Muscle relaxation time as outcome measure for myotonia in patients with non-dystrophic myotonic syndromes (NDMs)

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In this study we want to investigate if relaxation time (RT) measured by handgrip myometry and muscle- elastography is a reliable outcome measurement for the quantification of myotonia in NDM patients. Mean RT values en standard deviations of NDM...

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

Summary

ID

NL-OMON35097

Source
ToetsingOnline

Brief title
Muscle relaxation time measurements in NDMs patients

Condition

- Muscle disorders
- Neuromuscular disorders

Synonym
inherited skeletal muscle stifness, non-dystrophic myotonic syndromes (NDMs)

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: echo muscle elastography, handgrip myometry, myotonia, relaxation time

Outcome measures

Primary outcome

Average muscle relaxation time (RT) of the right underarm flexors measured by

- (1) handgrip myometry

- (2) muscle- elastography (measured with echography)

Secondary outcome

none

Study description

Background summary

Non dystrophic myotonic syndromes (NDMs) form a heterogeneous group of rare diseases caused by either chloride or sodium channelopathies that exclusively affect skeletal muscles. The key symptom of NDMs is myotonia, i.e., a delayed muscle relaxation after voluntary or evoked muscle contraction.

Recently the neurology department of the Radboud University Nijmegen Medical Centre (RUNMC) defined the Dutch genotypes and phenotypical characteristics of NDMs, by conducting a nation wide survey. They concluded that a good quantative outcome measure for myotonia does not exist yet.

There is no evidenced based, effective therapy for NDMs patients. Therefore, the only drug treatment option left for NDMs patients is prescription of off label drugs, for example mexiletine, a sodium channel blocker. There are serious plans for setting-up a mexiletine trial using the N of 1 methodology. For determination of the effectiveness of mexiletine, a reliable and validated outcome measure is needed which can quantify myotonia. In this study we want to investigate if relaxation time (RT) measured by handgrip myometry and muscle-

elastography is a reliable outcome measurement for the quantification of myotonia in NDM patients.

Study objective

In this study we want to investigate if relaxation time (RT) measured by handgrip myometry and muscle- elastography is a reliable outcome measurement for the quantification of myotonia in NDM patients. Mean RT values en standard deviations of NDM patients measured in this study, will be used in sample size calculations for a future double blind, randomized, placebo controlled, cross over intervention study with Mexiletine versus placebo. In this study the therapeutic effect of Mexiletine will be evaluated by measurement of the RT of the fingerflexors, as a new method for quantification of myotonia.

Study design

observational study without invasive interventions.

Study burden and risks

There are no direct advantages for the participating subjects. Disadvantages for participating subjects: the study wil cost the subjects two times 45 minutes, 3 days prior to the measurments they will have to follow a potassium low diet and they have to fast 2 hours prior to the measurement.
Risks are minimal, some extra muscle stiffness or muscle pain of the right hand due to the measurement of muscle strength of the right hand can occur.

Contacts

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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 the skeletal muscle sodium channel (SCNA4) or skeletal muscle chloride channel (CLCN1) and healthy volunteers (18-65 years old).

Exclusion criteria

Neurological or metabolic comorbidity which affect the muscles. The use of medication which affect myotonia or muscle strength (mainly sodium chanel blockers). Presence of renal or cardiac diseases that do not allow the participant to follow a potassium restricted diet. For the same reason, pregnant woman are not allowed to participate in this study.

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:	Recruitment stopped
Start date (anticipated):	06-04-2010
Enrollment:	30
Type:	Actual

Ethics review

Approved WMO

Date:	06-04-2010
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	NL30366.091.09