Lambert-Eaton Myasthenic Syndrome

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Risk

- Unknown true incidence. Few studies of very specific regions outside USA report prevalence between 0.48 and 3.42 per million.
- 60–84% of LEMS patients have SCLC.
- LEMS mostly affects middle-aged adults, with rare occurrence in children.

Perioperative Risks

- Increased risk for fall when ambulating, due to proximal lower extremities weakness
- Hypotension due to autonomic dysfunction
- Prolonged emergence secondary to persistent muscle weakness
- Respiratory compromise or collapse after extubation

Worry About

- Failing extubation, necessitating unplanned ICU admission.
- Exacerbation of muscle weakness postoperatively.
- Concomitant presence of SCLC may complicate respiratory function.

Overview

- Autoimmune disorder affecting the presynaptic NMJ.
- Most patients present with slow progressive lower extremities muscle weakness.
- LEMS is different from MG:
  - LEMS affects the proximal lower extremities more than MG.
  - Primarily affects presynaptic mechanisms, whereas MG primarily affects postsynaptic mechanisms.
  - Muscle weakness transiently resolves with activities in LEMS.
  - LEMS is strongly associated with SCLC.
- Pathophysiology: Antibodies attack VGCC, diminishing the release of calcium and subsequent reduction in the release of acetylcholine in the presynapse.

Etiology

- LEMS is an autoimmune disease. IgG antibodies target the P/Q type VGCC at the presynaptic endplate of the NMJ.
- The strong prevalence of SCLC in patients with LEMS suggests the presence of the same antigen in SCLC and at the presynaptic NMJ.
- Cerebellar degeneration may be present in some patients with LEMS. This is likely due to the presence of the P/Q type VGCC in the cerebellum.
- Acetylcholine is necessary for the autonomic function and its reduction in LEMS may result in autonomic nervous system dysfunction.

Usual Treatment

- First line treatment with 3,4-diaminopyridine, which blocks potassium at the neurosynapse, allowing for the release of acetylcholine. It also directly facilitates neurotransmission at VGCC.
- Removing or treating the SCLC tumor may be considered if diaminopyridine is ineffective in alleviating symptoms.
- In cases of severe weakness prednisone and azathioprine may be added to treatment therapy. This treatment must be initiated slowly because it may cause acute weakness prior to having their positive impact.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Ptosis	Drooping of eyelid	Cranial nerve assessment	Symptoms reported by pt
RESP	Respiratory weakness	Dyspnea	Supplemental O_2 dependence Difficulty taking deep breaths	PFTs, including inspiratory force measurements. Phrenic nerve stimulation recordings CXR, CT scan
CNS	Autonomic dysfunction	Dry mouth Erectile dysfunction	Reduced saliva Impotence	Symptoms reported by pts
MS	Proximal lower limbs weakness	Aching, fatigued or stiff muscles affecting gait	Depressed or absent DTRs	Plasma levels of P/Q type VGCC antibodies Electromyography

Key References: Titulaer MJ, Lang B, Verschuuren JJ: Lambert–Eaton myasthenic syndrome: from clinical characteristics to therapeutic strategies, *Lancet Neurol* 10(12):1098–1107, 2011; Weingarten TN, Araka CN, Mogensen ME, et al.: Lambert-Eaton myasthenic syndrome during anesthesia: a report of 37 patients, *J Clin Anesth* 26(8):648–653, 2014.

Perioperative Implications

Preoperative Preparation

- Confirm adequate respiratory function. Verify use of supplemental oxygen at home.
- Prepare for possible ICU admission if unable to extubate due to muscle weakness.
- Consider use of monitored anesthesia care or regional anesthesia whenever possible instead of general anesthesia.
- Assess baseline strength of extremities.

Monitoring

- Standard monitoring
- Arterial line in patients presenting with respiratory compromise or significant autonomic dysfunction
- Nerve stimulation monitoring

Airway

- Avoid intubation whenever possible.
- Consider use of supraglottic airway instead of endotracheal intubation to avoid muscle relaxation.

Preinduction and Induction

- Depolarizing and nondepolarizing muscle relaxants can be used if muscle relaxation is required.
- If unable to avoid endotracheal intubation, consider intubating without muscle relaxants. Avoid neuromuscular relaxants whenever possible. Remember that inhaled anesthetics each have efficacy in neuromuscular blockade.

Maintenance

- Both IV and inhalational anesthetics have been given safely to LEMS pts. Vigilance to administer minimum requirements of anesthetics will likely minimize perioperative complications.
- Hypotension may occur as a result of autonomic dysfunction; vasopressors may be given as boluses or continuous infusion.
- Pts on prednisone should receive a perioperative dose of hydrocortisone.

Extubation

- Ensure return of safe cognitive function, muscle strength, and ventilatory function before extubation.

Postoperative Period

- Judicial use of opioids in the PACU to avoid respiratory compromise.
- Increased susceptibility to respiratory failure and reintubation.
- Consider continued monitoring of pulse oximetry after discharge from PACU.

Anticipated Problems/Concerns

- Muscle weakness
- Respiratory compromise

Landouzy-Dejerine Dystrophy (Facioscapulohumeral Muscular Dystrophy)

Francis Veyckemans

Risk

- Prevalence estimated to be 1:20,000
- Affects equally males and females
- Third most common familial myopathy after myotonic dystrophy and Duchenne muscular dystrophy

Perioperative Risks

- No greater risk of malignant hyperthermia than normal population.
- Possible risk of acute rhabdomyolysis if succinylcholine and/or a halogenated agent is used. However, the dystrophin-glycoprotein complex is not involved in Landouzy-Dejerine dystrophy, and no case of anesthesia-induced rhabdomyolysis has been reported so far.
- Perioperative respiratory complications
- Supraventricular paroxysmal tachycardia

Worry About

- Muscle weakness

Overview

- Muscular dystrophies are a heterogeneous group of genetic muscle disorders leading to progressive weakness and muscle wasting. They were grouped together based on a common histologic picture: variations in fiber size and areas of muscle necrosis