Neuromuscular Disease in the Neurointensive Care Unit

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KEYWORDS

• Neuromuscular diseases • Acute inflammatory demyelinating polyneuropathy
• Guillain-Barre syndrome • Amyotrophic lateral sclerosis • Myasthenia gravis
• Critical illness polyneuropathy • Critical illness myopathy

KEY POINTS

- Knowledge about the management of critically ill patients with neuromuscular disease is required for anesthesiologists, because these patients are frequently encountered in the intensive care unit and operating room.
- Acute inflammatory demyelinating polyneuropathy, that is, Guillain-Barre syndrome, is a rapidly progressive peripheral neuropathy that may affect multiple organ systems. Mainstay therapies include intravenous immunoglobulins or plasma exchange.
- The most common cause of death from amyotrophic lateral sclerosis is respiratory failure. This disease requires special attention to airway management and prevention of aspiration.
- Myasthenia gravis encompasses multiple known subtypes, all of which respond differently to the variety of available treatments. If symptomatic treatment with an acetylcholinesterase inhibitor is unsatisfactory, immunosuppressive therapy should be started.
- Critical illness polyneuropathy and myopathy are conditions of significant morbidity, including prolonged mechanical ventilation. Management is mainly preventative and supportive.

NEUROMUSCULAR DISEASE IN THE NEUROINTENSIVE CARE UNIT

Neuromuscular diseases are different syndromes that affect nerve, muscle, and/or neuromuscular junction (**Fig. 1**, **Table 1**). Afflictions encompassed in this category present a myriad of challenges for anesthesiologists in the operating room, pain clinic, and intensive care unit (ICU). These challenges and advances in management will be reviewed for neuromuscular diseases most commonly encountered.

This work is published in collaboration with the Society for Neuroscience in Anesthesiology and Critical Care.
Disclosures: None.
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Anesthesiology Clin 34 (2016) 601–619
http://dx.doi.org/10.1016/j.anclin.2016.04.010 **anesthesiology.theclinics.com**
1932-2275/16/$ – see front matter

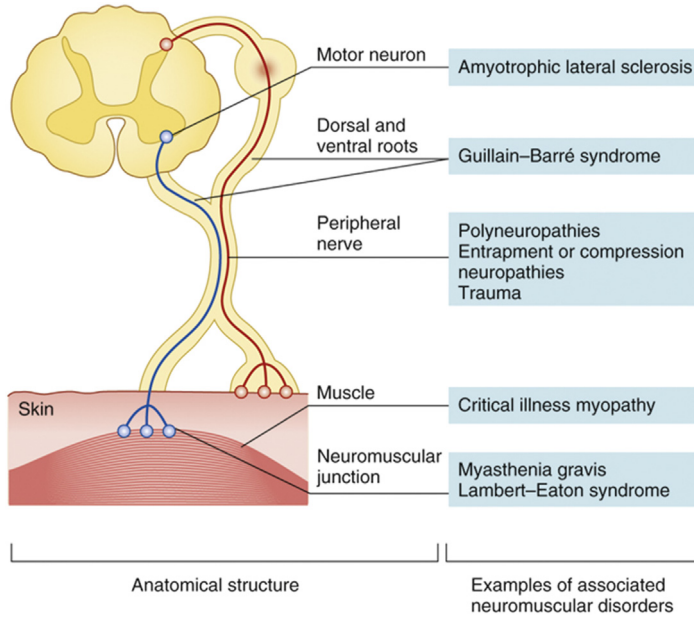

Fig. 1. Sites of neuromuscular disease in the spinal cord, peripheral nerves, and muscle. (*Adapted from* Horlings CG, van Engelen BG, Allum JH, et al. A weak balance: the contribution of muscle weakness to postural instability and falls. Nat Clin Pract Neurol 2008;4:504–15; with permission.)

Anesthetic Considerations

The anesthetic plan for any individual involves thoughtfully weighing risks against benefits, often from a range of choices. When the potential risks and benefits of possible interventions are not well known or misunderstood, decisions regarding the appropriate course of action are fraught with the possibility of unintended consequences. One such instance is the decision to provide general versus neuraxial or regional anesthesia to a patient with neuromuscular disease. General anesthesia with endotracheal intubation provides unconsciousness with a secure airway. However, general anesthesia often requires the use of neuromuscular blocking agents (NMBA); thus, removal of the endotracheal tube after the end of surgery may be challenging in patients with advanced neuromuscular disease, especially those with pre-existing respiratory or bulbar involvement. Neuraxial techniques generally preclude endotracheal tube placement but may cause profound respiratory impairment in patients with neuromuscular disease with only minimal involvement of accessory respiratory muscles. Perhaps the greatest concern regarding regional (including neuraxial) anesthesia in this patient population is the uncertainty of its effects on disease progression and symptoms. Regional anesthesia has been documented as safe and without neurologic sequelae in the setting of pre-existing neuromuscular disease by multiple sources.[1–3] However, neuromuscular disease exacerbations have been reported, although possibly influenced by a variety of confounding factors.[3–5] A causal link between regional anesthesia and neuromuscular disease exacerbation has not been established.

Regardless of anesthetic type, patients with neuromuscular disease may present to the ICU after surgery and anesthesia. Preoperative optimization of the patient may greatly decrease the need for postoperative mechanical ventilation and improve