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Pain Location and Intensity Impacts Function in Persons with Myotonic Dystrophy Type 1 and Facioscapulohumeral Dystrophy with Chronic Pain

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Abstract

Introduction—We examined the effects of pain site and intensity on function in patients with myotonic dystrophy type 1 (DM1) and facioscapulohumeral muscular dystrophy (FSHD) and chronic pain.

Methods—Questionnaires assessing pain sites, pain extent (number of sites), pain intensity, and pain interference were completed by 182 individuals with DM1 (43%) or FSHD (57%) and chronic pain.

Results—There was a positive association between pain extent and intensity with pain interference, and a negative association with psychological functioning in both DM1 and FSHD. Pain intensity at specific sites had differential impact beyond the effects of pain intensity alone. Head pain intensity independently affected psychological functioning, whereas leg, foot, hip, and knee pain contributed independently to the prediction of pain interference.

Conclusions—Pain site and intensity differentially modulates the effect of chronic pain on function in DM1 and FSHD patients. Researchers and clinicians should consider these factors when assessing and treating pain.

Keywords

facioscapulohumeral muscular dystrophy; myotonic dystrophy type 1; pain assessment; pain management; rehabilitation

Chronic pain is a significant problem for many patients with chronic neuromuscular diseases (NMDs), including as many as 70–90% of those with myotonic dystrophy type 1 (DM1) and facioscapulohumeral muscular dystrophy (FSHD).[1–12] Studies have shown that as many as 70% and 90% of patients with DM1 and FSHD, respectively, report pain, which negatively impacts quality of life (QoL) and increases disease burden.[7–26] Yet, very little is known

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about the nature of pain in DM1 and FSHD (i.e., how bad and where it hurts) and how pain contributes to overall function.

A recent study in this patient population confirmed that, as expected, QoL was reduced significantly by many physical domains, including pain.[22] The available literature suggests that disease severity is the best determinant for physical domains of QoL, with mood and illness perception being the main determinants for the psychological domains of QoL.[23–26] However, the interplay between the physical and psychological domains of QoL in patients with neuromuscular disease is complex.

Adjustment to chronic pain appears to depend as much on pain site as on intensity. However, there are no studies of the relative importance of pain extent (number of body areas with pain) and site to patient functioning in individuals with any NMD, including DM1 and FSHD.

This study was designed to test our hypotheses that: (1) pain extent would be associated negatively with psychological functioning and associated positively with pain interference; and (2) pain intensity in critical physical areas (i.e., low back, arms) would evidence stronger associations with pain interference and psychological functioning than pain at other sites. For the purposes of this study, chronic pain was defined as the reported experience of: (1) any pain in the past 3 months; and (2) persistent, bothersome pain at 1 or more sites of the body at the time of survey completion.

Methods

Participants

All protocols were approved by the human subjects committee and medical ethics review board of the University of Washington prior to initiation of the study. Participants were recruited primarily from the National Institutes of Health (NIH)-funded Registry of Myotonic Dystrophy and Facioscapulohumeral Muscular Dystrophy Patients and Family Members (http://www.urmc.rochester.edu/nihregistry/).[27] Informed consent was obtained from all subjects prior to participation. Registry patients must have medical records containing documentation of diagnosis confirmation before joining. A small number of patients recruited locally had their diagnosis confirmed by one of the investigators (G.T.C.). The Registry consists of de-identified information collected at baseline and annually thereafter. The Registry protocol is as follows: individuals who have been diagnosed with FSHD or DM1 by a neuromuscular specialist contact the Registry and provide the Registry with demographic information and permission to access their medical records. The Registry then abstracts and de-identifies the information in the medical records and assists with subject recruitment. Inclusion criteria for this study were: (1) primary diagnosis of DM1 or FSHD; (2) age ≥18 years; (3) ability to read and write English; and (4) cognitive ability to comprehend the questionnaires.

Upon approval of the proposed study by the scientific advisory committee of the Registry, the data manager extracted potentially eligible members from the database and wrote them a letter about the study. Members of the Registry were invited to call or research personnel if

they were interested in participating (http://www.urmc.rochester.edu/nihregistry/). The remaining participants were recruited from the Neuromuscular Disease Clinic of the University of Washington or affiliated regional centers. A total of 395 surveys were mailed to potential participants, and 12 were returned (4 due to ineligibility, 6 due to being deceased, and 2 due to wrong address). Of the 383 surveys sent out to appropriate respondents, 298 were returned, for an overall response rate of 78%. Participants were compensated $25 for completing the survey, which took approximately 1 hour to complete. Of the 298 surveys returned, 182 were from participants who: (1) had a specific diagnosis of DM1 or FSHD; (2) indicated they were experiencing or had experienced any pain in the past 3 months (other than occasional headaches or menstrual cramps); (3) indicated they were experiencing persistent, bothersome pain at 1 or more sites at the time of survey completion; and (4) had valid data. These 182 participants comprised the study sample.

Measures

Demographic Information—The following demographic information was collected from study participants: age; gender; educational level; employment and marital status; and racial group. NMD-related information was also assessed and included: type of NMD diagnosis and the date at which the person received the diagnosis of DM1 or FSHD.

Pain Intensity—A 0–10 numerical rating scale was used to gather information on the participant's average global pain intensity in the past week. A “0” indicated “no pain” and a “10” indicated “pain as bad as it could be.” A great deal of evidence supports the reliability and validity of numerical rating scales as measures of pain intensity.[28]

Pain Interference—A modified version of the Brief Pain Inventory (BPI) pain interference scale was used to assess the degree of pain interference in the past week.[29,30] With the BPI, respondents rate the extent to which pain interferes with 7 different activities of daily living (general activity, mood, walking ability, normal work, relations with other people, sleep, and enjoyment of life) on a 0–10 scale, where a 0 indicates that “pain does not interfere with that activity” and a 10 indicates that “pain completely interferes” with the activity. As in previous studies, we modified the original scale to adapt the items to consider the unique characteristics of the study population.[29,30] Specifically, to accommodate the respondents who are not ambulatory we changed item 3 (“walking ability”) to “mobility (ability to get around).” In addition, 3 items related to self-care, recreational activities, and social activities were 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. Analogous to the original BPI scale, the modified 10-item version has shown high internal consistency values (Cronbach alpha = 0.89–0.95) and validity properties in previous studies of pain interference in persons with cerebral palsy, limb loss, and spinal cord injury.[31–33] In this sample, the internal consistency of the modified pain interference scale was 0.91.

Pain Site and Extent—Participants reported the location(s) where they experienced pain and the average pain intensity experienced in the past week in that particular location.

Specifically, they were asked to indicate if they experienced any persistent, bothersome pain in any of 17 different body locations or sites (as well as any "other" location not covered by these 17, so there were 18 location categories total): head; neck; shoulder; upper back; lower back; arms; elbows; wrists; hands; buttocks; hips; chest; abdomen/pelvis; legs; knees; ankles; feet; and other. A pain extent score for each participant was created by summing the total number of pain sites (possible range 0–18).

Psychological Functioning—The 5-item Mental Health Scale of the 36-item Short Form Health Survey (SF-36) was used to assess psychological functioning.[31,32] This measure has shown good reliability (as shown by internal consistency and test–retest stability analysis).[34,35] The association of the SF-36 with other measures of psychological functioning health supports its validity as a measure of psychological functioning.[34,35] Scores on the SF-36 psychological functioning scale range from 0 to 100, with higher scores indicating better psychological functioning.

Statistical Analysis

We first computed means and standard deviations of the study variables for descriptive purposes. Next, we used regression analyses to determine whether differences existed between the 2 NMD diagnoses in how pain extent or pain site was related to the criterion variables. We entered global pain intensity in step 1 (as a control variable), diagnosis (DM1 vs. FSHD, dummy coded) in step 2, pain extent and intensity at each site in step 3, and diagnosis × pain extent and diagnosis × pain site interaction terms in step 4. We examined the significance levels of diagnosis main effect and the interaction terms to determine whether diagnosis was associated with either criterion variable or had a moderating influence on the associations between the predictors and criterion. In the event of significant effects related to diagnosis, which would indicate that the 2 groups differed with respect to levels of pain interference or psychological functioning (indicated by a significant main effect for diagnosis) or the associations between the predictors and criterion variables (indicated by a significant interaction), we planned to examine these associations separately for each group. In the event of no significant interactions, we planned to collapse across the 2 diagnoses for the study analyses. We then planned to compute zero-order correlation coefficients between average global pain intensity in the past week, the pain extent score, and average pain intensity in the past week at each pain site, and the study criterion measures of pain interference and psychological functioning to test the hypothesized associations between these variables. For the correlational analyses involving pain site, we used 2 samples: (1) only the individuals who reported that they experienced at least *some* pain (i.e., 1 or more on the 0–10 scale) at the site in question in the past week (ranging from 19 participants who reported at least some pain in the chest to 128 participants who reported at least some pain in the low back); and (2) *all* of the study participants, with the pain intensity at each site coded as "0" for those who did not report any pain at that site. Finally, we used regression analyses to determine whether pain extent and pain intensity at specific sites explained unique variance in the criterion variables. The criterion variables in these analyses were pain interference and psychological functioning. Global pain intensity was entered in step 1 (as a control variable), followed by stepwise entry of pain extent and pain intensity at each pain site.

Results

Sample and Study Variable Description

The majority of study sample patients were white (96%), married (65%), and had at least a high school education (79%). Fifty-seven percent were women. Forty-three percent had a diagnosis of DM1, and 57% had a diagnosis of FSHD. The average age of participants when they completed the survey was 48.9 years (SD = 12.4, range = 19–83 years). Time since NMD diagnosis was 16.1 years (SD = 12.0 years, range 0.6–52.2 years). Additional descriptive information about the study sample and the means and standard deviations of the key study variables are listed in Table 1.

Main and Moderating Effects of Diagnostic Group

In the first set of regression analyses to determine whether diagnosis had a main or moderating effect on the associations between the predictors and criterion variables, neither the main effects nor interaction terms involving diagnosis were statistically significant. *F*-change (1.179) values for the main effects for diagnosis predicting pain interference and psychological functioning were 0.90 and 1.34, respectively [*P* = not statistically significant (NS)]. *F*-change (19.142) values for the predictors as a block were 0.76 and 0.65 (*P* = NS), and the *t*-values for the interaction terms ranged from 0.06 to 1.82 (predicting pain interference; all *P*s = NS) and 0.02 to 1.24 (predicting psychological functioning; all *P*s = NS). Thus, diagnostic group had no significant influence on the criterion variables or their associations with the predictors. Therefore, the data were collapsed across the diagnostic groups for all subsequent analyses.

Associations between Pain Extent and Pain Site, and Pain Interference and Psychological Functioning

The zero-order correlations between pain extent and pain intensity at each of 18 pain site categories and the study criterion variables are listed in Table S1 (refer to Supplementary Material available online). Pain extent was positively and statistically significantly correlated with pain interference and negatively associated with psychological functioning. An examination of the coefficients associated with the measures of pain intensity at each site showed that most were in the moderate (0.30–0.50) or strong (>0.50) range. Stronger associations between pain intensity at specific sites and the criterion variables tended to be found in the samples of participants who reported at least some pain at the site in question, although significant associations emerged for both samples. Low back and arm pain did not demonstrate the strongest associations with the criterion variables. Rather, the strongest associations ($r \geq 0.50$) were found for the associations between pain intensity in the wrist, hand, buttock, hip, leg, and knee (for those reporting at least some pain at these sites) and pain interference, and for the association between leg pain and pain interference for the sample as a whole.

Independent Effects of Pain Extent and Pain Site

In the regression analysis predicting psychological functioning, overall pain intensity explained 9% of the variance. After controlling for overall pain intensity, higher scores for pain intensity of the head predicted an additional 2% of the variance ($P < 0.05$).

In the regression analysis predicting pain interference, overall pain intensity explained 39% of the variance (Table 2). Specific pain sites accounted for an additional 11% of variance in the criterion. Those sites that contributed significantly to the prediction of pain interference were the legs, feet, hips, and knees. Pain extent did not emerge as a significant unique predictor of either criterion variable.

Discussion

These data document the importance of body site when one assesses pain and its impact in patients with DM1 or FSHD and chronic pain. Pain interference and psychological functioning also appear to be associated with pain intensity at different sites. However the pain sites that matter most to patients with DM1 or FSHD and chronic pain differ from those with other chronic pain conditions.

Prior studies in patients with musculoskeletal problems support the idea that pain extent is important to consider when evaluating a patient with chronic pain.[36] Our study further supports this concept. Overlooking any given pain site and the pain intensity associated with that site may result in a failure to capture factors that are important in an individual's adjustment to pain and ultimately in finding a successful treatment strategy.

The sites related to functioning in our DM1 and FSHD subjects are those related to ambulation (i.e., legs, feet, hips, and knees). This makes intuitive sense given that the muscles in these areas are particularly taxed in physical terms and thus would be susceptible to contraction-induced injury.[37–39] This also supports the idea that dystrophic muscle, as an end-organ, behaves similarly with regard to clinical symptoms (i.e., pain), despite a differing genotypic etiology (DM1 vs. FSHD). Thus, a potentially important area for study may be in the development specific strategies or activities that could improve strength, flexibility, and endurance of those muscles and related areas. The data indicate that experiencing pain in an “other” location (other than the 17 specific locations listed) also contributes to the variance of pain interference. A small subgroup of participants ($N = 19$) in this study reported pain in locations other than those indicated in the survey. This suggests that there are other locations to consider beyond the 17 pain site locations assessed.

There are noteworthy limitations to our study, given that all of the data were self-reported. Thus, some of the significant associations among measures could potentially be related to shared method variance. Future studies could use more objective measures of patient functioning, such as ratings made by spouses or significant others, or objective measures of activity (e.g., actigraphy). We also computed a large number (68) of correlation coefficients without controlling for alpha inflation. In addition, although we used a stepwise selection procedure to identify the significant predictors in the regression analyses (as a way to limit the number of predictors that were in the final equation), a large number of potential

predictors (19; 18 pain site categories and 1 measure of pain extent) were considered in addition to the measure of pain intensity. Therefore, it is possible that some of the significant associations found were due to chance alone (e.g., type I error). However, if there was no association between the study predictors and criterion variables, then only 2 (5%) of the statistical tests would have been significant by chance alone. The number of significant associations found (20% or 55%) far exceeds this, supporting the importance of pain site as contributing to the prediction of patient functioning over and above global pain intensity alone. In addition, our sample was comprised entirely of patients with DM1 and FSHD who agreed to participate in the survey. We do not know how representative these individuals were of the sample of patients involved in the Registry used for recruitment, or even from the population of individuals with these conditions. Moreover, even if the study participants were representative of individuals with DM1 and FSHD, the findings do not necessarily generalize to individuals with other forms of NMD. Thus, replication of the findings in additional samples of individuals with DM1 and FHSD and in individuals with other forms of NMD is needed to help establish their reliability. Larger, prospective studies may also help elucidate whether these preliminary results are representative of the population. International multicenter studies may be instrumental in this endeavor.

The similarity in pain symptom profiles between DM1 and FHSD subjects with chronic pain suggests that dystrophic muscle in slowly progressive NMD produces similar clinical symptomatology with regard to disease burden and pain, as documented previously.[40–42] Conversely, it could also reflect an insensitivity in the questionnaires, although this is not likely given their extensiveness. Finally, our data may only reflect, and be applicable to, United States and North American populations. Earlier studies on QoL have shown differences in response patterns of subjects from the USA with regard to certain aspects of QoL compared with subjects from the UK.[41] This may have implications if our data were used in the design of multinational clinical trials.

Despite the study limitations, the findings provide evidence that supports the value of assessing specific pain qualities in addition to overall pain intensity measures in persons with DM1 and FSHD and chronic pain. Our data also document the importance of pain location as a pain domain factor contributing to function in these patients. Thus, despite the fact that pain intensity is an important pain domain contributing to patient dysfunction, pain location is a domain that appears to play a significant role in quality of life in this setting. These aspects of pain should be incorporated in the design of future rehabilitation paradigms for DM and FSHD.[43]

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Abbreviations

BPI	Brief Pain Inventory
DM1	muscular dystrophy, myotonic type 1
FSHD	facioscapulohumeral dystrophy
NMD	neuromuscular disease
NRS	Numerical Rating Scale
QoL	quality of life
SF-36	36-item Short Form Health Survey

Table 1

Study measures (N = 182).

	Mean (SD)		Mean (SD)
Global average pain intensity (0–10 NRS, 0–10)	4.50 (2.60)	Pain interference (BPI, 0–10)	3.22 (2.53)
Psychological functioning (MHI-5, 0–100)	66.92 (19.02)	Pain extent (0–18)	6.43 (3.75)
Average pain intensity at each site (0–10) *			
Head	0.74 (2.09)	Neck	2.23 (2.80)
Shoulder	2.78 (2.85)	Upper back	1.68 (2.63)
Lower back	3.87 (3.25)	Arms	1.80 (2.76)
Elbows	0.71 (1.85)	Wrists	1.11 (2.32)
Hands	1.76 (2.77)	Buttocks	0.91 (2.13)
Hips	2.18 (2.79)	Chest	0.51 (1.65)
Abdomen/pelvis	1.03 (2.37)	Legs	3.62 (3.19)
Knees	2.39 (3.11)	Ankles	1.67 (2.85)
Feet	1.99 (3.14)	Other	0.56 (1.93)

0 – 10 NRS, 0 to 10 Numerical Rating Scale of pain intensity; BPI, Modified Brief Pain Inventory pain interference scale; MHI-5, Mental Health Scale from the SF-36.

* For all study participants (N = 182), including those who reported no pain at the site.

Table 2

Multiple regression analyses predicting pain interference from pain extent and pain site

Step and variables	Total R^2	R^2 change	F change	Beta
1. Average pain intensity in previous week	0.39	0.39	116.16†	0.61
2. Legs pain intensity	0.45	0.06	20.01†	0.23
3. Feet pain intensity	0.47	0.02	5.37*	0.12
4. Hips pain intensity	0.49	0.02	5.53*	0.13
5. Knees pain intensity	0.49	0.01	4.95*	−0.12

*P < 0.05.

†P < 0.001.