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# Pain Management in the Ehlers-Danlos Syndromes

## FOR NON-EXPERTS

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Long-term (chronic) pain in the Ehlers-Danlos syndromes (EDS) appears early, is common, and may be severe. There has not been enough research on treatments to properly guide how pain in EDS should be managed. Causes and contributors to pain in EDS can include joints coming out of position, previous surgery (often done to treat pain), muscle weakness, improper movement in the neck and back, or issues with sense of joint position. People affected can come to a doctor with general body pain, tiredness, headaches, or pain in the stomach, genitals, face, or jaw. Management can focus on treating the cause (like joints coming out of position) and lowering the sensation of pain. Methods of pain management in EDS include physiotherapy, medication, cushions, compressive clothes, and braces, as well as behavioral adaptation.

## A Review of Literature About Pain in the Ehlers-Danlos Syndromes

Pain in the Ehlers-Danlos syndromes (EDS) may be linked to the level of joint mobility, how often joints come out of position (dislocate or sublux), how prone the person is to injury, previous surgery, muscle pain, and may become long-term. Pain can be limited to muscles or can be more widespread and may occur over a short period or continue over long periods of time. Pain may interfere with many aspects of daily living and can affect sleep quality (which is common in EDS), contributing to other difficulties independent of the level of tiredness.

Research has consistently found pain to be common in those with EDS and hypermobile type EDS (hEDS). In EDS, pain often begins in joints or limbs, which is influenced by factors such as lifestyle, sports activities, previous damage or surgery, and existing conditions. Many patients report their first painful sensations in relation to an incident such as dislocations, sprains as well as "growing pains" mostly localized to the knees or thighs. The majority of patients with hEDS were female. Children may not be believed by doctors about their pain and can be wrongly diagnosed or dismissed based on this. Children can also come to the doctor with recurring unexplained bruising or multiple joint problems that lead to accusations of child abuse.

Neck pain is a common feature of hEDS and is often associated with headaches. Loose ligaments connecting the skull and neck as well as a hypermobile neck can lead to problems: swallowing, speech difficulty, changes in walking, bodily weakness, uncontrolled muscle contractions, altered senses, and changes in automatic (autonomic) bodily functions like heart rate, blood pressure, body temperature, and digestion. Headaches often occur in EDS and were more common and more disabling in those with hEDS. A recent study reported headaches occurring in a third of patients with EDS. Issues with tension and stress or problems with the jaw may be major causes of headaches.

## Chronic Pain

Chronic pain (pain that lasts a long time), is one of the major symptoms showing in patients with hEDS and is experienced by a very large proportion of hEDS patients. Chronic pain can often be a general body pain, affecting almost every part of the patient. Loss of sense of joint position could be an important factor in hEDS-related chronic pain. The detection of position, movement, and interaction of the body with the environment is needed to allow balance and prevent damage to joints and ligaments. Loss of this sense may be due to excessive joint movement damaging the

ability to sense, or due to pain lowering the ability to sense. Improving this positional sense may help improve functions like balance as well as chronic pain.

The evidence to show the precise cause of pain in hEDS is weak. Proposed mechanisms are drawn from other chronic pain conditions and require further study. Investigation into the biological causes of pain in EDS suggests it is not caused by nerve damage, but an increased sensitivity, possibly sharing mechanisms with a condition called fibromyalgia. Treatment of chronic pain should be in treating the underlying cause of the pain.

## Pain Management

Management of chronic pain in hEDS is made difficult by a lack of evidence-based research specific to EDS showing effective approaches. Pain management adapts and alters methods used in non-EDS patients. Generally, pain management focuses on treating the cause of pain (such as dislocation of a joint) and minimizing the sensation of pain.

### Key Points in the Management of Pain in Hypermobile Type EDS

1. Successful pain management requires several approaches working together.

2. Physiotherapy: evidence suggests patients who receive exercise-based treatments improve over time. Stretching exercises should be very gentle to avoid causing joint injury.

3. Cognitive behavioral therapy: may be helpful for all patients, especially those whose pain is difficult to control.

4. Medication and devices: These are varied and each can have benefits and drawbacks.

    - Drugs for pain. These may be used depending on the type of pain such as pain from inflammation, or for immediate relief from severe pain. Some drugs used for treating pain such as opioids or NSAIDs (ibuprofen, naproxyn) can be harmful over long periods of time or in EDS patients with a condition called mast cell activation syndrome. Drugs for nerve pain may not be suitable in hEDS because of the effect they have on bodily functions already badly affected. Lidocaine is a

short-lasting drug for local pain relief and can be useful when joints misalign, for gum pain, and in serious cases of painful intercourse.

- Hormonal control. For those with painful periods or worse symptoms around menstruation.
- Transcutaneous neuro stimulator (TENS) for pain: a device to block pain signals.
- Cushions and mattresses: ease discomfort during sitting/lying.
- Treatments for positional sensing: compressive (tight) clothing, physiotherapy, shoe inserts.
- Drugs for uncontrolled muscle contractions: some drugs or combinations of drugs may improve uncontrolled muscle contractions, pain, and tiredness.
- Treatment of fatigue and pain: as fatigue and pain are linked when it comes to disability, treating fatigue can help with pain, and managing pain can help with fatigue.

## Future Directions

The management of the often severe, changing, debilitating pain in patients with hEDS is currently not good enough. Commonly-used pain medications do not help treat most patients, probably because the cause of pain is different to most cases. We think that more research into pain management, fatigue management, and uncontrolled muscle movements is needed, as well as a better understanding of how the patient uses energy and their positional sensing.

Research into the following subjects are urgently needed:

- Identifying the differences between fibromyalgia and hEDS. The cause of pain may be different in fibromyalgia and hEDS, meaning that we might need to use different treatments. Many people are diagnosed improperly with fibromyalgia and not getting ideal treatment.
- Early diagnosis. Diagnosing early could lead to better quality of life. Following on from this, looking at better ways of diagnosing EDS in children would be helpful.
- Research into using anti-inflammatory drugs (NSAIDS).

- Oxygen therapy. Giving oxygen with a face mask can help with certain serious migraines, and decreases pain in EDS.

- Positional sensing problems. Looking at how positional sensing relates to pain and fatigue.

- Hormonal changes and pain. More work needs to be done to understand how hormones alter pain management in EDS.

- Treating uncontrolled muscle contractions: can increase joint problems and therefore pain.

- How the body uses energy in hEDS. Problems with how energy is used by cells may be contributing to fatigue, but needs more attention.

- Low Dose Naltrexone (LDN). A drug called naltrexone may be helpful in some patients with specific kinds of pain and has been reported as helpful for managing chronic pain in hEDS.

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*This article is adapted from:* Chopra P, Tinkle B, Hamonet C, Brock I, Gompel A, Bulbena A, Francomano C. (2017). Pain management in the Ehlers-Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C:212–219. http://bit.ly/2obxv5v

*[Updated Feb. 17, 2020]*