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355 Lexington Avenue, 15th Floor
New York, NY 10017-6603
800-541-5454 (Helpline)
212-297-2158 fax
MGFA@Myasthenia.org

www.Myasthenia.org   

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EMERGENCY MANAGEMENT OF MYASTHENIA GRAVIS

Information and Guidance for First Responders and Emergency Care



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EMERGENCY MANAGEMENT OF MYASTHENIA GRAVIS:

Information and Guidance for First Responders and Emergency Care

Clinical Manifestations of Myasthenia Gravis (MG)

Myasthenia gravis is a rare neuromuscular disorder which causes fatigable weakness of voluntary muscles. Symptoms of myasthenia can include ptosis (eyelid drooping), disconjugate eye movements and double vision, slurred speech, difficulty chewing and swallowing, and neck, arm and leg weakness. When weakness is severe, there may be trouble walking and breathing. This muscle weakness may fluctuate over time and throughout the course of the day. Weakness can be mild or severe. Individuals with MG usually feel stronger in the morning or after a period of rest. Prolonged activity or repeated use of affected muscles can increase myasthenic weakness. Treatments for myasthenia include symptomatic therapy and/or immunosuppressant medications.

Triggers which may worsen MG muscle weakness include:

Medications

- High doses of steroids
- IV magnesium
- Some antibiotics
- Certain heart/blood pressure medications
- Some general anesthetics and paralytics
- Botulinum toxin
- Stopping or reducing medications used to treat MG

Illness or infection

Heat

Stress from trauma or surgery

Myasthenic crisis

- A potentially life-threatening complication of myasthenia gravis. Respiratory failure occurs due to weakness of respiratory muscles and mechanical ventilation is required.
- Respiratory failure may also develop due to weakness of muscle that keep the airway open. BiPAP may be sufficient or the patient may need endotracheal intubation.
- **Careful assessment and monitoring is required as myasthenic crisis presents differently from other forms of respiratory failure.**
- Prompt recognition of impending myasthenic crisis may prevent fulminant crisis from developing.



PRE-HOSPITAL RESPIRATORY ASSESSMENT AND MANAGEMENT:

Check for tachypnea:

- Rapid shallow breathing is seen in 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.

Inspect for use of accessory muscles of respiration:

- Check for retraction of supraclavicular fossa and intercostal spaces as indicators of respiratory accessory muscle usage. Patients may also use neck and abdominal muscles. 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 and inability to lie supine or speak more than a few words are indicators of diaphragm weakness.
- Weak neck flexion also correlates with diaphragmatic dysfunction. Neck flexion strength can be tested by having the patient lie supine and attempt to lift his/her head off the stretcher and tuck his/her chin.
- Severe slurred speech and difficulty managing secretions are also signs of potential impending MG crisis.

Single breath count test:

- Single breath count test is a good bedside measurement of respiratory function which can be performed quickly and without additional equipment
- To perform, ask patient to count out loud after maximal inspiration. Ability to reach 50 indicates normal respiratory function. Single breath count of less than 15 typically correlates with low forced vital capacity (FVC) and respiratory muscle weakness.

Immediate management:

- **Elevate head of the stretcher, keep patient cool and have suction available**
- 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 BiPAP.
- Invasive ventilation is needed when airway patency cannot be maintained or when noninvasive ventilation is unsuccessful.
- Transport patient immediately. Alert ED to the patient's history of MG. Bring medical history paperwork if patient has it readily available.

In-Hospital Initial Assessment and Management:

- Measure forced vital capacity (FVC) and negative inspiratory force (NIF) at baseline and trend, typically every 6 hours or more or less frequently, as needed. 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 typically prompts BiPAP or intubation. BiPAP may be indicated earlier for FVC less than 20 ml/kg or NIF worse than 30 cm H₂O if patient is able to clear his/her secretions and has adequate bulbar strength.
- Careful observation (tachypnea and use of accessory muscles) and bedside measurements (forced vital capacity, single breath count) are much more informative than pulse oximetry or ABG results. Measuring FVC upright and supine can sometimes provide insight, as decline in the supine position may indicate neuromuscular weakness
- **Pulse oximetry and arterial blood gas (ABG) measurements are NOT good indicators of respiratory strength in MG patients as abnormalities often develop only after potentially life threatening respiratory failure has already occurred.**
- **Do not wait for ABGs 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.

- BiPAP is an alternative to intubation in MG patients without hypercapnia who are able to clear secretions. Patients may have their own BiPAP or NIV equipment. Depending on local guidelines, patients may be able to use this if it remains medically appropriate for the presenting clinical scenario.

Next Steps:

- MG patients with impending or actual MG crisis should be admitted to an intensive care unit. Signs of impending crisis with need for ICU admission include: FVC less than 2 ml/kg, NIF less than 30, serial reductions in these numbers, significant bulbar dysfunction, orthopnea and/or rapid shallow breathing.
- Consult neurology for specific treatment options (e.g. plasma exchange, IVIG, corticosteroids etc). Consult with neurology regarding continuation of pyridostigmine if patient is intubated. Due to possibility of increased secretions, continued use may predispose patient to aspiration and ventilator associated pneumonia and is typically avoided.
- Contact the patient's outpatient neurologist for input regarding care of worsening myasthenia.
- **Review medication list and minimize medications which can worsen MG**
- Identify and address triggers that may have exacerbated myasthenia (see above).