Guillain-Barre Syndrome

Acute idiopathic inflammatory demyelinating polyneuropathy characterized by ascending progressive muscle weakness and areflexia, usually associated with spontaneous remission.

ANESTHETIC CONSIDERATIONS:

- **Autonomic Dysfunction**
  - including cardiac dysrhythmias & sudden death
  - epidural preferred vs. SAB for autonomic dysfunction
- Neuromuscular weakness
  - **aspiration risk** with depressed bulbar reflexes, paralytic ileus
  - **respiratory failure** secondary to respiratory muscle weakness
- Altered pharmacology
  - **Avoid SCh** - hyperkalemia response to SCh 2° to up-regulation of nicotinic ACh receptor
  - NMB - increased sensitivity 2° to loss of motor units and channel blockades at NM junction
  - Sensitivity to LA with neuraxial / regional / local anesthesia
  - Alpha-adrenergics cause exaggerated response given autonomic dysfunction

ANESTHETIC GOALS:

- Prevention of complications of autonomic dysfunction (profound hypotension or hypertension, asystole / arrhythmias, sudden death)
- Aspiration prevention
- Delivery of safe anesthetic given altered NMB response

HISTORY

- Standard anesthetic Hx
- Course of illness (duration, progression, severity, Tx and current symptoms)
- Baseline neurological function
- Autonomic dysfunction (postural hypotension, syncope, GERD, early satiety, or dysrhythmias - palpitations)
- Bulbar dysfunction (dysphagia, bilateral facial weakness, poor airway maintenance in supine position) = aspiration risk

PHYSICAL

- **HEENT** – dry eyes
- **CVS** – orthostatic BP / pulse (autonomic dysfunction)
- **RESP** - respiratory exam should focus on VC, adequacy of respirations (air entry & chest excursion), signs of pneumonia, and cough strength
- **GI** – bowel exam (risk of bowel obstruction)
- **CNS** – distal limb sensory deficits (absence of sensory level)
- **MSK** – symmetric flaccid paralysis or weakness of lower limbs, trunk, upper limbs

INVESTIGATIONS

- **Labs**
  - CBC, lytes, BUN, Cr for evaluation of active infection, increased potential for renal dysfunction, and SIADH
  - LFTs for increased potential for liver dysfunction
  - Coags if plasmapheresis considered
  - ABGs
- **Imaging**
  - ECG for autonomic dysfunction
  - CXR for respiratory status and pneumonia
- **Special**
  - PFTs (esp. vital capacity)
  - Special tests for diagnosis of GBS (anti-GM antibody, CSF proteins w/ normal cell count, neurological studies, and infections agent testing)

OPTIMIZATION

- Neurology consult to document baseline neurological status and direct care
- Aspiration prophylaxis
- IVIG or plasmapheresis Tx
- DVT prophylaxis

ANESTHETIC OPTIONS

- Consider extreme autonomic dysfunction
- Consider lower motor neuron lesions
- Local, regional, neuraxial, GA, with goal to minimize autonomic impact
- Altered physiologic response to anesthetic drugs:
  - NMB:
    - SCh – hyperkalemia response to SCh 2° to up-regulation of nicotinic ACh receptors
    - NDNMB - increased sensitivity 2° loss of motor units and channel blockades at NM junction
  - Alpha-adrenergics cause exaggerated response given autonomic dysfunction
- RA is not contraindicated, but GBS pts are sensitive to LA 2° to the presence of Na-channel blocking factor
- Epidural better than SAB b/c of autonomic instability and speed of onset of RA

ANESTHETIC SETUP

- **Drugs**
  - Standard w/o NMBA if possible
  - Pressors / dilators for autonomic dysfunction
- **Equipment**
  - CAS w/ 5 lead ECG for autonomic dysfunction

- PNS
- A-line b/c of severe autonomic dysfunction
- Others as per pt co-morbidities / procedure

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Consider IV preload for autonomic dysfunction
  - Consider **modified RSI w/out SCh** for aspiration risk
  - Consider need for NMB and altered response to NDNMB vs. NM free anesthetic
- **Maintenance**
  - Tight volume control (IV fluids, insensible losses, 3rd spacing, and blood loss) for autonomic dysfunction
  - Minimize peak airway pressures for autonomic dysfunction
  - Tight temperature control for autonomic dysfunction
  - Minimize positioning changes for autonomic dysfunction
- **Emergence**
  - Ensure NMB reversal

DISPOSITION & MONITORING

- Requires post-op step-down or ICU monitoring for mechanical ventilation (even if pre-op not ventilated)
- Neurology f/u

COMPLICATIONS

- Severe HD instability and sudden death 2° autonomic dysfunction
- Respiratory failure 2° respiratory muscle weakness
- Aspiration 2° poor airway tone & reflexes + inability to clear airway
- Altered response to NMB and alpha-adrenergics
- Thromboembolic events 2° to immobilization
- Fecal impaction
- Stress ulcers
- Prolonged ICU care and resources

PREGNANCY

- During 3rd trimester, risk of exacerbation
- For labor a regional anesthetic indicated to avoid exaggerated hemodynamic response to pain from autonomic dysfunction
- Aspiration pneumonitis and respiratory failure may result in premature labor and maternal mortality
- For C/S a regional anesthetic contraindicated even for patient with mild respiratory involvement

PATHOPHYSIOLOGY

- GBS is an acute demyelinating polyneuropathy which typically occurs as an autoimmune response approximately 1 month following a GI or respiratory infection
- Most common cause of acute generalized paralysis
- Assoc infectious agents:
  - Campylobacter jejuni (most common & assoc w/ axonal degeneration as well as demyelination which results in more severe debilitating form of disease)
  - Mycoplasma pneumoniae
  - EBV
  - CMV
  - HIV
- Epidemiology:
  - Incidence = 1 in 33,000 per year in Western world
  - All ages affected w/ slight bimodal distribution in young adults and elderly
  - Men > Women
  - Children affected less severely
- Types of GBS:
  - Acute inflammatory demyelinating polyradiculopathy (AIDP) (most common type seen in developed countries)
  - Acute motor axonal neuropathy (AMAN) (most common seen in developing countries - c. jejuni type)
  - Acute motor sensory axonal neuropathy (AMSAM) (similar to AMAN but w/ more sensory component and more prolonged)
  - Miller Fischer syndrome (ataxia, areflexia and ophthamoplegia which may be assoc w/ other symptoms)
- Clinical features:
  - Progressive motor weakness ascending from legs w/ proximal > distal muscles
  - Areflexia
  - Facial palsy and bulbar weakness
  - Ophthalmoplegia
  - Sensory symptoms (often gloves and stockings)
  - Severe pain (often girdle area)
  - Weakness of respiratory muscles leading to respiratory failure
  - Autonomic dysfunction causing under or over activity of SNS & PNS leading to:
    - Arrhythmias and sudden death (case reports of asystole after eyeball pressure, carotid sinus massage, and suctioning)
    - Postural hypotension (case reports assoc w/ head lifting from pillow) and hypertension
    - Resting tachycardia
    - Profuse diaphoresis
    - Peripheral vasoconstriction
    - Urinary retention
    - GI ileus
- Pathogenesis:
  - Segmental demyelination may be the only pathologic process w/ recovery occurring w/in several weeks
  - Nodes of Ranvier and Wallerian degeneration cause axonal damage and result in longer, more severe course of illness

- Na-channel blocking factor has been identified in the CSF of GBS pts which could contribute to the paralysis
- Tx:
  - Supportive:
    - ETT for bulbar weakness and inability to protect A/W
    - Mechanical ventilation for vital capacity < 15 ml/kg (25% of pts)
    - Careful BP control for autonomic dysfunction
    - Tube feeds for nutritional support
    - APS / CPS for neuropathic pain of proximal muscles and immobility pain
    - DVT prophylaxis
  - Dz modifying:
    - IVIG (0.4 mg/kg OD x 5 days)
    - Plasmapheresis (5 exchanges substituting 250 mL/kg of plasma w/ 5% albumin)
    - **Steroids ARE NOT useful**
- Prognosis:
  - Normally long progressive course w/ slow gradual recovery taking months
  - 10% die from complications
  - 10% suffer long term neuro morbidity and physical dependence (i.e.- unable to walk unaided)
  - Adverse outcome is assoc w/:
    - C. jejuni infection
    - Old age
    - Mechanical ventilation
    - Poor nerve studies
    - Abnormal CSF proteins

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