FROM THE EDITOR’S DESK: PAIN & MANAGING
PAIN FROM AN EDS PATIENT’S VIEW

I HAVE been studying Ehlers-Danlos syndrome since I was diagnosed more than ten years ago, but I still find it helpful at times to think through from the fundamentals. EDS is a collection of genetic collagen defects. Each type of EDS is defined as a distinct problem in making or using one of the types of collagen. Collagen is the most abundant protein in the body, so effects can be wide-ranging in both location and severity. Collagen is what the body uses to provide strength and elasticity to tissue; normal collagen is a strong protein that allows tissue to be stretched, but not beyond its limit, and then safely returned to normal. In most EDS types, the affected collagen is badly structured, which means that any tissue using the mutated collagen can be pulled beyond its normal limits, causing damage.

What can still surprise me is how widespread the effects of having a mutated type of collagen can be. There are five main collagen types that are affected by EDS, as far as is known to date; not all the genetic causes of EDS are known, and in particular, EDS Hypermobility Type is still largely undetermined (although a tenascin-X mutation accounts for a small percentage). One of the fundamental keys we are missing is why EDS expresses in such a wide range of severities; even in the same generation of a family with the same mutation, it can be mild in one person and severe in another. EDS was not named until 1936, so it is still a young disorder; we have learned most of what we know about EDS since the last diagnostic and nosology conference in 1997, but there is still so much we do not understand. There will be new information after the next nosology, to be formulated at the First International EDS Symposium in Belgium, September, 2012.

EDS pain can be severe and widespread. Built with collagen, the fascia is the sort of “Saran Wrap” that holds our bodies together, a connective tissue that surrounds muscles, groups of muscles, blood vessels, and nerves. The result of an overly-stretchable fascia can be literally body-wide because it is wrapped around nearly everything.

Chronic pain is a well-established and cardinal manifestation of EDS and it is common for pain to be out of proportion to physical and radiological findings. The etiology of EDS pain is not clearly understood, but some of the likely causes include muscle spasm (tender points are sometimes present) and degenerative arthritis; neuropathic pain is also common. (EDNF’s Pain Management Medical Resource Guide http://www.ednf.org/index.php?option=com_content&task=view&id=2123&Itemid=88889247)

Chronic frequently debilitating pain of early onset and diverse distribution is a constant feature in most individuals affected with different types of EDS.

- 90% of EDS population has chronic pain for more than 6 months
- 84% feel it worsened over their lifetime
- 89% had chronic pain before adulthood
- 88% have taken pain medication
- 51% have taken narcotics

Chronic pain, distinct from that associated with acute dislocations or advanced osteoarthritis, is a serious complication of the condition and can be both physically and psychologically disabling. ([Ehlers-Danlos Syndrome: Hypermobility Type](http://www.ncbi.nlm.nih.gov/books/NBK1279/))


Types of pain in EDS: headache, musculoskeletal pain—acute and chronic, gastrointestinal/abdominal pain

Headache in EDS and other Hypermobility syndromes: migraine, ‘low pressure’ headaches, ‘high pressure’ headaches, myofascial pain, sinus pain, temporo-mandibular joint problems, dental pain

Musculoskeletal Pain in EDS and other Joint Hypermobility Syndromes: joint laxity predisposes to acute injury, muscle spasm in response to acute injury or as a means to stabilize unstable joints, myofascial trigger points develop in response to joint laxity, nerve impingement may result from disc disease or joint laxity causing muscle spasm as well as neuropathic pain

Gastrointestinal Pain in EDS: dysmotility, constipation, irritable bowel syndrome [and food allergies], sphincter of Odi dysfunction


Of course, there is an element of the traditional view of chronic pain, synaptic plasticity, in which the body learns pain and increases pathways for pain, setting up a cycle that remodels the nervous system. “A cardinal feature of EDS is pain. At first there are just small pains; but acute pains may accumulate, then become continual and chronic. There may be a major dislocation of injury to start the cycle; but without adequate treatment, persistent pain can change the nervous system in a process that is difficult to reverse. By lowering the threshold for pain signals, chronic pain becomes harder to treat.” (EDNF’s [Pain Management Medical Resource Guide](http://www.ednf.org/index.php?option=com_content&task=view&id=2123&Itemid=88889247))

But there is an added element to EDS pain that we have started recognizing these past few years: we suffer what might be called chronic acute pain. Some of us suffer frequent dislocations, and most of us endure at least joint subluxations. We do not injure a joint once and then recover. We injure a joint over and over, because if we are in motion, our joints are moving out of alignment. As a personal example, my shoulders separate with about four pounds of force, so my arms are continually, literally, falling out of the socket, even at the computer. I once kept a journal for a week of how often joints slipped or dislocated; when I was sitting still, I still managed ten or so an hour, and there were many more when I was up and moving about doing things.

Every time a joint slips or dislocates, the soft tissue connected to that joint—tendons, ligaments, muscle, fascia, blood vessels—all stretches. Because of the structural defect
in collagen, all that connected tissue is stretched beyond what normal should be; high-resolution MRIs reveal microtrauma, microscopic tears that start up the inflammation and coagulation cascades. But because each joint can subluxate over and over in just a single day, these microtraumas happen over and over in the same tissue without healing successfully. This was first pointed out as an EDS problem in July 2011 by Dr. Clair Francomano in her comments while presenting at the EDNF Conference: “Microtears are not visible with ordinary MRIs but are experienced by EDSers, causing pain and instability.” (From my own notes, verified by notes at http://edsers.com/2011_Conference_Notes.html; Dr. Francomano’s presentation is a great summary, Ehlers-Danlos Syndrome Update 2011: What We Know—And What We Don’t Know, available from http://bit.ly/HQDNwk.)

Tissue damage has the potential to elicit mechanisms that can create disabling, refractory, chronic situations that may prolong and even outlast the period of healing. (Hedderich & Ness, 1999)

Shoulder instability is a relatively common occurrence, reported in 2% to 8% of the population. The condition may arise from a single traumatic event (ie, subluxation or dislocation), repeated microtrauma, or constitutional ligamentous laxity, resulting in deformation and/or damage in the glenohumeral capsule and ligaments. (http://blue.regence.com/trgmedpol/surgery/sur100.html)

We postulate that the propensity for tendon degeneration in Ehlers-Danlos syndrome coupled with repetitive microtrauma might lead to fraying of the tendon fibers and, consequently, triggering. (http://www.mendeley.com/research/multiple-triggering-girl-ehlersdanlos-syndrome-case-report/)

When a joint has more movement than is ideal, stress and strain are placed on the joint, causing microtrauma to the joint, and this can result in pain. (http://www.hypermobilot.com.au/uploads/7/0/8/7/7087979/pregnancy_a4.pdf)

No one really knows why this happens, but one theory is that every-day overuse or misuse of hypermobile joints causes “microtrauma” to the joint tissues. Eventually this “microtrauma” can build up and cause pain—a sort of “the straw that broke the camel’s back” situation.” (http://hypermobilityhope.blogspot.com/p/what-is-hms.html)

It has been speculated that when tendon is overused it becomes fatigued and loses its basal reparative ability, the repetitive microtraumatic processes thus overwhelming the ability of the tendon cells to repair the fiber damage. The intensive repetitive activity, which often is eccentric by nature, may lead to cumulative microtrauma which further weakens the collagen cross-linking, non-collagenous matrix, and vascular elements of the tendon. Overuse has also been speculated to cause chronic tendon problems, by disturbing the micro- and macrovasculature of the tendon and resulting in insufficiency in the local blood circulation. Decreased blood flow simultaneous with an increased activity may result in local tissue hypoxia, impaired nutrition and energy metabolism, and together these factors are likely to play an important role in the sequence of events leading to tendon degeneration. (Histopathological findings in chronic tendon disorders [Järvinen et al] http://www.ncbi.nlm.nih.gov/pubmed/9211609)

It is thus reasonable to postulate that excessive laxity of ligaments may predispose an individual to repeated microtrauma during even normal activity. A possible reason for recurrent joint trauma in hypermobile persons may be the recently demonstrated impairment of proprioception noted in both small and large joints. It therefore follows that recurrent microtrauma to ligamentous structures in some hypermobile individuals will lead to repeated pain experience and may trigger disordered pain responses. ...A more immediate question might be the association of hypermobility and soft tissue complaints. It seems reasonable to hypothesize that loose ligaments may lead to skeletal structural instability, and predispose the body to repeated minor or more serious traumatic episodes. (Is Hypermobility a Factor in Fibromyalgia? [Fitzcharles] http://www.jrheum.com/abstracts/editorials/200106.html)

Within the physiological range, particularly towards the higher range, microscopic degeneration within the tendon may start to occur, especially with repeated and/or prolonged stressing. This can eventually lead to a symptomatic tendon with altered mechanical properties as a result of repeated microtrauma.
An added problem, suggested by one of my doctors, may be that axons do not stretch; with each dislocation or subluxation, neural pathways may be pulled apart and then reconnected, adding a neuropathic pain to the mix.

Because of the constant reinjury from joint laxity to tissue and neural pathways, I have been using the term “chronic acute pain,” acute pain that is recurring chronically. From these recurring problems comes much of EDS fatigue, I would suspect, although orthostatic intolerance and dysautonoma can account for much of the fatigue as well.

Symptoms Of Orthostatic Intolerance: Lightheadedness, Dyspnea, Syncope, Chest Discomfort, Diminished concentration, Palpitations, Headache, Tremulousness, Blurred vision, Anxiety, Fatigue, Nausea, Exercise intolerance, Nocturia

Working hypothesis: Connective tissue laxity in blood vessels allows increased vascular compliance, promotes excessive pooling during upright posture, leading to diminished blood return to the heart, and thus to OI symptoms (Rowe PC, et al. J Pediatr 1999;135:494-9)


- 273 patients with EDS
- 77% severe fatigue
- 57% reported fatigue as 1 of their 3 most important symptoms
- Severe fatigue was more common in hypermobile than classical EDS (84% vs. 69%; P=.032)
- Fatigue had a greater impact on daily function than did pain

Fatigue is a frequent and clinically relevant problem in EDS (Voermans et al. Semin Arth Rheum 2010; 40:267-74)


Severe fatigue afflicts 75% of those with EDS http://doctorstevesbanjo.com/severe-fatigue-afflicts-those-ehlersdanlos-syndrome/

One other major contributing factor to EDS pain is the spine. The spine is, of course, a series of joints, and like any joint in the body of someone with EDS, the vertebrae are potentially hypermobile, as is the junction where the skull sits on the spine. The effects can range, like everything in EDS, widely: Chiari Type I malformation, tethered cord, syringes, cerebrospinal fluid abnormalities, syringes, and craniocervical settling.

Hereditary disorders of connective tissue may present with Chiari I malformation, occipitoatlantoaxial hypermobility, and functional cranial settling [Francomano et al.] http://www.ednf.org/index.php?option=com_content&task=view&id=1618&Itemid=88889208


I have EDS and was just diagnosed with Chiari, is this related to EDS? http://www.ednf.org/index.php?option=com_content&task=view&id=1580&Itemid=88889064

Spine abnormalities are correlated with back pain in young persons with Ehlers-Danlos syndromes [Bangura et al.]

Urologic Manifestations of Tethered Cord Syndrome: Clinical and Urodynamic Findings [Murdock]

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**Physical therapy**

Physical therapy can be an important part of EDS pain management, with the goal of reducing muscle spasm and toning muscles around joints. There are some important factors to physical therapy for people with connective tissue disorders, and EDS specifically. In many ways, this approach is the opposite of traditional physical therapy. Instead of aggressive, short-term building of muscle mass through rapidly increasing weight or resistance, physical therapy for EDS has to be focused on long-term, gentle building of muscle tone. Special attention has to be paid to ensure that physical therapy does not continue to injure the person with EDS, that motion does not exceed normal range of motion and that joints are not ever hyperextended or slipping in place. While some degree of muscle ache is part of building muscle, one has to be aware with EDS and connective tissue disorder patients that any pain during physical therapy is probably signaling joint instability and subsequent tissue damage.

A physical therapist who is highly knowledgeable and deeply understanding about connective tissue and joint dysfunction can be useful to long-term health:

- use low resistance exercise to gradually increase muscle tone and to help stabilize loose joints;
- minimize joint trauma by avoiding joint hyperextension and any high resistance or impact activities;
- just as it often takes years for joint laxity to cause significant pain and instability, it can take at least months and usually years to gradually reverse the process via muscle toning exercise.


Physical therapy:

- Myofascial release (any physical therapy modality that reduces spasm) provides short-term relief of pain, lasting hours to days. While the duration of benefit is short and it must be repeated frequently, this pain relief may be critical to facilitate participation in toning exercise for stabilization of the joints. Modalities must be tailored to the individual; a partial list includes heat, cold, massage, ultrasound, electrical stimulation, acupuncture, acupressure, biofeedback, and conscious relaxation.
- Low resistance muscle toning exercise can improve joint stability and reduce future subluxations, dislocations, and pain.

...Improved joint stability may be achieved by low-resistance exercise to increase muscle tone (subconscious resting muscle contraction, as opposed to voluntarily recruited muscle strength). Examples include walking, bicycling, low-impact aerobics, swimming or water exercise, and simple range-of-motion exercise without added resistance. Progress should be made by increasing repetitions, frequency, or duration, not resistance. It may take months or years for significant progress to be recognized.


Manual Techniques

- Slow non-thrust manipulations
  - Sustained stretching
» Passive oscillatory movements (neural mobs)
» Muscle energy techniques
• Gentle indirect techniques
• Myofascial release
• Strain and counter-strain
• Cranio-sacral therapy


Some of the neurological features associated with this type is; primary muscular hypotonia may occur and may cause delayed motor development, problems with ambulation, and mild motor disturbance. Fatigue and muscle cramps are relatively frequent. Physical therapy can teach certain exercise techniques that can strengthen muscles around joints and may help to prevent or limit damage. Hydrotherapy (taking place in water) may be less damaging to joints.


Presently physical therapy management of patients diagnosed with Ehlers-Danlos syndrome focuses upon a compensatory/prevention approach to address the presence of specific signs and symptoms of the condition that vary specifically to each affected individual. Unfortunately, no protocol of physical therapy interventions exists to address the impairments and functional limitations associated with EDS, due largely in part to the varied presentation of the condition for each affected individual. Therefore, each physical therapy plan of care must be specially created for the patient depending upon the subtype of EDS and the signs and symptoms of that patient. In general, physical therapy intervention focuses on increasing joint stability through a low resistance, high repetition resistance training program, preventing excessive loading through weightbearing joints, avoiding excessive use of involved joints for heavy lifting, adaptive equipment to accomplish ADLs without accentuation of symptoms, and pain relief/pain management.

Resistance training
• Low resistance, high repetition activities
• Goal is to improve static and dynamic muscle tone to promote increased joint stability during weightbearing and functional activities
• Avoidance of recurrent joint subluxations/dislocations due to increased muscle tone to counteract presence of excessive joint, ligament, tendon, and muscle laxity

Aerobic training
• Walking
• Bicycling
• Low-impact aerobics and/or water aerobics
• Swimming
• ROM exercises
• Goal is to promote increased static and dynamic muscle tone to prevent acute joint subluxations/dislocations by minor trauma or stimulus
• May function as pain relief mechanism for individuals experiencing chronic joint and muscle pain associated with EDS

Myofascial release techniques
Pain relief (immediate - several hours)
- Allows pain free participation in resistance training or daily activities
- Goal is to reduce the presence of muscle spasms that result in intense pain in muscles and surrounding ligaments, tendon, and joints

Modalities
- Hot/cold pack
- Massage
- Ultrasound
- Electrical stimulation
- Acupuncture
- Acupressure
- Goal is to provide pain relief to the patient, who may/may not be experiencing chronic muscle and joint pain from frequent joint subluxations/dislocations, myofascial spasms, and trigger points associated with EDS
- Selection of proper modality is dependent upon patient preference

Adaptive Equipment
- Wheelchair/scooter
- Walker/Crutches/Cane (should be used with caution and discretion due to increased weightbearing through upper extremities with use)
- Modified eating utensils (prevents excessive strain placed on small joints of hands and fingers)
- Modified writing utensils (prevents excessive strain placed on small joints of hands and fingers)
- Modified sleeping surface (air mattress, viscoelastic foam mattress, pillow mattress)
- Goal is to allow daily functioning and promote increased quality of life by decreasing pain or chance of joint subluxation/dislocation

Despite diagnostic differences between Hypermobility Syndrome and genetic disorders (characterized by generalized joint hypermobility), such as Ehlers-Danlos Syndrome, similar treatment approaches and interventions remain relevant and appropriate between the two diagnostic categories. Russek advocates the use of education and exercise as potential interventions for Hypermobility Syndrome. Education on ergonomics and body mechanics may result in decreases in musculoskeletal pain as well as assist in joint protection strategies. Splints, braces, and taping may also function as viable options to protect vulnerable joints. Russek suggests that therapeutic exercises, such as strengthening, proprioceptive activities, balance, and coordination to affected and surrounding joints as a means for treatment of Hypermobility Syndrome.

Pain treatment and management

There are many solid reasons to treat and reduce pain, not least of which is simple quality of life: if someone is in pain constantly, how long before it becomes unbearable? There are known effects on cardiovascular and respiratory systems that can be more serious in someone with EDS, who might have inherent problems in these areas to which the effects of pain are added:

- adrenergic stimulation (sympathetic nervous system);
- hypercoagulation, leading to blood clots;
- increased heart rate;
- increased cardiac output;
- increased myocardial oxygen consumption;
- reduced pulmonary vital capacity;
- reduced alveolar ventilation;
• reduced functional residual capacity;
• arterial hypoxemia;
• suppression of immune functions, predisposing patients to wound infections and sepsis.

There are a multitude of approaches to treating pain in EDS. As addressed in Dr. Francomano’s Comprehensive Pain Management in Ehlers-Danlos Syndrome (http://www.ednf.org/images/2010conference/Handouts/Francomano_ppt_2_slides_per_page.pdf), the approaches are generally:

• reduce muscle spasm;
• myofascial trigger point release;
• integrative medicine approaches;
• pharmacologic therapy;
• surgical approaches.

Realize that pain-free is not possible for almost anyone, whether or not they have EDS. But it is possible to reach lower levels that allow someone with EDS to enjoy life. Try each as needed, and keep what works. Each technique that works is a step closer to pain-free.

The best thing is to find a multi-modal pain doctor, one who uses all the available tools including pharmaceutical to help you. These are outlined in EDNF’s Pain Management Medical Resource Guide available from http://www.ednf.org/index.php?option=com_content&task=view&id=2123&Itemid=88889247; the Multi-Modal Pain Management chart is reprinted at the end of the article (page 26).

There are TENS units. There is massage, specifically myofascial release. Without well-built collagen to minimize the effects of movement, the fascia (and muscle, and anything attached to each joint that hyperextends) can stretch too far and suffer innumerable micro-tears, part of the chronic acute pain that keeps happening over and over. Releasing the tension held in the fascia can allow joints to realign into their natural position. Trigger points are particularly a problem in EDS, like they are in fibromyalgia, and anything that can break up the trigger points to get blood and oxygen flowing through them will help your muscular pain and fatigue. It is possible to get trigger point injections, which may or may not be helpful depending on your own resistance to anesthetic; dry needling can be helpful, too, but not to everyone. Acupuncture helps many.

Gentle chiropractic might work, using activators rather than manipulation. Chiropractic manipulation can be dangerous to hypermobile people; neck manipulation is completely out-of-bounds according to the World Health Organization (World Health Organization, 2005, Guidelines on basic training and safety in chiropractic, Geneva), and should certainly be avoided for joints that are dislocated or prone to subluxation.
There is little evidence that spinal manipulation is an effective treatment for headaches, according to the authors of a new study. ... The authors are concerned about the potential complications associated with spinal manipulation. A 1996 RAND study found that the rate of severe complications—including strokes—was just three for every two million neck manipulations. However, in another study Dr. Ernst found more than 700 unreported severe complications, suggesting that the rate is potentially much higher. ... given the lack of quality evidence supporting spinal manipulation for headaches due to neck strain, “patients should be advised to use other therapies,” Dr. Ernst said.


Chiropractic manipulation of the neck carries the risk of tearing the vertebral artery that leads to the brain, thereby causing stroke or transient ischemic attack. Although the risk is low, it does happen, and physicians and patients should be aware of spinal manipulation therapy as a rare but potential risk factor for stroke. ...

A 2003 study in the American Academy of Neurology's journal *Neurology* confirmed the connection between cervical manipulation and stroke. Wade S. Smith, M.D., Ph.D., director of the Neurovascular Service at the University of California at San Francisco and an AAN member, found that patients under age 60 who had strokes from tears in the vertebral artery were six times more likely to have had spinal manipulative therapy in the 30 days prior to their stroke than patients who had strokes from other causes ... patients and physicians need to realize that a significant increase in neck pain or neurologic symptoms within 30 days following spinal manipulative therapy warrants immediate medical evaluation. Dr. Smith recommends that ... chiropractic practitioners performing spinal manipulative therapy should warn patients of the risk of tearing the vertebral artery. “I encourage patients to discuss with their chiropractor the types of spinal manipulations that are practiced and whether they pose a risk of stroke,” Dr. Smith says.

*Hidden Hercules: Actor Kevin Sorbo reveals his secret battle with stroke* [Childers] [http://journals.lww.com/neurologynow/Fulltext/2011/07050/Hidden_Hercules__Actor_Kevin_Sorbo_reveals_his.15.aspx](http://journals.lww.com/neurologynow/Fulltext/2011/07050/Hidden_Hercules__Actor_Kevin_Sorbo_reveals_his.15.aspx)

Distraction works: find enjoyable activities and focus on them. Meditation is helpful; I personally find Zen and other Buddhist practices particularly effective.

There are also binaural apps and programs that use sound waves to synchronize your brain into particular states similar to meditation; they can induce relaxation, help with pain relief and sleep. One I find successful is Brainwave, which allows you to combine binaural beats with environmental sounds in a wide range of tailored programs ([http://itunes.apple.com/us/app/brain-wave-25-advanced-binaural/id307219387?mt=8](http://itunes.apple.com/us/app/brain-wave-25-advanced-binaural/id307219387?mt=8)); there are others. Brain machines add lights to sound and can help change your brain waves more dramatically and faster than the sound programs alone—some can be found at [http://www.iproducts.ws/mind-machines.htm](http://www.iproducts.ws/mind-machines.htm) and [http://www.mindplace.com/Light-and-Sound](http://www.mindplace.com/Light-and-Sound).

There are biofeedback and hypnosis techniques that also produce similar results.

My doctor suggested psychological counseling because he felt EDS would not be an easy disorder to manage, which proved very helpful to me as a way of discovering it was not all in my head and there were ways of handling my own reactions and my life that made EDS easier to endure.
There are the usual pharmacologic approaches to pain, codified in the WHO escalating therapy for chronic non-cancer pain, starting with non-steroidal anti-inflammatory drugs, then adjuvants and integrative approaches, topical therapies, and tramadol or other opioid agonists. For much more information about these established treatments, see Dr. Howard Levy’s *A Primer on Pain Medications* (http://bit.ly/HQDNwk). These drugs work variably in EDS. Some of us have reactions to NSAIDs that range from ineffectiveness to bleeding.

Local anesthetics can produce unexpected results; normal doses may not work, but once an effective dose is reached, the results may last longer than usual.

- Anesthetic/corticosteroid injections for localized areas of pain and inflammation are often helpful, but cannot be repeated indefinitely; "dry needling" without injection of any material sometimes provides similar benefit.
- Anesthetic nerve blocks can provide temporary relief of neuropathic pain. These are sometimes followed by surgical nerve root destruction and/or implantable stimulators (sensory or motor), with variable results.

Patients suffering from Ehlers-Danlos syndrome type III do not respond to local anaesthetics (http://www.ednf.org/index.php?option=com_content&task=view&id=1337&Itemid=8889208)

Local anaesthetic failure in joint hypermobility syndrome (p. 84, Hakim, Grahame et al.) (http://www.ednf.org/images/stories/pdfs_medical/Local_anaesthetic_failure_in_joint_hypermobility_syndrome.pdf)

There have been studies showing that subcutaneous infiltration of lidocaine has a reduced time of action and that transcutaneous application of a eutectic mixture of lidocaine 2.5% and prilocaine 2.5% (EMLA, AstraZeneca, Södertälje, Sweden) did not produce sufficient analgesia.


Opioids can also have surprising results in EDS patients; what works may be different with each EDS patient, and effective doses may vary as widely in EDS as pain does. When dealing with opioids, it may become important to remember a few distinctions.

*Tolerance is not addiction.* Tolerance is: “The capacity to endure continued subjection to something, esp. a drug; diminution in the body’s response to a drug after continued use” (New Oxford American Dictionary 2007).

*Physical dependence is not addiction.* Physical dependence is: “A state of adaptation that often includes tolerance and is manifested by a drug class specific withdrawal syndrome that can be produced by abrupt cessation…. Physical dependence on and tolerance to prescribed drugs do not constitute sufficient evidence of psychoactive substance use disorder or addiction. They are normal responses that often occur with the persistent use of certain medications” (Definitions related to the medical use of opioids: Evolution towards universal agreement [Savage et al.] I, 2003, 26:655-67).
Addiction is: “When a person has lost control over and continues use even when such use is doing them or others harm. (New Oxford American Dictionary 2007).

The choice in EDS may be as simple as dependence on drugs or dependence on pain.

Pharmacologically, there are alternatives in Lyrica (pregabalin, http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0000327/) and Cymbalta (duloxetine, http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0000274/), both approved by the FDA for musculoskeletal pain, and used successfully by some of us. Lyrica is the frontline for fibromyalgia, and is not an anti-depressant; Cymbalta is an anti-depressant, which can be an added benefit in dealing with the perception of pain. Please read up on these (and any drug) before beginning them; they can cause some serious side effects, and knowing what to look for is a major advantage to minimizing them.


Ensure your vitamin D levels are good by having the blood test done fairly regularly; low vitamin D can make our pain worse, and restoring it to a strong, healthy level may reduce pain. Read more about vitamin D in the Winter 2010 issue of Loose Connections, which you can get free from the archives http://bit.ly/w7aHxc.

American diets are largely deficient in magnesium (http://ods.od.nih.gov/factsheets/magnesium-HealthProfessional/, which lists the Recommended Daily Allowance). Magnesium can be increased through diet or through Epsom salts, which are the best approaches because it is difficult to get too much magnesium in those ways. Usually the kidneys take care of any excess, but if you take supplements for magnesium, it is possible to take too much and get into toxic levels. Magnesium is the fourth most abundant mineral in the human body; a deficiency can result from low blood levels of calcium, which is why the two are frequently recommended together, and malabsorption, which seems to be a fairly common EDS problem (http://www.nlm.nih.gov/medlineplus/ency/article/002423.htm). Magnesium deficiency is implicated in a lot of problems: fatigue, insomnia, muscle spasms, poor memory, and heart/cardiovascular problems including rapid heartbeat. The mineral is particularly recommended with EDS for muscle pain: people who do Epsom salts baths reported their rheumatic pain disappeared. Baths are good for us because magnesium absorbs easily through the skin, and gets directly into the blood and muscles. The hot baths feel good, too. Maximum benefit seems to be two or three times a week with 500-600g Epsom salts each time (http://george-eby-research.com/html/absorption_of_magnesium_sulfate.pdf).

There are other food sources of relief, particularly white tea, rose, and witch hazel (Antioxidant and potential anti-inflammatory activity of extracts and formulations of white tea,
rose, and witch hazel on primary human dermal fibroblast cells [Thring et al.] http://www.journal-inflammation.com/content/8/1/27.)

Nerve blocks are another possibility. “Temporary nerve blocks are achieved by combining a local anesthetic (such as lidocaine) with epinephrine, a steroid (corticosteroid), and/or opioids. Epinephrine produces constriction of the blood vessels which delays the diffusion of the anesthetic. Steroids can help to reduce inflammation. Opioids are painkillers. Injection nerve blocks can be either single treatments, multiple injections over a period of time, or continuous infusions. A continuous peripheral nerve block can be introduced into a limb undergoing surgery, for example, a femoral nerve block to prevent pain in knee replacement. Permanent nerve block can be effected using other drugs or methods including alcohol or phenol to selectively destroy nerve tissue, cryoanalgesia to freeze nerves, and radio frequency ablation to destroy nerve tissue using heat. Nerve blocks are sterile procedures that are usually performed in an outpatient facility or hospital. The procedure can be performed with the help of ultrasound, fluoroscopy (a live X-ray) or CT. Use of any one of these imaging modalities enables the physician to view the placement of the needle. A probe positioning system can be used to hold the ultrasound transducer steady. In addition, electrical stimulation can provide feedback on the proximity of the needle to the target nerve.” (http://en.wikipedia.org/wiki/Nerve_block)

As a last resort, nerve endings can be severed surgically (http://seniorhealth.about.com/library/conditions/blchronicpain7.htm).

A new area that might prove helpful is low-level laser therapy (LLLT), a medical treatment using low-level lasers or light-emitting diodes to reportedly affect cellular function. “LLLT is controversial in mainstream medicine with ongoing research to determine the ideal location of treatment (specifically whether LLLT is more appropriately used over nerves versus joints), dose, wavelength, timing, pulsing and duration. The effects of LLLT appear to be limited to a specified set of wavelengths of laser, and administering LLLT below the dose range does not appear to be effective. Despite a lack of consensus over its ideal use, specific test and protocols for LLLT suggest it is effective in relieving short-term pain for rheumatoid arthritis, osteoarthritis, acute and chronic neck pain, tendinopathy, and possibly chronic joint disorders. The evidence for LLLT being useful in the treatment of low back pain, dentistry and wound healing is equivocal.” (http://en.wikipedia.org/wiki/Low_level_laser_therapy, also Efficacy of low-level laser therapy in the management of neck pain: a systematic review and meta-analysis of randomised placebo or active-treatment controlled trials http://www.ncbi.nlm.nih.gov/pubmed/19913903 and A systematic review of low level laser therapy with location-specific doses for pain from chronic joint disorders http://www.ncbi.nlm.nih.gov/pubmed/12775206).

Do not ignore the role of sleep; good sleep should be restorative and help reduce pain levels, but in EDS, that frequently is not what happens. Read Sleep Disorders in the Hypermobility Syndromes [Pocinki] available from http://bit.ly/HQDNwk.
A particularly useful resource in general is *The Intractable Pain Patient’s Handbook for Survival* [Tennant](http://pain-topics.org/pdf/IntractablePainSurvival.pdf).

It still surprises me to hear of doctors claiming EDS has no associated pain. In fact, chronic pain is a **minor diagnostic criterion** for EDS Hypermobile Type. How could the effects of hypermobility *not* be painful? EDS is a collagen disorder, not a nervous system disorder; it does not have some neurological effect that negates pain.

Pain treatment may be the single most important assistance that can be offered to someone living with EDS, and no one technique will work. But each one that works adds to the total reduction in pain. If one can find small reductions here and there, the combined effect can be significant reduction in overall pain levels. A life of out-of-control pain that seemed daunting and insufferable can become one filled with joy and accomplishment. It simply takes someone willing to listen and help.

**Mark C. Martino**

Editor-in-Chief

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**Approaches to pain management in EDS and other hypermobility syndromes**


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**REDUCE MUSCLE SPASM**

- Physical therapy to reduce muscle spasm
- Physical therapy to tone and strengthen muscles around lax joints
- Magnesium
- Muscle relaxants

**Physical Therapy to reduce muscle spasm**

- Passive therapy
- Deep heat
- Ultrasound
- Massage
- Transcutaneous Electrical Nerve Stimulation (TENS)

**Physical Therapy to tone and strengthen muscles**

- Active therapy
- Warm water therapy is often most helpful initially
- Tone and strengthen muscles around lax joints
- Tone and strengthen core muscles (neck, back, abdominal muscles)
- Pilates
- Alexander technique
- Feldenkrais technique

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**Magnesium**

- Crucial element for muscle relaxation
- Usual source is fruits and vegetables
- Soil has become depleted of magnesium in many parts of the US
- The body maintains blood levels of magnesium at the cost of intracellular depletion
- Too much oral magnesium is disturbing to the GI system (abdominal pain, cramping, diarrhea)
- Magnesium is well absorbed through the skin
- Repleting magnesium
  - Epsom salts are magnesium salts
  - Epsom salt baths (2 cups of salts in a bath of warm water, soak for 10–15 minutes)
  - Epsom salt foot baths (1 cup salts in a tub, soak feet for 10–15 minutes)
  - Magnesium lotion (www.selfhealthsystems.com); use 2 tsp daily

**Trigger point therapy**

- Dry needling of myofascial trigger points
- Injection of local anesthetic (lidocaine)
- Massage of trigger points
- *Trigger Point Therapy Workbook* – Claire Davies
- Myofascial trigger point therapy
Importance of staying active

- Exercise prescription
  - 20–30 minutes daily
  - moderate intensity
  - low-impact exercise
  - 5 days a week
- Exercise
  - Reduces stress
  - Lowers blood pressure
  - Improves circulation
  - Releases endorphins

Mind/Body Skills and Headache

- Hypnosis vs. Propanolol for Migraine
  - Prospective crossover-hypnosis, placebo and propanolol
  - Significant decrease in frequency of HA with self-hypnosis group only
  - Olness & MacDonald, 1987, Pediatrics
- Biofeedback for TT and Migraine HA
  - SEMG with bifrontal placement
  - Peripheral temperature biofeedback
  - Heart Rate Variability Biofeedback
  - Neurofeedback
  - Andrasik & Schwartz, 2006, Behavior Modification

Mind/Body Skills and Headache

- Adjunctive CAM Therapies (safety but unclear efficacy)
  - Massage, Aromatherapy

Myofascial Trigger Point Release

INTEGRATIVE MEDICINE APPROACHES

- Mind-body medicine
  - Mindfulness based stress reduction
  - Relaxation techniques
  - Yoga
  - Hypnosis
  - Autogenic training
  - Biofeedback – particularly helpful for Raynaud’s, migraine
- Acupuncture
  - Immediate relief from pain
  - Lasts 2–4 days
  - “Better than morphine”
  - Weaned self off of narcotics
  - Acupuncture analgesia – Opioid involvement
  - Naloxone blocks acupuncture analgesia
  - Patients with fewer opioid receptors experience less acupuncture analgesia
  - Endorphins increase in CSF with acupuncture
  - Can provide acupuncture analgesia with cross circulation in animal studies
- Integrative Approach to Headache
  - Adjust all lifestyle factors
  - Sleep, diet, overscheduling, exercise
  - Review medications
  - Analgesic rebound, polypharmacy
- Primary CAM Therapies (safety and efficacy)
  - Mind/Body, Acupuncture, Psychotherapy

“Bad things do happen; how I respond to them defines my character and the quality of my life. I can choose to sit in perpetual sadness, immobilized by the gravity of my loss, or I can choose to rise from the pain and treasure the most precious gift I have: life itself.”

Walter Anderson
Multi-Modal Pain Management for EDS
reprinted from Pain Management Medical Resource Guide
http://www.ednf.org/index.php?option=com_content&task=view&id=2123&Itemid=88889247

INFREQUENT PAIN EVENTS
(less than four days per week)

- **ANALGESICS**
  May be ineffective as single agents or require excessive doses
  ↓ if needed
  Short-acting opioids
  Muscle relaxants

- **PHYSICAL THERAPY**
  Reconditioning
  Exercise (gradual toning for stability, not strength\(^1\))
  Flare-up management by:
  - Distraction techniques
  - Trigger-point/myofascial therapy
  - Heat/cold application
  - Ultrasound/electrical stimulation

- **PSYCHOLOGICAL THERAPY**
  Relaxation
  Stress management (incl. Mindfulness-Based Stress Reduction)
  Group therapy
  Cognitive restructuring (incl. hypnosis)

- **PSYCHIATRIC THERAPY**
  Medication for mood/pain
  Individual counseling

- **SLEEP THERAPY**
  Sleep disturbance is common in EDS (contributes to poor pain recovery; incl. apnea)
  Tricyclic antidepressants (TCAs)

- **COMPLEMENTARY/ALTERNATIVE**
  Meditation
  Acupuncture
  Massage
  Yoga

- **MAINTAIN GENERAL HEALTH**
  Avoid weight extremes & smoking\(^2\)

CHRONIC PAIN
(constant & disturbing pain)

- **MANAGE ACUTE EVENTS**
  Treat acute events quickly and aggressively to defend against central neuronal plasticity (“learned” chronic pain)
  ↓ if needed
  Short-acting opioids (breakthrough pain)
  Long-acting opioids (baseline pain; see at bottom)

- **PREVENT RECURRING INJURY**
  Hypermobile joints and fragile connective tissues endure chronic acute injury
  Bracing may be helpful (maintain toning exercise)
  Orthopedic surgery should be avoided
  Almost never provides long-term improvement
  Added risk of soft tissue fragility and poor wound healing, esp. Classical & Vascular EDS

- **OCCUPATIONAL THERAPY**
  Delay disability and encourage activity as long as possible
  Body mechanics/ergonomics
  Intervention in workplace and work simplification
  Pacing skills

- **NEUROPATHIC PAIN**
  Common in EDS, also called neuralgia (different from usual nociceptive pain); described as numbness, tingling, burning, electrical, hot/cold
  Incl. disease/dysfunction of peripheral nerves (numbness, weakness), nerve injury, axonal loss
  Antidepressants (TCAs/SNRIs)
  Antiseizure medications
  Topicals (capsaicin)

- **CONTRIBUTING PROBLEMS**
  Additional genetic factors/disabilities
  Side effects from treatments, incl. overuse or ineffectiveness of analgesics or short-acting opioids

- **LONG-ACTING OPIOIDS** (methadone, morphine, oxycodone, fentanyl; oral/transdermal/pump)
  Tolerance for pain-killing builds up readily, but not to side effects incl. GI dysmotility & reduced consciousness

- **SURGICAL PAIN INTERVENTION**
  Injection (trigger point, nerve root)
  Implantable nerve stimulators
  Medication pumps (subarachnoid cavity)

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\(^{1}\)The key is slow improvement in muscle tone, **not strength**. Tone is the degree of muscle contraction at rest; strength is voluntary force that can be invoked at will. Build tone with non-resistance exercise that gradually increases repetitions. Avoid hyperextension, resistance and impact. Elastic bands may make things worse because they combine increased resistance with joint hyperextension. Toning is a life-long strategy; if exercises are stopped, pain usually recurs. (Howard Levy, MD, PhD)

\(^{2}\)Quit smoking (pain, depression and addiction are all modulated by the same nicotinic and muscarinic receptors). Maintain normal weight; obesity or thinness seem to present problems. (Mark Lavallee, MD, CSCS, FACSM)
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