, only central apneas associated with a reduction of heart rate of less than 50 bpm for at least 5 seconds, or less than 60 bpm for at least 15 seconds are considered.

4. Hypopnea: Reduction of at least 30% of the amplitude of the pressure tube signal for two respiratory cycles accompanied by desaturation of ≥ 3% or arousal. When breathing effort is maintained, obstructive hypopnea is considered to be present [2].

In children, for the diagnosis and classification of SDB, no effect of the first night responsible for erroneous stratification of the disease was observed. The night-to-night variation of AHI in consecutive PSG or PSG performed at intervals of up to 50 days does not seem to be significant in children aged 2 to 17 years [23, 24, 25, 26, 27, 28]. In this respect, the recording of one night is usually adequate for the diagnostic evaluation of SDB.
Few studies have specifically assessed the accuracy of PSG for the diagnosis of OSAS in children. The fragility of the correlation between PSG parameters and the remaining aspects of the disease, such as the clinical itself does not necessarily indicate poor validation of PSG, since these aspects may not have the reliability or stability needed to represent a useful comparative measurement. Also, test-test tests after intervention studies have provided moderate to strong evidence of the validity of PSG for the characterization of childhood SDB. Also, reliability and reproducibility tests provide good to excellent support for the use of PSG in the evaluation of ventilatory parameters in infants and children. [17, 28] 

In summary, the PSG exam in children is probably useful, valid and reproducible and, when interpreted in the light of clinical data, it represents the gold standard for the diagnosis of SDB also in the childhood age range.

5. Treatment

Pharmacological Treatment. Since the major cause of OSAS in children is adenotonsillary hypertrophy [22], the initial treatment should approach these structures.

For children with adenoid hypertrophy alone, the initial treatment could be the use of topical nasal corticosteroids. The use of mometasone furoate, for example, has been effective in reducing the dimensions of the adenoids and in improving the obstructive symptoms. [29, 30, 31] There is no evidence about treatment with Montelucast alone.

Adenotonsillectomy should be considered in cases in which there is association with hypertrophy of the palatine tonsils and in cases that did not respond adequately to clinical treatment.

Adenotonsillectomy. Adenotonsillectomy is considered to be the main treatment of OSAS in childhood. [2, 32] This is a procedure with a high benefit/risk ratio[2], since it is highly efficient and presents a low prevalence of complications. Major complications are bleeding, infection, anesthetic complications, respiratory decompensation, velopharyngeal incompetence, subglottic stenosis and, rarely, death.

Despite the low postoperative risks in general, there is a pediatric population that is especially susceptible to complications: patients younger than 3 years, with severe OSAS, with cardiac complications, difficulty in gaining weight, important craniofacial changes, genetic syndromes, and neuromuscular diseases. All of these children should be submitted to adenotonsillectomy in a tertiary hospital, where prompt admission to the pediatric ICU would be possible.[33] In addition, the American Academy of Otolaryngology-Head and Neck Surgery recommends that children with AHI ≥ 10/h and/or Nadir of SATO₂ < 80% be admitted for observation after adenotonsillectomy. [34]

Partial tonsillectomy is not indicated since it may cause greater perioperative bleeding, maintenance of repeated infections and recurrence of obstruction due to new tissue growth. [2]
Relative contraindications of adenotonsillectomy for OSAS are: a small tonsil and adenoid size, acute infection of the upper airways, untreated hemorrhagic disease, or other clinical conditions that cause patient instability for the surgical procedure.

Adenotonsillectomy has proved to reduce AHI significantly when compared to preoperative values. [32, 35, 36, 37] The rates of cure obtained with adenotonsillectomy vary according to the definition of OSAS used, to the definition of cure criteria and to sample differences, such as the proportion of obese children among the subjects operated. For AHI ≥ 1/h the rates of OSAS persistence after adenotonsillectomy vary from 19 to 73%. Consistent risk factors for residual OSAS reported in the literature are obesity and severity of preoperative OSAS. The absence of postoperative snoring represents a good parameter for reevaluation, although it is not 100% specific. Thus, PSG should be performed after surgery in children at risk for residual disease.

In addition to improving AHI, adenotonsillectomy is associated with an improvement of the quality of life, of behavior, of cognitive function, and of oral motricity. [38, 39, 4, 41] Another benefit, mainly observed in children of preschool age, is the reversal of some craniofacial changes: in some studies, adenotonsillectomy led to greater transverse palatal growth, to compensation of anterior crossbite and to a reduction of mandibular inclination.[42, 43, 44, 45, 46, 47]

Rapid maxillary expansion. Rapid maxillary expansion (RME) is an orthodontic procedure for the enlargement of the transverse diameter of the hard palate by the redimension of the palatine suture, which may be an alternative for children with maxillary constriction and malocclusion.

RME is only indicated when the children present concomitant maxillary atresia, preferably associated with unilateral or bilateral crossbite and when the maxillary symphysis has not yet undergone fusion. In some studies conducted by the Stanford group, [48, 49, 50] RME was associated with a significant improvement of apnea indiceses and with an improved quality of life.

Despite this proven improvement of PSG indices, there still is some controversy about the effect of RME on the enlargement of nasal dimensions: while some studies have demonstrated an increased volume and a reduced nasal resistance, [51, 52, 53, 54] others have not been able to demonstrate this effect.[55, 56, 57] The same conflict occurs regarding enlargement of the pharynx: while Iwasaki et al.[58] observed an increased pharyngeal volume by cone-beam tomography, Ribeiro et al.[59] and Langer et al.[60] detected no effect of RME on nasopharyngeal volume.

Thus, the effect of RME on childhood OSAS needs to be better elucidated for a better understanding of the mechanism responsible for this clinical improvement and for the confirmation of its real benefit.

Positive Pressure Therapy. Despite the treatments described above, some degree of residual OSAS persists in many children, who continue to experience apnea even after optimized clinical/surgical treatment. [32, 61] This persistence is mainly observed in older children, in
children with associated obesity and asthma, and in children with more severe apnea during the preoperative period. [32, 36] In cases of residual OSAS in children with craniofacial anomalies, skeletal treatment (clinical, with orthodontic braces or surgical) can optimize the improvement and, in many cases, reverse the persistence of OSAS. However, the treatment most indicated for cases of moderate to severe residual OSAS is continuous positive airway pressure (CPAP). [2]

CPAP is used in general in children with persistent moderate to severe disease after surgical correction, especially obese children, children with craniofacial anomalies or children with contraindication of surgery. Treatment with CPAP is associated with improvement of clinical symptoms and of PSG parameters.

Despite a significant improvement in respiratory parameters and in quality of life, a problem with the use of CPAP in children is the rate of adherence: according to Marcus et al, [62] one third of the children abandon the use of the device by six months after its indication. Thus, the success of therapy depends on greater efforts for obtaining adequate nasal or oronasal interfaces, education, support and parental counseling.

Bilevel positive airway pressure (BiPAP) therapy is indicated for children with comorbidities that lead to the absence or insufficiency of the ventilatory drive, such as sequelae of cardiac-respiratory arrest and Moebius Syndrome, or hypoventilation secondary to neuromuscular diseases or chest wall deformities.

**Special Conditions.** Children with craniofacial abnormalities, genetic syndromes, sequelae of a hypoxic-ischemic insult and neuromuscular diseases should receive individualized treatment that might contemplate adenotonsillectomy, specific treatment of the base disease, when present, procedures for facial deformities such as mandibular distraction, and therapy with positive pressure. Tracheostomy is indicated when CPAP/BiPAP treatment is impossible or fails in children with very severe OSAS, or may be performed transitorily during the perioperative period in airway surgeries in children at risk for respiratory insufficiency.

6. Future research

The described flow diagram for the therapeutic approach to OSAS is not so simple, with many children who do not present the principal risk factors continuing to have OSAS after surgery, while others continue to have mild symptoms of low clinical importance for their parents.

At present, these children pose the greatest difficulty of conduct:

- Should partial polysomnographic improvement be treated even when the child continues to be asymptomatic?
- If so, which treatment should be indicated?
- May mild residual OSAS predispose this child to becoming an adult with OSAS?
All of these questions, although occasionally answered, should be further explored in the near future so that more appropriate therapeutic conducts can be offered to the pediatric population.

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