Which muscles are critical for abnormal posture, balance and gait in patients with FSHD?

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

Summary

ID

NL-OMON34404

Source
ToetsingOnline

Brief title
Posture, balance and gait in FSHD patients.

Condition

- Muscle disorders

Synonym
facioscapulohumeral dystrophy / Landouzy-Dejerine syndrome

Research involving
Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Sint Radboud
Source(s) of monetary or material Support: FSHD stichting

Intervention

Keyword: Balance, FSHD, Gait, posture

Outcome measures

Primary outcome

Maximum EMG amplitude, expressed as a percentage of the MVC during isolated contraction, is the primary outcome measure of the posture, balance and gait tasks.

For the MRIS measurements, the primary outcome will be the cross-sectional area of the various leg and trunk muscles.

Secondary outcome

Posture: the joint angles of the lower back, hips, knees, and ankles in two positions.

Gait: gait velocity; spatiotemporal variables (e.g. step lengths and cadence); joint angles and joint torques of the lower back, hips, knees and ankles during the gait cycle.

Balance: spatiotemporal variables (e.g. displacement of the centre of mass) joint angles and joint torques.

MRI: fatty degeneration on a four point scale proposed by Lamminen (1990).

Study description

Background summary

Facioscapulohumeral dystrophy (FSHD) is the third most common inherited neuromuscular disorder. It is an autosomal dominant slowly progressive myopathy with a variable age of onset, mostly in the second or third decade of life (Kissel, 1999). The pattern of muscle weakness is often asymmetrical, and the

rate and extent of progression may vary considerably with sudden periods of unexplained rapid disease progression. FSHD may eventually lead to serious disabilities of speech, swallowing, reaching, standing and walking, even in early adulthood. Twenty percent of the patients become wheelchair bound (Padberg 1982; Lunt & Harper, 1991; Tawil & van der Maarel, 2006).
The semimembraneous, tibialis anterior, biceps femoris, medial gastrocnemius muscles and adductor group appeared to be most affected, whereas the psoas, vasti, gluteal and peroneal muscles were relatively unaffected (Olsen et al. 2006). As a result, patients develop specific problems with posture, balance and gait.
Lastly, several case reports of infantile FSHD indicate the importance of excessive lumbar lordosis for losing the capacity to ambulate. Excessive lordosis would be related to weakness of the pelvic extensors, in particular gluteus maximus (Shapiro et al., 1991; Dorobek & Kabzinska, 2004; Lee et al., 2009). Hyperlordosis is often attributed to weak abdominal muscles and relatively strong back extensor muscles (Padberg 1982; Shapiro at al., 1991), but this notion has not yet been substantiated with quantitative data.
In general, one can conclude that the typical hyperlordotic posture and the marked balance and gait abnormalities in FSHD are poorly understood, which makes it difficult to develop effective intervention strategies such as bracing (Shapiro et al., 1991) or fall prevention programmes (Horlings et al., 2009).
It is expected that with a better understanding of the mechanisms that are responsible for the balance and gait disabilities, it should be possible to improve rehabilitation strategies.

Study objective

The primary aim of this study is to identify the muscles that are most critical to the abnormal standing posture, standing balance and gait in FSHD patients and to elucidate the most important mechanisms of falls. The ultimate goals are (1) to establish novel hypotheses that can be tested in larger groups of patients and (2) to find clues to improve bracing and training strategies in order to optimize individual functioning and prevent falls in FSHD patients.

Study design

Cross-sectional, explorative patient-control study with laboratory assessments of posture, balance and gait in combination with structural magnetic resonance imaging (MRI).

Study burden and risks

The extent of the burden for a person in time is about 6 hours, distributed over 3 sessions. After the measurements there might be a risk of fatique, which depends on the physical limitations of the person.

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

- Diagnosis FSHD based on a positive (family) history, examination, genetically confirmed.
- Age between 18-80 years.
- Being able to stand independently and walk for ten meters without aids.

Exclusion criteria

- Other neurological or musculoskeletal disorders.
- Use of medication affecting balance (e.g. neuroleptics, antidepressants, anticonvulsants).

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:	Basic science

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	10-03-2011
Enrollment:	20
Type:	Actual

Ethics review

Approved WMO

Date:	10-11-2010
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	NL32158.091.10