

Connecting the dots in Ehlers Danlos Syndrome

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Disclosure

- ▶ Director, Center for Complex Conditions
- ▶ Trained at the Harvard Medical School
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- ▶ Medical Advisory Board of multiple non profit organizations
- ▶ No financial disclosure



Disclaimer


- ▶ I have no actual or potential conflict of interest in relation to this presentation or program
- ▶ This presentation will discuss “off-label” uses of medications
- ▶ Discussions in this presentation are for a general information purposes only. Please discuss with your physician your own particular treatment. This presentation or discussion is NOT meant to take the place of your doctor.



Ehlers Danlos Syndromes

➤ Important things to remember:

1. Weak connective tissue
2. Poor joint position sense (proprioception)
3. Co-existing medical conditions – Dysautonomia, Mast Cell Activation syndrome, headaches, spine problems



It is important to understand the cause of the pain before deciding on treatment options.

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To manage pain first find out what is the cause of the pain

- Find out what's broken
- For example, pain in the shoulder joint can be from:
 - Dislocated shoulder joint,
 - Muscle spasms
 - Nerve or blood vessel impingement or damage
 - All the above
- The treatment of each of these is different

Principles of managing pain in EDS

- ▶ Use a mix of treatments. There is no one simple solution
- ▶ For example - Knee pain due to instability
 - ▶ Stabilize knee with braces
 - ▶ Strengthen muscles around the knee
 - ▶ Medications – NSAID's, topical etc.
- ▶ Follow the 10% rule






Head pain

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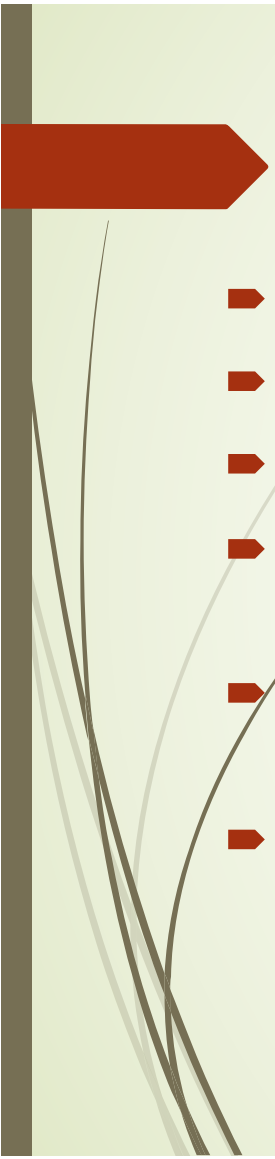
Common causes of headaches in EDS

1. Migraines
2. Chiari malformation
3. Cervicogenic Headaches – from muscles
4. Temporo Mandibular joint dysfunction (Craniofacial pain)
5. POTS / Dysautonomia
6. Tethered Cord syndrome (TCF)
7. Spontaneous CSF (Cerebrospinal) leak
8. Cranio Cervical Instability (Instability of the neck and head)
9. Idiopathic Intracranial Hypertension

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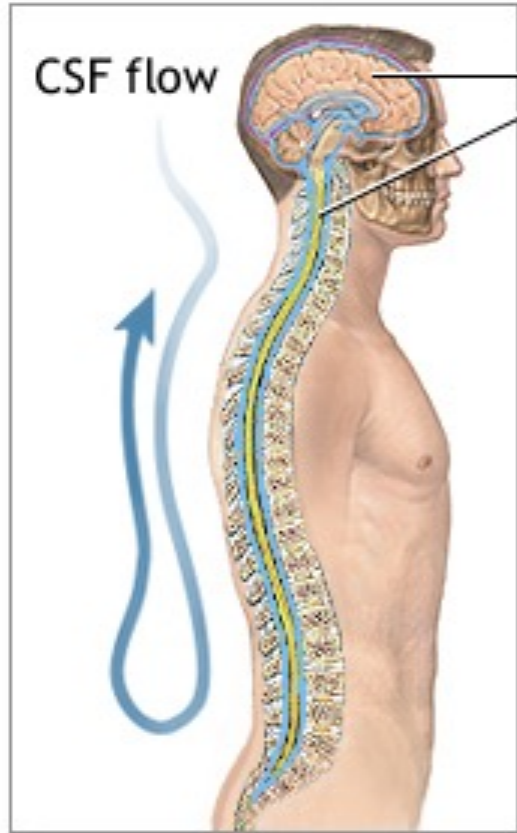


“My whole head hurts, I see double, I can feel throbbing in my ears, it increases when I laugh or cough ”




Increased Intracranial Hypertension (IIH) – increased pressure inside the head

- ▶ Raised pressure inside the head from different reasons
- ▶ double vision
- ▶ Ringing in the ears which pulses (pulsatile tinnitus)
- ▶ Maybe because of narrowing of blood flow (venous sinus stenosis) or Chiari Malformation
- ▶ Diagnosis: ~~spinal tap~~, eye exam, MR venography, Upright MRI of neck for Chiari, Bolt
- ▶ Treatment: medicines to decrease fluid pressure in the head, VP shunt to drain excess fluid, stent, surgery for Chiari



Central nervous

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“My headache gets worse when I stand, and it almost goes away when I lie down”

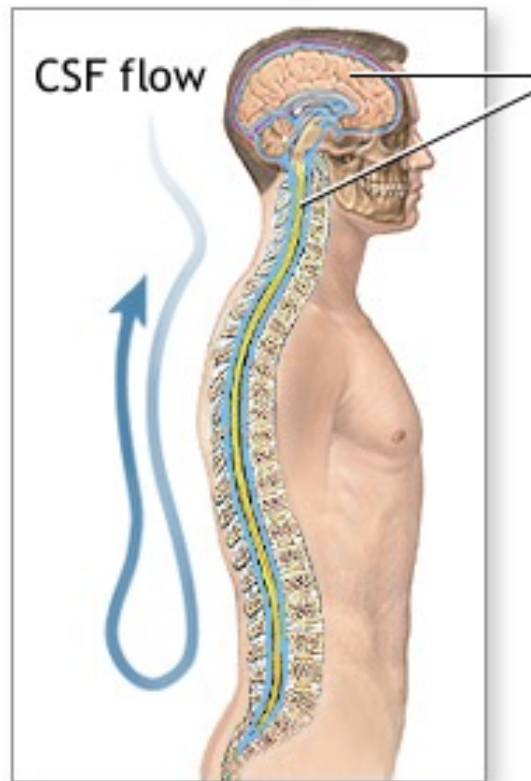
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CSF leak Headaches

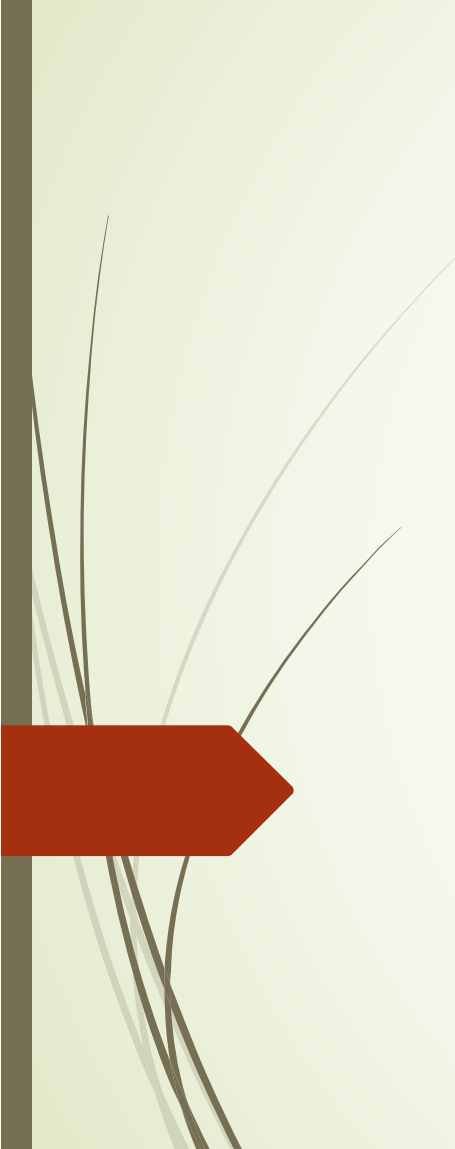
- ▶ The headache is very intense and starts as soon as the person stands up.
- ▶ It resolves fully with lying flat.
- ▶ The headaches are usually in the back of the head

Spontaneous CSF leak



Treatment of CSF leak headaches

- Increase oral fluids
- Caffeine
- Corset or abdominal binder
- Epidural blood patch
- Epidural fibrin glue
- Surgical repair (if after surgery)



“My headache gets worse when I cough, I have tingling in my hands and feet, I have difficulty swallowing”



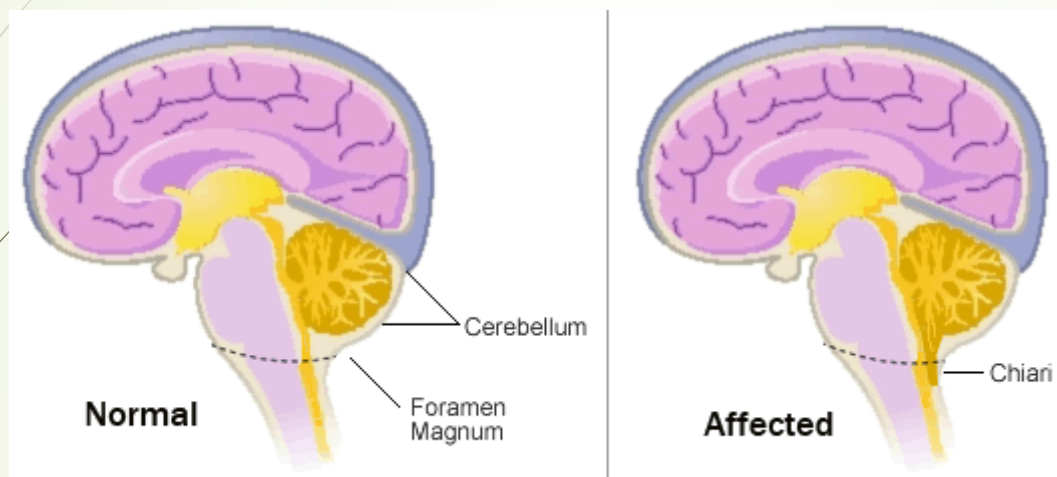
Chiari Malformation

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Chiari malformation



Small posterior fossa pushes the brainstem and cerebellar tonsils through the foramen magnum into the spinal canal

<http://www.craniofacial.vcu.edu/conditions/chiari.html>

Symptoms of Chiari Malformation

➤ COMMON FEATURES

- Headaches made worse by coughing or straining
- Neck pain
- Balance problems
- Numbness or paresthesia's to arms or legs
- Dizziness
- Difficulty swallowing
- Poor Hand co-ordination
- Ringing or buzzing in the ears
- Hearing loss
- Nausea, vomiting
- Pressure headaches in the back of the head (suboccipital)
- Muscle weakness
- Vision problems



Chiari Malformation and EDS

A diagnosis of Chiari Malformation depends on:

1. Good history and physical examination
2. Upright MRI of the neck in patients with EDS



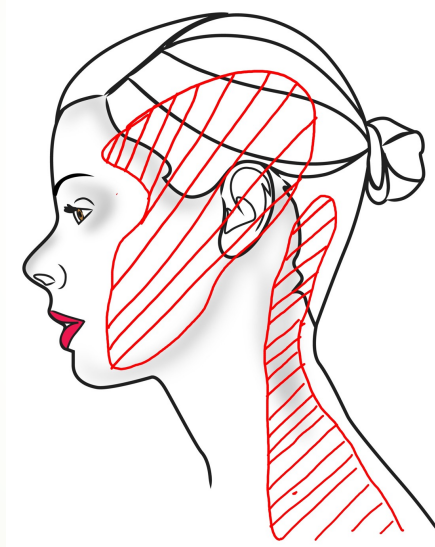
Temporo Mandibular Joint Dysfunction

TMJ Pain

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Temporo-Mandibular joint pain



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Temporo-Mandibular joint pain

- Gentle massage to the muscles
- Trigger point injections
- Botox injections



Upper back pain

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Upper back pain in EDS

- Shoulder instability
- Rib subluxation
- PoTS “coat hanger distribution”
- In women, it can be the weight of the breast tissue



Upper back pain in EDS - treatment

- Correcting PoTS
- Sports bra with racer back
- Compressive vest to support the spine

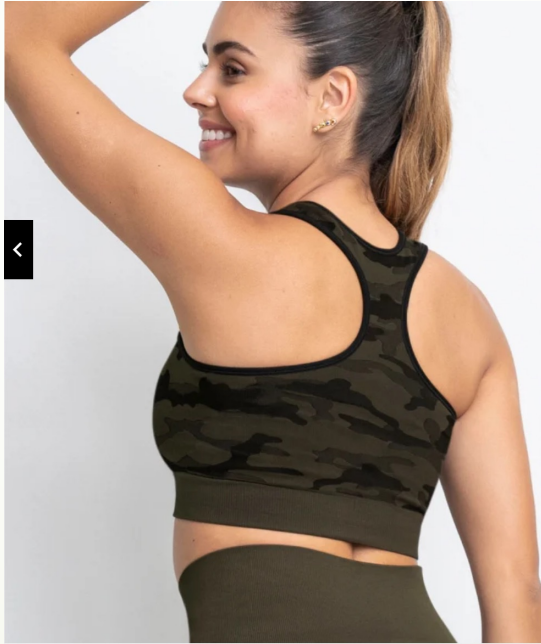
Compression garments to improve proprioception



www.dmorthotics.com

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Leonisa and CW-X



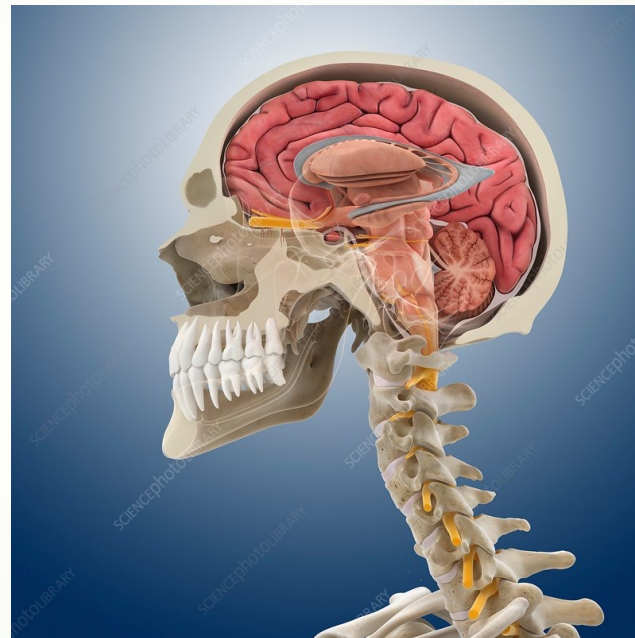


Neck pain

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Reasons for neck pain in EDS



Neck pain

- The head is supported and held in place on the neck by ligaments and muscles.
- In EDS, the ligaments may be lax or loose.
- As a result, the muscles in the neck must work hard to hold up the head.
- Patients often find themselves sitting with their chin supported in their hands
- A supportive neck collar will help

https://www.123rf.com/photo_13121454_portrait-of-smiling-woman-resting-her-chin-on-hand.html



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Cranio Cervical instability in EDS

- ▶ The head and neck are stabilized by ligaments
- ▶ In EDS, the ligaments are lax (like elastic bands instead of rope)
- ▶ Because of lax ligaments in the head and neck, the skull settles down on the spine – cranial settling
- ▶ This cranial settling may also push the lower part of the brain through the hole at the bottom of the skull – Chiari malformation.




Imaging for Cranio Cervical Instability

- Need functional imaging technology
- Static pictures are not helpful
- Digital motion x-ray (DMX)
- Functional computerized tomography (fCT scan)
 - **Flexion.**
 - **Rotate neck left 90 degrees.**
 - **Rotate neck right 90 degrees.**

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Cranio Cervical Instability - management

- ▶ Mild to moderate:
 - ▶ Neck muscles strengthening exercises
 - ▶ Hard cervical collar (Vista Aspen collar)
- ▶ Severe Instability:
 - ▶ Surgical fusion

Cervical fusion

- ▶ There are 2 thoughts on how Cervical fusion is done
- ▶ The best technique is to place a traction on the head (invasive cranial traction) and find out at what position is the best result
- ▶ During the surgery, the same traction weight is applied, and the neck is fused in the best fusion
- ▶ The other method is to look at CT scan and MRI and fuse the spine in whatever position it is.



Vista® MultiPost Therapy Collar – an improved design



Miami J collar



Cervical collar – what does it do?

- One of the problem of having ligament laxity in the neck was cranial settling
- The neck brace takes the load of the head off the spine.
- No, it does not weaken the neck muscles.
- The neck brace does not prevent your neck muscles from moving

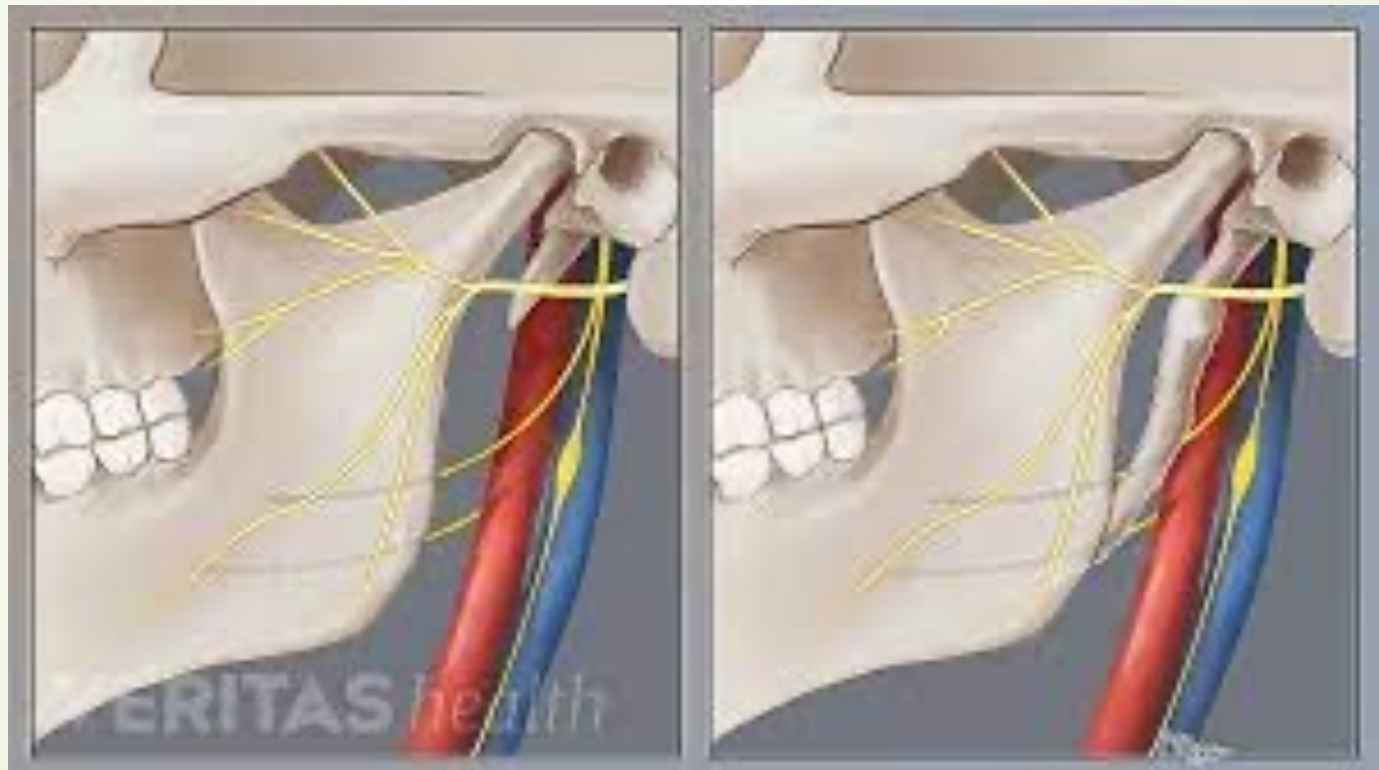
Prolotherapy

- ▶ Not a good idea in EDS
- ▶ Irritating substances like glucose is injected in the tissue.
- ▶ This causes scarring
- ▶ Unfortunately, in EDS, the scar tissue formed is weaker than the original tissue.
- ▶ Repeated studies have shown that there is no evidence that it is effective
- ▶ The benefits are temporary.

Facial pain

- ▶ Eagle syndrome
- ▶ Often misdiagnosed as Trigeminal Neuralgia
- ▶ Sharp nerve like pain in the jaw, back of the throat and base of tongue
- ▶ Pain worsened by swallowing, moving the jaw or turning the neck

Eagle Syndrome



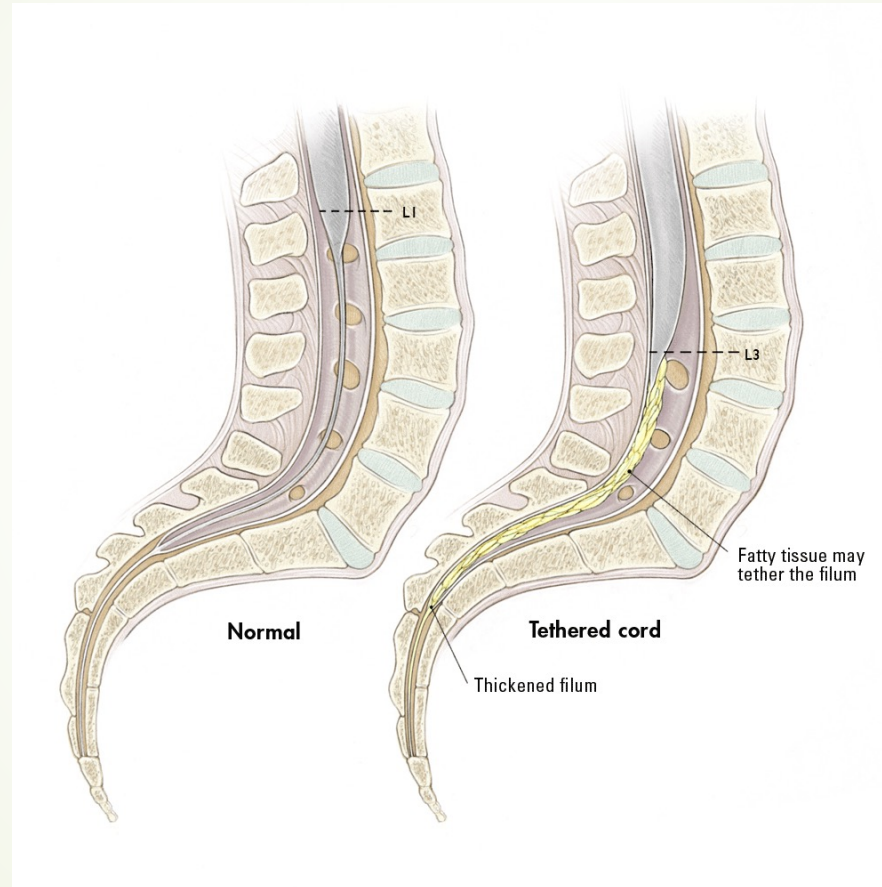


Numbing medicine injections

- ▶ Commonly used numbing medicine injections (Lidocaine) may be ineffective
- ▶ Marcaine or Carbocaine work well in EDS

Tethered Cord Syndrome

Tethered Cord Syndrome (TCS)



<http://javid1985.hubpages.com/hub/Tethered-Spinal-Cord-Syndrome-What-You-Need-To-Know>

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Clinical symptoms

- ▶ Low back pain
- ▶ Neurogenic bladder
- ▶ Leg weakness and sensory loss
- ▶ Diffuse pain in both legs



Tethered Cord syndrome

- Aching, burning pain in low back, legs and feet.
- Weakness in legs
- Legs – heaviness, stiffness, tightness, cramps
- Tingling or numbness in pelvic area or legs
- History of toe walking

Neurogenic bladder

- ▶ **Bladder symptoms (Neurogenic bladder)**
- ▶ Urinary hesitancy
- ▶ Increased (decreased) frequency,
- ▶ Urgency
- ▶ Sense of incomplete emptying of the bladder
- ▶ Incontinence

Tethered cord syndrome and Chiari Malformation

- ▶ People with EDS who have Chiari Malformation should also be checked for Tethered Cord Syndrome

Tethered Cord Syndrome and EDS

- ▶ MRI is NOT a useful tool for diagnosing TCS
- ▶ MRI may be done to rule out other problems
- ▶ Diagnosis is based on clinical history and examination
- ▶ A urodynamic study maybe helpful in case of urinary symptoms / neurogenic bladder

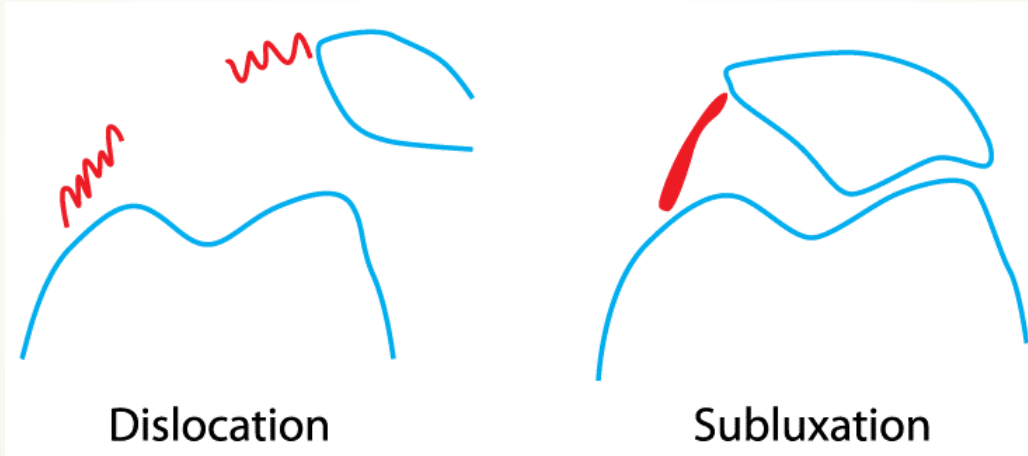


Subluxations and Dislocations

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Subluxations and Dislocations



Pain in subluxation and Dislocations

- ▶ When a joint subluxes or dislocates, the pain is usually from muscle spasms around the joint.
- ▶ Pain from capsular stretch
- ▶ Not as much from the bones



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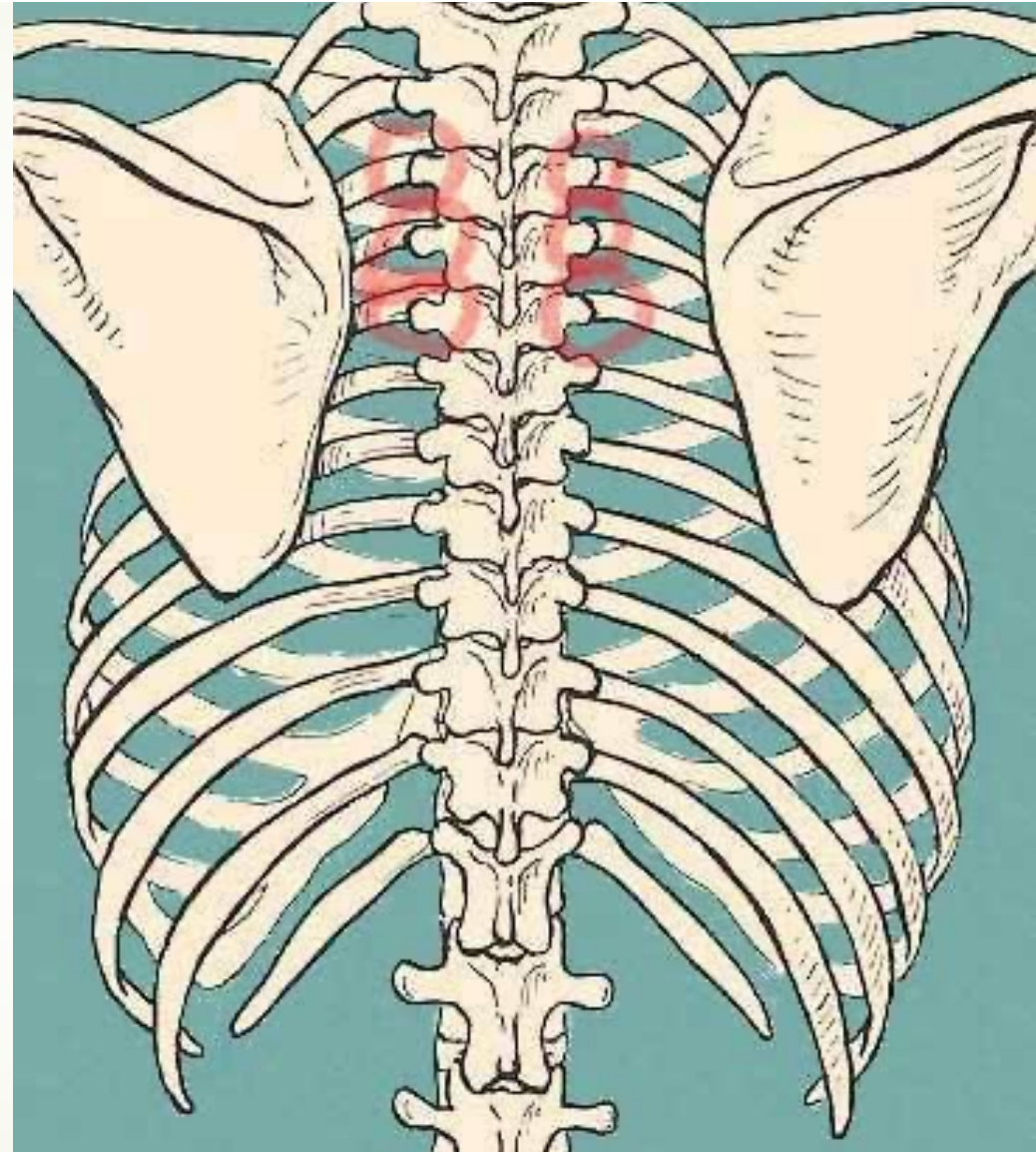
Rib pain

Rib pain

- ▶ Rib subluxations
- ▶ Costo chondritis

Ribs and thoracic spine

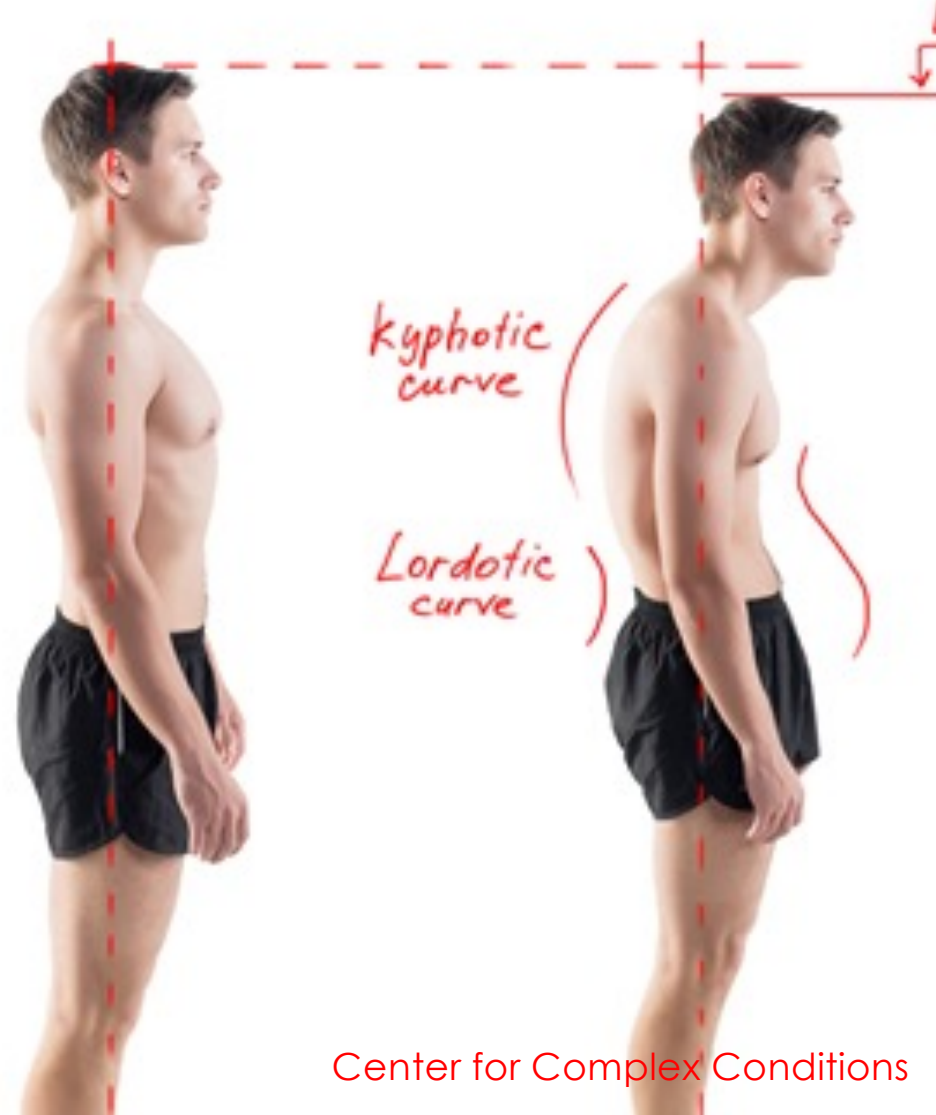
- ▶ Ribs are connected to the thoracic spine by joints



Rib subluxations

- ▶ With slouched posture the ribs tend to pop out of place
- ▶ In EDS having a slouched over posture is easy because of ligament laxity

<https://nationalallergyandinjuryclinic.com/how-posture-affects-your-health/>



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Using a Roller for rib subluxations



<https://www.ptandsr.com/foam-rollers-back-spine-relief/>

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Cat stretch



DM Orthotic vests



www.dmorthotics.com

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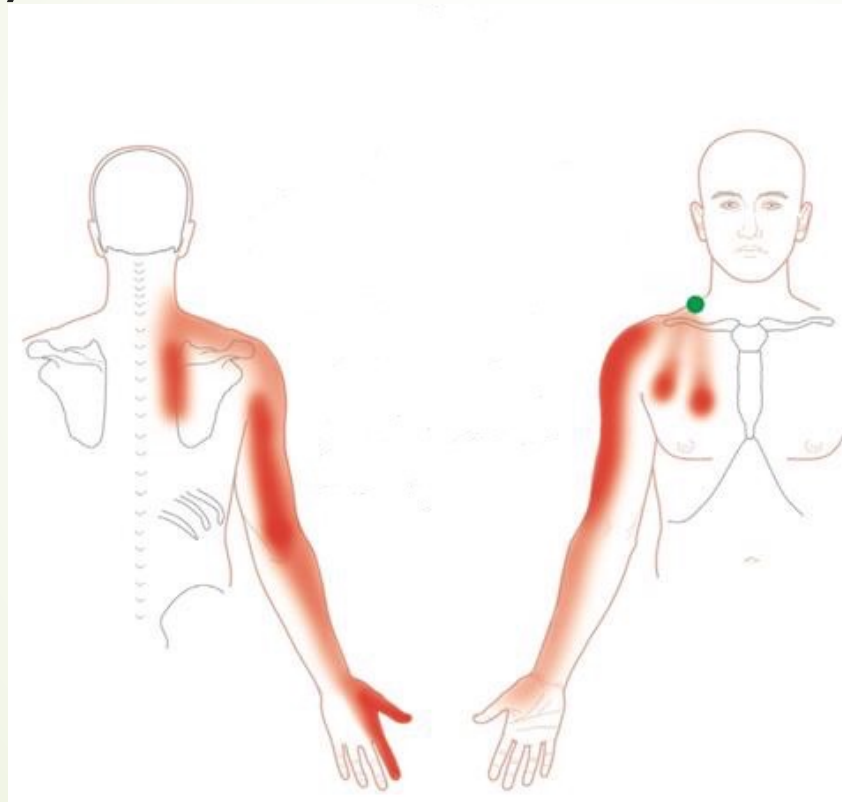


Shoulder pain

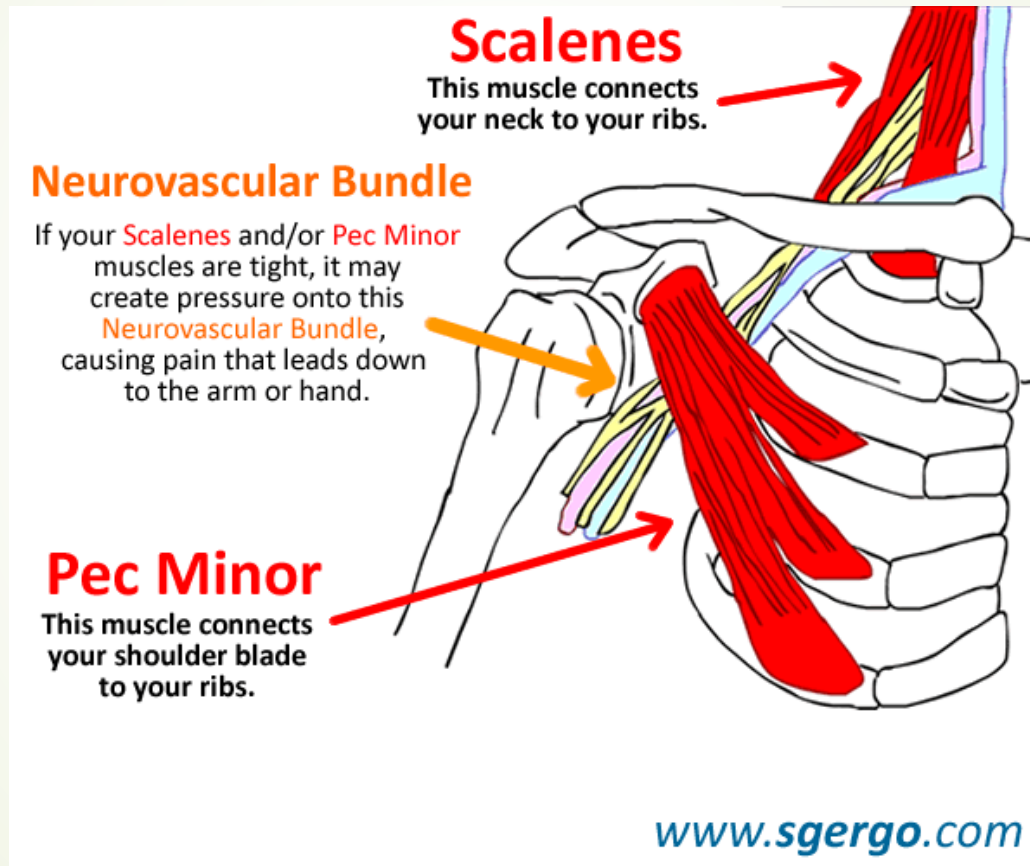
- ▶ The shoulder joint is one of the most unstable joint. It has very few muscles around it to support it (as compared to the hip joint)
- ▶ Laxity of the shoulder joint causes the muscles (rotator cuff) around the shoulder to spasm
- ▶ Thoracic Outlet syndrome.

Arm pain in EDS

Pain patterns in Thoracic Outlet syndrome



Thoracic Outlet Syndrome

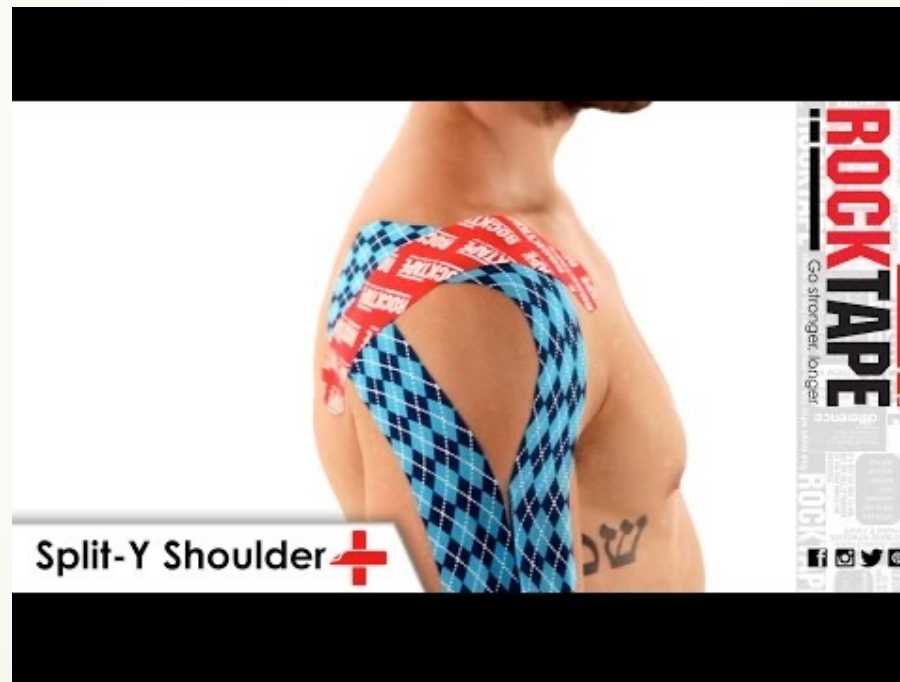




Thoracic Outlet Syndrome

- ▶ Kinesio taping
- ▶ Botox injections
- ▶ Surgical correction, may need to consider doing a shoulder stabilization surgery also (Neer Inferior Capsular Shift)

Neuromuscular Taping for shoulder joint pain



Joint Position sense

Proprioception

The body's ability to sense movement of the joints and their position

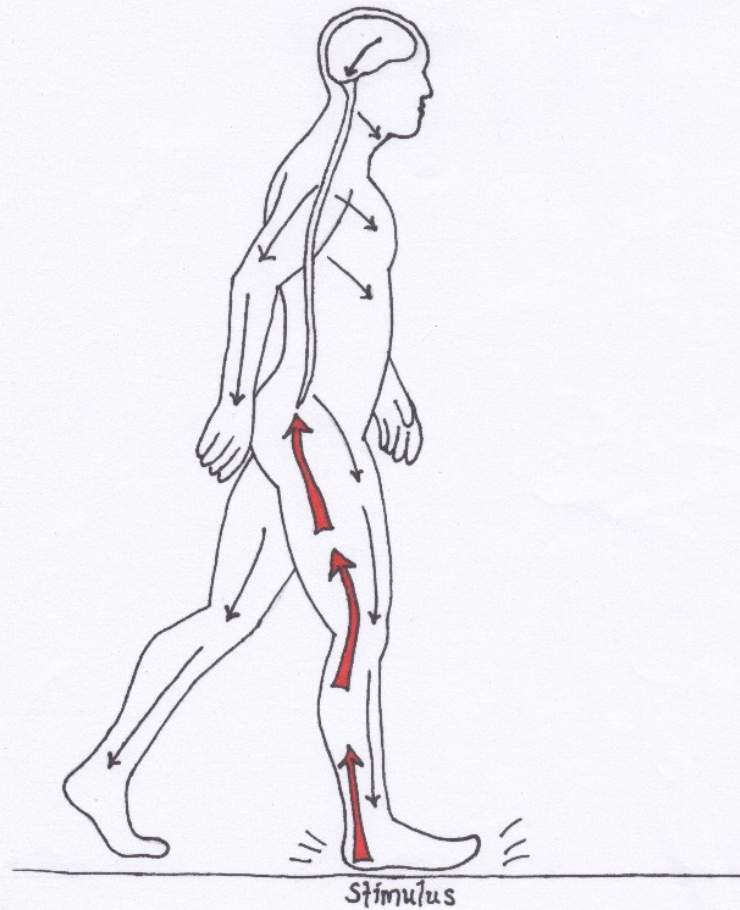
Proprioception (Joint Position Sense)

- There are sensors in our joints, tendons, ligaments, muscles and skin that send a message to our brain as to the exact position of the joint
- The brain in turn sends signals back to manage our body

Proprioception – Joint sense

- ▶ The brain constantly gets information from the joints as to the exact position of the limbs in space.
- ▶ It helps us walk, use our arms, maintain our posture without tipping over.
- ▶ Protects our joints from overextending and our muscles from over stretching
- ▶ EDS – poor proprioception.

Proprioception – Joint sense



Made-to-Measure



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CW-X



Shoulders and rib subluxations - Leonisa



Hand and wrist pain

- ▶ Because of poor proprioception people with EDS, the fingers do not send enough signals to the brain
- ▶ As a result, they drop things because the brain does not realize they are holding something
- ▶ In some cases, they hold things too tightly (pen, cooking).
- ▶ Their fingers are bendy so they must use even more muscle strength to hold things
- ▶ They press down hard when they write
- ▶ All of these cause pain in their hands and wrists

Fingerless compression gloves



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Ring splints to stabilize the joints



Where to get your ring splints

➤ <https://www.zebrasplints.org>

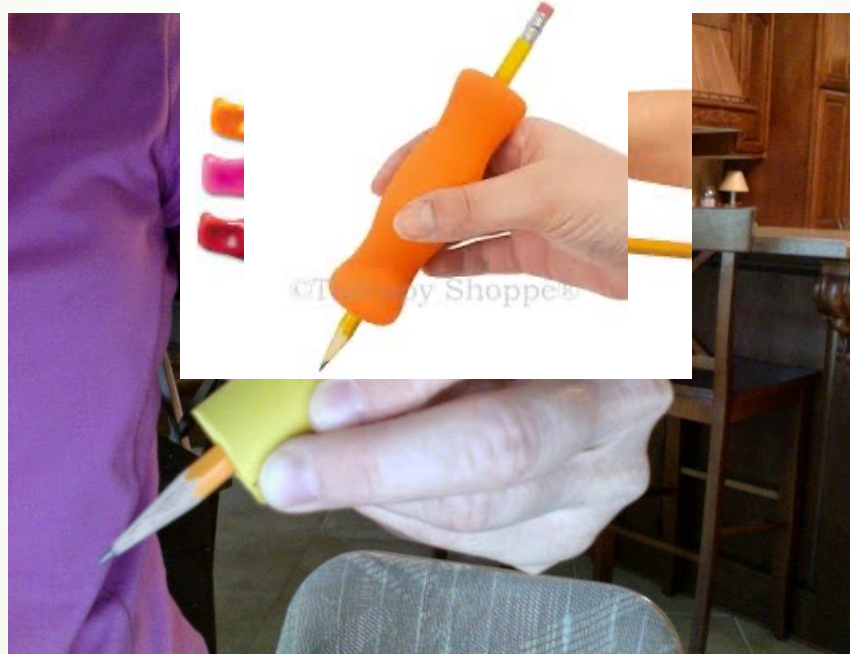
➤ Zebra splints

Thick handles made by Oxo™



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For pencils or pens





Do braces make your muscles weaker?

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Do braces make your muscles weaker?

 **NO**

- ▶ It's a common misconception
- ▶ No brace is tight enough to stop muscles from moving
- ▶ In fact, braces stabilize joints so your muscles can move the joints more efficiently.



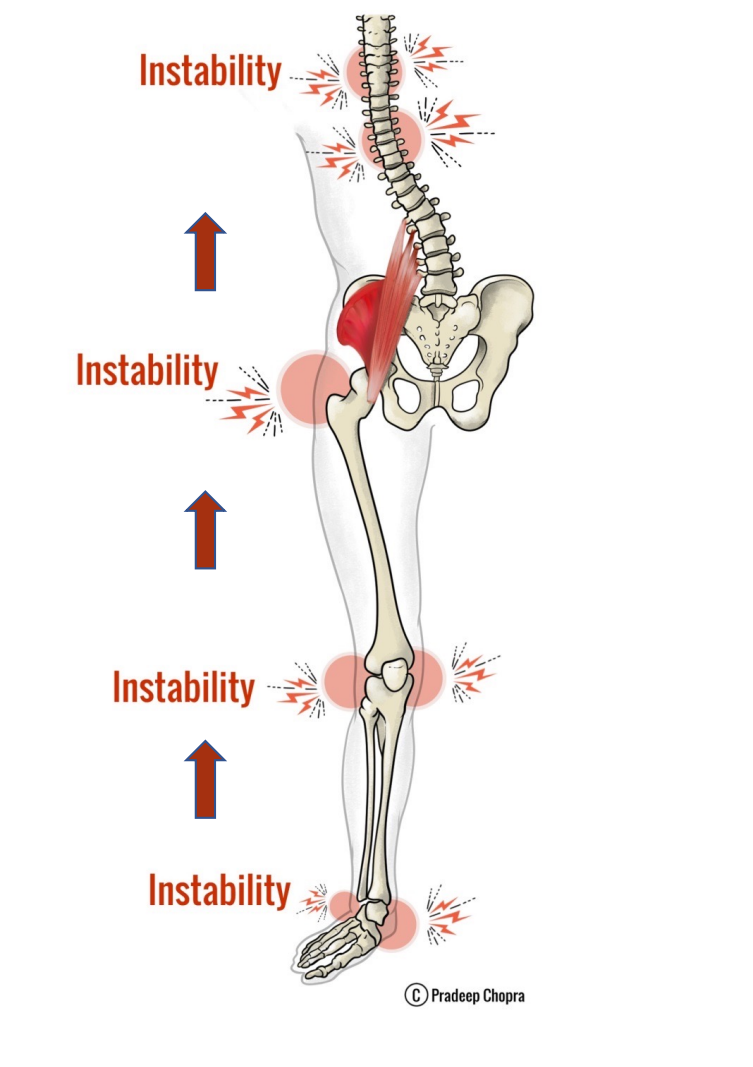
Pain in lower half of the body

- ▶ If the feet and ankles are unstable, they make
- ▶ The knees even more unstable, which then
- ▶ Makes the hips unstable, which then
- ▶ Throws the pelvis and spine off

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Feet and ankles

- ▶ There are usually 2 problems
- ▶ Unstable ankles
- ▶ Flat feet

Unstable ankles

- People with unstable ankles tend to roll their ankles when walking.
- This causes a nerve near the knee to become pinched.
- This causes pain on the side of the lower leg.
- They tend to trip on their toes



High top shoes for ankle instability

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Knees

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Problems with knees in EDS

- ▶ Hypermobile knee cap
- ▶ Hyperextension of the knee joint

Hypermobile knee cap

- ▶ A hypermobile patella can make the knee unstable
- ▶ It causes pain in the thigh muscles that support the patella



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<https://thekneeresource.com/conditions/patellar-dislocation/>

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Hyperextension of knee joint

- ▶ Hyperextension of the knee joints is very common in EDS
- ▶ This puts extra stress on the ligaments of the knee
- ▶ It feels unstable, and may buckle under you at times
- ▶ Banana knees



<https://www.thegaitguys.com/thedailyblog/tag/anterior+menisconfemoral+impingement>

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Treatment options for knee pain in EDS

- ▶ Stabilize the feet and ankles, first
- ▶ Strengthen muscles around the knee
- ▶ Knee brace keeps the knee cap in place and
- ▶ Prevents the knee from hyperextending
- ▶ Preferred brand:
- ▶ MedSpec™ Gripper Hinged knee brace

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EDS and children

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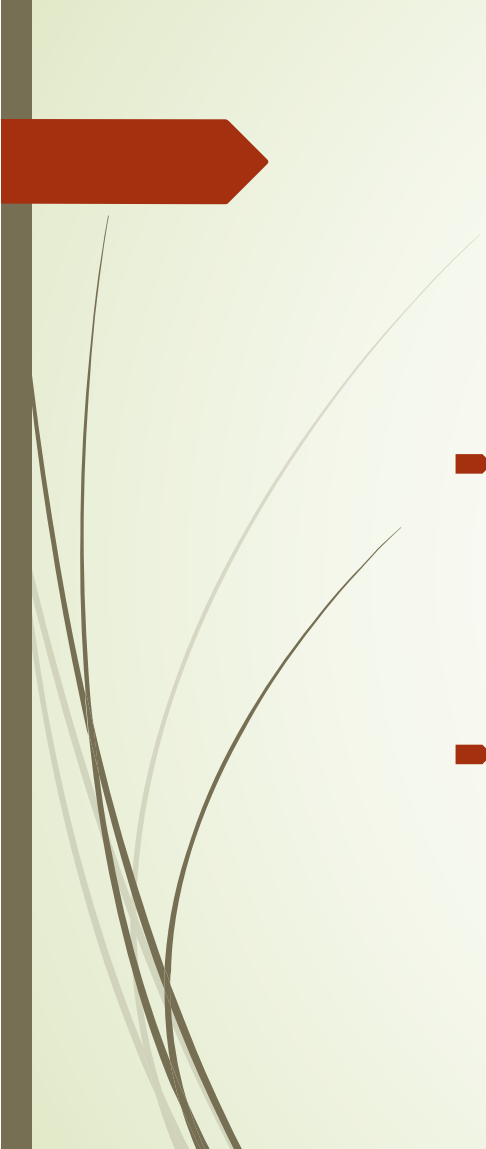
EDS in children

- ▶ Its best to start managing their hypermobility early in life.
- ▶ Excessive hyperextension can lead to life long issues
- ▶ They do have a rough time during growth spurts.



Dangers of EDS and children

- ▶ The biggest risk is from ignorant hospitals and doctors
- ▶ Unexplained bruises,
- ▶ dislocated joints,
- ▶ chronic body wide pain
- ▶ to the ignorant health care provider it may look suspicious.

- 
- ▶ Things people with EDS have to hear when they look for help: “EDS does not hurt” “EDS is very rare, you can’t have it”, “EDS what?” “your skin is not very stretchy”
 - ▶ The diagnosis of Conversion Disorder or Munchausen by Proxy are often made by providers with no training in Psychiatry and vice versa psychiatrists have no training in pain conditions.




Misdiagnosis

- ▶ Please check with other patients to get feedback before you go to a hospital.
- ▶ If you sense that the doctors are skeptical of your complaints, consider going somewhere else.
- ▶ When you go to a big academic hospital, chances are that the first few doctors who you see are interns or residents who are the least qualified to manage complex medical conditions.
- ▶ If you are able, choose a concierge physician.

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Stay away from children's hospitals unless it is an emergency, or you definitely know a doctor there who understands your child

Have your pediatrician on your side. Keep him/her updated on your child's health



It is important to understand the cause of the pain before deciding on treatment options.



Dysautonomia / POTS (Postural Orthostatic Tachycardia Syndrome)

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Postural Orthostatic Tachycardia syndrome (PoTS) - Symptoms

- Lightheaded
- Heart racing (Palpitations)
- Fatigue
- Headaches
- Cold hands and feet
- Poor concentration “brain fog”
- Feeling of constant anxiety

Diagnosis of PoTS

- Measure blood pressure and heart rate:
 - after lying flat for 5 minutes
 - standing up - immediately
 - Standing for 10 minutes

- Q-sweat test

- Tilt Table Test

Diagnosis of PoTS

- ▶ Increase in heart rate by 30 beats/min within 10 minutes of standing
- ▶ heart rate of 120 beats / min within the first 10 minutes of standing
- ▶ No significant change in blood pressure
- ▶ Lightheaded
- ▶ In children an increase of 40 beats/minute

Causes of Dysautonomia / PoTS

- ▶ Blood pooling
- ▶ Cranio Cervical instability
- ▶ Autoimmune Dysfunction
- ▶ MALS (median arcuate ligament syndrome) ?

Anxiety in EDS – it not from the brain. It's an overactivity of the autonomic nerves

- ▶ Patients with EDS and POTS (Dysautonomia) are often over diagnosed to have anxiety
- ▶ Symptoms of undiagnosed palpitations, fatigue, dizziness, chronic pain are attributed to 'anxiety'.
- ▶ In most cases its simply Dysautonomia – malfunction of the autonomic nervous system.
- ▶ The symptoms of 'anxiety' are more from dysfunction of the flight and fight mechanism

Management of PoTS

- ▶ Increase salt by mouth. Try a salty diet first, then try salt pills.
- ▶ "Salt Stick Vitassium" is a good brand to try. Easy to swallow. Contains potassium
- ▶ Electrolyte fluids – at least 16 oz per day.
- ▶ "Liquid IV" is a good brand. Other brands: Nunn, Banana Bag, Drip Drop
- ▶ Increase water intake – at least 2 liters / day
- ▶ Compression tights up to thighs
- ▶ Medications to try: Ivabradine,

Exercise in PoTS

- ▶ Exercise while lying flat or reclining
- ▶ Do not push yourself.
- ▶ Exercise whatever you can

Technology

- ▶ Amazon's Echo "Alexa" setup in different rooms of the house.
- ▶ Should you feel lightheaded, use it to call for help
- ▶ Apple Watch 4 and after: will record heart rate and has a fall warning. It can detect a fall and call for help



Mast Cell Activation Syndrome

MCAS

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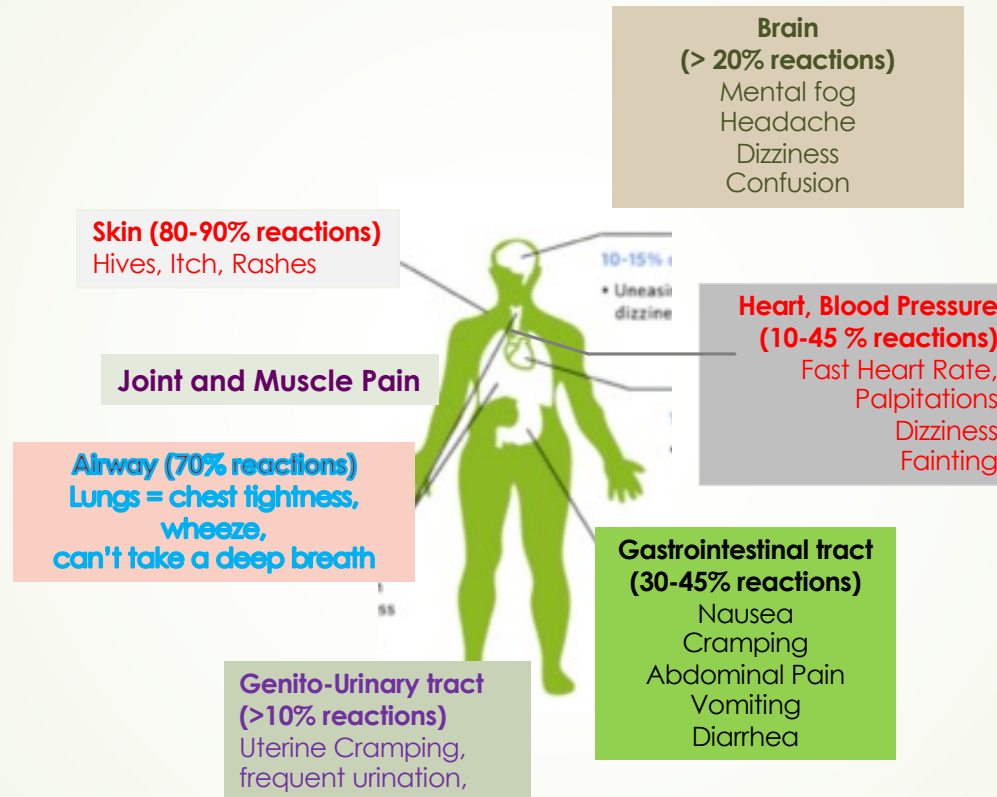
Mast Cell Activation Syndrome and EDS

- It feels like having flu – tired, cold, sick and no appetite.
- It adds to the fatigue
- Causes loss of appetite
- It causes diffuse body wide pain
- Worsens PoTS

Mast Cell Activation Syndrome (MCAS) and EDS

- ▶ Mast cells are normally found everywhere in the body
- ▶ They are activated when the body's immune system detects a danger
- ▶ In EDS, the mast cells start to misbehave
- ▶ They release many chemical agents like histamine, cytokines that increase pain

Signs and Symptoms of Mast Cell Activation Disorders



Management of MCAS – in 3 parts

- ▶ Antihistamines such as famotidine, Benadryl, Zyrtec
- ▶ Mast cell stabilizers: Cromolyn, Ketotifen, Quercetin
- ▶ Look for mast cell triggers: food or air that you are breathing
- ▶ Avoid drugs or food with color dye such as red or blue dye

Mast Cell Triggers

- Air quality
 - Look for mold
 - Dust
 - Avoid rapid changes in temperature
 - Place a HEP room air filter in the room
- Foods:
 - Gluten
 - Seasonings,
 - Dairy
 - Beef products



Fatigue in EDS

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Fatigue

- EDS
- Postural Orthostatic Tachycardia / Dysautonomia
- Mast Cell Activation Syndrome
- Drugs
- Pain
- Poor sleep
- secondary mitochondrial dysfunction

Mitochondrial dysfunction

- ▶ Rechargeable batteries
- ▶ The batteries (mitochondria) do not function well and cause fatigue
- ▶ Mineral supplement / mito cocktail



Abdominal pain

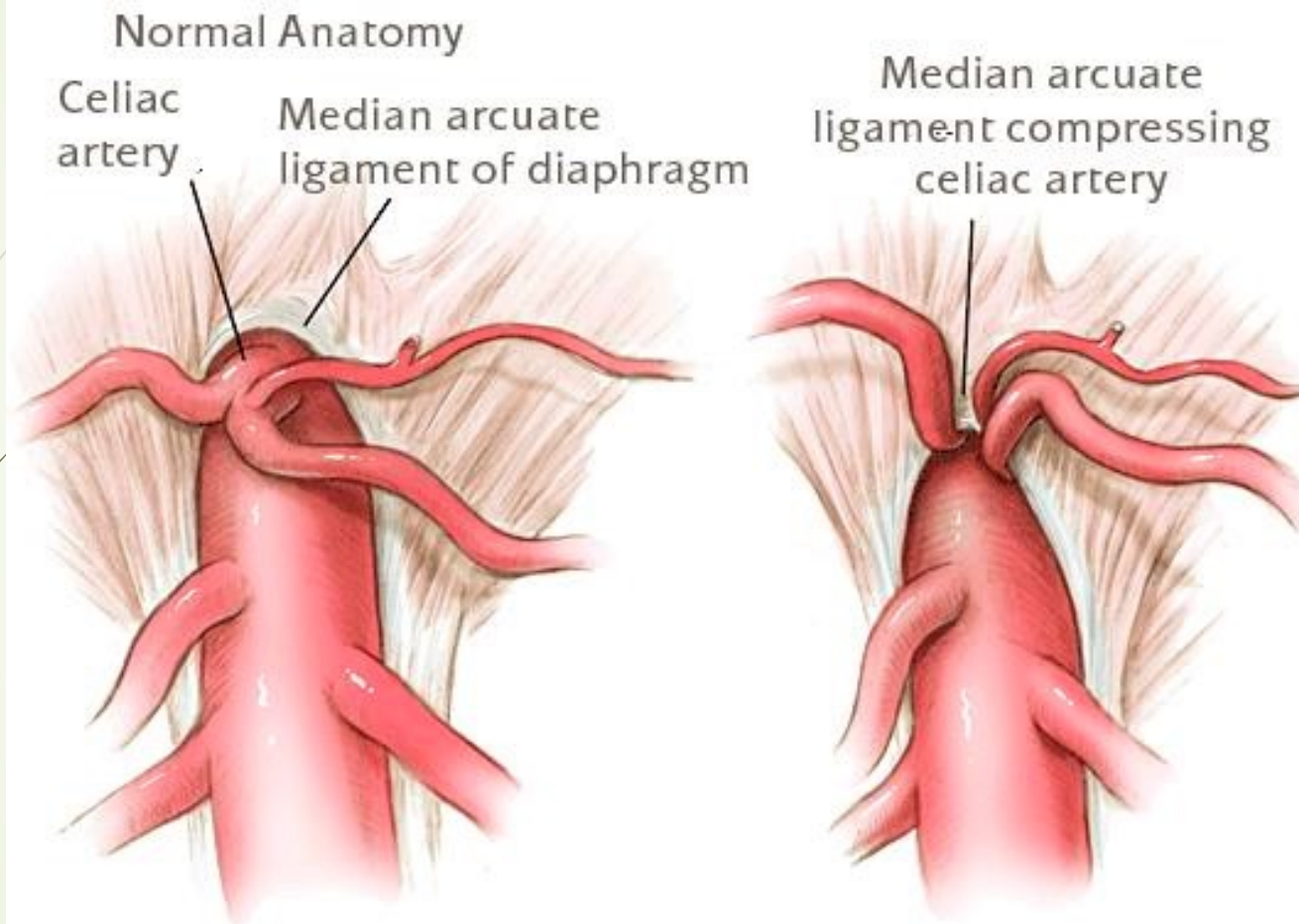
- Very common in EDS
- Gastroparesis (slowing down of the stomach)
- Slowing down of the intestines
- Alternating diarrhea and constipation
- PoTS –causes nausea, acid reflux, bloating and constipation
- Diverticulosis, prolapse

MALS (Median Arcuate Ligament Syndrome)

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Abdominal pain – MALS

- ▶ Upper Belly pain after eating food.
- ▶ Lethargy after eating a meal
- ▶ Exercise makes the pain worse
- ▶ Pain gets worse with standing
- ▶ Pain is better with lying on side




Abdominal pain – MALS

- ▶ Treatment is surgical decompression
- ▶ The results of surgery done by an experienced surgeon are good.

Gastrointestinal

- ▶ Gastroparesis from MCAS and PoTS
- ▶ Resolor (Canada) or Motegrity (USA) is a good drug to try for gastroparesis
- ▶ $\frac{1}{2}$ to $\frac{1}{4}$ tablet may be sufficient
- ▶ Managing your MCAS is really important



It is important to understand the cause of the pain before deciding on treatment options.

Muscle pain

- ▶ EPSOM salt bath – 2 large cups of EPSOM salt in a bathtub
- ▶ Magnesium Lotion
- ▶ IV Magnesium
- ▶ Carbidopa+Levodopa (Sinemet)

Menstrual Periods

- ▶ Talk to your OB/GYN about completely stopping menstrual periods
- ▶ They can worsen MCAS, PoTS, body wide pain, pelvic pain

Ketamine

- ▶ Ketamine is a strong pain killer at low doses.
- ▶ It works best when given as IV
- ▶ It can be used as a nasal spray or under the tongue.
- ▶ Only good for neuropathic pain
- ▶ Not much value in EDS pain

Ketamine for chronic pain

- Its useful in patients who have severe nerve pain
- It can be used as a nasal spray or under the tongue, but the best results are with IV administration
- Its not useful for EDS joint and muscles pain but if the pain is because a nerve has been affected such a Complex Regional Pain Syndrome (CRPS)

Opioids

- Reasonable to try
- More useful in acute flare ups
- Stay at low doses. They work better
- Some patients with EDS do not respond well to opioids.
- For surgical pain, dilaudid is a good option.
- The problem with opioids in EDS is that they worsen MCAS, GI issues. Hence, take them as needed.

Opioids and addiction

- ▶ According to the CDC, only 0.6% of patients are at risk of addiction to opioids at low doses
- ▶ 6% are at risk when on high doses.

Opioids and addiction in chronic pain is almost non-existent

versus no opioid prescription (22). Rates of opioid abuse or dependence diagnosis ranged from 0.7% with lower-dose (≤ 36 MME) chronic therapy to 6.1% with higher-dose (≥ 120 MME) chronic therapy, versus 0.004% with no opioids prescribed. Ten fair-quality uncontrolled studies reported

Opioids and addiction

- ▶ A person must be 'hard wired' to become addicted.
- ▶ They have other addictions like alcohol, illegal substances.
- ▶ Media – the deaths from opioids in the media are not from opioids only. These people have been taking other dangerous street drugs with alcohol.
- ▶ The CDC has not said that opioids cannot be used for pain. They have cautioned against high doses or giving with other drugs.
- ▶ Unfortunately, medical boards have made it difficult for physicians to write opioids by burdening them with paperwork and subtle threats.

Work with your physician

- ▶ Build a trust with your doctor.
- ▶ Start low and go slow
- ▶ Avoid excessive phone calls / emails about increasing the dose
- ▶ Most insurance companies deny opioids. This means hours of phone calls and lots of paperwork for your doctor.
- ▶ To help your doctor, the cash price / coupons are faster and cheaper. Talk to the pharmacist.

Undergoing surgery

- Before you go through surgery talk to the surgeon about your pain management after discharge.
- Some surgeons have a policy of discharging a patient after a major spinal surgery on Tylenol.

Marijuana

- ▶ Works well in EDS
- ▶ Does not affect MCAS or Gastrointestinal system
- ▶ Equal amounts of CBD and THC work best
- ▶ Avoid hemp CBD

What is LDN?

LDN (low Dose Naltrexone)

- ▶ I published the first paper in 2012 on the use of LDN for pain
- ▶ It helps with pain and fatigue by increasing endorphin levels
- ▶ Normal dose 4.5mg per day
- ▶ May start at 0.5mg per day and increase slowly
- ▶ Give it 6 months trial

What is Low Dose Naltrexone?

- ▶ LDN is a competitive opioid receptor ANTAGONIST.
- ▶ It blocks the effect of opioids
- ▶ Not an opioid
- ▶ Not addicting

LDN

- ▶ LDN is a disease modifying agent versus drugs that do not modify condition.
- ▶ It fixes what is broken and not band aid it

What does LDN help?

- ▶ Chronic pain
- ▶ Neuropathic pain
- ▶ Autoimmune dysfunction
- ▶ Mast Cell Activation Syndrome (MCAS)
- ▶ Fatigue
- ▶ Brain Fog

Practical tips

- ▶ Start at 2mg in AM for 2 weeks.
- ▶ Increase to 4.5mg per day
- ▶ May take it in morning or night.
- ▶ Usual side effects: headaches, insomnia and colorful dreams.
- ▶ Side effects go away after some time. Rarely, colorful dreams may persist

Practical tips

- ▶ It may take a few weeks to see a response.
- ▶ Try for at least 6 months
- ▶ If patients complain of significant side effects, may start at 1mg or even 0.5mg per day
- ▶ Must be ordered from a compounding pharmacy
- ▶ Can be a pill, liquid or cream

Who can take LDN?

- ▀ Adults
- ▀ Children

What if I am chemically sensitive?

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- ▶ Start with a very small dose such 0.1mg per day.
- ▶ If it helps – great
- ▶ You can increase it by 0.1mg every weeks or months .
- ▶ Check with the compounding pharmacy that they are not including any preservatives, colors, binding agents

Compounding pharmacies

- ▶ Every state has lots of them, you can go to any of them
- ▶ My preference for ordering LDN are:
 - ▶ Belmar Pharmacy in CO
 - ▶ Johnson Compounding in MA
 - These pharmacies are aware of patients with chemical sensitivities.
 - There may be others, but I don't know of them.

TAKE LDN ONLY ONCE A
DAY ONLY

TAKE LDN ONLY ONCE A
DAY ONLY

Fillers and LDN

- Avicel (acidophilus and calcium carbonate), sucrose, lactose and other fillers are used, although some patients are intolerant of certain fillers, and some patients claim to respond better to certain fillers.
- My preference – no fillers if it is possible

LDN and opioids

- ▶ Patients on LDN can be administered opioids in an emergency.
- ▶ The number of mu receptors blocked is small and transient, it does not affect the effectiveness of opioids.

LDN and opioids

- ▶ If a patient needs to take an opioid pill for acute pain, it is reasonable to take it
- ▶ Preferably 4 hours before or 4 hours after opioid pill

LDN and opioids

- ▶ No reported cases of withdrawal in patients taking LDN and given a low dose of opioid.

LDN and hospitals

- ▶ Hospitals are full of very ignorant physicians.
- ▶ They do not know, understand and care about LDN
- ▶ They will continue your “LDN” at 50mg because the software does not have the option for 4.5mg
- ▶ The hospital pharmacy will not compound it

LDN and hospitals

- ▶ Then there are other doctors who assume you are a drug addict because you take LDN
- ▶ There are some doctors who will give you a pediatric dose of opioid after spine surgery and claim that its not working because you are on LDN

LDN and hospitals – my advice

- ▶ Hold off taking LDN if you are going to the hospital
- ▶ If elective surgery, stop LDN 24 hours before
- ▶ Restart LDN 24 hours after the last opioid dose

TAKE LDN ONLY ONCE A
DAY ONLY

Practical tips

- ▶ Ointment is usually used in children with Autism and sensory issues.
- ▶ LDN needs to be taken once day – does not matter whether in morning or night
- ▶ If there is no response to 4.5mg, it may be reasonable to go to higher doses 6mg to 12mg
- ▶ LDN should be compounded for immediate release.

TAKE LDN ONLY ONCE A
DAY ONLY

Web resource

- ▶ LDN Research Trust
- ▶ <https://www.ldnresearchtrust.org/>

Perioperative care of EDS

- ▶ Chopra P, Bluestein, L: Perioperative Care in Patients with Ehlers Danlos Syndromes. January 2020. [Open Journal of Anesthesiology](#) 10(01):13-29
- ▶ DOI:[10.4236/ojanes.2020.101002](https://doi.org/10.4236/ojanes.2020.101002)



Poor concept of exercising in pain

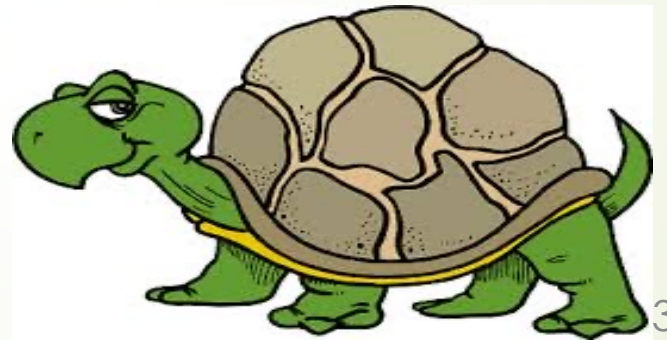


➔ No pain, no gain



Starting treatment -
medicines and exercise

Start low, go slow





Start low, go slow

- ▶ In EDS, healing is very slow.
- ▶ When doing exercises / Physical Therapy go very slow.
- ▶ If you push yourself excessively, you may cause damage to your fragile tissue that will take a lot longer to heal.
- ▶ If you feel that your PT is not aware of the special requirements for EDS – then do your own exercises.
- ▶ Its important to exercise at a low level daily than to do a lot one day and risk injury.

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Service dogs





Service Dogs - invaluable

- ▶ POTS – they can sense when their owner is having an episode of dizziness or seizure
- ▶ EDS and pain - they protect the limb from being injured or touched
- ▶ Helps boost confidence in their owners, making them more independent
- ▶ Help with balance, call for help, open doors, switch on lights, pull wheelchairs, anxiety,

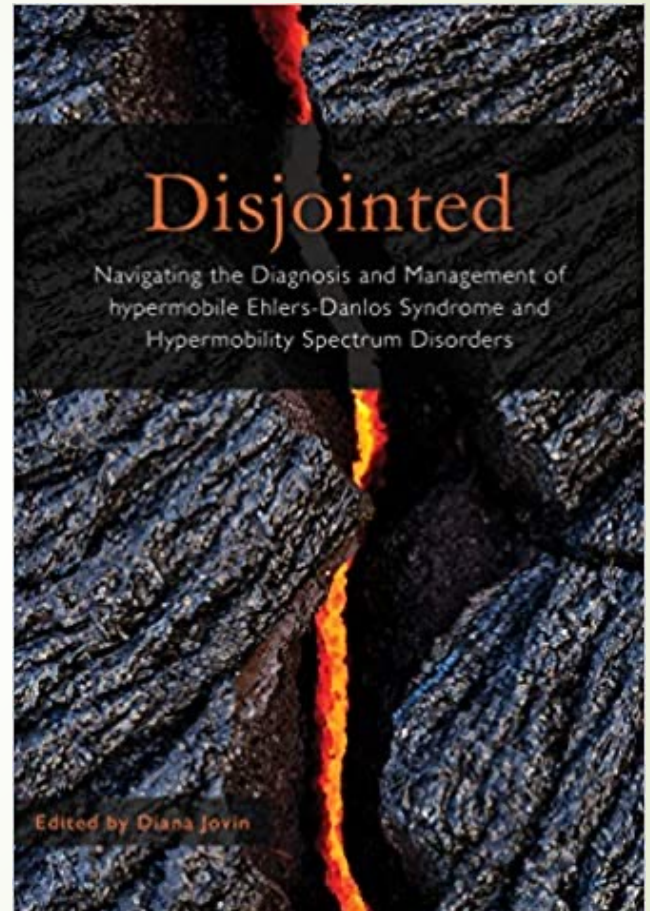
Some organizations that can help get a service dog

- ▶ <https://www.pawswithacause.org>
- ▶ <http://www.neads.org>
- ▶ <http://freedom servicedogs.org>



More information

- ▶ “Disjointed” is a book written on EDS by specialists.
- ▶ The information is credible and very helpful.



Lipedema and EDS

Lipedema

- ▶ This is a condition that causes excess fat to accumulate in the lower part of the body
- ▶ They have loose connective tissue with increased nodules or fibrotic fat tissue
- ▶ It often involves the buttocks, thighs and calves
- ▶ The upper arm can also be affected
- ▶ The fat is painful

Lipedema collects in the buttocks, legs



Lipedema and EDS

- ▶ A lot of women with lipedema have been noted to have hypermobility or EDS
- ▶ The fat can be painful when pressed
- ▶ Often associated with joint pain and hypermobility of the lower legs
- ▶ <https://www.lipedema.org>
- ▶ <https://www.fatdisorders.org>

Lipedema

- ▶ Lipedema fat cannot be reduced by diet or exercise or weight loss surgery.
- ▶ It may be associated with Lymphedema

Dercum's disease

- ▶ Patients develop multiple, painful growths of fatty tissue (lipomas)
- ▶ Many patients with Dercum's disease also have joint hypermobility and EDS





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Long Covid

Symptoms of Long-COVID

- Breathlessness
- Palpitation
- Chest discomfort
- Fatigue
- Pain
- Brain fog
- Poor sleep
- Orthostatic intolerance

Symptoms of Long-COVID

- Pins and needles and numbness
- Abdominal discomfort
- Nausea
- Joint and muscle pain
- Symptoms of anxiety
- Skin rashes
- Headache

- ▶ Two things that stood out as more definitive treatments:
 - ▶ Dysautonomia
 - ▶ Mast Cell Activation Syndrome (MCAS)

Robin Gorna, Nathalie MacDermott, Clare Rayner, Margaret O'Hara, Sophie Evans, Lisa Agyen, Will Nutland, Natalie Rogers, Claire Hastie. Long COVID guidelines need to reflect lived experience. The Lancet. Vol 397 February 6, 2021

Treatment of Long Covid

- Manage the symptoms of PoTS more aggressively including IV fluids, stimulants, mineral supplement for mitochondrial dysfunction, home oxygen,
- MCAS: not all patients with Long Covid present with MCAS. If they do, then managing the MCAS more aggressively helps.



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Pharmacogenomics

Pharmacogenomics

- This is a test that looks changes or variants in genes that may determine how your body processes medications.
- All medications are processed by the body – a term known as metabolized.
- Depending on how your body processes or metabolizes drugs, will determine how you respond to them

The Four Metabolizer types

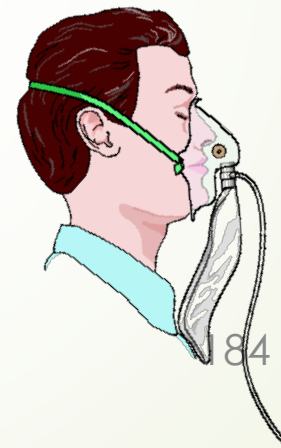
1. Poor Metabolizer: Medication is broken down very slowly. May experience side effects at standard doses.
2. Intermediate Metabolizer: Slow rate of metabolism. May have too much medication at standard doses, potentially causing side effects.
3. Extensive Metabolizer: Normal rate of metabolism. Has normal amount of medication at standard doses.
4. Ultrarapid Metabolizer: Medication is rapidly broken down. Medication may be removed from a patient's system too quickly to provide symptom relief.


How does that affect us?

- ▶ Poor metabolizers need to take lower doses of the drug
- ▶ Ultra rapid metabolizers may need to take higher doses.
- ▶ Sometimes an ultra metabolizer may increase the metabolism of another drug you may be taking at the same time.
- ▶ Get a pharmacogenomic test done from a company that actually tells which drugs are affected, not just the enzymes
- ▶ To be safe, always start your medications low and go up very slowly

Oxygen supplementation

- Anecdotal experience
- Oxygen by non-rebreather mask 3 liters to 5 liters, 20minutes per day up to twice a day.
- May administer as needed for severe symptoms of fatigue or dizziness.





Levodopa-carbidopa

- ▶ Anecdotal experience
- ▶ Significant relief for muscle spasms and dystonia
- ▶ Low dose
- ▶ Sinemet ®



Poor concept of exercising in pain



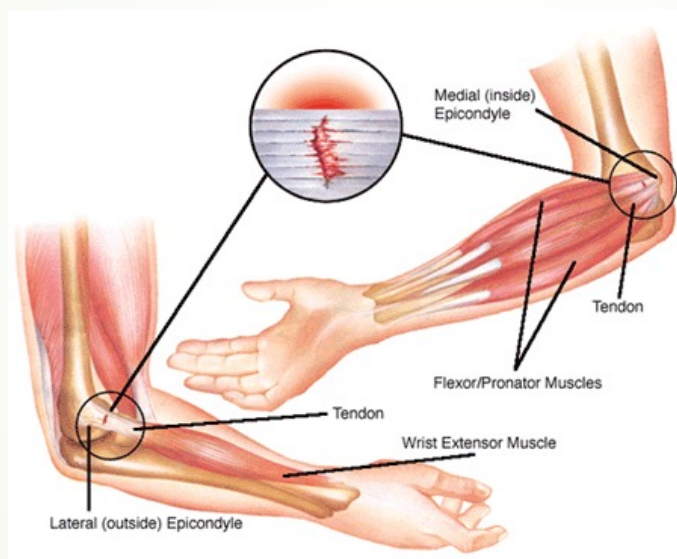
➔ No pain, no gain



Types of tissue injury in EDS

- ▶ Macro trauma – a large event or trauma resulting in injury. Dislocation or fracture
- ▶ **Micro trauma** – small repetitive traumas resulting in constant tissue breakdown. Invisible tissue breakdown
- ▶ For example – overstretching muscles and ligaments. Vacuuming, chopping food, dusting

Micro trauma





Exercises for EDS

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Exercises for EDS

- ▶ Muscles toning without stressing the joints – for stabilizing joints
- ▶ Proprioception exercises.
- ▶ Exercising consistently is more important than the degree of exercise



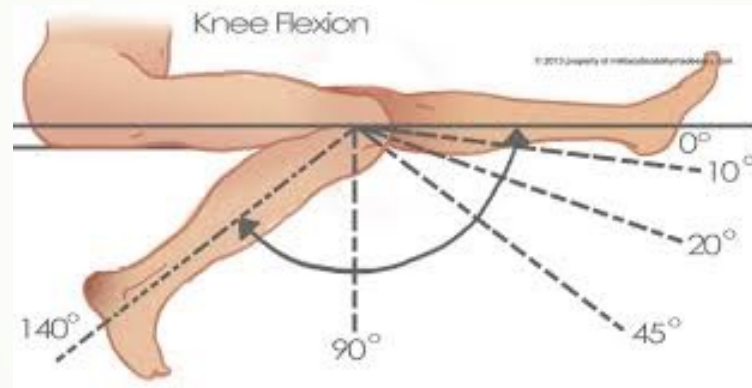
Muscle strengthening exercises

- Avoid joint loading or joint stressing
- Start very low, really very low
- Focus on muscle toning
- Avoid stretching

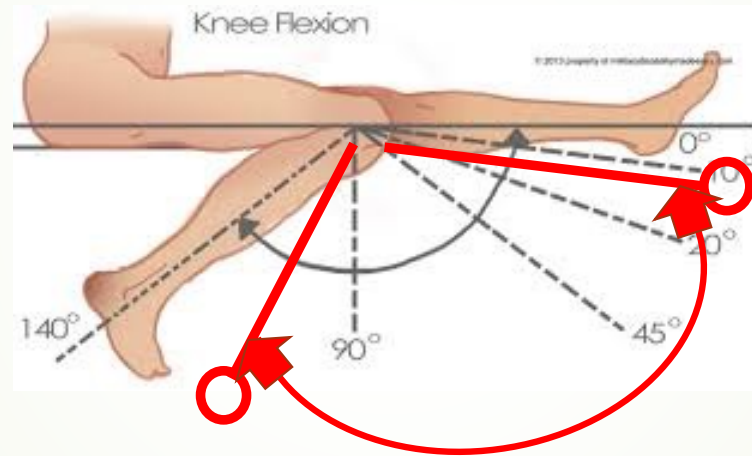
Exercise for strengthening muscles in EDS

- Not a good idea
- Patients with EDS have soft tissue.
- They undergo more micro and even macro trauma
- They take longer to heal.
- Exercise of muscles should be limited to improving the tone rather than strength.
- Its more about moving the muscles than focusing on strengthening them

Keep limb movements within extreme range of motion



Keep limb movements within extreme range of motion





Aquatic therapy

- ▶ Best form of exercise in EDS
- ▶ The contact of water with the skin helps the brain move muscles more efficiently, improves proprioception
- ▶ The water makes us weigh less which takes the load off the joints allowing us to exercise freely
- ▶ Avoid swimming – it strains the joints of the neck and shoulders.

Avoid swimming – injury to shoulder and neck





The Feldenkrais Method

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The Feldenkrais Method

- It is a type of physiotherapy that helps repair impaired connections between the brain and the body
- Patients with EDS and chronic joint conditions develop inefficient or strained habitual movement patterns
- **It uses slow repetition to teach correct and safe movements in EDS**
- The movements are gentle, slow, repeated movements.

The Feldenkrais Method

► <http://www.feldenkrais.com>

- Can be done sitting or lying down
- Each session consists of comfortable, easy movements within the limits of





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CGRP antagonists

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CGRP antagonists

- ▶ This class of drugs have been approved for migraines only.
- ▶ They have very few side effects.
- ▶ Research has shown that they work for nerve pain.
- ▶ It will not be long before they get approved for nerve pain also
- ▶ Drugs such as: Nurtec, Ubrelvy, Qulipta.

Talking to your Primary Care doctor about your EDS

- ▶ Do not
- ▶ Keep them updated but please do not burden them with managing your POTS or MCAS or neurological symptoms
- ▶ Let PCP's do what they do best – take care of routine problems
- ▶ Please keep them updated
- ▶ Make sure you see an MD only. You have a complex medical condition which most people do not care to read about. Seeing PA's, RNP, Residents, Interns and medical students will not be helpful.

Electronic Medical Records (EMR)

- All EMR's share your information.
- Please ask your physicians to block that feature, so your medical information cannot be shared with out your permission.
- Reason: very often patients get misdiagnosed (e.g., eating disorder), this gets passed on to all subsequent doctors.

Good resources for understand EDS

- Book: “Disjointed”
- Website: :Chronicpainpartners.com”



Thank you

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