

Ask The Experts

QUESTION: When is a sleep study necessary in people with neuromuscular disorders (NMD) in order to diagnose breathing problems? Are the screening devices such as ApneaLink™, for example, useful in diagnosing sleep apnea?

ANSWER: Barbara Phillips, MD, MSPH, FCCP, Professor, Division of Pulmonary, Critical Care and Sleep Medicine, and Director, Good Samaritan Sleep Center, University of Kentucky College of Medicine, Lexington, Kentucky

Screening studies such as ApneaLink and others are very good at ruling IN people with obstructive sleep apnea. However, hypoventilation and central sleep apnea, which are frequently seen in people with neuromuscular disorders are NOT well identified by portable or home sleep studies. Portable oximetry (simply measuring the oxygen level with a skin clip at night) can help screen and identify a person who is having significant drops in oxygen at night, but this may be a late sign. A person who has a neuromuscular disorder should consult with a respiratory sleep specialist (usually a pulmonologist who is board-certified in sleep) if he/she has shortness of breath at rest or with trivial exercise, if he/she wakes up gasping for breath, light-headed, with a racing heart or a headache. Sometimes simple pulmonary function testing can give us an idea about when more extensive testing, such as a sleep study, will be necessary.

ANSWER: Lisa Wolfe, MD, FCCP, Assistant Professor, Division of Pulmonary & Critical Care, Northwestern University Feinberg School of Medicine, Chicago, Illinois

NMD patients will most commonly develop nocturnal hypoventilation and REM-associated central sleep apnea and, less commonly, obstructive apnea. These ventilatory issues disrupt sleep quality, decrease oxygen saturation (SaO₂) in the blood and elevate carbon dioxide (PaCO₂) in the blood. The symptoms (see sidebar) are gradual, and waiting for all the symptoms to manifest is not recommended, because then the patient can be in respiratory failure. Reduction in lung function **or** low oxygen saturation **or** elevated carbon dioxide is sufficient to qualify an NMD patient to start ventilatory support.

Guidelines from Medicare do not require a sleep study in order for a physician to prescribe a ventilator. In some cases, polysomnography is still recommended for diagnostic purposes, but most sleep labs have no protocols for the treatment of those with NMD.¹ Recently the American Academy of Sleep Medicine has released standards for the care of those with hypoventilation. If an NMD patient is referred for a study, they should confirm that the lab is aware of the new standards and is prepared to follow the guidelines.²

Many devices are now available to facilitate home testing for obstructive sleep apnea, and although they are very accurate for both screening and diagnosis, these devices have not been tested in and are not indicated for those with NMD. This does not mean they have no role in NMD. If oxygen saturation is 88 percent or less for more than 5 minutes during the recording, this would suggest that hypoventilation is present, and ventilatory support should be initiated.

It is important to remember that home apnea testing is not meant to replace pulmonary function testing as the primary screening tool in NMD. ▲



SLEEP AND BREATHING TERMS

OSA or obstructive sleep apnea occurs when a person stops breathing periodically during sleep because muscles in the throat collapse and block the airway.

CSA or central sleep apnea occurs periodically when the brain fails to send the appropriate signals to initiate breathing. There is neither air flow or chest wall movement.

Mixed apnea is a combination of OSA and CSA.

Apneas are repeated episodes during sleep when breathing ceases for at least 10 seconds. More than 10 apnea episodes per hour indicate a need for further evaluation and treatment. **Hypopneas** are repeated episodes during sleep when air flow in and out of the lungs occurs, but is reduced. The apnea-hypopnea index (**AHI**) counts the number of apneas and hypopneas that occur per hour of sleep and is useful in quantifying the severity of sleep apnea.

Polysomnography is the study of an individual's sleep cycles and stages with a device that records information such as air flow, brain activity, blood oxygen levels, body position, breathing efforts and eye movement.

Hypoventilation or underventilation typically begins during sleep as the level of oxygen in the blood decreases and is offset by an increase in the level of carbon dioxide. This is due to the inability of weak inspiratory muscles (the muscles of breathing in) to allow the individual to take a deep breath. Signs and symptoms include: fatigue, shortness of breath, morning or continuous headaches, daytime drowsiness and falling asleep frequently, sleep awakenings with shortness of breath or heart racing, difficulty swallowing, poor concentration and impaired cognition, frequent nightmares, frequent arousals from sleep to urinate and heart failure, such as cor pulmonale, due to breathing problems.³

Are you a ventilator user or health professional with a question about home mechanical ventilation?

Send it to info@ventusers.org, and IVUN will find experts to answer it.

REFERENCES

1. Finder JD, Birnkrant D, Carl J, Farber HJ, Gozal D, Iannaccone ST, Kovesi T, Kravitz RM, Panitch H, Schramm C, Schroth M, Sharma G, Sievers L, Silvestri JM, Sterni L. Respiratory care of the patient with Duchenne muscular dystrophy: ATS consensus statement. *Am J Respir Crit Care Med* 2004; 170(4):456-65
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3. Bach JR. *Guide to the Evaluation and Management of Neuromuscular Disease*. Philadelphia: Hanley & Belfus, Inc, ©1999.