Neuromuscular disorders: relevance to anaesthesia and intensive care

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Abstract

Neuromuscular diseases are relatively rare but it is important for both anaesthetists and intensivists to have a working knowledge of the common diseases, as they may complicate general anaesthesia and result in neurogenic respiratory failure. The most common neurological condition seen in the intensive care unit is that of critical illness neuropathy; this subject is covered elsewhere in the journal. The diseases most commonly encountered in general anaesthetic practice include motor neurone disease, Guillain-Barré syndrome, botulism, myasthenia gravis and the muscular dystrophies.

Keywords Anaesthesia; botulism; Guillain-Barré; motor neurone disease; muscular dystrophy; myasthenia gravis; neuromuscular disease; neuromuscular junction

Royal College of Anaesthetists CPD Matrix: 3F00

Learning objectives

After reading this article, you should:

- have a working knowledge of the anaesthetic and intensive care unit (ICU) implications of the more common neuromuscular diseases
- understand that, although relatively rare, neuromuscular disorders can cause respiratory and bulbar failure
- know that Guillain-Barré syndrome is the most common form of generalized paralysis in the western world and a common cause of ICU admission

Common neurological disorders encountered in the intensive care unit (Table 1)

Anterior horn cell disease

The most common anterior horn cell disease worldwide is poliomyelitis but effective vaccination programmes have made this condition rare in the UK; where motor neurone disease is more common.

Motor neurone disease (MND)[1]

The aetiology of this progressive pre-junctional disorder is unknown but degeneration of motor neurones in the motor cortex, brainstem nuclei and the anterior horn cells of the spinal cord characterize it. Prevalence is 6/100,000 of population, peaking at 50–70 years of age and affecting more men than women. It can affect the upper motor neurones (primary lateral sclerosis), the lower motor neurones (progressive spinal atrophy) and both (as in amyotrophic lateral sclerosis). Loss of muscle innervation leads to muscle atrophy and extra-junctional acetylcholine receptors. Ninety per cent of patients die within 5 years of diagnosis.

Clinical features: presentation is variable, though brisk reflexes are seen in all forms. In the lower motor neurone variant, patients classically present with fasciculation, weakness and atrophy of the small muscles of the hand, lumbar symptoms (foot drop). The upper motor neurone form typically presents with spastic weakness of limbs. There is no sensory loss or cranial nerve involvement. May also present with bulbar or pseudobulbar symptoms (dysphagia, dysarthria).

Management: domiciliary non-invasive ventilation is increasingly being used to manage patients with severe respiratory muscle weakness. Bulbar and respiratory muscle dysfunction are common leading aspiration pneumonia to be the most common cause of death.

Anaesthetic considerations: suxamethonium should be avoided and doses of non-depolarizing neuromuscular blockers reduced, due to increased sensitivity. Due to poor bulbar and respiratory muscle function, postoperative ventilatory support is commonly required. There is significant risk of pulmonary aspiration, infection, atelectasis and difficulty weaning from mechanical ventilation.

Peripheral nerve disorders

Guillain-Barré syndrome (GBS)[2,3]

GBS is an acute inflammatory polyneuropathy and the most common cause of acute neuromuscular paralysis. The incidence of GBS is 1–3/100,000 of the population and is more common in the young, elderly and in males (M:F 1.5:1). In two-thirds of

Common neurological disorders encountered in the intensive care unit

Anterior horn cell disease	Motor neurone disease
Peripheral nerve disorders	Guillain-Barré syndrome
Disorders of the neuromuscular junction	Lambert Eaton myasthenic syndrome
	Botulism
	Myasthenia gravis
Muscle disorders	Muscular dystrophies
	Myotonic syndromes

Table 1

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cases, the weakness is preceded by an upper respiratory or gastrointestinal tract infection. Possible infective triggers include *Campylobacter jejuni*, cytomegalovirus, *Myocoplasma pneumoniae*, Epstein–Barr virus and the human immunodeficiency virus. The peripheral neuropathy may be caused by damage to the myelin sheath or axon itself. It is thought to be immune mediated, with half of patients having detectable antibodies. Antibodies raised against the infecting agent are thought to cross-react with the host's neural tissue. The acute inflammatory demyelinating polyradiculoneuropathy (AIDP) form accounts for 95% of cases in Europe and the US, in which the myelin sheath is affected. Other forms targeting purely motor or sensory axons, or both, are rare.

Clinical features: the condition is classically progresses over hours to weeks and is characterized by a symmetrical, ascending neuropathic weakness accompanied by areflexia. Patients present with weakness of the limbs and a glove and stocking distribution paresthesiae. The legs are usually affected earlier than the arms and the weakness is more prominent in proximal muscles. In 90% of patients the weakness reaches its nadir within 3 weeks and recovers in 2–4 weeks after progression ceases. If weakness progresses after 4 weeks then a diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP) should be considered. Although 25% of patients remain mobile, approximately 30% of patients with GBS will require mechanical ventilation. Half of patients present with pain and affects 90% overall. The cranial nerves are affected in 75% of cases, causing facial and bulbar weakness and ophthalmoplegia. In the Miller Fisher variant of GBS, areflexia, ophthalmoplegia and ataxia co-exist; the limbs may be relatively spared. Autonomic features are seen in about 75% of patients, more commonly those severely affected such as those requiring ventilatory support. Manifestations include tachyarrhythmias, bradyarrhythmias, labile blood pressure, ileus, urinary retention and abnormal sweating.

Diagnosis: diagnosis is based on history, clinical features, examination of the cerebrospinal fluid (CSF) and neurophysiological studies. The essential and supporting criteria for diagnosis of GBS are listed in Table 2.

Management: this consists of immunotherapy to shorten the acute phase and supportive treatment on the intensive care unit. Immunotherapy is either given in the form of plasma exchange (PE) or intravenous immunoglobulin (IVIg), and should be started early. Plasma exchange usually consists of five single volume plasma exchanges over 7–14 days and requires central access, specialist equipment and specialist staff. Co-existing infection is a relative contraindication, as sepsis may be worsened. Intravenous immunoglobulin has equal efficacy to PE but is easier to administer and has fewer side effects. It is usually given at a dose of 0.4 g/kg/day for 5 days. No overall benefit has been found for corticosteroids in GBS, and if given via the oral route are probably harmful.

Intensive care management involves ventilatory support, treatment of autonomic instability, pain management and supportive care. Respiratory muscle weakness leads to atelectasis and failure of secretions clearance, whereas bulbar dysfunction can cause aspiration. Indications for ventilatory support are listed in Box 1. In patients likely to require prolonged periods of mechanical ventilation early tracheostomy is advised. Tracheostomy should also be performed if bulbar function is severely compromised. Respiratory weaning is unlikely to be successful until vital capacity is >15–20 ml/kg. Autonomic dysfunction usually occurs early and its peak coincides with peak weakness. Pain usually affects the back and legs and often requires both conventional and adjuvant analgesic drugs, such as gabapentin and carbamazepine.

Good supportive care includes early enteral feeding, avoidance of pressure sores, passive physiotherapy and venous thromboembolism prophylaxis. Lower respiratory tract and urinary tract infections are common.

Prognosis: the mortality of GBS is approximately 5% (7–20% in those mechanically ventilated), with lower rates seen in specialist units. Causes of death include respiratory failure, aspiration, ventilator associated pneumonia, sepsis and thromboembolic events. Although the majority of patients make an almost complete recovery within 1 year, about 20% remain severely disabled. Poor prognostic factors include increasing age, severity of disease and axonal disease.

Neuromuscular junction disorders[4,5]

These can be divided into pre-synaptic (e.g. Lambert Eaton myasthenic syndrome, botulism) or post-synaptic (e.g. myasthenia gravis).

Criteria for diagnosis of Guillain-Barré syndrome

Essential criteria	Supporting criteria
Progressive weakness of limbs due to neuropathy	Clinical features including motor weakness, mild sensory signs, cranial nerve involvement, autonomic involvement
Areflexia	Cerebrospinal fluid shows raised protein (>0.55 g/dl) after the first week with <10 white cells/ml
Duration of progression of weakness <4 weeks	Neurophysiological tests suggest either demyelination or axonal loss

Table 2

Criteria for invasive mechanical ventilation in GBS and myasthenia gravis[7]

Major criteria

- Signs of respiratory distress
- VC <15 ml/kg, PImax or PEmax <25 cmH_2O
- $PaCO_2$ >6.4 kPa
- PaO_2 <7.5 kPa (FiO_2 = 0.21)

Minor criteria

- Expectoration insufficient
- Severe bulbar dysfunction
- Atelectasis

Box 1

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