Charcot-Marie-Tooth UK

Fact Sheet: Anaesthetics

Together we are Stronger

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CMT and Anaesthetics

Anaesthetics – general or local – are powerful drugs that have a particular effect on your nervous system and muscles. Because CMT is a condition that affects part of your nervous system, you need to take especial care that the anaesthetist understands that you have CMT and well in advance of any operation. This holds true even if you only have mild symptoms, no symptoms at all or have a family history of CMT.

By making sure you have told the anaesthetist well in advance, he or she can work out with you what type of pain relief is best for you. As an extra precaution, you will probably be asked to stay in hospital longer after the operation to make sure everything is okay, which is why people with CMT are often not able to have day treatment.

Help yourself:

Use this checklist to make sure you get the right treatment for you:

- Does the anaesthetist and the doctor carrying out the operation – usually a surgeon – and their team understand that you have CMT?
- Do they all understand what CMT is?
- Are they aware of the list of drugs that should not be used on people with CMT?
- Have you told them about all the drugs and treatments you are taking, including any over the counter treatments (eg cough mixtures) and any complementary or alternative remedies?
- Have they discussed with you all the possible options (including no treatment) and their benefits and risks? This is known as "informed consent" and the Department of Health recognizes that you have a "fundamental legal and ethical right to determine what happens to [your body]." This means that everything needs to be explained in a way that you feel comfortable with.

Anaesthetics

People with neuromuscular disorders (NMD) must take great care if they are to have a local or general anaesthetic. Even someone very mild 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 and proper care after the operation can be arranged.

Written by Dr.P.J.Halsall and Professor F.R.Ellis for the Muscular Dystrophy Campaign

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 NMD around operations or treatment under local anaesthetic.

Which are the neuromuscular disorders (NMDs)?

Neuromuscular disorders include all the muscular dystrophies plus: myotonic disorders; congenital myopathies including minicore, central-core and multi-core disease plus nemaline and myotubular myopathies; mitochondrial myopathies; lipid storage myopathies; inherited metabolic myopathies including glycogen storage disease; familial periodic paralysis; inflammatory myopathies including infective myositis; autoimmune myositides including polymyositis and dermatomyositis; spinal muscular atrophies; hereditary and idiopathic peripheral neuropathy (**HMSN also known as Charcot Marie Tooth disease**); inflammatory, autoimmune and toxic neuropathies including Guillain Barre syndrome and CIDP; disorders of the neuromuscular junction including myasthenia gravis.

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). Patients with neuromuscular disorders (NMDs) 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 how they will affect the heart that is itself a muscle. A

skeletal deformity such as scoliosis, or curvature of the spine, can also affect the way the patient responds to anaesthesia so it is important to consider that too.

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, and by taking a blood test to measure levels of a muscle enzyme, creatine kinase (CK). Any anaesthetic agent who affects the muscles will also affect the muscles 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 patients with NMDS, 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 NMD patients 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 NMD 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 anaesthetic agents are placed. For minor procedures, such as stitches for cuts, they are probably the first choice for patients with NMD because they have few if any side-effects. However for major local anaesthetic techniques, e.g. spinal or epidural, careful assessment of the patient is needed and the type of NMD considered well before the operation.

Changes in body temperature and pre- operative 'starvation'

Patients with NMD 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 minimise these problems by keeping the patient warm and well hydrated using drips.

To sum up ...

- Clearly anaesthesia in NMD is not to be undertaken lightly. Such patients should expect the anaesthetist to make a careful and thorough assessment of their particular condition and their current state of health.

- They are not 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 NMD should inform the anaesthetist even if there are only minor symptoms or no symptoms at all. Occasionally a neuromuscular disorder in a person who had no symptoms has come to light only because of an unexpected problem with anaesthesia, particularly in young children. The anaesthetist should also be warned if there is an inherited NMD in the family, even if the individual has no symptoms.

- If possible ask for the anaesthetist to be forewarned before admission to hospital and consider wearing a Medic Alert bracelet or similar in case of accidents.

- It is always a good idea to make sure hospital staff have copies of Fact Sheets about your condition, and if you are going to have an anaesthetic you could show your anaesthetist this Fact Sheet.

NEUROTOXIC MEDICATIONS

Before taking any medication, be sure to discuss it fully with your doctor or pharmacist for possible side effects. Ask them to look for the words "could cause peripheral neuropathy" in the drug description. In almost all the conditions in which these drugs are used, an alternative is available.

Drugs which are toxic to the peripheral nervous system and may be harmful to a person with CMT, but please note, with the exception on Vincristine and Taxols, there is little or no scientific data on the potential risk:

Further information can be found here: http://bit.ly/drugslist2015

Definite High Risk (including asymptomatic CMT)

- Vinca alkaloids (Vincristine)
- Taxols (paclitaxel, docetaxel, cabazitaxel)

Moderate to Significant Risk

- Amiodarone (Cordarone)
- Bortezomib (Velcade)
- Cisplatin & Oxaliplatin
- Colchicine (extended use)
- Dapsone
- Didanosine (ddI, Videx)
- Dichloroacetate
- Disulfiram (Antabuse)
- Eribulin Mesylate (Halaven)
- Gold salts
- Ixabepilone (Ixempra)
- Leflunomide (Arava)
- Metronidazole/Misonidazole (extended use)
- Nitrofurantoin (Macrodantin, Furadantin, Macrobid)
- Nitrous oxide (inhalation abuse or Vitamin B12 deficiency)
- Perhexiline (not used in U.S.)
- Pyridoxine (Although megadoses [10 times or more the RDA] of Vitamin B6 may be harmful, high intakes of vitamin B6 from food sources have not been reported to cause adverse effects.) NIH Fact Sheet
- Stavudine (d4T, Zerit)
- Suramin
- Thalidomide
- Zalcitabine (ddC, Hivid)

certain or Minor Risk

- 5-Fluoracil
- Adriamycin
- Almitrine (not in U.S.)
- Chloroquine
- Ciprofloxacin (Cipro)
- Cytarabine (high dose)
- Ethambutol
- Etoposide (VP-16)
- Fluoroquinolones
- Gemcitabine
- Griseofulvin
- Hexamethylmelamine
- Hydralazine
- Ifosphamide
- Infliximab
- Isoniazid (INH)
- Lansoprazole (Prevacid)
- Mefloquine
- Omeprazole (Prilosec)
- Penicillamine
- Phenytoin (Dilantin)
- Podophyllin resin
- Sertraline (Zoloft)
- Statins
- Tacrolimus (FK506, ProGraf)
- Zimeldine (not in U.S.)
- a-Interferon

Negligible or Doubtful Risk

- Allopurinol
- Amitriptyline
- Chloramphenicol
- Chlorprothixene
- Cimetidine
- Clioquinil
- Clofibrate
- Cyclosporin A
- Enalapril
- Gluthethimide
- Lithium
- Phenelzine
- Propafenone
- Sulfonamides
- Sulphasalzine

Charcot-Marie-Tooth UK asked our Neurological Advisor, Dr David Hilton-Jones, MD, FRCP, FRCPE, to review these for us and to give us his advice.

Dr Hilton-Jones says: "All of these drugs can cause nerve damage (ie. neuropathy). There is no evidence that having CMT makes you more susceptible to such damage (except in the case of Vincristine), even if that may seem an obvious possibility. I suggest:

1. Avoiding these drugs if a safer alternative is available and equally effective.
2. Not to panic if you have taken one of them for a serious problem.

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Supporting people affected by Charcot-Marie-Tooth Disease.

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