**Myasthenia Gravis in the ED**

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Case Presentation: 43 year-old woman with a history of MG presents to the ED with complaints of dysphagia and weakness that is worse in the evening. She denies difficulty breathing and states that she is compliant on her medications, pyridostigmine 60mg q6h and prednisone 5mg qd.

Pathophysiology of Myasthenia Gravis: Autoimmune disease characterized by **muscle weakness and fatigue**, which is seen especially when using **voluntary muscles repetitively**. Acetylcholine receptor (AChR) antibodies impair receptor function at the postsynaptic end plate of the neuromuscular junction, resulting in decreased muscle fiber potential amplitudes. This autoimmune response is usually caused by either **dysfunction of the thymus gland (75% of people) or an immune response to infectious agents**.

Clinical Features: Generalized weakness, most notable in the **proximal muscle groups, neck extensors and facial or bulbar muscles**. Most common presenting symptoms are **ptosis and diplopia**. Also seen are oropharyngeal symptoms such as **dysphagia, dysarthria, and dysphonia**. Internuclear opthalmoplegia as well as end-gaze nystagmus can also be seen. May also present as extreme weakness of the respiratory muscles resulting in respiratory failure. This is termed Myasthenic crisis.

What to worry about in the ED:

1. Myasthenic Crisis: this should not be taken lightly. If you suspect a patient is in myasthenic crisis immediately order a **NIF** (negative inspiratory force). This is commonly done by the respiratory therapist in the ED. A value **less than 20** indicates that the patient needs to be intubated (some sources say 30). Also check the patients cough and gag reflexes as well as their ability to swallow. Be careful, because the generalized weakness these patients present with **can sometimes mask the normal responses to respiratory distress**. This can commonly be caused by certain antibiotics (**aminoglycosides, fluoroquinolones, macrolides**), cardiac drugs (**beta-blockers**, procainamide, and quinidine), and **magnesium**. Avoid the administration of depolarizing and non-depolarizing NMB agents in patients with MG Crisis.
2. Cholinergic Crisis: side effect of prolonged or excessive anticholinesterase medication is weakness, which can be hard to recognize **whether this is due to the medication or worsening myasthenia**. Cholinergic crisis is rarely if ever seen with dose limitation of pyridostigmine to less than 120 mg every three hours. Cholinergic crisis is so rare that it should not be the presumed cause of increasing weakness unless the doses taken are known to significantly exceed the 120mg every three hours range. Otherwise, **even in the presence of cholinergic side effects, it should be assumed that the patient's underlying myasthenia gravis is worsening** and appropriate treatment should be initiated.

Rapid Therapies

Once your patient's **airway** is stabilized, the mainstay of treatment for MG is **plasma exchange (PLEX) and IVIG**. You of course want to get immediate consultation from your neurology and hematology colleagues before initiating these therapies.

Sources // Further Reading

1. Tintinalli J, Stapczynski J, Ma OJ et al. Tintinalli's Emergency Medicine: A Comprehensive Study Guide, Seventh Edition (Book and DVD). Mcgraw-hill; 2010. Pgs 1166-1168.
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