Sleep Studies in Children

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Sleep-related breathing disorders (SRBD), also referred to as sleep-disordered breathing (SDB), are common sleep disorders in children. They can be broadly divided between central and obstructive sleep-disordered breathing with or without associated hypoventilation. In most cases, SRBD are associated with adenotonsillar hypertrophy (obstructive SDB) which are classified as simple. SRBD can co-exist with an underlying condition like obesity, genetic syndromes or neuromuscular disorders which are classified as complex. Polysomnography (PSG) is the gold standard for diagnosing sleep disorders. However, it is time-consuming and requires trained technician to acquire and interpret signals. Attended in-lab respiratory polygraphies are easier to conduct and provide respiratory data equivalent to a PSG. Similar to adult sleep services, overnight unattended home respiratory polygraphies are becoming more widely used. These require careful patient selection and good parental education programs to be most successful in children. Overnight oximetry has limitations but can be a useful tool for screening children with obstructive sleep apnea and prioritizing treatment. This review aims to discuss these various diagnostic methods to assess sleep disorders in children.

Keywords: *Adenotonsillar hypertrophy, Diagnosis, Polysomnography Sleep-related breathing disorders.*

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The International Classification of Sleep Disorders 3 (ICSD-3) broadly classifies sleep disorders into insomnia, sleep-related breathing disorders (SRBD), central disorders of hypersomnolence, circadian rhythm sleep-wake disorders, parasomnias, sleep-related movement disorders and other sleep disorders [1].

Sleep-related breathing disorders (SRBD) commonly referred to as sleep disordered breathing (SDB) is an umbrella term for chronic conditions with partial or complete cessation of breathing occurs many times throughout the night. This leads to sleep fragmentation and impacts gas exchange with night-time symptoms, daytime symptoms and long-term deleterious health effects. SDB is divided into obstructive sleep disordered breathing and central sleep disordered breathing with or without hypoventilation. The subtypes of SDB are not exclusive and several subtypes can be present in a child depending on the clinical situation.

The commonest type of SDB is obstructive sleep apneas/ hypopneas syndrome (OSAHS). OSAHS is a disease spectrum varying from prolonged partial airway obstruction (snoring and upper airway resistance) to intermittent complete upper airway obstruction (obstructive sleep apnoea or OSA). OSAHS can be classified as simple or uncomplicated when it occurs only in association with adenotonsillar hypertrophy. It can be classified as complex or complicated when it is associated with other medical disorders (e.g., neuromuscular diseases, chronic lung diseases, sickle cell disease), genetic syndromes (e.g., Down syndrome, obesity syndromes, craniofacial anomalies, Pierre Robin sequence) and other high risk comorbidities such as obesity.

Children with complex OSAHS often have multilevel airway obstruction related to their craniofacial morphology and upper airway tone. Morphological features include midface hypoplasia, flat nasal bridge, retrognathia, glossoptosis amongst others. In this complex group, it is not unusual to have a multifactorial complex SDB picture with a combination of obstructive, central and hypo-ventilation components [2].

Sleep negatively affects control of breathing, lung mechanics and respiratory muscle contractility with reduction in functional residual capacity and minute ventilation. Upper airway resistance also increases during sleep. Hence, SDB is seen in many chronic illnesses where detailed sleep evaluation and management should be considered. These include intrinsic cardiopulmonary disorders (advanced cystic fibrosis, bronchiectasis, bronchiolitis obliterans etc), chest wall abnormalities like kyphoscoliosis or thoracic dystrophy, neuromuscular diseases, spinal cord injury or genetic disorders like Prader Willi syndrome [3,4].

EVALUATION

Evaluation and diagnosis of childhood sleep disorders may include sleep questionnaires, sleep diaries, actigraphies and sleep studies. This review will detail these with an emphasis on pragmatic sleep diagnostic tools for developing countries.

Sleep Questionnaire

Many questionnaires have been used for diagnosis of simple OSAS, with pediatric sleep questionnaires (PSQ) by Chervin, et al. [5], being used most widely. PSQ has a sensitivity of 0.85 and specificity of 0.87 in otherwise well children aged 2-18 years for identifying SDB confirmed by polysomnography. Others have shown a moderate sensitivity and specificity to diagnose SDB. As such, they are useful as a screening tool in primary care. However, it is not a good screening tool for OSAS in children with complex underlying disorders e.g., neuromuscular disorders, cranio-facial anomalies and Down's syndrome [8].

Sleep Diary

Sleep diary is a simple and inexpensive screening tool where parents make a two week daily record of the child's daily sleep routine and sleep related activity. This is done by shading times where child is asleep including during the day and adding visual aids to see when the child went to sleep or woke up. This provides interesting and useful visual representation of sleep. Any additional information provides a useful complement e.g., how refreshing the night sleep was, the amount of exercise or medications and caffeine/food intake particularly in the period before bedtime. Sleep diary is also a useful tool to measure treatment outcomes. However, sleep diaries have their limitations as self-reporting has a subjective element prone to systematic biases. For example, parents had reported total sleep times (TSTs) that were significantly higher (by an average of 1-2 hours) than those reflected in actigraphy recordings of their child [7]. They are; however, easy to implement and can be useful in understanding sleeping patterns. Salient points to be noted on a pediatric sleep diary are shown in **Web Fig. 1**.

Actigraphy

Actigraphy devices are worn on the wrist and record movements (movements and light exposure both in the more modern devices) that can be used to estimate sleep parameters (sleep onset, sleep duration, wake time) with specialized algorithms in computer software programs. It has the advantage of providing objective information on sleep habits in the patient's natural sleep environment.

Actigraphy is well validated for the estimation of night-time sleep parameters across age groups. In patients reporting significant sleep disruption, it can objectively document sleep patterns and evaluate treatment outcomes. They provide a visual map of a child's sleep and are less biased than the sleep diary. Typically two weeks of recording is recommended; however, this may vary depending upon the sleep parameters to be evaluated. Once the monitoring period is over the device is removed and data downloaded for evaluation. Actigraphy can be quite useful in evaluating hypersomnias, insomnias and circadian rhythm disorders [8].

SLEEP STUDY

A sleep study is a test that records physiological parameters while the child is asleep. It is usually done in a special sleep laboratory with the equipment to measure all the various physiological parameters including good video and audio recording. Ideally, a trained sleep physiologist sets up the study and monitors the child during the complete study. A good sleep study should have a total sleep time of at least six hours [9]. Sleep study using a non-standard equipment or untrained technicians have poorer data quality and are not routinely recommended. Unobserved studies currently have poorer data quality which may be overcome with the development of trained home sleep services. Recommendations for pediatric sleep investigations are currently the subject of British Thoracic Society guidelines with release expected in the near future.

Broadly, sleep studies can be classified as diagnostic or ventilation titration studies [non-invasive (NIV) or invasive via tracheostomy (long term ventilation or LTV)]. Diagnostic studies include simple overnight oximetry, oxy-capnography, cardiorespiratory polygraphy (RPG) or a complete polysomnography (PSG). Details of how these studies are performed, scored and interpreted are available from the American Academy of Sleep Medicine (AASM) [10].

Diagnostic Sleep Studies

Oximetry: Overnight oximetry studies are unobserved downloadable studies done using a timed pulse oximeter. These studies might be domiciliary or in-hospital. The use of an appropriate oximeter is paramount for accurate data interpretation. For example, averaging time is a crucial setting in assessing the diagnostic efficiency of the oximeter. Longer averaging times (8-16s) may reduce signal artefact (e.g. from motion) but also reduce the ability to detect the rapid change in saturation (SpO2) often seen with central or obstructive events (apnea/hypopnea). Therefore, a pulse oximeter with an averaging time of 2-3 seconds should be used to maximize diagnosis efficiency rather than the routine ICU pulse oximeters with usually longer averaging times. The oximeters should have the facility to download and review data in a way that is useful for interpretation of SDB. These studies are easy to perform and cost effective. They are useful in evaluating oxygenation in children on domiciliary

oxygen and help with weaning supplemental oxygen. Recent technical guidelines are available on overnight oximetry in children [1]. Overnight oximetry can also be useful to screen children with non-complex OSAHS for moderate to severe disease and help prioritize treatment. McGill's scoring [12], is done using the pulse oximetry trace. At least three clusters of desaturation events, and at least three SpO2 drops below 90% in a nocturnal oximetry recording are indicative of moderate-to-severe OSAHS [12]. Abnormal oximetry had 97% positive predictive value to detect OSAHS diagnosed by in-laboratory PSG; however, sensitivity was 43%, indicating that patients with an inconclusive oximetry could still have OSA. Therefore, in the context of significant symptoms, an inconclusive oximetry is not enough to rule out OSA [13]. In children with OSAHS and co-morbidities, positive predictive value of the McGill score is significantly lower. The higher number of false positives in children with medical comorbidities may be due, to central apneas.

Cardiorespiratory sleep studies or respiratory polygraphy (RPG): This is a limited channel study involving respiratory channels (nasal airflow, thoraco-abdominal movements, oximetry, end-tidal or transcutaneous CO2), cardiac channels (ECG, pulse oximetry) and body position channel. These studies are technician attended in-lab studies with a full audio and video recording and scored manually. Since the number of channels in a cardiorespiratory study are reduced [electroencephalogram (EEG), electro-oculogram (EOG), chin electromyogram (EMG)] it makes the set up and scoring less complex, less time consuming and provides almost the same respiratory information as a complete PSG. RPG can be scored using adapted rules as per the AASM 2012 guidelines for the scoring of sleep and associated events [10]. RPG have previously been demonstrated to be an accurate tool for the detection of SDB [14]. Sleep stages are scored as either wake, active sleep or quiet sleep in 30 second epochs by visual analysis of the cardiorespiratory parameters based on heart rate and respiratory rate variability and amplitude of breathing patterns [15]. It however, provides limited information on sleep architecture. Recent evidence suggests that unattended respiratory polygraphy after being set up in doctor's clinic is feasible, technically acceptable and interpretable in between 81-87% of pediatric patients [16]. However, drawbacks from these particular studies were the absence of audio, video recording, technician monitoring and carbon-dioxide channel. More evidence is currently being collected about home respiratory polygraphies and many centers, following the lead of adult sleep specialists, are now gathering experience [17,18].

Polysomnography (*PSG*): This is a complete study involving the respiratory and cardiac channels previously described for RPG with additional neurological channels (EEG, EOG, chin EMG, leg EMG). Other channels like extended montage EEG, 24h esophageal ph/impedance, diaphragmatic EMG can be added as per clinical need. The details of the channels used in both types of studies are shown in **Table I**.

Scoring a sleep study involves scoring sleep stages and then scoring respiratory events. Sleep staging involves identification of REM and NREM stages (N1, N2, N3) and arousals based on EEG, EOG and chin EMG. Respiratory scoring involves identification of apneas, hypopneas, and hypoventilation as per the AASM guidelines definitions [10]. Apnea is defined as cessation of flow >90% of the baseline for >2 breaths or >10 seconds while hypopnea is defined as flow reduction by ≥30% for >2 breaths or >10 seconds with either a ≥3% oxygen desaturation or an arousal (on EEG). If the events are associated with snoring, flattening of nasal flows or thoraco-abdominal paradox they are classified as obstructive events. Central, obstructive or unclassified events are scored separately.

Table I Channels Used in Polysomnography and Cardiorespiratory Sleep Study or Respiratory Polygraphy

Channel	*Purpose*	*Cardiorespiratory sleep study*[a]	*Polysomnography*
Nasal cannula or thermistor or both	Detects apnea and hypopnea	Yes	Yes
Thoracic and abdominal belts	Respiratory effort	Yes	Yes
Body position sensor	Body position	Yes	Yes
Microphone	Snoring	Yes	Yes
Video recording	Body movements, position, etc	Yes	Yes
Electrocardiogram	Cardiac rhythm	Yes	Yes
Oxygen saturation	Desaturations	Yes	Yes
CO2: Transcutaneous or end tidal	Hypoventilation	Optional	Optional
EEG, EOG and chin EMG	Presence and stage of sleep	No	Yes
Leg EMG	Periodic limb movement	No	Yes

[a]*or respiratory polygraphy. CO2-carbon dioxide; EEG-electroencephalogram; EOG-electro-oculogram; EMG-electromyogram.*

The most important parameter defining SDB in a sleep study (PSG or RPG) is the apnea hypopnea index (AHI) which is defined as number of apneas and hypopneas per hour of total sleep time [10]. The AHI can be further subdivided in OAHI (obstructive apnea-hypopnea index), CAHI (central apnea-hypopnea index) and UAHI (unspecified apnea-hypopnea index for events difficult to characterize). An AHI<1 is considered normal, an OAHI between 1-5 represents mild OSA, 5-10 moderate OSA, and >10 severe OSA. This classification only applies to OAHI and OSA and cannot be extrapolated to central SDB. Additional information is available from video and audio like snoring, gasps, pauses, apneas, work of breathing and sleep posture. **Web Figs. 2-4** shows respiratory poly-graphy epochs of obstructive apnea, obstructive hypopnea and central apnea, respectively. **Web Fig. 5** shows a polysomnography epoch.

Non-Invasive Ventilation Titration Studies

Children deemed to require CPAP or a Bi-level PAP require a ventilation titration study to ascertain the adequacy of ventilation (and optimize the CPAP or bilevel PAP support (determination of optimal pressure settings and assessment of synchronization with the ventilator). During the sleep study, mask fitting and unintentional leak can also be assessed. NIV titrations studies are ideally preceded by a phase of mask fitting and acclimatization. Split night studies are not usually done in children for initiating NIV. They can be done in children when improvement of underlying SDB is suspected and removal of NIV is being considered. Most NIV titration studies can be done as RPG or even oxy-capnography in compliant patients.

Multiple Sleep Latency Test (MSLT)

MSLT is another type of sleep study performed to evaluate children with excessive daytime sleepiness (EDS) and are often diagnostic for narcolepsy. It measures how quickly a child falls asleep in a quiet and dark environment during the day (over 5 naps of 20 minutes separated by 2 hours break) and how often and quickly they enter into REM sleep during those naps. This is usually done after a diagnostic overnight PSG to confirm the absence of OSA as a cause of EDS and to ensure the quality of sleep the night before was decent and will not influence the result of the MSLT [20].

INDICATIONS

Currently, the majority of care givers make a diagnosis of SDB or OSAHS on clinical parameters that include night-time and daytime symptoms of OSAHS in the presence of a predisposing clinical condition like adenotonsillar hypertrophy, obesity, craniofacial syndromes etc. In this context, sleep studies are seldom used as they are deemed expensive, burdensome and are often unavailable for children.

However, contrary to the general belief, the correlation between clinical symptoms and severity of OSA is poor. A meta-analysis on seven models of OSA questionnaires presented moderate sensitivity (0.04-0.94) and specificity (0.28-0.99). Some clinical features such as excessive daytime somnolence and observed apneas had a better specificity but have poor specificity unlike snoring and tonsillar hypertrophy, which had poor specificity [21]. The gold standard for diagnosing SDB and OSAHS is a sleep study. Delayed diagnosis of SDB and OSAHS can significantly lead to increased morbidity. The clinical signs and symptoms further, have poorer sensitivity and specificity in children with complex disorders.

The American Academy of Pediatrics recommends a sleep study in children having regular snoring and any of the additional complaints or findings suggestive of OSA like laboured breathing during sleep with gasps/snorting noises/ observed episodes of apnea, sleep enuresis, sleeping in a seated position or hyperextended neck posture, morning headaches, excessive daytime somnolence, attention-deficit/ hyperactivity disorder and any learning problems. Examination findings include being underweight or obese, having tonsillar hypertrophy or adenoidal facies, micrognathia/ retrognathia, high-arched palate and/or hypertension [22]. The size of tonsils poorly correlates with SDB severity [23]. A sleep study is such situations clarifies the severity of SDB and assists therapeutic decision making [24]. The indications for sleep study are detailed in **Box I** and conditions with complex sleep apneas are detailed in **Web Table I**.

Symptoms of OSAHS in children with genetic syndrome can often be subtle and non-specific. These children often have multifactorial SDB and multilevel airway obstruction indicative of need for an in-lab cardiorespiratory study (RPG) or a PSG is recommended [25]. Children with neuromuscular disorders will often develop nocturnal hypoventilation early which can eventually progress to diurnal hypoventilation. Sleep study is an important component of their evaluation and follow up [26-28].

Most typical parasomnias like confusional arousals, sleep walking and night terrors can be diagnosed based on clinical presentation ideally supplemented with a good video recording of the event. Sleep study is not necessary for diagnosis. A comprehensive in-laboratory video-PSG is recommended to evaluate parasomnias which are: *i*) unusual or atypical because of the patient's age at onset; the time, duration, or frequency of occurrence of the behaviour; or the specifics of the particular motor patterns in question (e.g., stereotypical, repetitive, or focal); *ii*) potentially injurious or have caused injury to the patient or others; and/or *iii*) potentially seizure-related but the initial clinical evaluation and a standard EEG are inconclusive.

Box I Indications of a Diagnostic Sleep Study

Respiratory indications of a sleep study (respiratory polygraphy or complete polysomnography)

Suspected sleep disordered breathing in association with adenotonsillar hypertrophy

Complicated sleep disordered breathing in the setting of:

- Genetic syndromes like Craniofacial syndromes (Apert, Crouzon, Pfeiffer), achondroplasia, Down syndrome, Obesity syndromes (e.g., Prader Willi), Mucopolysaccharidoses, Pierre Robin sequence etc.
- High risk groups like chronic lung diseases (eg CLD of prematurity, bronchiectasis, CF), obesity, Chiari malformation, sickle cell disease, airway disease (vocal cord palsy, tracheo-bronchomalacia)

Neuromusular disorders with suspected nocturnal hypoventilation

- Spinal muscular atrophy (SMA) Type 1, 2, 3, SMA with respiratory distress (SMARD)
- Duchenne muscular dystrophy (after loss of ambulation)
- Congenital muscular dystrophy: Ulrich, Rigid spine
- Congenital myopathies: minicore, nemaline rod, myotubular
- Myotonic dystrophy type I
- Congenital myasthenic syndromes
- Hereditary sensory motor neuropathy (HSMN)
- Mitochondrial myopathy

Non-respiratory indications of a sleep study (complete polysomnography)

- Atypical or potentially harmful parasomnia vs nocturnal seizures when the initial clinical evaluation and standard EEG are inconclusive (expanded EEG montage)
- Periodic limb movement disorder (PLMD)
- Children suspected of having restless legs syndrome (RLS) who require supportive data for diagnosing RLS
- Hypersomnolence (suspected narcolepsy) (PSG followed by a MSLT)
- Insomnias and circadian rhythm disorders to confirm an underlying SDB, RLS or PLMD

Restless legs syndrome in children can be diagnosed on clinical presentation and a sleep study is usually not necessary. Sleep study might be required to assess sleep quality (including apnea) which may worsen RLS or to assess periodic limb movements in sleep as a supportive tool for making a diagnosis of RLS [29,30]. Most children with insomnias and circadian rhythm disorders can be diagnosed with a sleep diary supplemented with an actigraphy. PSG might be required to confirm an underlying SDB, RLS or PLMD. Children with excessive daytime somnolence and suspected narcolepsy require a PSG to rule out a SRDB and ensure quality of sleep prior to a MSLT the day after.

LIMITATIONS OF A SLEEP STUDY

Often the sleep study result of a single night is taken into account for decision making. However, this may not be reliable as the patient's sleep can be affected by the unfamiliar surroundings leading to a poor night sleep. This often requires a second sleep study. Sporadic events like parasomnias and seizures can also be missed on a single night study. The family and the child undergoing a sleep study have to remain in a sleep laboratory hooked up on sleep study equipment that can sometimes affect sleep quality. Sleep studies also require laboratory set up, training of sleep technologists and adequate staffing to conduct, score and report sleep studies.

Pediatric sleep centers require multi-disciplinary involvement with a pediatric pulmonologist and sleep specialist and ideally, a pediatric neurologist, a craniofacial surgeon, a pediatric ENT surgeon, a pediatric endocrinologist and a pediatric cardiologist. It also requires well trained paraclinical team of sleep physiologists, child psychologists and play therapists. The diagnostic options need to be prioritized in clinical context for the best outcome of a sleep study.

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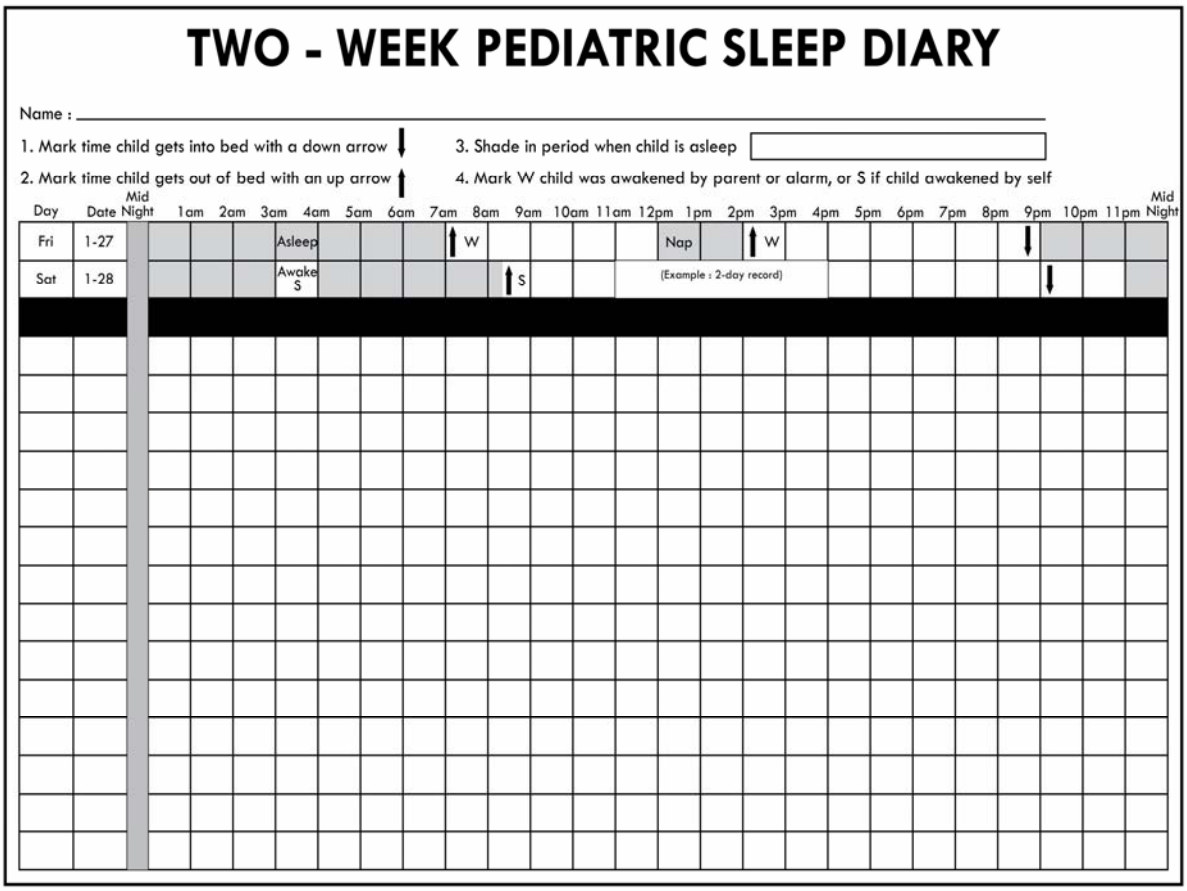
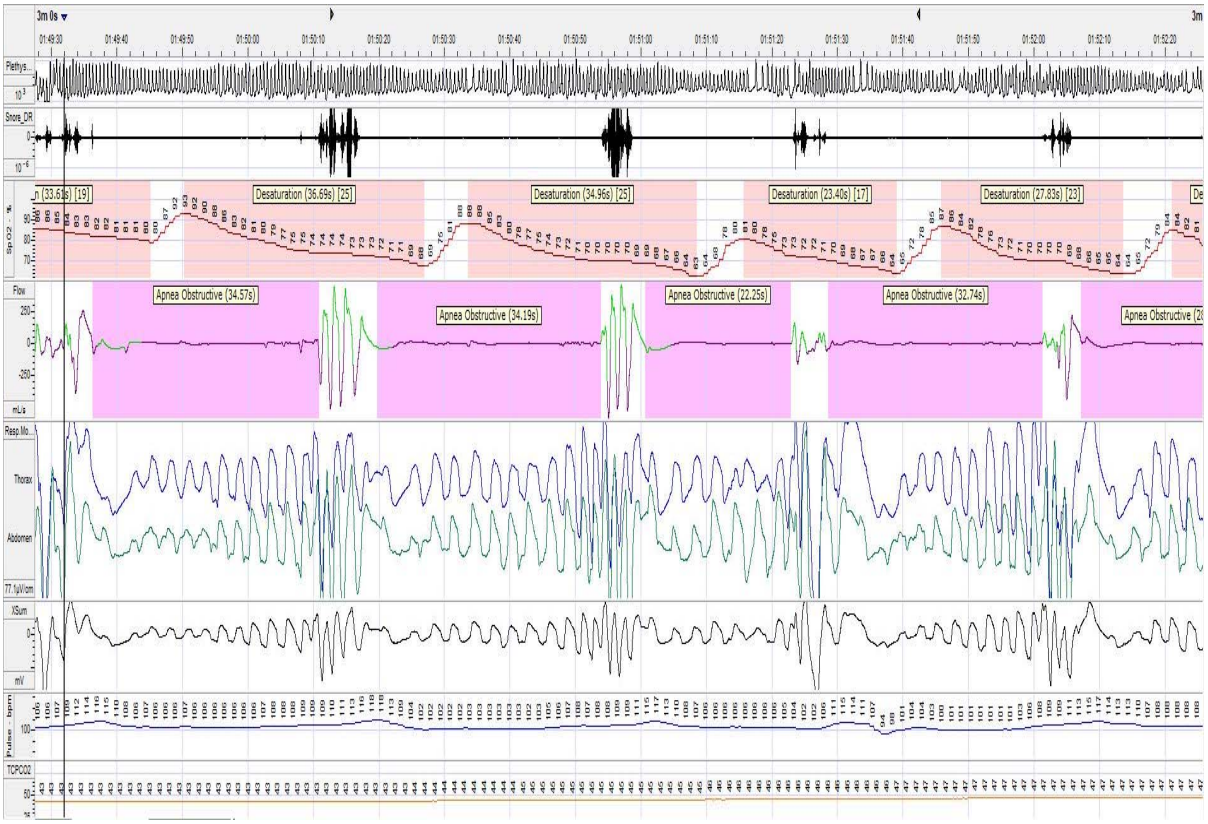
Web Table I Complex Sleep Disordered Breathing in Children

Condition	*Factors predisposing to SDB*	*Recommendation for testing*
Craniosynostosis and Craniofacial syndromes (Apert, Crouzon, Pfeiffer, Muenke and Saethre Chotzen)	Midface hypoplasia. Chiari malformation is a frequent finding in Crouzon (70%) & Pfeiffer (50%) syndromes and can have associated central sleep apnea	Yearly screening in patient with Apert, Crouzon and Pfeiffer syndromes. Muenke and Saethre Chotzen syndromes evaluate only if they become symptomatic
Down's syndrome	Relative macroglossia, midface hypoplasia, reduced muscle tone, small upper airway, adeno-tonsillar hypertrophy, obesity	All children before age of 4 years. In all children pre adenotonsillectomy
Cleft lip/palate	Small size of pharyngeal airways & craniofacial dimensions. Oropharyngeal musculature is disrupted by the cleft, which impacts on speech and swallow, as well as adversely affects maintenance of airway patency during sleep.	Sleep study is recommended if child is symptomatic, in the presence of syndromic cleft lip/palate or pre pharyngoplasty and pharyngeal flap surgery.
Syndromic micrognathia (i.e. Pierre Robin Sequence, Stickler syndrome, etc)	Micrognathia, glossoptosis, midface hypoplasia	All children should be screened
Obesity syndromes (eg Prader Willi, Bardet Biedl)	Obesity, hypotonia, micrognathia, small naso and oropharynx, scoliosis leading to OSA. Central sleep apneas, nocturnal hypoventilation, hypersomnolence/cataplexy also seen	Yearly sleep studies are recommended. Sleep study pre growth hormone therapy & post 3-6 m post therapy & afterwards if symptoms of SDB reappear. MSLT if hypersomnolence
Mucopolysaccharidosis	Cranial and spinal abnormalities (e.g., flattened nasal bridge, short neck, mandibular abnormalities) and glycosaminoglycans deposition in the mouth, nose, throat	All patients should be evaluated at diagnosis
Achondroplasia	OSA: Facial hypoplasia, retruded position of chin, adeno-tonsillar hypertrophy. Central sleep apneas also seen.	Should be screened at least once from 1 yr and then subsequently (or prior) if symptoms develop
Duchenne muscular dystrophy, limb-girdle muscular dystrophy, fascio-scapulo-humeral	Decreased respiratory drive, muscle weakness; pharyngeal muscle weakness, scoliosis, obesity, recurrent lung infections. OSA, central apneas & hypoventilation seen	FVC <60% predicted, symptoms of nocturnal hypoventilation or when children become non-ambulatory. Follow up annually.
Spinal Muscular Atrophy Congenital Myopathies Congenital Muscular Dystrophies Myotonic Dystrophy Congenital myasthenic syndromes Hereditary Motor Sensory Neuropathy	Same as above. OSA, central apneas & hypoventilation seen	Sleep study advised if: profound weakness with non ambulation, weak cry, ineffective cough, swallowing difficulties, repeated chest infections, poor lung function, scoliosis & chest wall deformity Episodic apnoea of infancy and childhood (sometimes life-threatening) described in CHAT and RAPSN mutations. Progressive respiratory muscle weakness in COLQ and DOK7 mutations. Sleep Study recommended if these mutations present and symptoms suggestive of SDB. Required if weakness is severe and persistent or if symptoms suggestive of SDB

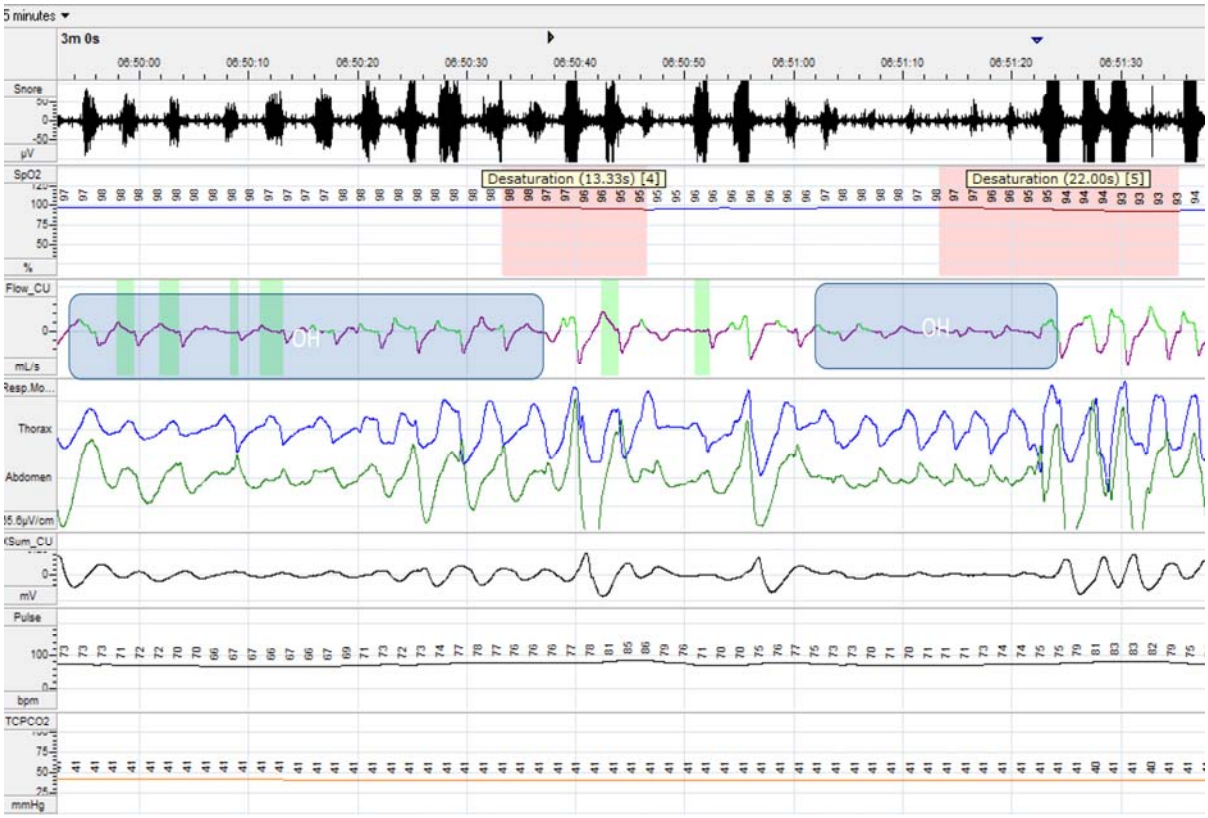


Record of sleep and naps in 24 hours; A record of 2 weeks is useful; Time at which child goes to bed is marked as ↓; Time at which child gets out of bed as ↑; Time period during which child is asleep is shaded; W signifies child is awake, either by parents or alarm or S if awakened by self.

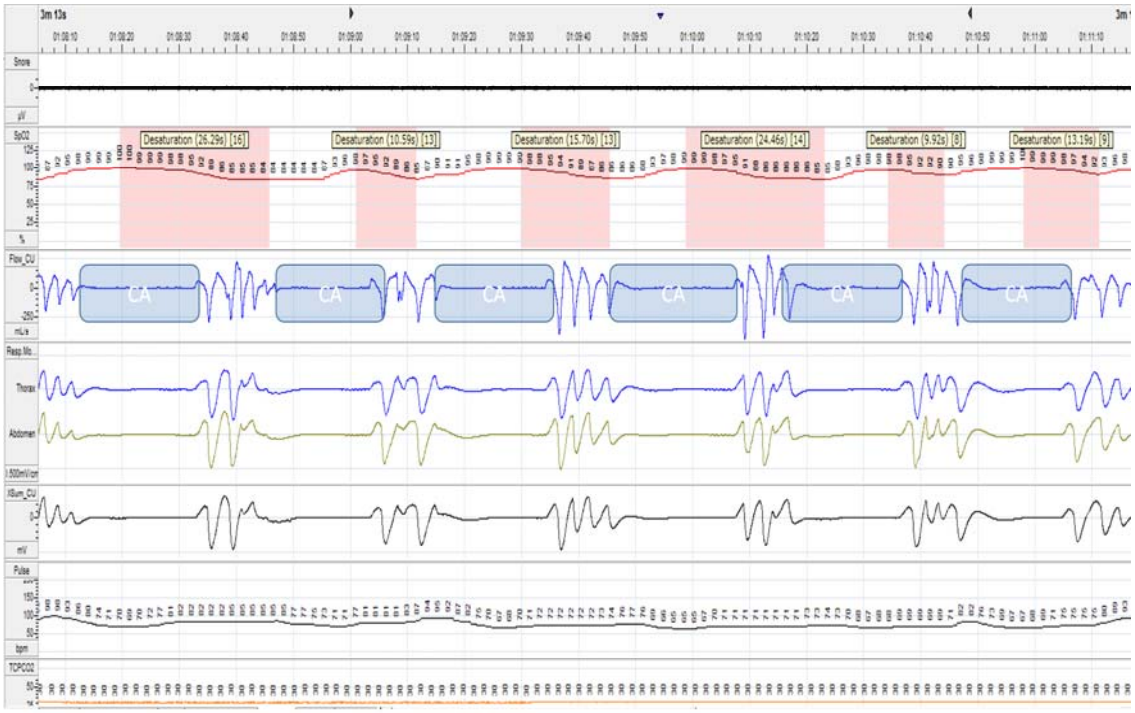
Web Fig. 1 Sleep Diary



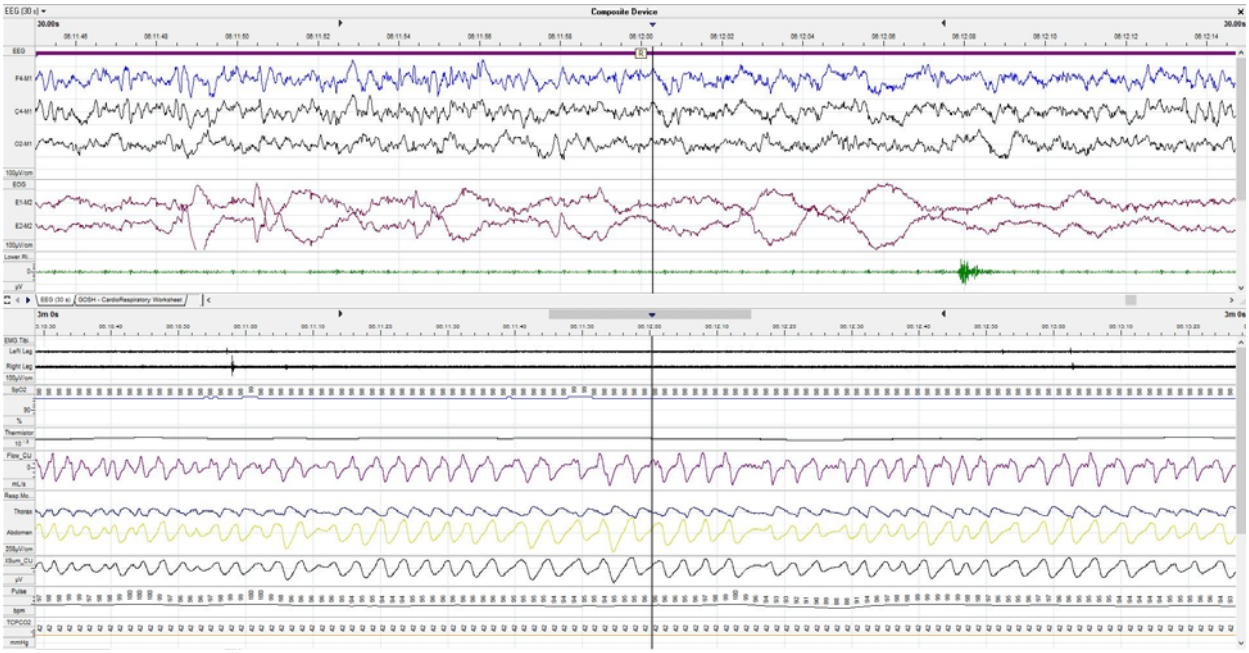
Web Fig. 2 A 3 minute epoch of a Cardiorespiratory Sleep study showing multiple Obstructive Apneas (scored on the nasal flow trace). Note the accompanying desaturations and paradoxical breathing in the thoraco-abdominal bands.



Web Fig. 3 A 3 minute epoch of a Cardiorespiratory Sleep study showing obstructive Hypopnea. Note that the flow is reduced >30% associated with desaturation and the thoraco-abdominal bands show paradoxical efforts.



Web Fig. 4 A 3 minute epoch of a Cardiorespiratory Sleep study showing multiple Central Apneas. Note that the thoraco-abdominal bands show no efforts compared to paradoxical efforts in obstructive apneas.



Web Fig. 5 A 30 s epoch of a Polysomnography. Note the additional neurological channels (EEG, EOG and chin EMG)