Psychological parameters impact health-related quality of life in mental and physical domains in adults with muscular dystrophy

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Abstract

The impacts of potentially treatable psychological parameters on quality of life are relatively unreported in adults with Facioscapulohumeral, Becker and Limb-girdle muscular dystrophy. The purpose of this study was to compare quality of life, psychological parameters, and physical function between adults with muscular dystrophy and controls, and to examine relationships among these parameters in muscular dystrophy. Twenty-one adults with muscular dystrophy (n=7 Becker, n=8 Facioscapulohumeral, n=6 Limb-girdle) and ten age-matched controls participated. Outcome measures were health-related quality of life, depressive symptoms, trait anxiety, self-esteem, physical self-worth and six-minute walk distance. Quality of life scores were lower in the muscular dystrophy groups than the control ($p < .05$). Depressive symptoms had the greatest association with quality of life in the Mental Health domain ($r= -.89$, $p < .001$). Depressive symptoms also had the most associations with quality of life (7 of 10 domains), followed by trait anxiety (6 of 10 domains), physical self-worth (5 of 10 domains), self-esteem (4 of 10 domains) and six-minute walk distance (3 of 10 domains). Psychological parameters and, to a lesser extent, physical function impact quality of life in muscular dystrophy. This study provides a rationale to include psychological assessment and treatment within muscular dystrophy healthcare.

Keywords: Mental health, Depression, Self-esteem, Anxiety, Physical function

1. Introduction

Muscular dystrophy (MD) is a collection of inherited neuromuscular disorders caused by mutations in various genes [1]. Such mutations result in either a problem with, or lack of, one of the proteins within muscle cells[2]. Facioscapulohumeral (FSHD), Becker (BMD) and Limb-girdle (LGMD) MD are associated with defects in the proteins of the sarco-glycan complex [3-5]. Despite variations in genotype between MD conditions and condition-specific presentations of physical impairment, all MDs cause progressive reductions in muscle strength and a worsening ability to perform daily functional tasks such as walking [6].

Low health-related quality of life (QoL) scores have consistently been reported in individuals with impaired muscle function [7]. QoL scores in MD populations however are varied. Studies have reported lower QoL scores in children and adolescents with Duchenne MD [8,9], a combined group of children and adults with Duchenne MD, BMD and LGMD [10] and adults with FSHD [11,12] compared to controls or normative reference values, in QoL domains related to both physical and mental aspects. Other studies have found lower QoL scores in physical domains compared to controls or normative reference values but not in the mental health domain of QoL [13-16]. These latter findings have been shown in Duchenne MD [13-15] and recently in adults with LGMD and Duchenne MD [16], and they reflect the Disability Paradox Phenomenon [17] whereby self-reported QoL scores are high despite substantial physical disability.

The relationship between MD and QoL is evidently complicated. A range of disease-specific variables have been found to be associated with QoL scores in physical domains in individuals with MD. These include age [8,12,18], condition severity [12,19], functional status [9], wheelchair dependence/use [20] and age at disease onset [18]. Other parameters are

associated with both physical and mental domains of QoL in MD, including pain [12,16,21] and fatigue [16,20,22]. Whilst disease-specific parameters, such as age at disease onset, provide insight into QoL they cannot be treated, whilst aspects like pain and fatigue can be treated. It is important to establish the impact of other potentially treatable parameters on QoL scores, such as psychological aspects, as they may be as, if not more, important than physical ones.

Symptoms of depression and anxiety are known to negatively affect QoL scores in medical conditions such as diabetes [23-26], but their impact on QoL in FSHD, LGMD and BMD remain relatively unreported. There is however some evidence that QoL scores are affected by psychological parameters in MD. In adults with Duchenne MD, a negative relationship was found between anxiety and both physical and mental domains of QoL, and between depression and physical domains of QoL [27]. In adults with LGMD, a moderate negative association was found between depression and overall QoL [18], but details regarding which QoL domains were not provided. In adults with FSHD, depressive symptoms were shown to be negatively related to both physical and mental domains of QoL [12]. The impact of depressive symptoms on QoL is yet to be reported in BMD and the impact of anxiety on QoL remains unreported in FSHD, BMD and LGMD.

In addition to depression and anxiety, a person's self-perceptions play an important role in their mental wellbeing [28]. Self-esteem, a general positive or negative self-regard [29], has been shown to positively impact QoL scores in adults with multiple sclerosis [30], women with cervical cancer [31] and women with fibromyalgia [32]. The impact of self-perceptions on QoL has received little attention in MD. One study measured general self-efficacy in adults with Duchenne MD, FSHD and BMD and found small to moderate associations with QoL [16], but

no assessments of self-esteem or physical self-worth have yet been conducted in adults with MD.

Muscle deterioration is a characteristic feature of FSHD, LGMD and BMD [6]. It is therefore unsurprising that maximum knee extensor strength was found to be moderately associated with QoL scores in the Physical Function and Social Function domains in adults with BMD [16], and that hip flexor muscle strength was moderately associated with overall QoL scores in adults with LGMD [18]. These findings highlight the impact of physical disease progression on QoL scores in MD, but whether physical function has a greater impact on QoL scores than psychological health in adults with MD is undetermined. The six-minute walk (6MW) test is an established and advocated outcome measure of physical function in MD [33], which may provide insight into the effect that physical function has on QoL scores in MD.

This study will contribute to a greater understanding of how QoL scores in adults with FSHD, BMD and LGMD differs from non-dystrophic adults, and it will highlight parameters that impact QoL in adults with MD to inform future healthcare.

1.1 Aims

1) To compare QoL scores of adults with FSHD, BMD and LGMD to a non-dystrophic age-matched control group. 2) To compare between the groups for psychological parameters (depressive symptoms, trait anxiety, self-esteem, and physical self-worth) and physical function (6MW distance). 3) To examine associations between QoL scores and psychological parameters and between QoL scores and physical function in adults with MD.

2. Materials and Methods

2.1 Participants

Twenty-one adults diagnosed with one of three variants of MD (n = 7 BMD, n = 8 FSHD and n= 6 LGMD) took part, along with ten age-matched control adults. The participant characteristics of the control group, MD sub-groups (BMD, FSH and LGMD) and a pooled average for MD are described in Table 1. All participants were of sound intellectual status. Control participants were free from any known health problems, illnesses or injuries and were recruited from the general population. MD participants were in otherwise good health without any uncontrolled co-morbidity or cardiac issues and were able to walk at least seven metres with or without assistive walking devices. They were recruited from a Neuromuscular Centre (Cheshire, UK), where they receive physiotherapy (weekly, bi-weekly or monthly) to maintain function. All participants self-reported that they were un-trained and did not undertake more than one hour of intense physical activity or three hours of low-moderate physical activity per week. The Manchester Metropolitan University Ethics Committee granted ethical approval and all participants provided written informed consent before participating.

2.2 Procedures

Participant characteristics, which were age (years), stature (cm), body mass (kg), muscle strength (N.m) and work status, were collected initially. Stature and body mass were measured using a stadiometer and digital scales, and isometric maximum voluntary contraction torque during knee flexion and knee extension was measured using an isokinetic dynamometer (Cybex Norm, Cybex International Inc., NY, USA), as described in a previous study [34]. Participants then completed a 6MW test and five questionnaires, which assessed QoL scores, symptoms

of depression, trait anxiety, self-esteem, and physical self-worth. Each questionnaire was explained to participants, and investigators were available if participants required assistance.

2.2.1 Six-Minute Walk Test

Sixteen MD participants completed a 6MW test (n = 6 FSHD, n = 4 BMD, n = 6 LGMD). Five participants did not complete this test as they expressed a fear of falling. An adapted version of the protocol set out by the American Thoracic Society [35] was utilised. Participants were instructed to walk at their self-selected pace, instead of as far as possible as prescribed by the American Thoracic Society [35], due to safety concerns in the current population.

The test took place along a straight corridor. Each end of the 25 m course was marked with tape on the ground. Participants were instructed to turn clockwise at each end and walk in a straight path. One investigator walked behind the participant, keeping at least a 2 m distance, to provide encouragement (in accordance with The American Thoracic Society guidelines) and assess fall risk. Participants used their walking aids during the test if they typically used them to walk for a period of six minutes. After five minutes' seated rest, participants were readied at the start line and the timer was started once they began to walk. After six minutes, participants were seated and the distance walked was measured.

2.2.2 Quality of Life -The 36-item Short Form Health Survey Version 2

The 36-item Short Form Health Survey is a self-report generic survey that covers 36 questions on functional health and wellbeing to assess QoL from the participant's perspective [36]. Each question has a multiple-choice scale on which participants rate themselves. The answers are summarised into four physical domains (Physical Function, Role Physical, Bodily Pain and General Health), four mental health domains (Vitality, Social Function, Role Emotion and

Mental Health) and two component health scores (Physical Component Score and Mental Component Score). Scores can range between 0 - 100 on each domain. Higher scores indicate a better QoL score. The scale has excellent test-retest reliability with intraclass correlation coefficients (ICC) above .80 [36].

2.2.3 Depressive Symptoms -The Beck Depression Inventory

The Beck Depression Inventory is a 21 question, multiple choice, self-report questionnaire that measures the severity of depressive symptoms. It has excellent reliability, with a test-retest ICC of .96 [37]. The questionnaire includes items relating to symptoms of depression and participants score each question from 0 (least) to 3 (most), with a total sum score indicating overall depressive symptom severity from 0-63. Higher scores indicate increased severity of depressive symptoms. The cut scores for The Beck Depression Inventory are as follows: ≤ 10 indicates zero to minimal depressive symptoms, 11-20 indicates mild depressive symptoms, 21-30 indicates moderate depressive symptoms, and ≥ 31 indicates severe depressive symptoms.

2.2.4 Trait Anxiety - State-Trait Anxiety Inventory

The trait sub-scale of the State-Trait Anxiety Inventory [38] is a 20 question multiple choice self-report questionnaire that measures trait-anxiety (a predisposition to long-lasting and persistent feelings of anxiety that are not restricted to particular circumstances). The trait sub-scale has excellent test-retest reliability, with a reported ICC of .86 [38]. The scale includes statements for participants to score between 1 (not at all) and 4 (very much so). Total scores can range between 20-80, with higher scores indicating more severe anxiety symptoms.

2.2.5 Self-Esteem - Rosenberg Self-Esteem Scale

The Rosenberg Self-Esteem Scale is a measure of global self-esteem [39]. The scale has excellent reported reliability, with an ICC of .90 [40]. It has 10 questions that assess both the positive and negative feelings towards the self. Participants rate themselves on a 4-point Likert scale, with total scores ranging from 10 to 40. The higher the score the higher the self-esteem.

2.2.6 Physical Self-worth -The Physical Self-Perception Profile

The Physical Self-Perception Profile is a reliable (test-retest ICCs between .74 and .91) self-report questionnaire [41] that comprises thirty questions that assess physical self-worth in the global domain and self-perceptions in four sub-scales: sport competence, body attractiveness, physical strength and physical conditioning and exercise. An additional eight questions relate to perceived importance of the four sub-scales and provide a measure of importance attached by the participant to the respective sub-scale.

2.3 Data Analysis

Each questionnaire was scored according to the relevant manual. QoL data were scored using Health Outcomes Scoring Software 4.5 (Quality Metric Health Outcomes™, Lincoln, United Kingdom).

Data are presented as means and standard deviations for the control group, pooled MD group and each MD sub-group. Statistical analysis is between the MD subgroups (FSHD, LGMD and BMD) and the control group, and because no difference in primary outcome measures were

found between the MD sub-groups, statistical analysis was also conducted between the pooled MD group and the control group.

2.4 Statistical Analysis

All statistical analysis was completed using IBM SPSS Statistics 22 software. The critical level of significance was set at $p < .05$.

Age, stature and body-mass data met the parametric assumptions. For these data, one-way ANOVAs were conducted to test for differences between the three MD subgroups and the control group, and independent t-tests were conducted to test for differences between the pooled MD group and the control group.

6MW data were not normally distributed and all questionnaire data were interpreted as non-parametric. For these data, Kruskall Wallis tests were conducted to test for differences between the three MD subgroups and the control group, with Mann-Whitney U post-hoc pairwise comparisons where appropriate. To test for differences between the pooled MD group and the control group, Mann-Whitney U tests were conducted. For 6MW data, n = 16 MD and 10 control participants to compare between the control and pooled MD group, and n = 6 FSHD, n = 4 BMD, n= 6 LGMD and n = 10 control to compare between the MD subgroups and the control group.

To examine associations, Spearman's Rank correlations were performed between each domain of quality of life and each outcome variable. As no difference was found between the three MD subgroups for the primary outcome variables, the FSHD, BMD and LGMD subgroups were merged into one MD group for correlational analysis. Correlations were interpreted

such that ≤ .40 = small, .41 - .60 = moderate, .61 to .79 = large and ≥ .80 = very large [42]. For 6MW data, n = 16 MD participants for correlational analysis.

3. Results

3.1 Participants

No significant difference in age, stature or body mass was found between the MD subgroups or between any MD group and the control group (Table 1; $p > .05$).

3.2 Quality of Life

QoL scores between the groups are presented in Table 2.

No difference was found among the MD subgroups (FSHD, BMD and LGMD) in any QoL domain ($p > .05$). A main effect of group was found in the following QoL domains: Physical Function ($p \leq .001$), Role Physical ($p \leq .001$), Bodily Pain ($p = .019$), General Health ($p = .026$), Vitality ($p = .005$), Social Function ($p = .002$), Role Emotion ($p = .036$) and Physical Component Score ($p \leq .001$). No difference was found in the Mental Health domain or Mental Component Score ($p > .05$). Post hoc comparisons found that QoL scores in the following domains were lower in each MD subgroup compared to the control group: Physical Function (FSHD,LGMD, BMD: $p \leq .001$), Role Physical (FSHD, LGMD, BMD: $p \leq .001$), General Health (FSHD: $p = .044$, LGMD: $p = .008$, BMD: $p = .027$), Vitality (FSHD: $p = .006$, LGMD: $p = .002$, BMD: $p = .022$), Social Function (FSHD: $p = .005$, LGMD: $p = .002$, BMD: $p \leq .001$), Role Emotion (FSHD: $p = .011$, LGMD: $p = .024$, BMD: $p = .022$) and Physical Component Score (FSHD, LGMD, BMD: $p \leq .001$). In the Bodily Pain domain, scores were lower in the FSHD ($p = .002$) and LGMD ($p = .044$) groups than the control group.

In the pooled MD group, QoL scores were lower than the control group in the Physical Function ($p \leq .001$), Role Physical ($p \leq .001$), Bodily Pain ($p = .002$) General Health ($p = .002$), Vitality ($p \leq .001$), Social Function ($p \leq .001$), Role Emotion ($p = .003$) and Physical Component Score ($p \leq .001$). No difference was found in the Mental Health domain or Mental Component Score ($p > .05$).

3.3 Psychological Parameters and 6MW

Psychological parameters and 6MW distance are presented in Table 3.

No difference in any of these measures were found between the MD subgroups ($p > .05$). A main effect of group was found in physical self-worth ($p = .006$) and 6MW distance ($p = .005$). Depressive symptoms, trait anxiety and self-esteem did not differ between the groups ($p > .05$). Post hoc comparisons found that physical self-worth was lower in the FSHD ($p = .016$), LGMD ($p \leq .001$) and BMD ($p =.026$) groups than the control group. Distance walked during the 6MW test was lower in the BMD ($p =.005$) and LGMD ($p =.009$) groups than the control group.

In the pooled MD group, depressive symptoms were greater ($p = .011$), and physical self-worth ($p \leq .001$) and 6MW distance ($p \leq .001$) were lower than the control group. Trait anxiety and self-esteem did not differ between the groups ($p > .05$).

3.4 Quality of Life Correlations

Correlation coefficients showing the strength of association between QoL scores and psychological parameters/6MW in pooled MD participants are presented in Table 4.

Depressive symptoms were negatively associated with seven of the ten QoL domains. Moderate associations were found between depressive symptoms and Bodily Pain ($r = -.45$, $p = .041$), General Health ($r = -.43$, $p = .049$) and Vitality ($r = -.45$, $p = .043$). Large associations were found between depressive symptoms and Social Function ($r = -.66$, $p \leq .001$), and between depressive symptoms and Role Emotion ($r = -.78$, $p \leq .001$). Very large associations were found between depressive symptoms and Mental Health ($r = -.89$, $p \leq .001$) and the Mental Component Score ($r = -.87$, $p \leq .001$). Physical Function, Role Physical and the Physical Component Score were not associated with depressive symptoms ($p > .05$).

Trait anxiety was negatively correlated with numerous QoL domains. Trait anxiety was moderately correlated with Role Physical ($r = -.44$, $p = .045$) and Vitality ($r = -.48$, $p = .027$). Trait anxiety was largely associated with Social Function ($r = -.74$, $p \leq .001$), Role Emotion ($r = -.85$, $p \leq .001$), Mental Health ($r = -.81$, $p \leq .001$) and the Mental Component Score ($r = -.82$, $p \leq .001$).

Self-esteem was positively associated with various QoL domains. Self-esteem had a moderate association with Social Function ($r = .47$; $p = .033$), and Role Emotion ($r = .68$, $p \leq .001$). Mental Health ($r = .77$, $p \leq .001$) and the Mental Component Score ($r = .74$, $p \leq .001$) were largely associated with self-esteem. No other QoL domains were associated with self-esteem ($p > .05$).

Physical self-worth was positively correlated with QoL in the Role Physical ($r = .49$, $p = .033$), General Health ($r = .50$, $p = .029$), Vitality ($r = .52$, $p = .023$), Mental Health ($r = .58$, $p = .009$) and Mental Component Score ($r = .55$, $p = .014$).

6MW distance was moderately correlated with the Vitality domain (r = .51; p = .043) and highly correlated with Physical Function (r = .65; p = .007) and the Physical Component Score (r = .64; p = .008). No other domains were associated with 6MW distance (p > .05).

4. Discussion

This study presents a comparison of QoL scores, psychological parameters and 6MW distance between adults with MD to matched control adults, and factors that impact QoL in adults with MD are identified. QoL scores and physical self-worth were lower in adults with FSHD, BMD and LGMD than controls, and 6MW distance and depressive symptoms were poorer in the MD group than controls. Psychological parameters and 6MW distance impact QoL, with depressive symptoms showing the greatest and most consistent impact on QoL.

4.1 Quality of Life

QoL scores were poorer in various physical and mental domains for the MD groups compared to the control group, except in the Mental Health domain and the Mental Component Score. This finding agrees with previous research that reported no difference in QoL scores in the Mental Health domain in children and adults with Duchenne MD [13-15], and with a more recent study that found no difference in the Mental Component Score of QoL between adults with Duchenne MD, FSHD, BMD and LGMD compared to control adults [16].

The findings in this study reflect the Disability Paradox [17], as QoL scores in the Mental Health domain was comparable across all groups despite physical disability. This finding had until recently only been reported in Duchenne MD [13-15], which had prompted suggestions that those with Duchenne MD may cope better than those with other types of MD because of earlier onset and limited experience of life without MD [13]. The MD participants in this study

were recruited from a Neuromuscular Centre, where they receive regular physiotherapy and other support. This association may support the development of coping mechanisms, or perhaps it negates some of the factors that impact QoL in the Mental Health domain. Despite comparable QoL scores in the Mental Health domain, QoL scores were poorer than controls in seven other domains for each MD subgroup and eight other domains for the FSHD and LGDM group. Therefore, improving QoL scores in adults with FSHD, BMD and LGMD should be at the forefront of healthcare.

4.2 Psychological Parameters and Physical Function

Differences in psychological parameters and physical function were evident between the MD and control adults. 6MW distance was significantly reduced compared to the control group in both the LGMD and BMD groups, and depressive symptoms were significantly greater in the pooled MD group than the control group. No difference in trait anxiety or self-esteem was evident between the MD groups and the control group, but physical self-worth was considerably reduced in all MD groups compared to the control group. Self-esteem is a multidimensional construct [43] said to be influenced by various domain levels of self-worth such as physical, academic, cognitive and social. The current findings highlight that adults with FSHD, BMD and LGMD exhibit reduced physical self-worth but comparable self-esteem to control participants. The implications of this are twofold: 1) increasing physical self-worth may improve global self-esteem for adults with MD and 2) increasing self-worth in other areas could limit the impact of poor physical self-worth on self-esteem in MD. It is important to note that a large proportion of the MD participants in this study (91%) were in, or had previously been in, paid employment, and sociodemographic factors such as work status may influence self-esteem and other psychological parameters in this population.

4.3 Associations with Quality of Life

QoL scores were consistently associated with psychological parameters including depressive symptoms, trait anxiety, self-esteem and physical self-worth. The size of associations was typically large with mental domains of QoL and moderate with physical domains of QoL. Depressive symptoms showed the most consistent association with QoL (7 of 10 domains), followed by trait anxiety (6 of 10 domains), physical self-worth (5 of 10 domains) and self-esteem (4 of 10 domains). Depressive symptoms also showed the largest association with QoL scores in the Mental Health domain, followed by trait anxiety in the Role Emotion domain. These results agree with findings in patients with other medical conditions such as diabetes [23-26], fibromyalgia [32] and cervical cancer [31]. Previous research has reported a small and a moderate relationship between depression and QoL scores in adults with FSHD [12] and adults with LGMD[18], respectively, but this is the first study to demonstrate associations between QoL and self-esteem, QoL and physical self-worth and QoL and trait anxiety in adults with FSHD, BMD and LGMD.

It is clear that symptoms of depression, trait anxiety, self-esteem and physical self-worth are important determinants of QoL scores in adults with FSHD, BMD and LGMD. Importantly, depressive symptoms, trait anxiety and physical self-worth impact the physical domains of QoL as well as mental domains. Therefore, despite the progressive physical nature of MD the management of psychological health may prove highly beneficial in improving QoL across the board in this population.

Despite reduced physical function being one of the main characteristics of MD, the impact of 6MW distance on QoL scores was limited to three domains. 6MW distance was associated with QoL in Physical Function, Vitality and the Physical Component Score. This finding is

similar to previous research in BMD [16] which found knee extension muscle strength was associated with QoL in the Physical Function and Social Function domains only.

4.4 Clinical Implications

Psychological parameters and 6MW distance impact QoL in adults with MD. Interventions that target either, or preferably both, of these areas could be highly advantageous for improving QoL in this population. Exercise may be a valuable intervention given that it targets both physical and mental health. Future research should seek to establish if exercise can improve psychological and physical parameters in adults with MD and, in turn, improve QoL scores.

4.5 Study Limitations

All MD participants were recruited from a Neuromuscular Centre, where they regularly receive physiotherapy and other support services. This support network may account for the similarities in Mental Health QoL found in this study between the MD and control participants, compared to other studies that reported differences between these groups. However, this finding does highlight the importance of such support networks on QoL in this population. It is also important to acknowledge that a reduced number of MD participants completed the 6MW test because they expressed a fear of falling. As such, there is less confidence in the findings regarding the 6MW test than the psychological parameters in this study, as sixteen participants completed the test compared to twenty-one in the other parameters. Further research with a larger sample of participants would therefore be advantageous to substantiate these findings.

5. Conclusion

In adults with FSHD, BMD and LGMD, QoL scores in mental and physical domains were lower compared to age-matched controls. Symptoms of depression and anxiety have a negative impact on mental and physical domains of QoL in adults with MD. Additionally, physical self-worth, self-esteem and, to a lesser extent, 6MW distance are important determinants of QoL in this population. This study provides a rationale for the assessment and treatment of psychological health alongside physical health for adults with MD. Interventions that target psychological health should be employed, especially where QoL scores are low.

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Declarations of Interest

None

Authors' Contributions

All authors contributed to the design of this paper. DO and EB contributed to the data collection, analysis and the writing of the manuscript. CP, CM and DS supervised the project. PO contributed to participant identification and recruitment. All authors critically revised the manuscript and approved the final version.

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Table 1: Participant characteristics. Maximum voluntary contraction torque is expressed as a percentage of the control group mean value.

	MD	BMD	FSHD	LGMD	CTRL
n	21	7	8	6	10
Age (years)	45.4 ± 10.5	42.1 ± 7.6	46.9 ± 12.6	47.3 ± 11.3	46.8 ± 10.2
Stature (cm)	177.4 ± 8.6	179.9 ± 9.4	180.0 ± 4.9	170.9 ± 9.0	174.4 ± 7.5
Mass (kg)	86.9 ± 17.4	93.0 ± 16.7	85.3 ± 17.3	82.0 ± 19.4	79.4 ± 12.3
KF MVC (%)	37 ± 31	27 ± 15	53 ± 43	25 ± 15	100 ± 28
KE MVC (%)	33 ± 34	11 ± 7	59 ± 42	21 ± 18	100 ± 38
Unemployed (%)	9	14	13	0	0
Employed (%)	67	57	62	83	80
Retired (%)	24	29	25	17	20

MD: muscular dystrophy; BMD: Becker; FSHD: Facioscapulohumeral; LGMD: Limb-girdle; CTRL: control; KF MVC: knee flexion maximum voluntary contraction torque: KE MVC: knee extension maximum voluntary contraction torque.

Table 2: Quality of life scores in each group. Maximum score in each QoL domain is 100.

	MD	BMD	FSHD	LGMD	CTRL
Physical Function	22 ± 17***	18 ± 10***	26 ± 21***	22 ± 17***	98 ± 4
Role Physical	54 ± 27***	46 ± 29***	61 ± 20***	54 ± 35***	99 ± 4
Bodily Pain	58 ± 29**	57 ± 35	52 ± 29**	67 ± 22*	89 ± 10
General Health	53 ± 21**	55 ± 20*	56 ± 25*	48 ± 19**	77 ± 15
Vitality	47 ± 21***	47 ± 27*	51 ± 21**	42 ± 13**	71 ± 8
Social Function	64 ± 28***	57 ± 33***	67 ± 30**	69 ± 21**	99 ± 4
Role Emotion	77 ± 25**	76 ± 28*	73 ± 28*	82 ± 21*	98 ± 3
Mental Health	71 ± 18	74 ± 18	71 ± 23	70 ± 14	84 ± 6
PCS	35 ± 8***	33 ± 10***	37 ± 5***	35 ± 10***	57 ± 3
MCS	52 ± 12	52 ± 14	51 ± 13	52 ± 8	55 ± 3

MD: muscular dystrophy, BMD: Becker, FSHD: Facioscapulohumeral, LGMD: Limb-girdle, CTRL: control, PCS: Physical Component Score, MCS: Mental Component Score. * denotes significant difference from control $p < .05$, ** denotes significant difference from control $p \leq .010$, *** denotes significant difference from control $p \leq .001$.

Table 3: Psychological parameters and 6MW distance in each group. Maximum score for each parameter is indicated in the 1st column.

	MD	BMD	FSHD	LGMD	CTRL
DS /63	10.9 ± 7.9**	11.3 ± 10.5	10.9 ± 7.9	10.3 ± 5.6	4.2 ± 3.0
TA /80	39.2 ± 11.4	39.9 ± 13.1	38.6 ± 11.6	39.2 ± 11.1	32.6 ± 8.4
SE /40	20.9 ± 5.6	21.0 ± 5.4	21.8 ± 6.3	19.5 ± 5.7	24.3 ± 5.2
PSW /24	10.8 ± 3.3***	10.8 ± 3.8*	11.5 ± 3.7*	9.8 ± 2.6***	16.7 ± 3.9
6MWD (m)	333 ± 112***	321 ± 77**	397 ± 102	276 ± 122**	481 ± 72

MD: muscular dystrophy, BMD: Becker, FSHD: Facioscapulohumeral, LGMD Limb-girdle, CTRL: control. DS: depressive symptom score, TA: trait anxiety score, SE: self-esteem score, PSW: physical self-worth score, 6MWD: six-minute walk distance. * denotes significant difference from control $p < .05$, ** denotes significant difference from control $p \leq .010$, *** denotes significant difference from control $p \leq .001$.

Table 4: Associations between QoL and psychological parameters/6MW distance.

	DS	TA	SE	PSW	6MWD
Physical Function	-	-	-	-	.647**
Role Physical	-	-.442*	-	.490*	-
Bodily Pain	-.450*	-	-	-	-
General Health	-.434*	-	-	.500*	-
Vitality	-.445*	-.482*	-	.518*	.511*
Social Function	-.664***	-.744***	.466*	-	-
Role Emotion	-.783***	-.854***	.680***	-	-
Mental Health	-.894***	-.811***	.765***	.584**	-
PCS	-	-	-	-	.640**
MCS	-.874***	-.822***	.743***	.554*	-

DS: depressive symptoms, TA: trait anxiety, SE: self-esteem, PSW: physical self-worth, 6MWD six-minute walk distance. PCS: Physical Component Score, MCS: Mental Component Score. – denotes non-significant relationships, * denotes significant difference $p < .05$, ** denotes significant difference $p \leq .010$, *** denotes significant difference $p \leq .001$.