Upper limb function and activity in people with facioscapulohumeral muscular dystrophy: a web-based survey

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ABSTRACT

Purpose To investigate the upper extremity (UE) at the level of impairments and related activity limitations and participation restrictions in people with facioscapulohumeral muscular dystrophy (FSHD).

Methods The study was conducted using web-based questionnaires that were distributed amongst people with FSHD in the Netherlands. Eighty-eight respondents started the survey, and 71 completed it. The questionnaires covered the following dimensions: Function, Activity and Participation of the International Classification of Functioning Disability and Health.

Results More than 40% of the respondents experienced pain in one arm or both the arms. Increased pain and stiffness scores and longer disease duration were associated with increased limitation scores. For basic activities, lifting the arm above shoulder-level was most frequently reported as most limited, coherent with the clinical picture of FSHD. Among the respondents, 50% indicated restrictions at school, 78% indicated restrictions at work and more than 80% indicated restrictions whilst participating in sports, hobbies, household activities and romantic relationships.

Conclusions This study has shown that alongside the well-known problem of lifting the arms above shoulder-level, UE activities below shoulder height during vocational and occupational activities are also problematic in patients with FSHD. Alongside disease duration, pain and stiffness are associated with UE activity limitations.

IMPLICATIONS FOR REHABILITATION

- Attention is needed for pain and experienced stiffness in the upper extremity as it is frequently present in patients with FSHD.
- Rehabilitation professionals need to be aware that patients with FSHD not only experience problems with activities above shoulder height, but also with activities below shoulder height.
- At least 50% of the patients with FSHD experience restrictions in participation as a result of limitations in their UE.

Introduction

Facioscapulohumeral dystrophy (FSHD) is an autosomal dominant, slowly progressive type of muscular dystrophy, with an estimated prevalence of 1:21 000.[1,2] FSHD is, thus, one of the most common inherited muscular dystrophies with the first signs of weakness occurring in the muscles of the face and shoulder girdle.[3] Fatty infiltration with loss of muscle fibres gradually results in weakness of nearly all skeletal muscles.[4] Despite the typical pattern of muscle involvement described in the literature,[5] the upper extremity (UE) impairments are highly variable among people with FSHD with regard to both proximal and distal muscle weakness.[6,7] In addition, no clear correlations have been found between muscle strength and gender, age or duration of symptoms.[8]

Although the clinical picture of muscle weakness is variable, most persons with FSHD experience activity limitations when elevating the arms above shoulder-level.[9,10] It is also known that the execution of many activities of daily living (ADL) takes considerably longer in persons with FSHD compared to healthy subjects.[6,9] Most people with FSHD are unable to raise their hands above their heads, but are able to bring their hands to their mouths. Hand function itself remains largely intact.[8] Interestingly, increased muscular co-contraction has been observed in the UEs of people with FSHD,[4,9] as well as enhanced electromyography activity in the shoulder muscles during arm elevation movements.[9]

To develop tailored training interventions and new supportive aids, it is not only important to better

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understand UE impairments and limitations in basic UE activities, but also to obtain knowledge about limitations in more complex, instrumented UE activities and restrictions in social participation. Currently, little is known about the UE capacity and performance of daily activities in people with FSHD. Hence, the aim of this study was to obtain a better understanding of the UE impairments and related limitations in activities and restrictions in participation in people with FSHD. In a previous study of boys and men with Duchenne muscular dystrophy (DMD), a web-based survey that consisted of a set of questionnaires covering all relevant dimensions of the International Classification of Functioning Disability and Health was used. The same web-based survey was used in this study. Based on the clinical picture of people with FSHD, it was hypothesized that especially activities above shoulder-level would be limited, but also that several participation restrictions would be present, independent of working above shoulder-level. In addition, it was hypothesized that UE activity limitations would be associated with disease duration, pain and stiffness.

**Methods**

**Participants and procedure**

A web-based survey (QuestionPro; Survey Analytics LLC, Seattle, USA) was designed. This survey included questions concerning pain and stiffness, as well as items to assess basic and daily (instrumented) UE activities. The survey was sent via the Dutch patient organization, "Spierziekten Nederland", to their Dutch-speaking members with FSHD in the Netherlands, in December 2011. The recruitment period was from December 2011 until February 2012. Ethical approval was obtained from the local Research Ethics Committee (Ethical Committee Arnhem-Nijmegen, 2008/341).

**Outcomes**

Outcomes were divided into four categories: (1) participant characteristics; (2) impairments; (3) activity limitations; and (4) participation restrictions.

**Participant characteristics**

The participant characteristics assessed were: age, age of diagnosis, disease duration, hand preference, wheelchair confinement and use of assistive devices for the arms. For the analyses, the originally preferred arm was used.

**Impairments**

The questions on pain and stiffness were adapted from the University of Michigan Upper Extremity Questionnaire. Both pain and stiffness scores were divided into the aspects frequency (range: 0–6, with the higher the score, the higher the frequency), severity (range: 0–10, with the higher the score, the more severe) and disablement due to pain or stiffness (range: 0–10; with the higher the score, the more disabling). Combination scores were calculated for each part of the arm, by taking the sum of the three aspects (range: 0–26). A similar approach was used previously in the study of Janssen et al. Along with the scores for each segment of the arms, total sum scores for both the right and left UE were calculated (range: 0–182). For each segment, percentages of respondents who experienced pain and stiffness (combination score >1) in either the right or left UE were determined.

**Activity limitations**

The Brooke scale was used to classify UE activity, the Capabilities of Upper Extremity Questionnaire (CUE) was used to assess basic UE activities and the ABILHAND-plus to assess UE capacity to perform daily activities. The CUE contains 17 items (of which 15 one-handed, scored on both right and left arm and 2 two-handed), yielding in 32 scores with a seven-point scale (1 = unable to perform, 7 = can perform without difficulty). The CUE questionnaire in this study had 16 items in total, because the item “hold a hammer” was missing. The ABILHAND-plus questionnaire contained 26 items (22 items that were described by Vandervelde et al., and 4 additional items that were indicated as important by boys with DMD that were scored on a three-point scale (0 = impossible, 1 = difficult and 2 = easy)). Participants were also asked to select the five items that were most important to them and to report the activities in daily life that caused most problems due to UE impairments or activity limitations.

**Participation restrictions**

To assess the level of participation, a set of open questions was used. Respondents were asked if they went to school, had a job, practiced sports, had hobbies, participated in household activities, performed activities with friends and/or were in a romantic relationship. Respondents who reported that they participated in a specific role were asked if they were restricted due to UE impairments or activity limitations.

**Statistical analysis**

The mean and standard deviation were calculated for age, age of diagnosis and disease duration. Because not
all data were normally distributed, we used non-parametric statistics. The median and inter-quartile ranges (IQR) of the pain and stiffness combination scores and of the Brooke, CUE total, CUE right/left and ABILHAND-plus scores were determined. Moreover, the percentages of respondents who reported a CUE activity to be very, extremely or totally limited, the percentages of respondents who reported an ABILHAND-plus activity to be difficult or impossible, and the percentage of respondents who mentioned an ABILHAND-plus activity as important were calculated. Answers to the open question “What are the most important problems you encounter in daily life due to limitations in your arms and/or hands?” were categorized. The activities that restricted the performance of specific social roles were determined and categorized for each of these roles.

Wilcoxon signed rank tests were used to compare differences in pain, stiffness and CUE scores between the right and left UEs and between the preferred and non-preferred UEs. Spearman rho correlation coefficients of the pain, stiffness, Brooke, CUE and ABILHAND-plus scores with disease duration and age were calculated, as well as Spearman rho correlation coefficients of CUE and ABILHAND-plus scores with the Brooke scale. Finally, backward multivariate linear regression analyses were performed to determine the effect of disease duration (in years), pain and stiffness on basic arm activities measured with the CUE. All statistical analyses were carried out using IBM SPSS Statistics Version 20 for Windows (IBM®, Somers, NY). If a participant did not fully complete the survey, all completed items were included in the analysis.

Results

Participant characteristics

A total of 88 respondents began the survey, of whom, 71 (81%) completed it. As a result, the number of respondents varied between the different domains of the questionnaires (see tables). The mean age of all respondents (N=88) was 51.1 ± 14.8 years, the mean age of diagnosis was 32.0 ± 16.0 years and the mean disease duration was 19.1 ± 13.1 years. The percentage of participants with FSHD that were confined was 17.6%. The same percentage (17.6%) of participants with FSHD that were completely wheelchair-confined was 17.6%. The same percentage (17.6%) of participants with FSHD that were confined was 17.6%. The same percentage (17.6%) of participants with FSHD that were confined was 17.6%. The same percentage (17.6%) of participants with FSHD that were confined was 17.6%

Impairments

Seventy-one participants (81%) were right-handed. Ten participants (11%) reported that they switched their preferred arm (nine from right to left, and one from left to right). The median pain and stiffness scores of the right UE were generally higher than that of the left UE. However, these differences were far from statistically significant for the shoulder and arm (p > 0.5). For the wrist (p = 0.04), thumb (p = 0.06) and fingers (p = 0.08) the differences were (nearly) significant. The overall pain combination score was significantly higher for the right arm (p = 0.03). The overall stiffness combination score did not reach statistical significance for the left-right-difference (p = 0.06). When the preferred arm was considered, no significant differences were found between the summed pain and stiffness scores of both arms (p = 0.25 and 0.31, respectively). Pain was most severe in the shoulders and upper arms. The percentage of respondents who experienced pain (combination score > 1) in one arm or both the arms varied between 43.9% and 89.0% for the different segments (Table 1). The overall pain score correlated with disease duration (rs = −0.28, p = 0.01), but not with age (rs = −0.09, p = 0.45). Stiffness was also most severe in the shoulders and upper arms. The percentage of respondents who experienced stiffness in one arm or both the arms varied between 40.3% and 66.2% for the different segments (Table 1). No significant correlations were found between stiffness scores and disease duration or age.

Activity limitations

The median Brooke scale was 2 (IQR 2−3). The Brooke scale (rs = 0.43, p < 0.01) and the CUE score (rs = −0.37, p < 0.01) correlated with disease duration, but there was no correlation between disease duration and the ABILHAND-plus sum score (rs = −0.11, p = 0.35). No significant correlations between Brooke, CUE and ABILHAND-plus scores for age were found. The Brooke scale significantly correlated with the CUE score (rs = −0.48, p < 0.01) and the ABILHAND-plus score (rs = −0.31, p < 0.01).

The median CUE total score was 155.5 (IQR 117.8–176.3), which was 70% of the maximum possible score. The median of both the CUE right and CUE left score was 75.0 (73% of the maximum possible score). Wilcoxon Signed-Ranks Tests indicated that the CUE right score was significantly lower than the CUE left score (p = 0.02) and that the CUE score of the preferred arm was lower compared to the non-preferred arm (p = 0.03) (Table 2). However, the medians of the CUE right/left scores as well as the CUE preferred/non-preferred scores were equal, indicating a minimal scale of the difference. The percentage of respondents who reported CUE activities being very, extremely or totally limited was higher for the items considering the right arm as for the left arm items.

The median ABILHAND-plus sum score was 44.0 (IQR 35.0–50.5), which was 84% of the maximum possible
score. The three activities that were most often indicated as difficult were “buttoning up a shirt” (56%), “taking the cap of a bottle off” (47%) and “fastening the zipper of a jacket” (44%). The three activities that were most often reported as important were “using the keyboard of a computer” (66%), “using a knife and fork” (52%) and “fastening the zipper of a jacket” (45%) (Table 2).

Multivariate regression analysis for the right arm revealed that the time since diagnosis, pain and stiffness explained 34% of the variance of the CUE. For the left arm these factors explained 40% of the variance. Table 3 shows that for both the arms, pain, stiffness, and disease duration were all independently associated with limitations in basic UE activities as assessed with the CUE.

The answers to the open questions revealed that reaching for and lifting of objects above shoulder level caused most problems, followed by personal care and carrying objects (Table 4).

### Participation restrictions

Table 5 shows the percentage of respondents who reported participation restrictions, as well as the UE activities they experienced as most limited within different participation domains. Of all respondents, 11% went to school, 51% had a job or did voluntary work, 30% participated in sports, 94% had a hobby, 75% participated in household activities and 81% had a romantic relationship. Fifty percent of the respondents indicated restrictions at school and 78% reported restrictions at work. More than 80% indicated restrictions whilst participating in sports, hobbies, household activities and romantic relationships. Carrying study materials, using a computer and performing household activities above shoulder height (such as cleaning windows and hanging laundry) were specific activities that were reported as limited in at least 25% of the participants, who were involved in school, work and household activities, respectively.

### Discussion

To the best of the researchers’ knowledge, this is the first study to explore upper extremity (UE) pain, stiffness, activity limitations and related participation restrictions in people with facioscapulohumeral muscular dystrophy (FSHD). The most important result is the frequent presence of pain and stiffness in the shoulders and upper arms, which was associated with UE activity limitations as assessed with the CUE. Besides pain and stiffness, time since diagnosis was independently associated with basic UE activity limitations. Together, these factors explained a substantial amount of the CUE variance (34–40%). The top three activities that were most often reported as being limited were “use of computer keyboard”, “use of knife and fork” and “fastening zipper of jacket”. Half of the respondents indicated restricted participation at school in relation to UE activity limitations, 78% indicated restrictions at work and more than 80% whilst participating in other social roles.

With regard to pain and stiffness, it is well known that reaching upwards causes increasing difficulty in people with FSHD, when muscle weakness progresses.
The most often applied strategy is making a ballistic movement of the arm, by means of the trunk muscles, through which the arm is “thrown upwards”. This rapid compensatory movement may easily lead to overburdening or even micro-damage of structures in and around the shoulder, which in turn, may explain the pain and/or stiffness scores of this body segments.

A difference was found between the right and left arms with regard to the prevalence of pain complaints (medians 39.0 vs 31.5, \(p = .003\)). Because the majority of

| Brooke scale | 76 | 3 (2–3) |
|--------------|----|---------|
| CUE score total (max score = 210) | 74 | 155.5 (117.8–176.3) |
| CUE score right (max score = 98) | 74 | 75.0 (55.5–84.0) |
| CUE score left (max score = 98) | 74 | 75.0 (59.5–85.0) |
| CUE score both (max score = 14) | 74 | 6.0 (3.8–8.3) |
| ABILHAND-plus score (max score = 52) | 73 | 44.0 (35.0–50.5) |

| CUE N | % respondents who answered the activity to be very, extremely or totally limited (right/left/both arms) |
|-------|----------------------------------------------------------------|
| Reach forward at shoulder-level | 74 | 41.9/36.5/* |
| Arms over head | 74 | 79.7/73.0/* |
| Reach to the floor | 74 | 39.2/36.5/* |
| Raise a five pound object over the head | 74 | */*/62.2 |
| Slide a light object towards you | 74 | 10.8/9.5/* |
| Slide a ten pound object towards you | 74 | 40.5/37.8/* |
| Slide a light object away from you | 74 | 12.2/8.1/* |
| Slide a ten pound object away from you | 74 | 37.8/29.7/* |
| Push up in chair | 74 | */*/58.1 |
| Curl wrist upward | 74 | 18.9/13.5/* |
| Supination | 74 | 14.9/8.1/* |
| Hold a hammer | 74 | 14.9/10.8/* |
| Pick up a small object with thumb and first two fingers | 74 | 14.9/10.8/* |
| Hold a small object between thumb and index finger | 74 | 13.5/8.1/* |
| Hold/open a two pound object with the tips of the fingers | 74 | 25.7/20.3/* |
| Manipulate a small object with the fingers | 74 | 17.6/12.2/* |
| Push a button with tip of the index finger | 74 | 9.5/5.4/* |

| ABILHAND-plus N | Difficult or impossible (%) | Important (%) |
|-----------------|-----------------------------|--------------|
| Take the cap off a bottle | 73 | 46.6 | 24.7 |
| Cut nails | 73 | 38.4 | 21.9 |
| Button up a shirt | 73 | 56.2 | 38.4 |
| Fasten the zipper of a jacket | 73 | 43.8 | 45.2 |
| Turn a key in a keyhole | 73 | 21.9 | 37.0 |
| Fasten a snap e.g. from jacket or bag | 73 | 32.9 | 16.4 |
| Open a pack of chips | 73 | 31.5 | 2.7 |
| Open a pack of biscuits | 73 | 31.5 | 5.5 |
| Insert a key in keyhole | 73 | 20.5 | 41.1 |
| Turn off a tap | 73 | 21.9 | 35.6 |
| Turn on a tap | 73 | 21.9 | 37.0 |
| Fill a glass with water | 73 | 17.8 | 19.2 |
| Sharpen a pencil | 73 | 17.8 | 2.7 |
| Open a lunch box | 73 | 21.9 | 4.1 |
| Squeeze toothpaste onto a toothbrush | 73 | 15.1 | 20.5 |
| Spread butter on a slice of bread | 73 | 17.8 | 37.0 |
| Open a toothpaste tube | 73 | 21.9 | 12.3 |
| Count banknotes | 73 | 20.5 | 8.2 |
| Handing out cards | 73 | 28.8 | 8.2 |
| Unwrap a chocolate bar | 73 | 16.4 | 4.1 |
| Dry hands | 73 | 15.1 | 17.8 |
| Wash hands | 73 | 16.4 | 32.9 |
| Eat with a spoon | 73 | 28.8 | 27.4 |
| Use knife and fork | 73 | 38.4 | 52.1 |
| Drink a glass of water without straw | 73 | 24.7 | 42.5 |
| Use keyboard of a computer | 73 | 27.4 | 65.8 |

ABILHAND-plus = measure to assess UE capacity to perform daily activities, Brooke scale = measure to classify UE activity, CUE = measure to assess basic UE activities, IQR = Inter Quartile Range, * = not applicable.

aPercentage of respondents who answered the activity to be difficult or impossible.

bPercentage of respondents who identified the activity to be important. Respondents were asked to identify the five most important activities.
people are right-handed, this right/left difference may be explained by slight overuse of the right arm. When arm preference was, however, considered, no side difference between the pain scores was found when comparing the preferred and non-preferred arms. This could indicate an asymmetric involvement of the disease that is independent from arm preference. This would be consistent with the results presented by Rijken et al., who demonstrated more fatty infiltration on the right body side compared to the left independent of arm preference, in 70 patients with FSHD, using computed tomography (CT) scans.[5] The presented results did not show a clear side difference in limitations of basic activities (measured with the CUE).

Activities such as sliding an object over a table and manipulating a light object with the fingers were less difficult, probably because these types of activities make less use of proximal muscles. However, a striking finding was the large percentage of participants that reported difficulties with desk-based activities, such as writing or using a computer. One explanation for this finding may be that the participants performed these activities without proper ergonomic positioning, whilst using their weakened arm and shoulder muscles too much. Another explanation could be that these activities are often performed over a longer time period, inducing muscle fatigue. Both explanations would support a rehabilitation strategy, in which the arms are supported by (adaptive) arm supporting devices during desk-based activities.

Both the Brooke and the CUE score were moderately ($r_s = 0.43$ and $r_s = -0.37$, respectively) correlated with disease duration. This is to be expected, as both scales assess the ability to lift the arms, which is a major problem for many persons with FSHD. In contrast, no correlation was found between the disease duration and the ABILHAND-plus. This can be explained by the fact that most activities assessed by the ABILHAND-plus involve a combination of arm and hand use, in which people can apply compensation strategies in order to succeed. It is also possible that the ABILHAND-plus is less sensitive to ‘minor’ activity limitations than the CUE, since the CUE contains questions aiming at basic activities with the arm or hand only. At the impairment level, only pain was associated with disease duration, but this association was only weak ($r_s = -0.28$), suggesting a more indirect relationship between pain and progression of muscle weakness.

The mean age was 51 years, which clarifies that a low percentage of the respondents indicated going to school (11%) and a higher percentage indicated having work (51%). The total percentage of respondents who indicated participation in either school or work was 62%. It is possible that some respondents do not participate in a given domain at all due to limitations, although they would like to. In fact, it is likely that they are most restricted. The actual percentage of restrictions may therefore be higher than presented in this study.

### Study limitations

Of all respondents who started the survey, 81% completed the survey which may have caused bias. Since the questionnaire was sent not only by email, but also via a link in a public digital newsletter, it is unknown how many persons received the invitation. This may also have caused bias. Such response bias may have obscured the experiences of either more severely or less severely affected persons. Moreover, the results of this study are based on a survey among the Dutch population with FSHD. Because the standards of care are relatively high in the Netherlands, generalization of these results to FSHD patients in other countries requires caution. Another limitation of this study is that the gender distribution of the respondents remained unknown. The applied questionnaire was originally designed for people with DMD. Since DMD is extremely

| Factors | CUE Right arm | Beta | Standard error | p Values | 95% Cl |
|---------|--------------|------|----------------|----------|--------|
| Duration | -0.67 | 0.15 | <0.01 | -0.98 | -0.36 |
| Pain right | -0.14 | 0.06 | 0.03 | -0.27 | -0.02 |
| Stiffness right | -0.10 | 0.05 | 0.07 | -0.21 | -0.01 |
| CUE Left arm | -0.63 | 0.13 | <0.01 | -0.90 | -0.37 |
| Pain left | -0.16 | 0.06 | 0.01 | -0.27 | -0.05 |
| Stiffness left | -0.09 | 0.05 | 0.07 | -0.19 | -0.01 |

* $R^2 = 0.34$,  
* $R^2 = 0.40$.

Table 3. Variables independently associated with limitations in basic upper extremity activities as assessed with the CUE based on linear multivariate regression analyses.

| Activities | %a |
|------------|----|
| Reach/lift objects above shoulder level | 45 |
| Personal care activities | 32 |
| Carry objects | 26 |
| Eat/drink | 19 |
| Get dressed | 19 |
| Prepare food/household tasks | 12 |
| Using the computer/use telephone/write | 11 |
| Open cans/bottles | 7 |
| Use the toilet | 3 |

*aPercentages of respondents who mentioned the activity when asking the open question “What are the most important problems you encounter in daily life due to limitations in arms and/or hands?”.

Table 4. Most limited upper extremity activities as reported by people with FSHD ($N = 73$).
rare in women, gender was not questioned. Erroneously, this adjustment was not corrected in this study. The results of this study provide little information about young persons with FSHD. Both the mean age of the study sample and the mean age of diagnosis were relatively high compared to other studies.[8] Only one participant was younger than 18 years. One explanation could be that children and teenagers do not yet experience severe UE activity limitations or do not want to be confronted with these limitations by participating in this survey.

**Recommendations**

For future studies in patients with FSHD, a set of measures including the Brooke classification, pain and stiffness questionnaires and the CUE are recommended. In addition, a patient-centred participation scale, such as the Canadian Occupational Performance Measure (COPM) should be considered, to provide more insight in patient specific limitations. To evaluate the effects of individually tailored exercises and supportive devices, objective measures are recommended, that provide also quantitative kinematic and electromyography parameters. Such measures include e.g. 3D-kinematic movement analysis[17] and the use of accelerometry to assess actual motor performance activities in daily life.[18]

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