Management of Chronic Pain in Ehlers Danlos Syndrome

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Disclosure and 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.

To manage pain first find out what's broken

- It is important to understand the <u>cause of the pain</u> before deciding on treatment options.
- For example, pain in the shoulder joint can be from:
 - Dislocated or subluxing shoulder joint,
 - Muscle spasms from the shoulder muscles
 - Pinched nerve at the shoulder
 - All of the above
- The treatment of each of these is different

Types of tissue injury in EDS

- Micro trauma when you use your joints repetitively there is microscopic tissue breakdown.
- In EDS tissue breakdown happens faster and healing is slower
- Pace yourself, slow down on activities that cause more tissue damage
- Activities that cause micro trauma overstretching muscles, repetitive use of joints, sitting in a weird position, subluxing joints
- Macro trauma a large event or trauma resulting in injury.

For example - Dislocation or fracture

Principles of managing pain in EDS

• Use a mix of treatments

- For example Knee pain due to instability
 - Stabilize knee with braces
 - Strengthen muscles around the knee
 - Medications NSAID's, topical etc.





Explanations

- POTS (Postural Orthostatic Tachycardia Syndrome):
 - Feeling of constant anxiety
 - Brain fog
 - Fatigue
 - Headaches
 - Gastroparesis fullness after eating a small meal and nausea
- MCAS (Mast Cell Activation Syndrome)
 - Like you have the flu all the time (tired, cold, hot, pain)
 - Brain fog,
 - Fatigue
 - Gastroparesis
- MCAS can worsen / cause POTS

Fatigue in EDS

- Postural Orthostatic Tachycardia Syndrome (POTS) big reason
- Mast Cell Activation Syndrome (MCAS) big reason
- EDS
- Poor sleep
- Medications
- Pain

Fatigue – managing it

- POTS Metoprolol, clonidine, electrolytes, compression clothes
- MCAS cold medicines, zantac, cromolyn, ketotifen.
- Potassium by mouth pill, drink
- Avoid low blood sugar before bedtime
- A small dose of stimulant like Adderall in the morning
- LDN Low Dose Naltrexone

Mitochondrial dysfunction

- Think of Mitochondrial dysfunction (secondary)
- Mitochondria are the rechargeable batteries in our body.
- A problem with the rechargeable batteries can cause fatigue
- Treatment: Mitochondrial cocktail.

Muscle Spasms

- Sinemet (25/100mg) helpful. Low side effects
- Myofascial release
- Gently rolling a tennis ball or golf ball to break the spasm
- Most muscle relaxants do not really break spasms.
- Muscle relaxants add to fatigue and brain fog

Pain in EDS by body regions

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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
- 5. PoTS / Dysautonomia
- 7. Spontaneous CSF (Cerebrospinal) leak low pressure inside the head
- 8. Idiopathic Intracranial Hypertension (raised pressure inside the head)
- 9. Cranio Cervical Instability (Instability of the neck and head)

"My head feels too heavy to hold up"

Chin poking forward position



Stand Against a Wall to Discover Your Proper Posture



1 6

Neck pain and headaches

- A common cause of neck pain is posture
- 'Chin poking forward position'
- Before looking at other reasons, correct this first
- If there are other reasons like cervical instability, Chiari malformation etc – these need to be addressed

Pain from a poor posture



Common reasons for incorrect head posture in EDS

- Vision Blurry vision. Usually intermittent
- Postural Orthostatic Intolerance (POTS)
- Loose ligaments in the spine
- Instability of the head on the neck (Cranio Cervical instability)

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Managing neck pain and headaches from poor posture

- Place index finger in front of chin and push back head gently till ears are in line with shoulders
- Large monitor, sit well balanced while working on a computer
- Vision correction
- Manage POTS



Upper back pain

Upper back pain in EDS

- Usually poor posture hunched over
- Shoulder instability
- Rib subluxation
- Doing a repetitive task with hands or arms (typing, vacuuming) -Repetitive Strain Injury
- In women, it can be the weight of the breast tissue dragging the upper torso forward and the muscles of the upper back attempting to stabilize the torso

Upper back and Shoulder pain in women with EDS

- Helps upper back, shoulder and Thoracic outlet pain
- Sports bra with:
- racer back (cross straps).
- Wide straps.
- Front closure
- Proper fitting recommend getting it done professionally.
- Shoulder stabilizing brace



Shoulder pain

• Laxity of the shoulder joint causes the muscles (rotator cuff) around the shoulder to spasm

• Thoracic Outlet syndrome.

Pain patterns in Thoracic Outlet syndrome



Thoracic Outlet Syndrome



www.**sgergo**.com

Thoracic Outlet Syndrome

- Thoracic Outlet exercises
- Kinesio taping
- Botox injections
- Stabilize shoulder
- surgery

"My whole head hurts, I see double, I can feel my heart beat in my ears," Headaches – Idiopathic Intracranial Hypertension (IIH)

- Raised pressure inside the head
- Vision problems double vision, sensitivity to light
- Pulsating ringing in the ears (pulsatile tinnitus)
- Maybe because of narrowing of blood flow (venous sinus stenosis)
- Diagnosis: spinal tap, eye exam, MR venography
- Treatment: medicines to decrease fluid pressure in the head, shunt to drain excess fluid, stent

"My headache gets worse when I stand and it almost goes away when I lie down"

Low pressure inside the head- Spontaneous CSF leak



3

CSF leak headaches

- Avoid spinal tap, spinal injections unless absolutely necessary
- May happen after spinal surgery
- Drink more fluids
- Abdominal binder (corset)
- Caffeine
- Epidural blood patch, may need to be repeated

CSF leak headache

- When CSF leaks, it causes the pressure in the head to decrease
- In some cases, an increase in pressure inside the head may be the cause of the CSF leak.
- Management would be to stop the leak and see if that helps the headaches
- If it does not stop the headaches, then it should be investigated for increased pressure inside the head

Neck pain
Cranio Cervical instability in EDS

- The neck is stabilized by ligaments
- Laxity of the ligaments causes the joints in the neck to move more, which causes the neck muscles to tighten
- Excessive movement of the joints in the neck causes cranio cervical instability

Cranio Cervical instability (CCI

- Neck pain / stiffness
- Headaches
- Dizziness
- Paresthesia to face
- Fatigue
- Poor sleep
- Nausea

- Poor vision
- Anxiety
- Lightheaded
- Poor balance
- Difficulty swallowing

Remember a lot of these symptoms overlap with other conditions

Imaging for Cranio Cervical Instability

- Functional computerized tomography (fCT scan)
 - Flexion.
 - Rotate neck left 90 degrees.
 - Rotate neck right 90 degrees.

Cranio Cervical instability in EDS – MRI scan findings

- These measurements have to specifically asked for when getting an MRI.
- 1. Clivo-axial angle (normal 140 to 160 degrees)
- 2. Harris Measurement (instability if > 12mm)
- 3. Grabb, Mapstone and Oakes measurement (> 9mm suggests high risk of ventral brainstem compression)

Cranio Cervical Instability management

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

Vista[®] MultiPost Therapy Collar – an improved design



Soft neck collars are useless

"neck warmers"

Numbing medicine injections

- Commonly used numbing medicine injections (Lidocaine, Bupivaciane) may be ineffective
- CARBOCAINE may work well in EDS

Abdominal pain

- Can be from POTS, MCAS and EDS
- Gastroparesis (slowing down of the stomach)
- Slowing down of the intestines
- Alternating diarrhea and constipation
- PoTS, MCAS causes nausea, acid reflux, bloating and constipation

Abdominal Pain – things to look for

- Small Intestinal Bacterial Overgrowth (SIBO)
 - Breath test
 - Antibiotics
- Median Arcuate Ligament Syndrome (MALS) rare, but can happen
 - Surgery

Treatments for GI pain

- Low FODMAP diet for bloating, pain and diarrhea
- Treating PoTS and Mast Cell Activation Syndrome is important
- If POTS then try eating in a recliner (feet elevated) 30 minutes before, during and 30 minutes after
- Compression garment such as the Spinal Q Vest by AlignMed may help.
- Drugs to try:
 - Mestinon,

Resolor,

Iberogast,

Erythromycin,

domperidone

Taking medications with gastroparesis

- Recommend crushing all pills, except extended release medications. Patients with Gastroparesis tend to have poor absorption.
- May take sublingual or topical medications where possible.

Pain in the back

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Spinal Instability

- The spine is made up of multiple joints held together by ligaments and muscles.
- Spinal instability with reflex muscle spasms may happen at any level

Spinal Instability

- Thoracic spine subluxations where the ribs meet the spine (costo vertebral joints)
- Lumbar spine subluxations of the facet joints.
- Sacroiliac joint pain (SI Joint) maybe more from uneven posture or pain from joints in the legs
- Kyphosis (spine poking backwards), scoliosis (spine sideways)
- Maybe a symptom of Tethered Cord syndrome

Back pain can be from unstable joints in the spine, causing muscles to spasm.



Spinal pain

- If the pain is from the joints in the spine postural correction, compression garments, muscle strengthening
- Steroid injections not very helpful but can be used in very select cases

Brace for lumbar and Sacroiliac joint



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Tethered Cord Syndrome

Clinical symptoms

- Low back pain
- Neurogenic bladder
- Leg weakness and sensory loss
- Musculoskeletal abnormalities

Pain from Ribs in EDS

Breathing and rib pain

- Patients with EDS often complain of not being able to take a breath in or difficulty breathing
- Pain from ribs subluxing (very painful condition)
- All the tests for heart and lungs are normal.
- Each rib has 3 joints in the back

Breathing and rib pain

- Loss of proprioception from the ribs, muscles of breathing and diaphragm gives a feeling of not having taken a full breath in or full breath out
- Similar to the uncoordinated movement of the rest of the joints in the body
- Some of the muscles of breathing are also part of the lower back

Exercise to help with chest wall pain (and lower back)

- Lie on your back
- Book or sand bag should be over the belly button
- Breathe in and breathe out for 20 minutes a day – lifting the book up



www.NormalBreathing.com

http://www.normalbreathing.com/

Chest wall pain and rib subluxations



- Singing (high and low notes), wind instrument like a flute or recorder
- May help with chest muscle and rib pain <u>and</u> strengthening lower back
- Rib subluxations happen with uncoordinated breathing (remember poor proprioception), and poor posture
- Kinesio taping try



The EDS way of holding a pen

- Poor proprioception makes patients grip a pen with as many fingers as possible
- They hold the pen very tight and press down hard on paper (poor haptic feedback)
- Puts abnormal pressure on the muscles and joints of the hand and wrist



- Dense foam padding (Ableware[®]) or wrap a foam padded tape – for pens, tooth brush, forks, knives
- Compression half finger gloves
- Brace for unstable joints





Splints for fingers



Splinting and braces in general

- Braces maintain joint in neutral position
- Avoid hyper extension
- Braces help with joint position awareness (proprioception)
- Start using them gradually
- Gradually decrease their use as you gain strength

Braces do NOT make muscles weaker



- They do not cause muscle weakness
- It's a common misconception
- Braces are not tight enough to stop muscles from moving
- In fact, braces stabilize joints so the 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



Knees

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Knee pain - causes

- Hyperextension of the knees
- Hypermobile knee cap
- Ankle instability
- Flat feet
- These are almost the same reasons for hip pain and SI joint pain

Treatment options for knee pain in EDS

- First, stabilize the feet and ankles
- Strengthen muscles around the knee
- Knee brace
 - Two straps above knee
 - Two straps below knee
 - Patella stabilizer
 - Metal strut to prevent hyperextension



Knee Gripper brace – wraps around the knee – good for thicker thighs





7 knee strengthening exercises


Kinesio taping – EDS knee

 A combination of two strips of 25 cm in length and 2.5cm in width along the collateral ligament (sides of the knee) using 50% tape tension applied distally (furthest) to proximal, a horizontal tape below the patella 25 cm in length and 2.5 cm in width applied with 25% tension and lastly a Y tape 30 cm in length and 5cm in width cut with 5cm in initial base applied laterally to the patella with no tape tension.



Ther Adv Musculoskelet Dis. 2015 Feb; 7(1): 3–10. doi: 10.1177/1759720X14564561 PMCID: PMC4314299 The effects of neuromuscular taping on gait walking strategy in a patient with joint hypermobility syndrome/Ehlers–Danlos syndrome hypermobility type Filippo Camerota, Manuela Galli, Veronica Cimolin,corresponding author Claudia Celletti, Andrea Ancillao, David Blow, and Giorgio Albertini Author information

Lower leg pain and you keep tripping on your own feet

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Leg pain – often missed cause

- Site of pain from the proximal Tibiofibular joint (a joint close to the knee)
- It can inflame the peroneal nerve which causes pain down the side of the leg and even foot drop



Pain in leg and foot drop

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Pain in knee with foot drop

- Increased pain around the knee with walking
- Tripping over your own toes
- Stabilize ankles
- AFO brace (Spry Step[®])



Strengthening exercises for feet

- Barefoot walking, where safe and comfortable helps with conditioning of muscles under natural loads
- Repeated rising on tip toes strengthens the muscles in foot and with proprioception
- Ankle raises by lifting heel (not leaning forward)
- Descend in a slow controlled way

Footwear - shoes

- Extremely important to wear proper footwear
- Help with unstable ankles, hypermobile feet
- Cushioned mid sole
- Good, strong heel counter provides stability
- Fastenings should be over the mid-sole for better support
- Sneakers !!



Orthotics

- Start using them slowly one hour a day for a few days, two hours a day for a few days.....
- Give your feet a chance to adjust
- Pedag shoe inserts (Check Amazon) or pedagusa.com



Ankle brace to stabilize the ankle joint





Sacroiliac joint pain

- Very common cause of low back pain
- Usually around the lower back, radiates into the buttock region and may even go down the legs (but not below the knee)
- Groin occasionally



SI joint and Hip pain • Leg length discrepancy Longer Leg LLD and Lateral Hip Shift

Brace for lumbar and Sacroiliac joint



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EDS and children

EDS and children

- dislocated joints,
- chronic body wide pain
- Easy bruising
- Abdominal pain
- Fatigue
- Unidentified bleeding (intracranial, abdominal)
- To the ER physicians they look suspicious of Medical Child abuse

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", "ED what?"
"your skin is not very stretchy"

• The diagnosis of Conversion Disorder or Munchausen by Proxy are often made by providers with little training in Psychiatry and vice versa most 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 an academic hospital, chances are that you will be seen by a junior doctor / Resident – who is the least qualified to manage complex medical conditions.
- If you are able, choose a Concierge physician or Private Physician

Oxygen supplementation

- Anecdotal experience
- Oxygen 2-3 liters, 20minutes per day up to twice a day.
- May administer as needed for severe symptoms of fatigue or dizziness.



Exercises for EDS

Keep limb movements within range of motion



Aquatic therapy

- Best form of exercise in EDS
- The contact of water with the skin helps the brain move your muscles more efficiently
- 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



- Muscle strengthening without straining the joints.
- Keep joints within range of motion
- May use braces while exercising to keep joints aligned

• Exercise in water – walking, treading but avoid swimming

Finger print pattern



Kinesio[™] taping - uses

- Reduces pain
- Improves Proprioception
- Relaxes muscles
- Stabilizes joints
- Supports weak joints
- Reduces swelling

How to prevent skin reactions to K-tape

- Cavilon cream[®] by 3M
- Milk of magnesia



Medicinal Marijuana

- Yes, its worth trying it
- Does not worsen MCAS, gastroparesis.
- Equal ratio of CBD and THC
- Avoids the hassles of getting a prescription for pain killers

Opioids for pain

- Reasonable to try for acute flare ups.
- They do worsen MCAS and gastroparesis

Other drugs for pain

- Ibuprofen, naproxen avoid the oral form. Use only in flare ups
- Gabapentin not helpful
- Cymbalta[™] Avoid
- Tylenol[™] safe to take for some pains or in addition to other meds
- Topicals good to try (ibuprofens, cannabis)
- Epsom salt bath or soak very helpful
- Palmitoylethanolamide (PEA) 2% ointment helpful. Sold as Soothamide

Pharmacogenomic testing

- How your body processes medications
- Testing for genetic polymorphism of drug response and drug metabolism, to rule out possible interaction
- Genetic testing for MTHFR mutation
- MTHFR is an enzyme in our body that processes folic acid

Starting treatment- medicines and exercise

Start low, go slow



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,



Low Dose Naltrexone

Low Dose Naltrexone (LDN)

- Dose can vary anywhere between 1.75mg to 4.5mg
- May cause insomnia, mild headaches initially.
- Patients report increased physical activity, flare ups not as acute, better tolerance to pain.
- Recommend a trial of at least 6 months
- To avoid all opioids or tramadol.

Connecting the dots.....

- Fatigue (EDS, POTS, MCAS)
- Diffuse pain (EDS, POTS, MCAS)
- Headaches (EDS, POTS, MCAS, Tethered cord, Chiari malformation)
- Joint pain (EDS, Tethered cord)
- Dizziness (POTS, Chiari malformation)
- Poor balance (EDS, POTS, Tethered cord, Chiari malformation)
- Gl issues (EDS, POTS, MCAS)
- Feeling anxious (POTS, MCAS)

Resources

<u>http://www.chronicpainpartners.com/webinars/</u>

• Excellent videos from speakers on EDS
Thank you

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