

EHLERS-DANLOS SYNDROME

Collagen is the most abundant protein in the human body.

It provides the structural strength in most human tissue, including the heart and blood vessels, eyes and skin, cartilage and bone.

What happens when this basic building block is flawed?

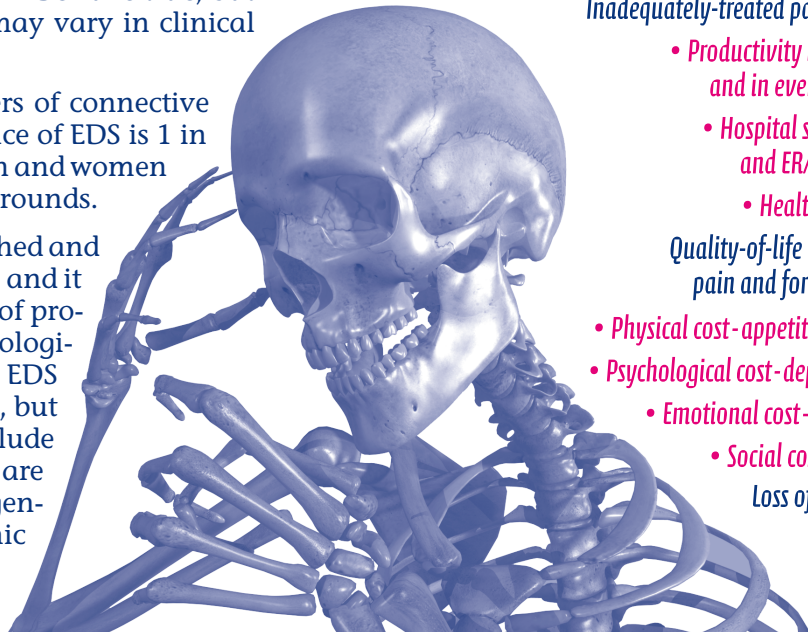
When muscles, ligaments, tendons and even large organs are built with structurally defective collagen there is systemic weakness and instability evident throughout the body.

There is Ehlers-Danlos Syndrome.

Excessively mobile joints, chronic pain and skin softness characterize Ehlers-Danlos syndrome (EDS). At least six types of EDS have been identified; clinical manifestations vary according to type and may also include poor wound healing with atrophic scars, easy bruising, chronic pain and generalized connective tissue fragility. Each type is thought to involve a unique defect in connective tissue, although not all of the genes responsible for causing EDS have been found. Within each family the type of EDS runs true, but individual family members may vary in clinical severity and manifestations.

As a group of genetic disorders of connective tissue, the estimated prevalence of EDS is 1 in 5,000. It is known to affect men and women of all racial and ethnic backgrounds.

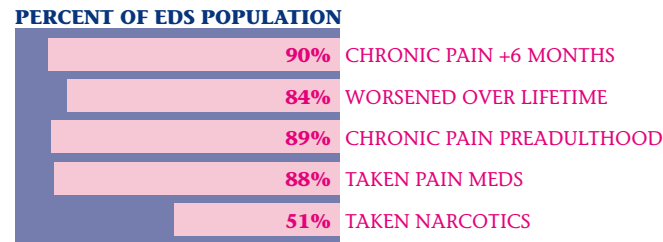
Chronic pain is a well-established and cardinal manifestation of EDS and it is common for pain to be out of proportion to physical and radiological findings. The etiology of EDS pain is not clearly understood, but some of the likely causes include muscle spasm (tender points are sometimes present) and degenerative arthritis; neuropathic pain is also common.



EDS AND PAIN MANAGEMENT

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 or injury to start the pain cycle. 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.

Probably the most important element of a successful therapeutic intervention for EDS is bringing that pain under control. **When a person with EDS reports pain, even if out of proportion to physical findings, they should be listened to and believed.**



STATISTICS FROM *Chronic Pain is a Manifestation of the Ehlers-Danlos Syndrome* (Sacheti et al, 1997); full text available at ednf.org

WE ALL PAY WHEN PAIN IS UNTREATED

Inadequately-treated pain affects everyone:

- Productivity is decreased at work and in everyday tasks at home
- Hospital stays are lengthened and ER/clinic visits increase
- Health care costs increase

Quality-of-life worsens for those in pain and for those around them:

- Physical cost - appetite loss and weakness
- Psychological cost - depression and anxiety
- Emotional cost - suffering and anger
- Social cost - lost relationships

Loss of independence, self-worth and financial stability can be an ultimate price

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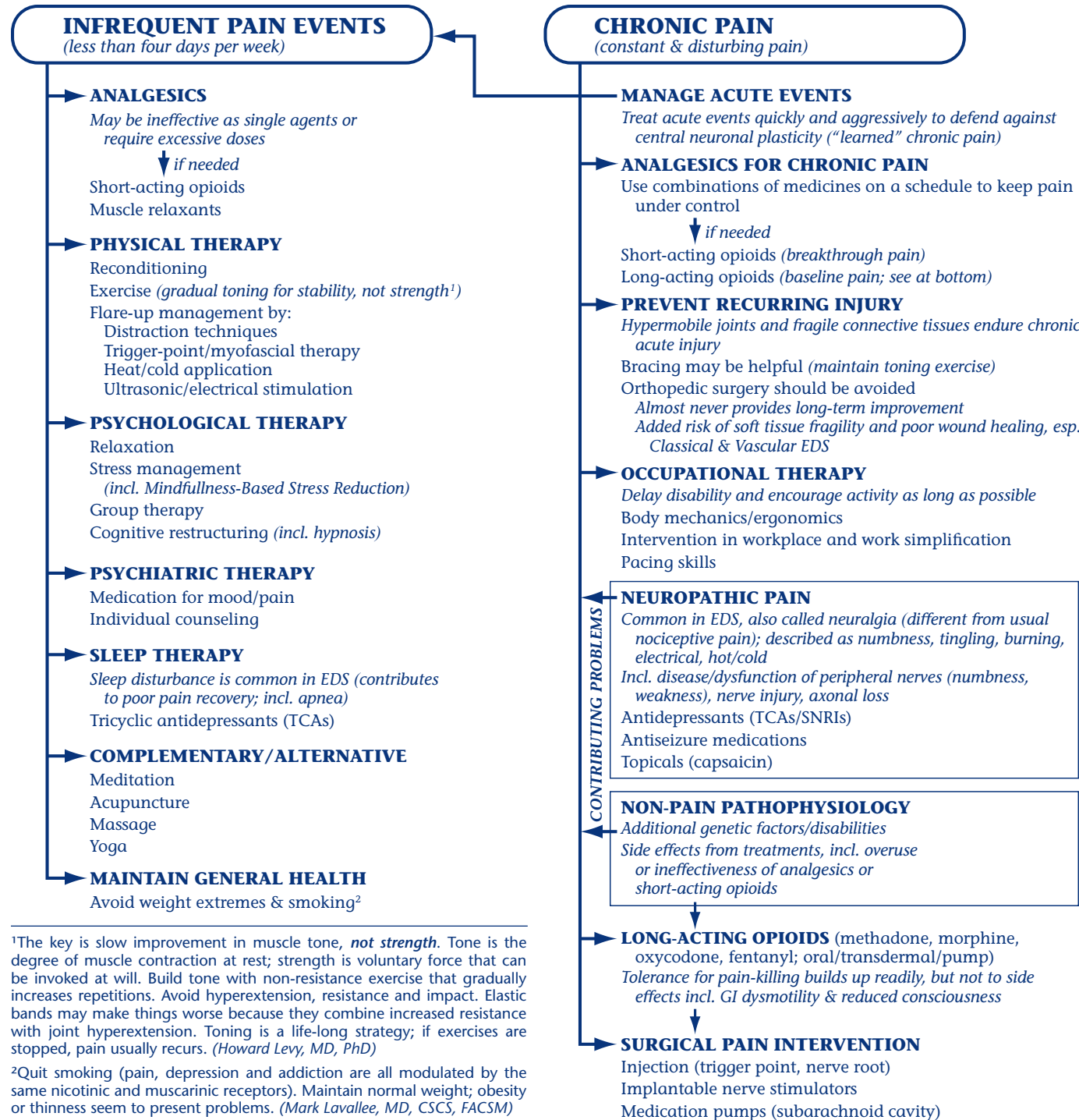
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PAIN MANAGEMENT

Medical Resource Guide



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¹The 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)

²Quit 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 Lavalley, MD, CSCS, FACSM)

Tolerance is not addiction.

tol·er·ance |'täl(ə)rəns|
The capacity to endure continued subjection to something, esp. a drug; diminution in the body's response to a drug after continued use. (From New Oxford American Dictionary 2007)

Physical dependence is not addiction.

phys·i·cal de·pend·ence |'fizikəl di'pendəns|
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. (From "Definitions related to the medical use of opioids: Evolution towards universal agreement"; Savage et al., Journal of Pain and Symptom Management, 2003, 26:655-67)

The choice may be simple: dependence on drugs or dependence on pain.

ad·dic·tion |ə'dik sh ən|
The fact or condition of being addicted to a particular substance, thing, or activity, when a person has lost control over and continues use even when such use is doing them or others harm. (From New Oxford American Dictionary 2007)

The chance of addiction is very low when these medications are taken as directed by a doctor and used for pain.

URGENT INFORMATION on Vascular EDS

ARTERIAL RUPTURE IS THE MOST COMMON CAUSE OF SUDDEN DEATH.

- Arterial or intestinal rupture commonly presents as acute abdominal or flank pain that can be diffuse or localized.
- Spontaneous arterial rupture is most likely to occur in a person's twenties or thirties, but can occur at any point in life.
- Cerebral arterial rupture may present with altered mental status and be mistaken for drug overdose.
- Mid-size arteries are commonly involved.

Arterial, intestinal, or uterine fragility or rupture usually arise in EDS Vascular type, but should be investigated for any EDS type.



Pain Management Medical Resource Guide brings you highlights of information found at ednf.org, available whenever and however you need it.

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