The Cognitive Functioning of Patients with Limb-Girdle Muscular Dystrophy Type 2A (LGMD2A): A Pilot Study

Published: 01-06-2006
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This study aims to make a cognitive profile of dutch LGMD2A patients.

Ethical review	Approved WMO
Status	Pending
Health condition type	Congenital and hereditary disorders NEC
Study type	Observational non invasive

Summary

ID

NL-OMON29740

Source
ToetsingOnline

Brief title
Cognitive Functioning of LGMD2A Patients: A Pilot Study

Condition

- Congenital and hereditary disorders NEC
- Muscle disorders
- Neuromuscular disorders

Research involving
Human

Sponsors and support

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

Intervention

Keyword: calpain 3, cognitive functioning, LGMD2A, memory

Outcome measures

Primary outcome

Deviations from the norm for the cognitive functions mentioned in the

background.

Secondary outcome

Not applicable

Study description

Background summary

LGMD2A is a muscle dystrophy caused by changes in the calpain 3 gene. This gene is also found in the hippocampus and the corpus callosum of rats' brains. In humans, the hippocampus and the corpus callosum play an important role in such cognitive functions as arousal, attention and concentration, declarative memory, and visuospatial and visuoperceptual abilities.

Research findings suggest that changes in calpain 3 can lead to changes in the functions of the concerned tissues and systems. Based on this, this study will test the hypothesis that lack of or changes in calpain 3 in the hippocampus and the corpus callosum lead to deviations in cognitive functions associated with these areas.

Study objective

This study aims to make a cognitive profile of dutch LGMD2A patients.

Study design

Neuropsychological research:
The research consists of a series of neuropsychological tests. The selected tests are aimed to test thec ognitive functions mentioned in the background. The following tests will be conducted: Verbal Learning and Memory Test (VLGT); key search and shift-rule test of the Behavioural Assessment of Dysexecutive Syndrome (BADS); picture arrangement, digit span, and picture completion of the

Weschler Adult Intelligence Scale (WAIS); visual memory span of the Weschler Memory Scale (WMS); face recognition and picture recognition of the Rivermead Behavioural Memory Test (RBMT); word fluency of the Groninger Intelligence Test (GIT); and the Stroop Colour-Word test.

Data analys: The data will be analysed quantitatively and qualitatively.

Research duration: 5 months.

Feedback: Participants will receive an abstract of the final report.

Protocol in case of patient resistance: A patient can at any moment decide to stop with the research.

Privacy: Results will be coded in a way that they would not directly lead to the identity of the individual patient.

Research organisation:
The research team consists of the following:
Principal researcher:
Dr. Anneke J. van der Kooi, M.D., Ph.D., Academic Medical Centre, University of
Amsterdam
Executing researchers:
Prof. Dr. Erik J.A. Scherder, Ph.D., Professor Clinical Neuropsychology,
Vrije Universiteit Amsterdam
Lyzel S. Elias-Sonnenschein, M.A., Department of Clinical Neuropsychology,
Vrije Universiteit Amsterdam
Vladimir Vladimirov, M.A., Department of Neuroscience, Vrije Universiteit Amsterdam

Study burden and risks

Testing takes about 2,5 hours and will be done at the patients' home.
There are no risks involved in this research.

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

LGMD2A is genetically proven
Dutch citizen
Dutch speaking
Physically able to participate in the reseach

Exclusion criteria

LGMD2A not genetically proven
Not able to understand and express in dutch sufficiently
Physically unable to participate in the research

Study design

Design

Study type: Observational non invasive

Masking:	Open (masking not used)
Control:	Uncontrolled

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	30-04-2006
Enrollment:	30
Type:	Anticipated

Medical products/devices used

Registration:	No

Ethics review

Approved WMO

Application type:	First submission
Review commission:	METC Amsterdam UMC

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	NL12097.018.06