

National Hospital for Neurology and Neurosurgery

Neurophysiological testing in patients with suspected or confirmed muscle channelopathies

Centre for Neuromuscular Diseases

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1. Introduction

Your neurologist has recommended for you to have neurophysiological tests to help make a diagnosis of your condition; this may also help decide what the best treatment is for you.

This leaflet explains what nerve conduction studies (NCS) and electromyography (EMG) are, how they are performed, and what to expect. It will also discuss why these investigations are necessary in the diagnosis of a muscle channel disorder.

2. What are nerve conduction studies (NCS) and electromyography (EMG)?

Nerve conduction studies (NCS) involve the use of small electric currents which are applied to nerves. The electrical pulses are conducted by your nerves and this can elicit a sensation in part of your hand or foot and/or cause a muscle twitch. The electrical pulses are delivered through a stimulator placed on the skin and the responses from your nerves or muscles are recorded a short distance away from the stimulation via electrodes attached to the skin.

EMG stands for electromyography. This is the capture of electrical signals that occur in muscles either at rest or during muscle contraction. Usually this requires the insertion of a small needle into the muscles.

3. What are muscle channelopathies?

To be able to feel, hear, see and move our body we need nerves and muscles. The normal functioning of these tissues depends on their ability to conduct electrical signals. Tiny electrical currents occur at the membrane of muscles and nerves due to small pores that allow electrically charged ions (salt particles) to pass between the inside and outside of these cells.

In patients with muscle channelopathies one type of these pores in muscle tissue is defective allowing either too much or too little current to flow. Depending on which pore is affected this can manifest in a variety of symptoms including attacks of weakness or muscle stiffness.

4. What specific tests are performed for muscle channelopathies?

In muscle channelopathies there may be factors that trigger attacks or worsen symptoms. These include exercise and low temperature. We use a standardised protocol to test and monitor the changes in the electrical properties of a muscle (usually in the hand) in response to exercise and cooling.

5. How can this test help?

The electrophysiological tests can help in making a diagnosis of a muscle channelopathy and distinguish between different types of muscle channelopathies. This in turn is useful to direct further testing (e.g. genetic tests) and advise on treatments.

6. I have had nerve conduction studies and EMG testing before, do I need to have them again?

Yes, the majority of patients referred to the muscle channelopathy service undergo electrophysiological testing even if they had some tests done previously. Tests performed elsewhere often do not include specific exercise and cooling procedures and may not have been carried out to the standardised protocol that we use. At times the tests we use are also helpful in monitoring your condition and assess how effective your current treatment is.

7. What happens during the test?

The doctor or clinical physiologist performing the test will ask you to lie or sit on an examination couch. We will initially test the general function of your nerves and muscles with nerve conduction studies and EMG usually in one arm and one leg. We will then ask you to perform a little exercise with your hand. This involves repeated brief bouts of exercise (pushing with your little finger against resistance) and we will monitor the changes in the electrical properties of the hand muscle (short exercise test at room temperature). We will repeat this test after cooling the same muscle in your other hand by placing the side of your hand on a bag of ice for a few minutes (short exercise test after cooling). The final test involves a more prolonged exercise over five minutes followed by 50 minutes of monitoring the muscle properties by using an electrical pulse every one-two minutes (long exercise test, also called McManis test). The whole study usually takes between two- two and a half hours.

8. Are there any risks or side-effects?

This is a safe test with no major long-term risks or complications.

The insertion of the EMG needle (similar to an Acupuncture needle) into the muscle can cause mild discomfort and a small amount of bleeding or bruising. Very rarely more significant bleeding can cause a small blood clot. This can lead to local swelling, pain and discoloration but does not require any specific treatment and should improve with rest within a few days. We would usually expect you to be able to return to normal activities immediately after the test.

The use of electrical pulses during the test can cause discomfort but most people tolerate this well. Due to the possibility of interference with implanted electrical devices such as a cardiac pacemaker we need to use know about these in advance so that we can modify our test protocol accordingly.

9. What if I choose not to have NCS & EMG?

If you chose not to have the NCS and EMG it may be more difficult to confirm or exclude the muscle channel disorder.

10. Are there any alternative tests?

There are no alternatives to the NCS and EMG, however we may be able to investigate the suspected channel disorder through the genetic investigations. The neurophysiology can help guide our genetic investigations.

11. How do I prepare for the test?

- You can eat and drink and take your routine medication as normal unless we contact you and instruct you otherwise.
- Please avoid using any cream and moisturiser on your arms and legs on the day of the test.

• It also helps if you wear loose fitting clothing which reduces the need to undress for the test.

• You should receive a questionnaire asking whether you have an implanted electrical device (in particular a cardiac pacemaker or implantable cardiac defibrillator (ICD)) or whether you are on anticoagulation (blood thinning) treatment such as Warfarin or Heparin. Please ensure that we receive this information before your appointment so we can plan the test accordingly.

• If you have an allergy against latex or plaster tape let us know on the day of the test.

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12. Asking for your consent

We do not require formal consent for the NCS and EMG, we will discuss all the risks with you before the tests and if you are happy to proceed verbal consent is sufficient.

13. Receiving the results

Some of the findings may already be clear at the time of testing and the doctor involved may be able to explain these at the end of the study. You are welcome to ask any questions. If you are seen in the muscle channelopathy clinic the same or next day we will liaise directly with the clinical team to inform them about the results. You will have the opportunity to ask more questions about this or any other investigation in the muscle channelopathy clinic. A formal report will be issued within a few days of completing the test.

14. Contact details

National Hospital for Neurology and Neurosurgery Queen Square London WC1N 3BG

Switchboard: 0845 1555 000 Extension: 88009 Direct line: 0203 448 8009 Fax: 0203 448 3633

The test will be performed in the Department for Clinical Neurophysiology, 3rd floor Albany Wing within the main hospital building.

For general enquiries you can contact the neuromuscular nurse specialist on 0845 1555 000 extension 88009, between 9am and 5pm Monday to Friday. During working hours our answer phone is checked at least twice each day. Please leave a message with your name and a contact number and we will get back to you as soon as possible. For enquiries regarding booking/changing appointment and appointment times please contact the NCG Channel Coordinator on 0845 155 5000 extension 88030. Alternatively you can contact the administrative office of the Clinical Neurophysiology Department on 0845 155 5000 extension 84752.

15. Further information

You may find the following organisations useful:

Centre for Neuromuscular Diseases Website www.cnmd.ac.uk

Muscular Dystrophy UK www.musculardystrophyuk.org

Space for notes and questions

If you would like this document in another language or format, or require the services of an interpreter, please contact the clinical nurse specialist. We will do our best to meet your needs.



First published: June 2015

Date last reviewed: July 2018

Date next review due: July 2020

Leaflet code: UCLH/NHNN/MUS/MC-NEUROPHYSIOLOGY TESTING/1 © University College London Hospitals NHS Foundation Trust 2015

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