

# EMERGENCY PREPAREDNESS

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Every person with MG should be prepared for the possibility of an MG Crisis. Although you may never experience a crisis – being prepared is an essential. Here's what you should know.

Myasthenic crisis can develop slowly or quickly. It is important for patients to get medical care right away when symptoms of myasthenic crisis develop. Sometimes it is hard to tell if shortness of breath is due to anxiety or MG muscle weakness. Patients can do simple assessments at home to check respiratory function.

Signs that breathing function is worsening include:

- Cannot lay flat in bed without feeling short of breath or gasping for air
- Rapid shallow breathing (especially more than 25 breaths/minute)
- Having to pause in the middle of a sentence to take a breath
- Weak cough, especially when mucus/saliva cannot be cleared from the throat
- The muscles between the ribs and around the neck pull in during breathing
- Cannot count out loud past 20 after a full breath of air (single breath count)
- Sweating even when the room is not too warm
- Waking up frequently during the night gasping for air
- Feeling restless, agitated, drowsy or confused
- Breathing worsens even after taking MG medications
- The chest wall moves inward instead of expanding when air is inhaled
- Feeling too tired to keep breathing

Patients should seek immediate medical help when feeling short of breath with the warning signs listed above.

## Emergency preparedness plan

Every MG patient should have an emergency plan in place in order to make decisions and provide critical information to healthcare professionals.

## Calling for emergency assistance

Some 911 call centers now accept text messaging and some 911 centers can register patient medical information in their database in case of emergency.

Some medical alert systems can

call 911 when the help needed button is pushed.

Patients and caregivers need to check to see what options are available in their area. If the patient is unable to speak when calling 911, the operator will identify the location and send help. An ambulance should be called if the driver is anxious, the patient is too ill to speak, breathing assistance is needed or traffic will cause delays.

First responders must easily be able to see the house address from the street, especially at night. A flashing or colorful front house light can help first responders identify where to go. New electronic light switches can be activated by smart phone apps or voice-activated assistants like Google Home or Amazon Alexa. If the front door

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has an electronic key pad lock, the access code should be given to the 911 operator. Some medical alert programs can put a coded lock box on the front door. Lock boxes can also be purchased and mounted. In case of emergency, paramedics will be provided with the access number so that they can enter the home.

MG patients should wear some type of medical alert jewelry at all times to quickly notify healthcare professionals about their health history in case of emergency. Paramedics and hospital personnel are trained to look for medical identification jewelry, especially when caring for patients who are unable to speak. Various types and styles of bracelets and pendants are available. Some have USB chips or QR codes that store important medical information. Others have fall detection sensors and 24/7 emergency response services to provide medical history to providers.

MG patients should also have emergency contacts listed in their cell phones indicated by ICE in the cell phone directory. The ICE and medical history can be added to password-protected phones so that the information can be accessed by emergency room personnel without having to unlock the phone first. Emergency medical information apps can also be

downloaded to smart phones for use by healthcare providers. Patients can use their providers' patient portals to gather pertinent medical history and test results. Patients should check <https://www.healthit.gov/providers-professionals/faqs/what-patient-portal> and <https://www.healthit.gov/patients-families/your-health-data> for more information.

MG patients and their families should be prepared to educate providers about MG as it is a rare illness that presents differently than other types of respiratory failure. Shortness of breath from myasthenia gravis is due to weak respiratory muscles- a concept that many providers are not familiar with. Pulse oximetry is NOT a good test for breathing function in MG patients unless the

All MG patients should have a packet of medical information ready in case of emergency in a packet on the refrigerator and glove box of personal vehicles. The packet must be kept organized and up to date in a brightly colored, clearly labeled envelope. It should include:

Contact information for all physicians

Medical history & hospital records

Emergency contact list

Health insurance cards

Information about any implanted devices (i.e. IV port, pacemaker)

Advance directive papers and medical power of attorney (POA) forms

Complete medication list – including dosage, frequency and reason for all supplements, vitamins, over-the-counter and prescription medications.

List of allergies – including medications and other substances (foods, latex, chemicals)

Medical History – all surgeries, conditions and recent test results

MG information for providers  
<http://www.myasthenia.org/HealthProfessionals/EmergencyManagement.aspx>

MG medication precautions list  
<http://www.myasthenia.org/LinkClick.aspx?fileticket=zmLaFItarOQ%3d&tabid=318>

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patient has underlying lung problems such as COPD or pneumonia. It is not uncommon for MG patients in acute respiratory failure/myasthenic crisis to have pulse oximetry levels > 95%. Careful observation of respiration and bedside measurements (forced vital capacity, single breath count) are more reliable indicators of respiratory status than pulse oximetry in MG patients.

The ER physician should call the neurologist to guide treatment decisions. A family member or friend should be present at all times to act as an advocate. If the patient has a BiPAP machine at home, it should be brought to the hospital so that respiratory support will be provided without interruption.

An ambulance should be called if the driver is upset or if the patient is too ill to speak. An ambulance will be able to provide breathing assistance during the trip to the hospital and avoid delays from traffic congestion. Patients should bring their own ventilator or BiPAP to the hospital to ensure continuous breathing support although it may become necessary to switch to a different type of machine depending on the patient's respiratory status. While it may be helpful to go to the emergency room where the patient's neurologist practices, the ambulance may need to go to the nearest hospital to obtain immediate care for the patient.

Once the patient arrives at the emergency room, the patient's advocate should be prepared to communicate effectively with healthcare personnel. Key medical information should be organized and ready for providers. The patient's symptoms and medical history should be related in a concise manner using correct terminology to avoid missed information or errors. The patient's advocate should also be ready to clarify contradictory information between physicians, nurses and other providers.

Having an emergency preparedness plan in place allows MG patients to quickly receive appropriate medical care when myasthenic crisis develops. It also enables MG patients to have some control and direction in what happens during their treatment.

## MGFA RESOURCES:

- Emergency Alert Card (1) – Get this card for your wallet, it states that you have MG; gives your details (name, address, etc.); who to contact in an emergency; and First Responder guidance.
- Emergency Alert Card (2) – Provides guide on Drugs to be Avoided as well as your details (name, address, etc.); who to contact in an emergency. Ask for both cards by calling or writing the MGFA office.
- Emergency Management I brochure is directed to medical personnel and gives an overview of the issues – also discussed in this article.
- Emergency Management II is directed to patients, families and caregivers. Get both brochures to share with your family, add to your emergency kit and share with caregivers.

## IMPORTANT TERMS TO KNOW – MYASTHENIC CRISIS

### PATIENT SUPPORT

**Patient advocate** = an individual or group that assists patients by giving voice to concerns, helping resolve issues with insurance and healthcare providers, and ensuring that the patient's wishes are respected.

**ICE** = acronym for In Case of Emergency

**Living will** = a written statement specifying the treatment plan to be followed by healthcare providers when the patient is unable to communicate. This usually includes CPR, tube feeding, and other types of interventions provided at the end of life. Also known as an Advanced Directive.

**Medical guardian** = A person who is authorized to make healthcare decisions for a patient who is unable to do so.

**Hospital advocate** = Hospital employee who addresses problems and voices concerns about care provided in the facility. Also known as a Patient advocate.

**Medical durable power of attorney** = Legal document naming a person or persons to make healthcare decisions for the patient when the patient is unable to do so. It also provides instructions about the type of medical care the patient wishes to receive. Also known as a Health Care Proxy.

**Resuscitation** = treatment to revive a patient when the breathing has stopped and/or the heart has stopped beating.

## GENERAL

**Respiratory failure** = dangerous state in which the lungs are unable to pass enough oxygen into the blood and/or the lungs are unable to remove enough carbon dioxide from the blood. This can happen to MG patients when the respiratory muscles are too weak to move enough air in and out of the lungs.

**Cholinergic crisis** = increased muscle weakness due to overdose of MG medication (mestinon/Pyridostigmine)

**Myasthenic crisis** = the respiratory muscles have become so weak due to worsening MG that a ventilator is needed to breathe for the patient. Crisis should not be confused with exacerbation, which is a general worsening of MG symptoms.

**Exacerbation** = worsening of symptoms

**Conversion disorder** = physical symptoms that occur without a physical disease to cause them. Instead, symptoms are caused by an ongoing mental health problem such as depression or anxiety

**Respiratory arrest** = absence of breathing because the lungs have failed

**Refractory MG** = MG that does not improve with standard treatment

## SYMPTOMS

**Diplopia** = double vision

**Ptosis** = droopy eyelid

**Dyspnea** = shortness of breath

**Orthopnea** = difficulty breathing when lying flat in bed

**Tachypnea** = rapid breathing rate

**Hypophonia** = very quiet speech that is hard to hear

**Dysarthria** = slow, slurred speech that is hard to understand

**Dysphagia** = difficulty swallowing

**Diaphoresis** = breaking out into a sweat

**Bulbar symptoms** = weakness of the muscles in the neck and jaw making it hard to talk, chew, swallow and hold up the head. Bulbar weakness can lead to choking and aspiration

**Aspiration** = inhaling food, fluids or other substances into the lungs

## HOME RESPIRATORY ASSESSMENT

**Single breath count** = the patient counts out loud after breathing in as deeply as possible. Patients with normal respiratory function can count to 50. A single breath count less than 15 indicates dangerously poor respiratory function.

**Interrupted speech** = pausing to take a breath after every few words, due to inability to inhale enough air to speak in complete sentences. Also known as staccato speech.

**Use of accessory muscles** = Muscles around the collarbone and neck are used to help move air in and out of the lungs when the diaphragm becomes very weak.

**Paradoxical breathing** = Weakness of the diaphragm leads to abnormal chest movements with the rib cage contracting inward as air is inhaled into the lungs,

## HOSPITAL RESPIRATORY ASSESSMENT

**Pulse oximeter** = a small device used to measure blood oxygen levels

**ABGs** = Arterial Blood Gases. Used to measure oxygen and carbon dioxide levels in the blood.

**Respiratory acidosis** = poor respiratory function leads to too much carbon dioxide in the blood, which increases the acidity of the blood

**Hypoxemia** = not enough oxygen in the blood

**Hypercarbia** = too much carbon dioxide in the blood due to poor respiratory function Also known as hypercapnia.

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## PULMONARY FUNCTION TESTS

**VC** = Vital Capacity. Largest amount of air that a patient can exhale after inhaling as much air as possible.

**FVC** = Forced Vital Capacity. Largest amount of air that a patient can exhale as forcefully and quickly as possible after inhaling as much air as possible.

**NIF** = negative inspiratory force. Used to measure strength of respiratory muscles for inhaling air. Also known as MIP = Maximum Inspiratory Pressure.

**MEF** = Maximum Expiratory Force. Used to measure strength of respiratory muscles for exhaling air. Also known as MEP = Maximum Expiratory Pressure.

## TREATMENTS

**CPAP** = Continuous Positive Airway Pressure. Breathing machine used by patients with sleep apnea to keep their airways when they are asleep. Continuous, steady air pressure is used to splint the airways and keep them open so the patient can breathe during sleep.

**BiPAP** = Bi-level Positive Airway Pressure. Breathing machine used by patients with MG when respiratory muscles become weak. BiPAP machines have two settings: a high air pressure is used to help the patient inhale and a lower air pressure is used to allow the patient to exhale easily.

**Plasmapheresis** = a procedure similar to dialysis in which blood is removed from the body and returned after disease-causing antibodies are carefully removed.

**IVIG** = Intravenous Immunoglobulin, a concentrated solution of antibodies given via IV to treat autoimmune diseases.

**Cholinesterase inhibitor** = medication that prevents the acetylcholine enzyme from being destroyed in the muscles. This helps relieve muscle weakness due to MG. Examples include: Pyridostigmine (Mestinon), Caffeine, Huperzine A, and Tensilon.

**Mestinon** = Primary cholinesterase inhibitor used in the treatment of MG. Also known as Pyridostigmine

**Intubation** = insertion of a breathing tube into the airway so that a ventilator can breathe for the patient

**Mechanical ventilation** = use of a breathing machine to breathe for a patient ✨