A study of the effects of balanced shoulder therapy on shoulder function, local fatigue and pain symptoms in children with FSHD type 1

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Primary: To describe shoulder function, local fatigue and pain symptoms around the shoulders in children with FSHD type 1, before and after balanced shoulder therapy.
Secondary: To gain insight into the experience of children and parents and the...

Ethical review	Approved WMO
Status	Pending
Health condition type	Muscle disorders
Study type	Interventional

Summary

ID

NL-OMON57269

Source
ToetsingOnline

Brief title
Research on Better Shoulder Movement in Children with FSHD

Condition

- Muscle disorders

Synonym
Facioscapulohumeral muscular dystrophy type 1; FSHD

Research involving
Human

Sponsors and support

Primary sponsor: Radboud Universitair Medisch Centrum

Source(s) of monetary or material Support: FSHD Stichting

Intervention

Keyword: FSHD Type 1, Occupational Therapy, Physical therapy, Shoulder Function, Shoulder Therapy

Outcome measures

Primary outcome

The Pediatric/Adolescent Shoulder Survey (PASS).

A patient reported outcome measure (PROM) with 13 questions on symptoms, impairments, compensatory strategies and emotional stress related to shoulder dysfunction (10 minutes).

- Canadian Occupational Performance Measure (COPM).

The COPM is administered by the occupational therapist and identifies the main problems experienced by the child and measures changes in the child's perception of his actions and his satisfaction with the change during the treatment process (30 minutes).

Secondary outcome

Qualitative study:

- Experiences of children, parents and therapists, from which themes are determined. The experience concerns the content of therapy, the individual elements of therapy, the perceived effect, the balance between putting energy into it and what it yields. There is also room to share unexpected experiences.

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Functional level:

Muscle strength of upper extremity:

- Hand Held Dyanometry (HHD (Microfet))

Duration: 10 minutes.

Scapular kinesia:

- Visual analysis of scapular kinesia (VASK) by means of a film recording on a normal camera of a cell phone in which there is analysis of the scapula in static and dynamic position

Duration: 1 minute.

Activity Level

Active Range of Motion within activities:

Reachable Work Space (RWS).

Duration: 10 minutes.

Degree of upper extremity complaints and limitations during activities:

- Quick Disabilities of the Arm, Shoulder and Hand (Quick DASH), including module on sports and playing musical instruments.

Duration: 10 minutes.

- 4 Items of upper extremity (Overarm throwing, Underarm throwing, Chest pass, Lifting a box) of the Functional Strength Measurement (FSM) with Numeric Rating Scale (NRS) of pain and fatigue of upper extremity

Duration: 10 minutes.

Quality-of-Life:

- Neurology Quality-of-Life: Pediatric Version. Upper Extremity- Fine Motor,

ADL. [Hatch et al. 2020] (validated for children [Lai et al. 2012]).

Duration: 10 minutes.

Study description

Background summary

The shoulder (scapulo) and arm (humerus) function, in addition to the face (facies), is affected in the hereditary muscle disease fascioscapulohumeral muscular dystrophy (FSHD) type 1; movements of shoulders and arms become more difficult and require more energy. In a natural course study of children with FSHD type 1, pain, fatigue, problems in endurance, and decreased quality of life are reported, threatening overuse. In addition, 96% of adults with type 1 FSHD report shoulder pain.

In adults and children with type 1 FSHD, we often see compensatory strategies, with abnormal scapula (scapula) position at rest and during movement. Any clinically observable change from normal scapular position and movement is defined as scapular dyskinesia. The etiology is multifactorial, most often scapular dyskinesia occurs as a result of changes in the activation or coordination of the muscles that control movement. Scapular dyskinesia indicates an altered motor profile and is frequently seen in the clinical setting in FSHD type 1, which can lead to pain, fatigue, and increasingly difficult movement of the arm, suggesting more rapid loss of function. There is scientific evidence that targeted shoulder therapy in adults with other muscle disease leads to a reduction in symptoms, but so far there is no evidence in children with FSHD type 1.

We have, based on the above scientific evidence in adults and our clinical expertise built in the Children's Muscle Center developed a therapy tailored to children. Movements to regain motor control of scapular movements are elicited in ball games and other exercise activities along with pediatric physical

therapy. The therapy also focuses on teaching energy-saving strategies during daily activities, along with pediatric occupational therapy.

This ''balanced shoulder therapy'' is already being used in the children's muscle center at Amalia Children's Hospital in children with various neuromuscular disorders including those with FSHD type 1. Subjectively, they experienced improvement in various domains (pain, fatigue, shoulder function) and it proved easy to incorporate this therapy into daily life. A scientific evaluation for this balanced shoulder therapy is so far lacking to identify the effects and experiences with this therapy

Study objective

Primary: To describe shoulder function, local fatigue and pain symptoms around the shoulders in children with FSHD type 1, before and after balanced shoulder therapy.

Secondary: To gain insight into the experience of children and parents and the therapists,with balanced shoulder therapy.

Study design

Design: Mixed Method- Embedded design: multi case study limited efficacy testing, gecombineerd met kwalitatieve studie - beschrijvende fenomenologische studie.

Intervention

Balanced shoulder therapy consists of 6 sessions of pediatric physiotherapy for training scapula-coordinating function and 4 sessions of pediatric occupational therapy for education of child and parents on shoulder load, optimizing sitting-sleeping posture and teaching energy-saving strategies. This takes place over a period of 4.5 months in the Radboudumc.
After completion of the therapy semi-structured interviews will take place, with child, parents and therapists, on the experience of this therapy.

Study burden and risks

See the entire study protocol for full details and comprehensive information.
Specifics on time load: table 2 of study protocol page 21.

Measurement instruments:
Three months before the start of training, 3 questionnaires (PASS, QuickDASH, NeuroQoL, total 30 minutes) will be administered by telephone. These questionnaires are relatively short (10 minutes each) the first two lists are patient reported outcome measure (PROM) with questions about symptoms,

limitations, compensation strategies and emotional stress related to shoulder dysfunction. They are also asked to take a 1minute film to map scapular kinesia (VASK).

Intervention takes place within 4.5 months, 6 sessions of pediatric physical therapy and 4 sessions of pediatric ergotherapy.
Prior to session 1 of the intervention, the measurement instruments are administered on the same day (3 questionnaires: PASS 10 minutes, QuickDASH 10 minutes, NeuroQoL upper extremity 10 minutes; severity of FSHD type 1: FSHD Clinical Score 5 minutes; video observation of shoulder movements (VASK) 1 minute; muscle strength measurements (HHD and FSM) 20 minutes; functional shoulder movements with Kinect (RWS)10 minutes.

mmediately after the last training session the same measuring instruments will be used and the semi-structured interview (<12 years maximum 30 minutes, 12 years or older: maximum 60 minutes) with the child and the parent(s) will take place.
Three months after the end of the intervention the child comes back once more for evaluation, during which the above measuring instruments are taken again (total duration: 1.5 hours).

Intervention
Pediatric Physical Therapy
6 sessions of 1 hour by the pediatric physiotherapist of the Radboudumc with an individual training program.

The children are given exercises more for at home, supported by films. 4 x per week, 20 minutes each time.
The children are given a short list that is easy to fill in. On this they keep track of whether and how long they have practiced and if there are any peculiarities. They take this back during the therapy session and then they get a new one.

Pediatric Occupational Therapy
4 individual sessions by the Radboudumc pediatric occupational therapist.

Interview
On last day of the intervention, interviews will be held with the parents and children and involved pediatricergo- and pediatric physiotherapist. The interview lasts up to 30 minutes for children <12 years and up to 1 hour for children 12 years and older and their parents/caregivers.

Risks:
There is no major risk associated with participating, as the children are instructed to perform the exercises in a comfortable and controlled manner.

The tests that are done are also not risky, these are tests that are also frequently performed within current regular care in children with FSHD.

We expect that the children themselves will benefit from this form of therapy, both now and in the future. In addition, we expect them to have fun during the therapy (we received this as feedback from the children who have already done this form of therapy). There is a burden of coming to the Radboudumc and doing exercises at home, but experience shows that children and their parents are happy to do a lot to improve the function of their shoulders. They also now have the added value of being able to use the expertise of experienced therapists within this target group (which is not available in their neighborhood).

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)
Children (2-11 years)

Inclusion criteria

- Children with clinically or genetically confirmed FSHD type 1 aged 8-18 years.
- Children attending normal regular education.
- Children with scapular dyskinesia (VASK score >=20).

Exclusion criteria

- Other neurological disorder or neuromuscular disorder affecting shoulder function.
- Orthopedic condition that affects shoulder function.
- Children who have had this type of therapy before.
- Children who are not motivated or able to perform this therapy.
- Children who are non-ambulatory.

Study design

Design

Study type: Interventional

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

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	02-09-2024
Enrollment:	6
Type:	Anticipated

Ethics review

Approved WMO

Date:	20-01-2025

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	NL87566.091.24