

NMJ disorders			
	Myasthenia gravis (MG)	Lambert Eaton	Botulism
Classification	Postsynaptic (Ach.R)	Presynaptic (VGCC)	Presynaptic (SNARE protein)
Clinical manifestations	<p><b>Ocular:</b></p> <ul style="list-style-type: none"> <li>Fluctuating fatigable weakness</li> <li>Binocular <b>diplopia</b></li> <li><b>Asymmetric ptosis</b></li> </ul> <p><b>Generalized:</b></p> <ul style="list-style-type: none"> <li><b>Bulbar:</b> dysarthria, dysphagia, fatigable chewing, breathy nasal speech, regurgitation</li> <li><b>Facial:</b> bell's phenomena, sneer, expressionless face</li> <li><b>Neck and limbs:</b> (Symmetric Proximal &gt; distal, <b>arms</b> &gt; legs) <b>neck flexors</b>, deltoid, triceps, <b>finger extensors</b>, hip flexors and ankle dorsiflexors, neck extensors (dropped head)</li> <li><b>Respiratory:</b> restrictive PFT, SOB, respiratory failure (<b>myasthenic crisis</b>)</li> </ul> <p>Pupils are Not affected</p>	<p><b>Proximal &gt; distal legs weakness.</b></p> <p><b>Mild</b> ptosis, dysarthria, dysphagia.</p> <p><b>Autonomic:</b> Dry mouth, erectile dysfunction</p> <p><b>Sensory:</b> paresthesia, myalgia, mild distal feet sensory loss</p> <p><b>Hyporeflexia</b></p> <p>Respiratory weakness (Rare)</p> <p>Pupils are Poorly reactive</p>	<p><b>Nausea and vomiting</b> in foodborne botulism</p> <p><b>Acute Symmetric descending weakness (Proximal &gt; distal)</b></p> <p><b>Symmetric ptosis &amp; EOM weakness</b></p> <p><b>Bulbar:</b> Dysarthria, dysphagia, fascial muscle weakness</p> <p><b>Respiratory weakness</b></p> <p>NO fever No sensory deficits except <b>blurred vision</b> Normal or slow heart rate</p> <p>Pupils are Fixed dilated</p>
Investigation			
Bedside test	1. Edrophonium (Tensilon) test	2. Ice pack (ocular cooling) test	3. Fatiguing Maneuvers
Fatigue >	Improves with rest	Improves with exercise	Improves with exercise
Serology	<b>Ach. R Ab.</b> > if -ve, female, bulbar > <b>MuSK Ab.</b>	VGCC P/Q-type Ab.	-
Repetitive nerve stimulation	>10% decrement	Increment	Increment
EMG	Rarely used		
CT chest	Thymic hyperplasia (young, females) Thymoma (elderly, males)	Small cell lung carcinoma	
Others	Screen for autoimmune thyroid disease		
Management			
	<ol style="list-style-type: none"> <li><b>Physostigmine (cholinesterase inhibitor)</b></li> <li>Prednisolone</li> <li>Methotrexate and azithroprine</li> </ol> <p><b>Myasthenic crisis:</b></p> <ul style="list-style-type: none"> <li>Plasmapheresis</li> <li>IVIg</li> </ul> <p><b>Thymectomy:</b></p> <ul style="list-style-type: none"> <li>AchR Ab. +ve pt. &lt;65 y.o.</li> <li>Thymoma</li> </ul>	<p><b>Aminopyridines</b> (3,4 DAP) + physostigmine</p> <p>+/- guanidine HCl</p> <p>Initial IVIG &gt; SCLC resection</p>	<p>Respiratory Support</p> <p>Clostridium botulism A, B &amp; E antitoxin</p> <p><b>Infantile botulism:</b> Human IVIG</p> <p><b>Wound Botulism:</b> Antibiotics (penicillin G or metronidazole if resistant)</p>