Somatosensory profile of pain in myotonic dystrophy type 2 and fibromyalgia

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1. What is the somatosensory profile of DM2 patients?2. What are the similarities and differences between the somatosensory profile of pain in myotonic dystrophy type 2 and fibromyalgia patients?

Ethical review	Approved WMO
Status	Pending
Health condition type	Chromosomal abnormalities, gene alterations and gene variants
Study type	Observational non invasive

Summary

ID

NL-OMON38498

Source ToetsingOnline

Brief title

Characterising pain in myotonic dystrophy type 2 and fibromyalgia

Condition

- Chromosomal abnormalities, gene alterations and gene variants
- Muscle disorders
- Neuromuscular disorders

Synonym Myotonic dystrofie type 2; rare hereditair muscle disease PROMM

Research involving

Human

Sponsors and support

Primary sponsor: Canisius Wilhelmina Ziekenhuis

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Source(s) of monetary or material Support: Ministerie van OC&W

Intervention

Keyword: Dystrophia myotonica 2, Fibromyalgia, Pain, Quantitative Sensory Testing

Outcome measures

Primary outcome

The main study parameter of this study is the description and profile of pain

in DM2, compared to patients with fibromyalgia and healthy controls.

Secondary outcome

nvt

Study description

Background summary

DM2 is a rare dominantly inherited multisystemic disorder with a heterogeneous phenotype. In the Netherlands there are 54 known patients with DM2. The clinical features of DM2 were first described in 1994 [Ricker et al 1994]. In 2001, the genetic defect of DM2 was identified as an unstable CCTG tetranucleotide repeat expansion in the ZNF9 gene on chromosome 3q21.3 [Liquori et al 2001]. The core symptoms of DM2 are proximal muscle weakness, muscle pain, myotonia and cataracts [Day et al 2003]. Other characteristics are sleep disturbances, fatigue, gastrointestinal symptoms and involvement of the heart, endocrine and autoimmune system [Tieleman et al 2008, 2010; Wahbi et al 2009].

Pain is an important and early feature of DM2. According to Suokas et al, the prevalence of pain in DM2 is 76% [Suokas et al 2012]. In up to a third of DM2 patients, pain is the most disabling feature [George 2004], and it tends to increase with disease duration. 69% of DM2 patients with nocturnal sleep disturbances report pain as the cause [Tieleman 2010]. DM2 patients with pain show a lower quality of life compared to those without pain [Suokas et al 2012]. Moreover, according to Suokas et al both depressive symptoms and insomnia occur significantly more frequently among DM2 patients with pain compared to those without pain.

Several studies have tried to characterize the pain by means of questionnaires. George et al reported that the spectrum of coexisting types of musculoskeletal pain in DM2 is wide [George et al 2004]. They found that the pain is widespread, most pronounced localised in thighs, back and proximal upper limbs. Ricker et al reported the pain to be fluctuating, sometimes lasting for hours, days or even weeks and then disappearing for days or weeks [Ricker et al 1995, Ricker et al 1999]. Several studies reported that the pain is induced by cold and reduced by warmth [Day 1999, George 2004, Suokas et al 2012]. Furthermore, it seems to increase during and after exercise, and is relieved by rest [George et al 2004; Suokas et al 2012]. A lot of patients report tenderness in the muscles, aggravated by palpation [George et al 2004, Ricker et al 1995]. George et al and Ricker et al did not find a relation between the severity of pain and myotonia [George et al 2004, Ricker et al 1995]

There is no curative treatment for DM2, and the symptomatic effect of pain medication has not widely been studied, only effects in a few cases have been described. In single cases, a positive effect on pain is described in treatment with NSAIDs, carbamazepine, phenytoin, a short-term course of corticosteroid therapy, baclofen, tizanide and gabapentin [Ricker et al 1999, Meola et al 2000, Sander et al 1996, Udd et al 2006]. The pain and stiffness have been improved in some patients after mexiletine [Day et al 1999; Moxley et al 2002]. However, mexiletine is not used extensively, because of concern about cardiac arrhythmias. In conclusion, no consistently effective treatment is known for pain in DM2.

A modern concept in treatment of pain is *mechanism based treatment* [Jensen et al 2003]. In this concept not the underlying disease is target of treatment (as many underlying diseases are unknown or not curable), but the mechanism that causes the pain. Quantitative sensory testing

(QST) is a useful tool to quantify pain processing. It assesses different aspects of pain. It examines the sensory pathway, from the function of peripheral nerve fibers (large Ab fibers and small C fibers), the spinothalamic pathways to the cortical regions in the brain [Rolke et al 2006]. Nociceptive pain is caused by stimulation of peripheral nerve fibers that respond to stimuli approaching or exceeding harmful intensity. It results in altered sensory processing by the central nervous system. Ongoing nociceptive input might lead to central sensitisation. However, in comes cases changes in central pain processing are so pathological that central pain perception becomes autonomous and thus independent of peripheral pain inputs [Buscher et al]. With QST several pain thresholds are determined and a somatosensory profile is made. Approximately, there are two main profiles: patients with normal central pain processing and with disturbed central pain processing.

The Pain Treatment Center of UMC St. Radboud has a regional function for regular treatment of pain and a supra-regional function for advice of complex pain syndromes. Quantitative Sensory Testing (QST) is frequently performed, both in daily practice as well as in scientific research, according to a standardized protocol [Buscher et al 2005].

In conclusion, pain is common in DM2 and for up to a third of patients the most disabling feature. The mechanism that causes pain in DM2 is unknown. So far, no trials have been performed to investigate the effect of treatment in DM2. The goal of this study is to characterize pain in DM2 more extensively, by accomplishing quantitative sensory testing as well as questionnaires. We expect this will lead to a better understanding of the pathophysiological nature of pain in DM2, and eventually to a mechanism based treatment. Moreover, we will also perform the same tests in fibromyalgia patients, to compare the results. If similarities in the nature of pain in both patient groups are found, treatment strategies of fibromyalgia might be used in DM2 as well.

Study objective

1. What is the somatosensory profile of DM2 patients?

2. What are the similarities and differences between the somatosensory profile of pain in myotonic dystrophy type 2 and fibromyalgia patients?

Study design

Observational study

Study burden and risks

The burden for the patient is fulfilling five questionnaires and one visit to our outpatient department for QST testing. QST testing is harmless, although some patients will find this test unpleasant. There is no immediate advantage for the patient. However, if this study leads to a better understanding of the pathophysiology of pain in DM2, we are closer to an adequate treatment of this disabling feature of DM2.

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

Three different groups with subjects:

1. Adult patients with genetically proven myotonic dystrophy type 2

2. Adult patients with fibromyalgia, who fulfill the criteria by Wolfe et al 2010 for

fibromyalgia, in who myotonic dystrophy type 2 is excluded

3. Adult healthy controls, matched for age and sex

Exclusion criteria

- Age younger than 18 years
- Severe illness (apart from DM2 or fibromyalgia)
- Major depression
- Moderate to severe neuropathy
- Recent (< 6 weeks) major surgery

Study design

Design

Study type: Intervention model: Observational non invasive Other

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Allocation:Non-randomized controlled trialMasking:Open (masking not used)Primary purpose:Basic science

Recruitment

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NL	
Recruitment status:	Pending
Start date (anticipated):	01-03-2013
Enrollment:	120
Туре:	Anticipated

Ethics review

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Approved WMO	
Date:	02-05-2013
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

ID: 22162 Source: Nationaal Trial Register Title:

In other registers

Register CCMO OMON ID NL43167.091.13 NL-OMON22162