

Guide to Respiratory Care for **NEUROMUSCULAR DISORDERS**



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Vision

To find a cure for neuromuscular disorders in our lifetime.

Mission

Muscular Dystrophy Canada's mission is to enhance the lives of those affected with neuromuscular disorders by continually working to provide ongoing support and resources while relentlessly searching for a cure through well-funded research.

For information about our programs and services, please visit our website—www.muscle.ca—or contact the office in your area (see back cover for complete list).

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Gender neutrality

This guide was written to be gender neutral. The information presented within can be applied equally to both sexes.

We'd like to hear from you

E-mail us at info@muscle.ca if you have comments or suggestions to help us make this booklet more useful for you and other readers.

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DISCLAIMER

This document is intended for general information and awareness. Muscular Dystrophy Canada will not be held responsible for misuse of information or any damages incurred as a result of its use. This document is not meant to replace consultations with your physician or to provide medical advice, diagnosis or treatment. For information specific to the condition affecting you or your family, please consult your physician.

1 Introduction

While neuromuscular disorders do not impair the lungs themselves, they can affect the muscles involved in breathing and coughing. This can lead to the impairment of these functions over time, making respiratory complications common in people with neuromuscular disorders.

Although breathlessness (a key symptom of breathing impairment) may not be obvious because of reduced physical activity or wheelchair use, people with neuromuscular disorders may still find that their breathing is significantly impaired. Issues can be detected if the appropriate breathing tests are performed, but symptoms may not be apparent until there is a complication (such as a lung infection). These symptoms can include recurrent chest infections, chronic headaches, ever-present fatigue and increased muscle weakness.

There are over 150 different types of neuromuscular disorders under the umbrella of Muscular Dystrophy Canada, and the majority of people with neuromuscular disorders are at risk of developing breathing problems. The degree and timing of respiratory complications vary, depending on the individual's diagnosis and overall health status. For instance, there are some neuromuscular disorders—such as Charcot-Marie-Tooth disease, which mainly affects the lower extremities—that only occasionally result in serious respiratory impairment. It is important to speak with your physician to understand the risks for respiratory complications associated with your particular diagnosis.

How to use this guide

This guide is intended to provide people who have neuromuscular disorders with the information they need to make informed choices about their respiratory care. It is meant for people of all ages, but we recognize that

parents and guardians are responsible for the care of a child. As a result, although we have used "you" to simplify the language, we are referring to anyone who must make a decision about respiratory care for someone with a neuromuscular disorder, whether it is for themselves or a loved one.

We encourage you not only to share and discuss the information contained in this guide with your family, but also to seek expert help and advice. Be your own advocate! As a patient, you decide the level of care that you wish to receive.

You may choose to read this guide from start to finish, or you may turn to the section that is most relevant to you. The following list of topics and chapter references will help you navigate the information contained within this guide.

I want to....

- understand the basics of breathing and how respiratory muscle weakness can affect my breathing (Chapter 2)
- learn how to monitor my respiratory health, and how to recognize and manage a respiratory crisis (Chapter 3)
- read more about pulmonary hygiene that will help keep my airways and lungs clear, and prevent breathing problems (Chapter 4)
- be informed about the signs and symptoms of complications, what might happen in a respiratory crisis and how to prepare for such an event (Chapter 5)
- understand the principles of mechanical ventilation and the available options (Chapter 6)
- learn about methods of mechanical ventilation, including those that are non-invasive (Chapter 7) and invasive (Chapter 8)
- be alerted to the risks of oxygen therapy and anaesthesia, or learn more about traveling, hydration and other important issues (Chapter 9)
- get advice on how to develop a plan that addresses my respiratory needs (Chapter 10)

2 The Respiratory System

The primary function of the respiratory system is to exchange oxygen and carbon dioxide within the body. The air we breathe is made up of several gases, primarily nitrogen (78%) and oxygen (21%). The human body requires oxygen (O_2) on a continuous basis for survival, and it is the job of the respiratory system not only to bring that oxygen into the body, but to exchange it for the carbon dioxide (CO_2) that our bodies produce as waste. This occurs through **respiration**—although most people simply think of it as “breathing.”

The muscles that we use for breathing

Breathing consists of two separate steps: **inhalation** and **exhalation**. Each step requires a number of different muscles, although not all of these muscles are solely dedicated to breathing, nor are they all used in both inhalation and exhalation.

When you breathe in (or inhale), the **diaphragm**—a dome-shaped muscle positioned between the chest and the abdomen—contracts, moving downwards and flattening out. At the same time, the **intercostal muscles** between the ribs contract to pull the rib cage up and out (see Figure 2.2). These two actions create a partial vacuum that causes fresh air to rush down the **trachea** (windpipe), through the **bronchi** (or small air tubes) in the lungs, and then into millions of tiny microscopic air sacs in the lungs known as the **alveoli**. The alveoli pass fresh oxygen into the bloodstream. Red blood cells then carry the oxygen throughout the body, where it is used by organs and tissue, producing carbon dioxide (see Figure 2.3).

When you breathe out (or exhale), the diaphragm and intercostal muscles relax. This reduces the volume of the chest cavity,

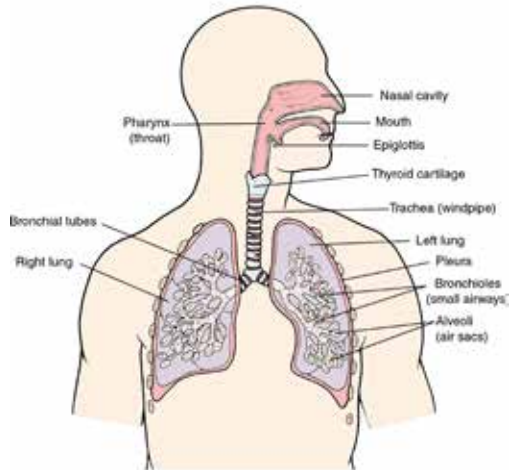


Figure 2.1
The respiratory system

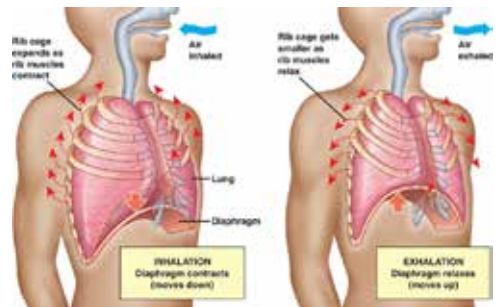


Figure 2.2
Inhalation and exhalation

increasing the pressure in the lungs. Air is forced out of the lungs and you exhale. The carbon dioxide, which the red blood cells have carried back to the lungs and passed to the alveoli, is then expelled from the body as part of the used air that is exhaled.

If you are breathing heavily, two additional muscle groups come into play. When you take a deep breath, muscles in the neck that attach to the collarbone and upper ribs assist in breathing, and when you force a breath out, the abdominal muscles help to push up the diaphragm.

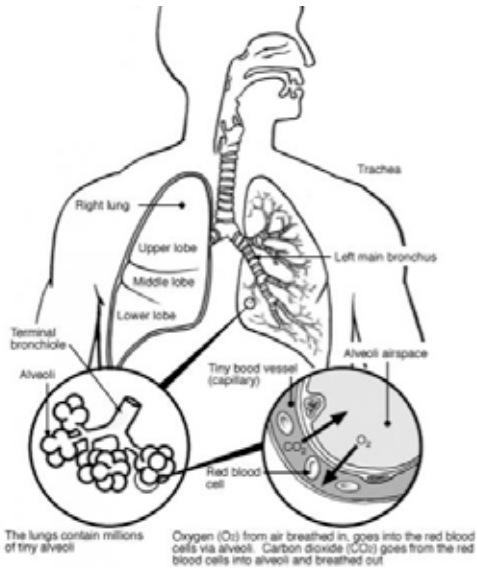


Figure 2.3
Lung showing alveoli

The amount of air we inhale with each breath (roughly 300–500 ml of air) is called the **tidal volume**, while the number of breaths per minute is called the **respiratory rate**. Adults typically breathe approximately 12–20 times per minute, while the respiratory rate of children is age-dependent and faster than that of adults.

Minute ventilation is the amount of air that must move in and out of the lungs every minute for someone to inhale enough oxygen and exhale enough carbon dioxide. Higher minute ventilation is necessary in situations where the body is producing elevated levels of carbon dioxide (such as during exercise or infection) or the lungs are not working at full capacity (due to disease or other impairment).

Coughing

We all cough from time to time and probably think nothing of it, but it is an important body function that keeps the airways and lungs clear of excess mucus and foreign bodies (such as dust). In people with neuromuscular disorders, however, weakened respiratory muscles can impair the ability to cough. This is a serious problem, because if your cough is ineffective and you are retaining mucus in your lungs and airways, you are at increased risk of pneumonia and respiratory failure.

It is important that you maintain the ability to bring up phlegm (expectorate). This can be done through effective coughing or with the help of therapies and assistive devices, as required.

Therapies to help with a weakening cough are discussed in detail in Chapter 4. It is particularly important to begin therapy before you have an ineffective cough (as that may make it more difficult to address potential pulmonary complications) or before you develop a respiratory infection (so that you can be familiar with the techniques when they are needed).

Muscle weakness and impaired respiration

Since neuromuscular disorders can cause muscle atrophy (wasting) and weakness, people with these disorders are at risk of developing breathing problems due to weakened respiratory muscles. This is a cause for concern because impaired breathing can result in low levels of oxygen in the blood (**hypoxemia**) and high levels of carbon dioxide (**hypercapnia**).

For more information about the symptoms and effects of impaired respiration, please see Chapter 5.

How much we need to breathe

The number of breaths we take each minute and the size of each breath (how much air we take in) vary from person to person. It depends on a number of factors, including the person's size, age, cardiopulmonary health and metabolism.

3 Monitoring Your Respiratory Status

Importance of monitoring

Respiratory monitoring provides information about your current respiratory status, as well as how quickly your respiratory muscles are weakening. Early and regular monitoring of breathing and coughing capacities is essential to detecting problems.

Begin by having a series of pulmonary function tests to assess how your breathing is right now. After that, your health-care team should set up a testing schedule to monitor your respiratory status.

The frequency and type of testing depends on your neuromuscular disorder and medical history. For example, you initially may be sent for annual or semi-annual tests, but the frequency may increase when there is evidence of progressive weakness or if you begin to experience symptoms of respiratory complications.

PULMONARY FUNCTION TESTS

Pulmonary function tests involve blowing into a tube or performing a series of breathing manoeuvres. These tests provide information about how much air your lungs can hold, how deeply you can breathe, how much air moves in and out of your lungs at rest, and/or the maximum amount of air you can move in and out of your lungs over the course of a minute. The results give your health-care team detailed information about your respiratory system, and they may point out its potential weaknesses.

RESPIRATORY MUSCLE STRENGTH

Tests to measure the strength of your respiratory muscles may include:

- maximum inspiratory pressure (MIP)
- maximum expiratory pressure (MEP)
- sniff nasal inspiratory pressure (SNIP)



Values below certain thresholds suggest a weakening of respiratory muscles and the ability to cough.

PEAK COUGH FLOW

Your health-care provider (e.g. physician or respiratory therapist) may measure your cough capacity to determine its effectiveness. The test consists of coughing forcefully into a tube or chamber (pictured above). Results showing decreased values may indicate a weakened ability to cough.

CARBON DIOXIDE MEASUREMENT

It is important to measure and track the carbon dioxide level in your blood as your breathing muscles become weaker. If your breathing is too shallow, the carbon dioxide level will rise. This is an indication that you may require breathing support.

Carbon dioxide is reliably measured by capillary blood gas (a finger prick) or arterial blood gas (a needle prick in the wrist area). It can also be measured with a small nasal probe or skin probe.



POLYSOMNOGRAPHY (SLEEP STUDIES)

Various levels of sleep studies can be performed to see how you breathe while sleeping. Some tests can be done at home with a small monitor (such as an oximeter) to painlessly measure the amount of oxygen in your blood through a sensor on the finger (pictured above). Other tests must be performed in a sleep lab (pictured right).

CARDIAC FUNCTION ASSESSMENT

It is important to evaluate cardiac function. This is typically done by echocardiography, which uses sound waves to create a moving picture of the heart.



VIDEO FLUOROSCOPY (SWALLOW TEST)

This in-hospital test is done to evaluate how your mouth and throat function when swallowing food and liquids.

CHEST X-RAY

If you are experiencing new symptoms, your physician may order a chest x-ray to assess the status of your lungs and chest wall.



From left to right: Douglas McKim, MD; Kathy Walker (RRT); Joan Norgren (RRT); Sue Stevens (Admin Assistant); Carole LeBlanc (RRT, Professional Practice Leader); Paula Baxter (RRT).

Missing from picture: Mary Irven (RRT) and Renée Longpré (RRT).

Interview with Douglas McKim MD, FRCPC, FCCP, DABSM

Medical Director of the Ottawa Hospital CANVent Program, Director of the Respiratory Rehabilitation Program at the Ottawa Hospital, and Associate Professor of Medicine at the University of Ottawa

Q: How important is establishing a baseline and ensuring ongoing monitoring?

A: It is very important and unfortunately does not happen routinely. That is why it is critical for patients and family members to know what to look for and ask about. For example, a peak cough flow test result should be approximately 270 litres/minute or 4.5 litres/second. Your health-care provider will interpret your rates based on factors such as age, weight and gender. There is no consensus about the right numbers for other respiratory tests (e.g. maximum inspiratory pressure and maximum expiratory pressure). It depends on one's condition, but monitoring various aspects of lung capacity every six months—or sometimes more frequently—is appropriate in most cases. A test of oxygen (pulse oximetry) overnight is also very important, as respiratory problems often begin during sleep and may not be associated with any symptoms.

Self-monitoring

Nobody knows your breathing better than you. That is why an important part of any monitoring plan is being self-aware. Tell your health-care team if you are experiencing any of the following symptoms, any of which could be a sign of a respiratory problem:

- restlessness while trying to sleep
- anxiousness or shortness of breath when lying down
- feeling unrefreshed in the morning
- recurring chest problems
- morning/daytime headaches
- changes in the amount or colour of the phlegm or mucus that you cough up
- periods of confusion or not making sense when communicating
- increased fatigue, unexplained sleepiness or difficulty waking up
- sudden shortness of breath (note: if you are experiencing significant or severe shortness of breath, seek emergency medical attention)

For a detailed description of the symptoms of weakened breathing muscles and respiratory failure, please refer to Chapter 5.

4 Pulmonary Hygiene Therapies

This chapter describes some exercises and therapies that will help you to maintain good pulmonary hygiene and produce an effective cough, both of which are crucial to your respiratory health.

Before you begin any of these therapies, you must consult an experienced and qualified health-care professional to determine which approach is best for you and to receive personalized training.

Pulmonary hygiene and ventilation therapy: What is the difference?

Pulmonary hygiene therapy is not the same as ventilation therapy. Pulmonary hygiene keeps your airways clear and lungs stretchy. This helps you cough and prevent infection. Ventilation therapy (discussed in detail in Chapters 6–8) helps you get oxygen in (and carbon dioxide out of) your body.

Maintaining good pulmonary hygiene



Jeff Sparks demonstrates breath stacking using a lung volume recruitment device with assistance from his attendant, Chris Cormier

TIP Begin therapies early, when you notice the first signs of a weakening cough.

With respiratory monitoring, your physician or therapist can advise you when this is happening, or you may notice it yourself. **DO NOT WAIT** until your cough is noticeably weak (or non-existent).

Breath stacking

Breath stacking therapy (also known as **lung volume recruitment**) consists of periodically filling your lungs to their maximum capacity, beyond what your weak muscles can achieve on their own. Breath stacking may increase your lung volume, cough effectiveness, lung and chest suppleness and speaking volume. It may also decrease **atelectasis**, which occurs when areas of the lung are not routinely filled with air. Atelectasis can lead to chest infections and poor air exchange in the lungs.

Once prescribed, breath stacking therapy is performed using a lung volume recruitment device, which consists of a manual resuscitation bag, a one-way valve inserted into tubing, and either a mouthpiece or a mask. The lung volume recruitment device (also called a breath stacking device) can be assembled by a knowledgeable health-care professional or purchased as a pre-assembled kit. (Note: the device should be clearly labelled so that it is not confused with a manual resuscitator, as it cannot be used for resuscitation.)

Using a lung volume recruitment (LVR) device

1. This technique is best performed while you are sitting, but it can also be done lying down, with your head slightly elevated. It should be performed at least 60 minutes after a meal.
2. Take a deep breath, then place the mouthpiece in your mouth (or the mask over your nose and mouth) to create a pressure seal. When a mouthpiece is used, a nose clip can also be used to prevent air from escaping.
3. Take another breath as you gently squeeze the resuscitation bag. This will push the air into your lungs.
4. Hold the previous breaths and allow the bag to re-inflate. Squeeze successive breaths into the lungs, the breaths on top of each other, until you feel your lungs are completely full of air. You may feel a stretch in the chest or slight discomfort, but this is normal.
5. Once your lungs are full, release the seal by letting go of the mouthpiece (opening your mouth), by pulling the mask away from your face or by signalling to your attendant to break the seal.
6. Once the seal is broken, you will automatically exhale completely.
7. Pause 30–60 seconds.
8. Repeat the procedure three to five times per session.

Note: A knowledgeable health-care professional **MUST** initiate this therapy and periodically review your technique. Be sure to mention if you have a history of emphysema, spontaneous pneumothorax or heart conditions before beginning this therapy.

If you have limited use of your arms and hands, an attendant can assist you. Be sure to establish a signal with your attendant to indicate when to stop introducing air or to break the pressure seal. Alternatively, adaptations are available for use with a foot pump for people with weak hands.

Research shows that people with Duchenne muscular dystrophy who use LVR twice a day demonstrate a slower decline in pulmonary function.¹

¹ McKim D et al. "Lung volume recruitment slows pulmonary function decline in Duchenne muscular dystrophy," *Archives of Physical Medicine and Rehabilitation*, March 2012, 93(7): 1117–22.

Self-assisted cough

The objective of the **self-assisted cough** is to give your cough a little boost. Begin using this procedure when you notice your cough becoming less effective. An example of this would be difficulty expectorating (bringing up phlegm) with a cough.

For this technique to be effective, you will need to have good muscle strength, the

ability to hold your breath and the capacity to cough (to some degree) without assistance.

Warning: This technique is not recommended if you are pregnant or have had recent abdominal surgery (such as placement of a feeding tube).

Performing a self-assisted cough

1. While sitting comfortably, fold your arms over your abdomen and below your rib cage. Hold one wrist with the other hand at your middle.
2. Take a deep breath and hold it. You can use breath stacking (i.e. breathe in and hold, breathe in again without breathing out, breathe in again) to achieve maximal lung volume. If you have difficulty holding your breath or cannot take a deep breath, use a lung volume recruitment device (LVR) to help inflate your lungs.
3. Hold the breath until you are ready to cough.
4. As you begin to cough, forcefully lunge your upper body forward and downward against your hands. You are aiming to have your hands push inward and upward against your belly. This helps push the diaphragm upward, creating an extra little push to force the air out of your lungs as quickly as possible.

It takes some practice to get this technique right. Do not be afraid to experiment a little to get the best results. It should produce a cough that is strong enough to bring up phlegm from your airway.



If you feel pain or significant discomfort at any point while performing this therapy, stop immediately. Either re-adjust your position or seek professional advice.

Caregiver-assisted cough

If your arms are weak, your caregiver can help you with an assisted cough. The technique is similar to the self-assisted cough, but it uses the assistance of a caregiver to help push the air out of your lungs.

There are two ways of doing this: a **manual abdominal thrust** or a **costal lateral compression**. You may find one technique more beneficial or appropriate than the other.

Performing a manual abdominal thrust

Warning: If you are pregnant or have had recent surgery in the abdominal region, do not use this procedure. Obese individuals may not benefit from this technique.

1. You can be sitting or lying down. Choose the position that is most comfortable and that produces the best results.
2. Fill your lungs with as much air as you can hold (using the breath stacking device, if necessary).
3. With the palm of one hand on your upper abdomen—but well below the bottom of your breastbone and rib cage—your attendant or caregiver will push rapidly inward and upward. This will rapidly expel the air from your lungs.

It often takes more than one manoeuvre to get results. Repeat the procedure two to three times, as necessary and tolerated.

Performing a costal lateral compression

Warning: This technique is not recommended if you have significant curvature of the spine, rib cage injuries, osteoporosis, or if you have had recent chest surgery.

1. You can be sitting or lying down.
2. If you require the LVR device to inhale and hold a deep breath, you will need a second attendant because your first attendant will need both hands for the compression.
3. The attendant positions their hands on either side of your lower rib cage with their thumbs in the front of your chest, pointed toward your breastbone.
4. Fill your lungs with as much air as you can hold (using the breath stacking device, if necessary).
5. Your attendant (with their hands on your rib cage) instructs you to cough while rapidly pushing inward and upward on both sides of your chest.

It often takes more than one manoeuvre to get results. Repeat the procedure two to three times, as necessary and tolerated.



Mechanically assisted cough (mechanical insufflator-exsufflator)

As your respiratory muscles become weaker, you may be unable to bring up phlegm effectively or maintain clear airways and lungs, even with the cough exercises described earlier in this chapter. If you do not have the strength to produce an effective cough, consider using a **mechanical insufflator-exsufflator**. This device quickly inflates the lungs by delivering a deep breath. It then rapidly deflates the lungs with a vacuum, moving the phlegm up the airway into the mouth, where it can be removed.

The marketplace for insufflator-exsufflators is always changing, and different brands and models may be available. These devices are often referred to by the brand name "CoughAssist™" and are manufactured by Philips Respironics (pictured above).

Using a mechanical insufflator-exsufflator is very helpful for pulmonary hygiene because it helps with airway clearance and inflates areas of the lungs that may not otherwise open fully for proper air exchange.

GETTING STARTED

Many people need between two to four weeks before they notice results from a mechanically assisted cough technique. During this transition,

you can continue using non-mechanical cough techniques (as described above).

A health professional must provide training in the mechanically assisted cough technique, and they should also continue to follow your progress to ensure that you are consistently getting good results.

The effective pressures required to inflate and deflate the lungs vary from person to person, but they generally fall between 35–50 cm H₂O. Most people cannot tolerate these pressures when they begin; it is best to start with much lower pressures (10–15 cm H₂O) and work up to an effective therapeutic pressure over a period of time. Lower pressures may be appropriate for younger children (typically 20–40 cm H₂O.)

If you have good control of your arms, you may be able to perform the mechanical insufflation-exsufflation procedure alone, although it may take practice. Those with limited use of their arms and hands will need assistance.

If you have good strength in your facial muscles and good control of your lips, you can try using a mouthpiece. Otherwise, a full-face mask is required to create a pressure seal over your mouth and nose.

Many find that a full-face anaesthesia mask with an inflatable and adjustable air cushion seal works well. You can adjust the amount of air in the seal with a syringe to obtain a good seal. Later on, if your facial contours change with muscle loss, you can make adjustments to the seals.

THE TREATMENT

One cycle of this technique includes

1. the machine pushing air into your lungs (the inhalation phase);
2. the machine forcing the air out with a vacuum (the exhalation phase); and
3. a pause of two to five seconds (as required).

Repeat this cycle three to five times until you bring up phlegm and clear your airway.

Use this treatment daily as a maintenance therapy, increasing the frequency during acute infection or if you are experiencing severe respiratory weakness.

CoughAssist™ and pulmonary hygiene



Van Bernard using a mechanical insufflator-exsufflator with support from his mother, Julie Clegg

Van has spinal muscular atrophy type 2 and has been using a mechanical insufflator-exsufflator to help clear secretions and maintain optimum pulmonary health. Prior to obtaining this device when he was 16 months old, Van was hospitalized 12 times in one year for double lung pneumonia. Since then, however, he has had minimal hospitalizations due to respiratory infections. Van and his family believe that there is a direct correlation between using his CoughAssist™ twice a day and his current healthy respiratory state.

For Van, this vital piece of equipment has literally been a life saver!

USING A MECHANICAL INSUFFLATOR-EXSUFFLATOR WITH A TRACHEOSTOMY

If you have a tracheostomy, you are likely using an aspirator (also known as a suction machine) with an endotracheal catheter to clean your upper airway of mucus. While endotracheal suctioning is effective for clearing out the trachea and upper airway,

it does not move mucus from the lower airways up into the larger airways or trachea. Using a mechanical insufflator-exsufflator, however, will help to move mucus into the larger airways, where it can be suctioned by a catheter. The mechanical insufflator-exsufflator will also promote lung volume recruitment and help prevent lung infection.

If you have tracheostomy cuff, it should be inflated while using the mechanical insufflator-exsufflator to enhance the effectiveness of the therapy.

Please refer to Chapter 8 for more information about tracheostomies and invasive ventilation.

The Home Mechanical Ventilation Guidelines published in 2011 by the Canadian Thoracic Society contains additional details on lung volume recruitment and airway clearance in at-risk and ventilator-assisted individuals. It also offers specific recommendations for people with neuromuscular disorders.

A copy of this document can be obtained from www.respiratoryguidelines.ca or www.muscle.ca.

Will this equipment be covered by my insurance?

Funding for mechanical insufflator-exsufflators, ventilators and other breathing equipment will depend on your provincial health plan and/or insurance coverage.

Muscular Dystrophy Canada helps clients obtain this equipment.

Contact us for more information.

www.muscle.ca
1.866.687.2538

5 Breathing Complications

People affected by progressive neuromuscular disorders often experience weakness in the muscles that are necessary for breathing and coughing (as described in Chapter 2).

Symptoms of weakening breathing muscles gradually appear over time as muscles weaken, and they may include:

- more frequent or pronounced shortness of breath on exertion
- occasional mild shortness of breath while at rest
- shortness of breath while lying on your back
- difficulty sleeping
- waking with shortness of breath or with a headache that goes away after being awake for awhile
- dizziness during or after exertion
- ringing in your ears

Note that shortness of breath may not occur if muscle weakness in the legs or arms prevents you from exercising or engaging in strenuous activity.

Respiratory failure

Progressive weakness in the breathing muscles can lead to **respiratory failure**, a severe condition that can be fatal if untreated. Respiratory failure occurs when gas exchange (the process of taking oxygen into the blood and removing carbon dioxide) in the lungs is significantly impaired. This results in oxygen levels that are significantly lower than normal (hypoxemia). It can also be accompanied by an increase in carbon dioxide levels.

Symptoms of respiratory failure may include:

- sleeping more often
- constant fatigue
- occasional confusion
- difficulty concentrating
- new muscle twitching
- periodic or constant headache
- significant shortness of breath at rest
- problems lying down due to breathlessness
- difficulty sleeping and/or waking
- unconsciousness (in extreme cases)

Once respiratory failure begins, the respiratory muscles try to work harder, and this makes the muscles even more tired.

Depending on your pulmonary reserve (the maximum increase in minute ventilation that you can maintain without exhausting your respiratory muscles), respiratory muscle exhaustion can occur within hours or days. If you experience symptoms of respiratory failure, seek immediate emergency medical attention. **The key to surviving respiratory failure is receiving prompt mechanical ventilation and airway clearance to boost your breathing.** Appropriate clinical assessment and follow-up should identify such difficulties early and prevent any respiratory crises.

Nicolas Reny's respiratory crisis



Nicolas on ventilation in the hospital

In April 2013, Nicolas Reny—a young man with Duchenne muscular dystrophy—caught a simple cold that became worse. While a respirologist prescribed some medication for the cold, he also recommended Nicolas start lung volume recruitment exercises. A respiratory therapist came to Nicolas' house to train him in these respiratory exercises and to conduct a few tests. The therapist noticed that Nicolas' blood oxygen levels were very low, that he was coughing a lot and that the medication hadn't decreased his lung secretions.

On April 12, Nicolas was admitted to Cité de la Santé in Laval. After attempts were made to reduce his cough and lung secretions with intravenous antibiotics and bronchoscopies, Nicolas was transferred to the intensive care unit (ICU). In the ICU, Nicolas was intubated—a flexible plastic tube was passed through his mouth into his trachea in order to maintain an open airway and facilitate ventilation of the lungs using mechanical ventilation. He remained intubated for 12 days. His lung secretions were so abundant that the staff recommended that he undergo a tracheotomy, a surgical procedure that creates an opening for an artificial airway. Nicolas and his whole family were surprised that the situation had become so serious so quickly.

The operation took place at the end of April. At the beginning of May, Nicolas was transferred to the Montreal Chest Institute and enrolled in their ventilator assistance program. The Institute's staff monitored his recovery, prescribing both lung volume recruitment exercises four times a day and nightly use of a bi-level positive airway pressure ventilator. At the end of May, Nicolas' tracheostomy tube was removed; he was back home by the beginning of June to the great delight of his entire family. The Institute has continued to support the family by providing them with equipment, training, at-home care from a respiratory therapist and a 24-hr helpline. Nicolas—who has continued his lung exercises and the use of his bi-level device at night—is enjoying life!!!

Pneumonia

A respiratory infection, even a common cold, can place you at risk of developing pneumonia. If you notice symptoms—such as a change in the colour or consistency of your mucus—see your physician for assessment and possible treatment.

Pneumonia can be viral or bacterial, and antibiotics are typically prescribed to treat bacterial infections.

Left untreated, pneumonia can rapidly lead to respiratory failure if your body cannot sustain the increased work of breathing during acute illness. In this case, you will likely require emergency ventilatory assistance. Seek emergency medical attention if you are experiencing symptoms such as fever, changes in your breathing and/or shortness of breath.

Intensive care

If you develop acute, life-threatening breathing complications that are so severe that you cannot breathe on your own, you will need to receive assistance from a machine to breathe (for more on mechanical

ventilation, see Chapter 6). In order to receive mechanical ventilation, you will be admitted to the intensive care unit (ICU). There you can expect to receive continuous attention and care from doctors, nurses and respiratory therapists.

There is a significant risk of further complications (such as additional infections and blood clots) for any patient who is admitted to the intensive care unit. In particular, your muscles will grow weaker while in the ICU. As a result, it may be more difficult for you to breathe without ventilator assistance, even after you recover from the initial breathing problem.

Some people with muscular dystrophy and similar problems require several weeks on the ventilator before they can breathe without assistance, and some remain permanently ventilated. Many of these people, however, still enjoy life after they leave the ICU, even if they require continued ventilator assistance.



Caution: Oxygen therapy

In the general population, hypoxemia (low levels of oxygen in the blood) is commonly treated with supplemental oxygen. **For people with neuromuscular disorders who are experiencing respiratory difficulty, however, oxygen therapy must be used with great caution. Mechanical ventilation (preferably non-invasive) should be the initial therapy for people with neuromuscular disorders who experience hypoxemia.**

If you are sent to the emergency department with respiratory complications, be prepared to communicate to the health-care team that you have a progressive neuromuscular disorder and that receiving supplemental oxygen *without ventilatory assistance* can worsen the situation.

There may be other confounding conditions (i.e. pneumonia) that necessitate the use of supplemental oxygen *in addition to* mechanical ventilation. The health-care team must be able to closely monitor your carbon dioxide levels (with a blood gas or transcutaneous monitor).

Refer to Chapter 9 for further details.

Being prepared in case of a respiratory crisis

In order to prepare for the possibility of respiratory failure or ICU admission, consider your desired quality of life and goals, and discuss your wishes with a physician who can explain both the types of interventions that may be available to treat respiratory failure and the subsequent consequences of each action.

You may consider such questions as:

- Do you wish to receive non-invasive ventilatory assistance before invasive mechanical ventilation?
- Do you wish to receive mechanical ventilation through an endotracheal tube (artificial airway)?
- Do you wish to be resuscitated (i.e. chest compressions, emergency ventilation, drugs to keep your heart pumping) should your heart stop working?

After carefully considering and discussing your choices, communicate your wishes to your caregivers, family members and health-care providers.

It is also advisable to prepare a legal document that clearly states your wishes for treatment and care, and that appoints someone you trust to make decisions on your behalf should you become unable to do so. For more information, please see Chapter 9.

A note to parents

When your child is stable, find a time to discuss (with family and the child, if appropriate) what is in his or her best interests. Consider what the child would want in the event of a crisis with respect to acute management (resuscitation) and long-term management (e.g. tracheostomy and ventilation). Although these decisions can change over time, it is best to discuss them before a crisis occurs. Families have found it very difficult to process information, communicate with the child and other family members, and make decisions during stressful times.

Prevention tips

Even a common cold or flu can lead to serious respiratory problems, especially if you already have impaired respiratory function. Here are some tips to help you limit your risk of getting a respiratory infection.

Get immunized

- Keep routine immunizations up to date (such as those against polio, measles and mumps).
- Get an annual flu shot.
- Get vaccines and boosters to protect against pneumonia (Pneumovax) and whooping cough.

Limit your exposure to germs

- Try to avoid direct contact with people who have a cold or respiratory infection.
- Ensure that you and your caregivers are practicing proper and regular hand washing.

Take care of your overall health

- Make sure your diet is meeting your nutritional needs and that you are sleeping well. See Chapter 9 for more information about how nutrition affects your breathing.

6 Mechanical Ventilation

Planning for your breathing needs



Luke and Kayla Melchior on their wedding day

It is never too early to think about your breathing needs—don't wait until a health crisis. Luke Melchior has used a non-invasive ventilator for over 20 years. He was able to become accustomed to his ventilator and CoughAssist™ while his pulmonary function numbers were borderline (rather than waiting for symptoms of respiratory failure to appear). Luke strongly recommends starting ventilation before problems occur and explains that a ventilator isn't the "scary device" you think it is. He says he is "living with a ventilator—not dying with one."

Respiratory monitoring (discussed in Chapter 3) is important for determining the progression of your respiratory muscle weakness and the overall status of your breathing. The therapies described in Chapter 4—including LVR and assisted coughing—will help to maintain respiratory function and reduce the risk of acute infection. As respiratory muscles weaken, however, it can become increasingly difficult to maintain adequate minute ventilation or to prevent a respiratory crisis due to an acute infection (as discussed in Chapter 5). If such a situation occurs, your health-care team may prescribe **mechanical ventilation**.

What is mechanical ventilation?

Mechanical ventilation is a method of assisting or replacing your own effort of breathing through mechanical means. Most often, mechanical ventilation will involve a machine called a **ventilator**, a piece of equipment that is designed to provide breathing assistance to individuals who are unable to breathe sufficiently (or at all) on their own.

Many individuals with a progressive neuromuscular disorder begin mechanical ventilation part-time. It is often prescribed for use while you sleep (when breathing is most shallow) to give your respiratory muscles a rest. If your respiratory muscles continue to weaken, you will likely require daytime or continuous ventilator support.

If you develop respiratory failure and choose not to receive any form of mechanical ventilation (because of your values or a personal assessment of your quality of life), you likely will not survive. Research, however, has shown that ventilator users describe themselves as healthy and enjoying a high quality of life. They report that ventilatory assistance enhanced their independence, energy and overall health.¹

In this document and your related reading, you may find that terms like “ventilatory assistance,” “assisted ventilation,” and “mechanical ventilation” are used interchangeably. They all refer to the act of breathing with the support of equipment. You may also see the term “home ventilation.” While this also refers to mechanical ventilation, it specifically means ventilation with support from equipment at home or on the go.

Types of mechanical ventilation

There are two ways of providing ventilation: non-invasively and invasively.

Non-invasive ventilation (NIV) uses a mask or other type of interface to deliver air from a ventilator. This does not require a surgical procedure. Non-invasive ventilation is discussed in greater depth in Chapter 7.

Invasive ventilation delivers air via tubing from a ventilator through a tracheostomy, which is inserted through a surgically created hole in the windpipe (trachea). Invasive ventilation is discussed in greater depth in Chapter 8.

The right kind of ventilation depends on the person and their medical needs. For example, non-invasive ventilation can be an effective means of treatment for some people, while others may be advised to have a tracheostomy because of advanced bulbar palsy (the weakening of muscles used for speaking, chewing and swallowing), severe lung infection or other factors.

Planning and decision making

Even before you develop respiratory symptoms, you should become informed about treatment options and their implications. You

¹ Brooks D et al. “Ventilator user’s perspectives on important elements of health: Related quality of life.” Available online from <http://www.post-polio.org/res/QofLFINALREPORT-Sept2002.pdf>.

have already taken the first step in doing so by reading this document.

Next, you may want to review the information available from reputable organizations (see Appendix C for a list of resources), consult with medical professionals who are experienced in treating patients with neuromuscular disorders, and speak with people who have experience with both invasive and non-invasive ventilation. These sources will give you a range of perspectives on the subject.

This information—and discussions with your health-care team and family members—will help you to explore the plan of care that is right for you. Ultimately, decisions regarding mechanical ventilation need to take into account your medical needs, best practices, and the availability of services and funding, as well as your personal situation, quality of life and wishes.

Even if non-invasive treatment is not initially discussed when you consult with your health-care team, you may wish to learn about this type of ventilation. You can then evaluate potential treatment plans and, if necessary, advocate for the treatment and services that are best suited to your personal situation.

If you decide against receiving ventilatory assistance altogether, ask your physician or health-care team about the palliative care services that are available in your community. Palliative care focuses on maintaining a patient's quality of life through managing pain and symptoms rather than treating the underlying condition. This kind of care can also provide a range of services, including emotional, spiritual and practical support for patients and their families, or assistance with advanced care planning and end of life decision making.

Body positioning

People living with neuromuscular disorders have reported that their body position affects their breathing. The following strategies may help you cope with your breathing difficulties if the symptoms are minimal, mechanical ventilation assistance is unavailable or you choose not to pursue this type of therapy.

- Limit raising your arms for extended periods of time, as this may cause shortness of breath.
- Avoid bending or leaning over, as this also may cause shortness of breath.
- Rest in a semi-sitting position (with your feet up and your torso partially reclined) for comfortable breathing.
- Sleep with several pillows under your torso, head and shoulders, or elevate the head of the bed mechanically or with a wedge. This will help minimize instances when you wake with shortness of breath.

Frog breathing

Frog breathing, the common name for glossopharyngeal breathing (GPB) due to its frog-like gulping technique, can be a life-saving skill if your respiratory muscles are weak. When performed well, GPB can support your breathing needs (in the event of a sudden ventilator failure, for instance) and help clear secretions independently.

The technique involves using the tongue and throat muscles to gulp small volumes of air into the lungs. After each gulp, the glottis (throat) closes, preventing air from escaping. One breath usually consists of six to nine sequential gulps, and up to 20 gulps can provide enough air for you to cough or blow your nose.

Non-invasive & invasive assisted ventilation

The following chart provides a summary of some of the factors related to non-invasive and invasive methods of ventilation.

Factor	Non-invasive	Invasive System
Air exchange	Can be effective even with no respiratory function (if adequate bulbar function is maintained).	Effective even when respiratory and bulbar muscles are non-functional.
Appearance	Depends on type of interface used (e.g. full-face masks are very noticeable; mouthpiece interfaces are less obtrusive).	Some tracheostomy tubes can be camouflaged; others are highly visible.
Coughing	User can cough without assistance and/or use lung volume recruitment, manually assisted coughing or a mechanical insufflator-exsufflator.	User will require coughing assistance and a suction machine to remove secretions from respiratory tract. A mechanical insufflator-exsufflator is also effective.
Infection risk	No increased risk of infection (if device and interface are properly cleaned and maintained)	Increased risk of infection (related to tracheostomy)
Living arrangements and support	User is able to live at home (or in a community setting) with appropriate supports. If family support is unavailable, caregivers/support workers must be hired according to the individual care needs of the patient (which are typically less extensive than those of people with invasive ventilation).	Some users live at home, but if family support is unavailable, caregivers/support workers must be hired according to the individual care needs of the patient (which are typically more extensive than those of people with non-invasive ventilation). For full-time or almost full-time ventilatory assistance, 24-hour attendant care is required. More complex care requirements, costs and other factors may necessitate individuals live in a hospital or long-term care facility that accommodates ventilator users.
Mechanism	Air is delivered through the mouth or nose, and no surgical intervention is required.	Air is delivered through a tracheostomy tube that is inserted through a surgical opening in the windpipe (trachea).
Protection of airway	Effective unless swallowing muscles are too weak to prevent inhalation of liquids or solids, or when the upper airway is obstructed.	A cuffed tracheostomy tube helps to protect the airway from liquids or solids.
Speaking	Minimal interference (depending on interface)	Ability to speak is altered; speaking valves and other devices improve speaking ability
Swallowing	Doesn't interfere with swallowing	May interfere with swallowing



Brandon McCarthy receives non-invasive ventilation using the CANVent Arm (co-developed by respiratory therapists of The Ottawa Hospital Rehabilitation Centre and London Health Sciences)

Interview with Douglas McKim MD, FRCPC, FCCP, DABSM

Medical Director of the Ottawa Hospital CANVent Program, Director of the Respiratory Rehabilitation Program at the Ottawa Hospital, and Associate Professor of Medicine at the University of Ottawa

Q: You and your hospital have a strong track record of choosing non-invasive ventilation (NIV) over tracheostomy and ventilation. Why is that?

A: I am a firm believer that patients should have the choice of what type of ventilation they wish. They should not be made to feel that NIV is not a safe alternative. There are many advantages to NIV, such as less reliance on a caregiver and fewer hospital admissions due to health crises. Patients and families must be firm and clear in their desires, but they also must recognize that this expertise is not widely held and that physicians cannot really be forced to provide a therapy when they are unfamiliar and uncomfortable with it. Strong encouragement should be provided for physicians to seek expert advice on NIV because it is likely to be less complicated than tracheostomy ventilation. While some patients may need a tracheostomy because they do not have the muscle strength or understanding to allow them to use a mouthpiece, it is ultimately the patient's choice. In my experience, patients actually prefer NIV, especially in the early stages of respiratory support, and they find it offers tremendous comfort and satisfaction.

7 Non-invasive Ventilation

Now that the two different types of mechanical ventilation—non-invasive and invasive—have been introduced, this chapter will describe some of the features and devices that are commonly used in non-invasive ventilation.

Getting started with non-invasive ventilation (NIV)

A ventilator provides assistance by boosting your breathing and giving your respiratory muscles a rest. A vigilant respiratory monitoring program will help you and your physician decide the best time to begin ventilation therapy. Commonly, individuals experiencing symptoms of respiratory complications (described in Chapter 5) will start out using a bi-level positive airway pressure device (such as the ResMed VPAP ST, pictured right) or a ventilator at night.

Bi-level positive airway pressure devices

A bi-level positive airway pressure device is a relatively small, quiet machine that creates air pressure and airflow that is in tune with your own breathing. These devices are commonly referred to as a "BiPAP®" (which is actually the brand name of bi-level devices manufactured by Philips Respironics).

A bi-level device is set to deliver inspiratory positive airway pressure (IPAP) when you breathe in, pushing air into the lungs. This is followed by a significantly lower expiratory positive airway pressure (EPAP) that allows you to exhale. The prescribed pressures are determined to give you the boost you need to obtain a normal-sized breath.

An example of bi-level therapy

Joe started bi-level therapy with a prescribed pressure of 14 cm of water pressure on inhalation and 5 cm of water on exhalation. These settings gave him a tidal volume of 500 mls with each breath. After nine months, Joe's respiratory muscles were weaker, and so the inhalation pressure of 14 cm of water pressure only delivered a tidal volume of 425 mls. At this point, he began to experience shortness of breath at rest and trouble sleeping. Joe made an appointment with his respiratory therapist, who assessed his breathing and adjusted the settings on his bi-level device to ensure that he would be receiving the correct tidal volumes.



Monitoring adequate bi-level pressure settings

Bi-level positive airway pressure devices do not guarantee a specific tidal volume. If you require a bigger boost of air, the pressure settings can be adjusted to accommodate your changing needs. For example, if you have a lung infection, the prescribed pressures may become inadequate to deliver

the proper amount of air. As a result, you may require a change to the settings of your bi-level device, or you may be temporarily switched to a more advanced ventilator to ensure your ventilation needs are met.

Keep in mind that symptoms such as a slight shortness of breath or other signs of respiratory complications also may be caused by a leaky interface (mask) or mucus buildup, either of which can prevent you from achieving a full breath under the existing settings of your bi-level device. While you should seek professional help and advice if these symptoms begin to occur, it is also important to be monitored on regular basis (every three months) by an experienced respiratory therapist or clinician. This will ensure you are receiving sufficient ventilatory support.

Bi-level devices generally work well in the earlier stages of ventilatory problems, when ventilatory support is only required for part of the day or night. If you require continuous ventilatory support, however, you will need a device that has advanced features that are currently unavailable on many bi-level devices (although features vary between brands and models).

Ventilators

If you require continuous ventilatory support, you will need a ventilator with features that are more advanced than those found on many bi-level devices. Ventilators are more sophisticated—and more expensive—than bi-level devices. While the exact features will vary between brands and models, many ventilators offer a number of useful features.

- A lightweight internal battery and the ability to auto transfer functions between power sources, an important safety feature for people who require continuous ventilation.

- Different modes or ways of triggering and delivering breaths.
- Superior breath sensing and synchronizing capacity.
- Multiple alarms to warn of a potential or impending problem.

Some ventilators are incompatible with certain interfaces, so be sure to check the manufacturer recommendations when matching an interface with a ventilator.

Non-invasive interfaces

The air from the bi-level device or ventilator travels through tubing that connects to a mask or a mouthpiece interface.

There are many brands and types of interfaces (such as the ResMed Swift™ LT nasal pillows, pictured opposite). The interface is just as important to the delivery of non-invasive ventilation as the machine itself, so be sure to explore your options during the initial set-up of your new bi-level device or ventilator. Try a mask under relatively low pressure for a few minutes to get accustomed to it, then try other interfaces to see which one you like best. The system that works best for you may depend on your facial and neck muscle strength.

For more information about the different types of non-invasive interfaces, please refer to Appendix B.

See Appendix A for troubleshooting tips for non-invasive ventilation.



Use and care of your non-invasive interface

To avoid complications and ensure that your non-invasive ventilation is working efficiently, be sure to take the following steps.

- Clean the interface (mask, nasal pillows or mouthpiece/straw) daily with dish soap and warm water, following the manufacturer's recommendations. If your interface is not cleaned regularly, you may be exposed to bacteria or viruses and potential infection, and you will shorten the life of the mask.
- Only tighten headgear enough to create a seal. This requires instruction and practice, but it is an important step in preventing pressure sores from forming on the bridge of your nose or other areas of your face and head (such as above your ears). If you do develop redness and pain around the bridge of your nose, consult your health-care team. Don't put this off—such a wound can be difficult to heal and may limit the use of certain interfaces.
- Replace your interface when required. Typically, masks and headgear used 24 hours a day will last six to twelve months, but be sure to replace them whenever they begin to fail to create a seal.
- If you begin to notice symptoms of respiratory complications while using non-invasive ventilation, consult your health-care team immediately. You may need to adjust the settings on your ventilator, or you may be experiencing other complications.

8 Invasive Ventilation

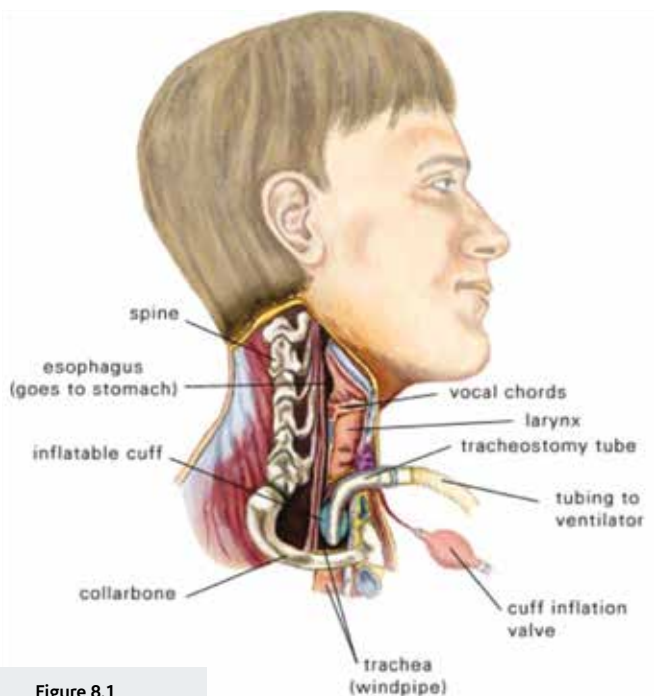


Figure 8.1
Diagram of a tracheostomy

In Chapter 6, we discussed some of the factors that influence how and when assisted ventilation is initiated, while Chapter 7 examined non-invasive ventilation. This chapter will consider the other form of mechanical ventilation: invasive ventilation.

Getting started with invasive ventilation

When beginning assisted ventilation, your medical needs and quality of life should be considered in conjunction with best practices, availability of funding programs and services, and your personal wishes.

There are certain situations in which invasive ventilation may be advised over NIV, including the following:

- you are experiencing severe bulbar dysfunction
- you are unable to clear airway secretions
- you have had an acute deterioration of respiratory function that has required an ICU admission with intubation (life-saving, temporary invasive ventilation)
- you have chosen to proceed with invasive ventilation as the “next step” in escalation of care because 24-hour non-invasive ventilation has failed to provide sufficient ventilatory support

Living with a tracheostomy



Scott Parlee at home

Scott Parlee is 42 years old and lives in Fredericton, New Brunswick, with his mother and father. Scott, who has Duchenne muscular dystrophy, has had a tracheostomy and been using a ventilator for seven years. He has relied on mechanical ventilation ever since a severe respiratory infection landed him in the ICU for two months.

Luckily, his family has been a strong advocate for Scott and his care. When there were questions about whether or not Scott would be able to move out of the ICU and live at home, Scott's father, Allen, made a strong case that the costs of home ventilation should be covered by the provincial government. Not only would home be a happier environment for Scott, but it would actually cost the health-care system less than if Scott had to remain in the hospital.

In addition to the ventilator, Scott now uses a CoughAssist™ mechanical insufflator-exsufflator to help with his breathing and secretion

release. His parents have noticed a tremendous difference in Scott's respiratory health since he began this therapy, and they encourage others with respiratory muscle weakness to learn more about it. They believe that there is not enough awareness about the benefits of the CoughAssist™ device. In fact, Scott and his family have encountered many situations where health-care professionals did not know how to use the CoughAssist™ machine and had to be taught by Scott's father.

Scott and his parents advise others not to be fearful of getting a tracheostomy. They acknowledge that it has taken getting used to and that there is an adjustment period following the surgery and initiation of invasive ventilation. For Scott, however, getting a tracheostomy was life saving, and it has not prevented him from leading a fulfilling and productive life. He travels extensively, and over the past few years, he has attended AC/DC, U2 and KISS concerts.

If you and your health-care team decide to initiate invasive ventilation, you will be booked for a tracheotomy. A tracheotomy is surgical procedure performed under general anaesthesia. A surgeon makes a small incision in the neck to access the trachea (windpipe) and inserts a tracheostomy tube. The tracheostomy tube remains in place to allow the delivery of air from the ventilator. It is also referred to as an artificial airway or a trach (rhymes with “rake”).

If possible, you should engage suppliers and health-care professionals before the surgery; planning that is done in advance will inform your decision about proceeding with a tracheostomy and may shorten your hospital stay. Some individuals, however, are not given the opportunity to plan ahead because they receive a tracheostomy in an acute crisis or emergency situation. Many individuals without sufficient financial or personal resources will be unable to return home as a consequence of the level of care required for invasive ventilation. These individuals may spend significant time in an ICU before potentially moving to a permanent residence in a long-term care facility that accommodates ventilator users.

Getting used to a tracheostomy

If you require ventilatory assistance at the time of your tracheotomy procedure, you will wake up in the recovery room with your tracheostomy tube connected to a ventilator. Once the anaesthesia and other medications wear off, you will recognize that you do not have to do any work to breathe. The ventilator will be delivering all the air you need.

Your health-care team will carefully monitor your progress and make any necessary adjustment to the ventilator settings. In most cases, your home ventilator will be used while you are in hospital to ensure that you are comfortable with the device before you are discharged.

Many people who are admitted to hospital for a tracheotomy and invasive ventilation remain there for several weeks or months. During this time, you and your family members and caregivers will receive training, and your home will be set up with the necessary equipment.

Before you leave the hospital, you and your caregivers should be provided with training and information on how to do the following:

- operate the ventilator
- perform routine, daily tracheostomy care
- respond to a ventilation, tracheostomy or airway emergency
- do suctioning via tracheostomy tube
- troubleshoot and perform daily maintenance and inspections of the ventilator and aspirator (commonly known as a portable suction unit)
- perform manual ventilation with a bag

Receiving a tracheostomy and beginning invasive ventilation at the same time can be overwhelming. It will take time for you and the people assisting you to become comfortable with the procedures needed to operate the equipment and maintain your new daily care routine.

Tracheostomy safety tips

The following precautions may be helpful or necessary, depending on your particular situation or health needs.

- use caution around water—use shower guards
- avoid turtleneck tops and plastic bibs
- use care with gauze padding; it can become an obstruction
- control dust, lint, mold, pet hair and smoke in the home
- drink plenty of water to stay hydrated
- avoid aerosol sprays
- practice infection control by getting immunized, washing hands frequently, proceeding with caution in crowds and screening visitors
- watch for a change in secretions; this may be a sign of infection
- disinfect respiratory equipment with white vinegar diluted with distilled water

Equipment for home

The following list is meant to represent some of the materials that you may need at home. It is not exhaustive, and individual needs may vary.

- spare tracheostomy tubes (same size and smaller sizes)
- suction catheters (pictured below)
- suction machine
- sterile water
- resuscitation bag and face mask—properly sized
- bulb syringes
- heat and moisture exchangers
- compressor for humidification



- tracheostomy collar and tubing
- Q-Tips
- hydrogen peroxide
- gloves
- oxygen source
- pulse oximeter
- ventilator
- emergency tracheostomy kit
- standby or alternate power source
- tweezers and blunt scissors
- tracheostomy wedge and gauze
- normal saline packs

For a list of equipment to take with you when leaving the house, please see Chapter 9.

Resources

Contact Muscular Dystrophy Canada to find out if you are eligible for financial support through our equipment program: **1-866-687-2538** or visit www.muscle.ca

For a directory of resources for ventilator-assisted living, visit www.ventusers.org

9 Other Important Considerations

Appropriate use of oxygen therapy

Mechanical ventilation should be the first course of action to treat respiratory failure. Receiving supplemental oxygen (delivered through nasal prongs or a mask) may cause further complications unless mechanical ventilation is prescribed simultaneously.

The use of supplemental oxygen is appropriate under the following circumstances:

1. You are receiving adequate ventilator assistance to maintain appropriate carbon dioxide levels, but an acute illness or complication causes your oxygen levels to drop below optimal levels. A physician may prescribe supplemental oxygen to be added to your ventilatory assistive device on a temporary basis until the acute illness is corrected.
2. You do not wish to have any ventilatory assistance and are experiencing shortness of breath. Oxygen is administered to help decrease the shortness of breath, usually with other medications. This is considered comfort care at or near the end of life.

Why can oxygen therapy alone be dangerous for people with neuromuscular disorders in respiratory distress? Simply put, adding oxygen will only fix one side of the equation—it will not help you exhale enough carbon dioxide.

In fact, if you receive supplemental oxygen without also receiving mechanical ventilation, your body's oxygen sensors may send a signal to reduce your tidal volume and/or your respiratory rate. This will cause the amount of carbon dioxide in your blood to further increase, possibly to a point where life is unsustainable. It does not happen to

everybody, but there is a significant risk that it will occur in people with progressive neuromuscular disorders.

Unfortunately, there currently is not enough awareness about the risks of oxygen therapy for people with neuromuscular disorders. If you are taken to the hospital, you and your caregivers need to clearly communicate that you have a progressive neuromuscular disorder causing respiratory muscle weakness, and that treatment of supplemental oxygen without proper ventilator support may result in a further rise in your carbon dioxide levels.



Anaesthesia

People with a progressive neuromuscular disorder require special attention and planning when undergoing general anaesthesia. This is because general anaesthesia causes reduced lung capacity in patients after the procedure is complete. This reduction in capacity is temporary (a few days or weeks), but it can be too much for those who have existing respiratory muscle weakness. Be aware that you do not have to have respiratory-related symptoms to be at risk after general anaesthesia. Tell your anaesthetist about your condition so your risk can be properly assessed and contingencies can be put in place.

Some people require temporary ventilatory assistance after general anaesthesia. If you are currently using non-invasive ventilatory support at night or at any point during the day, you likely will require ventilatory assistance immediately following general anaesthesia. Lung volume recruitment and airway clearance are also important following the operation.

Sometimes neuromuscular blocking agents (such as succinylcholine) are used during procedures requiring anaesthesia. People with muscular dystrophy can experience rhabdomyolysis (a rapid breakdown of muscle fibre) and hyperkalemia (high potassium in the bloodstream) following the use of these agents. This can subsequently cause life-threatening cardiac complications. Be sure to discuss this with your anaesthetist before the procedure.

Ventilation equipment and mobility

If you use a wheelchair and require ventilatory assistance, there are options for customizing your chair to make the respiratory equipment available while you are on the go.

Many people who require constant ventilation support have their equipment mounted on



the back of the wheelchair (pictured above). Almost all ventilators and bi-level devices can run off of a 12V battery.

Most ventilators also have internal batteries for a back-up. Bi-level devices, however, generally do not have internal batteries. They must be powered by either a 110V wall source or a 12V battery source. Special proprietary cables are required to obtain power from external battery sources.

Tips for getting your ventilator set up with your wheelchair:

- Have your respiratory therapist and occupational therapist coordinate their efforts and discuss the best way to mount the ventilator, taking into account weight, balance, continued usage and access to the ventilator in recline.
- Do not use the wheelchair battery as an external battery for your ventilator. Best practice requires that a medical system have its own sole source battery. Have a second deep-cycle battery installed in your chair to power the ventilator.

- Consider the size of the battery. Large size or weight may be a limitation to chair mounting. A compromise between ventilator battery life and chair installation may be necessary.
- Have a separate charging system for the battery. Never charge the battery when it is connected to the ventilator.
- a spare tracheostomy tube (and inner cannula, if appropriate)
- heat and moisture exchangers to provide moisture when not using a heated humidifier
- portable aspirator (suction machine) with appropriate attachments
- suction catheters for endotracheal suctioning

Travel

If you are receiving pulmonary hygiene therapy and ventilatory assistance, have a standard set of equipment with you whenever you leave your home. The equipment you carry will depend on your type of ventilation (invasive vs. non-invasive). Your kit may include:

- a breath stacking device
- an extra battery power source for your ventilator
- a spare circuit for extended travel, remote travel or hazardous mobility where there is a risk to the ventilation circuit/tubing

If you have a tracheostomy, you may also need:

- a manual resuscitator and the interface to connect to your tracheostomy tube
- a mask for the resuscitator, in case the tracheostomy tube gets dislodged

TIP

If your mechanical insufflator-exsufflator is stationary at home, consider carrying a portable aspirator (suction machine) and a breath stacking device (as described in Chapter 4). Ensure your attendant is trained in assistive coughing exercises.

Before you travel, consider the following things.

- If you are travelling by air, check with your physician. Cabin air (even in pressurized cabins) has less oxygen when at altitude. While this is generally not a problem, it may become an issue if your respiratory status is impaired and you are not currently receiving support through mechanical ventilation.
- Most airlines have medical desks that require passengers with medical needs to receive pre-approval before they can fly. Your physician will have to complete a questionnaire and send it to the airline's medical department.
- Always have two sources of power (a primary and a back-up) for a ventilator that you depend on for your breathing.
- Always take a travel kit with you and ensure that it is accessible to you or your attendant at all times.
- Verify voltage and power outlet compatibility when traveling outside of mainland North America.
- Talk to your physician about immunization if you are traveling abroad.
- Check with your home-care provider about service centres or companies that can provide you with service at your destination.

Nutrition and hydration

What you eat, how much you eat, what you drink and how much fluid you take in all affect your breathing.

Your muscles require adequate nutrients to work, especially given the muscle weakness caused by your disorder. If you do not take in the correct nutrients or enough of them, your muscles may weaken unnecessarily. This in turn may change your breathing and other muscle functions. Talk to a registered dietician about getting the proper nutrition to maintain your existing muscle strength.

If you have bulbar palsy (see sidebar) and are having difficulty managing your food or swallowing, talk to your physician or dietician to learn ways to prepare your food so that it is easier to manage. People in this situation often find thicker fluids and soft or pureed solids are easiest to consume. Thinner fluids like water are more likely to be aspirated (when secretions or foreign material enter the trachea and lungs) because they move quickly down your throat, and you may be unable to react quickly enough to swallow them properly.

Some people receive their required nutrition via tube feeding, where a tube is implanted through the abdominal wall into your stomach. This procedure may be recommended by your physician if you are having difficulty swallowing, or if you have significant bulbar palsy progression. Sometimes the procedure is advised early in the course of bulbar palsy to reduce the risk of complications.

As long as you have the muscle strength to chew and swallow, a tracheostomy should not prevent you from receiving your nutrition by mouth.

Adequate hydration is essential for both optimal body functioning and pulmonary hygiene. If you become dehydrated, the

mucus in your lungs and airways may thicken, making it more difficult to cough up. This condition could worsen and begin blocking areas of your lungs, decreasing ventilation. Too much hydration, however, may cause significant breathing concerns. Discuss this with your health-care team and be aware of your fluid consumption and elimination (for example, through urination and perspiration).

What is bulbar palsy?

The bulbar muscles are responsible for speaking, chewing and swallowing. They are located in and around the jaw, throat and neck. In some progressive neuromuscular disorders (such as ALS), these muscles can weaken over time. This condition is called bulbar palsy.

Bulbar palsy is more likely to develop in some neuromuscular disorders than in others. Talk to your physician about the possibility of this developing for you.

With progressive bulbar palsy, your speech can become slurred and your ability to cough is reduced. You may experience drooling and saliva management issues that affect your ability to manage food and swallow.

Some people with progressive bulbar weakness experience choking episodes when food or liquid goes "down the wrong pipe" into the trachea (windpipe). Since your cough is ineffective, you may feel a sensation of choking. Most of the time, air will actually continue to move in and out of the lungs, but difficulty coughing up the foreign matter may lead to aspiration pneumonia, which can be life threatening.

If you find you are having difficulty swallowing or managing your food, consult your health-care team about things you can do to minimize the risk of complications.



Speaking and communicating

People considering their options for respiratory care often wonder if they will be able to speak with ventilatory assistance.

The answer to this question is complex, and it depends on your current abilities and the degree of your muscle weakness.

Some people with fairly good control of their bulbar muscles find it difficult to have a conversation because they cannot muster an adequate breath due to weak respiratory muscles. In this circumstance, ventilatory assistance helps by providing a larger exhaled breath, which may strengthen vocal and conversational abilities.

The addition of ventilatory assistance or any other respiratory therapy, however, cannot correct or prevent speech deterioration caused by bulbar palsy or muscle weakness in and around your mouth and vocal cords.

Your speech also may be affected by the type of ventilator interface you use. For example:

- Nasal pillows allow for speech if you have good muscle control of your glottis (muscles in and around your voice box) and the back of your mouth.
- Full-face or hybrid masks allow for speech, but it will be muffled and may be difficult to understand, especially if you have some existing speech impairment.
- With sip or straw interfaces, you can carry on a conversation between sips. You may experience shortness of breath the longer you speak; this is a sign that you should stop speaking momentarily and take a sip to get your breath back.
- If you have a tracheostomy, you will likely be able to speak with the proper tracheostomy tube and specialty valves that attach to the ventilator circuit. People using invasive ventilation tend to speak in cycles with the ventilator. If you have a tracheostomy and are only using invasive ventilation some of the time, you can talk by blocking the air from the tracheostomy tube or by using a special one-way valve.



Legal documents concerning your wishes for care

A personal health-care directive or living will (referred to in some jurisdictions as a Power of Attorney) is a legal document that appoints someone you trust to make decisions on your behalf if you are unable to do so yourself.

This document may contain your instructions regarding treatment and care. For instance, you can indicate whether or not you wish to receive basic life support (CPR) or advanced life support (which generally includes mechanical ventilation). You also may specify the type of medical response you desire—such as ventilation only, medication only or no resuscitation efforts—in the event that you suffer cardiac arrest or respiratory failure. Prior to making these decisions, it is important to consult with a physician who can explain the treatments and the potential consequences of your choices.

The person you have appointed should have a copy of this document, and you should have it readily available to give to first responders and emergency room staff in the event of an emergency.

The laws regarding personal health-care directives vary with each province and territory. Consult a lawyer in your own jurisdiction in order to comply with the relevant legal formalities. Without the appropriate legal documentation, someone may have to apply to a court to be appointed as your legal guardian, which involves time and expense, and in an emergency situation, time is critical.

You can revoke or amend your directives at any time (provided you are mentally competent to do so). It is advisable to review and update your personal health-care directives on a regular basis.

10 Developing a Plan

While the treatment of respiratory issues has improved drastically over the past decades, practices and resources often vary across Canada.

Nonetheless, most clinicians specializing in neuromuscular disorders believe that early and regular monitoring of breathing capacity is crucial to detecting and preventing problems. Being self-aware about your own breathing capacity enables you to establish strategies (such as airway clearance and assisted coughing) to reduce the risk of respiratory failure. It also helps you to make informed decisions about the treatment options that are available to you.

Now that you have some information about how your breathing works, how progressive muscle weakness affects your breathing and what choices you have for respiratory care, take the next steps.

1. Discuss your respiratory care plan with your physician or health-care team. Ask questions related to what you have learned or seek even more information.
2. Share information with your physician. Speak to Muscular Dystrophy Canada if you want additional information and resources, or if you need assistance with your self-advocacy.
3. Investigate the options available to you based on your medical needs, financial situation, family support and available resources.
4. Discuss your options with your loved ones and explain your wishes.
5. Share your decisions with your physician and proceed with developing a respiratory care plan.
6. Ensure your caregivers and family understand your plans and your wishes regarding emergency care.
7. As your disorder progresses, continue to re-evaluate your wishes for care. Make sure your health-care team, family/caregivers and legal documentation are updated accordingly.
8. Remember that you are in the driver's seat—you control your level of care. It is the responsibility of the health-care system to ensure your wishes are respected.





My story

By: Danielle Peers
December 2012

Most of the protocols in this book have never been offered to me. If they have, I have had to fight tooth and nail for them. Over the last six years of dealing with respiratory issues without the benefit of a coordinated neuromuscular health team, I have learned to rely on other strategies of care.

First, I take charge of my own health care. I keep a copy of all my medical records and take them to each appointment.

I read up on findings and protocols. I ask my community

for alternatives. I get second or third opinions. I self-advocate. I make medical choices based on how they fit with my life goals and desires. Muscular dystrophy comes in many types and variations, and people come with different desires, passions, life situations and opportunities. No one can balance the knowledge of my body and my life priorities like I can.

Second, I use strategies that are not offered by my doctors: massage and heat for the recovery of sore respiratory muscles; postural support in the form of backrests, knee support and wedges; adapted restorative yoga for deep breathing and flexibility; medically acknowledged supplements for my immune system (vitamin C & D), for easier lung clearing (n-acetyl cysteine), and for muscle recovery (magnesium). I seek out tools that allow me greater mobility and access with less energy expenditure (such as my Viper motorized wheelchair attachment, courtesy of Muscular Dystrophy Canada's equipment program). I also seek community that supports my care and shares strategies and experiences.

While I (still) struggle for the kind of progressive medical respiratory care outlined in this document, these strategies keep me breathing, working, playing and fighting for the care we all deserve.

Glossary

Airway: The passage that allows air to move to the lungs.

Alveoli: Small air sacs in the lung that give the tissue a honeycomb appearance and expand its surface area for the exchange of oxygen and carbon dioxide.

Aspiration: The drawing of a foreign substance, such as mucus or stomach contents, into the respiratory tract during inhalation.

Aspirator: Also known as a suction unit, this instrument uses suction to remove substances, such as mucus or serum, from a body cavity.

Atelectasis: A condition where alveoli in the lung have collapsed or do not open for air. Parts of the lung that have developed atelectasis do not participate in gas exchange, may be at risk of infection and can contribute to low oxygen levels.

Bi-level positive airway pressure device: A relatively small, quiet machine that provides non-invasive ventilation by creating air pressure and airflow that is in tune with your own breathing. Frequently referred to as a "BiPAP[®]," the brand name of a bi-level device manufactured by Philips Respironics.

Bronchus, Bronchi and Bronchioles: The airways of the lungs. The trachea divides into the left and right bronchus, which further divide into smaller airway passages called the bronchi. As the airways enter the lung tissue, the passages become smaller and are called bronchioles. These terminate in the alveoli, which is where gas exchange occurs.

Bulbar palsy: A condition wherein the bulbar muscles become progressively weaker. The

bulbar muscles are located in the jaw, throat and neck.

cm H₂O: A centimeter of water, a unit of pressure used during mechanical ventilation.

Hypercapnia: High levels of carbon dioxide in the blood.

Hypoxemia: Low levels of oxygen in the blood.

Interface: Mask used to deliver air from a bi-level device or a ventilator.

Living will: See "Power of Attorney."

Manual resuscitation bag: A device that is used to push air into the lungs through a face mask or a connection to a tracheostomy.

Mechanical ventilation (also called ventilatory assistance or assisted ventilation): Used to boost or completely supply ventilation. It includes the use of equipment that supports ventilation by means of volume or positive pressure, either invasively (tracheostomy) or non-invasively (masks, nasal pillows and mouthpieces).

Mid-face hypoplasia: Abnormal growth of the cheek bones resulting in cosmetic abnormalities and orthodontic problems.

Minute ventilation: The amount of air that one must move in and out of the lungs every minute in order to inhale enough oxygen and exhale enough carbon dioxide.

Non-invasive mechanical ventilation: Mechanical ventilation with a non-invasive interface (such as a nasal or face mask) that does not use an artificial airway.

Power of Attorney for personal decisions and medical care: Also called a living will, this is a document authorizing one person to take legal actions related to personal care and medical decisions on behalf of another.

Pulmonary: Anything that affects, occurs within or relates to the lungs.

Pulmonary reserve: The maximum increase in minute ventilation that you can maintain without exhausting your respiratory muscles.

Respiratory failure: A condition in which respiratory function is inadequate to meet the body's needs.

Respiratory rate: The number of breaths per minute that we take. An adult typically breathes 12–20 times per minute. Children breathe faster than adults (although the rate is age-dependent).

Respiratory therapist: A health professional who treats people who have breathing or cardiopulmonary problems.

Respirologist: A medical specialist who diagnoses and treats lung diseases (also known as pulmonologist, a term commonly used in the USA).

Suctioning: The process of removing secretions from the tracheostomy by applying suction through a catheter.

Tidal volume: The amount of air we inhale with each breath.

Trach: Short for "tracheostomy."

Trachea: Also known as windpipe, this cartilaginous and membranous tube descends from the larynx and branches into the left and right main bronchi.

Tracheostomy: A tracheostomy is a surgical opening in the windpipe (trachea), made with a surgical incision below the Adam's apple (below the vocal cords). A tube is placed in the opening, and air goes in and out through the tube instead of through the mouth and nose. For some, a tracheostomy is short term. For others, it is long-lasting or permanent.

Tracheostomy stoma: The opening in the neck where the tracheostomy tube is inserted.

Tracheostomy tube: A curved tube that fits into the tracheostomy stoma. It consists of an outer cannula and a flange that allows for tracheostomy ties to go around the neck to secure the tube in place. Some tracheostomy tubes also have an inner cannula and/or a cuff. There are many styles and sizes available from a variety of manufacturers.

Tracheotomy: A surgical procedure that creates an opening for an artificial airway to maintain an individual's ability to breathe.

Ventilation: The exchange of air between the lungs and the environment. It consists of inhalation and exhalation.

Ventilator: A device for giving artificial respiration or aiding in ventilation.

Appendix A

Troubleshooting and Tips for Non-invasive Ventilation

Getting accustomed to using non-invasive ventilatory support takes time. Work with your respiratory therapist to correct any concerns early, before they develop into larger issues.

The following table describes some common problems and provides suggestions on how to correct them.

Issue	Potential cause and remedies
My nose is congested and stuffed up; I cannot get the air I want.	Increase the humidity heater settings. Congestion is likely the result of nasal irritation caused by a lack of moisture in the inhaled air coming from the machine.
I have the humidity as high as it will go; my nose is still congested.	Check your mask/interface to ensure that it is not leaking. Talk to your health-care team about a nasal anti-inflammatory medication.
The mask/interface is leaking a lot.	There are several things that you can do if your mask or interface is leaking: <ul style="list-style-type: none"> • adjust the headgear (without making it too tight) • replace the headgear and/or mask if it is more than six months old • try a different size, brand or type of interface
My respiratory symptoms have not gone away or have returned weeks or months later.	Have an assessment done by your health-care team. You may need to have the prescribed settings on your ventilator adjusted.
The bridge of my nose is sore.	Your mask is either the wrong size or the headgear is too tight. Try adjusting the headgear, and make sure that the mask is just tight enough to cause a seal (no tighter). It takes experimentation to get it just right.
I have a pressure sore on my nose and cannot wear the mask.	This is a potentially serious problem because you need your ventilation. Try using nasal pillows instead of a mask. Get professional wound care as soon as possible. Do not put anything (including gauze) between the mask and the wound—it only will make it worse. You can try an approved nasal bridge gel pad from your mask supplier to see if that helps. You also need to fix what caused the pressure sore in the first place to avoid future re-occurrences.

Issue	Potential cause and remedy
<p>My sip/straw ventilation is blowing hard all the time.</p>	<p>Some machines are not made to deliver mouthpiece or straw ventilation. Be sure your machine is designed for that use.</p> <p>Ensure you are using an interface that is designed for this purpose and approved for use with your machine.</p> <p>Have an assessment done by your health-care team. You may need to have the prescribed settings on your ventilator adjusted.</p>
<p>Alarms on my sip/straw ventilation are going off all the time.</p>	<p>Ensure you are using an interface that is designed for this purpose and approved for use with your machine.</p> <p>Have an assessment done by your health-care team. You may need to have the prescribed settings on your ventilator adjusted.</p>
<p>When I wake up, I often find that my humidifier has run dry.</p>	<p>You may have a leak somewhere, either at the mask or the tubing. Most often it is the mask interface.</p> <p>To find the leak, have someone watch you while you sleep wearing your interface; talk to your therapist about correcting it.</p> <p>It is also possible that your humidifier may run dry due to low levels of humidity in your home. This is particularly the case in winter months, when the air typically has low levels of moisture.</p>
<p>I wake up with an intense headache that goes away after a few minutes.</p>	<p>You are sleeping with a nasal mask and your mouth is dropping open, or you are using a full-face or hybrid mask that has a large leak.</p> <p>Try a full-face mask or mouth/nasal hybrid mask that fits well with minimal leak. If you are using a nasal mask, try a chin strap to keep your mouth closed.</p>
<p>I can only keep the mask on for a minute or two. Then I rip it off, gasping for air.</p>	<p>If you are looking for more air, the pressure settings may be too low.</p> <p>If you cannot tolerate the high pressure, start getting used to lower pressure settings or reduce the flow settings that may be pushing the air too fast.</p> <p>Tell your therapist about this and ask them to have you try a few settings to see what you prefer.</p>

Appendix B

Types of Non-invasive Interfaces



FULL-FACE MASKS

Full-face masks are built to cover the nose and mouth to create a pressure seal. The mask is held in place with headgear and straps.

Advantages:

- Provides a good option for people who have significant bulbar or facial muscle weakness.
- Eliminates the need to learn to breathe through the nose only.
- Works well during sleep (when the lower jaw relaxes). Using a full-face or hybrid nasal/mouth mask at night will prevent or minimize mouth leak.

Disadvantages:

- Covers large portion of the face, muffling the voice (making speaking difficult). Some people may find it hot and cumbersome.
- May result in pressure sores on the bridge of the nose during extended periods of use. This can be minimized with proper fitting mask and headgear.
- A person with no upper extremity function cannot remove the mask voluntarily. If vomiting occurs and the mask cannot be removed quickly, there is a risk of aspiration.



NASAL MASKS

The nasal mask is similar to the full-face mask, except that it only seals over the nose. It is held in place with headgear similar to that of the full-face mask.

Advantages:

- Less cumbersome and obtrusive than a full-face mask.
- Lighter than a full-face mask.
- With some experimentation and practice, you can speak while using this mask.

Disadvantages:

- Cannot be used during sleep by some people with weak facial muscles due to mouth leak (although this may be resolved by using a chin strap).
- May cause pressure sores on the bridge of the nose and above the ears if not properly fitted or if the headgear is too tight.



NASAL PILLOW SYSTEMS

Nasal pillow systems have two small, unobtrusive pillows that fit into and seal the nostrils. When starting out using non-invasive ventilation periodically throughout the day, some people like to use the nasal pillow system because it can be a break from having to use the full-face or nasal mask. Like the other interfaces, there can be variations between brands.

Advantages:

- Lightweight and unobtrusive compared to a full-face or nasal mask.
- With some experimentation and enough muscle strength, you can speak with relative ease.
- Your face will not be as warm as with a full-face or nasal mask.
- No risk of pressure sores on the bridge of the nose.
- Sight is easier since the nasal pillow system causes fewer focus problems than a full-face or nasal mask.

Disadvantages:

- Cannot be used during sleep by some people with weak facial muscles due to mouth leak (although this may be resolved by using a chin strap).

- Extended use over months and years may cause nostrils to stretch in size.
- Requires fairly good muscle strength in the lower jaw, upper airway and facial muscles to keep the mouth closed during inspiration.



HYBRID NASAL/MOUTH SYSTEMS

Some manufacturers have a hybrid mask that combines a nasal pillow system with a mouth seal system. Some individuals prefer using this over a full-face mask while sleeping.

Advantages:

- Delivers pressure through both nose and mouth, so it has the same effect as a full-face mask.
- Can be used while sleeping.
- Provides no pressure on the bridge of the nose.
- Does not generate as much heat around the face as a full-face mask.
- Sight is easier since the nasal pillow system causes fewer focus problems than a full-face or nasal mask.

Disadvantages:

- May be difficult to maintain a seal around the mouth at higher pressures, depending on your face shape.

- May cause stretching of the nostrils if used extensively over months and years.
- Speaking is difficult while using the device.
- Not currently available for pediatric use.



SIP/STRAW OR MOUTHPIECE INTERFACE

This mouthpiece-shaped or straw-shaped device is mounted on the wheelchair in a position that is easily accessible by mouth. When you feel like you need a boost to your breathing, you place your mouth and lips around the mouthpiece, creating a pressure seal. After one or more assisted breaths, you release the seal and carry on with whatever you were doing.

In order to use this interface successfully, you need strength in the muscles of your face and lips. People with any significant bulbar weakness usually cannot use this interface. You must also have strength in your neck muscles to move your head to and from the interface.

This type of interface cannot generally be used with simple bi-level devices because the airflow will continuously blow while not taking a breath. It works best with a home ventilator system or an advanced bi-level device that has volume cycling. Consider selecting a ventilator that has settings for day and night.

Advantages:

- Provides ventilation whenever required, allowing the user to take a breath as needed.
- Does not require a mask to be held against your face.
- Allows daily activity, such as speaking, eating, etc.
- Eliminates risk of pressure sores on the bridge of the nose.
- Allows the patient to conduct lung volume recruitment independently and whenever desired.

Disadvantages:

- Requires good muscle control around the lips, face and neck.
- Needs to be connected to a home ventilator or advanced bi-level device with volume cycles.
- Cannot be used while sleeping.

Children who use masks over a longer period of time may be at risk of abnormal growth of the cheek bones (mid-face hypoplasia) that can result in cosmetic abnormalities and orthodontic problems. The risk of mid-face hypoplasia can be minimized by switching between different types of masks to avoid repeated pressure in the same area.

Appendix C

Additional Information & Resources

Muscular Dystrophy Canada • www.muscle.ca

We are a hub of information about respiratory care for people affected by neuromuscular disorders. Visit our website to find links to publications, standards of care and clinical practice guidelines, such as:

Birnkrant DJ *et al.* "The respiratory management of patients with Duchenne muscular dystrophy: A DMD care considerations working group specialty article." *Pediatric Pulmonology*, August 2010, 45(8): 739–48.

Canadian Thoracic Society. *Home mechanical ventilation: A Canadian Thoracic Society clinical practice guideline*. For detailed information for people with DMD, see pages 109–124; for other muscular dystrophies and myopathies, see pages 125–131; and for myotonic dystrophy, see pages 132–137.

Cure CMD. *The management of congenital muscular dystrophy: A guide for families*. 2013.

Finder JD *et al.* "Respiratory care of the patient with Duchenne muscular dystrophy: American Thoracic Society consensus statement." *American Journal of Respiratory Critical Care Medicine*, August 2004, 170(4): 456–65

Hull J *et al.* "British Thoracic Society guidelines for respiratory management of children with neuromuscular weakness." *Thorax International Journal of Respiratory Medicine*, 2012, 68(Suppl. 1): i1–i40.

Wang CH *et al.* "Consensus statement on standard of care in spinal muscular atrophy." *Journal of Child Neurology*, August 2007, 22(8): 1027–1049.

If you need further information or support, please call us toll-free and ask to speak with a member of our services team: 1-866-687-2538.

Provincial, National and International Websites

BC Association for Individualized Technology and Supports for People with Disabilities
www.bcits.org/

This website includes a learning centre where you can find self-help resources, training materials and explanatory videos.

Center for Noninvasive Mechanical Ventilation, University Hospital, Newark, New Jersey
<http://www.theuniversityhospital.com/ventilation/index.shtml>

This centre consists of physicians and other health-care professionals dedicated to the care of patients with neuromuscular weakness and respiratory impairment from any cause. Led by Dr. John Bach, the Center provides real hope for patients who are thought to be without any options through conventional management.

College of Respiratory Therapists of Ontario • www.crto.on.ca

The website of the organization that regulates the respiratory therapy profession in Ontario. On this site you will find information on emergency planning, customer service standards and practice guidelines for respiratory therapists. It also has an extensive resources section that provides fact sheets, e-learning modules and webinars that are of interest to both health-care professionals and caregivers.

Dr. John Bach • www.doctorbach.com

Dr. Bach is internationally recognized for his ground-breaking work in non-invasive mechanical ventilation. The author of over 250 publications, including seven books on neuromuscular pulmonary rehabilitation and non-invasive mechanical ventilation.

Institute for Rehabilitation Research and Development at The Rehabilitation Centre (Ottawa)
www.irrd.ca

This website contains online education modules for ventilation and respiratory care, including protocols specifically designed for neuromuscular disorders.

International Ventilator Users Network • www.ventusers.org/

This site contains resources, tools and information for people using ventilators. The “Take Charge, Not Changes” kit, developed specifically for people with neuromuscular conditions, includes emergency checklists and other documents that can be downloaded and customized to reflect your own needs and situation.

Muscular Dystrophy Association (USA) • mda.org

Muscular Dystrophy Campaign (UK) • muscular-dystrophy.org

West Park HealthCare Centre Long-Term Ventilation Centre of Excellence • www.ltvcoe.com

West Park HealthCare Centre provides information and online e-learning modules about long-term ventilation for individuals and their caregivers.

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For more information about neuromuscular disorders, as well as our programs and services, please visit www.muscle.ca or call us at 1-866-687-2538.