



Pain in FSHD and other Neuromuscular Diseases

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Pain in FSH and other Neuromuscular Diseases

- Pain is a well known symptom in chronic debilitating conditions, such as osteoarthritis, diabetes, post-polio syndrome
- Not been described as a significant clinical problem in slowly progressive neuromuscular diseases such as Facioscapulohumeral muscular dystrophy (FSHD)



Case Report of Pain in 4 FSH Patients – Bushby*

- Described pain character, persistence, severity, frequency, site, and precipitating and relieving factors
 - Pain was their most disabling symptom
 - 3 - 7 separate pain complaints.
 - None had more than one pain-free day per month
 - All complained of disturbed sleep.
 - Response to conventional first-line pain treatments (e.g., analgesics) was poor.
 - 1 patient responded well to morphine during bad pain episodes
 - 3 individuals reported benefit from swimming or hydrotherapy

*Bushby K et al. Neuromusc Disord 1998; 8:574-579.



Pain in FSH and other Neuromuscular Diseases

- Not a focus of clinicians who study neuromuscular diseases
- Pain is not cited as a common problem in textbooks on neuromuscular disease
- Our clinical experience suggested that chronic pain may be a serious problem for persons with NMD.
- Began a multi-center program of research to study the nature and scope of chronic pain and its impact on functioning in patients with NMD.



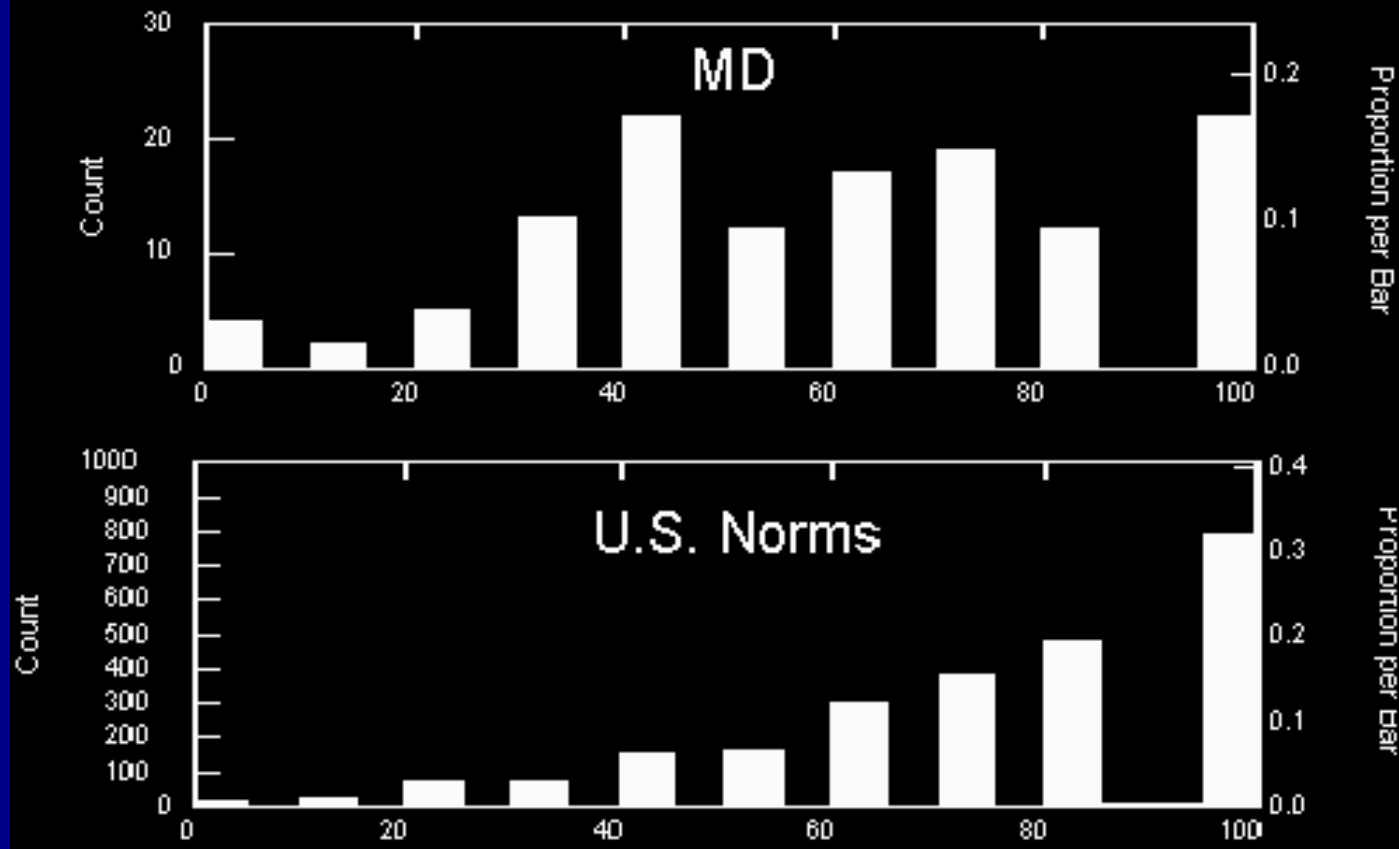
Quality of Life Survey: 811 subjects with NMD*

- 128 MD, 97 SMA, 232 CMT, 355 Post-polio
- Short Health Form-36 (sf-36)
- 83% of 811 participants had some pain
- 54% reported that their pain was moderate to very severe

* Abresch RT, Carter GT, Jensen MP, Kilmer DD. Am J Hosp Palliat Care 2002; 19:39-48.

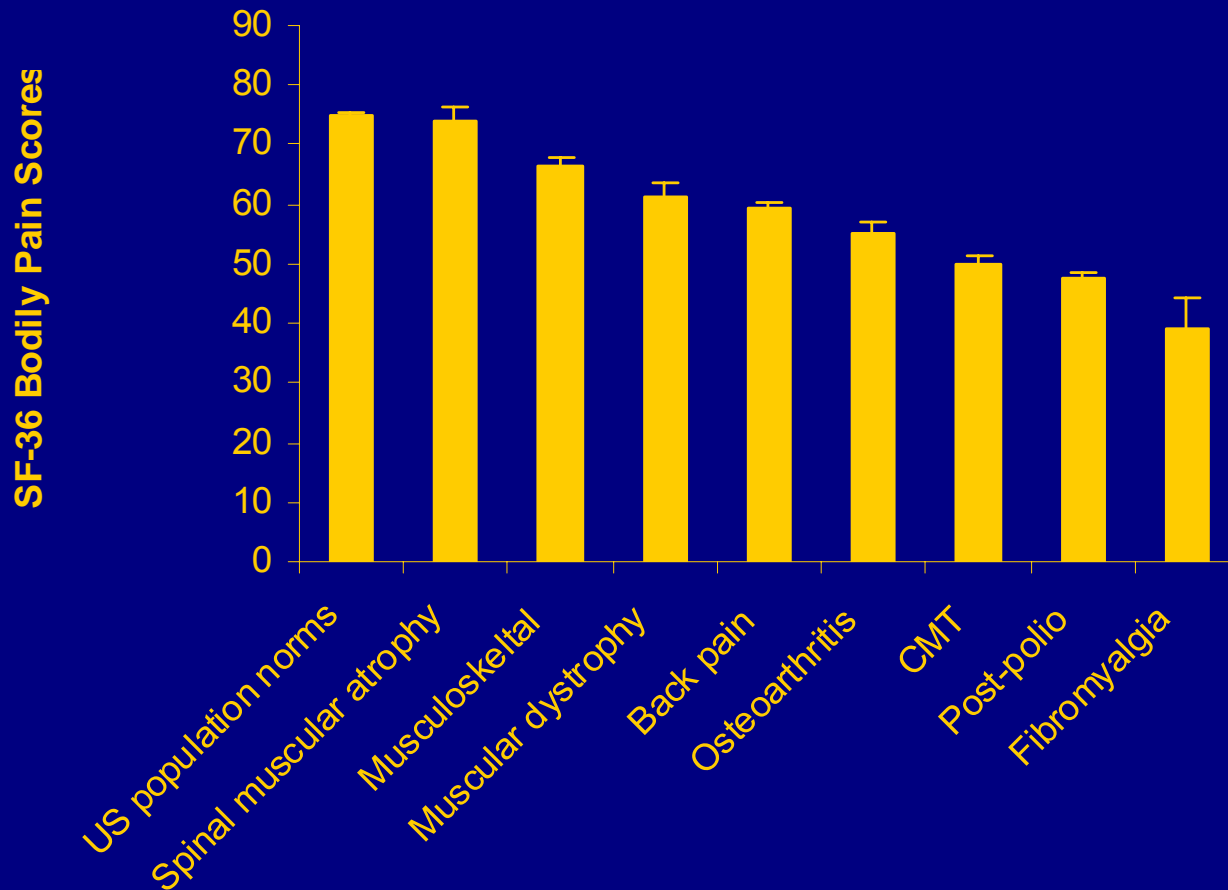


sf-36 Bodily Pain Scores





sf-36 Bodily Pain Scores





Correlation between sf-36 and Bodily Pain in MDs

	Correlation Coefficient
Physical Function	0.32
Role-Physical	0.51
General Health	0.55
Vitality	0.55
Social Function	0.58
Role-Emotional	0.4
Mental Health	0.39



Problems impacting QOL: Correlation with sf-36 Pain

	% with problem	Correlation with pain
Muscle weakness	96.5	-0.32
Fatigue	95.2	-0.53
Difficulty getting exercise	85.8	-0.35
Muscle spasms	71.1	-0.38
Sleep disturbance	68.1	-0.21
Problem controlling weight	67.4	-0.44
Coping with stress	66.2	-0.23



NIH Survey of MMD & FSHD

- Participants were recruited from the following sources: the NIH-funded National Registry of Myotonic Dystrophy and Facioscapulohumeral Muscular Dystrophy Patients and Family Members
- 395 surveys were mailed out
- 257 completed and returned survey



Demographics

	All subjects	MMD subjects	FSHD subjects
Number	257	130	127
Gender % (M/F)	45 / 55	42 / 58	48 / 52
Age	49 ± 13	47 ± 12	52 ± 14



Measures

- **Demographic characteristics and NMD-related information.**
- **Grading of Chronic Pain scale**
(0="no pain" to 10="pain as bad as could be")
- **Brief Pain Inventory Pain Interference scale**
Extent to which pain interferes with 12 specific activities during the preceding week on a 0 ("does not interfere") to 10 ("completely interferes") scale
- **Pain site(s) and intensity at specific site(s).**
- **Pain treatments and relief provided by pain treatments**



Pain Frequency & Severity

(0="no pain" to 10="pain as bad as could be")

	MMD subjects	FSHD subjects
Patients reporting pain	78%	82%
Average pain	4.5 ± 2.8	4.4 + 2.4
Subjects reporting severe pain	24%	23%
Duration of pain (years)	11.2	13.4



Pain Interference

0 = “does not interfere” to 10 = “completely interferes”

	MMD subjects	FSHD subjects	Correlation with pain intensity
Mobility	4.0	3.7	.63
Work	3.9	3.4	.59
Enjoyment	3.7	3.5	.52
General activity	2.9	2.9	.58
Relations with others	2.7	2.0	.51
Self-care	2.1	2.3	.41
Communication with others	1.6	2.2	.43



Pain Locations & Intensity

0="no pain" to 10="pain as bad as could be"

	MMD		FSHD	
	%	Intensity	%	Intensity
Headache	17%	4.8	14%	4.9
Shoulder	49%	4.4	69%	4.8
Lower back	66%	5.5	74%	5.6
Legs	64%	5.9	72%	4.9
Knees	43%	5.8	52%	4.6
Feet	48%	6.6	29%	4.0
Hands	47%	5.2	32%	3.8



Pain Treatments & Average Pain Relief

0="no relief" to 10="complete relief"

	All patients		Patients with severe pain	
	Tried / Still use	Relief	Tried / Still use	Relief
Ibuprofen, Aspirin	32% / 46%	5.3	41% / 42%	4.3
Heat	45% / 26%	5.2	44% / 35%	5.0
Massage	30% / 16%	5.2	28% / 16%	6.1
Narcotics	25% / 8%	6.5	30% / 16%	6.7
Physical Therapy	50% / 7%	3.6	49% / 16%	3.2
Marijuana	7% / 4%	6.0	12% / 9%	8.3
Chronic manipulation	28% / 6%	5.3	35% / 0%	5.5



Causes of pain in FSHD

- Musculoskeletal system pain generators
- Impaired mobility
- Joint instability secondary to muscle weakness, leading to joint destruction
- Mobility impairments lead to deconditioning and decreased pain tolerance



Causes of pain in FSHD

- Loss of range of motion, particularly in shoulders can cause complex regional pain syndrome such as frozen shoulder
- Joint contractures
- Immobility and neck and trunk spinal weakness leads to poor spinal support causing neck and back pain
 - Spinal bracing may be used to improve sitting posture and balance along with specialized wheelchairs



Treatment of pain in FSHD

- Begin with an initial diagnostic workup to determine if there is a modifiable pathophysiologic process that might underlie the pain
- Consider all ways a person is affected by pain
 - Pain intensity
 - Emotional impact of pain
 - Limitations to engage in daily activities
 - Limitations in social role such as employment
- Establish identifiable treatment goals
 - Measurable markers of pain relief
 - Improvements in social roles (return to employment)



Rehabilitation Measures for Treatment of Pain

- Best management is to proactively prevent the development of painful musculoskeletal syndromes
- Muscle weakness and progressive loss of mobility contributes to pain generation
- Generalized loss of cardiopulmonary fitness or “deconditioning” lowers pain tolerance and contributes to depression
 - ➔ **Low impact aerobic exercise & strengthening exercise**
 - ↑ cardiorespiratory fitness & ↓ fatigue
 - ↓ pain in patients with NMDs
 - ↑ mood, ↑ psychological well-being
 - ↑ appetite and ↑ sleep



Rehabilitation Measures for Treatment of Pain

- A good stretching program may help prevent painful musculoskeletal conditions such as joint contracture
 - If painful joint contracture develops more aggressive treatment may be indicated including corticosteroid injections to reduce inflammation
 - Topical heat and ice (short term measures may be effective)
 - Aggressive manipulation is contraindicated due to risk of joint damage
- Spinal deformities are manifestations of joint contracture causing back and neck pain (particularly if the patient is wheelchair dependent)
 - Spinal bracing may be used to improve sitting posture and balance, but it will not prevent or treat scoliosis



Rehabilitation Measures

- Any mobility device must be fitted carefully
 - Adequate lumbar support
 - Good cushioning (eg. Gelfoam)
 - Good overall pressure relief to avoid pain and pressure ulcers
 - Foam wedges to facilitate proper positioning
- Daily passive and active range of motion is helpful in treating pain from immobility



Conclusions

- Pain is a common problem in FSHD
- Most frequent pain sites were lower back and legs
- Pain problems are chronic lasting 11 – 13 years in our samples
- Highlights the need to identify and provide effective pain treatments.