Myotonic Dystrophy, PrOteiN and Diet study

No registrations found.

Ethical review	Positive opinion
Status	Other
Health condition type	-
Study type	Observational non invasive

Summary

ID

NL-OMON25314

Source
Nationaal Trial Register

Brief title
MD-POuND

Health condition

myotonic dystrophy type 1, neuromuscular disease, metabolism, body composition

Sponsors and support

Primary sponsor: Academisch Ziekenhuis Maastricht
Source(s) of monetary or material Support: Prinses Beatrix Spierfonds
Postbus 85810
2508 CM Den Haag

Intervention

Outcome measures

Primary outcome

The main study endpoints are:

- total energy expenditure measured by DLW and respiration chamber;

- resting metabolic rate (RMR) measured by respiration chamber;

- substrate selection at rest measured by respiration chamber.

Secondary outcome

Secondary endpoints include:

- Physical activity level (PAL) measured by accelerometer;

- Whole-body skeletal muscle mass measured by MRI;

- Quadriceps muscle cross-sectional area measured by CT;

- Maximal grip strength measured by dynamometer;

- Body composition expressed as leg and whole-body lean tissue mass measured by DEXA;

- Muscle tissue morphology (including fibre size, split fibres, location of nucleii, regenerating and degenerating fibres, connective tissue, inflammatory cells, inclusions and storage material), muscle fibre differentiation, muscle fibre type specific cross-sectional area and vascularization, all determined by microscopically performed histological muscle biopsy assessment;

- Muscle fiber biochemical analysis: cytochrome C oxidase enzym activity;

- Maximal oxygen uptake capacity (VO2 max) and aerobic and anaerobic ventilatory thresholds determined by cycling exercise test;

Study description

Background summary

Myotonic dystrophy type 1 (DM1) is an autosomal dominant disorder that affects the skeletal, cardiac, and smooth musculature and many other tissues. While muscle weakness and myotonia (inability to relax muscles) are the main characteristics of disease, patients with DM1 frequently experience marked loss of muscle, as well as issues regarding nutrition and weight. This prospective study aims to assess metabolism in 15 patients with DM1, taking into account muscle mass, and compared to 15 healthy age-, BMI- and gender-matched subjects. We hypothesize that DM1 is accompanied by impairments in whole-body and/or muscle metabolism.

Study objective

We hypothesize that DM1 is accompanied by impairments in whole-body and/or muscle metabolism.

Study design

total duration of 2 years

Intervention

none

Contacts

Public

Scientific

Eligibility criteria

Inclusion criteria

- Age 18 years or older.

- Legally competent adult.

- Defined DM1 of the adult subtype.

- Participants must be able to walk and to cycle (in order to perform exercise tests).

- Participants must give informed consent by signing and dating an informed consent form.

Exclusion criteria

- Implantation of pacemaker or ICD device, a implantable insulin device, a neurostimulator, internal hearing aid or artificial heart valve.

- Implantation of orthopaedic prostheses, screws or plates.

- Metal shreds or splinters inside the body.

- Vascular clips in the body.

- Big tattoo’s and/or permanent make-up.

- Claustrophobia.

- Use of medication interacting with muscle metabolism (such as steroids and statins).

- Diabetes mellitus.

- Weight loss of more than 3 kg in the last three months.

- Pregnant or lactating women.

- Use of protein supplements.

- Participation in an exercise program (for a study).

- Patients who are not able to perform basic activities of daily living such as walking, or patients who are suffering from other disabling comorbidity that seriously hamper physical exercise (e.g. heart failure, coronary artery disease, chronic obstructive pulmonary disease (COPD), orthopedic conditions).

- Body mass index (BMI) <18,5 or >35 kg/m2.

- Use of oral anticoagulants.

- In case of DM1 affected subjects: muscular impairment rating scale (MIRS) score of 5 (which represents severe proximal muscle weakness).

- In case of the healthy control group:
presence of a neuromuscular disorder or an abnormal neurological examination (specifically if muscle weakness is present), or other condition possibly interfering with muscle strength or muscle mass

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Masking:	Open (masking not used)

Control:	N/A , unknown

Recruitment

NL

Recruitment status:	Other
Start date (anticipated):	01-06-2018
Enrollment:	0
Type:	Unknown

Ethics review

Positive opinion

Date:	23-10-2018
Application type:	First submission

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
NTR-new	NL7374
NTR-old	NTR7582
Other	: METC182009

Study results