I. Guillain-Barré – autonomic dysfxn

Definition

Guillain-Barre Syndrome is characterized as an acute inflammatory demyelinating peripheral polyneuropathy secondary to an autoimmune response in association with an infectious process. Classically the syndrome is described as an ascending paralysis, starting in the legs and progressing cephalad. There are multiple variants of GBS; ascending weakness may or may not be accompanied by dysesthesias or in the most severe form autonomic dysfunction.

Autonomic Dysfunction is a common complication of GBS and is very important to identify early. Both the sympathetic & parasympathetic nervous systems may be involved. The most commonly involved organ systems are cardiovascular, gastrointestinal and sudomotor. **The cardiovascular complications are particularly important as they can be life threatening. Common manifestations include hypertension, hypotension, and brady/tachyarrytmias**. Identifying patients can be life saving in that some patients may require pacemaker placement. Gastrointestinal symptoms are less likely to be life threatening; however, they can delay diagnosis. Patients often experience constipation or diarrhea, abdominal bloating, urinary retention or bladder dysfunction that can masquerade as a viral illness. With respect to sudomotor involvement, some GBS patients have been shown to have decreased innervation of sweat glands. In and of itself, this is not necessarily lethal, but it does indicate that other small nerve fibers may be diseased in those patients.

PubMed

1. D W Zochodne Autonomic involvement in Guillain-Barré syndrome: a review. Muscle Nerve: 1994, 17(10);1145-55
2. Chun-Liang Pan, To-Jung Tseng, Yea-Huey Lin, Ming-Chang Chiang, Whei-Min Lin, Sung-Tsang Hsieh Cutaneous innervation in Guillain-

Barré syndrome: pathology and clinical correlations. Brain: 2003, 126(Pt 2);386-97

Succinylcholine in Guillain Barre Syndrome

Should be avoided due to a significant risk of hyperkalemia.

Ferguson et al described four patients with chronic/relapsing polyneuropathy who developed life-threatening arrhythmias following succinylcholine administration, although in this instance SCh was presumed, but not documented to be the cause (1). Reilly and Hutchinson described a case (2) in which a 51-year-old man developed unstable V-tach leading to cardiac arrest (K increased from 4.3 to 8.6) and death following SCh administration for an intubation during a GBS relapse. Most recently an occurence was documented in Belgium (3). Note that this risk may persist even after the disorder has run its course, as has been documented in at least one case report (4), in this case of a pregnant woman one month post-recovery. Vecuronium and rocuronium, both of which have minimal cardiovascular effects are recommended. Unfortunately, these patients may be either overly or under sensitive to non-depolarizing NMBDs.

Other Anesthetic Considerations in Guillain Barre

Impaired swallowing and ventilatory difficulty are common. Vital capacity should be assessed frequently, and if < 15 cc/kg, mechanical intubation may be indicated. There is a substantial correlation between the rapidity of paralysis and the need for mechanical ventilation, with rapidly deteriorating patients being more likely to require support.

Cardiovascular System

Autonomic dysfunction can create wide swings in cardiovascular variables, thus alpha and beta blockade may be indicated; however, some patients will manifest by lack of compensatory responses and will overreact to positional changes, blood loss, etc (and may require pressers).

PubMed

1. R J Fergusson, D J Wright, R F Willey, G K Crompton, I W Grant Suxamethonium is dangerous in polyneuropathy. Br Med J (Clin Res Ed): 1981, 282(6260);298-9
2. M Reilly, M Hutchinson Suxamethonium is contraindicated in the Guillain-Barré syndrome. J. Neurol. Neurosurg. Psychiatr.: 1991, 54(11);1018-9
3. J E Dalman, W I Verhagen Cardiac arrest in Guillain-Barré syndrome and the use of suxamethonium. Acta Neurol Belg: 1994, 94(4);259-61

J M Feldman Cardiac arrest after succinylcholine administration in a pregnant patient recovered from Guillain-Barré syndrome. Anesthesiology: 1990, 72(5);942-4

II. Multiple sclerosis: Periop mgmt

Definition

Multiple Sclerosis is an autoimmune disease of inflammation, demyelination, and axonal damage to the central nervous system . The disease progression may be subacute with relapses and remissions or chronic and progressive.

Treatments include corticosteroids, Interferon-beta, glatiramer acetate, azathioprine, and low-dose mexthotrexate. Although exacerbations can be triggered by physical and emotional stress, exacerbations and remissions often occur unpredictably.

In the pre-operative evaluation, a thorough baseline neurologic history and exam should be performed. Patients on corticosteroid therapy should continue therapy and may require stress dosing.

The anesthesiologist should closely monitor temperature and minimize increases above baseline as even slight increases in body temperature (increases as little as 1 C) may precipitate a decline in neurologic function postoperatively.

Succinylcholine - denervation may increase the risk of succinylcholine-induced hyperkalemia in these patients. Nondepolarizing neuromuscular blockers are safe to use although patients with multiple sclerosis may have altered sensitivity to these drugs in the setting of baseline limb weakness.

They may also have limited ‘physiologic reserve’ (neurologic and respiratory) and be less able to tolerate stressors such as a mild degree of post-operative residual muscle relaxant (Dorotta, 2002).

Some multiple sclerosis patients, such as those with baseline weakness or pharyngeal dysfunction, will require extended monitoring and care postoperatively.

As in other patients with chronic brain injury, patients with MS may be expected to have some MAC reduction and delayed emergence proportionate to the severity of their disease.

In terms of regional anesthesia, both spinal and epidural anesthesia has been successfully employed in parturients with multiple sclerosis (Perlas, 2005). In some studies, spinal anesthesia has been implicated in postop exacerbations whereas epidural and peripheral nerves blocks have not. One theory is that demyelination of the spinal cord makes it more susceptible to the neurotoxic effects of local anesthetics and the concentration of local anesthetic in the white matter of the spinal cord is higher following a spinal compared to an epidural. There is very little evidence to support or refute the concerns regarding spinal anesthesia (Martucci et al., 2011).

Finally, regardless of anesthetic technique, worsening of multiple sclerosis symptoms is experienced by 20-30% of women in the post-partum period. Whether this is due to a reversal of the pregnancy-induced 'immunotolerant' state or other factors is not entirely clear (Dorotta, 2002).

Multiple sclerosis: exacerbation

According to Miller, "*It has been speculated that general anesthesia and surgery may increase the risk for aggravation of MS. At present, there is no general consensus on this matter, and patients should therefore be informed of the potential for aggravated symptoms in the postoperative period*", and "*There has been no documented association with the type of anesthetic or particular anesthetic agents and exacerbation of disease*" (p 1031-21). According to Stoelting's Co-Existing Disease, the suggested reason that spinals (and not epidurals) have been implicated in exacerbation is that CNS demyelination may render the spinal cord overly sensitive to local anesthetics – epidural anesthestics result in lower concentrations of LA in the white matter.

Multiple Sclerosis Anesthetic Considerations / Concerns

- Possibility of exacerbation should be mentioned, although this is speculative at best
- No anesthetic technique has been shown to be superior to another, although spinals may exacerbate symptoms and are not recommended
- Impaired autonomic function
- High sensitivity to physical and emotional stress
- *May* be susceptible to hyperkalemia following SCh
- Avoid hyperthermia
- Have a low threshold for ICU admission post-operatively

III. Myasthenia: physiology

Definition

Myasthenia includes three groups of autoimmune disorders (myasthenia gravis, congenital myasthenic syndromes, Lambert-Eaton myasthenic syndromes) that affect the transmission of signals at the neuromuscular junctions that results in weakness and muscle fatiguability.

- With myasthenia gravis, ptosis, diplopia, dysphagia, and respiratory failure are often the initial symptoms. As the disease progresses, neck and limb-girdle muscle weakness becomes apparent. Diagnosis is confirmed with improvement in strength after intravenous injection of edrophonium. Thymectomy is treatment for young patients. Acetylcholinesterase inhibitors are used to treat symptoms, and plasmapharesis and IVIG are sometimes used to treat crises or to optimize preoperatively. MG is usually caused by antibodies against the nicotinic acetylcholine receptor.
- The congenital syndrome is extremely rare, and caused by inherited mutations in the synaptic vesicles, acetylcholinesterase, or nicotinic acetylcholine receptors resulting in either an increase or decrease in the response to acetylcholine. Immunosuppression and plasmapheresis are not effective treatment strategies.
- Lambert-Eaton myasthenic syndrome is an acquired disorder resulting from autoantibodies targeting the presynaptic voltage-gated calcium channels causing decreased Ach release. It causes proximal muscle weakness in the lower and upper extremities, fatigability, and autonomic dysfunction (but not bulbar or respiratory weakness).

Myasthenia gravis preop risk eval

Definition

Myasthenia gravis is an autoimmune disease that attacks post-synaptic nicotinic acetylcholine receptors at the NMJ. This leads to varying degrees of muscle weakness and fatigue and can affect ocular, bulbar (muscles involved in speech, chewing and swallowing), respiratory, and proximal skeletal muscles. Symptoms seem to be worse at the end of the day or after exertion. Classification of myasthenia depends on whether the patient has only ocular or ocular and non-ocular weakness.

These patients can pose a challenge during anesthesia not only because of their disease process but also from the medications used to treat them (acetylcholinesterase inhibitors, steroids, etc.). Preoperative evaluation should consider the recent course of the disease, muscle groups affected, drug therapy, and coexisting diseases. Perioperative management may also require a neurology consult.

- Patients should take their morning dose of acetylcholinesterase inhibitor though doing so may result in:
- Altered patient drug requirements following surgery
- Increased vagal reflexes
- Possibility of disrupting bowel anastomoses secondary to hyperperistalsis
- Significant side effects including salivation, miosis, bradycardia and even cholinergic crisis if given NMB reversal

Patients with involvement of respiratory or bulbar muscles tend to have a higher risk for aspiration so pretreatment with metoclopramide or H2 blockers may be helpful. Some MG patients are sensitive to respiratory depressants and premedication with opioids, BZDs, or barbiturates should be considered carefully or not at all.

Patients should be made aware that they might remain intubated after procedure is finished and will need to be awake prior to extubation. Predictive risk factors for post-operative ventilation support following thymectomy:

- Disease duration > 6yrs

- Concomitant pulmonary disease
- Peak inspiratory pressure < -25 cmH2O
- Vital capacity < 40 mL/kg
- Pyridostigmine dose > 750 mg/d

A steroid listed on a patient's medication profile indicates a higher risk and elective procedures should be postponed if possible.

Of note, IV pyridostigmine is 1/30 of the PO dose.

IVIG or plasma exchange can be used in emergent cases.

Diseases associated with MG include DM, thyroid disorders, SLE and RA.

Because there are functionally fewer nicotinic receptors, patients with MG are resistant to succinylcholine and are exquisitely sensitive to non-depolarizing NMBs.

NMBDs are further complicated by pyridostigmine (which can decrease the efficacy of SCh and non-depolarizing agents), and chronic steroid therapy, which can produce resistance to non-depolarizing agents (at least vecuronium). Thus, quantitative twitch monitoring and careful titration is prudent.

Myasthenia gravis: postop management

Definition

Ensure that the patient is reminded prior to induction of the possibility of a prolonged intubation.

Extubation: performed on awake patients and hopefully close to his/her baseline status. Reinstitute anticholinesterase medication, either by IV infusion or by reimplementation of the patient's oral regimen.

Leventhal criteria: Predictive scoring system for the need for postoperative ventilation

1) duration of disease for 6 years or longer

2) chronic comorbid pulmonary disease

3) pyridostigmine dose >750 mg/d

4) VC <2.9L

5) Other indicators include preoperative use of steroids, and previous episode of respiratory failure.

These predictors have not been widely validated. (1)

Drugs to avoid: Calcium Channel blockers, Magnesium, Aminoglycoside antibiotics as all of these may contribute to muscle weakness

Post-Op Bed: Patients should be monitored in either a ICU or step-down unit but NOT to a conventional surgical ward.

Myasthenic Synd vs Myas Gravis: Sx

Definition

Myasthenic syndrome and myasthenia gravis are two disorders of neuromuscular transmission that result in muscle weakness and fatigue. They differ in pathophysiology and their symptoms. **Myasthenia gravis (MG) is an autoimmune disorder caused by circulating antibodies that block acetylcholine receptors at the postsynaptic neuromuscular junction (NMJ).**

Myasthenic syndrome (sometimes referred to as Lambert-Eaton syndrome) is an autoimmune disorder where there are antibodies formed against the voltage-gated calcium channels in the NMJ. Roughly 60% of patients with LEMS have an underlying malignancy,

most commonly SCLC making this a “paraneoplastic syndrome.” Symptoms of LEMS improve with successful treatment of the underlying malignancy. Weakness in LEMS involves the arms and legs; with leg involvement more pronounced than in MG. Proximal musculature (hip and shoulder girdle) is predominately affected making climbing stairs difficult. Unlike MG eye muscle weakness is uncommon. 75% of LEMS patients have involvement of the autonomic nervous system resulting in ataxia, impaired sweating, orthostatic hypotension, and metallic taste in mouth.

A key difference between LEMS and MG is that the weakness and reflexes on neurologic examination improves with repeated testing