# Pain Assessment in Facioscapulohumeral Muscular Dystrophy

Pain can be distinguished, based on pathophysiology, in nociceptive (i.e. musculoskeletal pain) and neuropathic. The first originates from chemical, thermal, mechanical stimulation of pain receptors of A-delta and C fibers and it is secondary to tissue damage. The second arises as a consequence of a lesion or disease affecting the somatosensory system (Treede RD, Jensen TS, Campbell JN, Cruccu G, Dostrovsky JO, Griffin JW, Hansson P, Hughes R, Nurmikko T, Serra J. Neuropathic pain. Redefinition and a grading system for clinical and research purposes. Neurology. 2008 Apr 29;70(18):1630-5)

Chronic pain is defined by the IASP as constant or intermittent daily pain, persisting for greater than three months, (Classification of chronic pain. Descriptions of chronic pain syndromes and definitions of pain terms. Prepared by the International Association for the Study of Pain, Subcommittee on Taxonomy. Pain Suppl. 1986; 3:S1-226.) usually after the tissues are expected to have healed. There may or may not be a persistent irritant that continues to cause pain. Once pain becomes chronic, it no longer has a protective role, with the pain continuing despite a lack of obvious, ongoing tissue damage. It is thought that once nociceptive pain becomes chronic there is also a neuropathic pain component, with synapse changes that are established through persistent use of pain pathways. (Stephenson R. The complexity of pain: part 2. Pain as a complex adaptive system. Phys Ther Rev. 1999; 4:183-94)

A large variety of pain are common in neuromuscular diseases. The chronic pain is frequent in FSHD (Jensen MP, Hoffman AJ, Stoelb BL, et al. Chronic pain in persons with myotonic dystrophy and facioscapulohumeral dystrophy. Arch Phys Med Rehabil. 2008;89:320-8; Della Marca G, Frusciante R, Vollono C, Iannaccone E, Dittoni S, Losurdo A, Testani E, Gnoni V, Colicchio S, Di Blasi C, Erra C, Mazza S, Ricci E. Pain and the alpha-sleep anomaly: a mechanism of sleep disruption in facioscapulohumeral muscular dystrophy. Pain Med. 2013 Apr;14(4):487-97), and may be present in a vast majority of the patients (up to 89%) (Jensen MP, Abresch RT, Carter GT. The reliability and validity of a self-report version of the FIM instrument in persons with neuromuscular disease and chronic pain. Arch Phys Med Rehabil. 2005;86:116-22); it may be disabling, and it may interfere with several activities of daily living (Jensen MP, Abresch RT, Carter GT. The reliability and validity of a self-report version of the FIM instrument in persons with neuromuscular disease and chronic pain. Arch Phys Med Rehabil. 2005;86:116-22; Jensen MP, Hoffman AJ, Stoelb BL, et al. Chronic pain in persons with myotonic dystrophy and facioscapulohumeral dystrophy. Arch Phys Med Rehabil. 2008;89:320-8; Abresch RT, Carter GT, Jensen MP, Kilmer DD. Assessment of pain and health-related quality of life in slowly progressive neuromuscular disease. Am J Hosp Palliat Care. 2002;19:39-48; Bushby KM, Pollitt C, Johnson MA, Rogers MT, Chinnery PF. Muscle pain as a prominent feature of facioscapulohumeral muscular dystrophy (FSHD): Four illustrative case reports. Neuromuscul Disord. 1998;8:574-9).

Overall, chronic pain has a significant impact on physical, emotional, and cognitive function, on social and family life, and on the ability to work and secure an income (Breivik H, Collett B, Ventafridda V, Cohen R, Gallacher D. Survey of chronic pain in Europe: prevalence, impact on daily life, and treatment. Eur J Pain. 2006; 10: 287-333). Consequentially, assessment of long-lasting pain is therefore a more challenging task than assessing acute pain.

This is true both in clinical practice and when conducting trials of management of long-lasting pain.

(Melzack R, Katz J. Pain assessment in adult patients. In: McMahon SB, Koltzenburg M, eds. Wall and Melzack’s Textbook of Pain, 5th Edn., London: Elsevier, 2006; 291-304).

## Assessment of pain

A comprehensive assessment of any chronic pain condition requires documenting pain history, physical examination, and specific diagnostic tests.

### Pain history

A medical history is an important part of the pain history, often revealing key aspects of co-morbidities implicated in complex pain condition. The specific pain history must define location, intensity, pain descriptors, temporal aspects, and possible pathophysiological and aetiological issues.

1. Where is the pain?
2. How did the pain start?
3. How intense is the pain?
4. Description of the pain (e.g. aching, burning, stabbing, throbbing, etc).
5. What is the time course of the pain?
6. What aggravates the pain?
7. What relieves the pain?
8. How does pain affect?
   1. sleep?
   2. physical functions?
   3. ability to work?
   4. economy?
   5. mood?
   6. family life?
   7. social life?
   8. sex life?
9. What treatments have the patient received? Effects of treatments? Any adverse effects?
10. Is the patient depressed?
11. Is the patient worried about the outcome of your pain condition and your health?
12. Is the patient involved in a litigation or compensation process?

### Physical examination

1. General physical examination;
2. Specific pain evaluation (including specific evaluation of cold allodynia, heat allodynia, cotton wool and brush for dynamic mechanical allodynia, and a blunt needle for hyperalgesia and temporal summation of pain stimuli);
3. Neurological examination;
4. Musculoskeletal system examination;
5. Assessment of psychological factors.

### Specific diagnostic studies

1. Quantitative sensory testing (QST) with specific and well-defined sensory stimuli for pain thresholds and pain tolerance. (Shy ME, Frohman EM, So YT, Arezzo JC, Cornblath DR, Giuliani MJ, Kincaid JC, Ochoa JL, Parry GJ, Weimer LH; Quantitative sensory testing: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology. 2003 Mar 25;60(6):898-904)
2. Laser evoked potentials (Bromm B, Treede RD. Nerve fibres discharges cerebral potentials and sensations induced by CO2 laser stimulation. Human Neurobiol. 1984; 3: 33-40)
3. Conventional radiography, computerized tomography, magnetic resonance imaging.

## Chronic pain assessment tools.

Several assessment tools are developed; the following are documented to be reliable and valid in several languages.

### The Brief Pain Inventory

The Brief Pain Inventory (BPI) was developed from the Wisconsin Brief Pain Questionnaire. (Daut RL, Cleeland CS, Flanery RC. Development of the Wisconsin Brief Pain Questionnaire to assess pain in cancer and other diseases. Pain. 1983; 17:197-210)

The BPI assesses pain severity and the degree of interference with function, using 0–10 numeric rating scale (NRS). It can be self-administered, given in a clinical interview, or even administered over the telephone. Most patients can complete the short version of the BPI in 2 or 3 min.

Chronic pain usually varies throughout the day and night, and therefore the BPI asks the patient to rate their present pain intensity, ‘pain now’, and pain ‘at its worst’, ‘least’, and ‘average’ over the last 24 h.

Location of pain on a body chart and characteristics of the pain are documented.

The BPI also asks the patient to rate how much pain interferes with seven aspects of life: (1) general activity, (2) walking, (3) normal work, (4) relations with other people, (5) mood, (6) sleep, and (7) enjoyment of life.

The BPI asks the patient to rate the relief they feel from the current pain treatment (Wang XS, Cleeland CS. Outcomes measurement in cancer pain. In: Wittink HM, Carr DB, eds. Pain Management: Evidence, Outcomes, and Quality of Life. A Sourcebook. London: Elsevier, 2008; 361-76).

### The McGill Pain Questionnaire, short-form McGill Pain Questionnaire, and Adapted Versions Of The MPQ

The McGill Pain Questionnaire (MPQ) and the short-form MPQ (SF-MPQ) evaluate sensory, affective-emotional, evaluative, and temporal aspects of the patient’s pain condition.

The McGill Pain Questionnaire, a widely used multidimensional tool for evaluation of pain, consists of 78 verbal descriptors in 4 large groups (1-sensory, 2-affect, 3-evaluative and 4-miscellaneous) and distributed in 20 subgroups. Each descriptor has a specific rank value, ranging from 1 to 6 and represented in the results as subscript figures. The patient’s descriptor choice gave us two measures: the number of descriptors chosen and the pain level, obtained by adding up the chosen descriptors’ rank values (Melzack R. The McGill Pain Questionnaire: major properties and scoring methods. Pain. 1975 Sep;1(3):277-99).

The SF-MPQ consists of 11 sensory (sharp, shooting, etc.) and four affective (sickening, fearful, etc.) verbal descriptors. The patient is asked to rate the intensity of each descriptor on a scale from 0 to 3 (=severe). Three pain scores are calculated: the sensory, the affective, and the total pain index. Patients also rate their present pain intensity on a 0–5 scale and a visual analogue scale VAS (Melzack R, Katz J. Pain assessment in adult patients. In: McMahon SB, Koltzenburg M, eds. Wall and Melzack’s Textbook of Pain, 5th Edn. London: Elsevier, 2006; 291-304).

ADAPTED VERSIONS OF THE MPQ

The knowledge that there are differences in pain reports between languages and people from different cultures made researchers from different countries start the elaborate work of developing adapted versions of the MPQ. (Strand LI and Ljunggren AE. Different approximations of the McGill Pain Questionnaire in the Norwegian language: a discussion of content validity. J Adv Nurs. 1997 Oct;26(4):772-9.)