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**Fatigue in facioscapulothoracic muscular
dystrophy: a qualitative study of people's
experiences**

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IN PRESS AT DISABILITY AND REHABILITATION

ABSTRACT

Purpose: The aim of this article is to describe how fatigue affects the lives of people with facioscapulohumeral dystrophy (FSHD), how they experience fatigue and how they deal with it in order to attune rehabilitation care to patients' needs.

Method: A qualitative study, consisting of 25 semi-structured interviews with patients with FSHD and severe fatigue (as measured with the CIS fatigue questionnaire), was conducted to gain insight into the experiences of patients with fatigue. Data were inductively analysed.

Results: Patients describe fatigue as an overwhelming and unpredictable experience and they make a distinction between actual experienced fatigue and fear of becoming tired. Fatigue can, according to patients, be the result of weak muscles, physical overachieving or underachieving and stress. But most of the time patients do not know the actual causes of the fatigue, which makes it hard to deal with. The experienced fatigue has a large impact on participation, social contacts and the quality of life of patients, and patients try to use many strategies to adapt themselves to the constantly changing situations.

Conclusions: Fatigue is a severe problem in FSHD and has a huge impact on patients' lives. Patients should be helped to reduce fatigue, for instance by offering evidence-based therapies such as aerobic exercise training or cognitive behavioural therapy.

Keywords: facioscapulohumeral muscular dystrophy (FSHD), fatigue, patients' experience

BACKGROUND

Facioscapulohumeral dystrophy (FSHD) is a slowly progressive inherited neuromuscular disorder. The yearly incidence is 1:20,000 (1). In FSHD, muscle function declines over time. Currently, there is no treatment available to slow down this decline (2-5).

Besides muscle problems (1), severe fatigue is reported by a majority of patients (2). A recent study shows that 61% of patients with FSHD reported severe fatigue (defined as a score ≥ 35 on the subscale fatigue of the Checklist Individual Strength). Those patients had more problems with physical and social functioning, mental and general health, and planning and concentration than did patients without severe fatigue (2,6).

Patients living with FSHD thus often have to deal with fatigue. In the literature, many definitions of fatigue are given (7). For the purpose of this study, fatigue was defined according to the definition of Ream and Richardson: *'Fatigue is a subjective, unpleasant symptom which incorporates total body feelings ranging from tiredness to exhaustion, creating an unrelenting overall condition which interferes with individuals' ability to function to their normal capacity'* (8).

Many chronic illnesses are associated with fatigue, including, for instance, cancer (9), rheumatic disorders (10), post-polio syndrome (11) and COPD (7). Fatigue is often identified as one of the most challenging aspects of chronic diseases (12). It is a disabling symptom that has a substantial impact on patients' self-care activities (13) and overall quality of life (14).

The literature on chronic illnesses contains qualitative studies on the personal experiences of those living with fatigue related to a certain disorder (15-17). Insight into the experiences of people with FSHD considering fatigue is lacking. A better understanding of fatigue may lead to better management strategies and improved quality of life for people living with FSHD. It may also lead to care that is better attuned to patients' needs. The aim of this article is, therefore, to describe how fatigue affects the lives of people with FSHD and how they experience fatigue and how they deal with it.

METHODS

Design

A qualitative research design, using a content analysis approach, was used to gain insight into experiences of fatigue. This design was chosen because it allows the researcher to gain knowledge about various phenomena in real-life events (18) and is preferred for gaining an understanding of the experiences and lifeworld content of

people (19). Data were collected through semi-structured interviews (20) and inductively analysed by using a content analysis (21).

Respondents

All respondents were purposively selected based on relevant characteristics to gather relevant information (22). Patients had to be diagnosed with FSHD and at least 18 years old. All respondents reported severe fatigue (score ≥ 35 on the subscale severity of fatigue) as measured using the Checklist Individual Strength (CIS) (23,24). Scores up to 35 on the subscale severity of fatigue indicate high levels of fatigue (23). The CIS was chosen because of its good internal consistency and reliability (25). Patients with clinical depression as diagnosed using the reliable and valid Beck Depression Inventory for Primary Care (BDI-PC) were excluded (26,27).

The respondents were recruited via different medical centres and rehabilitation centres in the Netherlands. In total, 25 respondents were recruited for an interview. These respondents were first approached by their rehabilitation specialist, and, after giving consent, they were called by one of the researchers. During this phone call, the respondents received further information about the study and were asked to participate in the interview. If they were willing to participate, an appointment was made for the interview.

Data collection

For this study, semi-structured interviews were held at people's homes for comfort reasons and to create an open atmosphere. A topic guide was used to structure the interviews. Themes that were formulated in a topic guide (based on literature) and addressed in the interviews included personal experiences with illness and fatigue, causes of fatigue, strategies to deal with fatigue and restrictions they may be facing. Nevertheless, the interviews had an open nature and aimed to cover the various aspects of the respondents' experiences of fatigue. This open nature was reached by formulating broad topics (instead of specific questions) so as not to restrict respondents to talking about specific themes and to gather information about themes that the respondents addressed.

The process of data collection and analysis was iterative so that emerging themes could be further explored and validated over the course of the research (28). The first analysis showed that no new themes emerged after 21 interviews. This suggested data saturation (29). To confirm this saturation, four additional interviews were held. Since the additional interviews did not give new insights compared to the other interviews, saturation was met. The interviews were conducted by the first (psychologist) and second (sociologist) author. Both have followed courses on how to conduct qualitative studies and both

have carried out several qualitative studies before. They took general quality criteria for the semi-structured interviews into account, such as asking open-ended questions, using different probes and avoiding jargon (30). The interviews lasted between 60 and 90 minutes and were, with the respondents' consent, audio-recorded and transcribed.

Data analysis

The interviews were analysed using inductive content analysis for the purpose of providing knowledge and understanding of the phenomenon of fatigue. The transcripts were read and reread and coded by the first two authors both during and after the data collection. While reading the transcripts, phrases regarding fatigue were coded. New codes in the transcripts were added to the list of codes. All transcripts, including those that were already coded, were read and coded with the use of the new code list. Codes and coded segments were compared and grouped as main and subcategories. The different categories of both researchers were compared and discussed by the research team until consensus was reached. This was done to increase dependability by preventing distortions caused by the personal and professional background of the individual researcher. Relevant themes were agreed upon, and, for each theme, the most suitable quotes were selected for the final report. A mix of computerized ('MAXQDA') and manual techniques was used to facilitate data analysis.

Rigour

To enhance credibility, all respondents received an interpretation of their interview to check the accuracy of the interviewer's interpretation (member check) (28). Furthermore, different investigators were involved in the analysis process (investigator triangulation) (31). The investigators arrived at the same conclusions, which heightened our confidence in the findings (32). Different procedures, such as contextual description, have a positive influence on the transferability of the findings presented in this article (19).

Ethical considerations

The study was approved by the Medical Ethics Committee and the procedures followed were in accordance with the ethical standards of the Medical Ethics Committee of the VUmc. All respondents voluntarily took part and gave informed consent. Confidentiality was maintained by restricted, secure access to the data, destruction of audio tapes following transcription and de-identification of the transcripts.

RESULTS

Twenty-five interviews were held (11 women, 14 men) with respondents, aged between 24 and 77, with FSHD. All respondents were native Dutch and lived independently alone or together with their spouses. Nine respondents regularly or permanently used a wheelchair. Two respondents used a ventilation device during the night. None of the respondents received medical treatment for the experienced fatigue (see Table 1 for an overview of patient characteristics).

Table 1: characteristics respondents

Number	Gender	Age	Diagnosis since X years	Wheelchair	Work
1	M	38	18	-	Full time job
2	M	77	49	Wheelchair for longer distances	Retirement
3	M	62	3	-	Retirement
4	F	24	10	-	Full time student
5	M	51	10	-	Part time job
6	M	59	40	Wheelchair for longer distances	Part time job
7	M	71	5	Wheelchair for longer distances	Retirement
8	M	58	16	-	Alimony
9	F	55	2	-	Alimony
10	M	40	22	-	Full time job
11	F	57	32	Permanent wheelchair	No job
12	M	54	14	-	Part time job and alimony
13	M	66	8	-	Alimony
14	F	54	11	-	Alimony
15	F	38	9	Wheelchair for longer distances	Part time job
16	F	68	11	-	Retirement
17	M	44	29	-	Alimony
18	F	58	20	Permanent wheelchair	Alimony
19	F	40	17	Wheelchair for longer distances	Alimony
20	M	40	1	-	Alimony
21	F	51	14	-	Alimony
22	F	68	6	-	Retirement
23	M	62	4	-	Retirement
24	M	57	24	Wheelchair for longer distances	Part time job
25	F	53	15	Wheelchair for longer distances	Alimony

Four main themes were identified: 1) fatigue: an overwhelming, capricious and unpredictable experience; 2) fatigue: a complicated combination of causes; 3) fatigue: the far-reaching influences on my life; 4) fatigue: an ongoing process of adaptation. These themes will be explained below. Representative quotes are used to illustrate the findings. Each quote is followed by the number of the respondent and his or her gender and age.

Fatigue: an overwhelming and unpredictable experience

Respondents described their fatigue as an encompassing and overwhelming experience with both physical and psychological dimensions. The fatigue is first of all described as a physical experience. Respondents expressed an actual feeling of tiredness in their body as can be illustrated by a quote from one of them.

'My legs are heavy and it feels like being a pudding. My body is so tired.' (2,M,77)

The physical fatigue can be unpredictable and capricious. Sometimes people become overwhelmingly tired. Things can easily, without notice, be too tiring, and sometimes it is hard to know the causes of the fatigue. This makes the fatigue overwhelming and unpredictable for respondents.

'I don't trust my body. The fatigue can be there suddenly without any warning.' (4,F,24)

Furthermore, the physical, bodily fatigue has a serious, encompassing impact on the lives of patients with FSHD as will be explained later on. Along with the actual experienced physical fatigue, respondents described a more psychological element of fatigue: the fear of becoming extremely tired, which makes them sometimes reluctant to do something.

'I had a birthday of my cousin. I didn't want to go because I thought it would be too exhausting. But that's just fatigue in my head. It's a mental thing. Based on my physical tiredness there was no reason to stay at home.' (5,M,51)

Even thinking about the possibility of becoming tired can make people tired.

'I'm afraid of getting tired, even before I start. Having the assumption that I will become tired did, even if I didn't do anything at all, lead to more tiredness.' (3,M,62)

Fatigue: a complicated combination of causes

Respondents mentioned some factors that may influence the experienced fatigue. We will describe these factors below. The disease itself is, according to the respondents,

a first important cause of the fatigue. Having FSHD goes hand in hand with being tired.

'I'm very often tired, more than I wished to be. But I know it's the disease.' (7,M,71)

Because they see fatigue as a result of the FSHD, the respondents feel they do not have any influence on the fatigue.

'I can't change it. The FSHD is an unchangeable fact.' (7,M,71)

Secondly, the amount of physical activity is also mentioned as playing a role in the experienced fatigue. Some of the respondents described the pitfall of doing too much, which causes fatigue.

'If I do too much with my bad legs, I will become tired.' (9,F,55)

Other respondents, however, describe the negative influence of taking too much rest.

'I'm resting too much. I'm going to rest if I'm tired but by doing that I'm in a negative loop which makes me even more tired.' (5,M,51)

The third mentioned factor causing or influencing the experienced fatigue is stress.

'I had a lot of stress and I worried a lot. The fatigue disappeared when everything was arranged and the stress became less severe.' (13,M,66)

Many respondents, however, mentioned the fact that they are just extremely tired without knowing the exact causes.

'I think it [the fatigue] is the result of a combination of elements. But which ones? And how does it work? I really don't know.' (13,M, 66)

The unpredictable nature makes the experienced fatigue different from the normal fatigue respondents used to experience before the symptoms of FSHD.

'I used to know the causes of being tired. I was always able to explain it: being busy at work, having a bad night..... But now... I often can't explain it.' (13,M, 66)

Not knowing the causes and the unpredictable and capricious character of the fatigue make it quite difficult to deal with, as will be explained below.

Fatigue: the far-reaching influences on my life

Respondents described the far-reaching consequences of fatigue on their lives. In some cases fatigue, rather than decreased muscle strength, is experienced as the biggest impairment.

'For me, the fatigue is a bigger problem than the fact that I can't walk adequately.'
(9,F,55)

In general, respondents felt that the fatigue makes things that were possible in the past impossible now. They are no longer able to do the things they want to do.

'It is an awful disease. I can't do the things I would like to do. It is like having a Pac-Man in your life; he's eating and eating till nothing is left.' (9,F,55)

Respondents declared, furthermore, that they need more time and energy to participate in activities, which has an influence on the amount of things they can do.

'I used to do things in one day. Now I need a week.' (1,M,38)

More specifically, the fatigue may, according to respondents, influence their ADL activities, their social participation and participation in work.

'I used to be a skipper but started to do other work when my job became too exhausting. After some years, my body forced me to stop working. I had lost the fight against my tiredness.' (13,M,66)

The respondents furthermore described that the fatigue can also have an impact on their intimate relationships and may lead to an overburdening of their partners.

'I'm disabled and tired, but my husband is also restricted. He has to follow my tempo.' (9,F,55)

'I'm leaning too much on my wife now. It [the care] is becoming too aggravating for her.' (6,M,59)

Contacts with friends are also described as being influenced by the fatigue. Not being able to join in activities may lead to incomprehension.

'If friends ask me out for a date in a few days, I cannot say yes immediately. I have to decide it on the day itself; only then do I know how tired I am and sometimes I'm not able to go out.' (4,F,24)

The loss of possibilities and participation may, according to respondents, lead to frustration.

'It's taking a step backwards, and that's frustrating.' (17,M,44)

Fatigue: an ongoing process of adaptation

The overwhelming, capricious and unpredictable nature of fatigue forces patients to adapt themselves constantly to the actual situation. Fatigue is not the same as tiredness; taking some rest or having a good night's sleep is not enough to solve the exhaustion. Respondents explained their search to deal with the fatigue and find the right balance between doing too much and doing too little, setting borders and making choices. Finding such a balance can be difficult and exhausting as well; respondents explained that using the right strategy influences the fatigue in a positive way while using a suboptimal strategy can be seen as a significant additional cause of the fatigue.

Respondents mentioned several strategies that were used in order to prevent becoming tired or to make the fatigue less severe. One of the most important strategies is to find a balance between doing too much and doing too little. It is all about listening to your body and setting borders.

'I don't have a time-driven but an energy-driven agenda now.' (6,M,59)

However, respondents reported problems with listening to their bodies and setting borders. They often try to *'go on without giving up'* even if they should take some rest.

'I'm just going on. Sometimes I feel that things aren't going well but I really don't want to rest at such moments.' (1,M,38)

Not being able to set borders or crossing one's borders can negatively influence the fatigue.

'I did not say "No" very often. I just followed other people, but by doing that I got extremely tired. You should say "I can't do that".' (14,F,54)

It is thus difficult to find the right balance and many respondents are doing too much. Opposite to the strategy of just going on is the strategy of being very careful and taking rest.

'I'm afraid to cross the borders. For me it's all or nothing, so I don't do anything at all.' (1,M,38)

Respondents explained that it is rather difficult to find the balance between doing too much and doing too little because it is difficult to predict the fatigue.

'I have to find the borders, and I have to think and plan in advance. But that's hard, and it is difficult to anticipate the effects of activities.' (4,F,24)

Dealing with the unpredictable and capricious character of the fatigue and a constant lack of energy also requires other adaptation strategies such as omitting things.

'I have to make choices, and I have to set priorities.' (18,F,58)

However, making choices, according to respondents, is not always easy.

'Life is full of possibilities. It is such a rich life. As a result of that, I would like to do everything. I want too much...That's my problem.' (11,F,57)

Nothing can be taken for granted anymore and things that were doable in the past are not self-evident anymore. The lives of patients with FSDH are no longer controlled by themselves but by the (unpredictable) fatigue. It is not surprising that respondents try to stay as much in control as they can. This may, however, clash with the fact that they should ask for help in order to prevent or overcome their fatigue.

'I have problems with delegating activities because I want to stay in control. But I'm not able to do anything by myself anymore because of the tiredness.... I have to delegate.' (11,F,57)

Dealing with the all-encompassing influence of the fatigue on their lives requires a positive mind. Respondents try to think as positively and realistically as possible since they have the experience that this decreases the fatigue.

'I'm always looking for the positive aspects in my situation.' (9,F,55)

They also try to manage their expectations in order not to feel let down.

'I don't have any expectations anymore because expectations lead to disappointments and being tired as a result.' (6,M,59)

Furthermore, they try to have faith and avoid being worried since losing faith and being worried may lead to fatigue.

'I was losing my guts and that made me feel tired.' (2,M,77)

Dealing with their fatigue requires therefore a constant adaptation to the actual situation and the use of the right strategy. Respondents have the experience that using the right strategy helps them to decrease the fatigue. However, finding the right strategy and the right balance takes some time and may cause tiredness in itself.

'You have to search and adapt over and over again and that's making me extremely tired.' (9,F,55)

Respondents described furthermore how the progressive character of the disease makes acceptance and adaptation to the disease and the fatigue ever more difficult.

'It is an ongoing process of adapting to new circumstances. You have to change your priorities time and time again.' (18,F,58)

DISCUSSION

This is the first study to give insight into FSHD patients' experiences of fatigue. Four themes were identified: fatigue as an overwhelming and unpredictable experience, the complicated combination of the causes of fatigue, the far-reaching consequences of fatigue and the process of adaptation to fatigue. We will discuss these themes in more detail below.

With regard to the first theme, the overwhelming and unpredictable character of fatigue, it can be concluded that people with FSHD make a distinction between actual fatigue and the fear of becoming fatigued whereby the fatigue is experienced as unpredictable and sometimes unaccountable. Respondents did not make a distinction between physical fatigue and mental fatigue (such as memory and concentration problems), which is often made by other patient groups (16,33). There is also literature suggesting that there are various dimensions of fatigue, including physical/neuromuscular, emotional/affective and mental/cognitive aspects (34). We cannot explain why our respondents did not mention mental fatigue. The fact that they did not mention it does not mean that it does not exist.

People with FSHD described, secondly, several causes of fatigue. The fatigue can, according to respondents, be caused or influenced by the FSHD itself, the amount of physical activity and the amount of stress. These contributing factors are partly in line with the model of Kalkman and colleagues (36) in which perpetuating factors for fatigue in FSHD are described. The model is based on questionnaires, scales, self-observation lists and measurements of physical activity (36). Muscle strength/weakness as a result of the FSHD (36) seemed to be an important perpetuating factor influencing fatigue in FSHD. It is interesting to know whether the muscle strength/physical condition also affects fatigue in other similar diseases. The literature shows that physical condition does indeed seem to influence the fatigue in other diseases. A recent systematic review shows, for instance, the influence of spasticity of people with multiple sclerosis (MS), spinal cord injury (SCI) and stroke on fatigue (37).

Our study also suggests the influence of physical activity on fatigue. The influence of self-reported physical activities on fatigue in FSHD is also described in the model of

Kalkman and colleagues (36). Recent systematic reviews also describe the influence of exercise/physical activity on fatigue in similar diseases such as rheumatoid arthritis (38) and post-polio syndrome (39).

Our respondents also mentioned the influence of coping and stress on the experienced fatigue. These factors are not described in the model of Kalkman and colleagues (36). They included only measurements of depression and psychological distress in their study and did not investigate the potential influence of coping and stress on fatigue (36).

Research on stress, coping, fatigue and FSHD is, however, scarce and focuses only on the influence of coping on pain (40) and not on fatigue. Voet and colleagues do conclude that cognitive behavioural therapy can decrease fatigue in patients with FSHD, but it is not clear which elements of the therapy are effective and whether coping influences the experienced fatigue or not (41). Recent published systematic reviews on other similar diseases such as rheumatoid arthritis (42), juvenile idiopathic arthritis (43) and post-polio syndrome (39) confirm our finding: coping and/or stress may influence the experienced fatigue (39,42,43).

With regard to the third and fourth finding, the consequences of fatigue and the adaptation process, patients explained the large impact of fatigue on their lives. Our study shows that fatigue can even have a bigger impact than physical restrictions/complaints. Due to the progressiveness of the disease, people have to adapt themselves constantly to the changing situation. Patients described fatigue as being disabling as it sometimes leads to participation problems (e.g. in social life and work abilities) and changing relationships. Nowadays, society has high expectations regarding participation; people have to be able to keep up with a high tempo in terms of their work and social relations. Patients have to deal, consequently, not only with physical impairments as a result of decreased muscle strength but also with a disability that can be described as *'something imposed on top of our impairments by the way we are unnecessarily isolated and excluded from full participation in society'* (35). This means that a disability is not a state of the body but rather a social situation in which patients, in this case those with FSHD, are not able to fully participate due to the consequences of their disease.

The loss of possibilities and participation and the high expectations of the current society can lead to frustration and powerlessness. Dealing with the disease is needed. This can be done by setting borders, making choices and dividing energy in an adequate way. Doing this can lead to less fatigue and can also have an impact on patients' lives. The finding that severe fatigue often has a significant impact on the lives of patients with FSHD is in line with studies of other similar patient groups (e.g. 15,16). A former quantitative study on the physical and social functioning of people with FSHD also shows problems in these domains (6).

In conclusion, this study gives us important new insights into how patients with FSHD experience fatigue, which factors influence the fatigue, how fatigue affects the lives of patients with FSHD and how patients try to deal with it. These findings are partly in line with the experiences of people with other similar chronic diseases.

Critical readers may mention that this study is 'just' a qualitative study, which makes the results difficult to generalize. The sample size was indeed relatively small compared with that of a quantitative study, but it was very reasonable for a qualitative one. The results can be seen as valid since we have reached saturation (22,28). Nevertheless, the reader must decide whether the results are generalizable for their practice, and, while doing so, one should bear in mind that this study described the experiences of patients with severe fatigue. Although 61% of patients with FSHD experience severe fatigue (2), there are still patients who do not experience any fatigue at all.

CONCLUSION

Fatigue has a significant influence on the lives of patients with FSHD but patients are insufficiently supported in coping with their fatigue. Patients should be helped to reduce fatigue, especially because they experience a negative impact on their quality of life and participation. To the best of our knowledge, interventions aimed at reducing fatigue are not often described in the literature. One study shows that improving muscle strength by strength training and/or (anabolic) medication is not successful in patients with FSHD (41). Recently, an RCT showed that aerobic exercise training and cognitive behavioural therapy can be effective in reducing fatigue (41). Broader implementation of such interventions is needed to reduce fatigue and improve the quality of life of people with FSHD.

COMPETING INTERESTS

The authors declare that they have no competing interests. This study is funded by the Prinses Beatrix Fonds (PBF) (the Dutch Public Fund for Neuromuscular and Movement Disorders) and the Netherlands Organization for Health Research and Development (ID: ZonMW 89000003).

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