

While MND does not impair the lungs themselves the muscles involved in breathing and coughing may become weakened over time. Breathing may be significantly impaired without complaints of breathlessness due to the reduction of physically demanding activities imposed by MND. Impairments to breathing may not be detected until there is a lung infection, most probably occurring because of limited breathing ability.

This factsheet will explain the breathing process and provide overviews of symptoms of changes in breathing, tests that may be used to monitor breathing function (so you know what to expect), respiratory infections, airway management techniques, and options for decision you might want to make for the future. If your breathing is already affected by your MND you might also want to look at factsheet number 40 "The Use of Ventilation in MND," after you have read this one. Number 40 describes how your breathing can be supported, particularly overnight, something you might want to think about before it becomes a necessity.

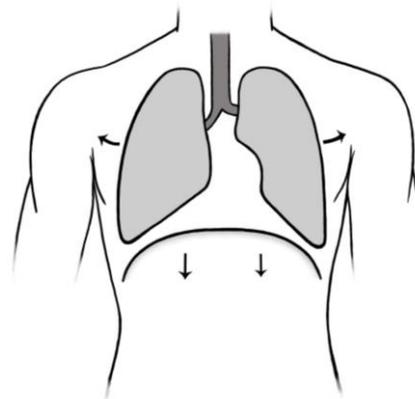
Poor breathing (respiratory) function leaves a person with MND fatigued, short of breath, and more prone to respiratory infections and, possibly, respiratory failure. Therefore, it is a vital part of the MND disease management plan to monitor breathing function throughout the course of the disease.

However, how and when to address respiratory issues in MND remains debatable and is practised differently

across the world. Methods used to monitor lung function also vary across the world. Nonetheless, most MND clinical specialists believe regular monitoring promotes early detection of breathing problems, prevention of unnecessary emergencies, and opens the door for you to consider whether or not you want to go down certain roads.

THE MECHANICS OF BREATHING

Normal "easy" breathing involves two main muscle groups. When you breathe in, a large dome-shaped muscle that separates your heart and lungs from your stomach, called the diaphragm, moves down; at the same time the muscles between your ribs contract to pull your rib cage up and out.



These two actions cause a partial vacuum in your chest cavity. Fresh air rushes down the windpipe into your lungs, inflating them to fill your chest and at the same time pass fresh oxygen (O₂) to the blood. This process is called inspiration.

MND Scotland is the working name of the Scottish Motor Neurone Disease Association, the only charity funding research and providing care and information for those affected by MND in Scotland.

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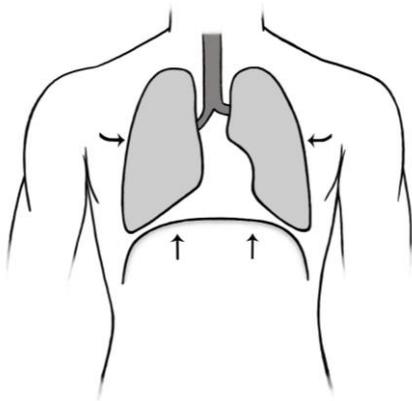
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When you breathe out, both diaphragm and rib muscles relax, and the lung recoils decreasing the size of your chest cavity. Used air that contains waste carbon dioxide from your bloodstream (CO₂) is pushed out of your lungs. This process is called expiration.

If you are breathing heavily, two



additional muscle groups come into play: when you take in 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, your abdominal muscles help to push up the diaphragm.

Many of the muscles involved in breathing can be under conscious control and are called “voluntary” muscles. Since MND is a disease that causes muscle atrophy (wasting) and weakness, it is easy to imagine how changes in breathing can occur, even though your lung tissue may be very healthy. People with pre-existing lung disease may have their breathing abilities much more affected by the weaknesses MND can bring.

SIGNS AND SYMPTOMS OF WEAKENED BREATHING MUSCLES

Shortness of Breath with Activity

Shortness of breath may be the first symptom of the weakening of breathing muscles. You may notice shortness of breath after rushing upstairs, carrying a load, etc. Shortness of breath can also occur with no particular exertion; you may not be able to walk or talk for long without becoming breathless. You should tell your doctor about these symptoms. When you experience breathlessness, stop what you are doing as the symptoms will disappear when activity is reduced.

If you are able, relax in one of the following positions and breathe slowly and deeply.

1. Sit at a table, lean forward with a straight back and rest your head and shoulders on a pillow on the table.
2. Sit on a chair or the edge of your bed, lean forward, keep your back straight and rest your elbows and forearms on your thighs.
3. Lean against a wall with your feet about 12 inches from the wall. Rest your lower back against the wall and lean your upper back away from the wall.

Fatigue

Fatigue, or tiredness, is a common symptom of MND. It is caused by a number of factors. As MND attacks your motor neurons, they become increasingly less able to send commands from your brain to the muscle cells that they control. A smaller number of muscle cells must then try to perform jobs usually done by the full number. The result is that your muscles tire before they normally would.

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When your respiratory muscles are affected by MND, you may be less able to fill your lungs as you were once able to do. In turn this makes it difficult to clear CO₂ from the bottom of your lungs and O₂ uptake may also be reduced. When activity increases, it becomes more difficult for the lungs to supply enough oxygen to the body causing other metabolic changes to take place so making you feel fatigued.

Besides the physiological changes that may cause fatigue, MND can produce many other changes in your life. Change often results in general stress that can also manifest itself as fatigue. Do what you can to prevent getting tired. Rest when you begin to feel tired. Try to keep your exertions within the limits that your body is now imposing. Pay attention to signals like fatigue. It may be necessary for you to change your priorities and forego less important activities. Plan your day to include regular rest periods.

Morning Fatigue or Headache

Some people with MND experience morning fatigue. You may wake up feeling tired, sometimes with a headache, and the feeling that you slept poorly. This may be caused by what is known as *obstructive sleep apnoea* and/or *under-ventilation*.

Obstructive sleep apnoea: Weakened (bulbar) muscles of the upper throat and pharynx may over-relax result in noisy breathing, snoring, or closing of the upper airway while sleeping. This is what is called sleep apnoea and is characterised by intermittent reduction or stopping of breathing. A decrease in the oxygen (O₂) saturation of the blood can result. Sleep apnoea can also put strain on your heart as you struggle to take in air through the closed airway. The body's reaction to low

blood oxygen is to cause you to waken, resulting in disturbed sleep patterns and daytime tiredness.

Hypoventilation: The relaxation of nerve and muscle functions during sleep may lead to under-ventilation (hypoventilation) causing carbon dioxide (CO₂) levels to rise.

For example, when you stand upright, the diaphragm moves down when you breathe in; but when you lie down, the organs in your abdomen press against the diaphragm pushing it up into your chest, and more strength is required for the diaphragm to be pressed down during breathing.

To help cope with this problem, you may want to try to raise your head and shoulders during sleep. It is best to raise the whole torso by using blocks 4" to 6" high under the head of your bed, a wedge pillow 6-8" at its highest point, or a motorised adjustable bed. If you do not have access to the best solutions, try putting two or more pillows under your head and shoulders until you do.

Obstructive sleep apnoea and under-ventilation often occur together; therefore a sleep study in which your blood oxygen levels are measured, (nocturnal oximetry) may be advised (see Monitoring Your Breathing Function). Blood that is rich in oxygen is bright red whereas blood that is low in oxygen is dark red. The precise colour of your blood is related to the amount of oxygen it contains. A device like the one used to measure your pulse in hospital is clasped to your finger tip before you go to sleep and it logs the changing colour of your blood at intervals throughout the night. The level of oxygen in your blood is then determined.

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Excess Mucus and Secretions

In some people, excess mucus and secretions can build up to the point where they cannot be easily cleared by coughing. This is particularly the case first thing in the morning when secretions have built up during the night. If you experience this problem, ask your doctor for a referral to a physiotherapist or respiratory therapist to teach you methods to loosen secretions and bring them to the mouth to be spat out.

One method of dealing with mucus and saliva build-up is suctioning. Suction equipment acts very much like a dentist's suction tube; it consists of a motor, a collection bottle, tubing and a mouthpiece to gather the saliva. However, some healthcare professionals believe suctioning may be counter-productive for persons with MND.

Talk to your MND Care Team Specialist or a respiratory specialist about whether suctioning may be right for you, and if there is anything else you can do to reduce mucus build-up, such as assisted cough techniques (see **Assistive Cough Techniques** later in this factsheet) or the use of medications.

Weak Cough

During a cough, the diaphragm and additional muscles draw air in to completely fill the lungs. Then, the abdominal muscles and intercostal muscles between the ribs contract quickly generating a high pressure against a glottis (larynx) that is kept closed by force. The glottis is then suddenly opened and a very high speed flow of air is pushed from the lungs and up the airway. Any mucus or food in one of the air passages is forced out as well.

People with MND have normal cough reflexes, but the muscles involved may be weakened and unable to produce a strong enough cough. You can strengthen your cough using techniques mentioned later in this section and described in more detail on the web site of The Ottawa Rehabilitation Centre at www.irrd.ca/education/slide.asp?RefName=e2r3&slideid=55 . You will need to consult with a skilled professional to train you and your family.

A Quick List of Signs and Symptoms of Breathing Problems

- Paleness
- Bluish colour to finger tips (cyanosis)
- Contraction of neck and other muscles to breathe
- Confusion
- Inappropriate sleepiness
- Inadequate cough

There are a variety of methods to choose from to monitor the breathing function of a person with MND. Physicians vary in their approach, so keep that in mind as you read this section. If you have not had your breathing function monitored you might want to use this factsheet as the starting point for a discussion with your doctor.

Since MND is a progressive disease, it is helpful to assess lung function early on in the disease, so that there is a baseline *pulmonary function test* (PFT) result, or starting point, with which follow-up assessments can be compared. Without monitoring, a person with MND might seemingly experience sudden respiratory failure without having had the benefit of advance planning regarding ventilation options. Plus, with use of appropriate

airway management strategies, quality of life can be so much better. Signs and symptoms of weakness in the breathing muscles can be subtle and missed if not specifically looked for.

Repeating PFTs every three to six months may be advised to assess any decline in the respiratory system and to guide your doctor about when to discuss certain interventions such as airway management techniques, or non-invasive ventilation in as timely a manner as possible. The most common breathing support is known as the BiPap™.

The following are some of the measurements used to assess lung function.

Forced Vital Capacity (FVC)

FVC is the volume of air that can be maximally, forcefully exhaled following maximal inspiratory effort. The test generally involves having you sit with your trunk elevated to between 30 to 90 degrees, putting a mouthpiece in your mouth, a nose clip on your nose, and breathing into a spirometer that may either be a simple hand held unit or a computerised machine. After taking as deep a breath as you can, you blow out quickly and as hard as you can for one-to-two seconds. If your lips are weak, you will be provided a lip seal mouthpiece or mask to promote an accurate measurement.

Peak Expiratory Flow Rate (PEFR)

PEFR is the highest rate of expiratory (out) airflow you can generate (peak expiratory flow) following maximal inspiratory (in) effort. Flow rate can be measured at the same time as FVC using spirometry with flow volume loops. This is closely related to Peak Cough Flow (PCF) which tends to be slightly greater.

PCF can also be measured at home using an inexpensive hand held peak flow meter commonly used by people with asthma.

Normal PCF values would be 360 to 720 Litres per minute (L/min). The minimum effective PCF is 180 to 200 L/min (recognising that some smaller individuals may manage with lower values). PCF is highly dependent on the function of those muscles in the mouth, throat and larynx (bulbar muscles/bulbar function). The greater the loss of function the lower these measures become.

Maximum Inspiratory Force (MIF)/ Maximum Expiratory Force (MEF)

Maximum inspiratory force (MIF) or maximum inspiratory pressure (MIP) is the greatest force generated by the muscles of inspiration (breathing in); this force comes mainly from the diaphragm, but is contributed to by the other breathing muscles.

A pressure measuring device (manometer) is attached to a mouth-piece and the individual is instructed to breathe in as forcefully as possible without using the cheeks (the cheek muscles can falsely indicate a much higher pressure).

Maximum expiratory force (MEF) or maximum expiratory pressure (MEP) is the greatest force generated by the muscles of expiration (breathing out), primarily the abdominal muscles. These too may be much weakened in MND. These muscles and their pressure measurement are important in the prediction of an adequate cough. Remember, an adequate cough helps to clear airways. Maximum pressures of less than 40 - 50 cm H₂O are of some concern as they may be a sign of less

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respiratory reserve and some limitation of ventilation or cough ability.

Arterial Blood Oxygen Saturation (SaO₂)

A non-invasive way to measure oxygen saturation is to use an oximeter device which involves attaching small sensors to a pulse point on a person's body (e.g., finger tip) to transmit oxygen saturation readings. Normal levels of oxygen saturation are above 95%. Lower than expected levels of oxygen saturation in the blood (SaO₂) may indicate areas of loss of volume in the lungs (atelectasis) or when associated with illness may indicate pneumonia. Less commonly a low saturation can indicate that not enough CO₂ is being cleared (hypoventilation). Hypoventilation often begins, or is worse during sleep.

Nocturnal oximetry, is an over-night assessment used as an alternative to a full sleep study (polysomnogram) when problems associated with night-time (nocturnal) apnoeas (short periods of time when breathing stops), or hypoventilation are evident. Sleep studies, conducted over-night in a sleep laboratory, are not commonly performed in MND management, but in some cases of suspected obstructive sleep apnoea and nocturnal hypoventilation they may be advised.

COMPLICATIONS OF RESPIRATORY INFECTION

Common respiratory illnesses can cause complications in persons with MND. To reduce your odds of infection, try to avoid contact with people who have colds or flu and to practise regular hand-washing.

Bronchitis

Bronchitis is an inflammation and irritation of the bronchial tubes in the lungs

characterised by a cough. This condition may be caused by viruses, bacteria, environmental irritants such as cigarette smoke, or allergies. In response to the inflammation, airways secrete a sticky mucous. If a person is unable to produce an effective cough to clear the lungs, there is a risk of developing pneumonia.

If you do get a fever with thick discoloured or bloody mucus coughed from the chest, or if you notice that the mucus has changed from clear white to yellowish, tell your doctor immediately. These could be symptoms of a respiratory infection that could worsen if your breathing is impaired.

Pneumonia

Pneumonia can be caused by bacteria or viruses, and may be a complication of a cold, flu, bronchitis, or the aspiration (breathing in) of food or liquid. Pneumonia causes the very small air sacs in the lungs (alveoli) and surrounding tissues to fill with inflammatory cells (pus) and mucus which prevents the normal exchange of O₂ and CO₂. When this occurs oxygen is not taken up sufficiently by the blood and a low oxygen level may result.

Aspiration

Aspiration occurs when liquids or solids get into the airways blocking the airflow and preventing gas exchanges (O₂/CO₂). Aspiration can also cause irritation, and might carry infection from the mouth and gums into the lungs. Small amounts of aspirated material can be coughed out.

If aspiration is frequent and involves large amounts of material, an evaluation by a speech and language therapist and/or a respiratory consultant is advised. Changes in nutrition and swallowing techniques may be recommended.

STAYING HEALTHY: PREVENTATIVE AIRWAY MANAGEMENT STRATEGIES

Preventative airway management measures initiated at an early stage can prevent unnecessary hospital emergency visits and intensive care unit admissions. Airway management strategies are used to either improve your ability to take air into your lungs or cough effectively. Strategies range from self-care techniques to the use of assistive devices. Using airway management strategies can give better lung function for longer, and so promote quality of life.

Lung Volume Recruitment (LVR) and Assisted Coughing Strategies

Before trying any assistive breathing and coughing techniques, make sure to discuss them with your healthcare professionals. Some exercises and techniques are more complex than others and require training and practice

For more information about many of these techniques talk to a respiratory therapist, respirologist, or physiotherapist. Baseline pulmonary function and a respiratory assessment are recommended if these haven't already been carried out.

Deep Breathing Exercise

Exercises to use full lung capacity can compensate for weakened muscles. This can be achieved with the following deep breathing exercise:

1. Sit at a table, hunched over slightly with your weight supported on your hands or elbows. This position expands the rib cage for larger breaths by allowing your shoulder and neck muscles to assist your breathing.
2. Breathe in as deeply as possible, hold

the breath for a few seconds, and then exhale.

3. Repeat several times.

Some doctors believe that patients who do deep breathing exercises are less prone to lung infections and partial lung collapse. Ask your physiotherapist to demonstrate an exercise routine and advise you about the use of exercise devices.

Assisted Cough Techniques Without Devices

Assisted coughing is a technique where a carer applies forceful pressure to the abdomen and occasionally the chest wall, timed to the effort of coughing. This action assists the weakened muscles responsible for an effective cough. These techniques are very helpful, effective and easily learned. They are very empowering and relieving for carers who, without them, feel powerless to assist their loved ones in their respiratory distress.

1. **Abdominal thrust:** Immediately prior to a timed cough the carer applies rapid and forceful pressure to the upper abdomen in an inward and upward fashion between the bottom of the breast bone and the navel (see 4).
2. **Lateral costal compression:** Sometimes combined with the abdominal thrust, or used alone when a PEG tube has recently been introduced. Firm but not too forceful pressure is applied by each hand on the lower rib cage below the breasts or at the sides of the chest timed to a cough effort.
3. **Self-assist:** This is most appropriate for paraplegic patients and would rarely be used by MND patients. The

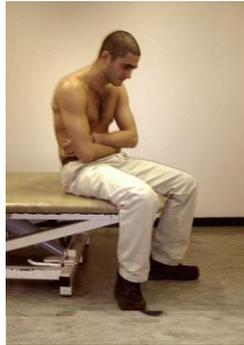
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individual in a stable wheelchair takes a full breath in, folds their arms across their abdomen and forcefully pitches their torso forward over their crossed arms. Once a significant pressure has been generated in the abdomen and chest they release the glottis and cough.

4. **Phlegm management:** With reduced respiratory muscle strength the normal movement of phlegm out of the lungs is reduced and this appears to be made worse by a lack of mobility. An inability to clear phlegm is distressing and increases the risk of chest infection. Advice for patients and carers on the technique of an assisted cough to help bring up phlegm can be sought from the physiotherapist. Treatment of chest infections with antibiotics may be appropriate.



In: the patient locks their arms across their body underneath the rib cage.



Out: the patient uses their arms to support the stomach muscles, bringing their arms into their body in time with the cough out.



(Photographs and description courtesy of King's College Hospital, London.)

Assistive Cough Techniques With Devices

1. **Modified resuscitation bag:** A special hand-held bag is usually used to maintain breath movements in a respiratory or cardiac arrest. This useful tool can be modified with the addition of one-way valve tubing and a mouth-piece (or mask) to allow the passive introduction (by yourself or a carer) of volumes of air into the lungs. This helps you regain breath sizes which have previously been lost due to muscle weakness. Its effectiveness is directly dependent on bulbar function. Once a larger volume has been taken in then an abdominal thrust (as above) can be added to enhance the effectiveness of the cough.
2. **"CoughAssist" (Mechanical in-exsufflation machine):** This is a very effective device for those who maintain sufficient bulbar function but have a weak cough. It is a vacuum that allows attachment through a face mask to both the positive pressure side (to help inflate the lungs) and the negative pressure side of the vacuum (to fill up the lungs and then forcefully empty them to mimic an effective cough). The high speed expiratory flow is effective in clearing airway secretions. Considerable experience is required in the assessment and introduction of this device and it is quite costly.
3. **Volume ventilator:** For people who are already using a volume ventilator for mouth-piece ventilation, the volume of an assisted breath can be

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held by the glottis and added to the next machine delivered breath achieving a breath-stacking effect. This improves both the volume (range of motion) of the lungs (and thorax) and the effectiveness of the cough. Again this is most effective with maintained bulbar function.

Lifestyle Strategies

Do not smoke

In addition to all of the other problems smoking can cause, it can reduce lung capacity and decrease the oxygen carrying capacity of the blood. A first cigarette of the day reduces the oxygen carrying capacity of the blood as carbon monoxide from the cigarette locks on to the same places where oxygen should be. However, carbon monoxide locks on tighter than oxygen and stays there for hours, so every cigarette you take thereafter further reduces your blood's capacity to carry Oxygen even further. Smoking can also cause increased phlegm that some patients can have difficulty clearing from their airways.

Be cautious with alcohol and sedatives

Alcohol and sedatives such as tranquilisers, anti-depressants and sleeping pills taken together can have what is called a "Synergistic effect" i.e. if alcohol has an effect of "2 units" and a sedative has an effect of "2 units," when they are taken together they might have an effect of "8 units" rather than the expected "4 units" and may consequently increase the risk of aspiration or hypoventilation during sleep.

Avoid allergens

If you are allergic to fur, feathers or dust mites, make sure your home is free of pets that cause you problems and is

regularly dusted. You might find hard surfaces such as laminate flooring or linoleum both reduce the number of allergens in a room and make it easier to negotiate if you are using a wheelchair when compared to fitted carpets.

Investing in an air purifier with a HEPA filter can help keep the air free of particles that irritate your airways.

Where to now?

This factsheet has been concerned with maintaining, and hopefully improving, breathing when the respiratory (breathing) muscles have been affected by MND. If these muscles continue to deteriorate breathing may become increasingly difficult, particularly while sleeping.

It is worthwhile emphasising that people with MND who waken with a headache, are tired during the day, have difficulty concentrating, are often confused or are difficult to rouse from sleep could well be suffering from breathing difficulties while asleep.

However, these symptoms could also be due to medications being taken to alleviate other symptoms of MND, so it is worthwhile discussing these symptoms and your medications with your MND Care Team Specialist or GP to try to get to the root of the cause. If the cause is weakened breathing muscles you might want to consider the use of overnight breathing support, which is very effective in curing morning headaches and the related symptoms of insufficient breathing while asleep.

After you decide you might like to try overnight breathing support (night-time ventilation) to see if it cures the

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headaches, makes you more alert and less confused, you need to consider the stage after that, which is where do you go

when your day time breathing is affected? Please consult factsheet 40 for more information about these questions.

Further Information

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|--------------|--------------------------------------|
| Factsheet 40 | Ventilation in MND |
| Factsheet 6 | Feeding Tubes |
| Factsheet 8 | Care Planning and End of Life Issues |
| Factsheet 24 | Powers of Attorney |

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