

Anaesthetics

Who should read this?

- Everyone who has a neuromuscular disorder, even if their symptoms are very mild
- Everyone who has, or had, a relative with a neuromuscular disorder
- Professionals involved with the care of people with neuromuscular disorders

Anaesthetics

People with neuromuscular disorders must take great care if they are to have a local or general anaesthetics. Even someone with very mild, or non-existent symptoms, or someone who has a family history of a disorder, needs to let the anaesthetist know well in advance so that tests can be carried out if necessary and proper care after the operation can be arranged.

Many people are afraid of having an anaesthetic, mainly through ignorance, but when we look at the rate of complications and even deaths arising from anaesthesia we see that it is, in fact, very safe. This safety is the result of a thorough understanding

of the patient's medical condition with a careful assessment before the operation, marked technical improvements in monitoring facilities during the operation, and the provision of good recovery facilities such as High Dependency Units (HDU) and Intensive Care Units (ICU).

People with neuromuscular disorders deserve special attention when it comes to anaesthesia because many of the agents used (gases and chemicals) have effects on both muscle and nervous tissue. The main areas of concern are how the anaesthetic agents will affect the muscle and nervous tissue including the heart, which is, itself a muscle. A skeletal deformity such as scoliosis, or curvature of the spine, can also affect the way a person responds to anaesthesia so it is important to consider that too.

Anaesthetics and the heart

People with Neuromuscular disorders could sometimes have associated heart disease. This can occur as a cardiomyopathy, when the heart muscle doesn't work effectively, or as a defect in the way the electrical activity of the heart is transmitted, a conduction defect. The anaesthetic vapours - the smelly agents such as ether and halothane - that are inhaled can reduce the effectiveness of the heart's muscle contractions and also aggravate any conduction defect. The vapours are all slightly different from each other, some having more effect on the heart

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than others. So it is important that the anaesthetist makes a good assessment of the heart's condition before the operation which would include the level of physical activity that the patient can manage, and an ECG. Occasionally a more extensive assessment is needed.

Anaesthetics and breathing

Doctors need to measure how weak the patient's muscles are, usually by assessing the amount of physical activity that the patient can perform, ordering lung function tests, and/or by taking a blood test to measure levels of a muscle enzyme, creatine kinase (CK). Any anaesthetics agent that affects the muscles will also affect the muscle we use to breathe. Strong analgesic or sedative agents will affect these muscles indirectly, and muscle relaxants will have a direct effect on them. As breathing (or respiration) may already be difficult for people with neuromuscular disorders, these drugs should be used cautiously, and monitoring of breathing after the operation is absolutely essential. As a result, the patient is usually best cared for in a High Dependency Unit or Intensive Care Unit immediately after the operation. The muscles used for swallowing can also be affected which is another reason why good post-operative care is important.

Muscle relaxants

Muscle relaxant drugs should only be used if essential because they tend to have a more profound and prolonged effect in people with neuromuscular disorders compared to other patients. One type of muscle relaxant, called suxamethonium, should usually be avoided. It causes the release of potassium ions (K⁺) from the muscle tissue into the blood. In normal patients this is usually of little practical significance. In patients with neuromuscular disorder the muscle may normally leak K⁺ so that a further increase in the levels of K⁺ in the blood

may cause abnormal heart rhythms. A pre-operative blood test to check K⁺ levels is therefore important.

Local anaesthetics

A local anaesthetic works by preventing the normal electrical activity in the nerve around which the anaesthetics agents are placed. For minor procedures, such as stitches for cuts, they are probably the first choice for people with neuromuscular disorders because they have few, if any, side effects. However for major local anaesthetics techniques, e.g. spinal or epidural, careful assessment of the patient is needed and the type of neuromuscular disorder considered well before the operation.

Changes in body temperature and pre-operative 'starvation'

People with neuromuscular disorders do not tolerate changes in body temperature or the starvation often associated with anaesthesia or surgery as well as normal patients, so steps need to be taken to minimize these problems by keeping the patient warm and well hydrated using drips.

Malignant hyperthermia (MH) and Central Core disease

Malignant hyperthermia (MH) is an inherited disorder, which causes an unexpected, sometimes fatal, reaction in the patient to certain anaesthetic drugs. Because some people with neuromuscular disorders have sometimes experienced similar problems during anaesthesia, there have been claims that people with neuromuscular disorders may also have MH. However, it is generally accepted that the only neuromuscular condition truly related to MH is Central Core Disease (CCD), although this is not always the case. People with CCD should be considered potentially susceptible to MH unless proved otherwise by a special type of muscle biopsy, which screens for MH.

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To sum up...

- Anaesthesia for people with neuromuscular disorders should not be undertaken lightly. People with these conditions should expect the anaesthetist to make a careful and thorough assessment of their particular condition and their current state of health.
- It is not usually suitable to be treated as day cases because doctors should carry out pre-operative investigations, and enough time and recovery facilities should be available after the operation.
- It is absolutely essential that the person affected by a neuromuscular disorder should inform the anaesthetist even if there are only minor symptoms or no symptoms at all. The anaesthetist should also be warned if there is an inherited neuromuscular disorder in the family.
- If possible, ask for the anaesthetist to be forewarned before admission to hospital.
- Consider wearing a Medic Alert bracelet or similar informational device in case of accidents.

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