Myasthenia Gravis

Myasthenia Gravis is a consequence of autoimmune attack on the acetylcholine receptor complex at the postsynaptic membrane of the neuromuscular junction.

- Results in weakness with a fluctuating pattern that is most marked after prolonged exertion.
- Typically involves ocular muscles producing ptosis and diplopia as well as bulbar muscle weakness resulting in dysphagia and dysarthria.
- The diagnosis should be considered in patients who have acute respiratory failure with these cranial nerve findings.
- Upper airway muscle weakness is a common mechanism leading to myasthenic crisis due to oropharyngeal and laryngeal weakness resulting in upper airway collapse with obstruction along with inability to swallow secretions leading to aspiration.
- Findings of bulbar myasthenia associated with upper airway compromise include:
  - Flaccid dysarthria with hypernasal, staccato or hoarse speech
  - Dysphagia sometimes associated with nasal regurgitation
- Cholinergic crisis includes:
  - Muscle fasciculations and prominent cholinergic symptoms including miosis, lacrimation, sweating, salivation, abdominal cramps, diarrhea, vomiting, increased bronchial secretions and bradycardia.
  - Other symptoms of cholinergic crisis include:
    - Muscle cramps, diarrhoea, vomiting, increased bronchial secretions and bradycardia
    - Chewing fatigue - jaw closure is often weak and cannot be maintained against resistance.
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- Patients with features of impending myasthenic crisis including severe bulbar weakness, vital capacity of less than 20-25ml/kg, weak cough with difficulty clearing secretions should be admitted to an intensive care unit.
- Criteria for intubation are similar to other neuromuscular conditions:
  - Failure to protect airway
  - Fatigue
  - Hypercapnic respiratory failure (the majority who become hypercapnic require intubation)

Plasma exchange:
- An effective short term treatment for myasthenia gravis and for surgical preparation in symptomatic myasthenic patients with significant improvements having been demonstrated in several case series (no controlled trials have been performed).
- Onset of improved strength is generally seen after 2-3 exchanges.

IVIG:
- May represent an alternative short term treatment for myasthenia candidates who are poor candidates for plasma exchange due to difficult vascular access or sepsis.
- Comparable efficacy with plasma exchange has been demonstrated in a small randomised controlled trial of IVIG at 1.2 and 2g/kg over 2-5 days; however, a larger retrospective study suggested plasma exchange was more effective.

Corticosteroids:
- Eg prednisolone 1mg/kg/day are occasionally used in prolonged myasthenic crises that fail to respond to treatment with plasma exchange or IVIG.
- Initiation of corticosteroids may lead to a transient increase in weakness.

Thymectomy:
- May result in long-term improvement in patients with a suspected thymoma or with a life expectancy of greater than 10 years.

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