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### Evaluation of Sleep Problems in Children

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#### Introduction

The prevalence of sleep disturbances in typically developing children ranges from 9% to 50% [1]. The range of sleep disturbances in children with neurodevelopmental disorders (NDDs), such as autism, may be as high as 75-80% [2]. Neurodevelopmental disabilities are defined as a collection of a large number of neurologic disorders that start in childhood and have different etiologies [2]. The Diagnostic and Statistical Manual of Mental Disorders V (DSM-V) categorizes these disorders as intellectual disabilities, communication disorders, autism spectrum disorders, attention-deficit/ hyperactivity disorder, learning disorders, motor disorders, and other neurodevelopmental disorders [3]. This wide range of disorders includes intellectual disability, epilepsy, cerebral palsy, Rett syndrome, and Down syndrome, to name a few [2]. The various types of sleep disorders are not unique to this population. However, sleep problems in these children can be more severe, chronic, and difficult to treat and more likely to relapse [4]. More than one sleep disturbance in a given child is common. Impaired sleep in general often has an impact on their physical health and predisposes children to mood, behavioral, and cognitive impairments; these consequences may be more pronounced in children with underlying neurodevelopmental disorders [2, 4]. In addition, the burden of sleep problems in caregivers of children with NDDs is often substantial and may add significantly to family stress [5].

The etiology of sleep disorders<sup>1</sup> in these children is often the consequence of underlying disease-related factors, which include the extent and location of brain abnormalities; severity of developmental delay; communication challenges; associated sensory loss such as visual impairments, concomitant health problems, and limited mobility; and pain. In addition, medications such as anticonvulsants and antidepressants prescribed for medical or psychological conditions can contribute to sleep problems in these children; for example, antidepressants can exacerbate restless legs syndrome symptoms and may contribute to sleep onset and maintenance insomnia [6, 7]. Children with neurodevelopmental disabilities often have comorbid mental health impairments such as mood or anxiety disorders, which can be exacerbated by sleep disorders and vice versa. Similarly, strong associations have been found between disruptive behaviors such as hyperactivity, aggression, and oppositionality and sleep disruption [4].

Biological factors can play a role in sleep disorders in children with NDDs. An example of this is the high prevalence of sleep disturbances in children with autism spectrum disorder. Circadian rhythm abnormalities have been postulated, and a number of studies have shown that a large portion of the studied children with autism spectrum disorder (ASD) had low plasma melatonin concentration. Melatonin is a hormone secreted by the pineal gland in response to decreased light, mediated through the suprachiasmatic nucleus, thus regulating sleep and wake cycles. Supplementing these children with exogenous melatonin has been shown to be beneficial in regulating sleep [7, 8]. Finally, environmental factors and issues related to caregivers also play important roles in these sleep disturbances. Environmental factors can include the child's sleeping space,



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<sup>©</sup> Springer Nature Switzerland AG 2019

J. A. Accardo (ed.), Sleep in Children with Neurodevelopmental Disabilities, https://doi.org/10.1007/978-3-319-98414-8\_2

<sup>&</sup>lt;sup>1</sup>While this term is used interchangeably for the purposes of this review with sleep disturbances, problems, issues, etc., it should be noted that sleep disorders are typically defined by specific diagnostic criteria as found in the American Academy of Sleep Medicine International Classification of Sleep Disorders (ICSD), 3rd edition, or the *Diagnostic and Statistical Manual (DSM) V*.

noise, room, bed sharing, family composition, as well as lifestyle issues such as parental work status, household rules, and socioeconomic status. Caregiver variables include discipline styles, parental mental health issues, family stress, and parental education level [2, 5].

#### Conceptual Framework of Sleep Disturbances in Children

The majority of sleep problems in children can be categorized by one or more of the following disturbances of sleep. These include insufficient sleep (inadequate sleep quantity), fragmented sleep (poor sleep quality), circadian rhythm disorders (disrupted regulation of sleep timing), and primary sleep disorders of excessive daytime sleepiness (EDS). Insufficient sleep results from a mismatch between the child's sleep needs and the amount of sleep actually obtained; however, this calculation is often based on recommended sleep duration ranges for typically developing children in different age ranges, which may not necessarily always be applicable to children with neurodevelopmental disabilities. Insufficient sleep can be the result of suboptimal time in bed (opportunity to sleep) but usually stems from difficulty initiating and/or maintaining sleep. Sleep fragmentation impacting sleep quality usually results from frequent and repetitive nighttime arousals, such as those associated with obstructive sleep apnea. Circadian rhythm disorders are characterized by inappropriately timed sleep periods that often interfere with daytime functioning (e.g., a child sleeps during the day and is awake all night), but the sleep quality and quantity is typically normal if the child is allowed to sleep on his or her preferred schedule. Primary disorders of EDS such as narcolepsy and idiopathic hypersomnia are relatively rare in children in general but may be more common in children with neurodevelopmental disorders such as Prader-Willi syndrome and Niemann-Pick disease [4].

#### **Screening for Sleep Problems**

While a broad range of sleep disorders including obstructive sleep apnea (OSA) and restless leg syndrome (RLS) are discussed in this chapter, the focus will be on the most common presenting sleep complaints in children with neurodevelopmental disorders including insufficient sleep, irregular sleeping patterns, bedtime resistance, delayed sleep onset, frequent and/or prolonged night wakings usually requiring caregiver intervention, and early morning wakings [4]. It should be noted that while often related, bedtime resistance and delayed sleep onset are not necessarily equivalent; i.e., a child may exhibit a prolonged struggle at bedtime but then fall asleep immediately after lights out or may go to bed willingly but remain awake for hours. Due to the high prevalence of sleep disturbances in these children, it is essential that clinicians both have a systematic approach to screen them at regular intervals for sleep problems and maintain a high index of suspicion.

The BEARS sleep screening tool is a brief five-item instrument that is easy for the provider to remember and user-friendly for families. The parent (and child, if developmentally appropriate) should be asked if the child experiences possible problems in each of the five items below based on the BEARS acronym.

- B Bedtime issues. This should include difficulty falling asleep and bedtime resistance.
- E Excessive daytime sleepiness. Specifically issues related to morning waking, napping during the day, and involuntary daytime sleepiness.
- A Night awakenings. Particularly if parental intervention is required.
- R Regularity and duration of sleep. This includes inquiring as to the specifics of bedtime and wake time and variability night-to-night and variability on weekend/weekdays.
- S Snoring. Questioning about nocturnal behaviors such as snoring, sleep disordered breathing sleepwalking/sleep terrors/confusional arousals, enuresis, bruxism and nocturnal seizure activity [9].

#### Components of the Pediatric Sleep Evaluation

The assessment should include a very detailed sleep history, medical history, developmental/school history, family history, psychosocial history, behavioral assessment, and physical examination [4].

#### **Sleep History**

A thorough sleep history is essential when assessing a child for a sleep disturbance and should include details of the presenting complaint(s), sleep-wake schedules, bedtime, nighttime behaviors, and daytime behaviors. It is helpful to obtain information about the presenting complaint from the caregiver, the child when possible, and if applicable/possible the individual who shares a room/bed with the child. The sleep complaint should include a detailed account of the onset, duration, severity, and night-to-night variability. Any events that preceded and possibly triggered the sleep problem should be elicited (e.g., new sibling, move, illness or death in family, changes in medication). It is also helpful to understand why the caregivers are requesting help at this particular time (e.g., school year starting, expecting new baby). It is important to inquire as to what methods the family has tried and to describe in detail (e.g., tried a "cry it out" approach for 2 days or inconsistent response to night wakings by each parent) and how they trialed those methods. Lastly, parental expectations regarding treatment goals must be discussed and understood to guide treatment recommendations [7].

It is important to understand the sleep schedules and sleep habits for any child presenting with a sleep problem. This can help the provider better understand the sleep problem in order to formulate etiologic hypotheses and thus generate potential interventions and to identify other coexisting sleep issues [7]. While clearly overlapping, for the sake of completeness and clarity, we have divided these various sleep components into (1) sleep schedule (bedtime and wake time, night-to-night and weekday/weekend variability), (2) evening behaviors, (3) bedtime routine, (4) the process of falling asleep (how and where the child falls asleep, including problematic sleep onset associations), (5) night wakings (including required parental interventions), (6) other episodic nocturnal behaviors (e.g., snoring and respiratory symptoms (choking, gasping, mouth breathing, etc.), sleepwalking/ sleep terrors/confusional arousals, enuresis, bruxism, nocturnal seizure activity), (7) sleep environment (noise, temperature, light), and (8) morning waking and daytime sleepiness, including scheduled napping and involuntary daytime sleepiness.

#### **Sleep Schedules**

The caregiver should be asked if the child has a set bedtime. Many children are allowed to fall asleep when they choose. It is important to evaluate the timing of bedtime for age and developmental level of appropriateness. The variability of bedtime should be elicited by asking for night-to-night variability as well as weekday, weekend, and vacation discrepancies. Some children will have a strict bedtime on weekdays but are allowed to stay up late and wake up late, thus contributing to difficulty falling asleep on their regular schedule. Since children with NDDs are more vulnerable to sleep and circadian rhythm disruptions, it is particularly important that they have regular and appropriate bedtimes and wake times [2]. It is also important to understand what parental involvement consists of at bedtime. For example, some children will only go to bed with one parent, suggesting possible limitsetting issues. Other parents go to bed before their child falls asleep, which can often be the case with teenagers, and thus are unaware of the child's activities after their own bedtime.

## Evening Behaviors (Including Bedtime Resistance)

It is helpful to ask the caregiver to describe activities from dinnertime until bedtime to best understand evening and bedtime behaviors. For example, engaging in vigorous exercise with several hours of bedtime may not only be energizing but may increase core body temperature, making it more difficult to fall asleep. Similarly, eating a heavy meal just before lights out may prolong sleep onset. In particular, children with NDDs can become easily over stimulated, as they may have difficulty processing large amounts of information resulting in an overload state [2]. Overstimulation may occur when children with NDDs are exposed to new or unexpected events, anxiety, excessive noise, cold or heat, vigorous exercise, hunger, large meals, pain, seizures, and certain drugs [2]. Other factors that can affect bedtime should be questioned, such as evening activities of the child/parent/family and family schedules. Some families schedule familycentered activities later in the evening, potentially further delaying bedtime to an inappropriate hour.

The provider should thoroughly review with the caregiver all refusal behaviors of the child at bedtime. The details can be obtained by asking about the characteristics of the refusals (coming out of room/bed, crying), the intensity of the behavior (calling out to parent or crying and screaming), how often the behavior occurs (daily or once a week), and how long the behaviors last at bedtime. A critical factor is whether the child behaves differently with various caregivers or family members, and the caregivers' response to the behavior, because a key element to reducing these behaviors is consistency, not only on the part of the caregiver with each interaction but also among all of the family members who are involved with bedtime [7].

Multiple studies have noted delayed sleep onset particularly in children with Asperger syndrome (AS). One hypothesis is that the anxiety that is often found in children with AS can contribute to difficulty falling asleep at bedtime. The study by Paavonen et al. [10] noted a high prevalence of sleep fears and negative attitudes toward sleeping in children with AS. Some other stimulating examples could include playing with siblings, watching a TV show, or participating in a physical activity close to bedtime. Certain textures can be difficult for children with autism spectrum disorder (ASD) to tolerate (such as pajamas or blankets), contributing to difficulty settling at bedtime; a weighted blanket may be beneficial [1].

The child's level of sleepiness at bedtime should be evaluated, sleep location (bed, couch) and variability of location as well as the transfer of a child from one bed to another once the child falls asleep [7]. Children with delayed sleep phase disorder, often seen in adolescence due to circadian preference, are not sleepy until later in the evening. Delayed sleep phase disorder is a circadian rhythm disorder, which involves a significant, persistent, and intractable phase shift in sleepwake schedule (later sleep onset and wake time) that interferes with environmental demands. This can often result in significant daytime sleepiness and academic and behavior problems if the child cannot adhere to their preferred sleep schedule [7].

#### **Bedtime Routine**

Caregivers should be questioned about the presence of a regular bedtime routine and what activities are included. Electronic media use such as television viewing, playing video games, or even using an e-reader as part of the bedtime routine should be elicited (especially the presence of electronic media in the bedroom) as this has been clearly shown to be associated with sleep problems such as difficulty initiating sleep and shorter sleep times in both typically developing children and those with NDDs [7]. In a study published by Pediatrics in 2013, for example, results indicated that having bedroom media decreased total sleep time at night. Access to a computer or television in the bedroom showed a stronger association to less nighttime sleep in boys with ASD as compared to boys with ADHD and typically developing children. This study also demonstrated the amount of time playing video games was associated with less nighttime sleep in boys with ASD [11]. This study confirms the importance of evaluating for media use in the bedroom within a sleep evaluation. Some households have televisions in each bedroom and are not aware of the consequences of bright light on their child's sleep. Exposure to light, including light-emitting devices (television, tablets, cellular phones), suppresses the release of the sleepfacilitating hormone melatonin. In turn this shifts the circadian clock to a later time, which contributes to difficulty falling asleep at bedtime [12, 13]. The provider may have to be creative and compromise with the family to allow for realistic expectations in such households.

An extended bedtime routine may point to limit-setting difficulties, e.g., a child demanding to be read five or more books at bedtime. However, the lack of a bedtime routine does not allow the child to transition as easily from wake to bedtime. Sleep routines in children with ASD may involve stereotypic and repetitive behaviors, and these children can have difficulty adapting to changes in their routines [4].

Some characteristics of children with ASD can contribute to delayed sleep onset and include difficulty with selfregulation, hypersensitivity to environmental stimuli, and repetitive thoughts or behaviors that interfere with settling [1]. For example, some children with ASD may find pacing calming at bedtime. However, this activity can be stimulating in itself and can contribute to difficulty with sleep onset. Calming activities can include a well-structured routine of quiet baths, listening to stories, prayers, or small snacks. The sleeping environment should be comfortable and cool with minimal light and sound [1, 7].

#### **Falling Asleep**

The provider should ask when lights are turned out and what time the child actually falls asleep. A child who has difficulty

falling asleep should be assessed for possible causes such as restless leg syndrome (RLS), anxiety, or delayed sleep phase syndrome. Children with ASD commonly exhibit symptoms of sleep onset insomnia [1]. Children with ASD are also susceptible to RLS, and some medications used by children with ASD (risperidone, selective serotonin reuptake inhibitors) can exacerbate RLS. Specifically the child with RLS would describe the urge to move the legs and leg discomfort that may include verbal descriptors such as "ants crawling on my legs" or terms such as squeezing, tingling, wiggling, itching, or "funny" feeling. Nonverbal children may be observed to move, shake, or rub their legs. These symptoms usually occur exclusively or are worse at rest such as in a lying or sitting position and in the evening. This uncomfortable leg sensation is accompanied by the urge to move the legs. The leg sensation diminishes with movement but returns when movement is stopped [7].

Sleep onset associations are behaviors that are required by the child in order to fall asleep at bedtime and return to sleep after night wakings. Examples of sleep associations, which are needed to help the child fall asleep, can include rocking, nursing, cuddling, or parental presence. This can be problematic as these can cause prolonged bedtimes and night wakings as well as disrupting other family members' sleep. However, some children with profound brain damage may require sleep-promoting cues such as gentle rhythmic movements, light massaging, or tightly tucked bedsheets [2, 7].

#### **Night Wakings**

A thorough evaluation of nighttime activity should be discussed. Night wakings should be evaluated by discussing when the night wakings started occurring, the frequency per week and per night, and the timing and duration. Possible triggers (getting up to void, external noise) should be reviewed as well as the child's behavior upon awakening (calling out, leaving room) and the parental response (ignoring, responding inconsistently). It should be noted whether the child needs help returning to sleep (rocking to sleep, television, bottle) [7]. Two studies, one by Schreck et al. [14] and one by Mayes and Calhoun [15], suggest that more severe autism symptoms predict more frequent night wakings and sleep disturbances [6].

#### **Episodic Nocturnal Behaviors**

The evaluation should include questioning whether the child experiences partial arousal parasomnias, including sleepwalking, sleep terrors, and confusional arousals. Sleepwalking commonly occurs at the beginning of the night during slow-wave sleep and is not remembered by the child. The child may appear dazed and occasionally agitated and may perform bizarre actions. Sleep terrors are a sudden arousal from slow-wave sleep and include autonomic and behavioral manifestations of intense fear. Confusional arousals are nocturnal episodes characterized by confusion, disorientation, grogginess, and at times significant agitation upon awakening from slow-wave sleep or following forced awakenings [7].

Other nocturnal events are sleep-related rhythmic movements (SRRMs) which include head banging, body rocking, and head rolling which are common among young children and often function as self-soothing behaviors. These episodes usually occur at sleep onset following normal nighttime arousals and can occur during night wakings. While most typically developing children outgrow SRRMs by elementary school, it is not uncommon in children with NDDs to have these behaviors persist, and they may require more aggressive management (including pharmacologic interventions in some cases) than "tincture of time." Bruxism is the repetitive grinding or clenching of teeth during sleep. Sleep enuresis involves involuntary voiding during sleep, at least twice a week beyond the age of 5 years [7]. Night screaming and laughing are particularly common in younger children with Rett syndrome, whereas night wakings are found among all age groups in girls with Rett syndrome [16]. Some studies have reported an increase in sleepwalking, bruxism, and other partial arousal parasomnias in children with ASD. There is also an increase in REM behavior disorder in children with ASD; this behavior is characterized as acting out their dreams due to the absence of muscle atonia normally present during REM sleep [4].

Nocturnal symptoms of sleep-disordered breathing include snoring, snorting, pauses in breathing, mouth breathing, restless sleep, sweating during sleep, and abnormal sleep positioning (i.e., neck extension or requiring multiple pillows). Daytime symptoms can include excessive daytime sleepiness, irritability, emotional dysregulation, internalizing behaviors, aggression, hyperactivity, inattention, and learning problems and academic problems. Some additional features that can be associated with sleep-disordered breathing include enuresis, growth failure, increase in partial arousal parasomnias, increase in seizure activity in predisposed children, and other comorbid sleep problems such as periodic limb movement disorder or RLS [7].

#### **Sleep Environment**

Bedroom space and location should be ascertained. This includes bed sharing or room sharing and with whom. The questions should include with whom the child co-sleeps, how long at night, when at night, how often per week, and reason for co-sleeping (i.e., "lifestyle" or "family bed" by

choice versus "reactive co-sleeping" in which the parent brings the child into bed in an attempt to solve a sleep problem such as prolonged night wakings) [7]. If the child sleeps in a separate room from the parent, the distance between their rooms should be determined. Some parents may place an alarm on the child's door if they are prone to sleepwalking or exiting their bedroom in the middle of the night. If the child sleeps in multiple sleep locations, such as with divorced parents or grandparents, the bedtime routine and sleeping arrangements at these alternate locations should be discussed. The caregiver or child should describe light exposure at bedtime, which includes a main light, small lamp, or night-light on, as well as exposure to outside light, including streetlights and use of blackout shades. Some children may be afraid of the dark and may require a small light source such as a night-light. Noise exposure from outside or from other family members as well as the time of day of the noise should be ascertained by the provider. Some children will benefit from a "white noise" machine if they are sensitive to noise. The ability to adjust room temperature and comfort in the room is important to review as well. The provider should ask what type of bed the child is using, crib versus bed, and what type of bedding [7]. Children with gastroesophageal reflux may benefit from an elevated head of bed. Immobile patients such as children with cerebral palsy may require frequent repositioning due to risk of bedsores. Some infants feel comforted by swaddling, whereas other children with ASD may prefer a weighted blanket [2].

#### Morning Waking and Daytime Sleepiness

The time of awakening for the day should be evaluated on weekdays and weekends and during vacation periods. In particular, the failure to awaken spontaneously at the desired time and the need for alarm clocks and especially repeated reminders and prompts from caregivers to get up suggest that a child is not meeting his or her sleep needs with the current schedule. Sleeping longer on the weekends also suggests that a child may be getting insufficient sleep during the week. Alternatively, children with a circadian phase delay will also tend to awaken later in the morning because their sleep-wake schedule is shifted later; they may revert to their preferred sleep schedule of going to bed and waking up later on weekends and on vacation/school break. Furthermore, if a child awakens sleepy and groggy despite apparently sufficient sleep, this may indicate a sleep disruptor such as obstructive sleep apnea or periodic limb movement disorder. Finally, some children and adolescents display extreme difficulty in waking on school days that appears out of proportion to their sleep schedule but rise without difficulty on nonschool days, suggesting that school avoidance may be a potential confounding factor [7].

The timing, frequency, location, and duration of both planned/scheduled and developmentally appropriate naps and involuntary sleep periods (i.e., "dozing off") should be assessed. For example, a planned nap in a preschool child in the late afternoon may interfere with falling asleep at bed-time. Since the majority of children over 5 years have stopped napping, planned or unplanned naps in an elementary-aged child suggest insufficient sleep or the presence of sleep disruptors [7]. Nocturnal events such as partial arousal parasomnias, nightmares, symptoms of sleep-disordered breathing, and symptoms of periodic limb movement disorder should be elicited as these sleep disruptors can contribute to daytime sleepiness [7].

Caregiver reports of a child falling asleep riding in a car and during passive activities or especially during stimulating activities such as being in school or during a conversation could be due to inadequate sleep, a sleep disruptor, or primary hypersomnia disorder such as narcolepsy. Younger children often exhibit behavioral manifestations of sleepiness such as irritability, aggressiveness, hyperactivity, or impulsivity rather than "classic" symptoms such as yawning or dozing off [7]. It is important to explain to parents that a mismatch between sleep needs and sleep obtained can be a major contributor to daytime sleepiness. Children with a sleep disruptor such as OSA or periodic limb movement disorder (PLMD) may display symptoms of overtiredness or sleepiness depending on the severity and their age. Once their sleep disruptor is diagnosed and adequately treated, their sleepiness should be reevaluated.

As noted above, some children may have daytime sleepiness due to a primary hypersomnia disorder such as narcolepsy. These symptoms include falling asleep during the day during passive or more importantly active activities despite adequate sleep at night or other sleep disruptors. The child may describe an intense sudden need to sleep or "sleep attack." The child with narcolepsy may also experience the intrusion of REM sleep, phenomena such as muscle atonia or dream mentation into wakefulness. The primary example is cataplexy, which is the abrupt, bilateral, partial, or complete loss of muscle tone in the face of retained consciousness, classically triggered by positive emotion. Cataplectic episodes last seconds to minutes, and the child recovers completely afterward without any residual effects. Other REM-related symptoms include hypnagogic (falling asleep) or hypnopompic (waking up) hallucinations. These hallucinations can be auditory, visual, or tactile and can be very frightening. Another example of REM intrusion into wakefulness is sleep paralysis, which is the inability to move or speak for a few seconds also when falling asleep or waking up. While it should be noted that hypnic hallucinations and sleep paralysis could occur in otherwise normal individuals who are sleep deprived, cataplexy is unique to patients with narcolepsy. Furthermore, children with narcolepsy often have difficulty sleeping at night with frequent and lengthy

night wakings. The definitive diagnostic tests for narcolepsy include an overnight polysomnogram in conjunction with a multiple sleep latency test the following day and/or a CSF measure of hypocretin levels; HLA testing for DQB1\*06:02 may be helpful but lacks specificity as a substantial percentage (25–35%) of controls are positive [4].

It is just as important to elicit daytime behaviors while evaluating for sleep issues. Insufficient sleep can have negative consequences on many functional domains including mood, behavior, attention, learning, school performance, school relationships, and overall health [7]. The parent should provide information pertaining to the child's mood such as grumpiness in the morning or at bedtime. The provider should ask about the child's behaviors, as to whether they demonstrate hyperactivity, impulsivity, inattention, or disrespect to parents or others. It is also important to ask about the child's school experience and any learning difficulties. If the child has been diagnosed with a learning disability, ask if an evaluation has been performed to provide the services the child needs. It is vital to understand school performance when diagnosing sleep issues. If a child is not getting adequate sleep, often their grades suffer, although some children are able to continue to perform well in school despite their tiredness. The child should be able to discuss how they relate to their peers. It is important to obtain information such as negative relationships at school that can be classified as bullying. Some children try to avoid school due to bullying, difficulty making friendships, or disliking school. This avoidance may be exhibited by a child who goes to bed quite late at night, is unable to wake up in the morning for school, or, if they do attend school, sleeps through their classes. The provider must evaluate how interested the child truly is in fixing a sleep pattern to return to school.

Some children with NDDs have health problems that can cause multiple hospitalizations or may require extended time away from school. It is important to understand how this has impacted the child as well as their education.

Of note, fatigue is typically defined as lethargy (i.e., without sleepiness) and is often described as subjective feelings of low energy and low motivation without an increased sleep propensity. Fatigue tends to be associated with medical or psychiatric disorders. It is important to investigate the child's medical and psychiatric history as well as medications that may contribute to fatigue or sleepiness. On the other hand, sleepiness is described as the tendency to fall asleep usually in inappropriate settings. However, this can be difficult to distinguish when first interviewing a child and family members [7].

#### **Medical History**

A medical history and review of systems should be elicited with an emphasis on past and current medical conditions, prior hospitalizations, surgeries, and medications [7]. The infant should be screened for conditions such as gastroesophageal reflux, colic, and milk allergies as these can be associated with sleep problems. Likewise the child should be screened for asthma, upper respiratory infections, and headaches which may contribute to sleep-disordered breathing [6]. The child should be screened for past history of concussion particularly if daytime sleepiness is a complaint. Surgical procedures should be recorded, particularly adenoidectomy or tonsillectomy in the child with symptoms of sleep-disordered breathing. A comprehensive medication list should be obtained as certain medications can worsen RLS/PLMD and contribute to daytime sleepiness or insomnia.

Risk factors for sleep-disordered breathing include asthma, allergies, gastroesophageal reflux, adenotonsillar hypertrophy, obesity, prematurity, craniofacial abnormalities, nasal septal deviation, neuromuscular disease, hypothyroidism, hypotonia, and family history of sleep-disordered breathing [7]. A number of congenital syndromes are also associated with an increased risk of sleepdisordered breathing. For example, children with Down syndrome have multiple risk factors for OSA including generalized hypotonia, midface and maxillary hypoplasia, glossoptosis, hypothyroidism, adenotonsillar hypertrophy, obesity with central adiposity, and gastroesophageal reflux [4, 17]. The AAP recommends that children with Down syndrome should be screened for sleep-disordered breathing at least once in their first 6 months of life and then yearly thereafter with a recommended overnight polysomnogram before their fourth birthday [18]. Children with Prader-Willi syndrome are also at high risk for sleep-disordered breathing including obstructive sleep apnea, hypoventilation, and central sleep apnea due to obesity, hypotonia, and adenotonsillar hypertrophy [4, 19]. A study in 2011 by Hagebeuk et al. noted that 50% of a small cohort of girls with Rett syndrome showed obstructive sleep apnea and more frequently displayed central sleep apnea on overnight polysomnogram [20]. Thus it is important to screen these girls for sleep-disordered breathing and perform an overnight polysomnogram if warranted. Children with cerebral palsy have a higher risk for sleep-disordered breathing not only due to adenotonsillar hypertrophy but also due to disproportionate midface anatomy, mandibular alterations, abnormality of upper airway tone, abnormal central control of respiration, obesity, and medications that increase the collapsibility of upper airway musculature and reduce the patency of the airway [21].

Symptoms of PLMD include non-restorative sleep, clinically significant sleep disruption, and/or daytime sequelae [7]. Children with ASD may be at higher risk for PLMD possibly due to limited dietary choices which can contribute to limited iron in their diet; low ferritin is a risk factor for PLMD [4]. Parents of children with Williams syndrome have noted increased leg movements when sleeping; these children may have a higher prevalence of PLMD [22].

#### **Developmental/School History**

A developmental history should include prematurity and/or birth complications, developmental delays, and impaired neurological functioning. A developmental history is necessary to provide the most developmentally appropriate sleep advice. Academic problems can occur with inadequate sleep [7]. Therefore a school history including grade, academic performance, extracurricular activities, and behavior with students and teachers is important to elicit. At times, a concern for daytime sleepiness stems from a teacher reporting a child falling asleep frequently in class.

#### **Family History**

It is helpful to obtain a family history as some sleep disorders have a genetic component including partial arousal parasomnias, restless leg syndrome, obstructive sleep apnea, and narcolepsy. Even if a family member has not been diagnosed, it is helpful to discuss whether family members have symptoms of a sleep disorder such as snoring [7].

#### **Psychosocial History**

A complete psychosocial history should be elicited and should include overall family functioning, effectiveness of parenting skills, family structure, parental psychological functioning, and family discord. If the parents are divorced, the provider should elicit how much time has lapsed since the divorce, visitation schedules, level of parental involvement, consistency of sleep patterns and habits, discrepancies in parenting styles, and level of stress on the child. The provider should elicit significant life events such as a death of a family member, change in school, or a recent move. Cultural influences on sleep behaviors should be evaluated as well as family values regarding health priorities and family beliefs about sleep and importance of sleep. The effect of the child's sleep problem on the family should be understood, such as the effect on the parent's sleep, impact on parent's daytime functioning, and impact on marital and family satisfaction [7].

#### **Behavioral Assessment**

A behavioral assessment is important as sleep disturbances can result in psychiatric symptoms such as mood changes and oppositional behaviors. Conversely psychiatric disorders can result in sleep disturbances. It is important to evaluate for symptoms of depression, anxiety, and other psychiatric disorders [7]. It is helpful to understand the temperament of the child, as children who have higher levels of physiological arousal and feel their lives are more stressful may have increased rates of insomnia due to increased vigilance, anxiety, and hyper-responsiveness to threatening environmental stimuli [6].

#### **Physical Examination**

A physical exam should be performed on all children presenting with sleep concerns. The exam, while comprehensive, should be targeted toward the presenting complaints and potential etiologies of the sleep problems, for example, focusing on the otolaryngologic exam in children who have concerns for snoring or other symptoms of sleep-disordered breathing, including signs of atopy such as allergic shiners or mouth breathing. A neurologic examination should be done if there is concern for daytime sleepiness or seizure activity. The child's general appearance should be taken into account. Growth parameters should be evaluated as both growth failure and BMI/overweight/obesity can be associated with sleep-disordered breathing. Additionally, as circadian delayed sleep phase disorder often develops at the onset of puberty, the child's pubertal status should be evaluated as well [7].

#### **Diagnostic Tools**

A sleep diary is a helpful tool when evaluating many sleep problems, particularly if the child or parent is uncertain regarding the amount of sleep a child is getting and the variability in the sleep-wake schedule. A daily sleep diary collects information including bedtime, latency to sleep onset, number and duration of nighttime wakings, time of morning waking, total sleep time, sleep efficiency, and duration and timing of naps, over a 1–2-week period, and can be filled out by the parent or by the older child/adolescent [7]. Documenting sleep-wake patterns on a sleep diary can provide helpful feedback for the child and parent, given that there may be a discrepancy between self-report or parent-report of "typical" sleep schedules and the actual day-to-day reporting.

An overnight polysomnogram (PSG) is indicated to diagnose and delineate sleep-disordered breathing, determine optimal treatment options for sleep-disordered breathing, document periodic limb movement disorder, and investigate potential etiologies for sleep-related daytime sleepiness as well as to delineate etiology [23–25]. Some children with NDDs may have difficulty tolerating the PSG due to the multiple leads that are applied and must stay on throughout the night. Some children benefit from a desensitization period of being exposed to the equipment used prior to coming in for the PSG. All children should be tested in an AASMaccredited pediatric sleep lab that has the expertise and experience to study children who may pose significant behavioral challenges. This is necessary to ensure the PSG is scored according to pediatric criteria but also to ensure this population of children are tested in a laboratory that is familiar with children and those with NDDs and meet their needs.

The multiple sleep latency test (MSLT) provides an objective quantification of daytime sleepiness as well as REM onset sleep periods, which is highly characteristic of children with narcolepsy. This test consists of five 20-min nap opportunities every 2 h. The data that are collected include the number of naps during which the child slept, the length of sleep time during each nap, and whether the child had a period of REM sleep during each 20-min nap opportunity. It is essential to perform a PSG the night before the MSLT to insure that the patient had sufficient sleep the night before and to determine whether there is another underlying explanation for daytime sleepiness such as obstructive sleep apnea. Medications that affect the central nervous system could skew the results of the MSLT as some suppress REM sleep (e.g., selective serotonin reuptake inhibitors). If possible, the child should be weaned off such medications prior to the test, with adequate time to avoid REM sleep rebound [7].

Actigraphy utilizes a wristwatch-like device called an actigraph that records and stores data regarding body movements over a period of time (usually 2 weeks) that is downloaded and then transformed using a software program into an approximation of sleep-wake patterns. In conjunction with a sleep diary, the data will show the approximate sleep duration, sleep latency, and the length of time during the night that the child is asleep and is awake. Actigraphy is particularly helpful in quantifying sleep amounts more objectively when subjective sleep duration seems implausible (e.g., 4 h of sleep for a 5-year-old) or determining whether a child has a circadian rhythm disorder [7].

#### Summary

Sleep problems, ranging from insomnia to circadian rhythm disruption to sleep-disordered breathing, are extremely common in children with NDDs. Therefore, the clinician needs to maintain a high index of suspicion and institute a systematic screening and evaluation process in clinical practice settings. While often more challenging to manage, successful identification and treatment of sleep problems in these children often improve the health and quality of life of the entire family.

#### 2 Evaluation of Sleep Problems in Children

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