Neuromuscular disorders: relevance to anaesthesia and intensive care

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Abstract
Although relatively rare, neuromuscular disease is important to both anaesthetists and intensivists as it may complicate general anaesthesia and result in neurogenic respiratory failure. The most common diseases that will be encountered in a general anaesthetic practice include motor neurone disease, Guillain–Barré syndrome, botulism, myasthenia gravis and the muscular dystrophies. The clinical features and anaesthetic implications for these conditions are discussed.

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

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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

Most neurological diseases are relatively rare and therefore are not commonly encountered in general anaesthetic and intensive care practice. However, it is important to have a working knowledge of the more common disorders as they may directly influence anaesthetic management.

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. This article concentrates on a number of the more commonly encountered neurological diseases (Table 1).

Anterior horn cell disease

Although worldwide poliomyelitis is the most common anterior horn cell disease, effective vaccination programmes have made this condition rare in the UK; its incidence has therefore been superseded by motor neurone disease.

Motor neurone disease (MND)

MND is a progressive condition of unknown aetiology characterized by degeneration of motor neurones in the motor cortex, brainstem nuclei and the anterior horn cells of the spinal cord. Its prevalence is 6/100,000 of population with peak age of 50–70 years and men are more affected than women. Ninety per cent of patients die within 5 years of diagnosis. Presentation is variable. In the lower motor neurone variant, patients classically present with weakness, wasting and fasciculation of the small muscles of the hand. The upper motor neurone form typically presents with spastic weakness of limbs. Reflexes in all forms tend to be brisk. Bulbar and respiratory muscle dysfunction is common and aspiration pneumonia is the most common cause of death.

Anaesthetic problems relate to poor bulbar and respiratory muscle function with significant risk of pulmonary aspiration, the hyperkalaemic response seen with suxamethonium and the difficulty of weaning from mechanical ventilation. Patients with severe respiratory muscle weakness are being increasingly managed with domiciliary non-invasive ventilation.

Peripheral nerve disorders

Guillain–Barré syndrome (GBS)[1]

GBS is the most common cause of acute neuromuscular paralysis. It is characterized by a progressive, usually ascending neuropathic weakness accompanied by areflexia. Approximately 30% of patients with GBS will require mechanical ventilation.

The incidence of GBS is 1–3/100,000 of the population and is more common in the young and the elderly. In the majority of cases, the weakness is preceded by an upper respiratory or gastrointestinal tract infection. A number of viral and bacterial agents have been implicated, including *Campylobacter jejuni*, cytomegalovirus, Epstein-Barr virus and the human immunodeficiency virus. It is suggested that antibodies raised against the infecting agent cross-react with the host's neural tissue. Although the most common form of GBS is demyelinating, axonal forms exist.

Clinical features: patients present with weakness of the limbs and a glove and stocking type of parasthesiae. The legs are usually affected earlier than the arms and the weakness is more prominent in proximal muscles. The weakness reaches its nadir within the first week of the illness in most patients. The cranial nerves (especially the facial and bulbar nerves) are affected in 75% of cases. In the Miller Fisher variant of GBS, areflexia, ophthalmoplegia and ataxia co-exist; the limbs may be relatively spared.

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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

Thirty per cent of patients with GBS will require tracheal intubation and mechanical ventilation. Autonomic features are seen in about 75% of patients; sinus tachycardia is the most common manifestation but other, more dangerous bradyarrhythmias may occur.

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

Management: this consists of supportive treatment and specific therapy. The former consists of good nursing and medical care with careful monitoring especially during the acute phase when autonomic dysfunction may require rapid intervention. Patients may require prolonged periods of mechanical ventilation and tracheostomy should be performed early if this is likely; similarly, tracheostomy should be performed if bulbar function is severely compromised.

Other aspects of general care that should be meticulously followed are the early establishment of enteral feeding, regular turning of the patient to avoid pressure sores, passive physiotherapy with the use of limb splints if necessary and the use of prophylactic anticoagulation to help prevent thromboembolic complications. Pain is common and often requires both conventional analgesic drugs and those that specifically target neuropathic pain (e.g. gabapentin).

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

Specific treatment for GBS consists of high-dose intravenous immunoglobulin (IVIg) or plasma exchange. Both treatments have been shown to decrease recovery times if instituted early in the course of the disease. IVIg is favoured by many as it is easier to administer and has fewer complications than plasma exchange (e.g. sepsis). Corticosteroids have no role to play in the treatment of GBS.

Prognosis: the mortality of GBS is approximately 5% with lower rates seen in units specializing in its treatment. Although the majority of patients make an almost complete recovery within 1 year, a small proportion of patients (and especially those with axonal loss) may remain permanently disabled.

Neuromuscular junction disorders[2]

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

Lambert Eaton myasthenic syndrome (LEMS)

This rare autoimmune disorder is caused by IgG autoantibodies cross-reacting with the pre-synaptic voltage-gated calcium channels at the neuromuscular junction (NMJ). This results in a decrease in the mobilization and subsequent release of acetylcholine (ACh) from the pre-synaptic terminal; this, in turn, produces less depolarization at the post-synaptic membrane and a decrease in muscle contraction.

Clinical features: fifty per cent of cases of LEMS are associated with small cell carcinoma of the bronchus and therefore many patients have signs of associated malignancy. The clinical picture is one of weak and often tender proximal limb muscles in the presence of depressed tendon reflexes. In addition, autonomic dysfunction is almost invariable resulting in postural hypotension, gastrointestinal slowing and dry mouth.

Diagnosis: consists of detection of the antibody directed towards the voltage-gated calcium channel and characteristic electromyographic findings; the latter consist of an incremental response of the compound muscle action potential on tetanic stimulation which is due to increased mobilization of ACh at this rate of stimulation.

Management: the majority of patients with LEMS will be receiving 3:4 diaminopyridine, an agent that increases pre-synaptic ACh release. Removal of the tumour can result in remission. Furthermore, many patients will be taking immunosuppressive therapy. Plasma exchange and IVIg are short-term treatments for exacerbation of weakness.

Anaesthetic considerations include the need for hydrocortisone 'cover' during surgery, and the possible worsening of hypotension on initiation of positive pressure ventilation due to autonomic dysfunction. In addition, patients with LEMS show extreme sensitivity to both depolarizing and non-depolarizing neuromuscular blocking agents.

Botulism[3]

Botulism is the clinical syndrome caused by neurotoxins produced by the anaerobic organism *Clostridium botulinum*. Although seven different neurotoxins have been identified,

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