

NARCOLEPSY AND CHILDREN

For children and adolescents, narcolepsy is a chronic, under-recognized condition that can have debilitating symptoms.

Awareness and recognition of narcolepsy as a pediatric condition is low among healthcare professionals. A survey of healthcare professionals revealed that the majority did not associate narcolepsy with children or adolescents. Yet, more than half of patients report experiencing symptoms before the age of 18.

Healthcare professionals may attribute narcolepsy symptoms, like excessive daytime sleepiness, to another pediatric disorder, such as attention-deficit/hyperactivity disorder (ADHD), leading to misdiagnosis. In addition, lack of awareness and under-recognition of narcolepsy and its symptoms among patients and caregivers may contribute to the delay in diagnosis.

UNDERSTAND

The potential psychosocial burden of narcolepsy in pediatric patients and the impact it has on their caregivers.

Narcolepsy Can Start in Childhood

Although narcolepsy is often thought of as a disease of adulthood, symptom onset most commonly occurs in childhood and adolescence.¹⁻⁴ More than half of patients report experiencing symptoms before 18 years of age. Studies indicate onset of symptoms occurs most commonly between 10 and 25 years of age, and recent data support onset occurring as early as age 1 to 5, with initial peak at approximately age 15.

Narcolepsy Is Under-recognized in the Pediatric Population

The prevalence of narcolepsy in pediatric patients is uncertain; however, the condition is probably underestimated. Patients and their caregivers typically do not seek help for narcolepsy symptoms for many years, and when patients do seek help, their symptoms may be misdiagnosed as a more common medical condition, further contributing to delayed diagnosis.

Patients may suffer for more than a decade before receiving an accurate diagnosis. A mean delay in diagnosis of narcolepsy of 15 years was demonstrated in one study. In another study, pediatric onset of symptoms (17 years of age or younger) was a strong predictor of delayed diagnosis.

Recognize Narcolepsy in Pediatric Patients

Symptoms of narcolepsy may present differently in pediatric patients. The 5 main symptoms of narcolepsy are referred to by the acronym CHES (Cataplexy, Hallucinations, Excessive daytime sleepiness, Sleep paralysis, Sleep disruption). All patients with narcolepsy experience excessive daytime sleepiness; however, not all narcolepsy patients experience all of the other symptoms.

As children become adolescents and adults, the symptoms of narcolepsy may manifest differently and change over time.

- The possibility of an evolving disorder, with cataplexy developing over time, should be considered in all pediatric patients with narcolepsy type 2.
- As a child with narcolepsy type 1 ages, cataplexy develops into the more typical form observed in adults.
- Hypnagogic/hypnopompic hallucinations and sleep paralysis may be age-dependent phenomena that slowly appear and progress along the disease course.

Therefore, ongoing monitoring for changes in symptom presentation and comorbid medical and psychiatric conditions is important when managing patients with narcolepsy.

Cataplexy

Cataplexy may manifest differently in pediatric patients. Cataplexy typically presents at the same time as or within a year from onset of excessive daytime sleepiness and can have a distinct presentation in young children at onset, by manifesting as a complex movement disorder. In some cases, onset of cataplexy may be delayed for years or decades. Therefore, the potential for cataplexy to present later in the disease course should be considered in all pediatric patients with narcolepsy type 2.

ASKING PATIENTS AND CAREGIVERS ABOUT CATAPLEXY

How often does your child experience drooping or loose/weak muscles when he or she is laughing or expressing intense emotions, such as when tickled?

Do you ever notice eyelid drooping, mouth opening, or tongue protruding for no apparent reason?

Recognizing Cataplexy

- Close to disease onset, cataplexy may manifest as a complex movement disorder, which includes:
 - “Negative” (hypotonic) motor features, such as head drop, persistent facial hypotonia, persistent eyelid droop, and tongue protrusion
 - “Active” movement abnormalities or dyskinesia, such as raising eyebrows, lip licking, lip biting, lip chewing, grimaces, and tongue protrusion
- Close to disease onset, children may present with cataplectic facies, a localized version of status cataplecticus, which:
 - Affects facial muscles, often without any clear emotional triggers
 - Manifests as repetitive mouth opening, tongue protrusion, and drooping eyelids
- As children age, cataplexy presentation changes into the classical forms observed in adults.

Hallucinations (Hypnagogic and Hypnopompic)

Hypnagogic (occurring at sleep onset) and hypnopompic (occurring while awakening) hallucinations include primary visual hallucinations, such as seeing shadowy figures, animals, people, and formed shapes in the room. Up to 66% of patients report experiencing hypnagogic/hypnopompic

hallucinations. Therefore, uncovering the presence of these hallucinations can be helpful when looking for narcolepsy.

Recognizing Hallucinations

- For younger children, hallucinations can be scary and, in some cases, may cause them to fear going to bed.
- Young children may be unable to distinguish hallucinations from reality, making obtaining an accurate history of this symptom difficult.³

ASKING PATIENTS AND CAREGIVERS ABOUT HALLUCINATIONS

How often does your child describe vivid dreamlike experiences?

How often does your child see or hear things that are not really there when falling asleep or waking up?

Excessive Daytime Sleepiness

Excessive daytime sleepiness may present differently in children. Excessive daytime sleepiness is the cardinal symptom of narcolepsy. All children and adolescents with narcolepsy experience excessive daytime sleepiness; however, they may not experience all of the other 4 symptoms. In children, excessive daytime sleepiness may be difficult to identify because its presentation can vary from that of adults. Because of its unusual presentation in children, excessive daytime sleepiness is often mislabeled or overlooked as laziness or inattention. When associated with irritability and hyperactivity, it is often misdiagnosed as another behavioral condition, such as ADHD.

Recognizing Excessive Daytime Sleepiness

- Younger children may present with elongation of daytime naps and nighttime sleep, with earlier bedtimes.
- Older children may restart regular daytime napping after naps have been discontinued.
- Nap durations are generally longer compared with adults and may be un-refreshing.
- Confusional arousals, or awakening without becoming fully aware,¹⁵ lasting up to 15 to 30 minutes, can occur upon awakening in the morning or from naps.
- Children and adolescents with excessive daytime sleepiness may present as aggressive, irritable, or hyperactive in an attempt to cope with or counteract sleepiness.
- Excessive daytime sleepiness is often not recognized as abnormal until cataplexy appears.

ASKING PATIENTS AND CAREGIVERS ABOUT EXCESSIVE DAYTIME SLEEPINESS

Is your child very difficult to wake up in the morning?

Is your child constantly tired or sleepy during the day?

Does your child take a nap during the day? If so, how long does it last and is it refreshing?

Have you noticed any hyperactivity or difficulty focusing?

Sleep Paralysis

Because children may have difficulty describing sleep paralysis and appropriately reporting the inability to move when falling asleep or waking up, it may be difficult to confirm sleep paralysis in children.

Recognizing Sleep Paralysis

- In addition to being unable to move when falling asleep or waking up, children with sleep paralysis may describe difficulty breathing, despite normal respiratory function.
- Sleep paralysis usually lasts for a few seconds to a few minutes and can end spontaneously or when the child is touched, shaken, or spoken to.
- Sleep paralysis may be an age-dependent phenomenon, as its appearance slowly progresses in young patients along the disease course.

ASKING PATIENTS AND CAREGIVERS ABOUT SLEEP PARALYSIS

How often does your child feel unable to move or speak when falling asleep or waking up?

Sleep Disruption

Children and adolescents with narcolepsy may have difficulty maintaining sleep, and they may have frequent nighttime awakenings.

Recognizing Sleep Disruption

- In children and adolescents, disrupted nighttime sleep may impact cognition, emotional regulation, and neurobehavioral functioning.
- Disrupted nighttime sleep may not be reported by pediatric patients or their caregivers unless targeted questions are asked.

ASKING PATIENTS AND CAREGIVERS ABOUT SLEEP DISRUPTION

Is your child very difficult to wake up in the morning?

How many times does your child wake up during the night? For how long?

If you have any questions, or have a pediatric patient with any of these symptoms, please call or send us a referral at Phone/Fax **1-888-884-9493**. For scheduling select extension 706. We specialize in children and adults from the age of 2 to 99.

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