Indications for polysomnography in children – practical recommendations for the paediatrician

Mieke Boon [a], Marijke Proesmans[a], Katrien Jansen [b]

[a] Paediatric Pulmonology, Department of Paediatrics, University Hospital Leuven
[b] Paediatric Neurology, Department of Paediatrics, University Hospital Leuven

mieke.boon@uzleuven.be

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Abstract

Sleep problems are frequent in children. The diagnosis of most sleep-related disorders starts with a detailed history and clinical examination.
Polysomnography (PSG) is the gold standard for objective measurement of neurologic and respiratory function during sleep, combining multiple data-collecting sensors.
Sleep-related breathing disorders are frequent, and PSG is needed for the diagnosis and quantification of obstructive sleep apnoea syndrome. Moreover, more rare disorders like central apnoea and hypoventilation can correctly be identified.
When narcolepsy is suspected, a multiple sleep latency test must be added to a PSG to objectify hypersomnolence.
This review describes the respiratory and neurologic indications for PSG in children and some practical and ethical considerations that must be taken into account when referring children for PSG.

Introduction

Sleep problems are frequent: 1/3-1/5 of the paediatric population experiences short or long term sleep problems. A real sleep disorder is present in 1-6% of healthy children, with an ever higher incidence in children with neurodevelopmental disorders.

The diagnosis of sleep disorders starts with a detailed history, both on sleep and daytime issues. The acronym BEARS describes the main issues to be discussed: bedtime behaviour, excessive daytime somnolence, awakenings during the night, the regularity and duration of sleep and the presence of snoring must be assessed. A general clinical history and physical examination are needed to detect underlying diseases, known risk factors or secondary effects of sleep disorders.

Polysomnography (PSG) is the gold standard diagnostic technique to evaluate sleep-related disorders, in combination with a full clinical evaluation. It is a complex test that evaluates all aspects of normal sleep (1,2).

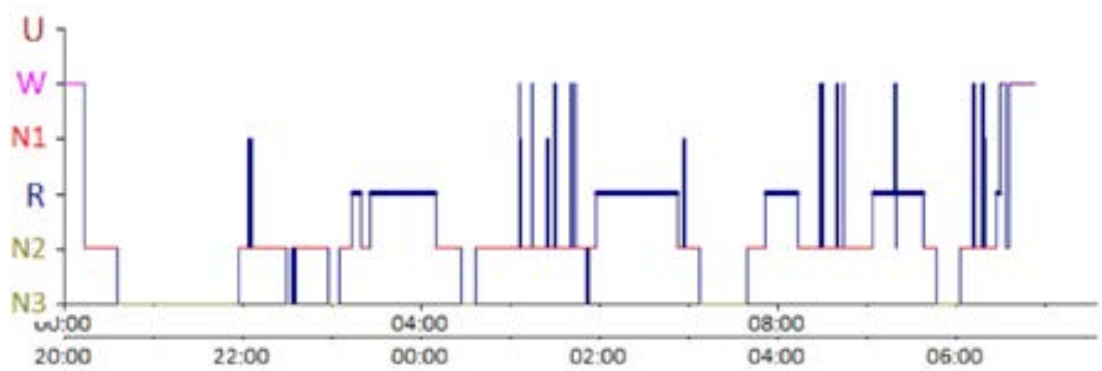
Because PSG is time-consuming, costly and quite invasive for young children, it is important to understand its strengths, limitations and clinical utility.

In this review, we will discuss the indications for childhood PSG apart from prematurity and infancy. We will not discuss the technical requirements and interpretation/scoring of paediatric PSG in detail.

What is measured?

The American Association of Sleep Medicine (AASM) has formulated strict guidelines on how to perform, evaluate and score polysomnography (3). First, a complete neurological evaluation is performed, with simplified electroencephalography (EEG) monitoring, mostly with 8-20 electrodes. Electrooculography (EOG) is performed with one electrode placed lateral from each eye, electromyography (EMG) mostly uses a submental and bilateral tibial electrode. Based on the EEG, EOG and EMG interpretation, sleep can be divided in sleep stages N1 (transition from wake to sleep, very short-lasting), N2, N3 (deep sleep) and REM (dream stage of sleep, characterized by rapid eye movements and minimal muscle tone). Arousals are short disturbances in sleep, due to varying causes. These different sleep stages appear in sleep cycles from 90-100 minutes. A hypnogram is the overview of all sleep cycles and sleep stages during the night and gives an overview of the sleep quality (Figure 1).

Figure 1: *Example of a hypnogram from a 7-year old child. The different sleep stages can visually be identified: wakefulness (W), stage N1, N2 and N3, and REM (R)-sleep. N3 is typically more abundant in the first part of the night, REM-sleep typically increases towards the morning. Short arousals are seen as very short periods of wakefulness.*



For the respiratory evaluation during sleep, respiratory muscle activity is evaluated by movement of thoracic and abdominal wall and by airflow measured at the nose and the mouth. In this way, interruptions of breathing (apnoea) can be detected by a drop of >90% in muscle activity or flow, hypopnea by a drop of 50-90% in muscle activity or flow. If the activity of all muscle groups decreases, the apnoea/hypopnea is of central origin, if thoracic and abdominal movement is preserved (can be paradoxical) with absent nasal and buccal flow, the apnoea/hypopnea is obstructive in origin. Age-appropriate scoring rules must be used. Additionally, oxymetry and electrocardiogram monitoring can detect subsequent desaturations and bradycardia, arousals can be identified on the EEG. Sound and video recordings (with infrared camera) are used to interpret noises (snoring, stridor,…) or abnormal movements (seizures, periodic limb movements,…). Registering body position can be helpful to identify position-related phenomena. The apnoea/hypopnea index (AHI) is defined as the number of detected apnoea/ hypopneas per hour of sleep and is most frequently used as objective measure of sleep related breathing disorders. If night time hypoventilation is suspected, additional percutaneous or end-tidal CO_2 monitoring must be applied.

More abbreviated versions (simple overnight oximetry polygraphy which is a full respiratory evaluation without EEG monitoring) can be used to screen for sleep disorders and can even be applied in home settings, but are less sensitive and less specific for diagnostic purposes and evidence for their use is limited.

Respiratory indications for polysomnography in children

Physiology of respiration during sleep (Figure 2)

In healthy individuals, normal sleep is characterized by a generalized decrease of muscle tone. In combination with a reduced lung compliance, this leads to a reduction of the functional residual capacity, leading to micro-atelectasis and ventilation perfusion mismatch. Besides, decrease of the pharyngeal and tongue muscle tone causes an increase of the upper airway resistance. On top of the generalized decrease in muscle tone, the central ventilator drive decreases, resulting in a decrease of minute ventilation by around 10%. Overall, this results in a slight increase of Pa_{CO2} of 2-8 mmHg and a decrease of Pa_{O2} of 3-11 mmHg compared to wakefulness. These effects are most pronounced during REM-sleep, as muscle tone is at its lowest level.

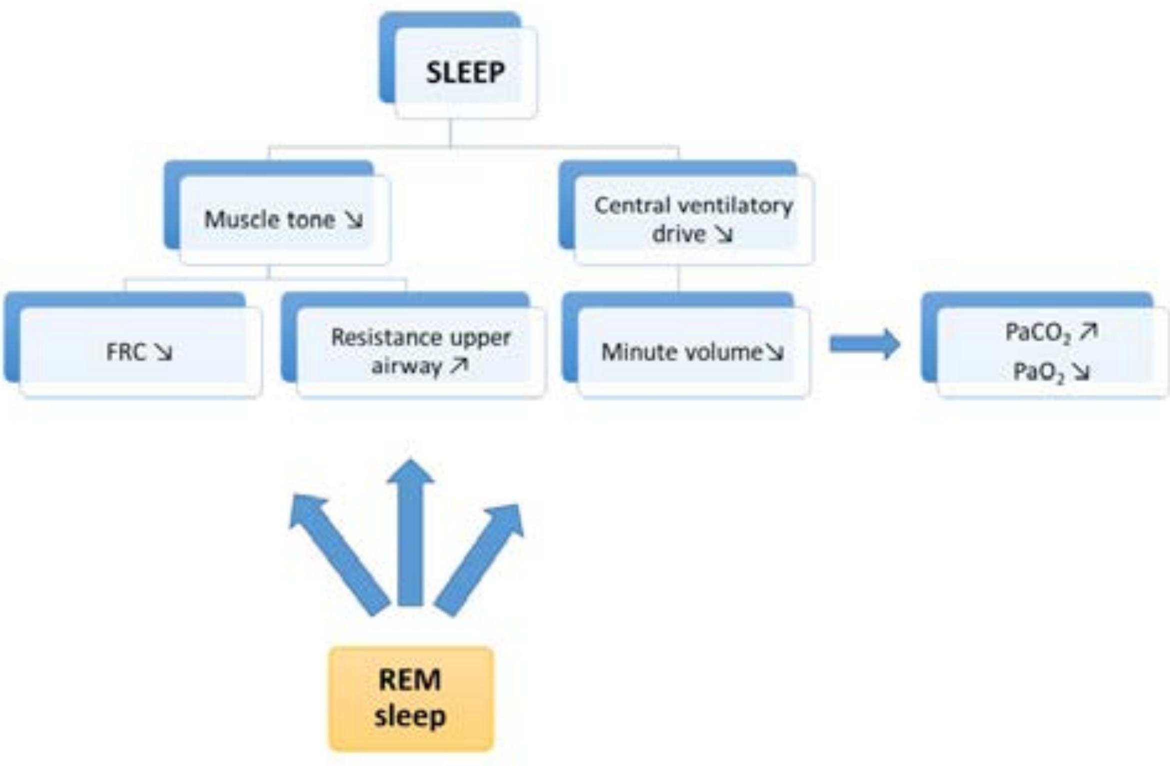
OSAS

Because of generalized decreased muscle tone during sleep compared to wakefulness, the resistance in the upper airway increases and this can lead to snoring (vibration of the weak tissues of the upper airway) or complete collapse of the upper airway (Figure 2). Sleep disordered breathing (SDB) is a spectrum of disorders caused by upper airway dysfunction during sleep, characterized by snoring and/or increased respiratory effort secondary to increased upper airway resistance and pharyngeal collapsibility (4). When recurrent events of partial or complete upper airway obstruction occur, resulting in disruption of normal oxygenation, ventilation and sleep pattern, this is defined as obstructive sleep apnoea syndrome (OSAS) (4). OSAS is frequent in the paediatric population, with a reported prevalence between 1 and 4% (5).

Symptoms of OSAS are snoring, open mouth breathing, abnormal sleep position with extended neck or witnessed apnoeas, although the correlation between the presence of snoring and OSAS is bad. The aetiology of OSAS is multifactorial, but the most frequent causes in otherwise healthy children are adenotonsillar hypertrophy and obesity. Additional risk factors (congenital or acquired) are described in table 1 (4,6) growth delay, behavioural problems.

Screening for OSAS by means of PSG is indicated if suggestive symptoms are present: snoring, witnessed apnoea, unexplained fatigue, excessive daytime sleepiness or concentration problems, morning headaches and nausea, disturbed or restless sleep, persisting enuresis. In isolated cases, OSAS can be an aggravating factor in growth retardation and feeding difficulties, bad asthma control, systemic or pulmonary hypertension, metabolic syndrome, frequent otitis media and attention deficit and hyperactivity disorder (ADHD). For

Figure 2: *Physiology of normal respiration during sleep.*



certain populations (marked by * in Table 1), screening PSG is recommended, sometimes even irrespective of symptoms (7,8)no recent evidence-based practice parameters have been reported. These practice parameters are the first of 2 papers that assess indications for polysomnography in children. This paper addresses indications for polysomnography in children with suspected sleep related breathing disorders. These recommendations were reviewed and approved by the Board of Directors of the American Academy of Sleep Medicine. Methods: A systematic review of the literature was performed, and the American Academy of Neurology grading system was used to assess the quality of evidence. Recommendations for PSG Use: 1. Polysomnography in children should be performed and interpreted in accordance with the recommendations of the AASM Manual for the Scoring of Sleep and Associated Events. (Standard.

It is important to quantify the severity of OSAS, as the risk of long-term morbidity (systemic and pulmonary hypertension, enuresis, cognitive and behavioural effects) are correlated to the severity of OSAS. An obstructive AHI ≥1 in the presence of suggestive symptoms is diagnostic of OSAS. Mild OSAS (obstructive AHI between 1 and 5) based on adenotonsillar hypertrophy can spontaneously improve, as was demonstrated in a large randomized controlled trial studying the effect of adenotonsillectomy in children with OSAS (9). If symptoms are invalidating and persisting, treatment can be considered. Moderate-to-severe OSAS (AHI>5) is unlikely to resolve spontaneously, predictive of long term morbidity (mainly cardiovascular) and needs intervention.

PSG can be performed to confirm the presence and severity of OSAS preoperatively before adenotonsillectomy is executed, as it is known that clinical symptoms badly correlate with the severity of OSAS (10). Contrary, there is a good correlation between the severity of OSAS and the perioperative risk for complications. PSG can be used for re-evaluation after adenotonsillectomy, for example in patients with moderate to severe OSAS, or to exclude incomplete resolution of OSAS in patients with additional risk factors (4). Those patients will need additional treatment, for example with continuous positive pressure (CPAP) or maxillofacial surgery.

In patients treated with CPAP therapy, PSG should be used for titration of the initial CPAP pressure, and for follow-up, as growth or changes in the airway morphology may impact the needed pressure for optimal control of OSAS. For patients needing extensive surgery like rapid maxillary expansion and more specialized craniofacial surgery, postoperative PSG is warranted to evaluate residual OSAS.

Central apnoea's

Congenital central hypoventilation (CCHS) is a very rare genetic condition, based on *PHOX2B* mutations, that is characterized by an abnormal central control of ventilation, resulting in central apnoeas, night time hypoventilation and other autonomic disorders (11). PSG with capnometry is indicated for diagnosis and follow-up of CCHS.

Patients with Chiari malformation or other intracranial causes of pressure on the brain stem (for example brain stem tumours, skeletal dysplasia, lysosomal storage diseases) are at specific risk for the development of central sleep apnoea, but also obstructive and mixed sleep apnoea. PSG can detect respiratory disturbances and can be used to evaluate the effect of neurosurgical intervention.

Night time hypoventilation

In children with neuromuscular disorders, further decrease of muscle tone during sleep can result in sleep-related hypoventilation (decrease of inspiratory respiratory muscle force) and/or OSAS (weakness of pharyngeal muscles resulting in upper airway collapse). A high prevalence of obesity due to immobility may further increase the risk of OSAS.

Symptoms of night time hypoventilation can be daytime fatigue or hypersomnolence, morning headaches, nausea or drowsiness, hyperactivity, cognitive problems, irritability, frequent arousals with poor sleep quality, recurrent respiratory infections, failure to thrive and cor pulmonale. However, symptoms are nonspecific and underreported.

Table 1: *Risk factors for OSAS in children can be divided in 3 categories: general risk factors, structural risk factors and factors influencing the upper airway tone. Factors marked in bold or the most frequent risk factors in otherwise healthy children.*

General risk factors	Structural risk factors	Upper airway tone
Allergic rhinitis	**Adenotonsillar hypertrophy**	**Obesity**
Prematurity	Nasal septum deviation	Neuromuscular disease*
Familial OSAS	Hypertrophic conchae	Cerebral palsy - epilepsy
♂ **>** ♀	Laryngotracheamalacia	Arnold-Chiari malformation*
African-American ethnicity	Subtle craniofacial abnormalities in non-syndromic children: – Dolichocephaly – teeth malocclusion – retrusive chin – steep mandibular plane – vertical direction of craniofacial growth	Prader Willi syndrome*
Passive smoking	Complex craniofacial abnormalities: – mandibular hypoplasia (f.e. Pierre Robin sequence*) – midfacial hypoplasia – Treacher-Collins, Crouzon, Apert, Pfeiffer* – Palatal cleft – Achondroplasia* – Mucopolysaccha-ridosis* – Choanal atresia – Beckwidth-Wiede-mann syndrome – Marfan Syndrome – Ehlers-Danlos	Down syndrome*

In children with progressive neuromuscular diseases, it is suggested that the risk for sleep disordered breathing dramatically increases when forced vital capacity (FVC) drops below 60% (12–14). Therefore, guidelines suggest to perform systematic screening PSGs in this group irrespective of the presence of symptoms, always in combination with CO_2 monitoring (15). Moreover, loss of (or inability to attain) ambulation, severe diaphragmatic weakness and rigid spine syndrome are regarded as additional risk factors for the development of night time hypoventilation (15).

The respiratory status in patients with neuromuscular disorders can further be compromised by progressive scoliosis, chronic aspiration, recurrent and chronic infections, etc. When invasive surgery with general anaesthesia is needed (e.g. scoliosis surgery), PSG is advised in the pre-operative evaluation in patients with severe restrictive lung disease.

Other patient groups with severe generalized hypotonia and risk of development of night time hypoventilation are: cerebral palsy, Prader Willi syndrome, Down syndrome, acquired hypotonia due to steroid myopathy, diaphragmatic paralysis, metabolic decompensation, excessive sedative drug use, high cervical lesion,….

Severe restrictive lung disease from respiratory (end-stage cystic fibrosis, bronchopulmonary dysplasia, interstitial lung disease, …) or non-respiratory origin (congenital diaphragmatic hernia, skeletal dysplasia, (kypho)scoliosis) can lead to night time hypoventilation and children with these disorders should be screened by PSG.

Once ventilation is started, PSG can be used for optimal titration of the ventilator settings (ventilatory pressure, frequency, synchronisation, to identify leaks, etc) and to adapt the settings in function of growth and improvement or worsening of the ventilatory capacity in function of the underlying disease.

Non-Respiratory indications for polysomnography in children

Narcolepsy

Narcolepsy is a neurologic disorder that is characterized by daytime hypersomnolence (recurrent attacks of irresistible daytime sleepiness) and cataplexy (sudden loss of muscle control in the legs, trunk, face or neck in response to emotional stimuli). Night time sleep is typically fragmented, and REM-sleep can occur abnormally early. The diagnosis of narcolepsy can be made based on a typical clinical history, in combination with a PSG with a multiple sleep latency test (MSLT) (16–18)and assessment requires a thorough history and in many cases, objective assessment in the sleep laboratory. These practice parameters were developed to guide the sleep clinician on appropriate clinical use of the Multiple Sleep Latency Test (MSLT. MSLT is feasible and can be interpreted reliably from the age of 5 years. After a normal night with PSG recording, the patient is given the opportunity to do 4-5 daytime naps of 20 minutes every 2 hours. The time needed to fall asleep (sleep latency) is measured and the time to REM onset is also evaluated. A mean sleep latency below 8 minutes in combination with at least one SOREMP (sleep onset REM periods) is diagnostic for narcolepsy, although normative data for children and adolescents are scarce. The PSG is needed to exclude other causes of hypersomnolence like severe OSAS and can illustrate the abnormal sleep architecture in these patients. PSG alone is not sufficiently sensitive for the diagnosis of narcolepsy.

An MSLT cannot be interpreted correctly in patients with sleep deprivation or in patients that are treated with neurostimulatory drugs.

Parasomnia

Parasomnia is a phenomenon of abnormal movements and behaviour during sleep, as a consequence of transition between different sleep stages or between sleep and wakefulness. It is relatively frequent in the paediatric population, but is mostly harmless and disappears with age.

A suspicion of parasomnia is not a primary indication to perform PSG. Mostly, a detailed history can sufficiently adequately diagnose typical parasomnia in children (e.g. sleep terrors, sleep walking, rhythmic movement disorders,…) (17). A video recording made by the parents can be very helpful to have a good view on the specific type of the parasomnia. Only when the parasomnia is atypical, dangerous, or suggestive of nocturnal epilepsy, or when there is a suspicion of concomitant OSAS, a PSG can be indicated.

Restless legs syndrome

Restless legs syndrome (RLS) is a clinical diagnosis that includes sensorimotor discomfort of the legs, worsening in the evening hours, particularly when the child is at rest. PSG can identify abnormally frequent (>5/h) periodic limb movements (PLMs) in most patients with RLS, and can support the diagnosis in this way, as parental reporting of movements is unreliable (17).

Delayed sleep phase syndrome

The diagnosis of circadian rhythm disorders such as delayed sleep-wake phase disorder is based on a detailed history in combination with a sleep diary of at least one week (weekend included). Actigraphy, a measurement of movement during the night, can be used to support the diagnosis. PSG does not show typical abnormalities and doesn't contribute to the diagnosis.

Insomnia

Insomnia is not an indication for PSG, only in exceptional cases, for example when there is a suspicion of a discordance between objective and perceived sleep duration. Sleep history with a detailed sleep diary is mostly sufficient for a correct diagnosis and adequate treatment.

Practical considerations and limitations

Performing a PSG of good quality is not easy in young children and it is regarded as quite invasive. Especially children with limited mental capacities are not able to understand the need for all applied electrodes. Therefore, the examination should be performed in optimal circumstances; good information should be provided in advance by a brochure or a visit to the sleep lab; presence of a parent at the moment of application of all electrodes and during the night; presence of a lab technician or nurse with experience and patience in dealing with children and a child-friendly environment are important factors to be considered.

PSG is typically a one-night examination, with the possibility of a reduced sleep quality because of external factors (noise, unfamiliar environment, fear) and the risk of missing non-frequent night time events. However, studies have shown that the variability in respiratory evaluation between different nights is limited and not clinically relevant in most cases.

In children, there is insufficient evidence for the use of home PSG for diagnostic purposes (19). Similarly, nap or abbreviated polysomnography has been proven insufficiently sensitive. In children needing oxygen treatment, full PSG is generally not indicated, unless there is an additional suspicion of sleep related breathing disorders or hypoventilation (7)no recent evidence-based practice parameters have been reported. These practice parameters are the first of 2 papers that assess indications for polysomnography in children. This paper addresses indications for polysomnography in children with suspected sleep related breathing disorders. These recommendations were reviewed and approved by the Board of Directors of the American Academy of Sleep Medicine. Methods: A systematic review of the literature was performed, and the American Academy of Neurology grading system was used to assess the quality of evidence. Recommendations for PSG Use: 1. Polysomnography in children should be performed and interpreted in accordance with the recommendations of the AASM Manual for the Scoring of Sleep and Associated Events. (Standard).

It must be realized that PSG is different from EEG: the electrodes used are different, another reading frame is used (10 sec for EEG versus 30 sec for PSG), and the reader is different. Clear seizures can be recognized on PSG, but subtle epileptic activity can be missed. If there is a suspicion of epilepsy, an extended EEG monitoring could be applied.

PSG provides a lot of information, but cannot give answers to questions such as 'Why does a child wake up?', 'Why is a child tired?', 'Why is sleep quality bad?', 'What is a child dreaming?', 'Are there night mares?'. Isolated fatigue, nightmares, fibromyalgia, parasomnia or a disturbed sleep without indication of OSAS, epilepsy or underlying disease are therefore no good indication for a PSG in children. Whenever a child is referred for PSG, a clear explanation of the indication will allow the reader to provide a correct and complete protocol.

Ethical considerations

When a PSG is planned, the impact on therapeutic decisions should be questioned in advance. For otherwise healthy children, it is obvious that an abnormal result will be followed by an adequate treatment.

For severely disabled children (for example cerebral palsy with severe mental disability), caregivers should evaluate whether the treatment plan would be changed in function of the result of the PSG. In patients with a limited life expectancy, would quality of life or prognosis change? Treatment options for OSAS in this population are often limited: a considerable number of patients need orthopaedic appliances for abduction of the hips and therefore choices have to be made between optimal sleep position for respiratory (lateral decubitus for optimal respiration and reduction of upper airway obstruction) or orthopaedic reasons (dorsal decubitus). Similarly, CPAP therapy is not always efficacious because of open mouth breathing, or may not be tolerated because of drooling, risk of aspiration, uncontrolled movements and epilepsy. A multidisciplinary approach is absolutely needed, and parents should be counselled in advance. The risk of treatment and abstinence of treatment should openly be discussed.

Conclusions

Paediatric PSG is a very useful technique that provides extensive information on the neurologic and respiratory activity during sleep. It is indicated for the diagnosis of OSAS and narcolepsy, and can be helpful for detecting other sleep-related disorders. Correct interpretation requires expertise in paediatric sleep medicine. Caregivers should be aware of the utility and limitations of PSG, especially in children with underlying disorders.

Conflict of interest

The authors have no conflict of interest to declare.

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