

Ehlers- Danlos Syndrome and Chronic pain: Pathophysiology and Management Principles

Abstract: Questions from patients about Ehlers-Danlos syndrome and associated chronic pain are answered to help the patients understand the mechanisms for pain in this connective tissue disorder as well as general treatment principles for chronic pain management.

Keywords: Ehlers-Danlos syndrome, hypermobility, chronic pain, rehabilitation

Question from a patient: I have Ehlers-Danlos Syndrome, Type 4, and have been in pain for 35 years. For the last 11 years I've been taking strong opioids, which haven't helped like I had hoped and make me very sleepy. I have been to many medical centers, to no avail. I have been pretty much housebound for the past 3 years. I'm only 47, but feel like 147! I need my life back! Can you shed any light on what could help me?

Ehlers-Danlos Syndrome (EDS) is a group of inherited disorders that are known for a defect in a protein called collagen. Collagen acts like "glue" and is one of the main components of connective tissue which help provide support to many structures in our bodies including skin, muscles and ligaments which support joints, and even our blood vessels and organs depending on the severity of the disorder. A defect in this "glue" leads to excessive laxity or "looseness" of the affected structures. While having more flexibility may be perceived as beneficial, this can actually be harmful. As an example, think of the tightened cables that hold a suspension bridge together. If the cables become loose, the bridge would potentially break down. Just like the bridge, our bodies can become damaged more easily and rapidly due to these "loose cables".

Commonly, "looseness" in our cables can lead to misalignment (also known as subluxation or dislocation) of the joints, even with little movement or trauma. Repeated episodes of subluxation can result in early and accelerated osteoarthritis or joint "wear and tear" due to joint instability and uneven stress on the joints. In addition to joint pain, patients may also experience pain from increased stress of the muscles and ligaments around a joint as well as neuropathic or nerve-related pain. A serious complication of these long-standing types of pain is chronic pain where the pain-sensing system in the spinal cord and brain has malfunctioned and residual pain signals from a site of a healed injury may persist or become amplified. This can be extremely disabling from physical, emotional, and social standpoints.

Unfortunately, there is no specific cure for EDS. Furthermore, the use of opioid medications in patients with chronic, non-cancer pain such as yourself remains controversial. While some patients may display noticeable improvements in both pain relief and ability to perform activities or exercise, other patients may only experience minimal or decreasing benefits from these medications due to developing physiological tolerance (a reaction whereby over time, larger doses of a medication are required to achieve the same effect) or having pain that is not responsive to opioids. In addition, side effects (such

as sleepiness or difficulty with thinking) can be caused by taking a dose that is too high or taking multiple drugs that interact with one another-- this can actually lead to a decrease in activity level. Given the fact that you have been taking strong opioids for 11 years with no significant benefits and sedating side effects, it may be important to have a discussion about ongoing short and long-term risks and alternatives of chronic opioid therapy with your prescribing physician.

Treatment plans that have traditionally been geared towards patients with diffuse, chronic joint disorders such as rheumatoid arthritis can be designed to improve quality of life and daily function. Seeking consultation by a specialist in Physical Medicine and Rehabilitation may be helpful in developing a successful rehabilitation plan.

This approach involves an individualized assessment of a patient's physical, mental, and emotional limitations and goals. Although a common goal is to improve pain relief, an equal and perhaps more important goal is to improve physical function. Goals can range from being able to dress without assistance to walking outside the home to socialize with friends.

An example of a multidisciplinary rehabilitation approach might include a combination of the following treatments:

- Physical therapy visits to develop individualized exercise, rest, and energy conservation programs to learn appropriate exercise activities and precautions to safely strengthen muscles, protect joints, and prevent further accumulation of damage.
- Occupational therapy visits to improve the ability to perform daily activities of living such as dressing, grooming, eating, and household chores
- Physical modalities such as heat, cold, ultrasound, and electrical stimulation to decrease pain during exercise or activity
- Bracing options to help protect and support joints, prevent or correct joint deformities, and improve the ability to perform activities safely with less pain
- Education and self management in the form of support groups to learn more about the disease process, pacing strategies, and other problem solving skills to reduce visits to healthcare providers and associated costs
- Pain psychologist visits to help identify psychological disorders such as depression or anxiety that can limit progress and learn tools to help cope with chronic pain such as biofeedback or relaxation
- Pain medications or injection therapies to reduce pain to thereby allow for improved activity level
- Surgical evaluations to correct or prevent worsening joint deformities
- Complementary alternative medicine therapies such as acupuncture

Since this approach involves different treatments which can be hard to coordinate, a referral to a comprehensive interdisciplinary pain management program may be worthwhile when other options have failed. These programs may last 7 to 8 hours per day for multiple weeks and involve a "team" of health care providers that meet regularly to improve communication and coordination of care. These

programs focus on the patient being an active participant in the rehabilitation process. After completing the program, patients are encouraged to continue using these learned techniques to further improve their independence and function.

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