Inspiratory muscle training in patients with nemaline myopathy

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The primary objective is to determine the effect of a 8-week inspiratory muscle training program on respiratory muscle function in nemaline myopathy patients. The secondary objective is to determine respiratory muscle function in nemaline myopathy...

Ethical review	Approved WMO
Status	Recruitment stopped
Health condition type	Musculoskeletal and connective tissue disorders congenital
Study type	Interventional

Summary

ID

NL-OMON46630

Source
ToetsingOnline

Brief title
NEMTRAIN

Condition

- Musculoskeletal and connective tissue disorders congenital

Synonym
Nemaline myopathy, rod myopathy

Research involving
Human

Sponsors and support

Primary sponsor: Radboud Universitair Medisch Centrum
Source(s) of monetary or material Support: A Foundation Building Strength en Prinses Beatrix Spierfonds

Intervention

Keyword: diaphragm, inspiratory muscle training, nemaline myopathy, respiratory muscle function

Outcome measures

Primary outcome

The primary outcome parameter is the change in maximal inspiratory pressure

(MIP) after active inspiratory muscle training.

Secondary outcome

Several secondary outcome parameters:

* Diaphragm ultrasound (thickness, thickening, excursion).

* Pulmonary function tests (peak cough flow, forced vital capacity, forced

expiratory volume the first second, peak expiratory flow, vital capacity)

* Respiratory muscle function tests (sniff nasal inspiratory pressure, maximal

expiratory pressure, twitch mouth pressure)

* Neurological examination

* Maximal voluntary contraction

* Rate of muscle relaxation

* Mobility: Rivermead mobility index, and Community Balance and Mobility Scale

* Falls: Falls efficacy scale and Falls History Questionnaire

Study description

Background summary

Nemaline myopathy is a group of congenital, hereditary neuromuscular disorders with variable symptoms such as muscle weakness, swallowing dysfunction, and dysarthria. Respiratory failure is the main cause of death in nemaline myopathy

and occurs even in ambulant patients who otherwise appear to be only mildly affected; respiratory muscle weakness may even be the presenting feature. Inspiratory muscle training has shown to increase inspiratory muscle strength in patients with other neuromuscular disorders. We hypothesize that inspiratory muscle training improves inspiratory muscle function in nemaline myopathy patients with respiratory muscle weakness.

Study objective

The primary objective is to determine the effect of a 8-week inspiratory muscle training program on respiratory muscle function in nemaline myopathy patients. The secondary objective is to determine respiratory muscle function in nemaline myopathy patients and its correlation with clinical severity and general neuromuscular function.

Study design

The study consist of two phases. Phase 1: A screening phase with an open design from which patients will be selected for the second phase. Phase 2: A controlled before-after trial of inspiratory muscle training. The 2 conditions tested are active IMT and sham IMT.

Intervention

Active IMT consists of 15 minutes of IMT, twice a day, 5 days per week for 8 weeks, at a training workload of 30% of MIP using a resistive inspiratory muscle training device. Sham IMT consists of a similar training regime using a resistive inspiratory muscle training device where the resistance has been removed.

Study burden and risks

The intervention of inspiratory muscle training is not associated with any risks, but can be challenging in patients with respiratory muscle weakness to perform. There will be four visits to the hospital in 16 weeks. The first visit will be approximately 3.5 hours (including breaks) and the other three visits 1.5 hour. During these visits several tests and physical examinations will be performed. Some of the tests may cause some physical discomfort, but none of them carry any risk. Patients may benefit from participating in this study by developing improved respiratory muscle function as a result of the inspiratory muscle training.

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)
Children (2-11 years)
Elderly (65 years and older)

Inclusion criteria

* genetically-confirmed nemaline myopathy
* informed consent from participant or legal representative

Exclusion criteria

* history of another condition that affects respiratory muscle strength or function (e.g. COPD)*

Study design

Design

Study type:	Interventional
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	Placebo
Primary purpose:	Treatment

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	10-10-2018
Enrollment:	23
Type:	Actual

Medical products/devices used

Generic name:	Inspiratory muscle training
Registration:	Yes - CE intended use

Ethics review

Approved WMO Date:	13-06-2018
Application type:	First submission
Review commission:	CMO regio Arnhem-Nijmegen (Nijmegen)
Approved WMO Date:	27-08-2018
Application type:	Amendment
Review commission:	CMO regio Arnhem-Nijmegen (Nijmegen)
Approved WMO Date:	02-01-2019
Application type:	Amendment

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	NL65214.091.18