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Review

Complications of anaesthesia in neuromuscular disorders

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Abstract

The purpose of this review is to alert non-anaesthesiologists to the various complications from which patients with neuromuscular disorders and those susceptible to malignant hyperthermia can suffer during anaesthesia. The patient's outcome correlates with the quality of consultation between anaesthesiologists, surgeons, neurologists and cardiologists. Special precautions must be taken, since many anaesthetics and muscle relaxants can aggravate the clinical features or trigger life-threatening reactions. Complications frequently occur in these patients, although anaesthetic procedures have become safer by the reduced administration of suxamethonium and the use of total intravenous anaesthesia, new volatile anaesthetics and non-depolarising relaxants. This review provides a synopsis of pre-operative anaesthetic considerations and adverse drug effects on skeletal, cardiac and smooth muscle tissue. It describes the pathogenetic aspects of typical complications and introduces anaesthetic procedures for the various neuromuscular disorders, including regional anaesthesia for patients in whom a restriction of respiratory and/or cardiac function is predicted.

Keywords: Anaesthetics; Muscle relaxants; Rhabdomyolysis; Malignant hyperthermia; Myotonia; Neuromuscular disease; Cardiac complications

1. Introduction

Anaesthesia in patients with neuromuscular diseases is a concern for anaesthesiologists, surgeons, neurologists, pediatricians, cardiologists, pulmonologists and sometimes also for geneticists. It is desirable to discuss with the patient and family members the risks and benefits of the various treatment options. Often the anesthesiologist is not left with a single absolute risk, but in many cases must balance conflicting management strategies, fully bearing in mind the possible deleterious outcomes even with the chosen course of action. In order to reduce patient risk to a minimum, pre-operative considerations in respect of these circumstances and a precise diagnosis (often per biopsy) in advance are essential. It is also important to identify and treat potential anaesthetic complications promptly. Lastly, adverse drug effects or specific risks associated with certain neuromuscular diseases should be taken into account.

This review is an overview of these elements. It deals with the pre-operative anaesthetic considerations, the typical anaesthesia-related exacerbation of skeletal, cardiac and smooth muscle weakness resulting in respiratory distress, cardiac complications and autonomic dysregulation. Adverse drug effects such as rhabdomyolysis, muscle spasms, malignant hyperthermia and similar reactions are discussed with respect to their pathogenesis. Afterwards, typical anaesthetic complications and their prevention and management are described for specific neuromuscular disorders: motoneuron diseases, peripheral neuropathies, neuromuscular transmission disorders, progressive muscular dystrophies, metabolic and mitochondrial myopathies, myotonias and periodic paralyses, and congenital myopathies (Table 1). Lastly, areas are indicated for non-anaesthesiologists to turn to, for more insight into the care of their patients. Further details may be found in [1].

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Table 1
Overview of anaesthetic considerations in neuromuscular diseases

Neuromuscular disease	Respiratory test	Cardiac exam	Volatile anaesth.	NDMR	Comments
Motoneuron diseases					
Amyotrophic lateral sclerosis	x		x	⇩	
Spinal muscular atrophy	x		(x)	⇩	
Neuropathies					
Inflammatory polyneuropathies					
Guillain–Barré syndrome	x	x	x	⇩	Note dysautonomia, consider PM
CIDP	(x)	(x)	x	⇩	
Hereditary polyneuropathies					
Charcot–Marie–Tooth syndrome	x	x	x	⇩	Decrease barbiturate
Friedreich's ataxia	x	x	(x)	x	
Toxic polyneuropathies		(x)	(x)	x	
Disorders of the neuromuscular transmission					
Myasthenia gravis	x		x	⇩	Increase Sx dose
Lambert–Eaton syndrome	x		x	⇩	Decrease Sx dose
Myopathies					
Progressive muscular dystrophies					
Myotonic dystrophy	x	x		⇩	Avoid aChE and respiratory depressants consider PM
Facioscapulohumeral dystrophy	(x)		x	x	
Limb girdle dystrophy		x		x	
Congenital myopathies	x	x		x	
Poly- and dermatomyositis	x	x	x	⇩	
Metabolic myopathies					
Glycogenoses	x	x	(x)	x	Difficult intubation, metabolic acidosis
Disorders of lipid metabolism	x	x		x	Avoid hypoglycaemia
Homozygous MAD deficiency		x		x	
Mitochondrial myopathies		x		x	Consider PM
Ion channel diseases					
Myotonia congenita				⇩	Avoid aChE
Potassium-sensitive myotonias and periodic paralyses	(x)			⇩	Avoid dyskalaemia, hypothermia, ache

x, may be used or should be performed; (x), x with restriction, Sx, suxamethonium; PM, pacemaker; aChE, anticholinesterases; NDMR, non-depolarising muscle relaxants; MAD, myoadenylate deaminase deficiency. Sx is contraindicated in all NMD except myasthenia gravis.

2. Pre-operative considerations

The most obvious pre-operative question is whether the benefit of surgery justifies the anaesthetic risk. Decisions are based on a clear diagnosis with a full workup including histopathological findings, genetic data, or at least family history, and a wider variety of metabolic tests. Since patients with neuromuscular diseases are very challenging and may appear deceptively healthy, it is important to document all choices and their rationale on the medical record for subsequent care-givers, future research in rare disorders, and for medico-legal protection of care-givers and institutions. The optimum result will only be achieved if the various disciplines involved work together closely.

In neuromuscular diseases, the thorough pre-operative examination should include the detection of associated cardiac and respiratory dysfunction [2,3]: a neurological examination for scoring neuromuscular disease symptoms, electrocardiogram, an X-ray of the thorax, echocardiography, pulmonary function tests with body plethysmography, arterial blood gas analysis, determination of serum Na^+, K^+, Cl^-, Ca^{2+}, Mg^{2+}, creatine kinase (CK), and myoglobin levels, and, in the case of respiratory distress, measurement of inspiratory muscle strength.

For pre-medication, substances leading to respiratory depression or decreased muscle tone are often are inadequate in severely affected individuals. In other cases, drugs such as benzodiazepines can be ideal, although only under close monitoring. A possible alternative could be clonidine at low dosage as this leads to sufficient anxiolysis without relevant cardiovascular side-effects. Regional or local anaesthetic techniques can be employed in patients with cardiac and/or respiratory dysfunction [4,5]. However, in patients with autonomic dysfunction, a potential sympathetic block resulting from regional anaesthesia requires careful control of blood pressure. In patients in whom volatile anaesthetics and regional techniques are not indicated, propofol and opioids are recommended; over the last 10 years they have replaced neurolept analgesia.