Reproducibility and correlation with clinical severity of the McManis long-exercise test in patients with hypokalemic periodic paralysis

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- To assess the reproducibility of the LET.- To define the relation between the LET and clinical severity in HypoPP patients.

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

Summary

ID

NL-OMON42874

Source
ToetsingOnline

Brief title
LET in HypoPP study

Condition

- Muscle disorders

Synonym
Hypokalemic periodic paralysis (HypoPP)

Research involving
Human

Sponsors and support

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

Intervention

Keyword: Channelopathy, Long-exercise test, Periodic paralysis

Outcome measures

Primary outcome

- The intraclass correlation coefficient (ICC) of the maximum percentage

decrease of the compound muscle action potential (CMAP) of all ten patients on

all four measurements

- The correlation coefficient between the clinical severity and the mean

maximum decrease of the CMAP measured over the four LET's.

Secondary outcome

Not applicable

Study description

Background summary

Hypokalemic periodic paralysis (HypoPP) is a rare, monogenetic muscle disorder caused by a mutation in the skeletal muscle calcium (CACNA1S) or sodium ionchannel gene (SCN4A). Clinically, these disorders are characterized by attacks of flaccid muscle weakness of the extremities with an accompanying shift in serum potassium from the serum compartment into the muscle. The long-exercise test (LET) is a well validated diagnostic test for HypoPP that is performed in between attacks. During the LET, a partial paralytic attack is provoked in a small muscle (m. abductor digiti minimi) of the little finger by five minutes of voluntary maximal exercise. After this exercise, the compound muscle action potential (CMAP) amplitude is measured every two minutes with evoked potential (EMG). The CMAP amplitude provides information on changes in the number of active fibres and their ability to depolarize and repolarize. Most studies in the field have only focused on the diagnostic quality of the test; no previous study has investigated the reproducibility of this test, nor its correlation with the clinical severity experienced by patients. More knowledge on these aspects of the test will tell us more about the reliability of the test as a diagnostic test as well as an objective assessment of

treatment status.

Study objective

- To assess the reproducibility of the LET.
- To define the relation between the LET and clinical severity in HypoPP patients.

Study design

Explorative, diagnostic study

Study burden and risks

Patients will be subjected to the LET four times, two times on study day 1 and two times on study day 2. A single test takes about one hour to perform. During the test, a transient paralytic attack in the musculus abductor digiti minimi of the little finger of the patient will be provoked by 5 minutes maximal use of this muscle. The paralytic attack only effects this muscle and therefore the patient will not experience much inconvience. During the following 50 minutes, a single electrical pulse to the ulnar nerve will be used to assess the level of paralyses every 1-2 minutes. These pulses are not painful to the patient. Next to this, patients will be asked to keep a diary of the paralytic attacks they experience during one month, noting both length and severity of each attack.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)
Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

1. At least 16 years of age
2. Genetically confirmed diagnosis of HypoPP

Exclusion criteria

1. Inability or unwillingness to provide informed consent
2.Other neurological conditions that might affect the assessment of the study measurements
3. Any reason why the tests cannot be performed on both hands

Study design

Design

Study type: Observational non invasive

Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Diagnostic

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	16-05-2016
Enrollment:	10
Type:	Actual

Ethics review

Approved WMO Date:	29-04-2016
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	NL57236.091.16