



EMERGENCY CARE BY FIRST RESPONDERS

Patients with myasthenia gravis (MG) can experience respiratory muscle weakness leading to respiratory insufficiency. Pending respiratory failure necessitating intubation is a life-threatening situation and defined as myasthenic crisis. Prompt recognition of impending respiratory paralysis is the key to the successful management of a myasthenic crisis.

Respiratory assessment

- Check for retraction of supraclavicular fossa and intercostal spaces as indicators of respiratory accessory muscle usage. Use of accessory respiratory muscle in MG patients is an important sign that respiratory effort may not be sustained. However, generalized muscle weakness in MG patients can at times mask accessory muscle usage.
- Paradoxical breathing, failure to lie supine and inability to speak more than a few words are indicators of diaphragm weakness. Weak neck flexion also correlates with diaphragm dysfunction.
- Tachypnea- rapid shallow breathing is seen with MG patients to compensate for weak respiratory muscles.
- Pulse oximetry is NOT a good indicator of respiratory strength in MG patients as abnormalities often develop only after life-threatening respiratory failure has already occurred. This is distinct from other causes of respiratory failure. 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.
- Single breath count test: Ask patient to count out loud after maximal inspiration. Ability to reach 50 indicates a normal respiratory function. Single breath count of less than 15 indicates a dangerous low forced vital capacity (FVC).

Immediate treatment

- Oxygen usage is helpful but does not alleviate respiratory distress in MG patients. Titrate to keep oxygen saturation at 94-98% on pulse oximetry.
- If breathing is inadequate, provide assistance with ventilation immediately. Non-invasive ventilation may be given via bag-valve mask (BVM) or CPAP.
- Invasive ventilation is needed when airway patency cannot be maintained or when non-invasive ventilation is unsuccessful.
- Transport the patient immediately to nearest emergency facility. Bring medical history paperwork if patient has them readily available.

EMERGENCY CARE BY PHYSICIANS

Pulmonary function testing

- Measure forced vital capacity (FVC) and negative inspiratory force (NIF) immediately and every two to six hours thereafter. FVC and NIF are primary parameters to measure respiratory muscle strength, especially in patients without obvious respiratory distress.

- Consider FVC and NIF values together along with clinical signs and symptoms of respiratory failure. MG is characterized by repetitive muscle weakness- the muscles become weaker as they are used. Trend of numbers over time is more important than individual test results. A declining NIF or NIF worse than 20 cm H₂O and FVC less than 10 to 15 mL/kg should prompt intubation.
- A 10% decrease in FVC from upright to supine position indicates diaphragmatic weakness. This is a more sensitive indicator of respiratory muscle weakness than upright FVC alone.
- FVC and NIF may be falsely low if the patient is unable to close mouth adequately around the mouthpiece due to bulbar weakness. A face mask may be used instead for these patients.
- Pulse oximetry and arterial blood gas (ABG) measurements are NOT good indicators of respiratory strength in MG patients as abnormalities often develop only after life-threatening respiratory failure has already occurred. Careful observation (dyspnea and use of accessory muscles) and bedside measurements (forced vital capacity, single breath count) are much more informative than pulse oximetry or ABG results.
- Single breath count test- Ask patient to count out loud after maximal inspiration. Ability to reach 50 indicates a normal respiratory function. Single breath count of less than 15 indicates a dangerous low forced vital capacity (FVC).

Decision to initiate assisted ventilation

- Do not wait for ABG to show hypoxemia or hypercapnia. These are late developing signs that appear only immediately prior to respiratory arrest in MG patients.
- Weak respiratory muscles may suddenly fatigue, producing precipitous respiratory collapse. Elective intubation is preferable to emergency intubation or respiratory arrest.
- The standard 20/30 rule (FVC less than 20 ml/kg or NIF less than 30 cmH₂O) are the best indicators of the need for ventilator support.
- BiPAP is an alternative to intubation in MG patients without hypercapnia who are able to clear secretions
- The use of neuromuscular blockers for intubation may impair neuromuscular function and delay extubation.
- The initial ventilator mode is typically “assist control/volume control.”

Indicators of need for ICU admission: MG patients with deterioration or impending crisis should be admitted to an intensive care unit.

- Orthopnea, dyspnea at rest, weak cough, prominent neck flexion weakness, difficulty in clearing secretions
- Rapid shallow breathing, use of accessory muscles, paradoxical abdominal breathing
- Baseline FVC less than 30 mL/kg of ideal body weight even if the patient does not show signs of respiratory distress
- Broken speech in need of pause and breathe after every a few words

Other considerations

- Discontinue cholinesterase inhibitors (mestinon) for intubated patients as they increase secretion production and risk of aspiration pneumonia. Glycopyrrolate should only be used with extreme caution as it can lead to mucous plugging.
- Discontinue all medications that exacerbate MG muscle weakness (antibiotics, beta blockers). For a detailed list of medications potentially exacerbating MG symptoms, please click [here](#).
- Consult neurology for specific treatment options (plasma exchange, IVIG, corticosteroids)
- Identify and address triggers that may underline myasthenic crisis (infection, tapering of immunosuppression etc.).

We greatly appreciate input from Sally O'Meara MSN, RN.