



## Australasian Sleep Association clinical practice guidelines for performing sleep studies in children



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### 1. Executive summary

This clinical practice guideline, intended for use by paediatric respiratory and sleep practitioners, provides best practice recommendations for conducting paediatric sleep studies in Australia and New Zealand; it is applicable for children up to the age of 18 years. In particular this guideline provides recommendations in relation to the investigation of sleep-related respiratory disorders; non-respiratory causes of excessive daytime sleepiness; sleep-related movement disorders and the occurrence of abnormal or unusual behaviours during sleep. Clinical indications for diagnostic, interventional or follow-up sleep studies including the multiple sleep latency test (MSLT) are outlined. The recommendations specified in this document represent the consensus view of a committee of experienced paediatric sleep practitioners selected by the Australasian Sleep Association (ASA) and will require regular revision and modification as sleep medicine and technology continues to evolve.

Standards and indications for polysomnography (PSG) in children were first published by the American Thoracic Society [1] and

more recently by the American Academy of Sleep Medicine [2]. These documents outline evidence-based recommendations regarding technical aspects of sleep study recording and clinical situations where polysomnography is or is not indicated. These publications were considered by the panel when formulating recommendations in the current document and where necessary modifications have been made for Australasian practice. Where available, randomised controlled trials have been cited to substantiate recommendations.

This clinical practice guideline is not intended as the only source of guidance in the assessment of sleep disorders in children. In particular it is noted that for all types of sleep studies the laboratory investigation is only one component of diagnosis. Clinical history and examination are as important and complementary to the sleep study. Treatment decisions should not be based on a test result alone. However, as is the case for any diagnostic test, it is crucial that the necessity and urgency of the test, and the subsequent results, are evaluated by a clinician with the necessary expertise and knowledge to interpret the findings accurately.

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### Abbreviations

AASM	American Academy of Sleep Medicine	MWT	Maintenance of Wakefulness Test
AHI	Apnoea–hypopnoea index	NATA	National Association of Testing Authorities
ALTE	Apparent life threatening event	NIV	Non-invasive ventilation
ANS	Autonomic nervous system	NMD	Neuromuscular disease
ASA	Australasian Sleep Association	NREM	Non-rapid eye movement
ASTA	Australasian Sleep Technologists Association	NPV	Negative Predictive Value
AT	Adenotonsillectomy	OAHI	Obstructive Apnoea–Hypopnoea Index
BPAP	Bilevel Positive Airway Pressure	OHS	Obesity Hypoventilation Syndrome
CCHS	Congenital Central Hypoventilation Syndrome	OSA	Obstructive Sleep Apnoea
CPAP	Continuous Positive Airway Pressure	PAP	Positive Airway Pressure
DS	Down Syndrome	PB	Periodic Breathing
ECG	Electro-cardiogram	PLMD	Periodic Limb Movement Disorder
EDS	Excessive daytime sleepiness	PPV	Positive Predictive Value
EEG	Electro-encephalography	PSG	Polysomnography
EMG	Electro-myography	PWS	Prader–Willi Syndrome
EOG	Electro-oculography	REM	Rapid eye movement
FRC	Functional Residual Capacity	RLS	Restless Legs Syndrome
FVC	Functional Vital Capacity	SDB	Sleep Disordered Breathing
GH	Growth Hormone	SIDS	Sudden Infant Death Syndrome
MSLT	Multiple sleep latency test	SpO <sub>2</sub>	Saturation of oxygen
		TcCO <sub>2</sub>	Transcutaneous carbon dioxide
		VNS	Vagal nerve stimulator

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### Terminology and intent

Throughout this document the term “*indicated*” is used for situations where it is generally accepted that, following clinical evaluation, PSG is an appropriate and valid investigation in the assessment and management of subjects with suspected sleep-disordered breathing or other specific sleep disorders.

### Summary of Recommendations

This clinical practice guideline makes the following major recommendations in relation to undertaking sleep studies in children:

#### **Obstructive Sleep Apnoea**

- PSG is indicated when clinical assessment suggests the diagnosis of obstructive sleep apnoea syndrome (OSA).
- Abbreviated testing is indicated in selected cases as a clinical adjunct tool for the assessment of children with suspected obstructive sleep apnoea (OSA) following paediatric sleep specialist review.
- PSG is indicated if clinical assessment suggests OSA and abbreviated testing is inconclusive for the diagnosis of obstructive sleep apnoea.
- PSG is indicated when the severity of OSA is in doubt or if there is significant risk of post-surgical complications (e.g., idiopathic thrombocytopenic purpura).
- PSG is indicated when clinical assessment suggests the presence of residual OSA following AT or other treatment modalities.
- PSG is indicated following AT in children where there is the presence of ongoing symptoms or other clinical features suggestive of persistent OSA, particularly in those with: moderate

to severe OSA pre-operatively, obesity, craniofacial abnormalities that pre-dispose to the development of upper airway obstruction, neurologic disorders (e.g., neuromuscular disorders, spina bifida, Chiari malformation, meningomyelocele) and high risk congenital conditions (e.g., Down Syndrome, Prader–Willi Syndrome). See Appendix 1.

#### **Central Sleep Apnoea and Non-Obstructive Hypoventilation**

- PSG is indicated if clinical evaluation or abbreviated testing suggests the presence of central apnoea that is: (i) unexpected for development/age (ii) of prolonged duration or (iii) associated with significant oxygen desaturation or frequent bradycardia, pallor change, sleep disturbance or other daytime sequelae.
- PSG is indicated in cases of apnoea of infancy that are unresponsive to treatment or unusually severe or persistent or if there is diagnostic doubt over the aetiology of events (i.e. obstructive versus central).
- PSG is indicated when clinical assessment suggests the diagnosis of Congenital Central Hypoventilation Syndrome (CCHS).
- PSG or abbreviated testing is indicated when clinical assessment or other investigations (e.g., pulmonary function tests) suggests the presence of non-obstructive hypoventilation due to neuromuscular disease (NMD), obesity, kyphoscoliosis or chest wall deformity.
- PSG or abbreviated testing is indicated pre-operatively in at-risk children with neuromuscular disorders undergoing major orthopaedic, thoracic or upper airway surgery to screen for undetected SDB which may be exacerbated by sedation or analgesics.

#### **Polysomnography for Children Receiving Respiratory Support**

- PSG is indicated for children receiving positive airway pressure support (CPAP or BPAP) to titrate the level of pressure support required and to periodically re-evaluate the appropriateness of settings, including synchrony between machine and patient.

- PSG or abbreviated testing is indicated for children treated with invasive or negative pressure ventilation to adjust ventilator settings.
- PSG is indicated for children with symptoms of sleep-disordered breathing following tracheostomy decannulation. PSG may be clinically useful in some children (e.g., achondroplasia, Pierre Robin Sequence) with tracheostomy capped or downsized as part of the evaluation prior to decannulation.
- PSG is indicated for children with tracheostomy for upper airway obstruction where the co-occurrence of another breathing disorder (e.g., central sleep apnoea) is suspected.
- PSG is not routinely indicated for the titration of supplementary oxygen in children with chronic lung disease except in patients who are known, suspected or considered at risk of developing hypoventilation, central apnoea or hypercapnia with supplemental oxygen.

### **Apparent Life Threatening Event (ALTE)**

- PSG is not indicated for infants as part of a routine evaluation for an uncomplicated ALTE.
- PSG is indicated for infants who have experienced an ALTE and where there is clinical suspicion of underlying sleep disordered breathing or if there is ongoing or unexplained oxygen desaturation.

### **Prader–Willi Syndrome (PWS) and Growth Hormone (GH) Therapy**

- PSG is indicated for children with PWS prior to and six to ten weeks following the commencement of GH treatment and thereafter as clinically indicated.

### **Children with underlying chronic lung disease**

- PSG is indicated for children with chronic lung disease (e.g., asthma, cystic fibrosis, bronchopulmonary dysplasia) or unexplained pulmonary hypertension if there is clinical suspicion of sleep-disordered breathing.

### **Movement Disorders**

- PSG is not routinely indicated to diagnose restless legs syndrome (RLS), with the diagnosis based on clinical features.
- PSG is indicated for children suspected of having periodic limb movement disorder (PLMD).

### **Parasomnias and Epilepsy**

- PSG is not indicated if clinical assessment is consistent with a typical childhood parasomnia.
- PSG is indicated if a parasomnia is atypical or if excessive daytime sleepiness or impaired daytime functioning is present that cannot be explained by other causes.

### **Hypersomnia and Narcolepsy**

- The MSLT preceded by overnight PSG is indicated in children as part of the evaluation for suspected narcolepsy and excessive daytime sleepiness.

## **2. Introduction and background**

The knowledge and practice of paediatric sleep medicine has expanded markedly over the past few decades. As a result there has been a concomitant increase in demand for the clinical investigation of sleep disorders, particularly with respect to overnight polysomnography (PSG). Polysomnography (also known as a Type 1 study) is the reference standard employed to diagnose and evaluate a range of sleep disorders and is defined as the simultaneous measurement and recording of multiple physiological and behavioural variables during sleep. Overnight PSG is generally conducted in a dedicated sleep laboratory or sleep centre, supervised by trained staff. During PSG physiological signals related to sleep state, ventilation, cardiac function, respiratory effort and body and leg movements are usually measured. Historically overnight PSG has been utilised in children largely to evaluate suspected sleep-disordered breathing. However, increasing recognition and understanding of the non-respiratory sleep disorders has seen the utilisation of PSG widen within the practice of paediatric sleep medicine.

This clinical practice guideline provides best practice recommendations for the respiratory and non-respiratory indications for conducting paediatric sleep studies in Australia and New Zealand and is applicable for children up to the age of 18 years. These recommendations represent the consensus view of a committee of experienced paediatric sleep practitioners selected by the Australasian Sleep Association (ASA). Individual conflicts of interest were declared before the review began and were vetted by the ASA Board and were declared to all other committee members.

This report highlights the expanding and evolving nature of sleep investigations. It stresses the central role of the expert clinician in establishing the indications for sleep investigations and in the interpretation of sleep study results. This document seeks to improve standards within Australian and New Zealand by encouraging an evidenced-based approach to the performance of sleep testing, by promoting an accepted and uniform set of definitions of sleep disordered breathing and by encouraging a high standard of laboratory quality control.

The document considers the clinical indications for paediatric sleep studies under the following topics:

- Types of sleep studies.
- Respiratory indications for sleep studies.
- Non-respiratory indications for sleep studies.
- Sleep laboratory facilities and personnel requirements.
- Methodological and technical considerations.

Detailed guidelines regarding the technical specifications for recording sleep and the scoring of sleep and related events are documented elsewhere [3,4].

### *2.1. Indications for a sleep study*

There are three broad indications for performing a sleep study:

- **Diagnostic:** to assist in the diagnosis and evaluation of the severity of a sleep disorder.
- **Assessment of an intervention:** to initiate, confirm the adequacy of, or titrate a treatment.

- **Follow-up/surveillance:** to evaluate the persistence, re-emergence or worsening of a sleep disorder or to evaluate the impact of a treatment over time as the child grows and develops.

### Diagnostic studies

Diagnostic studies can be used to:

- Identify sleep disordered breathing.
- Investigate sleep-related causes of excessive daytime sleepiness (EDS) such as narcolepsy or frequent arousals (e.g. periodic limb movements).
- Identify and investigate causes of abnormal sleep/wake patterns or sleep fragmentation.
- Delineate the aetiology of episodic, unusual or paroxysmal phenomena occurring during sleep (e.g. parasomnias versus seizures).

### Intervention studies

Intervention sleep studies are undertaken to initiate and/or titrate treatment for a sleep-related breathing disorder, such as continuous positive airway pressure or non-invasive ventilation. This enables the modality of treatment to be titrated directly in response to abnormal respiratory events observed during sleep so that the therapy provides optimal support for that patient.

### Follow-up studies

Where treatment for a sleep disorder has been instituted, it may be important to confirm the effectiveness of that treatment. Re-assessment may also be required if symptoms persist or reappear following treatment or if symptoms worsen in cases previously assessed as having primary snoring or mild obstructive sleep apnoea (e.g., if there is significant weight gain). For children receiving positive airway pressure support (e.g., CPAP, BPAP) routine follow up sleep studies are indicated to ensure that the efficacy of treatment is maintained with growth and development or to evaluate if therapy is still needed or can be withdrawn. In some cases follow up sleep studies may also be required to ensure that pharmacotherapy is effective or does not elicit or worsen a sleep disorder (e.g., growth hormone therapy in Prader–Willi Syndrome). Follow up PSG may also be indicated in some medical conditions where there is an increased risk of sleep-disordered breathing but where individuals may be asymptomatic (e.g., central sleep apnoea in achondroplasia).

Persistence of daytime sleepiness, despite treatment, may require additional or repeat testing to confirm satisfactory adherence to therapy, to establish objectively the level of ongoing daytime sleepiness and/or to rule out alternative causes of daytime sleepiness. The maintenance of wakefulness test (MWT) may be undertaken to evaluate the efficacy of treatment in overcoming daytime sleepiness.

### Other Sleep-related Tests

#### Multiple sleep latency test (MSLT)

As an adjunct to overnight PSG, a MSLT may be conducted to objectively evaluate an individual's level of daytime sleepiness and sleep architecture. The MSLT consists of 4 or 5 twenty minute nap opportunities given at 2 h intervals on the day following an overnight PSG. The basic parameters measured are (i) latency (time taken) to fall asleep (if sleep occurs), which is averaged over the naps to give a mean sleep onset latency and (ii) latency to REM sleep (if observed). No widely accepted norms for children currently exist. Adult norms for "pathologic" levels of sleepiness are generally applied to adolescents and older children but are not relevant or utilised for children <5 years of age. Normal mean sleep latency in a school age child is 16–18 min [5]. Pathologic levels of sleepiness are

generally associated with narcolepsy but may also occur as a result of significant chronic sleep deprivation, circadian misalignment or a number of other medical conditions (e.g., epilepsy, Kleine–Levin Syndrome) or due to certain medications/drugs (and hence a drug screen is undertaken as part of the protocol). A short mean sleep onset latency (<8 min) in addition to either (i) REM sleep occurring in 2 or more naps (sleep-onset REM sleep or SOREMs) during a MSLT or (ii) the presence of SOREM on the overnight PSG and in one nap during MSLT is highly suggestive of narcolepsy [6,111].

*Maintenance of wakefulness test (MWT)* – As the MWT is rarely performed in children refer to the adult document "Guidelines for sleep studies in adults – a position statement of the Australasian Sleep Association" [7].

#### 2.2. Types of sleep studies: levels of investigation

It is evident that the burden of disease associated with sleep disorders in children is great, given their high prevalence and significant associated morbidities [8]. This is especially the case with OSA, which affects 1–5% of children [9]. There is pressure on the specialized facilities throughout Australia and New Zealand to meet growing demands for diagnosis and management, with the result that the waiting list for sleep studies in many centres is very long. This high demand for investigation plus the relatively costly and labour intensive nature of attended PSG (type 1 study) has led to the development of a number of simplified or "abbreviated" sleep studies. These abbreviated studies (types 2–4) are classified according to the number of channels of physiological data that is recorded (classification is detailed below).

As objective testing is preferable to clinical evaluation alone [9–13], some centres may choose to carry out abbreviated studies particularly to prioritise the waiting list for PSG and/or adenotonsillectomy (as the first line treatment for uncomplicated OSA in most children) [14]. Given the potential for continuing adverse consequences of undiagnosed and untreated OSA (including neurocognitive, behavioural and cardiovascular impairment [9]), a pragmatic approach to diagnosis that expedites treatment for symptomatic children is appropriate.

In choosing which test or tests are to be used physicians should have a clear understanding of:

- (a) The question to be answered by undertaking the test (i.e., indications for testing);
- (b) The sensitivity and specificity of the test(s) to diagnose a particular sleep disorder;
- (c) The overall utility of the test, taking into consideration the prevalence of a given sleep disorder in the paediatric population;
- (d) The cost/benefit balance of the test in a given clinical setting(s);
- (e) The technical limitations of the monitoring signals utilised in each particular study type;
- (f) Comorbidities and other factors which may affect the reliability and interpretation of the result.

Ultimately the type of sleep study performed will be defined by local facilities, experience and expertise. If unattended and/or abbreviated sleep studies are all that is available it is essential that paediatricians reviewing the study report remain objective and realistic about the limitations of such tests. For sleep studies of any type, the raw data must be available for review by the sleep specialist reporting the study, to enable evaluation of the study quality and to ensure the technical adequacy and accuracy of scoring and interpretation of results.

Sleep studies may be divided into two broad categories: (1) a Type 1 or "comprehensive" study (attended polysomnography)

which is considered the reference standard against which other diagnostic methods are evaluated, or (2) home-based or limited channel sleep studies (Type 2–4) where fewer physiological variables are recorded (see below for more detail). Limited channel studies are usually undertaken using specific portable equipment and are often undertaken in the home or on the hospital ward. Sleep studies may be supervised (continuous attendance by medical, scientific/technical or nursing staff specifically trained in conducting sleep studies) or unsupervised (staff are absent during the recording period). The study duration may be:

- **A full night:** a study conducted over the entire normal sleep period, beginning at the usual bedtime and usually lasting more than 6 h.
- **A full night but split into two parts:** a diagnostic component (commonly characterised by identification of moderate or severe OSA) followed by a therapeutic intervention component (most commonly CPAP titration).
- **Of limited or restricted duration:** a study where the planned length of study is less than 6 h. Such studies would include “nap” studies conducted during the hours of daylight e.g. to investigate sleep disordered breathing in neonates.

### Type 1 Sleep Studies

A Type 1 sleep study (PSG) is considered the reference standard against which other diagnostic methods are evaluated [9,12,13,15]. A type 1 study refers to polysomnography carried out throughout the night in a sleep laboratory in the presence of trained technical staff and under the supervision of a qualified sleep specialist. It involves the continuous recording of multiple physiological variables so that detailed measures of sleep and breathing can be made. Recommended signals include: electroencephalogram (EEG), bilateral electro-oculogram (EOG), submental electromyography (EMG), electrocardiography (ECG), leg EMG, arterial oxygen saturation, respiratory thoraco-abdominal movements, nasal pressure and oronasal airflow, sound, a measure of carbon dioxide (transcutaneous or end tidal), body position and digital video recording. The reader is referred to the AASM [4] and ASTA/ASA guidelines [3] for details of the variables that should be measured during PSG and the technical recording specifications.

PSG enables measurement of sleep architecture (the amount and distribution of the various stages of sleep), quantification of sleep disturbance (arousals and awakenings) and accurate quantification of abnormal respiratory events against time spent asleep. Rapid eye movement (REM) sleep is frequently associated with exacerbation of a sleep-related breathing abnormality and, in some cases, sleep apnoea/hypopnoea may be confined entirely to REM sleep. Thus the quantification of different sleep stages during the night may be important. PSG distinguishes obstructive from central breathing abnormalities, determines the effects of body position on sleep disordered breathing and enables the diagnosis of alternative sleep disorders (e.g. periodic limb movement disorder, parasomnias). Polysomnography also provides information on sleep fragmentation and arousals which are likely important in the genesis of daytime symptoms.

Overall there is a large body of evidence supporting the reliability and accuracy of type 1 sleep studies for detecting the presence of OSA, and the test-retest reliability or consistency of results across multiple nights of PSG recordings has been demonstrated in several studies [9,10,13,16–18]. While it is recognised that some children may not sleep as well on the first night they are in a sleep laboratory because of anxiety, an unfamiliar environment and the attached sensors, research evaluating this so-called “first night effect” suggests few are misclassified on the basis of a single night PSG [17–21].

### Type 2 Sleep Studies

A Type 2 study refers to an abbreviated PSG that is unattended by trained sleep laboratory staff. It is usually undertaken at home but may also be carried out on a hospital ward. A higher success rate in terms of good quality signals may be achieved when the study is set up by experienced personnel rather than by parents or untrained health care professionals [19,22]. A type 2 or ambulatory study must record a minimum of seven channels including EEG, EOG, chin EMG, ECG or heart rate, airflow, respiratory effort and oxygen saturation. The important distinction from level 3 and 4 studies is that sleep itself is quantified as well as respiratory variables. Audio/video monitoring improves the reliability of unattended studies [23].

This type of testing allows for sleep staging and therefore calculation of total sleep time and respiratory indices (e.g. apnoea–hypopnoea index, AHI). One of the major limitations of unattended studies is the potential for data loss overnight, particularly airflow signals (thermistor or nasal pressure) which are crucial for determination of respiratory events. However, this potential concern regarding signal loss has not been borne out by recent studies showing high success rates in children [22]. However, studies to date have included uncomplicated normal mainly school-age children and excluded children with complex medical problems, neurodevelopment delay and significant co-morbidities (e.g., in Marcus et al., only 6 children were developmentally abnormal). Further research is required to compare the feasibility and accuracy of Type 2 with Type 1 studies in children with medical comorbidities who form a significant proportion of children referred to specialist sleep centres for evaluation of sleep disorders.

### Type 3 Sleep Studies

Type 3 or limited channel sleep studies, sometimes called cardiorespiratory sleep studies, have a more restricted number of parameters that are measured. Usually type 3 studies have at least four variables that are monitored (depending on the type of monitor used): oximetry plus a selection of respiratory effort (chest, abdominal, or both), airflow (nasal or oral by pressure or thermistor), head or body position, ECG, tonometry (a marker of autonomic control), actigraphy (vibration detection) and/or sound/video recording. These studies may be attended or unattended and can be performed in the sleep laboratory, in hospital or at home.

The diagnostic accuracy of type 3 studies is potentially limited by the need to estimate total sleep time using data other than EEG. Total recording time is often used as the denominator, resulting in an underestimate of the severity of sleep disordered breathing. Also sleep disordered breathing leading to arousal and secondary sleep disruption rather than oxygen desaturation may be missed by limited channel studies (type 3 and 4) where sleep and arousals are not quantified. Video recording may reduce this as body movement arousals can be scored visually [24].

While level 3 studies are of relatively low cost, there is limited evidence to date of their efficacy. They have a variable failure rate due to data loss, particularly when the setup is performed by caregivers rather than trained staff. Rosen et al. [25] using a level III device at home on children aged 8–11 years, demonstrated 94% were technically adequate and, when compared to a subset undergoing laboratory PSG, showed good sensitivity and specificity. However, although this cohort study reported ‘excellent agreement’, no actual data was reported. Zucconi et al. [26] evaluated a level III device in the hospital ward (children 3–6 year, N = 12) compared with laboratory PSG on an alternative night. This showed good sensitivity (89%) for increasing obstruction but poor specificity. Poels et al. [27] used an in-home recording device set up by the carer in 2–7 year old children (n = 24) prior to adenotonsillectomy and due to a high failure rate (25–30%) concluded

level 3 studies were of limited use. Conversely, Jacob et al. [24] reported that limited channel home testing plus video was an adequate (83%) and practical option in the evaluation of routine OSA in children ( $n = 21$  2–12 years) with adenotonsillar hypertrophy (Positive Predictive Value (PPV)  $\geq 70\%$  and Negative Predictive Value (NPV)  $> 90\%$ ). This latter study involved specialist staff setting up the unit at home. A recent study by Tan et al. found that clinical decisions at the extremes of sleep-disordered breathing (normal/primary snoring with obstructive apnoea–hypopnoea index (OAH)  $< 1$ , or severe OSA with OAH  $> 10/h$ ) were unaffected by the use of type 3 studies, whereas under-estimation of events on a type 3 study may have significant impacts on treatment decision in the middle range of OAH (1–10/h) [28].

Type 3 studies may be useful in cases where a child is too unwell to have a formal study undertaken in a sleep laboratory, or as a relatively simple way of determining the efficacy of treatment in the early stages of initiation of non-invasive respiratory support. However, overall the literature suggests that level 3 sleep studies typically have a high positive predictive value for the presence of OSA but a significant false negative rate (low negative predictive value) i.e., rule in but do not rule out OSA.

#### **Type 4 Sleep Studies**

A Type 4 study is one that incorporates only one or two measured parameters – for example oxygen saturation, heart rate, transcutaneous carbon dioxide or airflow. Type 4 studies are usually unattended and conducted in the patient's home or on a hospital ward. Oximetry is an accurate, quantifiable, reliable and informative signal [29]. The development of the multi-wave length oximetry and reduction in size has made oximetry a ubiquitous and accurate marker of hypoxemia. As for Type 3 studies, level 4 studies reliably rule in but do not rule out OSA. Focussing on dips in oxygenation will miss children with OSA with significant sleep disruption but not desaturation (high positive predictive value but low negative predictive value). As with all tests, it is critical to understand the indications for pulse oximetry, what the instrument measures and reports, its limitations and how to interpret results. Technical factors such as adequate signal acquisition, averaging time and storage sampling frequency are crucial to the reliability of oximetry data. Careful choices of equipment and software are needed for services undertaking oximetry for diagnostic purposes, and results should be interpreted by trained practitioners. Errors in interpretation may lead to inappropriate treatment decisions being made.

Limitations of type 4 studies are seen in cardiac, pulmonary and neurological patients where differentiation between obstructive and central sleep apnoea is especially important, as this is not possible using type 4 studies (the pattern of desaturation may be identical). Low baseline oxygen saturation will reduce specificity further, while supplemental oxygen therapy may negate the utility of signal interpretation. Other limitations include difficulty in identifying artefact, lack of positional data and the use of time in bed rather than actual sleep time in calculating the level of respiratory disturbance.

Despite these limitations, the appearance of the trend and the analysis of desaturation events measured on overnight oximetry may be helpful as a quick, inexpensive alternative to a PSG for the diagnosis of uncomplicated OSA [14]. In the context of a sleep laboratory with a high (60%) pre-test probability of OSA in referred patients, a positive oximetry trend graph (defined as 3 or more clusters of desaturation events and at least 3 desaturations  $< 90\%$ ) had a positive predictive values of 97% and could therefore reliably predict the need for adenotonsillectomy in uncomplicated patients without the need for further testing [29]. On the other hand a patient with a negative oximetry had a post-test probability of OSA of

47% and therefore required further testing to exclude or confirm the diagnosis of OSA. Children with frequent oxygen desaturation events  $< 80\%$  are at increased risk of requiring major airway intervention such as re-intubation in the post-operative period [30–32], a finding that facilitates planning of surgical intervention and perioperative care [30]. Recently Horwood et al. [14] demonstrated that oximetry studies evaluated with the McGill Oximetry score expedite diagnosis and treatment of children with adenotonsillar hypertrophy referred for suspected OSA and Pavone et al. [33] demonstrated good reproducibility of oximetry testing.

In the hospital environment Type 4 studies particularly oximetry and transcutaneous carbon dioxide may be useful for the acute assessment of children and young people with neuromuscular disease at risk of development of hypoventilation or as an easy method of checking adequacy of non-invasive respiratory support. For other medically fragile children where hypoventilation during sleep may be a component of their respiratory status (e.g. achondroplasia, spina bifida, severe obesity etc) assessment of the child's gas exchange may be helpful in prioritising further evaluation or planning for definitive PSG at a future date.

#### **Summary**

In summary it is important to highlight that no research has been conducted on the validity of home monitoring (level 2, 3 or 4 studies) for the range of children typically referred for PSG for suspected OSA. Most research studies utilising limited channel recording devices have excluded very young children and those with significant co-morbidities. It is therefore recommended that limited channel diagnostic devices be used in developmentally normal children who are  $> 2$  years of age with a high clinical suspicion of OSA following specialist review. A comprehensive clinical history and assessment is essential to help exclude conditions other than sleep disordered breathing e.g. PLMD. While research has not identified safety concerns for home unattended studies, they still require particular attention to safety (e.g., entanglement). Technically reliable information depends on adequate parental instruction, and when possible monitors should be setup by trained staff. Despite the availability of limited channel sleep studies, access to type 1 PSG studies remains vital for the comprehensive and definitive assessment of sleep disordered breathing in children, particularly those with complex health needs. However, irrespective of which type of test is utilised, it is crucial that the results are evaluated and reviewed by a clinician with the necessary expertise and knowledge to interpret the findings accurately and in the context of the clinical history and the developmental stage of the child (i.e., an accredited paediatric sleep physician).

### **3. Respiratory indications for sleep studies in children**

#### *3.1. Obstructive sleep apnoea*

#### **Background**

Sleep-related upper airway obstruction is a common disorder of childhood that is considered to represent a spectrum of severity ranging from primary snoring at the mild end to obstructive sleep apnoea at the severe end. OSA is defined as the presence of periods of partial upper airway obstruction and/or intermittent complete upper airway obstruction resulting in abnormalities in ventilation (hypoxia and/or hypercapnia) and/or sleep disruption [1]. Primary snoring is characterised by habitual snoring without ventilatory abnormalities and relatively preserved sleep architecture. OSA is estimated to affect between 1 and 5% of children [9]. It occurs across all age groups but peaks in the preschool years due to adenotonsillar enlargement at a time when the upper airway is still

relatively small [34]. In otherwise normal healthy children adenotonsillar hypertrophy is the most common underlying cause of OSA but other factors such as structural and neuromuscular factors may also play a role.

An increased prevalence of OSA has been reported in specific ethnic groups and in obese children. Particular clinical groups are also at increased risk of developing OSA. For example Down syndrome (DS), or trisomy 21, is the most common chromosomal disorder seen in childhood and is associated with a significantly increased prevalence of sleep-disordered breathing compared to the general population. Anatomical abnormalities including mid-face and mandibular hypoplasia, a small nasopharynx, relative or true macroglossia in addition to glossoptosis, generalised hypotonia and obesity, are characteristic features of DS, and predispose to the development of OSA which is often more abnormal on PSG findings compared to non-syndromic children [35,36]. Additional concerns regarding the management of OSA in Down syndrome include a poor correlation between parental report of OSA symptomology and PSG findings [37] and indications that there may be a reluctance or delay in referring DS children for PSG evaluation [36]. In view of this, parents of children with DS should be regularly questioned about symptoms of OSA including snoring, heavy breathing, witnessed apnoea, restless sleep, frequent night waking, unusual sleeping positions (e.g., neck splinting), daytime tiredness or behavioural problems that may be associated with disturbed sleep, with a low threshold for evaluation with PSG if symptoms of OSA are present [38,39].

Other pre-disposing risk factors for OSA include upper airway craniofacial anomalies (particularly those associated with mid-face hypoplasia or micrognathia), neuromuscular disease (including both hypotonia and hypertonia), structural abnormalities of the chest wall and neurological disorders including myelomeningocele and Chiari malformation [40]. A high prevalence of upper airway obstruction has also been reported in atypical presentations of laryngomalacia (e.g. no history of stridor) [41]. Thus children with a range of craniofacial, genetic or neurological disorders are at increased risk of developing OSA (see Appendix I), with more careful screening and a lower threshold for evaluation recommended for these clinical populations. The neurocognitive, behavioural and cardiovascular sequelae of OSA are well documented [9].

The presentation, pathophysiology and clinical findings of paediatric OSA are different to that observed in adults [34] and therefore different diagnostic criteria are applied to infants and children. The pattern of upper airway obstruction seen in children during PSG varies widely with prolonged periods of partial upper airway obstruction (obstructive hypoventilation) rather than frank obstructive apnoea often observed. The clinical presentation of OSA in children includes snoring, restless sleep, hyperhidrosis and poor concentration or neurobehavioral deficits. The prevalence of enuresis and parasomnias is increased in children with OSA [42]. While snoring is the cardinal symptom of OSA in older children, infants may have little or no snoring [1]. Furthermore habitual snoring occurs in 7–15% of children [43] but clinical history is unable to distinguish primary snoring from OSA [1].

While adenotonsillectomy (AT) is commonly the first line of treatment for children with OSA, other treatment modalities may include anti-inflammatory therapies, positive airway pressure support, nasopharyngeal airway, mandibular distraction, rapid maxillary expansion or mandibular advancement splint.

Although AT is effective in treating OSA in many children, some individuals and clinical populations are at increased risk of residual OSA after surgery. Estimates of residual OSA following AT vary (13–29%) in low risk groups but as high as 73% in high risk groups [9]; with increased risk of residual obstruction related to a range of factors including pre-existing co-morbidities and moderate to severe pre-operative OSA [44–46]. Incomplete resolution of OSA is more

common in children with craniofacial abnormalities that obstruct the upper airway [47], and neurologic disorders such as myelomeningocele [48] and Down syndrome [49]. Residual OSA is also more common in obese children [46,50,51]. In a large randomised controlled trial of AT for OSA, residual OSA was detected in 33% of obese subjects [51]. Recurrence of OSA one year following AT has also been demonstrated [44]. Given the high prevalence of residual OSA clinicians should evaluate children postoperatively for symptoms of OSA and parents advised of the potential recurrence of OSA.

### Consensus recommendations

- PSG is indicated when clinical assessment suggests the diagnosis of obstructive sleep apnoea (OSA).
- Abbreviated testing is indicated in selected cases as a clinical adjunct tool for the assessment of children with suspected obstructive sleep apnoea (OSA) following paediatric sleep specialist review.
- PSG is indicated if clinical assessment suggests OSA and abbreviated testing is inconclusive for the diagnosis of obstructive sleep apnoea.
- PSG is indicated when the severity of OSA is in doubt or if there is significant risk of post-surgical complications (e.g., idiopathic thrombocytopenic purpura).
- PSG is indicated when clinical assessment suggests the presence of residual OSA following AT or other treatment modalities.
- PSG is indicated following AT in children where there is the presence of ongoing symptoms or other clinical features suggestive of persistent OSA, particularly in those with: moderate to severe OSA pre-operatively, obesity, craniofacial abnormalities that pre-dispose to the development of upper airway obstruction, neurologic disorders (e.g., neuromuscular disorders, spina bifida, Chiari malformation, meningomyelocele) and high risk congenital conditions (e.g., Down Syndrome, Prader–Willi Syndrome). See Appendix 1.

### 3.2. Central sleep apnoea and non-obstructive hypoventilation

#### Background

Central sleep apnoea is characterised by absent respiratory effort occurring in a cyclical or intermittent pattern arising from a deficiency in central respiratory control. Non-obstructive hypoventilation is characterised by reduced pulmonary ventilation arising from diminished central respiratory drive, neuromuscular abnormalities or restrictive lung disorders. The aetiology of these abnormal respiratory patterns may be congenital or acquired [52,53] and can result in significant hypercapnia and/or hypoxia. Clinical indicators of non-obstructive sleep disordered breathing include witnessed apnoea, nocturnal dyspnoea, restless sleep or frequent arousals, hyperhidrosis, morning headaches, waking unrefreshed or tired, elevated awake PaCO<sub>2</sub> or base excess, changes in behaviour or neurocognition or daytime tiredness. However some patients may be asymptomatic, particularly in the early stages of non-obstructive hypoventilation.

#### Central sleep apnoea

Discrete respiratory pauses are a normal feature of sleep in all age groups and are typically observed in REM sleep or following sighs and body movements. These events are generally of brief duration (<20 s) and do not usually result in significant blood gas exchange abnormalities or sleep disruption [54]. Clinically relevant central apnoeas are those of prolonged duration (>20 s) or if associated with oxygen desaturation, cyanosis, cortical arousal or

bradycardia. Brief central apnoeas are common in term infants in the first few months of life (particularly in REM sleep), decreasing in frequency with postnatal development due to maturation of the respiratory system. Central apnoea can also be seen in some individuals at sleep onset due to instability of ventilatory control occurring at wake/sleep transitions, but these events typically resolve once sleep deepens.

Children with OSA sometimes display an increased frequency or severity of central apnoea [55] which can resolve following AT [56]. Children at risk of cervico-medullary or brain stem compression (e.g., Chiari malformation, meningomyelocele, spina bifida, achondroplasia) are at increased risk of developing central apnoea [40,57]. An increased frequency and/or severity of central apnoea has also been reported in children with a range of neurological disorders (e.g., Joubert syndrome) and those with neurodevelopmental disorders such as Down syndrome [58] and Prader–Willi syndrome [59], due to impaired ventilatory control.

#### *Periodic breathing*

Periodic breathing (PB) is defined as a series of three or more central pauses lasting  $>3$  s separated by no more than 20 s of normal breathing usually presenting in a crescendo/decrecendo pattern [4]. Periodic breathing is very common in premature infants reflecting the immaturity of the respiratory system. Periodic breathing can also be seen in term infants, peaking between 2 and 4 weeks of age, where up to 2–5% of total sleep time may be spent in this breathing pattern. Periodic breathing can be seen in both REM and NREM sleep but the pattern appears to differ between these two sleep states [54]. Elevated levels of PB in infancy or PB observed in older children is considered abnormal [54]. Periodic breathing is sometimes seen in children at sleep onset due to instability of ventilatory control occurring at the wake/sleep transition and this typically resolves once sleep deepens.

#### *Apnoea of prematurity*

Due to the immaturity of the respiratory control centre, central apnoea and PB are very common in premature infants, with the prevalence inversely related to birth weight and gestational age [60]. Other potential structural abnormalities of the respiratory system related to pre-term birth include narrow airways, chest wall distortion, diaphragmatic fatigue, poor neuromuscular control and reduced functional residual capacity (FRC) which can all be exacerbated during sleep, particularly active sleep [61]. Upper airway obstruction is therefore also a frequent feature of SDB in premature infants, with mixed apnoea the most common type of respiratory event observed [62]. Rapid and profound oxygen desaturation may occur with respiratory events of relatively short duration while significant bradycardia and pallor may also be associated with abnormal breathing. Given that apnoea of prematurity (central sleep apnoea or periodic breathing) is virtually ubiquitous in very premature infants, PSG would not be indicated in those cases [1] and the apnoea can be managed and monitored clinically. There are selected cases of post-term infants however where other medical disorders have been ruled out and PSG may be indicated to confirm a diagnosis of idiopathic apnoea of infancy [2], define the extent of gas exchange abnormality and rule out hypoventilation [1]. In this context PSG may be useful to determine the need for treatment and/or home monitoring.

#### *Congenital central hypoventilation syndrome*

Congenital Central Hypoventilation Syndrome (CCHS) is a rare disorder of ventilatory control associated with autonomic nervous

system (ANS) dysregulation [63]. The clinical presentation and severity of CCHS is variable but in general it is characterised by sleep-related respiratory insufficiency and a diminished ventilatory response to hypercapnia and hypoxia. A mutation in the *PHOX2B* gene which is involved in ANS development has been identified as the cause. While ventilation is usually most abnormal in NREM sleep (or quiet sleep for infants), REM sleep and wake ventilatory abnormalities can also occur [64]. Most children with CCHS typically present at birth or in infancy with severe central hypoventilation requiring ventilation, but a milder phenotype can also present later in older children or adults.

#### *Non-obstructive hypoventilation*

In children, non-obstructive hypoventilation is commonly seen in neuromuscular disorders (e.g., spinal muscular atrophy, Duchenne muscular dystrophy, myotonic dystrophy), in restrictive lung disorders (e.g. kyphoscoliosis, chest wall deformities) and sometimes in individuals with morbid obesity. In addition to hypoxia (which may be intermittent or sustained) the development of carbon dioxide retention may cause blunting of central chemoreceptor function further exacerbating sleep-disordered breathing [65,66]. Upper airway obstruction may also co-occur due to diminished muscle activity, abnormal neurological control or mechanical breathing disadvantages [67].

While the development and pattern of sleep-disordered breathing is highly variable in different neuromuscular diseases (NMD), respiratory muscle weakness and scoliosis are two major contributing factors to the development of non-obstructive hypoventilation [68]. It has been estimated that up to 62% of children with NMD have some degree of sleep-disordered breathing [69]. In children with progressive NMD, non-obstructive hypoventilation may develop gradually but daytime symptoms and clinical assessment such as pulmonary function testing frequently fail to identify the advent of non-obstructive hypoventilation until daytime respiratory abnormalities are present indicating advanced disease progression [70,71].

There is some evidence to suggest that commencement of non-invasive ventilation in the early stages of sleep-related hypoventilation slows disease progression and reduces hospital admissions [72]. Therefore it is recommended that children with progressive NMD have regular clinical assessment to review disease progression and symptoms of SDB which may need evaluation with PSG to determine if non-invasive ventilation should be considered [73,74]. In particular, the risk of sleep-related hypoventilation is increased if scoliosis or diaphragmatic weakness are present, and after the loss of ambulation in Duchenne muscular dystrophy [68,75]. Additionally, as non-obstructive hypoventilation may not be clinically evident in children with NMD, pre-surgery PSG should be considered if major surgical procedures are planned (e.g., scoliosis surgery) due to the risk of adverse responses to the administration of anaesthetics or sedatives. A forced vital capacity (FVC) below 1 L and/or 40% predicted [76] or the presence of daytime hypercapnia are also clinical indicators for conducting PSG.

Relatively few studies in children have examined the effect of restrictive lung disease on sleep-related respiratory patterns. The relatively small thorax of children with achondroplasia is believed to put them at increased risk of developing sleep-related hypoxia due to restrictive lung disease, although not all studies report this finding [77]. While restrictive lung disease has been reported in some children with achondroplasia [78,79] it appears to be less prevalent than OSA in this population. Adults with kyphoscoliosis are known to have breathing difficulties during sleep and the same finding may be expected in children but studies are lacking [34].



Non-obstructive hypoventilation due to morbid obesity (obesity hypoventilation syndrome) is typically characterised by a combination of symptoms including increased carbon dioxide retention during sleep, sleep-related hypoxia not caused by upper airway obstruction and daytime somnolence not explained by other causes. Some individuals may also show awake hypercarbia or other adverse cardiovascular abnormalities such as pulmonary hypertension [80]. The pathophysiology of obesity hypoventilation syndrome (OHS) is complex and not fully understood. The increased mass loading on the pulmonary system reduces chest wall compliance and FRC leading to increased work of breathing and reduced oxygen reserve. Blunted ventilatory drive has been demonstrated in adults with OHS [81] and though data are lacking for children, similar findings may apply.

### Consensus recommendations

- PSG is indicated if clinical evaluation or abbreviated testing suggests the presence of central apnoea that is: (i) unexpected for development/age (ii) of prolonged duration or (iii) associated with significant oxygen desaturation or frequent bradycardia, pallor change, sleep disturbance or other daytime sequelae.
- PSG is indicated in cases of apnoea of infancy that are unresponsive to treatment or unusually severe or persistent or if there is diagnostic doubt over the aetiology of events (i.e. obstructive versus central).
- PSG is indicated when clinical assessment suggests the diagnosis of Congenital Central Hypoventilation Syndrome (CCHS).
- PSG or abbreviated testing is indicated when clinical assessment or other investigations (e.g. pulmonary function tests) suggests the presence of non-obstructive hypoventilation due to NMD, obesity, kyphoscoliosis or chest wall deformity.
- PSG or abbreviated study is indicated pre-operatively in at-risk children with neuromuscular disorders undergoing major orthopaedic, thoracic or upper airway surgery to screen for undetected SDB which may be exacerbated by sedation or analgesics.

### 3.3. Polysomnography for children receiving respiratory support

#### Background

The treatment of sleep-disordered breathing by positive airway pressure (PAP) support has increased significantly in the paediatric population in recent years, particularly with the advent of suitable mask interfaces for infants and children. Continuous positive airway pressure (CPAP) and bi-level positive airway pressure (BPAP) are used to treat a range of sleep-related breathing abnormalities including upper airway obstruction and non-obstructive hypoventilation. An initial sleep study is generally performed on PAP to optimise treatment as pressure levels can be manually titrated directly in response to abnormal respiratory events observed during sleep. Subsequent repeat sleep studies are important in optimising long term management, particularly to determine if pressure requirements have changed as a result of the child's growth and development, if symptoms reappear while on PAP or if additional or alternate treatment has been instituted [82]. The frequency of follow up should be based on clinical symptoms, the child's growth rate, clinical stability or disease progression or other factors that may precipitate a worsening of sleep disordered breathing (e.g., significant weight gain). However, children receiving PAP should have at least an annual PSG, with young children and those with a rapidly progressive disorder often requiring studies more frequently (e.g. 4–6 monthly). Clinical or physiological improvement has been demonstrated in children

with SDB and neuromuscular disorders when BPAP was titrated during PSG [82–84].

Regular review of interface fit and suitability is also important. Following reports in the literature, clinicians should be mindful of the potential development of mid-face hypoplasia in children receiving PAP. Regular assessment and techniques to try and minimise the development of mid-face hypoplasia (e.g. mask rotation, nasal cushions) are recommended with a low threshold for referral to a craniofacial or orthodontic specialist. Furthermore, it is important to note that PSG is only one component in the treatment of sleep disordered breathing, with education of the patient and family and long term evaluation of the patient's symptoms, quality of life, adherence and side effects of the treatment being equally important. Patient education and motivation to use CPAP account for a greater variance in PAP adherence than standard biometric or anthropometric markers of OSA severity [85].

Other methods for treating SDB include tracheostomy, mechanical ventilation and humidified high-flow nasal cannula therapy. Polysomnography is potentially useful for assessing children using these treatments as respiration may worsen during sleep and awake ventilator settings may therefore be inadequate during sleep. There are no published papers for this indication. There are some studies that have demonstrated the clinical usefulness of PSG using a downsized or capped tracheostomy tube as part of the evaluation to assess readiness for decannulation in children with long-term tracheostomy [86,87]. Some children treated with tracheostomy for sleep-related breathing disorders may benefit from polysomnography as part of the evaluation prior to decannulation but this is not universally accepted. These children should however be followed clinically after decannulation to assess for recurrence of symptoms of sleep related breathing disorders.

### Consensus recommendations

- PSG is indicated for children receiving positive airway pressure support (CPAP or BPAP) to titrate the level of pressure support required and to periodically re-evaluate the appropriateness of settings, including synchrony between machine and patient.
- PSG or abbreviated testing may be beneficial in children treated with invasive or negative pressure ventilation to adjust ventilator settings.
- PSG is indicated in children with symptoms of sleep-disordered breathing following tracheostomy decannulation. PSG may be clinically useful in some children (e.g., achondroplasia, Pierre Robin Sequence) with tracheostomy capped or downsized as part of the evaluation prior to decannulation.
- PSG is indicated in children with tracheostomy for upper airway obstruction where the co-occurrence of another breathing disorder (e.g., central sleep apnoea) is suspected.
- PSG is not routinely indicated for the titration of supplementary oxygen in children with chronic lung disease except in patients who are known, suspected or considered at risk of developing hypoventilation, central apnoea or hypercapnia with supplemental oxygen.

### 3.4. Other respiratory indications for sleep studies

#### 3.4.1. Apparent life threatening event (ALTE)

The most widely used definition of an ALTE is an episode that is "frightening to the observer and that is characterized by some combination of apnea (central or occasionally obstructive), color change (usually cyanotic, but occasionally erythematous or plethoric), marked change in muscle tone (usually marked limpness), choking, or gagging" [88]. The majority of ALTEs occur in children under the age

of one year, with premature infants at increased risk [89]. It is recommended that the investigation and management of ALTE is predicated on the perceived severity of the presentation and the clinical history/physical examination of the child [54,90]. Guidelines for initial assessment and follow-up have been published [90]. Approximately 20% of ALTEs for which an explanation is found are believed to arise from respiratory causes including obstruction and central apnoea [54,90]. In general overnight polysomnography is only indicated in selected infants where there is suspicion of sleep-disordered breathing (including periodic breathing and obstructive sleep apnoea), evidence of oxygen desaturation or the presentation of repeated ALTEs of unknown aetiology [54].

Several studies [91–93] have shown subtle, nonspecific PSG abnormalities in some infants who had experienced an ALTE. The PSG findings were not predictive of recurrence of ALTE. Studies of infants who eventually succumbed to sudden infant death syndrome (SIDS) [94–96] demonstrated PSG abnormalities that were neither sufficiently distinctive nor predictive to support routine use of PSG for children at risk for SIDS. If, however, obstructive sleep apnoea is clinically suspected [97], or if bradycardia is demonstrated on cardiac monitoring in the absence of central apnoea, or if an underlying disorder of breathing control (e.g., CCHS) is suspected, consideration of a PSG should be discussed with a paediatric sleep centre. Thus PSG is not indicated as part of the routine evaluation of an ALTE unless the clinical indicators specified above are observed.

#### Consensus recommendations

- PSG is not indicated for infants as part of a routine evaluation for an uncomplicated ALTE.
- PSG is indicated for infants who have experienced an ALTE and where there is clinical suspicion of underlying sleep disordered breathing or if there is ongoing or unexplained oxygen desaturation.

#### 3.4.2. Prader–Willi syndrome and growth hormone therapy

Sleep and respiratory abnormalities are common in Prader–Willi Syndrome (PWS) and include central and obstructive sleep apnoea, impaired ventilatory control, excessive daytime sleepiness and altered REM sleep distribution [98,99]. Sleep-disordered breathing including OSA is frequently found in children with PWS in the absence of symptoms [100,101]. Children diagnosed with PWS are frequently prescribed human growth hormone (GH), which has been shown to have a multitude of beneficial effects including improved linear growth and body composition as well as improvements in cognitive and developmental trajectories [102]. However there have been reports of sudden death in PWS children having symptoms suggestive of OSA who were receiving GH supplementation. The mechanism underpinning the association between sudden death and OSA in children with PWS has not been completely elucidated, but an increase in the volume of lymphoid tissue in the upper airway following GH supplementation may be a contributing factor. In response to this increased risk of OSA and potential sudden death, screening for sleep-disordered breathing prior to commencement of GH in children with PWS has been instituted in many countries. Treatment of OSA is usually recommended prior to commencement of GH therapy. Similarly, PSG is recommended soon after GH treatment has commenced (usually 6–10 weeks), particularly in younger children [102,103], given evidence for the development of severe OSA in some children in this time period, prompting withdrawal of GH while OSA was treated [104]. In addition, for the duration of GH treatment, children with PWS should be regularly monitored for symptoms of OSA and PSG repeated in the event of the

development of new or worsening symptoms, as OSA can develop later in the course of treatment with GH [105].

#### Consensus recommendations

- PSG is indicated in children with PWS prior to and six to ten weeks following the commencement of GH treatment and thereafter as clinically indicated.

#### 3.4.3. Sleep-disordered breathing in children with underlying chronic lung disease

Children with underlying lung disease (e.g., cystic fibrosis, bronchopulmonary dysplasia, asthma) may be at increased risk of developing SDB. These patients have reduced functional reserve and may become hypoxic during sleep, particularly in REM sleep when accessory respiratory muscle hypotonia occurs [34]. Other factors that may exacerbate sleep disordered breathing in children with chronic lung disease include increased bronchoconstriction during sleep, reduced mucociliary clearing or cough and increased arousals or sleep fragmentation [34].

#### Consensus recommendations

- PSG is indicated for children with chronic lung disease (e.g., asthma, cystic fibrosis, bronchopulmonary dysplasia) or unexplained pulmonary hypertension if there is clinical suspicion of sleep-disordered breathing.

### 4. Non-respiratory indications for sleep studies in children

#### 4.1. Movement disorders

##### Background

Sleep-related movement disorders are characterised by involuntary, often stereotypical movements that disturb the initiation or maintenance of sleep. These disorders include restless legs syndrome (RLS), periodic limb movement disorder (PLMD), sleep-related leg cramps, bruxism and rhythmic movement disorder [106].

Restless legs syndrome (RLS) is a neurological sensorimotor disorder characterised by an irresistible urge to move the legs often accompanied by uncomfortable or painful sensations of the lower extremities. These symptoms are generally worse in the evening, prior to sleep onset or when inactive and are relieved by moving the legs [106]. Periodic Limb Movement Disorder (PLMD) is characterised by periodic episodes of repetitive, highly stereotyped periodic leg movements during sleep accompanied by a complaint of sleep disturbance or daytime fatigue/tiredness that cannot be accounted for by another primary sleep disturbance. Periodic limb movements during sleep often co-occur with RLS and can be exacerbated by certain medications. The symptoms of RLS and PLMD can be difficult for younger children to articulate [107] and are sometimes incorrectly categorised as “growing pains.”

Clinical evaluation should include a detailed history and physical examination. Special emphasis on complaints of leg discomfort, the occurrence of leg or body jerks, restless sleep, and reports of insomnia or excessive daytime sleepiness should be sought. Physical examination should focus on excluding a peripheral neuropathy that can mimic RLS. Serum ferritin, complete blood count, urinalysis, and biochemistry testing should be undertaken to look for secondary causes of RLS (e.g., iron deficiency anaemia, uraemia). Generally RLS is diagnosed from clinical history and therefore polysomnography is not routinely indicated except where

uncertainty exists in the diagnosis, in which case the presence of PLMs might be collaborative evidence [108]. Polysomnography is required to diagnose PLMD particularly as the individual is usually unaware of the muscle movements and as parental report is often unreliable with a low positive predictive value. A periodic limb movement index of more than 5 per hour is considered abnormal in children [109]. As night-to-night variability in periodic limb movements has been documented in some children a single study might not be adequate to establish the diagnosis or severity of PLMD [110].

### Consensus recommendations

- PSG is not routinely indicated to diagnose restless legs syndrome (RLS), with the diagnosis based on clinical features.
- PSG is indicated for children suspected of having periodic limb movement disorder (PLMD).

### 4.2. Parasomnias and epilepsy

#### Background

Parasomnias are common in childhood and are defined as “*undesirable physical events or experiences that occur during entry into sleep, within sleep or during arousals from sleep*” [111]. Depending on their presentation, parasomnias may be a normal feature of sleep (primary parasomnias) or may point to other underlying medical conditions or pathology (secondary parasomnias) [112]. There are three broad categories: parasomnias associated with NREM sleep (e.g., sleep walking, sleep terrors and confusional arousals); parasomnias occurring in REM sleep (e.g., REM-sleep behaviour disorder, isolated sleep paralysis, nightmare disorder); and other parasomnias (e.g., sleep-related enuresis, catathrenia, sleep-related eating disorder) [113]. Parasomnias can present as simple or complex motor behaviours, abnormal sleep-related movements or vocalisations, and are associated with varying degrees of consciousness or perception, autonomic activation and emotional expression.

The unusual or abnormal behaviours seen in some parasomnias can mimic sleep-related seizures and vice versa, as some of the motor and behavioural manifestations observed are common to both disorders [114]. In particular nocturnal frontal lobe epilepsy can present in a similar fashion to NREM arousal parasomnias, although clinical features are helpful in differentiating the two [115]. Co-existing sleep disorders (e.g., OSA) may worsen seizures occurring during sleep.

A careful clinical evaluation of the suspected parasomnia with emphasis on age of onset, time of event relative to sleep onset, frequency, regularity and duration of event, and a detailed account of the event itself, is often sufficient to diagnose common, uncomplicated, non-injurious parasomnias without the need for polysomnography. However, polysomnography may be indicated if clinical evaluation is inconclusive (e.g. REM-sleep behaviour disorder or seizure activity), if excessive daytime sleepiness or impaired daytime functioning is present (not explained by other causes) or if the presence of another sleep disorder (e.g. OSA, PLMD) is suspected, as these may be worsen parasomnias. For example the prevalence of sleep walking and sleep terrors has been found to be elevated in pre-pubertal children with OSA and restless legs syndrome [42]. Utilising PSG to investigate unusual or atypical events occurring during sleep should include the recording of EMG, good quality audio-video and if available, an expanded bilateral EEG montage. Liaison with a paediatric neurologist may be judicious in cases where doubt remains about the diagnosis or in the advent of unusual EEG findings which are not be routinely

encountered in the sleep laboratory. PSG (with an extended bilateral EEG montage if available) may be helpful for the diagnosis of seizures occurring during sleep but is generally not the first line investigation.

### Consensus recommendations

- PSG is not indicated if clinical assessment is consistent with a typical childhood parasomnia.
- PSG is indicated if a parasomnia is atypical or if excessive daytime sleepiness or impaired daytime functioning is present that cannot be explained by other causes.

### 4.3. Hypersomnia and narcolepsy – polysomnography, multiple sleep latency test (MSLT)

#### Background

Excessive sleepiness in a child may present as sleeping longer than peers, age inappropriate napping or a change in sleeping pattern to sleeping greater amounts. However due to a general lack of knowledge in the primary care setting with respect to age appropriate sleep patterns and developmental changes, sleepiness in children is often under-recognised or misinterpreted [116]. For example the diagnosis of narcolepsy in children is often delayed by up to 10–15 years despite debilitating daytime sleepiness often being one of the first symptoms [117]. While a common cause of excessive sleepiness is insufficient or poor quality sleep (for a variety of reasons including intrinsic sleep disorders such as OSA), hypersomnias of central origin are not typically characterised by insufficient overnight sleep or by misaligned circadian rhythms [106]. The third edition of the International Classification of Sleep Disorders (ICSD-3) recognises eight distinct types of hypersomnia of central origin, several of which may be encountered in paediatric practice [111].

#### Narcolepsy

Narcolepsy is a debilitating lifelong neurological disorder characterised by hypersomnolence and the pathologic manifestation of REM-sleep like phenomena. The clinical features of narcolepsy include chronic excessive daytime sleepiness, disturbed sleep, cataplexy, sleep paralysis and hypnagogic or hypnopompic hallucinations. Not all features may be present in all patients, and excessive daytime sleepiness is often the only feature in children. The diagnosis of narcolepsy in children can be challenging due to the highly variable presentation of this disorder, the inability of younger children to describe their symptoms and the sometimes atypical presentation of some features such as cataplexy [118]. Misinterpretation of symptoms and misdiagnosis are not uncommon. Objective diagnosis of narcolepsy is achieved by performing a multiple sleep latency test (MSLT) where the propensity to fall asleep across the day and the presence of sleep-onset REM periods during naps are examined.

While the diagnosis of narcolepsy may be reasonably certain based on a clinical history of daytime hypersomnolence and cataplexy (which is highly specific to this condition), a MSLT preceded by PSG is still recommended for children in order to obtain objective evaluation. Furthermore, although infrequent, attempts to secure stimulant medication or exaggerated parental report of symptomatology may elicit a false or misleading clinical history. Additionally because this condition is life long and its diagnosis may have significant implications for driving and vocational choices, and medications to treat this disorder may carry significant risk, objective testing is highly desirable.

Overnight PSG immediately prior to the MSLT is used primarily to exclude other causes of excessive daytime sleepiness (e.g. OSA), and to confirm that the patient had sufficient sleep the night prior to the MSLT. Repeat MSLT or performing a maintenance of wakefulness test (MWT) may be indicated to evaluate the effectiveness of treatment or to comply with driver licensing authority requirements. Additional details for performing MSLT or MWT are provided later. Of note, the most recent International Classification of Sleep Disorders (ICSD-3) has revised the diagnostic criteria for narcolepsy [111].

#### *Kleine–Levin Syndrome*

Kleine–Levin Syndrome is a rare condition sometimes seen in adolescents (male predominance) characterised by recurrent or periodic hypersomnia interspersed with normal periods of alertness and behaviour. The hypersomnolent periods may also be accompanied by cognitive and mood changes, hyperphagia and hypersexuality. Overnight PSG findings during the symptomatic period include decreased slow wave and REM sleep, shortened REM latency and reduced sleep efficiency [119]. Multiple sleep latency testing shows reduced sleep latency but no sleep-onset REM periods as seen in narcolepsy [114]. Of note, reduced CSF hypocretin levels have been demonstrated in one 14 year old subject while symptomatic that normalised when not in the hypersomnolent phase [120].

#### *Idiopathic Hypersomnia*

The diagnosis of idiopathic hypersomnia is one of exclusion. Overnight PSG is therefore required to rule out other common sleep disorders such as OSA that can cause hypersomnolence. An assessment of sleep-wake schedules using a sleep diary with or without actigraphy over a 2–3 week period can also be helpful to exclude chronic sleep restriction. MSLT should be performed to objectively confirm the presence of hypersomnolence. Other rare medical disorders (e.g. Prader–Willi Syndrome and Myotonic Dystrophy) are associated with pathological sleepiness (and may also be associated with sleep-onset REM periods) and may thus be part of the differential diagnosis of idiopathic hypersomnia or narcolepsy. However, these conditions are usually readily identified by careful clinical history and examination, supplemented by genetic testing in some instances.

#### *Other Causes of Hypersomnia*

Other causes or medical conditions associated with central hypersomnia include neurological disorders, head trauma or neurosurgery, medications, substance abuse, psychiatric disorders and menstrual-related hypersomnia [106].

### **Multiple Sleep Latency Test (MSLT)**

A MSLT must be performed immediately following in-laboratory PSG. The overnight PSG is conducted to exclude other potential causes of daytime sleepiness (such as OSA or PLMD) and needs to demonstrate that an adequate amount of sleep has occurred (minimum 6 h) prior to the MSLT. However the presence of these disorders does not rule out the existence of narcolepsy, as narcolepsy can co-exist with other sleep disorders. If excessive daytime sleepiness or other clinical features of narcolepsy remain after treatment for co-existing sleep disorders then a MSLT should be undertaken. Brain MRI may be useful to rule out structural lesions mimicking narcolepsy.

The MSLT provides an objective measure of the ability or tendency to fall asleep. Four to five evenly spaced 20-min daytime nap opportunities with the patient lying in a quiet darkened room are

provided, and the time to fall asleep and the occurrence of any REM sleep is quantified from EEG/EOG and EMG recordings. The patient is instructed to attempt to fall asleep at each nap and the mean sleep latency result is taken to be indicative of sleep propensity. The application of occipital leads in the EEG montage is strongly advised as the transition from wake to sleep is accompanied by attenuation of the alpha waveform which is more accurately measured by the use of occipital leads. An additional vertical EOG lead is also recommended as it may be helpful in documenting eye movements during REM sleep [121]. Measurement of respiratory parameters is not generally indicated in a MSLT. However where a MSLT is conducted on a patient known to have a sleep-related respiratory disorder and using treatment, the test should be conducted with the patient using treatment, for example using CPAP. If this is not done there is a risk of prolonging sleep latency through the occurrence of respiratory events at sleep onset. Sleep diaries or actigraphy prior to MSLT may be helpful in interpretation, particularly to ensure that the patient is not sleep deprived prior to the PSG. Urine drug screening should be routinely performed on the morning of the MSLT. The reader is directed to the AASM clinical practice guideline for a detailed description of the performance of the MSLT, including test interpretation and reporting of results which are applicable to adolescents [122]. The finding of a mean sleep latency of less than 8 min (in the absence of a history of chronic or acute sleep restriction and in the absence of sleep disordered breathing or RLS/PLMS on PSG), plus the presence of REM sleep in two or more naps of the MSLT is diagnostic of narcolepsy. However, the most recent International Classification of Sleep Disorders (ICSD-3) has revised the diagnostic criteria for narcolepsy, whereby the presence of REM sleep within 15 min of sleep onset on the overnight PSG (preceding the MSLT) may replace one of the sleep-onset REM periods of the MSLT for the diagnosis of narcolepsy (in conjunction with a mean sleep latency of less than 8 min and no other explanatory factors for the presence of SOREM such as sleep restriction) [111]. Although the MSLT for children has limitations in young children, there are consistent data [123–125] demonstrating the diagnostic utility of the MSLT in school-aged children as young as 5 years with a clinical diagnosis of narcolepsy with cataplexy.

There are too few studies in conditions with hypersomnia other than narcolepsy to demonstrate sensitivity, specificity, positive predictive value, or negative predictive value of this test. Nonetheless, the MSLT can provide important information regarding degree of sleepiness in children with suspected hypersomnia from causes other than narcolepsy such as hypersomnia associated with a medical or psychiatric condition, recurrent hypersomnia, and other hypersomnias.

### **Consensus recommendations**

- The MSLT preceded by overnight PSG is indicated in children as part of the evaluation for suspected narcolepsy and excessive daytime sleepiness.

### **5. Sleep disorders service procedures, facilities and personnel requirements**

The following sections outline technical and practical recommendations for performing attended paediatric polysomnography (type 1 sleep study) in order to facilitate uniformity in practice and to set the minimum standards of practice for the delivery of high quality sleep medicine services. While minimum standards for sleep disorders services have been published jointly by the ASA and the National Association of Testing Authorities (NATA) (2012) the information contained in the following sections highlights or

expands on some of the recommendations or considers items not specifically contained in the ASA/NATA accreditation document.

### 5.1. Policies and procedures

A paediatric sleep disorders service should have in place (or adhere to existing broader organisational) policies and procedures with respect to:

- Standardised methodology for conducting PSG and related procedures (including positive airway pressure (PAP) and supplementary oxygen titration protocols).
- Administrative processes (e.g., handling and management of referrals).
- Maintenance of patient confidentiality.
- Equipment testing and safety.
- Data and file management/security (for both paper and digital records/reports).
- Staff training, supervision and ongoing education/development including regular staff appraisal.
- Quality assurance and improvement including proficiency testing, auditing processes and document control.
- Identification of non-conformities to established practice or procedures and subsequent corrective action and resolution.
- Emergency procedures.
- Patient safety.
- Consumer feedback and complaint resolution.
- Occupational health and safety including manual handling.
- Infection prevention and control including cleaning of equipment, electrodes and sensors and other items used during PSG and related procedures.

All policies and procedures should be documented and must be readily accessible by staff in the form of a manual or in electronic format. Policies and procedures should reflect current knowledge and best practice for sleep medicine service provision and should therefore be reviewed at appropriate intervals. Processes should be in place whereby any changes or updates to policies and procedures are effectively communicated to relevant staff.

### 5.2. Referral for polysomnography and related procedures

All requests for PSG and related procedures should be evaluated prior to testing for appropriateness of investigation by a paediatric sleep physician. Prior to a diagnostic PSG each patient should be clinically evaluated by a paediatric sleep physician. Polysomnography and related procedures should only be conducted once the sleep laboratory has received an appropriately authorised test request completed by a qualified paediatric sleep physician or a medical officer under his or her supervision. At a minimum the referral should contain the following:

- Patient identifying details.
- Reason for referral.
- Relevant clinical history including current medications.
- If applicable, PAP settings or level of supplementary oxygen the patient is receiving.
- Type of testing to be undertaken (e.g., diagnostic PSG, titration PSG, MSLT) and if applicable, any specific instructions with respect to the conduct of the test procedure, particularly if there are any variations or additional requirements from normal testing protocols.
- Any special instructions or considerations in relation to testing (e.g., special staffing requirements).
- The time frame or urgency in which testing is to occur.

- Any pertinent patient information that may be helpful to overnight staff (e.g., physical limitations, developmental delay, anxiety or behavioural issues).

### 5.3. Sleep facility personnel requirements

Paediatric sleep studies should be performed by staff that are skilled in dealing with children of varying ages and developmental stages, and who may present with medical or behavioural comorbidities. Depending on the nature of the sleep disorders service, staff performing paediatric sleep studies may comprise nurses, technical officers/scientists or clinical physiologists. Paediatric PSG requires a higher staffing ratio compared to adult studies with a ratio of no less than one nurse/sleep technologist to two patients required overnight. However in some cases a higher staffing level may be required (e.g. for tracheostomy patients). All staff must be trained in performing paediatric cardiopulmonary resuscitation covering the age range of patients that are seen within the service, with regular updates. In addition, staff should be instructed in the relevant statutory legislation relating to child protection. A criminal history check prior to working with children is generally required by most state authorities. As some clinical groups (e.g., infants or children with neuromuscular disorders) are at increased risk of complications following respiratory infection, staff should be encouraged to keep up to date with relevant immunisations.

### 5.4. Sleep recording rooms and environmental conditions

A paediatric sleep facility should engender an environment that is non-threatening and child friendly. Patient recording rooms must be designated body-protected electrical areas. Bedrooms must be child safe with appropriate bedding for age (e.g., cots) that meets up-to-date industry standards. Staff must be trained in the safe operation of cots and beds. Bedroom and bathroom amenities need to accommodate wheelchair bound patients and patient lifters need to be available where needed to transfer the patient to and from the bed. Staff must be adequately trained in the safe operation of patient lifters (annual refresher training is recommended if use is infrequent).

Space and amenities for the parent or caregiver to sleep in the same room as the child during the test procedure should be available. This will enable the parent or caregiver to reassure and comfort the child if needed as well as provide routine night time care (e.g., toileting or feeding). Co-sleeping of a parent/caregiver and child in the same bed during a PSG is not ideal as it may (i) not be safe or practical in the laboratory setting, (ii) cause difficulties with staff accessing sensors, (iii) cause sleep disturbance to the child, (iv) result in signal artefact if the parent/caregiver lies on or makes contact with sensors, and (v) the parent may obscure the child from view of the video camera - all of which may impact on PSG interpretation. However, many children co-sleep with parents and may have difficulty sleeping if this is not permitted. The decision to permit co-sleeping should therefore be made at the local level depending on safety concerns, site specific policies and practicalities. Any indications of unsafe sleeping practices that may be happening in the home should be brought to the attention of the referring physician so that parents can be appropriately counselled. Alternative strategies include allowing the parent or caregiver to lie in the bed with the child until the child is asleep or moving the parent bed next to the child's bed. In some cases children with neurodevelopmental disorders may have an assistance dog which should be permitted to be stay with the child throughout testing.

Appropriate storage and heating facilities for milk or infant formula must be available with facilitation of overnight feeds for

infants/young children during PSG recording. Provisions must be made for the appropriate storage of patient medications. If stored in a central location, medication and milk/infant formula must be labelled with appropriate patient identifiers.

Bedrooms should have effective soundproofing, lighting and temperature control. The latter is particularly important for premature infants and neonates as they have poor thermal control. If a premature infant or neonate becomes too cool, motor activity increases resulting in restless/fragmented sleep. Furthermore, both low and high ambient temperatures have been shown to alter arousal thresholds as well as apnoea frequency and duration in infants [126,127]. High ambient temperatures may also degrade the quality of electrophysiological signals during PSG in the form of sweat artefact so it is important to be able to cool recording rooms if needed.

Provisions need to be made for medical emergencies including an emergency call system in patient areas and a complete range of age-appropriate resuscitation equipment as well as oxygen and suction located at the bedside. The patient's name, age and current body weight should be readily available in the event that medication needs to be administered in an emergency. A management plan should be in place for children with epilepsy in the event that a seizure occurs. Staff should consult with the parent or caregiver as to the typical presentation of the patient's seizures and any known triggers. A paediatric sleep/respiratory physician should be on call during sleep studies should advice be needed during the study. Clinical criteria for contacting the on-call paediatric sleep/respiratory physician during the study should be in place in the event of severely abnormal PSG findings or other concerns. Similarly a mechanism must be in place that alerts day staff (scoring analyst and/or the referring physician) that a child has had an abnormal PSG where urgent intervention or treatment may be required. Consideration should be given for a mock medical emergency exercise to be undertaken on an annual basis to ensure staff are aware of the correct emergency protocols and to ensure that emergency procedures are up to date and effective. This is particularly important if a sleep disorders service is situated in an isolated location or where relatively few staff may be present in the immediate vicinity outside of normal business hours.

## 6. Methodological and technical considerations for performing paediatric polysomnography

The AASM manual for the scoring of sleep and associated events (2007) together with associated documentation [128–131] provides guidelines for PSG recording and interpretation (including paediatric PSG) which have been valuable in standardising practice, particularly in North America. In 2011, the Australasian Sleep Technologists Association and the Australasian Sleep Society produced consensus guidelines for recording and scoring sleep studies in children in Australia and New Zealand. In 2012 the AASM guidelines were updated [132] and currently version 2.2 is the most recent published scoring manual produced by the AASM [4]. Read together, the AASM and ASTA/ASA documents provide practice parameters for recording and scoring of sleep studies in children, including technical requirements, recording specifications, scoring of sleep and respiratory events. The reader is referred to these documents as the information contained therein will not be repeated here.

### 6.1. Polysomnography – patient preparation

#### Preparation of patients prior to polysomnography

Polysomnography is a complex physiological test that is especially demanding of young children and parents and the methods

and facilities used for conducting adult sleep studies may not meet the needs of children. Successful testing depends on a child-friendly environment and staff experienced in caring for children in health care settings. Particular challenges for the paediatric sleep technologist will be dealing with children who are scared, too young to understand what is happening, developmentally delayed, have an aversion to tactile or somatosensory stimuli or who have undergone numerous or recent painful hospital procedures. Furthermore many children being evaluated for a sleep disorder will have neurobehavioural or medical co-morbidities which can be challenging to manage. Lastly, parents may also be very anxious about their child having a sleep study, particularly with respect to the application of electrodes and sensors.

Preparation and education of the child and family prior to the study enhances the quality of data obtained as it reduces anxiety and improves the likelihood of a successful study. Ideally this should be done at the time of the clinic visit including an explanation of why the child needs the test and what will be involved. In addition, families should be given printed information or photographs to take away which explain in simple terms what will happen during testing and any preparations they need to make. Provision for visiting the sleep laboratory or trying on some of the sensors beforehand may also be helpful in reducing anxiety in some children. Desensitisation activities prior to testing for children with tactile or other aversions (e.g., nasal cannula) may also be beneficial. A parent or adult care-giver should be present with the child throughout the test and age-appropriate distractions available to assist staff in setting up of the monitoring leads. Ideally the adult staying overnight should not snore although this is not always possible or practical. Encouraging children to bring their own pillow, wear their favourite pyjamas and bring a favourite toy can make the experience less alienating. Ideally the timing of the test should be appropriately geared to the bedtimes of young children. Where appropriate an interpreter may be helpful in explaining the test preparation and procedure to families from non-English speaking backgrounds.

#### Illness and Immunisation

In general polysomnography should not be performed while a patient is acutely unwell, as illness (e.g. fever, vomiting) may obscure PSG findings or the acute illness may exacerbate the true nature of sleep disordered breathing or other sleep problems. Furthermore certain respiratory symptoms (e.g., a runny or blocked nose) may create difficulties in obtaining good airflow signals during PSG leading to uncertainties in test interpretation and diagnosis. It is also advisable not to perform PSG on the same day as immunisations as immunisation may result in a fever and restless sleep.

#### Medications

Certain medications may alter normal sleep and respiratory physiology. For example:

- Hypnotics, sedatives and opioid based analgesics may worsen or induce sleep-disordered breathing and may alter sleep architecture and arousal thresholds.
- Antidepressant, antipsychotic and some antiepileptic medication may alter sleep architecture, arousal thresholds and features of the sleep EEG (e.g., spindles) and muscle tone.

Other medications that may be commonly encountered in the paediatric setting and which may influence PSG findings include caffeine, melatonin, nasal steroids, antihistamines, methylphenidate

or dexamphetamine and clonidine. Administration of sedating agents for the sole purpose of facilitating sleep during a PSG is not recommended. It is the responsibility of the sleep clinician ordering the test to decide whether the PSG should be performed on any medication that may impact respiratory or sleep physiology or whether the PSG should be performed following the withdrawal of confounding drugs. Considerations should include:

- Whether the medication is being used in the acute setting (e.g. to treat intercurrent illness) or in the chronic/long term setting thereby representing the patient's usual circumstances.
- Contraindications to medication withdrawal.
- The clinical indication for the PSG.

In some circumstances a study both on and off medication may be desirable. Withdrawal of medication prior to a PSG needs to be undertaken with appropriate medical supervision, and a suitable washout period should be instituted to minimise rebound effects. For MSLT studies, medications that could affect the results of the MSLT should be discontinued two weeks prior to the study (if not medically contraindicated). The paediatric sleep physician reporting the test should be fully aware of the condition of the patient at the time of sleep study including medication usage and interpret the result accordingly.

#### *Activities on the day of Polysomnography*

Patients should be instructed to follow normal activities of daily living prior to presentation for their PSG. Unless the study is being performed for a special purpose, patients should maintain their regular sleep habits prior to the study. It is advisable to ask patients to refrain from consuming caffeinated or energy drinks on the day of the PSG.

#### *Preparation of patients during polysomnography*

Preparing a paediatric patient for a sleep study requires a flexible and adaptable approach depending on the age or developmental level of the child and their medical history. While individual sleep facilities may differ with respect to patient preparation, at the start of PSG the following issues should be anticipated:

- Site specific protocols for obtaining informed consent should be followed.
- Sufficient information about each patient's clinical history should be made available to staff performing the PSG in the event that the on-call physician needs to be contacted or if a medical emergency should arise.
- The sleep technologist performing the test should be provided with the necessary patient and test information to maximise the quality and integrity of the PSG. Reviewing the child's last PSG, particularly with respect to how the child coped with the procedure, may be helpful in anticipating what might happen. Any potential medical emergencies or concerning events (e.g., breath holding spells) should be made known to staff before starting the study.
- If a considerable amount of time has elapsed between when the patient was referred by the paediatric sleep physician and the test date, then the sleep technologist may need to consult with the referring or on call sleep physician if there are concerns about changes in the patient's medical history or status.
- The patient should be assessed for any potential contraindications to PSG methodology. For example, care needs to be taken in cleaning electrode sites in children with haemophilia and EEG electrodes should not be applied on suture lines in children who have undergone cranial or neurosurgery.

- Prior to starting the PSG setup it is important to ascertain if the patient has any allergies to any of the tapes/adhesives, pastes or cleaning products used during the procedure. If there is uncertainty about a particular product apply a small amount on the underside of the patient's forearm and monitor for approximately 20–30 min for any signs of skin reaction. Patients should be screened for any allergy to products containing latex with consideration given to using only latex-free gloves in the unit.
- Attaching the necessary electrodes and sensors can be a challenging process for both the technologist and the child. Where possible, engaging the parent or caregiver during the PSG setup will help reassure an anxious or scared child. Having the child sit on a parent's lap or allowing the child to place a few electrodes on the parent or a doll/teddy can be helpful in eliciting cooperation. Useful distractions include stickers, interactive books, toys or games (e.g., hand held water games), colouring in or watching a non-stimulating video. Strategies for making PSG more child friendly and dealing with children with neurodevelopmental disorders have been published [133–135].

#### *Repeat Polysomnography*

Repeat PSG may be indicated in children who are receiving continuous positive airway pressure (CPAP) therapy or non-invasive ventilation (NIV) to assess whether therapy is still required or to check that current levels of therapy are appropriate. Growth and/or treatment (e.g., surgery, weight loss) may render CPAP/NIV unnecessary or may require changes to therapy settings. Alternatively a repeat “diagnostic” PSG may be required to assess the necessity of respiratory support in children who are non-compliant with therapy or who are having significant side effects. There is evidence in adults of a CPAP washout effect, i.e. CPAP therapy may have a residual beneficial effect on the severity of sleep-disordered breathing (temporarily less severe) for several days after withdrawal from treatment [136] but this has not been specifically studied in children. It has been recommended that CPAP be discontinued several days prior to PSG if considering discontinuation of therapy [137], however this should only be undertaken at the clinician's discretion. Some children may need to be progressively weaned from NIV therapy. The provision of an oximeter for the duration of the NIV withdrawal period may be a judicious safety precaution in certain cases.

#### *6.2. Polysomnography – test procedure*

##### *Equipment Calibrations*

The integrity of the high frequency physiological signals collected during PSG (EEG, EOG, EMG, ECG) is highly dependent on the amplifiers, gains and filters of the PSG recording system. It is therefore recommended that two types of machine calibrations are performed to check the accuracy of the high-frequency amplifiers and the high and low pass filters [138–140].

- An all-channel calibration where all the channels are set to the same gain and filter settings and a signal of known properties (usually a negative 50  $\mu\text{V}$  DC signal) is sent through all the channels simultaneously. This tests the integrity of the amplifier as each channel should respond the same way with respect to signal polarity, amplitude and decay time constant.
- A montage calibration where the gain and filter specifications used during the PSG recording are set for each channel as is appropriate for the physiological parameter being recorded. Once again a signal of known properties is sent through all the channels simultaneously and each channel is

checked to confirm the signal response is appropriate (if not gain or filter settings may not be set or working correctly). Recording channels with similar settings (e.g., EEG and EMG) should display an identical output.

Regular calibration of any ancillary monitoring equipment (e.g., DC output devices such as pulse oximeters or transcutaneous carbon dioxide (TcCO<sub>2</sub>) monitors) is essential to ensure accurate signal acquisition and processing by the PSG system. For continuous signals (e.g., pulse oximetry (SpO<sub>2</sub>), TcCO<sub>2</sub>, PAP pressure, sound level) a minimum of 3 points (if possible) should be calibrated that span the expected range of the measurement (as two points will always give a straight line!). In the case of step calibrations (such as body position and room light) a calibration of each level/type should be performed. All physical calibrations should be appropriately labelled and permanently recorded.

The frequency of equipment and signal calibrations depends on the stability of the transducer and the likelihood that an intrinsic or extrinsic factor could cause an error in the value reported. If the signal is critical to the interpretation of the study, for example SpO<sub>2</sub> or PAP pressure, it should be calibrated prior to each study. If loss of the signal would still allow interpretation and the transducer is stable, less frequent calibration is acceptable. In most systems the gain is stable over a long period of time and monthly calibration of these amplifiers is adequate.

#### Sensor and electrode application

Minimum technical and recording specifications for paediatric PSG including sensor and electrode placement are detailed in:

- The AASM Manual for the scoring of sleep and associated events [4].
- Rules for scoring respiratory events in sleep: Update of the 2007 AASM Manual for the Scoring of Sleep and Associated Events [132].
- The ASTA/ASA addendum to AASM Guidelines for recording and scoring of paediatric sleep [3].

The attachment of sensors and electrodes should be standardised to ensure consistent and accurate results for PSG interpretation. Variances from normal methodology should be clearly documented on the study log with an explanation of the reason for the variation. Any such variations should be disclosed to the reporting paediatric sleep physician. It is beyond the scope of this document to discuss the precise methods for electrode and sensor application and this information should be detailed in a protocols and procedures manual. However there are a number of considerations that are of particular relevance in the paediatric population:

- The skin of children (especially infants) is much thinner than in adults and care needs to be taken with cleaning electrode/sensor sites and removing adhesive tapes. Similarly, care must be taken when using sensors that produce heat – e.g., the oximeter and TcCO<sub>2</sub> probe. These sensors should be repositioned during the recording period to minimise the likelihood of patients receiving skin damage.
- EEG electrodes should not be applied over the fontanelle of infants.
- It is not uncommon for infants or children to have allergies or sensitivities to tapes and adhesives. Hypoallergenic products should be used if this is the case.
- Children with neurodevelopmental disorders often have sensory sensitivities particularly tactile or somatosensory aversions which can make applying electrodes and sensors challenging. This should be made known to staff prior to the PSG visit as it

may necessitate a higher staffing level at the start of the night. Engaging the parent/caregiver during the PSG process will be important as they best know their child. Desensitisation prior to the PSG may need to be considered if the sensitivities are severe.

- Despite the best efforts of staff there may be occasions when children are intolerant of the sensors and electrodes used during PSG. Guidelines should therefore be in place for these situations indicating the minimum sensors or electrodes that are needed to obtain clinically useful information.
- An extended bilateral EEG montage with continuous video monitoring should be considered for patients if the following are present or clinically suspected: seizures, parasomnias, unexplained hypersomnolence that does not appear to be classic narcolepsy, unusual sleep-related body movements or sleep-related paroxysmal events/arousals.
- Some children with refractory epilepsy may present with a vagal nerve stimulator (VNS) *in situ*. This should be noted on the study log and/or PSG report as VNS activation is known to cause reductions in airflow. The resultant flow limitation may be severe enough to warrant treatment by CPAP [141]. Activation of the VNS can usually be seen as artefact on the submental EMG which can be correlated with any reductions in airflow. Alternatively, an additional surface electrode can be applied close to the site of the VNS electrode in the neck to detect activation [135].
- Due to the higher risk of entanglement with electrode and sensor cables, infants and children must be under continuous visual surveillance while setting up and conducting PSG. Furthermore parasomnias such as sleep walking which are more common in children and injuries may result if children abruptly try to get out of bed. Most modern PSG systems now come with synchronised infra-red recording and cameras with zoom-in ability are invaluable for scoring sleep in infants where behavioural observations such as eye opening or closure are important adjuncts to the EEG in determining wake or sleep.

#### Electrode Impedance Verification

Once all sensors and electrodes are attached the sleep technologist should visually check all signals to verify they are displaying correctly and free of artefact. An impedance check should be conducted (and recorded) for all the AC recording channels (EEG, EOG, EMG, ECG) including the reference and ground channels. Ideally Impedances for individual electrodes should be below 5 KΩ. It is also important that impedances are as equal as possible between paired electrodes otherwise common mode rejection (elimination of electrical interference) will not be optimised. However it is not always possible to obtain low impedance levels in paediatric studies for a variety of reasons including patient tolerance and compliance. While striving to obtain low impedances is strongly encouraged, there will be situations where a compromise may need to be made in order to be able to collect some information rather than none at all. Discretion should be used in situations where the signal quality looks reasonable and can be interpreted but the entire PSG might be at risk if further attempts to obtain lower impedances are made. An impedance check should be performed again near the end of the PSG recording to confirm signal integrity or on any occasion during the study if it appears that signal quality has diminished or there is evidence of artefact in the signal.

#### Patient Bio-calibrations

Patient bio-calibrations are important for validating signal integrity and responsiveness and serve as a baseline scoring reference for each subject. Bio-calibrations should be performed at the beginning of each PSG once impedance checks have been done and corrective action taken if needed. Examples of commonly



performed bio-calibrations are shown below. Each manoeuvre should be conducted for an appropriate length of time to verify signal response. For younger or developmentally delayed children abridged bio-calibrations may be possible for the simpler instructions. For infants or toddlers signal responsiveness and sensitivity may be assessed by viewing the patient spontaneously performing various actions and confirming that the signals are responding appropriately. Bio-calibrations should be recorded as part of the PSG in the event they are needed during PSG scoring for signal referencing or corroboration.

Recording Parameter Verified	Patient Activity
EEG (alpha reactivity)	Eyes open, staring straight ahead
EOG	Eyes closed, keeping head still
EOG	Move eyes left/right, keeping head still
	Move eyes up/down, keeping head still
	Blink, keeping head still
Submental EMG	Grit teeth
	Swallow
	Smile
Respiratory bands	Hold breath
Nasal pressure	Deep/big inspiration
Thermocouple/thermistor	
Diaphragmatic/intercostal EMG	
Nasal pressure	Breathe through nose only
Thermocouple/thermistor	Breathe through mouth only
	Deep/big breath in
Anterior tibialis EMG	Flex left leg
	Flex right leg
Microphone/sound	Snoring sound
	Cough

### PSG Documentation

Adequate documentation by the sleep technologist during the PSG is crucial for interpreting the results. Even though the PSG system may have audio-visual recording capacity not everything may have been clearly captured during the recording process. For example, if snoring is coming through the audio-channel, is it that of the child or the accompanying parent/caregiver? Any technical difficulties encountered during the PSG should be documented including what action was taken to try and fix the problem. Therapy changes (PAP or supplementary oxygen) must be precisely documented. Any notable or unusual events or abnormalities observed by the sleep technologist should be also documented and disclosed to the scoring analyst and/or the reporting paediatric sleep physician. Significantly abnormal findings must be promptly reported to relevant staff following an overnight study so that appropriate intervention or treatment can be actioned.

Helpful information to collect or document includes:

- What time the child woke on the morning prior to the PSG and if any daytime napping occurred.
- If the child was displaying any respiratory symptoms upon presentation for the PSG.
- Any medication taken during the night and at what time.
- If the child fell asleep before the PSG recording started – this may explain any unusual features of the sleep architecture at the start of the night (e.g., reduced REM sleep latency).
- Any technical or procedural variations from normal methodology that may have occurred with an explanation of the reason for the variation.

- Any external disturbances that may have aroused the child during the night (e.g, overhead announcements on the public address system).
- Any observations or interactions between the sleep technologist and the patient/or family that may be relevant clinically relevant. For example, was the parent/caregiver not confident in applying the PAP mask to the child? Is a new mother displaying signs of post-natal depression?
- Did the child or parent feel that the individual's sleep was representative compared to what normally happens at home?

Additional documentation that should be chronicled or recorded with each PSG (or related test) includes: the referral information; the date and time of the test; the identity of all staff performing various aspects of the testing; the equipment used in testing including the name and version of software used for PSG acquisition and scoring.

### Study Duration and Split Night Studies

A PSG recording should be at least 8 h in duration and should ideally collect at least 6–8 h of sleeping data (depending on age). A PSG during which less than 6 h of sleep was observed or that contained less than 6 h of technically sound data should be interpreted with caution particularly if REM sleep was not observed.

Split night sleep studies including a diagnostic and treatment component are performed less often in children than adults. There are a number of reasons for this including potential difficulties with getting an accurate measure of the degree of sleep disordered breathing which may be confined mostly to REM sleep. Furthermore it is likely that most children will not respond well to being started on PAP therapy in the middle of the night without any prior preparation or acclimatisation. This may in turn lead to subsequent aversion to accepting treatment. However a split night PSG may be appropriate in children who have been receiving PAP support and are who are being evaluated for potential discontinuation of PAP therapy.

A split night PSG may also be appropriate in patients (generally infants) presenting with uncomplicated cyclical central apnoea/periodic breathing in order to institute and titrate supplementary oxygen therapy. Protocols should be in place for this clinical scenario.

### 6.3. Polysomnography analysis and reporting

Standards for scoring and analysing paediatric sleep studies are documented in:

- The AASM Manual for the scoring of sleep and associated events [4].
- Rules for scoring respiratory events in sleep: Update of the 2007 AASM Manual for the Scoring of Sleep and Associated Events [132].
- The ASTA/ASA addendum to AASM Guidelines for recording and scoring of paediatric sleep [3].

All studies should be reported by a qualified paediatric sleep physician who reviews the raw/scored data. The patient's relevant clinical data must be disclosed to the reporting physician to aid in interpretation of the test result. At a minimum this should include the age and associated medical condition(s) of the child and medications taken at the time of the sleep study. In addition the reporting physician should review or be aware of the technical observations made by the overnight and scoring technologist. All

studies should be reported in light of this clinical and technical information.

#### Severity Criteria for Sleep Disordered Breathing

The obstructive apnoea–hypopnoea index (OAH) has been used in clinical trials and epidemiological studies to classify patients as either having OSA or being normal, as well as to classify the severity of OSA.

The following classification of OSA severity is commonly used in clinical practice:

- Normal: OAH <1 events per hour of sleep
- Mild OSA: OAH 1 – ≤5 events per hour of sleep
- Moderate OSA: OAH 5 – ≤10 events per hour of sleep
- Severe OSA: OAH >10 events per hour of sleep

A classification for the severity of central and non-obstructive sleep disordered breathing has not been established.

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#### Conflict of interest

The following conflicts or potential conflicts of interest were declared by members of the committee:

All authors have no conflicts of interests.

1. Current or recent (last 3 years) involvement with a company or companies with a financial interest in devices or methods for performing sleep studies:
  - a. Direct financial interest
  - b. Employee, or engaged in a consulting capacity (including medical advisory boards, expert testimony)
  - c. Substantial research support
  - d. Sponsored attendance at national or international meetings
2. Financial benefit received (personally, spouse or dependents, or department) from performing or reporting sleep studies
  - a. Direct benefit received
  - b. Departmental benefit received

Individual COI statements are available from the secretariats of the Australasian Sleep Association and the Thoracic Society of Australia and New Zealand.

The ICMJE Uniform Disclosure Form for Potential Conflicts of Interest associated with this article can be viewed by clicking on the following link: <http://dx.doi.org/10.1016/j.sleep.2017.03.020>.

#### Appendix I

Children at risk of sleep-disordered breathing should be routinely assessed for clinical indicators of sleep problems with a low threshold for further investigation.

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Specific medical conditions associated with an increased risk for the development of sleep-disordered breathing

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- > Down Syndrome
- > Prader–Willi Syndrome
- > Achondroplasia
- > Neuromuscular disorders (e.g., Duchenne Muscular Dystrophy, Myotonic Dystrophy, Spinal Muscular Atrophy)
- > Craniofacial syndromes affecting the upper airway (e.g., Pfeiffer, Treacher–Collins, Crouzon & Apert’s Syndrome; Pierre Robin sequence)
- > Genetic disorders affecting respiratory control (e.g., Congenital Central Hypoventilation Syndrome, Rett Syndrome, Joubert Syndrome)
- > Metabolic Storage disorders (e.g., Hurler’s Syndrome)
- > Cerebral palsy
- > Leigh’s Disease
- > Chiari malformation
- > Spina bifida

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Risk factors which pre-dispose the development of sleep-disordered breathing

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- > Adenotonsillar hypertrophy
  - > Retrognathia
  - > Mid-face hypoplasia
  - > Macroglossia
  - > Structural abnormalities of the upper airway (e.g., laryngomalacia, cleft palate, choanal stenosis, choanal atresia, vocal cord palsy)
  - > Hypotonia
  - > Chronic lung disease
  - > Premature birth
  - > Chest wall deformity
  - > Scoliosis
  - > Epilepsy
  - > Face and neck burns
  - > Moderate to severe obesity
  - > Family history of sleep disordered breathing
  - > Disadvantaged socioeconomic status
  - > Ethnicity
- 

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