

Incidence of Obstructive Sleep Apnea Is Higher Among Children Who Have Cerebral Palsy and Epilepsy

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Obstructive sleep apnea (OSA) is a sleep disorder that occurs when the throat muscles relax during sleep, blocking airflow in the nose and mouth. Although largely associated with adults, the disorder also affects children, particularly those who have complex medical conditions causing hypertonia or hypotonia (abnormal muscle tone).

A number of childhood-onset disabilities can affect muscle tone, leading to development of OSA. Cerebral palsy is among the most common, particularly when the condition is severe or combined with an epilepsy diagnosis. Routinely assessing cerebral palsy patients for sleep disorders can facilitate accurate diagnosis and successful treatment of OSA in this population.

Heightened Risk Factors

Children who have cerebral palsy typically have hypertonia (high or tight muscle tone) in their extremities and hypotonia (low or loose muscle tone) in their midline. This tone combination is the opposite of typically developing children, who have relatively loose extremities and a rigid midline. As a result of their loose midline tone, children who have cerebral palsy may experience difficulty holding the muscles of their airway upright. Abnormal tone also affects pulmonary function, leading to decreased air reserves. Additionally, many children who have cerebral palsy exhibit decreased mobility, making it difficult or impossible to escape their episodes of apnea by repositioning during sleep. This is especially true for children who have more severe forms of cerebral palsy—such as levels III, IV or V—as measured by the Gross Motor Function Classification System (GMFCS). These children often spend their entire night supine, a vulnerable position for apnea as the tongue can more easily fall backwards into the airway.

KEY INSIGHTS

- Cerebral palsy is among the most common disabilities that can lead to development of obstructive sleep apnea (OSA).
- The likelihood of OSA increases with more severe forms of cerebral palsy, or when cerebral palsy is combined with an epilepsy diagnosis.
- Other groups at heightened risk for developing OSA include children who have neuromuscular conditions or craniofacial anomalies.
- The Pediatric Sleep Questionnaire (PSQ) is an effective tool for providers to assess patients for sleep problems.
- Treatment for OSA may be medical or surgical depending on the child's age, condition and other factors.

They might also have difficulty swallowing or handling excess secretions, putting them at greater risk for upper airway congestion.

Certain medications might also increase the likelihood of children developing OSA. A number of anti-seizure medications, for example, have a sedative effect, making it more difficult to wake from an apnea episode. Medications to address abnormal muscle tone, such as baclofen or diazepam, can affect airway tone as well.

Evaluating Patients Suspected of OSA

Primary care physicians and pediatricians should consider evaluating all patients for sleep disorders, paying special attention to patients who have moderate to severe forms of cerebral palsy or co-occurring cerebral palsy and epilepsy. Additional high-risk groups include patients who have other conditions affecting muscle tone, such as neuromuscular disorders (e.g. Duchenne muscular dystrophy or spinal muscular atrophy), and patients who have craniofacial abnormalities that might reduce airway size (e.g. cleft lip and palate, craniosynostosis or Apert syndrome). Obese patients and patients less than 1 year old should also be carefully evaluated. Certain typically developing children may have hereditary risk for developing OSA.

Evaluation varies depending on the provider's preferred approach. An open-ended question—"How is your (or your child's) sleep?"—followed by high-yield questions—"Is your child experiencing snoring or breathing pauses during the night?" can trigger productive dialogue about previously unexpressed concerns. The Pediatric Sleep Questionnaire (PSQ) is an effective tool for providers to consider, as well. It includes 20 standardized questions and is publicly available for use. A score of eight or more PSQ questions answered positively is associated with increased risk for OSA. A recent study found significantly more children with cerebral palsy (58 percent) or cerebral palsy and epilepsy (67 percent) exhibited increased PSQ scores than a comparison group (27 percent)¹.

Physicians who suspect OSA or another sleep disorder in a medically complex patient should refer to a specialty sleep center with pediatric expertise. There, a sleep medicine specialist will perform a polysomnogram to verify the diagnosis. Children who

have one or more complex conditions, such as cerebral palsy and epilepsy, may benefit from a combined polysomnography and video electroencephalogram (VEEG). Together, the tests can confirm the cause of a child's sleep concerns—abnormal muscle tone, seizures, or another factor—and inform treatment recommendations. Testing can also assess children for hypoventilation, another potential problem for children who have conditions resulting in abnormal muscle tone.

Treatment and Continued Assessment

Once a diagnosis of OSA is verified, treatment may be medical or surgical depending on the child's age, medical condition, and other contributing factors. An adenotonsillectomy, a routine procedure that can improve OSA, might be recommended when there is evidence of adenotonsillar hypertrophy, or enlargement of the adenoid tonsil. Continuous positive airway pressure (CPAP) or bilateral positive airway pressure (BiPAP) therapy can help keep the airway open during sleep and may be appropriate for certain patients. Medications such as steroid sprays may also be effective. A sleep medicine specialist can review test results and discuss treatment options with the child's family.

Providers should continue to assess medically complex patients for sleep problems during routine check-ups, as issues will often develop over time. Any change in medication, for example, has the potential to cause or exacerbate OSA. Maturity and facial shape changes can also cause problems, as some children's cerebral palsy prevents them from closing their mouths.

For children who have neuromuscular disorders, disease progression can result in new or worsening apnea issues. Typically developing children who have OSA, in contrast, will often improve with 'watchful waiting' and continued monitoring².

Treating OSA can improve a child's quality of life and prevent other complications, such as growth delays and social issues. In fact, research suggests that children who have cerebral palsy and are effectively treated for OSA can see nearly 20 percent improvement in overall health³. Successful treatment also improves quality of life for families and caregivers, the individuals most impacted by a child's poor sleep.

References

¹ Garcia, J., Wical, B., Wical, W., Schaffer, L., Wical, T., Wendorff, H. and Roiko, S. Obstructive sleep apnea in children with cerebral palsy and epilepsy. *Developmental Medicine and Child Neurology* 2016.

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BiPAP Therapy Improves Quality of Life

A 16-year-old boy who has spasticity and seizures caused by Bohring-Opitz Syndrome was referred to one of the authors when he developed nighttime snoring following two major surgeries. A polysomnography resulted in a diagnosis of obstructive sleep apnea (OSA). Bilateral positive airway pressure (BiPAP) therapy was recommended.

Within two weeks of beginning BiPAP treatment, his family noticed physical and emotional improvements. His post-surgical healing was expedited and he experienced fewer upper respiratory infections, to which he had previously been prone. Additionally, he became happier, more alert and more interactive with his family.

Sleep Study, Endoscopy Informs Treatment Plan

A 16-year-old girl who has Noonan syndrome and a seizure disorder was referred to Gillette after struggling with obstructive sleep apnea (OSA) for several years. After her first sleep study, a pediatric otolaryngologist performed an endoscopy and determined surgical intervention would not be beneficial. Continuous positive airway pressure (CPAP) therapy was recommended. A follow-up sleep study 24 hours later implemented continuous positive airway pressure. Her family, who previously had expressed concern about their daughter's potential to choke during sleep, reported "a real sense of relief" and improved sleep for the whole family.



John Garcia, MD

John Garcia, MD, a board-certified sleep medicine physician, sees patients who have disabilities and associated sleep disorders, including insomnia, obstructive sleep apnea, sleep-walking, hypersomnolence, circadian rhythm disorders and restless leg syndrome. He uses a combination of behavior management, medications, surgery and other therapies in his practice.

Garcia, a graduate of the University of Iowa School of Medicine, completed a residency in pediatrics and one year of fellowship training in behavioral/developmental pediatrics at Riley Hospital for Children in Indianapolis. He then completed a sleep fellowship equivalent at the Minnesota Regional Sleep Disorders Center in Minneapolis. His professional associations include the American Board of Sleep Medicine and the American Board of Pediatrics.

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Beverly Wical, MD

Beverly Wical, MD, is a board-certified pediatric neurologist. She has special expertise in epilepsy, seizures and the neurological problems of infants and young children. In addition to her work at Gillette, Wical serves as clinical faculty member in the Department of Pediatrics at the University of Minnesota.

Wical received her medical degree from Loma Linda University in Loma Linda, California. She also completed a pediatric residency and child neurology fellowship at Loma Linda University. Her professional memberships include the Child Neurology Society and the American Epilepsy Society.



Jennifer Maytum, DNP

Jennifer Maytum, DNP, is a doctor of nursing practice, pediatric nurse practitioner, and certified neuroscience registered nurse at Gillette. She works with patients who have sleep problems, including insomnia, excessive fatigue, parasomnias (night terrors, sleepwalking, and confusional arousals), and sleep disordered breathing (sleep apnea).

She has many years of experience working with children who have complex medical and neurological conditions.

Maytum obtained her doctor of nursing practice and master of science degrees from the University of Minnesota. She is certified in primary care by the Pediatric Nursing Certification Board. She has an additional certification in advanced interdisciplinary studies, with a focus on developmental disabilities, from the University of Minnesota. Maytum is a member of the American Association of Neuroscience Nurses, the National Association of Pediatric Nurse Practitioners, and the American Holistic Nurses Association.



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NEWS & NOTES

Phalen Clinic Now Open

Several key outpatient services, including neurology and sleep medicine, have moved to a new Gillette location. The Phalen Clinic is located on the second floor of 435 Phalen Blvd. in St. Paul, in the same building that houses the Gillette Lifetime Specialty Healthcare adult clinic. To assist with diagnosis and treatment, we have on-site electroencephalogram (EEG) and video electroencephalogram (VEEG). Overnight sleep studies will continue to be conducted at the main hospital campus located at 200 University Ave. E. in St. Paul. Pediatricians and primary care physicians can call the Gillette One-Call Access phone number at 651-325-2200 or 855-325-2200 (toll-free) to make a referral.

Motor Delay Clinic Facilitates Early Diagnosis, Intervention

When young patients aren't meeting certain motor milestones such as rolling over, crawling or walking, providers may want to consider a referral to the recently opened Motor Delay Diagnosis and Treatment Clinic at Gillette. At the clinic, a multidisciplinary team of specialists including a pediatric neurologist, pediatric rehabilitation medicine specialist and a pediatric orthopedic surgeon evaluate, screen and diagnose children who have a new or suspected diagnosis of cerebral palsy. Once a diagnosis is confirmed, the team works closely with families and primary care physicians to provide early intervention and establish a treatment plan. Call the One-Call Access phone number listed above to make a referral.

Musculoskeletal Conference Is Dec. 2

On Friday, Dec. 2, 2016, Gillette will present "Musculoskeletal Management in Pediatrics," an accredited continuing medical education course for medical providers. The conference will be held at the Saint Paul RiverCentre from 7:30 a.m. to 4:30 p.m. For a course description and to register, go to gillettechildrens.org/musculo16.