MAINTAINING PULMONARY FUNCTION WITH DUCHENNE MUSCULAR DYSTROPHY

Pulmonary function is how well your lungs work. With **Duchenne muscular dystrophy (DMD)**, muscles gradually weaken over time. This can affect the respiratory muscles that maintain pulmonary function.

The effects of DMD on pulmonary function depend on the stage of the disease. Most people with DMD do not have trouble breathing or coughing while they are ambulatory (able to walk). During this stage, individuals may not feel a difference in their breathing, but breathing tests may show a decrease in lung function. At the stage where individuals lose the strength to walk, typically the muscles around the lungs also weaken to the point that they can no longer move the chest wall.

It is important for people with DMD to see a pulmonologist even before any noticeable loss of strength so the doctor can follow them over time. The pulmonologist will monitor respiratory muscle functions and recommend the timely use of airway clearance, assisted coughing, and ventilation. Starting these therapies early can decrease respiratory complications, improve quality of life, and prolong survival.

Ethan

2022 MDA National Ambassador, living with DMD

MDA® Muscular Dystrophy Association

How DMD affects breathing

As respiratory muscles weaken, people with DMD may develop ineffective cough and decreased ventilation. These can cause respiratory muscle fatigue, mucus plugging, pneumonia, atelectasis (collapsed airways), and respiratory insufficiency — initially during sleep and eventually while awake. Many people with DMD don't realize when they no longer have an effective cough until a respiratory infection leads to a prolonged cough or pneumonia.

Breathing too shallowly during sleep can cause hypoxemia (low oxygen levels in blood) and hypercapnia (high carbon dioxide levels). These abnormal breathing patterns can disrupt sleep, impacting the ability to think, how the body grows, and quality of life. Individuals might experience morning headaches, fatigue, nightmares, difficulty staying awake, and unexplained weight loss.

Scoliosis (curvature of the spine) is relatively common in DMD. This can further affect respiratory function because the curve of the spine can impact the structure of the chest wall. Individuals with scoliosis should see an orthopedic specialist to discuss interventions for prevention and treatment.

Keeping lungs healthy

With developments in treatment and care, the stage at which a person with DMD needs ventilation is about a decade later than it used to be. Novel gene therapies have the potential to revolutionize DMD management even further.

In the meantime, it is important to have good hygiene and a healthy lifestyle, which can help prevent lung infections and slow down the decline in respiratory function.

Follow these tips to maintain lung health:

- Wash your hands often with soap and water. Alcohol-based hand sanitizer is a good alternative if you can't wash.
- Avoid crowds during cold and flu season.
- Avoid cigarette smoking, vaping, and other chemical inhalants. These can damage your lungs, worsen your airway clearance, and increase the chance of respiratory infections.
- Avoid exposure to secondhand smoke, chemicals in the home and workplace, and radon. These can cause or worsen lung disease.
- Make your home and car smoke-free zones.

MEDICAL EQUIPMENT YOU CAN USE AT HOME

During the **ambulatory stage** (generally from diagnosis to around age 10-12), most people with DMD do not need respiratory devices. If you are in the **nonambulatory stage** (unable to walk without assistance), depending on your symptoms, your healthcare provider might recommend using any of these devices at home.

Cough assist device

If your coughing becomes weak, manual or mechanically assisted cough techniques (e.g., insufflator-exsufflator, also called a cough assist device) can help you clear your airway and keep mucus and germs out of your lungs. Your healthcare provider might prescribe the cough assist device to be part of an airway clearance regimen, especially when you are sick.

Nebulizer

This machine turns liquid medicine into a mist that can be used for airway clearance inhalational therapies.

Vest therapy

This is called vest therapy because you wear an inflatable vest that delivers positive pressure air pulses to the chest wall, called high-frequency chest wall oscillation (HFCWO). The oscillations and vibrations in the chest can help loosen thick secretions and produce a cough. People with respiratory muscle weakness typically do not have abnormally thick mucus, so vest therapy is not used frequently for DMD.

Positive expiratory pressure (PEP) therapy

Portable devices like **Flutter** or **Acapella** might be used for regular airway clearance. These oscillating PEP devices work by providing resistance and vibration when you exhale through them. This helps move mucus from the surface of the airways so it can be coughed out.

Bi-PAP™

The most common device used for noninvasive ventilation assistance is bilevel airway pressure (Bi-PAP™). The Bi-PAP device delivers air into the lungs through a mask or mouthpiece to improve the oxygen and carbon dioxide levels in your body and to open up the airways, which can collapse because of weak respiratory muscles. This device often is used at night to help improve breathing during sleep, but it can also be used during the day. It takes some time to get used to assisted ventilation, so it's important to work with your pulmonary team to adjust your ventilation settings and try various interfaces until you find the most comfortable and safest regimen for you.

Humidifier

A humidifier is prescribed for people who use a ventilation device. It ensures the air you breathe is at an ideal temperature and humidity.

Tracheostomy tube

A tracheostomy involves creating an opening in the neck and inserting a tube in the windpipe that can be connected to a ventilator. The tracheostomy tube also is called an artificial airway or a trach. Invasive ventilation decreases the respiratory effort, secures the airway, and allows a caregiver to suction the airways. This is especially important when you have a respiratory infection. Having a tracheostomy is a personal decision between you and your pulmonologist. Some individuals prefer a tracheostomy because they cannot tolerate noninvasive ventilation, have swallowing issues, or find that noninvasive ventilation is not sufficient for their breathing problems. Newer home ventilators are small, portable, and lightweight, allowing you to maintain your daily living activities, including going to the grocery store or doctor's office, visiting family and friends, and traveling.

Pulse oximeter

This device clips onto the fingertip and measures the oxygen level in the body. It is prescribed for individuals who use any respiratory support device.

Other equipment

In the later stages of DMD, some people use a **suction device** and a **self-inflating bag and mask** for airway clearance and respiratory support when needed. **Oxygen delivery devices** (oxygen concentrator and/or oxygen tanks) are sometimes needed if ventilation devices are not enough to bring the oxygen levels in the body to an optimal level.

Although some devices need to meet certain criteria, these devices are covered by almost all insurance companies. Your pulmonary care team will help with obtaining insurance coverage when these devices are needed.

Learning to use pulmonary equipment at home

When you begin using a new type of respiratory device, your providers will offer training at clinic. In addition, a respiratory therapist from a home care company may come to your home to help set up your pulmonary equipment and train you and your caregivers on using it. Companies that supply pulmonary equipment have on-call providers to help with training and troubleshooting the devices. You can always reach out to your primary healthcare providers if you have any questions.

PREPARING FOR AN MDA CARE CENTER VISIT WITH YOUR PULMONARY TEAM

It is important for people with DMD to see their pulmonary care team at least once a year starting at 5 or 6 years old, and at least twice a year starting between 10-12 years old, or when they can no longer walk without assistance.

What to expect

During a pulmonary clinic visit, providers will take a medical history, perform a physical examination, measure lung function (pulmonary function tests), and perform other studies as needed. Pulmonary function tests involve breathing into a computerized machine through a mouthpiece while a clip blocks your nose. The machine measures lung capacity, muscle strength, and other indicators of how well the lungs are working.

Although you do not need to prepare for pulmonary function tests at home, be aware that your pulmonary care team will most likely perform these tests during your visit. Measuring lung capacity while an individual with DMD is still able to walk helps them become familiar with how to use the measuring equipment, and it can also help detect and manage any potential problems early.

Common tests

If your pulmonologist suspects you may be breathing too shallowly at night (nocturnal hypoventilation) or you may have sleep apnea, they may order a sleep study. Sometimes, the pulmonary team will perform other tests, including lung imaging, blood tests, oxygen/carbon dioxide measurements, swallowing/feeding evaluation, and microbiological studies. The most common symptoms indicating insufficient breathing (underventilation or hypoventilation) are fatigue, poor sleep, nightmares or night terrors, and headaches, especially right after waking. Reach out to your pulmonary team if you are experiencing any of these symptoms, if your cough gets weaker, or if you have any questions/concerns about your medications, medical equipment, or treatment plan.

What to bring

Bring any breathing equipment you use at home (Bi-PAP machine, cough assist machine, etc.) with you to your clinic visit. Your pulmonary team might adjust the settings of these devices while you are at the clinic and troubleshoot any possible issues.

How to prepare

Before your clinic visit:

- Confirm your visit with the clinic.
- Write a list of your questions and concerns and bring it with you.
- Gather any medical documents that you might need.
- Prepare any medicines, durable medical equipment, and emergency supplies you need to bring with you.
- Arrange your transportation.
- Request an interpreter if you need one.

MAINTAINING YOUR PULMONARY FUNCTION AT HOME

Breathing consists of two steps: inhalation and exhalation. Each step requires several different muscles. Breathing exercises, good posture, healthy eating, infection prevention, sufficient ventilation, and airway clearance are all important factors in maintaining good pulmonary function and quality of life for people with DMD.

Your healthcare provider may refer you to outpatient physical therapy (PT), a nutritionist, or other healthcare providers who can help with these factors. But there also is a lot you can do at home to promote good breathing capacity:

1. Practice breathing exercises and relaxation techniques on your own, if your medical team recommends it. Here's a simple exercise: Take five to 10 deep breaths with a short rest in between, several times a day. This strengthens the lungs and helps them expand fully.

2. Your provider might recommend breath stacking therapy, also called lung volume recruitment. The idea is to stack one small breath on top of another until the lungs are filled. Filling your lungs to their maximum capacity can increase lung volume, cough effectiveness, lung and chest suppleness, and speaking volume. Breath stacking can be done with a mouthpiece or face mask and an Ambu® bag (manual resuscitator bag).

3. Self-assisted cough is a technique that you can perform without a cough assist device if you have the ability to cough. Start in a sitting position and fold your arms below your rib cage, over your stomach. Take a deep breath and hold it until you have to cough. When you begin to cough, lean your upper body forward and downward against your hands. A family member or caregiver can assist if your arms are weak. Ask your healthcare provider if this technique is appropriate for you before trying it.

4. Good nutrition helps maintain an ideal body weight, which is key to long-term DMD management. Being underweight or overweight are both harmful to respiratory health. It is also important to stay well-hydrated and to be aware of vitamin and mineral levels, especially vitamin D and calcium to prevent osteoporosis. Constipation also can make it more difficult to take deep breaths. Tell your healthcare provider if you have constipation, weight loss or excessive weight gain, gastroesophageal reflux, or swallowing problems.

5. Staying active as much as you can and slowing down the progression of scoliosis are also important for maintaining respiratory strength and function. Regular physical therapy and walking, if you are able, can help with both.

6. A good night's sleep can give you more energy and improve concentration. In addition to using respiratory devices at night if you need them, try adjusting the light, temperature, and noise levels in your room for better sleep.

7. It is important to keep up with your vaccinations, including the pneumococcal and annual flu vaccine. Consult with your doctor about you and your entire family getting the flu shot every year.

8. Oral hygiene is important to prevent bacteria in the mouth that can cause chest infections. In addition to brushing and flossing your teeth twice a day, visit your dentist on a regular schedule.

9. In the case of a respiratory emergency, it is important to know what to do and who to contact. Make an emergency plan with your pulmonary team and family, and make sure your family members and caregivers have a copy.

FREQUENTLY ASKED QUESTIONS

When do people with DMD need ventilation?

Usually between 15 and 20 years old, but this can vary. Using steroids during childhood, having a healthy lifestyle, preventing infections, and doing breathing exercises and treatments regularly may extend this by a few years.

How do I know if I need ventilation?

Your pulmonary care team evaluates your lung function with pulmonary function tests, chest imaging, blood tests, and sleep studies as needed. They will alert you of any concerning test results. Tell your healthcare provider if you experience shortness of breath or it takes longer than usual to catch your breath and if you are snoring at night, feeling tired, having trouble paying attention during the day, or waking up with headaches in the morning. These might be signs of needing ventilation.

Can I travel with ventilation?

Yes, you can. Make sure you and your caregivers know what to bring with you and what to do in an emergency. Long trips and air travel are possible with careful planning involving you, caregivers, your medical team, and airlines or transportation providers.

How can I prepare for an emergency?

The more prepared you are for emergencies, the better you can handle them. Keep records of your recent visits to the doctor, test results, medications you take, doctors' names and contact information, and your insurance information. It is a good idea to have all this information stored as hard copies in a file and digitally on your smartphone or portable device.

Determine who your emergency contacts are. During visits with your healthcare team, make a plan to handle respiratory emergencies. Learn the emergency phone numbers of the clinic and the hospital. Prepare an emergency bag of clothes, medical supplies, and medications, and bring your respiratory device with you to the hospital if you can. Call 911 if you are experiencing significant respiratory symptoms.

What should I know about anesthesia?

When you have medical procedures, let your providers know that you should avoid all inhaled anesthesia, especially the drug succinylcholine and any of its derivatives, which can cause life-threatening complications. Most intravenous (IV) anesthesia is safe for people with DMD.

You should always have pulmonary and cardiac evaluations before surgeries. You may need some ventilation support while waking up from anesthesia, even if you are not using a ventilation device at home.

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