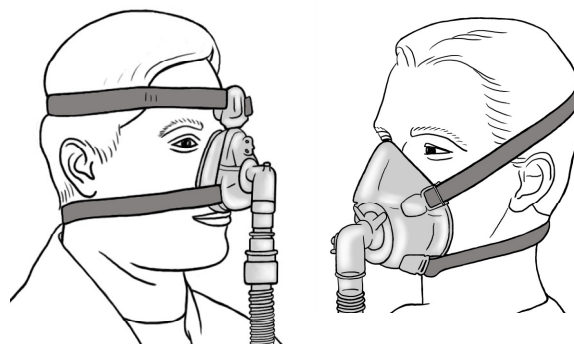


This factsheet is intended to deal with issues that may arise if your breathing is seriously affected by the progression of your MND. If you haven't already read factsheet 35 "Breathing Matters," you should read that before considering reading this factsheet. You should also read factsheet 6 "Gastrostomy and Tube Feeding," since the fitting of a feeding tube requires a small operation, which in turn requires that your breathing shouldn't be too badly affected at the time of the operation. If you have lost too much of your normal breathing abilities it may not be possible to fit a feeding tube, therefore consideration of these decisions is often raised well in advance and long before the decision is required.

If the breathing muscles are affected the early signs usually show themselves by causing problems due to lack of oxygen while sleeping. If this happens you might want to consider whether or not you want your breathing supported overnight by mechanical help. As MND progresses these problems can start to intrude into waking life until breathlessness can become a problem even with remarkably little exertion such as by simply holding a conversation.

The most common type of breathing support for people with MND in the UK consists of a tightly fitting flexible mask that can be fitted over the nose, or both the nose and mouth to supply a measured breath of air from a pump when you start the process of breathing in.

Many people, including people who do not have MND, find benefit in the use of such devices when they suffer from poor



Alternative types of face mask.

or disturbed sleep due to breathing problems. Overnight breathing support can help those suffering from sleep apnoea or night-time hypoxia (lack of oxygen) to return to a reasonable quality of life for very little inconvenience. A sufficient supply of oxygen and removal of carbon dioxide while you sleep prevents the morning headaches, confusion, tiredness and other problems caused by weakened respiratory (breathing) muscles.

Although the idea of sleeping with a mask fixed to the face can put many people off, those who have the will to persevere for three or four nights find the improvements to their quality of life so great that they very seldom decide that it is not for them. It is important to emphasise that it does need a couple of nights to get used to the mask and a decision should not be based on one night's experience alone.

Ultimately failure of the respiratory muscles can be an eventual result of

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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MND. The breathing muscles become too weak to expand the lungs and to provide an adequate cough, thus requiring assisted breathing support during the day as well as at night and the learning of assisted coughing techniques to clear phlegm from the lungs to prevent infections. The decision of whether or not to use breathing support is yours, but it is a decision you should make only after consulting with your family, doctor, and your other healthcare team members.

Choosing to use breathing support will have important consequences for your way of life and that of your family and carers. You will have to decide whether you are willing to accommodate your life and that of your family to the potential dependence on a breathing support apparatus. This decision must be made well before life-support ventilation is needed.

Although breathing support can be provided non-invasively or invasively it is normal in the UK to use a non-invasive positive pressure face mask as the first choice for respiratory support. As already mentioned, invasive support (tracheostomy) is done infrequently in the UK for people with MND and is usually done as an emergency procedure for people who suffer acute (sudden) respiratory failure.

Non-Invasive Ventilation

Non-invasive ventilation is achieved with a bi-level unit, a positive pressure volume ventilator, or much less commonly, a negative pressure ventilator. The most common breathing support is the bi-level (commonly known brand name product is the BiPap™). The bi-level helps you breathe by providing two levels of air pressure. A higher pressure when you inhale assists weak inspiratory muscles

and a lower pressure when you exhale. A circuit of tubing is used to deliver air from the bi-level pump to a mask over your nose or nose and mouth.

Non-invasive ventilation is usually started at night only. The practice of daily non-invasive lung hygiene techniques such as *lung volume recruitment* (see factsheet 35) with assisted cough is necessary when opting for non-invasive ventilation. As MND progresses increased breathing support may be required. This can be provided by using a mouth piece during the day with a volume ventilator and a different mask for night time use. A respiratory specialist can assist you in finding the appropriate masks for you.

Although some people using non-invasive ventilation may eventually come to rely on it 24 hours per day this is very difficult and cannot be sustained for extended periods of time. When someone does require non-invasive ventilation greater than 16 hours per day, it is probable they will soon need to consider the possibility that problems will set in making continued use uncomfortable or otherwise unsatisfactory. If these problems are encountered the person needs to consider whether or not they want to continue with assisted breathing.

One frequently encountered problem with prolonged use of face masks for non-invasive ventilation is damage to the skin resulting in ulceration where pressure is applied by the mask, particularly over the bridge of the nose. Not only is this ulceration painful when the cause (the face mask) is re-applied, it can also open a route for life threatening infections to enter the body. Complications like these, which might indicate a need to withdraw the ventilatory support, should also be considered when first making the decision to begin non-invasive ventilation.

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It is very easy to make the decision to use a ventilator for night time use only, then to gradually increase its use during daytime because it is there. People with MND considering the possibility of breathing support should also consider the possibilities that this support will be a temporary measure and that after a few months to a year or so it may need to be discontinued.

For many people in this situation invasive ventilation is a stage too far and they are not interested in having their life sustained "at any price," they therefore make the decision themselves as to how and when they should be weaned off their ventilator. Other patients might want to consider the possibilities of invasive ventilation; however the facts of this procedure are very uncomfortable for UK patients as it is only carried out infrequently in the UK since it raises many issues both about its practicality and subsequent quality of life

Invasive Ventilation

Invasive ventilation, which is rarely done in the UK for MND patients, requires a surgical procedure called a tracheostomy. The surgeon creates an opening in the neck connecting directly to the windpipe or trachea, into which is placed a curved plastic tube. This tube is connected by a hose to a volume ventilator, which assists with breathing. Whenever necessary, a suctioning catheter can be inserted into the tube in the trachea to remove secretions.

Most patients with a tracheostomy have already been fitted with a feeding tube (PEG tube) in anticipation of their breathing becoming too poor to permit the surgery to fit the PEG .

Provided you don't have severe bulbar

involvement speaking can continue after a tracheostomy through the use of a tracheostomy speaking valve such as the Passy-Muir® valve, which is used to direct airflow over the vocal cords when speech is required.

If you decide invasive ventilation is something you might want to consider then talk to your doctor about the full implications prior to making a decision.

Invasive ventilation will require full time, 24 hour support from trained carers. Even if you can count upon the assistance of family members it is a full-time job for three people, assuming eight-hour shifts and seven days per week commitment.

What about a power supply for the ventilator in the event of an extended power cut? There really are many issues to think about as this kind of intervention is truly a life-support situation.

Making Your Decision About Ventilation

In summary, it is best to learn what you can about ventilation options early on in the disease and prior to an emergency. Planning ahead enables you to be in more control. You should also communicate your wishes regarding the possible use of a ventilator in a personal healthcare advanced directive or a "Welfare Power of Attorney" and ensure that your attorney or advocate is agreeable to supporting your decisions. Be sure to provide your doctor with a copy of the directive and inform your family members where you keep it at home. Without a directive and making your wishes known, a tracheostomy might be carried out in the event of an unexpected respiratory crisis. If at any time you decide you are interested in any

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Mechanical Ventilation: Things to Consider		
Type of Ventilation	Non-Invasive Ventilation	Invasive Ventilation (with Tracheostomy)
Advantages	<p>Nose and mouth offer convenient routes for the delivery of breathing support and therefore do not involve any kind of surgical procedure.</p> <p>Easier to use than invasive ventilation.</p> <p>Usually more comfortable.</p> <p>Associated care is easier and with fewer complications.</p> <p>If using a mouth piece with the volume ventilator your cough will be more effective.</p> <p>Longevity may be increased.</p> <p>Little or no problem with aspiration</p> <p>With appropriate lung hygiene regimen, lung infection occurs less frequently.</p> <p>Easy to stop if you don't like it.</p>	<p>More secure system if you are ventilator dependent.</p> <p>Has been found to provide much longer survival.</p> <p>No masks required therefore the face is free of headgear, straps, and skin pressure problems.</p> <p>Doctors, nurses, and respiratory therapists tend to be more familiar with tracheostomy care and invasive ventilation.</p>
Challenges	<p>Finding and fitting the best face mask for you can take a bit of trial and error, specially with upper extremity paralysis.</p> <p>Custom-fitted silicone-moulded nasal masks which may provide a better seal at higher pressures, require additional time and expertise.</p> <p>Some people with MND find it difficult to adapt.</p> <p>If bulbar impairment is severe, non invasive breathing support may be more of a challenge to use.</p> <p>Air can get blown into the stomach causing bloating.</p> <p>Most centres are familiar with traditional invasive ventilation however, few have experience with day time mouth piece ventilation.</p> <p>Ulceration at pressure points can be problematic.</p>	<p>Some people feel it is too invasive, and increases their disability and dependence.</p> <p>The tracheostomy tube is a foreign object in the body and can increase secretion production and infections.</p> <p>Secretions require suctioning with a catheter through the tracheostomy, during the day and at night, which is uncomfortable.</p> <p>Coughing to clear the airways is difficult</p> <p>The tracheostomy site (stoma) can become infected, bleed, or develop granulations that need to be removed</p> <p>Associated care is more complex than non-invasive ventilation, therefore requiring more skills.</p> <p>Some people have difficulty with speech and swallowing.</p>

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form of breathing support, you should ask to be referred to a respiratory specialist.

Breathing management can play a major role in MND management and in enhancing quality of life. Monitoring your breathing function is very important for identifying problems early. Early detection is critical because there are things you can do to reduce the risk of infection, promote better ventilation for longer, and possibly extend your survival.

Many decisions about interventions and their withdrawal may need to be made along the way. Having good information about what is available to you and discussing options with your family and healthcare professionals will help you to make better decisions about what will be best for you

Further Information

Factsheet 35	Breathing Matters
Factsheet 6	Feeding Tubes
Factsheet 8	Advanced Care Planning and End of Life Issues
Factsheet 24	Powers of Attorney

International Ventilator Users Network <http://www.ventusers.org/index.html>

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