

# NMJ DISEASE

## Lambert-Eaton Myasthenic Syndrome

Autoimmune.

Autoantibodies to presynaptic voltage-gated Ca channels. May be associated with small cell lung cancer and some other cancers (paraneoplastic).

Sx:

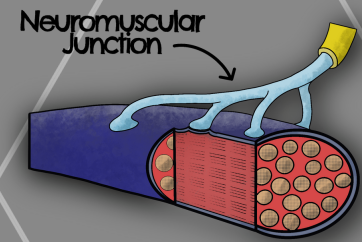
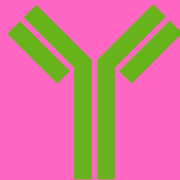
- Limb weakness
  - Proximal, legs more affected than arms
- Hyporeflexia
- Autonomic features
  - Dry mouth, impotence

Ix:

Increasing response on repetitive nerve stimulation is typical feature (not fatigueable as in Myasthenia Gravis).

Mx:

- Drugs facilitating ACh release (3,4-diaminopyridine)
- Pyridostigmine
- Immunosuppressants
- IVIG/plasma exchange



## Botulism

Exotoxin from *clostridia botulinum* irreversibly blocks ACh release.

Contaminated food/IVDU.

Bulbar muscles and autonomic nervous system effects give sx:

- Dilated, non-reactive pupils
- Diplopia >> blurred vision
- Flaccid paralysis
- Bulbar palsy
- Weakness begins with extraocular and oropharyngeal (bulbar) muscles, then generalised

Treat with Botulism trivalent antitoxin (early) and supportive care.

## Myasthenia Gravis

Autoimmune- antibodies attack nicotinic ACh receptors in skeletal muscle (AChR).

More common in women (as are most autoimmune conditions)

Fatigueable muscle weakness, fluctuating in severity- become fatigued with use and then improve on rest.

Muscles commonly affected:

- Eyes- ptosis and complex ophthalmoplegia, diplopia
- Face
- Neck
- Bulbar- dysphagia, dysarthria
- Respiratory- can be fatal

May be associated with other autoimmune conditions.

Ix:

- Electromyography
- CT thorax (thymoma)
- Autoantibodies to AChR or muscle-specific kinase
- Tensilon Test- IV edrophonium gives temporary relief of sx- no longer commonly used

Mx:

- Long-acting acetylcholinesterase inhibitors- pyridostigmine etc
- Prednisolone (immunosuppress)
- Plasma exchange/IVIG- temporary relief
- Thymectomy- thymoma is present in some cases

