Muscle relaxation time as outcome measure in patients with non-dystrophic and dystrophic myotonic syndromes; voluntary handgrip- versus peripheral nerve stimulation myometry

Published: 09-01-2012
Last updated: 30-04-2024

In this study we want to investigate if the muscle-relaxation time (muscle-RT), calculated from force profiles obtained with two different experimental methods (1) voluntary handgrip- and (2) peripheral nerve stimulation myometry, is a reliable and...

Ethical review	Approved WMO
Status	Pending
Health condition type	Muscle disorders
Study type	Observational non invasive

Summary

ID

NL-OMON35579

Source
ToetsingOnline

Brief title
Muscle-RT measurements in DMs and NDMs

Condition

- Muscle disorders

Synonym
inherited muscle stiffness, Myotonia

Research involving
Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Sint Radboud
Source(s) of monetary or material Support: Ministerie van OC&W

Intervention

Keyword: Muscle relaxation time, Myometry, Myotonia, Outcome measures

Outcome measures

Primary outcome

Outcome measures are the mean muscle-RT and intraclass-correlation-coefficient (ICC). Differences in mean muscle-RT between the group of healthy volunteers and each of the patientgroups will be a measure for the reliability and robustness of the muscle-RT as outcome measure for myotonia. This reliability will be calculated for both methods within the different patientgroups. To determine the reproducability of these measurements within both methods, the ICC will also be calculated.

Secondary outcome

n.v.t.

Study description

Background summary

Myotonia is defined as an elongation in skeletal muscle relaxation after voluntary muscle contraction or percussion and derives from a hyperexcitabel state of the sarcolemmal membrane. Myotonia is the cardinal symptom in dystrophic (DMs) and non-dystrophic myotonic syndromes (NDMs). Currently, there is no evidence-based, safe and effective treatment for patients with DMs and NDMs. This year, a multiple N-of-1 trial is scheduled to asses the safety and efficacy of mexiletine in NDM-patients. In the future, a trial with anti-sense oligunucleotide treatment to rescue the mRNA-defect in patients with DMs will be scheduled. In order to being able to measure de efficacy of both treatments

will be the availability of reliable, validated outcome-measures for the quantificatioin of myotonia, besides the already available subjective outcome measures.

Study objective

In this study we want to investigate if the muscle-relaxation time (muscle-RT), calculated from force profiles obtained with two different experimental methods (1) voluntary handgrip- and (2) peripheral nerve stimulation myometry, is a reliable and reproducable outcome measure for the quantification of myotonia in NDMs and DMs. Moreover, muscle-RT data of voluntary handgrip- and peripheral nerve stimulation myometry will be compared to determine which of these two methods can be used best in future medication trials.

Study design

Observational research without invasive measurements.

Study burden and risks

Advantages: Results from this study will be used to develop outcome measures for future treatment studies. Disadvantages: We ask study participants ±2 times 85 minutes of their time for performing the measurements. Furthermore, participants will be asked to eat or drink nothing (except for water) in the two hours prior to study measurements and to keep a low potassium diet in the three days before the study measurements and during study measurements. Risks are minimal and could consist of some extra muscle stiffness or muscle pain. Nerve stimulation could give an unpleasant sensation during measurements.

Contacts

Public
Universitair Medisch Centrum Sint Radboud

Reinier postlaan 1
Postbus 9101, 6500 HB Nijmegen
NL
Scientific
Universitair Medisch Centrum Sint Radboud

Reinier postlaan 1
Postbus 9101, 6500 HB Nijmegen
NL

Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

Patients with a genetically confirmed mutation in the gene encoding for the skeletal muscle sodium channel (SCN4A) or chloride channel (CLCN1) or a genetically determined mutation in the DMPK-gene, with clinical presence of hand (action) myotonia and between 18 and 65 years of age. Groups will be age and sex matched.

Exclusion criteria

Comorbidity in form of a neurological or metabolic disorder that can interfere with normal muscle function. Usage of medication that can influence myotonia or muscle force (such as sodium channel blockers as widespreas used for cardiological and neurological diseases, that might also influence skeletal muscle channels). Cardial or Nephrological comoborbidity that do not allow a potassium low diet. Pregnant women. Unwillingness or unable to sign informed consent forms or to place the hand or lower arm in de myometry test set-up.

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)

Control:	Active
Primary purpose:	Other

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	14-01-2012
Enrollment:	0
Type:	Anticipated

Ethics review

Approved WMO

Date:	09-01-2012
Application type:	First submission
Review commission:	CMO regio Arnhem-Nijmegen (Nijmegen)

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL38883.091.11