Liječ Vjesn 2023;145;suplement 5:3–6
https://doi.org/10.26800/LV-145-supl5-1

Pregled | Review

Sleep disordered breathing in children: what a pediatrician needs to know?

Poremećaji disanja u spavanju kod djece: što pedijatar treba znati?

Alessandro Amaddeo[1]✉

[1] Institute for Maternal and Child Health IRCCS Burlo Garofolo, Trieste, Italy

Keyword
SLEEP, OBSTRUCTIVE SLEEP APNEA, CONTINUOUS POSITIVE AIRWAY PRESSURE, NONINVASIVE VENTILATION

ABSTRACT. Obstructive sleep disordered breathing (SDB) is defined by the presence of recurrent partial or complete upper airway obstruction (hypopneas, obstructive or mixed apneas) with disruption of normal oxygenation, ventilation and sleep pattern. SDB is quite common in otherwise healthy children but the prevalence in children with congenital and genetic diseases is higher often with a more severe form of OSAS. Its consequences encompassed metabolic, cardiovascular and neurocognitive alterations. Clinical symptoms vary according to age, with some diurnal and nocturnal symptoms. No questionnaire had demonstrated a good reliability for the diagnosis of SDB and for the prediction of its severity. The mainstem of treatment is represented by adenotonsillectomy, with a good response in almost two tiers of patients. For those who failed to improve or who relapse other therapeutic options may be continuous positive pressure (CPAP) or noninvasive ventilation (NIV).

Definition and incidence

Obstructive sleep disordered breathing (SDB) is classically defined as a syndrome of "upper airway dysfunction during sleep, characterized by snoring and/or increased respiratory effort secondary to increased upper airway resistance and pharyngeal collapsibility "(1). Therefore, SDB should be considered as a continuum clinical spectrum, ranging from primary snoring (PS) to upper airway resistance syndrome (UARS), obstructive hypoventilation (OH) and finally obstructive sleep apnoea syndrome (OSAS).

The prevalence of SDB is somewhat difficult to ascertain as definitions varied over time.

PS is defined as the presence of habitual snoring for more than 3 nights per weeks, in the absence of respiratory events (namely apneas and hypopneas), sleep disruption and gas exchange abnormalities. Since there is no universally accepted standard definition of snoring, its exact prevalence varies according to the different perceptions of word's meaning. As a matter of fact, differences in loudness and frequency of the phenomenon (i.e. number of nights per week, number of hours per night) have led to differences in the estimation of the prevalence of PS. The prevalence of PS, gathered via questionnaires, ranges from 1.5% to 21% in paediatric population(2) according to the different definitions. However, recent meta-analyses estimated a median prevalence of PS in children worldwide of around 11-12% (3). As such, PS is common and, since it represents the less severe form of SDB, it has long time been considered as a benign disorder. However, recent data suggest a decrease of cognitive functions even in children with PS(4), rising the need for increased awareness regarding the consequences of the whole spectrum of SDB.

OSAS is the most severe form of SDB and is defined as the "recurrent partial or complete upper airway obstruction (hypopneas, obstructive or mixed apneas) with disruption of normal oxygenation, ventilation and sleep pattern"(2). The prevalence of OSAS in children varies widely from 0.1 to 13%(5). This variability probably derives from the different definitions and severity thresholds used over time. However, it is universally accepted that the prevalence of OSAS in children ranges between 2 to 4%(6). OSAS is thus a relatively common disease in children.

OSAS is classically associated with tonsillar hypertrophy in otherwise healthy children, and most of the first literature data concerned this population. However, over time OSAS and SDB in general have been studied in other paediatric populations like obese children and children with congenital or genetic diseases. This revealed that OSAS is far more common in children with associated conditions like congenital craniofacial malformations, neurological and neuromuscular diseases, and metabolic disorders. These children are often younger and present a multifactorial disorder that requires objective assessment and treatment of all underlying abnormalities that contribute to upper airway obstruction during sleep(7). Estimated prevalence of OSAS in these diseases are shown in table 1.

✉ **Correspondence address:**
Dr. Alessandro Amaddeo, Institute for Maternal and Child Health IRCCS Burlo Garofolo, Via dell'Istria 65/1, 34100 Trieste, Italy, E-mail: Alessandro.amaddeo@burlo.trieste.it

Neuromuscular patients often exhibit OSAS in association with nocturnal hypoventilation due to progressive respiratory muscles weakness, with a prevalence of 65% in children with congenital myopathy and up to 30% in adolescents with Duchenne dystrophy(8). Even patients with Ehlers-Danlos syndrome may have OSAS, with a prevalence ranging from 32 to 100% (9).

Clinical manifestations of SDB

Clinical manifestations of SDB in children differ from adults. Children tend to have a much more varied constellation of symptoms, often difficult to recognize. In contrast to adults, normal children snore infrequently. This is consistent with the better preservation of upper airway patency in case of subatmospheric pressure. Excessive daytime sleepiness, which is a hallmark of poor sleep quality in adults, is rarely observed in children with SDB(10). Rather, children tend to be hyperactive. Also, symptoms evolve and change with age as shown in Table 2. Some symptoms are observed at any age, such as night-time awakenings, while other symptoms are more common in some age groups. Clinical examination is important. Since tonsillar hypertrophy is a major contributor to SDB in children, tonsils size should be evaluated with the Mallampati classification and the Friedman score (figure 1 and 2).

Table 1. Prevalence of OSAS in different congenital or genetic diseases of childhood associated with craniofacial malformations.

	Incidence	Prevalence of OSAS
Craniofacial abnormalities		
– Pierre Robin Sequence	1/8000-14000	80-90%
– Goldenhar syndrome	1/5600	80-90%
– 22q11.2 deletion syndrome	1/4000	60%
– CHARGE syndrome	1/8500	65%
– Treacher Collins syndrome	1/50000	50-90%
Craniostenosis	1/2500	75-90%
Down syndrome	1/700	30-60%
Ehler-Danlos syndrome	1/10000	30-100%
Prader Willi syndrome	1/25000	50%
Achondroplasia	1/25000	60-90%
Mucopolysaccharidosis	1/65000	60-95%

Diagnosis

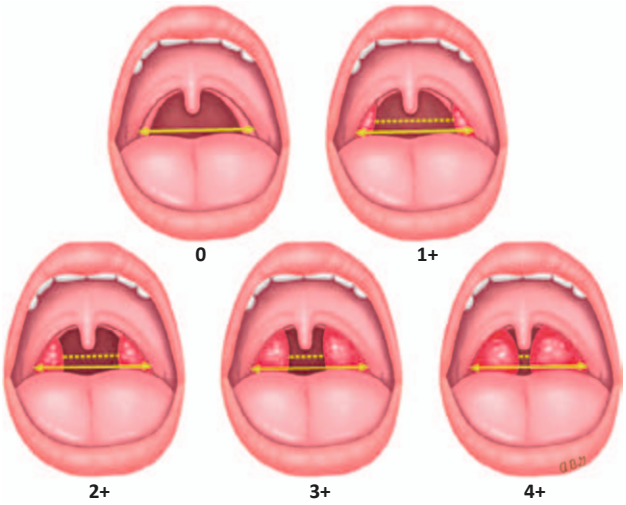
Nocturnal, attended, laboratory polysomnography (PSG) is the gold standard for diagnosis of SDB. However, due to limited resources and to simplify the diagnosis of OSAS, other simpler measurements have been proposed over time. Nevertheless, the diagnosis of OSAS in children derives from the combination of history, symptoms, clinical examination, and a sleep study. According to the recommendations of the AASM, the diagnostic criteria for OSAS in children, which includes also upper airway resistance syndrome and obstructive hypoventilation, are:

Clinical criteria ("A criteria") – The presence of one or more of the following clinical symptoms (reported by the parents or caregivers):

- Snoring
- Laboured, paradoxical, or obstructed breathing during the child's sleep
- Sleepiness, hyperactivity, behavioural problems, or learning problems

Table 2. Symptoms of sleep disordered breathing in children by age

Infants (3-12 months)	Toddlers (1-3 yr)	Pre-school (3-5 yr)	School (5-18 yr)
- Noisy breathing - Witnessed apneas - Frequent arousals - Difficult breathing - Mouth breathing - Nocturnal sweating - Failure to thrive - Nasal congestion - Hyperextended neck - Poor sucking - Stridor - Breathholding spells	- Snoring - Witnessed apneas - Frequent arousals - Difficult breathing - Mouth breathing/dry mouth - Nocturnal sweating - Failure to thrive - Nasal congestion - Hyperextended neck - Restless sleep - Sleep terrors - Confusional arousal - Irritability - Daytime sleepiness	- Snoring - Witnessed apneas - Frequent arousals - Mouth breathing/dry mouth - Nocturnal sweating - Failure to thrive - Nasal congestion - Hyperextended neck - Sleep terrors - Confusional arousal - Sleepwalking - Daytime sleepiness/ persistent naps - Restless sleep - Enuresis - Hyperactivity, inattention - Difficulty waking up in morning - Morning headache - Sleep in knee-chest position	- Snoring - Witnessed apneas - Frequent arousals - Mouth breathing/dry mouth - Nocturnal sweating - Failure to thrive - Nasal congestion - Hyperextended neck - Nightmares - Sleeptalking - Confusional arousal - Sleepwalking - Daytime sleepiness - Restless sleep - Enuresis - Hyperactivity, inattention - Difficulty waking up in morning - Morning headache - Insomnia - Learning difficulties - Delayed puberty - Mood disorders - Hypertension



Tonsil size is graded on a scale from 0 to 5:

0 – Tonsils are entirely within the tonsillar pillar, (or previously removed by surgery).

1+ – Tonsils occupy less than 25 percent of the lateral dimension of the oropharynx, as measured between the anterior tonsillar pillars (solid yellow arrow).

2+ – Tonsils occupy 26 to 50 percent of the lateral dimension of the oropharynx.

3+ – Tonsils occupy 51 to 75 percent of the lateral dimension of the oropharynx.

4+ – Tonsils occupy more than 75 percent of the lateral dimension of the oropharynx.

Adapted from reference[25].

Figure 1. Tonsils grading by the Mallampati score

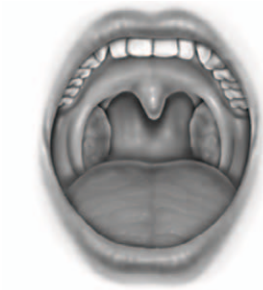


Fig 1. Friedman Palate Position I allows visualization of the entire uvula and tonsils/pillars.

Fig 2. Friedman Palate Position II allows visualization of the uvula but not the tonsils.

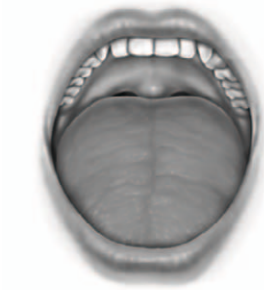


Fig 3. Friedman Palate Position III allows visualization of the soft palate but not the uvula.

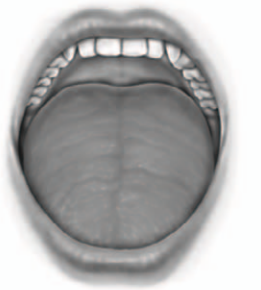


Fig 4. Friedman Palate Position IV allows visualization of the hard palate only.

Adapted from reference[26]

Figure 2. Grading of the pharyngeal space by the Friedman score.

Polysomnographic criteria ("B criteria") – The PSG demonstrates one or both of the following:

- One or more obstructive apnoea, mixed apnoea, or hypopnoeas, per hour of sleep.
- A pattern of obstructive hypoventilation, defined as at least 25 percent of total sleep time spend with hypercapnia ($PtcCO_2$ >50 mmHg) in association with one or more of the following:
- Snoring
- Flattening of the nasal pressure waveform
- Paradoxical thoraco-abdominal motion

AHI is calculated as the total number of apneas and hypopneas scored during the recording, divided by the total sleep time. This index is universally used to assess the severity of OSAS. Normative referenced values for healthy children have been published and have led to the following grading of OSAS severity.

The following categories are classically used as an index of severity(11):

- Mild OSAS – AHI 1 to 4.9 events/h,
- Moderate OSAS – AHI 5 to 9.9 events/h
- Severe OSAS – AHI >10 events/h

As these cut off values have not been validated against end-organ morbidity in children in the literature, especially in infants and toddlers, some authors consider that a AHI of more than 5 events/h should be considered as severe (1). The main raisons for this lower cut-off is that even children with an AHI >5 events/h may have an increased risk of elevated blood pressure as compared with healthy controls (12) and a lower rate of spontaneous improvement.

Treatment

Once the diagnosis of OSAS has been made, the decision to initiate treatment is taken on an individual basis. Anticipated benefits and risks of treatment must be carefully weighed. Classically, an AHI of more than 5/h is considered as the cut-off for treatment, irrespective of the presence of comorbidities. However, as stated before, recent evidence suggest that even primary snoring may be associated with neurocognitive dysfunction and cardiovascular stress. On the other hand, the decision to treat is challenged by the fact that a large group of pre-adolescent with OSAS (and without obesity) improve spontaneously over time (13). Finally, important considerations include the child's age, the severity of PSG abnormalities, and any underlying issue or complications related to OSAS. In conclusion, the decision to treat a child with OSAS relies on several factors and the best treatment strategy should be decided by a clinician having an expertise in pediatric sleep-related respiratory abnormalities. Since adenotonsillar hypertrophy is the main cause of OSAS in

otherwise healthy children, adenotonsillectomy represents the treatment of choice in this age group.

Children who fail to respond to this treatment may benefit from other therapeutic options like continuous positive pressure (CPAP) or noninvasive ventilation (NIV).

CONFLICT OF INTEREST

The author declare that he has no conflict of interest with this manuscript.

REFERENCES

1. *Kaditis AG, Alonso Alvarez ML, Boudewyns A, Alexopoulos EI, Ersu R, Joosten K, et al.* Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management. European Respiratory Journal. 2016 Jan;47(1):69–94.
2. *Marcus CL, Brooks LJ, Ward SD, Draper KA, Gozal D, Halbower AC, et al.* Diagnosis and management of childhood obstructive sleep apnea syndrome. Pediatrics. 2012;130(3): e714–55.
3. *Bonuck KA, Chervin RD, Cole TJ, Emond A, Henderson J, Xu L, et al.* Prevalence and Persistence of Sleep Disordered Breathing Symptoms in Young Children: A 6-Year Population-Based Cohort Study. Sleep. 2011 Jul;34(7):875–84.
4. *Smith DL, Gozal D, Hunter SJ, Kheirandish-Gozal L.* Frequency of snoring, rather than apnea–hypopnea index, predicts both cognitive and behavioral problems in young children. Sleep Medicine. 2017 Jun;34:170–8.
5. *Bixler EO, Vgontzas AN, Lin HM, Liao D, Calhoun S, Vela-Bueno A, et al.* Sleep disordered breathing in children in a general population sample: prevalence and risk factors. Sleep. 2009;32(6):731–6.
6. *Lumeng JC, Chervin RD.* Epidemiology of pediatric obstructive sleep apnea. Proc Am Thorac Soc. 2008 Feb 15;5(2):242–52.
7. *Kaditis AG, Alonso Alvarez ML, Boudewyns A, Abel F, Alexopoulos EI, Ersu R, et al.* ERS statement on obstructive sleep disordered breathing in 1- to 23-month-old children. Eur Respir J. 2017 Dec;50(6).
8. *Suresh S, Wales P, Dakin C, Harris MA, Cooper D (Gus) M.* Sleep-related breathing disorder in Duchenne muscular dystrophy: Disease spectrum in the paediatric population. Journal of Paediatrics and Child Health. 2005 Sep 1;41(9–10): 500–3.
9. *Guilleminault C, Primeau M, Chiu H yean, Yuen KM, Leger D, Metlaine A.* Sleep-disordered breathing in Ehlers-Danlos syndrome: a genetic model of OSA. CHEST Journal. 2013; 144(5):1503–11.
10. *Gozal D, Wang M, Pope DW.* Objective sleepiness measures in pediatric obstructive sleep apnea. Pediatrics. 2001 Sep;108(3): 693–7.
11. *Roland PS, Rosenfeld RM, Brooks LJ, Friedman NR, Jones J, Kim TW, et al.* Clinical practice guideline: polysomnography for sleep-disordered breathing prior to tonsillectomy in children. Otolaryngology–Head and Neck Surgery. 2011;145 (1_suppl):S1–15.
12. *Amin R, Somers VK, McConnell K, Willging P, Myer C, Sherman M, et al.* Activity-adjusted 24-hour ambulatory blood pressure and cardiac remodeling in children with sleep disordered breathing. Hypertension. 2008 Jan;51(1):84–91.
13. *Marcus CL, Moore RH, Rosen CL, Giordani B, Garetz SL, Taylor HG, et al.* A Randomized Trial of Adenotonsillectomy for Childhood Sleep Apnea. N Engl J Med. 2013 Jun 20;368(25): 2366–76.
14. *Brodsky L.* Modern assessment of tonsils and adenoids. Pediatr Clin North Am. 1989 Dec;36(6):1551–69.
15. *Friedman M, Ibrahim H, Bass L.* Clinical staging for sleep-disordered breathing. Otolaryngol Head Neck Surg. 2002 Jul; 127(1):13–21.