

Breathing Problems in Adults with Neuromuscular Weakness

Neuromuscular diseases may result in respiratory muscle weakness leading to breathing problems. Difficulty with breathing is often first noticed when lying flat and during sleep. It is important to treat breathing problems early. You should know what to look for and what to tell your healthcare provider to get the right treatment. You may also develop serious breathing problems and not have a lot of symptoms. Your healthcare provider will do tests to check your lung function and monitor your breathing.



What is neuromuscular weakness?

Neuromuscular diseases affect the function of muscles in your body. These conditions are usually due to problems with the nerves not sending information to your muscles and/or the muscles not working correctly. Some examples of diseases that cause neuromuscular weakness in adults include:

- ALS (amyotrophic lateral sclerosis, also known as Lou Gehrig's Disease)
- Muscular dystrophy
- Myasthenia gravis

How can neuromuscular weakness affect my breathing?

Many muscles are needed for normal breathing. These include your diaphragms (muscles that separate your lungs from your belly), the muscles between your ribs (intercostal), and muscles in your neck and throat. With neuromuscular weakness, some or all of these muscles may become tired (fatigued), making it difficult for you to breathe in and out. This weakness may cause you to take shallow breaths. You may feel like you are suffocating when lying down and sleeping. Shallow breathing from muscle weakness, known as hypoventilation, may lower your body's oxygen level and increase the carbon dioxide level in your blood. An increase in carbon dioxide can lead to headaches, confusion, and decreased awakening from sleep.

Neuromuscular weakness may also make it difficult for you to cough effectively. When you cannot cough well, you are not able to clear the mucus from your lungs. As a result, you may develop more frequent lung infections (pneumonia), or feel like it is harder to breathe due to mucus blocking your airways.

Sometimes the decrease in lung function and breathing problems comes on gradually. You may not have symptoms or realize how limited your breathing is. Your healthcare provider will help you watch out for breathing problems that may develop from your neuromuscular disease.

If your respiratory muscle weakness is getting worse, your healthcare provider will discuss tests and treatment options with you.

What kind of sleep problems should I look for if I have a neuromuscular condition?

Three problems that can affect breathing with neuromuscular weakness while sleeping are:

- Nocturnal hypoventilation
- Obstructive sleep apnea
- Central sleep apnea

You can have any or all of these problems depending on the type of neuromuscular disease you have.

Nocturnal hypoventilation

This problem is due to shallow breaths during sleep from muscle weakness. Hypoventilation leads to an increase in your blood carbon dioxide level and eventually a decrease in oxygen level. It is often worse during REM sleep (also known as "dreaming" sleep) when you rely on the diaphragm as your only muscle for breathing. The first sign you notice can be trouble breathing when lying flat. Your weakened respiratory muscles have trouble expanding your chest enough to take a normal sized breath. Some people describe the feeling of "not getting in enough air". Being overweight can make this problem even worse. Sleeping in a more upright position by adding extra pillows under your head can help lessen this problem.

Obstructive sleep apnea (OSA)

OSA is a condition during sleep in which the soft tissues in the back of the throat and tongue collapse and block (obstruct) air from flowing into the lungs normally. This can cause your breathing to stop or "pause" for several seconds or longer. Neuromuscular weakness can make underlying OSA worse. (For more information, see ATS Patient Information Series at www.thoracic.org/patients)

Central sleep apnea (CSA)

At times during sleep a person with neuromuscular disease may have problems with the brain not sending a signal to the lungs and muscles to breathe. If the normal cue from the brain to breathe is interrupted for a short time, it is called central apnea

during sleep. The central pauses in your breathing may cause drops in your oxygen level.

What are the symptoms associated with sleep disordered breathing from neuromuscular weakness?

Sleep disordered breathing may result in any of the following symptoms:

- snoring
- gasping or choking
- frequent awakening during sleep
- feeling tired or sleepy during the daytime
- morning headaches
- mood swings
- memory problems
- leg swelling

If you are experiencing any of these symptoms, you should talk with your healthcare provider.

What tests are done to look for sleep problems in neuromuscular disease?

There are several kinds of tests your healthcare provider may order to look for breathing problems due to neuromuscular problems even if you don't have symptoms.

Pulmonary function tests are helpful in looking for respiratory muscle weakness because they measure how much air you breathe in and out. (See also the ATS Patient Information Series fact sheet at www.thoracic.org/patients)

An arterial blood gas (ABG) is done using blood drawn from an artery, usually in the wrist. It measures the level of oxygen and carbon dioxide in your blood. A oxygen saturation monitor (pulse oximeter) only measures oxygen levels in the blood, but does not give levels of carbon dioxide.

Peak cough flow is a test that measures the strength of your cough. The higher the flow, the stronger your cough.

A sleep study (known as polysomnography) is done to look for sleep disordered breathing. Sleep studies can be performed at home or in a sleep center. (See also ATS Patient Information Series Fact sheet at www.thoracic.org/patients)

What can be done to treat breathing and sleep problems resulting from neuromuscular weakness?

If you have breathing problems during sleep, you benefit from using some type of non-invasive ventilation. Common types of non-invasive ventilation are CPAP (continuous positive airway pressure) and BPAP (bilevel positive airway pressure). The air pressure is delivered through a mask that fits over your nose and mouth, or just your nose. You will be fitted with the size and style of mask that fits your face.

These devices help you to breathe during sleep in several ways. They help relieve obstructive sleep apnea by using air pressure to keep your upper airway open. Positive pressure also helps you take a bigger, more normal sized breath, despite your weak respiratory muscles. If you have central apnea, a breathe rate can also be added to help assure you take a certain number of breaths each minute. Your healthcare provide may have you use supplemental oxygen. This may be given together with the non-invasive ventilation.

If you have a weak cough, you may benefit from using a cough assist device (also known as mechanical insufflator-exsufflator). This device simulates a deep cough by blowing air into your lungs

and then quickly sucking it out. You use it with a mouthpiece or facemask. Using it every day will help to keep your lungs clear of mucus. This device will also increase the volume of air in your lungs, which improves your oxygen level and reduces your risk of lung collapse and infection.

There are other things you can do to improve your sleep with neuromuscular weakness. It is important to follow a balanced diet. Your respiratory muscle strength will worsen with poor nutrition. Try to lose weight if you are overweight. Avoid smoking or vaping. Limit your alcohol intake, particularly before bedtime. Some medicines can affect breathing during sleep, so be sure to review with your healthcare provider any medication you are taking, including illicit drugs and over-the-counter medications.

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Rx Action Steps

If you have neuromuscular weakness, talk with your healthcare provider about your sleep problems and any concerns you may have about your breathing. Your healthcare provider may suggest tests to check your breathing function.

Let your healthcare provider know if you are having any of the following:

- ✓ Difficulty breathing when lying flat
- ✓ Snoring, gasping, or choking at night
- ✓ Morning headaches
- ✓ Confusion or "foggy" thinking
- ✓ Low oxygen level with a pulse oximeter reading
- ✓ Others notice that you have breathing pauses or a strange breathing pattern during sleep

Healthcare Provider's Contact Number:

Resources

American Thoracic Society

- www.thoracic.org
 - Obstructive sleep apnea in adults
 - PAP therapy in adults
 - Lung function testing
 - Pulse oximetry
 - Sleep studies

Muscular Dystrophy Association Website

- <https://www.mda.org/disease>

ALS Association Website

- <http://www.alsa.org/als-care/resources/publications-videos/factsheets/breathing-difficulties.html>

Breathe NVS Website

- <https://www.breathenvs.com/>

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