Published in final edited form as:

Am J Hosp Palliat Care. 2009 ; 26(4): 308–319. doi:10.1177/1049909109335146.

Impact of Biopsychosocial Factors on Chronic Pain in Persons With Myotonic and Facioscapulohumeral Muscular Dystrophy

Jordi Miró, PhD¹, Katherine A. Raichle, PhD^{2,4}, Gregory T. Carter, MD, MS², Sarah A. O'Brien², Richard T. Abresch, MS³, Craig M. McDonald, MD³, and Mark P. Jensen, PhD²

¹ ALGOS. Research on pain, Rovira i Virgili University, Carretera de Valls s/n, Catalonia, Spain

² Department of Rehabilitation Medicine, University of Washington School of Medicine, Seattle, Washington

³ Department of Physical Medicine and Rehabilitation, University of California at Davis, Sacramento, California

Abstract

To assess the role of biopsychosocial factors in patients with type 1 myotonic and facioscapulohumeral muscular dystrophy (MMD1/FSHD) with chronic pain. Associations between psychosocial factors were found to be important in other samples of persons with pain and both psychological functioning and pain interference in a sample of patients suffering from MMD/FSHD. Prospective, multiple group, survey study of 182 patients with confirmed MMD1 and FSHD. Participants completed surveys assessing pain interference and psychological functioning, as well as psychosocial, demographic, and injury-related variables. Analyses indicated that greater catastrophizing was associated with increased pain interference and poorer psychological functioning, pain attitudes were significantly related to both pain interference and psychological functioning, and coping responses were significantly related only to pain interference. In addition, greater perceived social support was associated with better psychological functioning. The results support the use of studying pain in persons with MMD/FSHD from a biopsychosocial perspective, and the importance of identifying psychosocial factors that may play a role in the adjustment to and response to pain secondary to MMD/FSHD.

Keywords

myotonic muscular dystrophy; facioscapulohumeral muscular dystrophy; chronic pain; biopsychosocial model

Introduction

Myotonic muscular dystrophy, type 1 (MMD1), and facioscapulohumeral muscular dystrophy (FSHD) are both hereditary, autosomal dominant, neuromuscular disorders that are slowly progressive and associated with significant disability and chronic pain. Both of these disorders have previously been shown to be associated with chronic pain.^{1–6} Myotonic muscular dystrophy type 1 and FSHD are the second and third most common forms of muscular dystrophy, after Duchenne muscular dystrophy (DMD).^{1,2}

Address correspondence to: Gregory T. Carter, 1800 Cooks Hill Road, suite E, Centralia, WA 98531; phone: (360) 330-8626; gtcarter@u.washington.edu.

⁴KAR is now at the Department of Psychology at Seattle University, Seattle, Washington

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Myotonic muscular dystrophy type 1, the predominant form of myotonic disorders, is caused by a repeated section of DNA on either chromosome 19.^{3,4} Rather than resulting in a missing protein product, this genetic defect results in abnormal RNAs, containing long expanses of CUG or CCUG repeats.³ This creates far-reaching abnormalities in cellular metabolism. The effects of MMD1 repeat expansions affect many different pathways, triggering a complex set of signs and symptoms. Myotonic muscular dystrophy type 1 is a multisystem disorder that affects the visual, cardiac, respiratory, gastrointestinal, and endocrine systems.^{1,4} The brain is also commonly involved and learning disabilities are frequently seen.¹ In skeletal muscle, there is impaired chloride channel conduction across the sarcolemma membrane.⁵ This results in delayed relaxation of muscles after contraction, muscle weakness, cramping, and difficulty with fine motor control.

Facioscapulohumeral muscular dystrophy is due to a genetic defect on chromosome 4 with a loss of a critical number of a repetitive element (D4Z4) in the 4q subtelomeric region.⁷ The loss of the repeats results in specific changes in chromatin structure, although the exact molecular and cellular consequences of this change are not yet known.⁷ The disease is characterized by skeletal muscle weakness and wasting, affecting facial muscles, and specific groups of muscles in the extremities.^{2,7} The heart is not typically involved in FSHD but respiratory failure may be seen.⁸

We have previously described chronic pain as a frequent, prominent symptom in persons with both MMD1 and FSHD.^{6,9} In these disorders, pain can vary in duration, location, and intensity and can greatly interfere with patients' daily activities.^{9–14}

Biopsychosocial models of chronic pain are very useful in understanding adjustment to pain in persons presenting with pain as a primary symptom.^{15–17} Importantly, these models identify factors that affect adjustment and are modifiable with treatment. However, there is a lack of research examining the association between biopsychosocial factors and the experience of chronic pain in persons with MMD1 and FSHD. Knowledge of these associations may be especially important, given the evidence that chronic pain is now known to be a commonly occurring problem in this population.

In this study, we chose to look at the 3 primary biopsychosocial domains known to be important in the setting of chronic pain: (1) attributions (guilt, self-blame, reduced self-efficacy, etc); (2) coping responses to the experience of pain (defined as both behavioral and cognitive efforts to manage specific external and/or internal demands that are appraised as taxing or exceeding the resources of the person); and (3) psychosocial and environmental factors contributing to the pain experience.^{15–18} Comparing results in our sample of MMD1 and FSHD patients to those already available for other pain populations, allowed us to determine which associations are similar across samples, and which might be unique to MMD1 and FSHD patients experiencing chronic pain, hypothesizing that attributions, coping responses, and social factors would all make independent, significant contributions.

Methods

Participants

The University of Washington Human Subjects Review Committee approved all study procedures. Eligibility criteria for this study included confirmed diagnosis, either via DNA testing or clinical parameters (history and physical examination, electrodiagnosis, muscle biopsy, muscle enzymes, etc), being at least 18 years of age, and ability to read and write English. Data from participants with no partner willing to collaborate were excluded from the analysis. All participants provided informed consent.

Study participants were recruited primarily from the NIH-funded National Registry of Myotonic Dystrophy and Facioscapulohumeral Muscular Dystrophy Patients and Family Members (<http://www.urmc.rochester.edu/nihregistry/>). Upon approval of the proposed study by the Scientific Advisory Committee of the Registry, the data manager identified potentially eligible members from the database and wrote them a letter informing the prospective participants about the study. Members of the Registry were instructed to call or email research personnel if they were interested in participating.

A total of 296 potential participants responded to the invitation and were mailed surveys, representing 51% of those enrolled at the NIH-funded National Registry. An additional 99 participants were also recruited directly from the University of Washington Muscular Dystrophy Association (MDA) Clinic. Thus, a total of 395 questionnaires were mailed to potential participants. All study participants who completed the questionnaire received compensation. Research assistants were available to all participants to answer any questions. Participants were contacted to clarify any unclear or omitted answers in returned surveys.

Of the 395 surveys sent, 2 were returned because the participant no longer lived at the address on record, 6 were deceased, and 5 were returned as ineligible. Of the remaining 382 surveys, 298 were returned, yielding a survey return-rate of 78%. Data from 5 participants could not be analyzed and were consequently excluded from further analysis. The current sample includes only participants with MMD1 and FSHD who indicated that they are currently experiencing or had experienced any pain in the past 3 months, other than occasional headaches or menstrual cramps ($n = 182$ of a total possible 257 participants with MMD1 or FSHD).

Among the 182 respondents with pain, 104 were female (57.1%) with a mean age of 48.91 years ($SD = 12.4$, range = 19–83). The majority of these participants were Caucasian (95.6%), married (64.8%), and unemployed (66%). All participants were at least high school graduates or had obtained their general educational development (GED). Of the respondents with pain, 57.1% were diagnosed with FSHD and 42.9% with MMD1. The participants averaged 16.14 years ($SD = 12.0$, range = 0.6–52.2) since their diagnosis. Although 11% of the participants had no mobility limitations, 61% were using some form of assistance for ambulatory circulation.

Measures

Pain intensity—Participants were screened for the presence of pain with the following question: “Are you currently experiencing, or have you in the past 3 months experienced any pain (other than occasional headaches or menstrual cramps)?” Those who indicated no pain were not included in this study. The participants who endorsed having pain, all of whom were included in the current study, were then asked to rate the intensity of their average overall pain in the past week on a 0 to 10 Numerical Rating Scale, with “0” indicating “no pain” and a “10” indicating “pain as bad as it could be.” Numerical rating scales evidence a strong association with other measures of pain intensity and stability over time thus demonstrating their validity and reliability as measures of pain.¹⁹ The mean and standard deviation of the Numerical Rating Scale (NRS) pain intensity measure are presented in Table 1.

Pain interference—The Brief Pain Inventory (BPI) scale was used to assess the degree of pain interference in the past week.^{20,21} The original 7-item BPI scale asks respondents to indicate the extent to which pain interferes with certain activities, such as general activity, mood, walking ability, normal work, relations with other people, sleep, and enjoyment of life. A 0 to 10 scale is used, where a 0 indicates that “pain does not interfere with that activity” and a 10 indicates that “pain completely interferes.” Similar to previous studies, we modified the original scale to adapt the items to consider the unique characteristics of the current study population.^{22,23} First, we changed item 3 (“Walking ability”) to “Mobility (ability to get

around)” to accommodate the respondents who are not ambulatory. In addition, 3 items relating to self-care, recreational activities, and social activities were also added, thus creating a 10-item version of the BPI scale. The addition of these 3 items allowed us to examine a broader range of factors that may be affected by pain. The average interference rating of the 10 items is used in the analyses, with scores ranging from 0 to 10. A higher score indicates a higher degree of pain-related interference. Analogous to the original BPI scale, the modified 10-item version has displayed exceptional internal consistency (Cronbach’s $\alpha = .89-.95$) and validity in previous research examining secondary pain in persons with cerebral palsy,²⁴ limb loss,²² and persons with spinal cord injury (SCI).²⁵ Mean, standard deviation, and Cronbach’s α values of the BPI interference scale are reported in Table 1.

Psychological functioning—Psychological functioning was assessed using the 5-item SF-36 Mental Health scale from the SF-36.²⁶ All survey participants answered the Mental Health items, regardless of presence of pain. This commonly used measure of psychological functioning has demonstrated good reliability, evidenced by high internal consistency coefficients (0.81–0.95) and test-retest stability coefficients (0.75–0.80).²⁶ Its validity as a measure of psychological functioning is supported by its association with other measures of psychological functioning.²⁷ Scores on the SF-36 Mental Health scale range from 0 to 100, with higher scores indicating better psychological functioning. Mean, standard deviation, and Cronbach’s α values of the SF-36 Mental Health scale in our sample are reported in Table 1.

Pain cognitions—The 57-item Survey of Pain Attitudes was used to assess pain-related cognitions (SOPA).¹⁹ The SOPA includes 7 subscales that measure specific types of pain cognitions, including Control (belief in one’s own control over pain), Disability (belief that one cannot function due to pain), Harm (belief that pain is a sign of damage and such activities should be avoided), Emotion (belief that emotions influence pain), Medication (belief that chronic pain may be treatable with medications), Solicitude (belief that others should be solicitous in response to pain behaviors), and Medical Cure (belief that a medical cure may be available for one’s pain). The range of possible answers is 0 (“This is very untrue for me”) to 4 (“This is very true for me”). Subscale scores include the average ratings for each set of subscale items. The SOPA has evidenced good internal consistency (Cronbach’s $\alpha = .71-.81$), test-retest reliability, and validity.^{28,29} Mean, standard deviation, and Cronbach’s α values of the SOPA scales are reported in Table 1.

Catastrophizing—Pain-related catastrophizing (symptom magnification, rumination, and feelings of helplessness) cognitions were assessed using the 6-item Catastrophizing subscale of the Coping Strategies Questionnaire (CSQ).³⁰ Items are scored from 0 (“Never do that”) to 6 (“Always do that”). The scale score is the mean of the 6 items, with higher scores indicating more frequent catastrophizing. The CSQ scale has evidenced excellent internal consistency reliability^{31–35} and the validity of the CSQ is supported by its correlation with depression and adjustment.^{23,36–40} Mean, standard deviation, and Cronbach’s α values of the CSQ Catastrophizing scale are reported in Table 1.

Coping—The 70-item Chronic Pain Coping Inventory (CPCI)^{41,42} was used to assess the frequency with which participants use 9 types of coping strategies for pain, including Guarding, Resting, Asking for Assistance, Relaxation, Task Persistence, Exercise/Strength, Seeking Social Support, Coping Self-Statements, and Pacing. The respondents were asked to indicate the number of days (0–7) during the past week during which they used strategies such as “Imagined a calming or distracting image to help me relax” and “I got support from a friend.” The subscale score is the mean of all of the items included in the subscale, ranging from 0 to 7, with higher scores representing greater use of that specific coping strategy. The validity of these scales is demonstrated by significant correlations with measures of depression and

adjustment to pain as well as significant correlations between patient and significant other versions of the scales.^{41,42} Mean, standard deviation, and Cronbach's α values of the CPCI scales are reported in Table 1.

Perceived support—The 12-item Multidimensional Scale of Perceived Social Support (MSPSS) can assess participants' perceptions of social support from 3 categories of relationships: family, friends, and a significant other. To minimize assessment burden, the current study only used the global perceived social support subscale score. The items are rated on a scale from 1 ("very strongly disagree") to 7 ("very strongly agree"). The subscales and total scale score of the MSPSS have evidenced excellent internal consistencies (Cronbach's $\alpha = .85$ to $.91$), and the scales have demonstrated strong test-retest stability over a 2- to 3-month interval ($r = .72$ to $.85$).⁴³ Validity of the total MSPSS scale has been demonstrated through its significant (negative) association with depression.⁴³ Mean, standard deviation, and Cronbach's α values of the MSPSS scale used in this study are reported in Table 1.

Statistical Analyses

Zero-order correlations, independent samples t tests, and regression analyses were computed and conducted to examine the extent to which demographic characteristics (ie, age and education), pain ratings (eg, pain intensity in previous week), and neuromuscular disorder (NMD)-related variables (eg, type of NMD) were related with the outcome variables of interest, including psychological functioning and pain interference. Education and pain intensity were significantly related with psychological functioning. The correlation analyses revealed that pain in the past week was significantly negatively correlated with psychological functioning ($r = -.28$, $P < .01$). Pain intensity was entered first in subsequent regression analyses to control for this variable. No other variables were significantly related to psychological functioning.

Average pain intensity in the last week was significantly positively correlated with pain interference ($r = .61$, $P < .001$). T tests revealed a significant difference in pain interference scores between male ($M = 2.29$, $SD = .26$) and female participants ($M = 2.45$, $SD = .24$; $t(180) = 2.52$, $P < .05$). No other demographic or pain-related variables were significantly related to pain interference.

The subscale scores of the CPCI and SOPA were subjected to principal components analyses (PCA) as a means of reducing the number of predictor variables in subsequent regression equations. We then performed univariate analysis for descriptive purposes, and to identify the specific psychosocial variables that are most closely linked to patient functioning. Principal components analyses maximizes variance extracted by orthogonal components, thus, was chosen over a number of different possible analyses.⁴⁴ Varimax rotation, an orthogonal technique, was chosen to maximize the variance of the loadings within the components and across variables, thus simplifying and aiding in the interpretability of the underlying components.⁴⁴ We used the scree test and the Kaiser criterion to determine the number of components.⁴⁵

The results of the PCA of the CPCI subscales indicated that the 2 components accounted for 57% of the variance (CPCI scores; eigenvalues = 4.10, 1.04, .94, .74, .63, .51, .41, .37, and .26). Five scale scores loaded on the first component, including Guarding (component loading = .75), Resting (.80), Asking for Assistance (.78), Seeking Social Support (.69), and Pacing (.64). Only 2 scale scores loaded on the second component, including Task Persistence (.73) and Exercise/Strength (.71). Relaxation and Coping Self-Statements were partially loaded onto both components (.41 and .55; .57 and .56). The first component was labeled Passive Coping and the second component was labeled Proactive Coping.

The PCA of the SOPA subscales indicated that the 3 components accounted for 65% of the variance (SOPA scores; eigenvalues = 2.10, 1.45, 1.02, .83, .62, .54, and .45). The subscale scores that loaded on the first component included Control (component loading = $-.77$), Disability (.82), and Harm (.76) scale scores. The second component included mostly the Medication (.76) and Medical Cure (.83) scales. The third component included the Emotion (.84) and Solicitude (.72) scales. The first component was labeled Disability and Harm Beliefs, the second component was labeled Pain as Illness Beliefs, and the third component was labeled Emotion and Solicitude Beliefs.

Included in the regression analyses were the component scores for the CPCI and SOPA, representing coping and pain beliefs, as well as perceived social support. Demographic and clinical variables that were significantly related to the outcome variables were entered first in the regression equations to control for their effects. To help better understand the specific factors that contributed to the significant effects found in the regression analyses, we examined the univariate relationships between specific psychosocial variables, including each subscale of the CPCI and SOPA scales, and the outcome variables of interest (pain interference and psychological functioning) using correlation analyses. Because of the large number of correlations performed (19 psychosocial predictor variables per criterion measure), we used a Bonferroni correction for each criterion measure ($.05/19 = .0026$) in the univariate analyses to determine whether each association was significantly different from zero.

Results

Associations Between Psychosocial Variables and Psychological Functioning

Pain intensity explained 9% of the variance in the criterion ($P < .01$). After controlling for pain intensity, the group of psychosocial variables accounted for an additional, significant ($P < .001$) 34% of the variance in psychological functioning. On whole (as a block), the group of psychosocial predictors was statistically significant ($P < .01$). Greater psychological functioning was significantly associated with increased MSPSS social support ($\beta = .40$, $P < .01$), lower scores on the Emotion and Solicitude beliefs component of the SOPA ($\beta = -.23$, $P < .01$), and lower catastrophizing ($\beta = -.15$, $P < .05$; see Table 2).

Obtaining component scores for the coping strategies and pain cognition variables was needed to be able to reduce the amount of variables to be included in the regression analyses. However, when using component scores as predictors, it is not possible to determine the univariate relationships between the psychosocial predictors and the criterion variables (psychological functioning and pain interference). Zero-order correlations were therefore computed between the psychosocial variables and the criterion for descriptive and explanatory purposes, using a Bonferroni correction for each criterion to control for α inflation (ie, 19 predictors per criterion; so $.05/19 = .0026$). No CPCI subscale emerged as significantly related to psychological functioning with the Bonferroni correction ($P < .0026$). However, several subscales evidenced significant trends. For example, lower scores on both Resting ($r = -.19$, $P = .009$) and Relaxation ($r = -.15$, $P = .047$) were associated with greater psychological functioning, whereas greater Task Persistence ($r = .15$, $P = .049$) was associated with greater psychological functioning. Numerous SOPA subscales were significantly related to psychological functioning. For example, higher scores on the SOPA subscales of Disability ($r = -.27$), Harm ($r = -.23$), Emotion ($r = -.29$), and Solicitude ($r = -.29$) were related with lower psychological functioning, while higher Control ($r = .31$) was related with better psychological functioning (all P s $< .0026$). Consistent with the multivariate analyses, both Catastrophizing ($r = -.43$) and MPSS social support ($r = .55$) were significantly associated with psychological functioning, with higher levels of catastrophizing associated with lower levels of psychological functioning and higher levels of MSPSS social support associated with higher levels of psychological functioning (both P s $< .0026$; see Table 3).

Associations Between Psychosocial Variables and Pain Interference

In the regression analysis predicting pain interference, pain intensity and gender explained 38% of the variance ($P < .001$) in pain interference, with pain intensity contributing the most to this prediction ($\beta = .60, P < .001$). The psychosocial variables as a whole accounted for an additional 19% of the variance ($P < .001$) in pain interference scores, after controlling for pain intensity and gender. Higher scores on the catastrophizing scale ($\beta = .25, P < .001$), the Disability and Harm beliefs subscale of the SOPA ($\beta = .17, P < .05$), and the Passive coping component of the CPCI ($\beta = .18, P < .001$) predicted greater pain interference, whereas lower scores on the MSPSS social support scale were related to higher levels of pain interference ($\beta = -.17, P < .01$; see Table 4).

The zero-order correlations identified a large number of significant associations between the psychosocial variables and pain interference. Of the nine CPCI subscales, 7 were significantly (and positively) correlated with pain interference: Guarding, Resting, Asking for Assistance, Coping Self-Statements, Relaxation, Seek Social Support, and Pacing ($r_s = .40, .39, .31, .30, .28, .25$, and $.23$, respectively, all $P_s < .0026$). Of the SOPA subscales, Disability, Harm, and Solicitude were all significantly positively correlated with pain interference ($r_s = .51, .46, .24$, respectively, $P_s < .0026$), while Control was significantly negatively correlated with pain interference ($r = -.32$). Consistent with the multivariate analyses, catastrophizing was positively and strongly associated with pain interference ($r = .58, P < .0026$), while MSPSS social support was negatively correlated with pain interference ($r = -.35, P < .0026$; see Table 4).

Discussion

The findings of this study support the use of a biopsychosocial model for understanding chronic pain and its impact in persons with MMD1 and FSHD. The hypothesis that attributions, coping responses, and psychosocial factors are all independent, significant contributions to chronic pain in this population is supported. The data also indicate that measures of psychological functioning and pain interference are associated with psychosocial variables in this patient population. Moreover, with the exception of positive associations between most of the coping responses (whether considered adaptive or maladaptive) and pain interference, the current study shows positive associations between patient functioning and psychosocial factors thought to be maladaptive (eg, guarding and resting coping responses, disability and harm beliefs), as well as negative associations between patient functioning and factors thought to be adaptive (e.g., perceived support and control beliefs). In particular, and in both the multivariate and univariate analyses, we found that greater pain-related catastrophizing (symptom magnification, rumination, and helplessness) was significantly related with both poorer psychological functioning and increased pain interference. This finding is consistent with many prior studies in persons with different pain conditions and disabilities, and across different age groups. Specifically, pain-related catastrophizing has been associated with poorer outcomes in children,⁴⁶ adults,^{16,27,47} and elderly patients with pain⁴⁸ as well as persons with chronic pain secondary to a number of disabilities such as multiple sclerosis,⁴⁹ cerebral palsy,^{50–52} spinal cord injury,^{17,18,23,53} and phantom limb pain.^{22,36}

To our knowledge, this is the first time that pain-related catastrophizing has been studied in patients with MMD1 and FSHD and pain. Given the correlational nature of the current study, it is not possible to draw conclusions regarding the causal nature of this relationship. Catastrophizing cognitions may influence and lead to greater dysfunction, dysfunction may influence and lead to greater catastrophizing, dysfunction and catastrophizing may influence each other, or both may reflect or be caused by a third variable (eg, a generally negative or pessimistic attitude). However, there is some evidence that decreases in catastrophizing are associated with improved outcomes for persons receiving multidisciplinary treatment for

chronic pain.²⁷ Moreover, an intervention that specifically targets catastrophizing has been shown to decrease reports of pain intensity in persons with disabilities.⁵⁴ Further studies are needed to determine the generalizability of our findings to other samples of persons with MMD1 or FSHD, and to confirm that changes in catastrophizing actually contribute to improved functioning, as opposed to merely reflect improved functioning, in persons with disabilities and chronic pain.

Our results showed that higher levels of perceived social support were associated with less pain interference and better psychological functioning, even after controlling for pain intensity and demographic variables. Again, it is not possible to determine the causal direction of the relationships found using correlational data. However, these findings are generally consistent with other research of persons with disabilities and chronic pain^{17,22,36,49} as well as with studies of persons with pain and other various health problems.^{16,53,55,56} The consistent associations found in this body of research raises the possibility that social support, or at least one's perceived access to support, may act as a buffer for maintaining physical health, and psychological functioning, in persons with disabilities and pain. Longitudinal and experimental research is needed to test and confirm this causal hypothesis.

The multivariate analyses revealed that the pain belief factor scores made statistically significant and unique contributions to the prediction of the criterion measures. Interestingly, while the Emotion and Solicitude beliefs factor was associated most closely to psychological functioning, the Disability and Harms Beliefs factor was related most closely to pain interference. Univariate analysis revealed that certain pain beliefs were significantly related to both psychological functioning and pain interference (ie, Disability, Harm, Solicitude, Control). These results are consistent with previous studies finding similar associations between pain beliefs related to disability and pain interference.^{17,49,52,57}

Moreover, the findings showed significant contributions of coping to the prediction of the criterion measures. However, only the Passive Coping factor showed statistically significant relationships with pain interference in the multivariate analyses. The univariate analyses, 7 of the 9 CPCI coping scales showed significant associations with pain interference (all associations were positive), with the strongest negative associations occurring among those coping responses considered to be maladaptive (eg, guarding, resting, and asking for assistance). These findings are consistent with research in other disability groups where pain coping has been found to be associated with pain and functioning. For example, guarding and resting have been found to be associated with increased pain interference and pain intensity in patients with phantom limb pain,³⁶ multiple sclerosis,⁴⁹ and SPI.¹⁷

The positive univariate associations found between 4 coping responses generally thought to be adaptive and pain interference helps to further illustrate the difficulty of identifying causal associations from correlational data. Such results could be obtained if these coping responses are, in fact, maladaptive in our sample. For example, it is possible that the coping strategies thought to be adaptive (eg, coping self-statements, seeking social support, and pacing) may be helpful at the outset of a disability or injury, but once maximum gains have been achieved, some of these strategies may become less effective or even hinder progress toward more independent functioning. However, it is possible that positive associations found between coping responses thought to be adaptive and pain interference occurred because people find them useful, and so they tend to be used more when patients suffer from higher levels of pain or pain interference; in the same way that one might expect a positive association between pain intensity and the use of effective analgesic medications. Thus, the current findings indicate that (1) pain coping responses predict and may be important to adjustment to pain in persons with MMD1 and FSHD and (2) research is needed to further explore the relationship of these variables with pain interference in this population, in particular, to help elucidate the presence

and extent of any causal associations. Such research would help guide clinicians in the development of effective interventions for persons with MMD1 and FSHD suffering from chronic pain. Future studies should also implement longitudinal designs to better understand these relationships and experimental designs that can help to elucidate causal relationships between all these factors.⁵⁴

Several methodological limitations of the current study should be acknowledged. A primary limitation, already discussed, is that the data are cross-sectional. Because of this, we cannot draw causal conclusions from the study; longitudinal and experimental research is needed to better understand the nature of the relationships between the variables, and to determine whether changes in specific psychosocial variables are associated with changes in pain-related criterion measures. Also, the use of component scores versus individual subscales of the SOPA and CPCI in the multivariate analyses represents a limitation. Although use of this strategy was necessary to ensure an adequate N to k ratio in the multivariate analyses, this approach limits the interpretability of the multivariate findings and increases the risk of type II error.^{17,27,58} A third limitation is that the sample was drawn primarily from persons with MMD1 and FSHD who were registered with a NIH-funded National Registry. This limits the generalizability of the findings. We are unable to determine whether and to what extent our findings generalize to other persons with MMD1 or FSHD not participating in the registry. Moreover, we do not know to what extent the participants who agreed to participate in this study are similar to or different from other patients who are listed in the NIH registry but did not choose to participate in the study. In addition, a large majority of the participants in this study were Caucasians. Thus, future studies should seek to replicate these findings in broader and more representative samples of persons with MMD1 and FSHD. Finally, the current findings may be affected by responder bias (eg, social desirability effects or memory failure), as the data are self-report. Moreover, using only self-report data increases the possibility that some of the significant associations observed could be due in part to shared method variance. Future studies should also include objective measures, and/or measures from other sources (eg, spouses/partners, health care providers) when possible.

Despite the limitations of the current study, the findings have important research and clinical implications; specifically regarding how chronic pain in individuals with these neuromuscular disorders should be understood and treated. Currently, chronic pain management in these populations is guided primarily by biomedical approaches, with research-limited success.^{59–63} Our study provides novel, empirical support for broadening the treatment approach to include biopsychosocial variables as potential treatment targets. Our data also point to a need for further research to determine if biopsychosocial interventions are as effective in these populations as they have been for other groups of individuals, both disabled and able-bodied, with chronic pain.

Acknowledgments

This research was supported by the National Institutes of Health, National Institute of Child Health and Human Development, National Center for Rehabilitation Research (grant no. P01HD33988), the National Registry of Myotonic Dystrophy and Facioscapulohumeral Muscular Dystrophy Patients and Family Members, and the National Institute for Disability Rehabilitation Research (grant no. H133B031118).

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Table 1

Descriptive Statistics of Study Measures

	Mean (SD)	Cronbach's α
0–10 NRS	4.50 (2.60)	NA
BPI Interference scale	2.96 (2.42)	.95
SF-36 Mental Health scale	66.92 (19.02)	.84
SOPA		
Control scale	1.89 (0.74)	.76
Disability scale	2.16 (0.76)	.73
Harm scale	2.21 (0.69)	.69
Emotion scale	1.54 (0.76)	.72
Medication scale	2.54 (0.90)	.76
Solicitude scale	1.37 (0.72)	.71
Medical Cure scale	1.69 (0.53)	.74
CSQ Catastrophizing scale	1.18 (1.24)	.89
CPCI		
Guarding scale	3.38 (1.83)	.82
Resting scale	3.25 (1.88)	.82
Asking for Assistance scale	3.20 (2.37)	.90
Relaxation scale	1.56 (1.12)	.66
Task Persistence scale	4.22 (2.00)	.86
Exercise/Strength scale	1.61 (1.74)	.92
Coping Self-Statements scale	2.55 (1.82)	.89
Seek Social Support scale	1.65 (1.60)	.84
Pacing scale	3.13 (2.07)	.85
MSPSS scale	5.30 (1.27)	.93

Abbreviations: 0–10 NRS, 0 to 10 Numerical Rating Scale of pain intensity; BPI, Modified Brief Pain Inventory Pain Interference scale; CPCI, Chronic Pain Coping Inventory; CSQ, Coping Strategies Questionnaire; MSPSS, Multidimensional Scale of Perceived Social Support; SF-36 MHS, Mental Health Scale; SOPA, Survey of Pain Attitudes.

Multiple Regression Analyses Predicting Psychological Functioning From Cognitions, Coping, and Perceived Social Support (n = 181)^a

Table 2

Step and Variables	Total R ²	R ² Change	F Change	β
1. Demographic and Clinical Variables	.09	.09	17.41 ^b	
Pain Intensity				-.30 ^b
2. Cognitions, Coping, and Social Support	.42	.34	14.31 ^b	
Catastrophizing				-.15 ^c
MSPSS Social Support				.40 ^b
Disability and Harm Beliefs (SOPA Factor 1)				-.08
Pain as Illness Beliefs (SOPA Factor 2)				.01
Emotion and Solitude Beliefs (SOPA Factor 3)				-.23 ^b
Passive Coping (CPCI Factor 1)				.01
Proactive Coping (CPCI Factor 2)				.03

Abbreviations: CPCI, Chronic Pain Coping Inventory; MSPSS, Multidimensional Scale of Perceived Social Support; SOPA, Survey of Pain Attitudes.

^aTotal n < 182 due to missing data.

^bP < .01.

^cP < .05.

Table 3

Zero-Order Correlation Coefficients between Subscale Scores of the Psychosocial Variables With Psychological Functioning and Brief Pain Inventory Scores^a

Belief/Coping Scores	Outcome Measures	
	Brief Pain Inventory	SF-36 Mental Health
CPCI subscales		
Guarding	.40 ^b	-.13
Resting	.39 ^b	-.19 ^c
Asking for Assistance	.31 ^b	-.04
Relaxation	.28 ^b	-.15 ^c
Coping Self-Statements	.30 ^b	-.05
Seek Social Support	.25 ^b	.08
Task Persistence	-.11	.15 ^c
Exercise/Strength	.05	.01
Pacing	.23 ^b	-.05
SOPA subscales		
Disability	.51 ^b	-.27 ^b
Harm	.46 ^b	-.23 ^b
Medication	.13	-.03
Medical Cure	.10	-.07
Emotion	.03	-.29 ^b
Solicitude	.24 ^b	-.29 ^b
Control	-.32 ^b	.31 ^b
CSQ Catastrophizing	.58 ^b	-.43 ^b
MSPSS	-.35 ^b	.55 ^b

Abbreviations: CPCI, Chronic Pain Coping Inventory; CSQ, Coping Strategies Questionnaire; SOPA, Survey of Pain Attitudes; MSPSS, Multi-Dimensional Scale of Perceived Social Support.

^aThese correlations and significant levels are presented for descriptive purposes. Given the large number of correlations performed on related variables, (18 psychosocial predictor variables per criterion measure), we used a Bonferroni correction for each criterion measure (.05/19 = .0026) to determine whether each association was significantly different from zero.

^b $P < .0026$, 2-tailed.

^cNonsignificant trend.

Multiple Regression Analyses Predicting Pain Interference from Cognitions, Coping, and Perceived Social Support (n = 181)^a

Table 4

Step and Variables	Total R ²	R ² Change	F Change	β
1. Demographic and Clinical Variables	.38	.38	54.59 ^b	
Pain Intensity				.60 ^b
Gender				-.07
2. Cognitions, Coping, and Social Support	.57	.19	10.84 ^b	
Catastrophizing				.25 ^b
MSPSS Social Support				-.17 ^b
Disability and Harm Beliefs (SOPA Factor 1)				.17 ^c
Pain as Illness Beliefs (SOPA Factor 2)				.01
Emotion and Solitude Beliefs (SOPA Factor 3)				.02
Passive Coping (CPCI Factor 1)				.18 ^b
Proactive Coping (CPCI Factor 2)				.01

Abbreviations: CPCI, Chronic Pain Coping Inventory; MSPSS, Multidimensional Scale of Perceived Social Support; SOPA, Survey of Pain Attitudes.

^aTotal n < 182 due to missing data.

^bP < .01.

^cP < .05.