

ACUTE NEUROMUSCULAR DISORDERS

(Last updated: 07/22/2019; Reviewed by: Catarina Aragon Pinto MD)

PRESENTING COMPLAINT: ascending weakness

FINDINGS

- **A** N or compromised airway
- **B** ↑ RR, accessory respiratory muscles use, paradoxical breathing, orthopnea
- **C** ↓/↑N BP, ↑/N HR
- **D** Symmetric focal deficits, paraparesis, bifacial weakness, ptosis, ↓reflexes
- **E** Cyanosis, rash (dermatomyositis)
- **L_{PC}** ABG ↓pH, ↓PO₂, ↑PCO₂, CBC, CRP, electrolytes
- **U_{PC}** Diaphragmatic paralysis

***V** (verbal), **P** (pain), **U** (unconsciousness), **D** (delirious)

U_{PC} (point of care ultrasound) **L_{PC}** (point of care labs)

OTHER HISTORY

- Sensorimotor (ascending paralysis and paresthesia; usually starts in the lower extremities); cranial nerves (diplopia, ptosis, dysphagia, dysarthria; autonomic (palpitations, postural lightheadedness, urinary retention, constipation); sphincter dysfunction is **not** usually present; myopathy and Myasthenic Syndromes do **not** have sensory symptoms; sensory level is highly suggestive of a transverse myelopathy, not of a neuromuscular disorder; pigmenturia; myalgia
- Weakness pattern
 - Proximal = distal: polyradiculoneuropathy
 - Proximal: myopathy or neuromuscular junction disorder
 - Proximal and oropharyngeal: neuromuscular junction disorder
- **Predisposing factors**
 - Trauma, surgery, infection, vaccine (GBS and MG), emotional distress (MG), wound infection, canned food (botulism), small cell lung carcinoma or other cancers (LEMS), mosquito exposure (WNV), excess pyridostigmine (cholinergic crisis), immunotherapy (inflammatory myositis)

DIFFERENTIAL DIAGNOSIS

- Peripheral Neuropathy (polyradiculoneuropathy): Guillain-Barre Syndrome, West Nile virus infection, poliomyelitis, arboviruses infection (Dengue, Chikunguya and Zika viruses), diphtheria, acute intermittent porphyria

- Neuromuscular junction disorder: Myasthenia Gravis (MG), Lambert-Eaton Myasthenic Syndrome (LEMS), botulism, hypermagnesemia, cholinergic crisis
- Myopathy (inflammatory myopathies)

OTHER INTERVENTIONS

- **Labs:** Blood: CBC, lactic acid, CK, NT-pro BNP baseline PT/APTT, ABGs; Urine: urinalysis
- Specific auto-antibodies:
GBS: Anti-GM1, anti-GQ1b, anti-GD1a, anti-GT1a; Myasthenia Gravis: Anti-Achr and anti-Musk;
Lambert-Eaton Myasthenic syndrome: Anti-VGCC; Inflammatory myopathies: Myomarker 3 panel and anti-HMGCR
- Monitoring: blood pressure, oximetry, and consider foley, especially in rhabdomyolysis
- Respiratory therapy assessment of negative inspiratory force (NIF), maximum expiratory pressure (MIP), vital capacity (VC) frequently: i.e. every 4-6 hours
- **Imaging:** Chest CXR (to rule out infection); If Guillain-Barre Syndrome, consider cervical Spine MRI (r/o transverse myelopathy)
- Nerve conduction studies and EMG with repetitive stimulation (especially in difficult cases)
- Swallow evaluation
- GBS: lumbar puncture with CSF analysis (cells, protein, glucose, gram and culture):
Albuminocytologic dissociation: CSF white cells <10, CSF protein >100
 - This characteristic lab finding occurs by day 4-5 into onset of symptoms
- WNV: positive CSF and/or serum West Nile PCR (send both)

THERAPEUTIC INTERVENTIONS

General

- Non-invasive ventilatory support: If neuromuscular respiratory weakness with symptoms without criteria for ET
- Consider endotracheal intubation if: hypoxemia, rapid and shallow breaths, paradoxical breathing, VC < 20 ml/kg, NIP > - 30 cmH₂O and/or MEP < 40 cm H₂O (20/30/40 rule); Consider intubation when a patient who initially presents with hypocapnia from hyperventilation, becomes normocapnia during ongoing weakness, which is a sign of impending respiratory failure
- Supportive treatment: IV fluids, correct electrolyte disturbances, infection prevention, LW heparin, gastric ulcer prophylaxis, O₂, if needed

Specific

- Guillain-Barre Syndrome (GBS): IVIg or plasma exchange, no benefit of steroids

- Myasthenia gravis (MG): IVIg or plasma exchange; High dose of steroids (caution in non-intubated patients), consider holding pyridostigmine temporarily if intubation is required
- WNV or poliomyelitis: No proven effective therapy, continuous supportive care
- GBS or Myasthenic Crisis (MG with respiratory failure)
 - Intravenous Immunoglobulin 2g/kg over 3 to 5 days; Check IgA levels, as IgA deficiency can lead to anaphylaxis; Contraindications: History of venous thromboembolism and active coronary artery disease. Adverse effects: Aseptic meningitis, renal failure, rash, TRALI
 - Plasmapheresis: Central venous access required; Consult transfusion medicine; Average 4-6 treatments scheduled daily or every other day; Hold any ongoing ACE inhibitors; Obtain daily fibrinogen, CBC and ionized calcium levels. Adverse events: TRALI, transfusion-associated bloodstream infection, hypotension, muscle spasms

ONGOING TREATMENT

- GBS: Daily abdominal auscultation to monitor for bowel silence; If the patient develops a paralytic ileus, give erythromycin or neostigmine
- MG: CT chest for thymoma or enlargement of thymus gland
- DVT prophylaxis: Subcutaneous fractionated or unfractionated heparin and support stockings
- Neuropathic pain: Gabapentin, pregabalin, carbamazepine
- Respiratory therapy assessment every 4-6 hours: NIF, MEP and VC
- Rehabilitation: Acute phase should include gentle strengthening, involving isometric, isotonic, isokinetic, and manual resistive and progressive resistive exercises

CAUTIONS

- **Complications:** Residual weakness, numbness or tingling, fatigue, DVT, neuropathic pain
- **Caution**
 - Be aware of the numerous medications that could potentially exacerbate myasthenia
 - Too much pyridostigmine can also cause myasthenic patients to present with weakness. This anticholinergic crisis can be differentiated from a myasthenic crisis by the presence of increased secretions, cramps, fasciculations and diarrhea
 - GBS patients can have autonomic instability in up to 70% of patients. **Autonomic disturbances can result in cardiac arrhythmias, blood pressure fluctuations, ileus, urinary retention**

REFERENCES & ACKNOWLEDGMENTS

Acknowledgement: *Marcus V Pinto, MD, MS; Joseph Zachariah, DO; Sara Hocker, MD; Alejandro A. Rabinstein, MD*

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