Pain management in Ehlers Danlos Syndrome – 2015

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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.
Introduction

• Training and Fellowship, Harvard Medical school
• Pain Medicine specialist
• Assistant Professor – Brown Medical School, Rhode Island
• Special interest in complex pain conditions
• A typical evaluation with me takes anywhere between 5 hours to 6 hours.
• I am going to try my best to squeeze all that in 1 hour
Types of pain in EDS
Types of pain in EDS

- Mechanical pain or structural pain (Nociceptive pain)
- Inflammatory pain
- Nerve pain
- Muscle pain.
- Headaches
Joint sense

Proprioception
The body’s ability to sense movement of the joints and their position
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 over extending and our muscles from over stretching.
• EDS – poor proprioception. Klutzy 😇
Proprioception

• Really important to work on improving proprioception.
• EDS’ers learn to protect and avoid injury to their joints properly by not overextending them.
Proprioception exercises

• Juggling
• Balance board or wobble board
• Stork standing (stand on one leg)
• Stand up paddle board (SUP)
• Sitting on exercise ball
• Exercise in water – walking, treading but NO swimming
Compression garments, taping

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Neuromuscular taping (Kinesio™)
Kinesio™ taping - mechanism

• Mimics the superficial layer of skin – after 10 minutes you can not feel it.
• Designed to stretch.
• Porous – allows for drying easily. You can take a shower with it on.
• The adhesive is applied in a wave like pattern to mimic the qualities of fingerprints.
Finger print pattern
Kinesio™ taping - mechanism

• The tape stimulates the sensors in the skin as we move – improves proprioception

• Helps reduce swelling
Kinesio™ taping - uses

- Reduces pain
- Improves movement patterns
- Relaxes muscles
- Stabilizes joints
- Supports weak joints
- Reduces swelling
Kinesio taping helpful for

- Neck
- Upper back
- Lower back – SI joints, muscles
- Wrist
- Shoulders
- Knees
- Ankles and feet
Kinesio taping – EDS knee

- A combination of two strips of 25 cm in length and 2.5 cm 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 5 cm in width cut with 5 cm in initial base applied laterally to the patella with no tape tension.

Taping for lower back in EDS

- Two tapes of 25 cm in length and 5 cm width applied with no tension to the tape, laterally to the spine from the intergluteal fold to the last dorsal vertebra with the patient maintain a lumbar flexion at 45 degrees in a standing position during the complete application.

Headaches and neck pain in EDS
Headaches and neck pain

• Headaches may be caused by neck pain
• Headaches may be from
  – inside the head (Migraines) or
  – outside (chronic daily headaches or tension type headaches)
• Treatment for both is different
Most headaches are caused by muscle contraction or blood flow problems.
Causes of headaches 1

1. Arnold Chiari malformation
2. Cervicogenic HA – from muscles
3. TMJ dysfunction (aka Craniofacial pain)
4. Vision – blurry
5. POTS / Dysautonomia
6. Tethered Cord syndrome
7. Trigeminal neuralgia
8. Spontaneous CSF (Cerebrospinal) leak
Causes of headaches 2

7. Instability of the neck – Cranio Cervical Instability
8. Atlanto – axial hypermobility
9. Cervical facet joint arthritis (joints in the spine)
10. Occipital neuralgia (nerves in the back of the head)
11. Sinusitis
Chiari malformation.....connecting

• Lower brainstem symptoms:
  – Nausea
  – Dysphagia
  – Throat tightness
  – Sleep apnea
  – Shortness of breath
  – Palpitations
  – Face pain
  – Double vision
  – Syncope (fainting)
TMJ Pain

Temporo Mandibular Joint Dysfunction
TMJ Pain

- Very closely related to neck issues
- Clicking noises
- Clenching, grinding
- Pain with chewing
- Difficulty opening mouth wide (eating an apple)
- Jaw locking up
Trigger points for TM Joint pain

A  Sternal division

B  Clavicular division

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Temporo-mandibular joint dysfunction (TMJ)

- Present in 70%
- Treatment: avoid excessive mouth opening, caution when yawning,
- Orthodontist specializing in TMJ
- Avoid over the counter mouth guards
Kinesio taping for TMJ
Neck pain
Neck pain and headaches

- The most common cause of neck pain is posture
- Chin poking forward position
- Correction is easy.
- Before looking at other reasons, correct this first
- There are other reasons like cervical instability, Chiari malformation etc – these need to be addressed
Poking chin posture

Good Posture
- Correct Head Posture
- Square Shoulders
- Centre of gravity of the body/S2
- Level Pelvis
- Sacrum

Poor Posture
- Headaches
- Neck Pain
- Back Pain
Head forward or poking chin posture

- This posture can cause pain in the jaws (TMJ), face, upper back, neck
- Blurry vision (POTS, EDS, Chiari)
- Laptops
- Brain Fog (POTS, Chiari, MCAS)
Poking chin posture / Forward head position

- Shortens the muscles in the back of the head and neck
- Weakness of muscles in front of neck (flexors)
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

• Post it note on monitor to remind you

• Vision correction

• Manage POTS
Different types of taping for neck pain and headaches.
Wrist and fingers
Muscles of the hand

• The small muscles of the hand are stressed to compensate for hypermobility

• Small intrinsic muscles fatigue easily – short, pain free sessions
The EDS way of holding a pen

- Poor proprioception makes EDS’ers grip a pen with as many fingers as possible
- They hold the pen very tight
- Puts abnormal pressure on the muscles and joints of the hand and wrist
Hands and wrist pain

- Damage to the 1st thumb joint (1st CMC joint)
- Hold pen too tightly (poor proprioception)
- Fingerless compression glove
- Foam padded pen (Ableware™)
- Splinting for subluxations
- Please do not voluntary sublux fingers and thumbs to show off
• Dense foam padding (Ableware®) or wrap a foam padded tape

• Fat pens are not helpful

• Compression half finger gloves
Thumb pain

- It’s the 1st CMC joint
- Hyperextension of this joint is common
- Splint should stabilize the joint but not cross the wrist crease (so as not to interfere with wrist movements)
Muscles of the hand – proprioception training

• Weight bearing in neutral position – eyes open, eyes closed
• May take months but does make the joints more stable and ligaments taut
• Goal should be to decrease fatigue and increase proprioception – wrap coban™, foam pad etc. for pens. But retrain to apply appropriate amounts of pressure.
Splinting and braces in general

- Braces maintain joint in neutral position
- Avoid hyper – extension
- Braces help with joint position awareness (proprioception)
- Gradually decrease their use as you gain strength
- Kinesio taping is a good option
Breathing difficulties in EDS

- May feel like running out of breath, need to take deeper breaths, working to breathe
- It almost feels like one has to take a breath voluntarily
- There are proprioceptor receptors in the muscles of the chest, joints of the ribs.
- Compression shirt
- Taping over the ribs.
Compression garment for breathing difficulties in EDS
Feet and ankles in EDS
Pain in lower half of the body

• Pay attention to feet and ankles.
• 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 off – Sacroiliac joint pain, lumbar pain
Flexible flat feet – common in EDS
The feet in EDS

• 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

- Custom made orthotics
- Walk Fit™
- Start using them slowly – one hour a day, two hours a day.....
- Give your feet a chance to adjust
Kinesio taping for arch of foot
Knees in EDS
Knee - patella

• The knee cap is held in place by a fine balance of thigh muscle forces

• Muscle imbalance around hips, pelvis, abnormal gait can cause pain around the patella
• Area of pain from a Hypermobile patella

http://www.mygeofit.com/member/Front-Thigh-(Quadricep)-Exercises.html
Knees - hyperextension

- Hyper-extension of knees
- Hypermobile patella – unstable knee
- Strains the ligaments inside the knee
- Keep knee in mid-range – avoid hyper-extending the knee
- Compression knee sleeve or tights
- Kinesio tape
Hyperextension at the knees
Hyperextension at the knees

- Avoid hyperextending the knees when standing – ‘break’ your knees slightly
- Compression knee sleeve or tights
- Kinesio tape
Compression knee sleeve

- Protects and stabilizes the knee joint by improving proprioception (joint sense)
EDS and POTS

• For patients with POTS, a full length compression tights

• POTS – Postural Orthostatic Tachycardia Syndrome
Kinesio taping for the knee
Knee pain – often missed cause

- The proximal tibio-fibular joint is on the outside of the knee.
- Like all joints it is prone to subluxations or arthritis.
- Subluxations are more common in athletes.
- A subluxing PTF joint affects the Peroneal nerve, which affects the side of the leg and causes a foot drop.
Knee pain - Proximal Tibiofibular joint (PTF)
Knee pain – often missed cause

- Site of pain from the proximal Tibiofibular joint
- It can inflame the peroneal nerve which causes pain down the side of the leg and even foot drop

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The TiboFibular joint – proximal and distal

- Pain along the lateral aspect of the leg below the knee
- Knee pain, especially with squatting
- Pain in thigh (Ilio-tibial tract)
- Foot drop – may be present
- May mimic DVT
Ilio Tibila band (Fascia Lata Fascitis)

• The IT band is a very common cause of pain in Ehlers Danlos Syndrome
• Pain along the side of the thigh up to the knee.
• Worsened if the tibiofibular joint is unstable.
Shoulders and upper back
Shoulders in EDS

• Quality of proprioception decreased
• One of the weakest joints in the body
• Not much pain from dislocation
• Pain due to
  – muscle fatigue,
  – surfaces of the shoulder rubbing against each other,
  – impingement
Shoulders in EDS

- Intense pain from muscle spasms
- Be assured that is if the shoulder is displaced because of abnormal muscle control, there is unlikely to be any harm to the structures
- Fear of not moving the shoulder will cause more harm
Shoulders in EDS – Initial step

• Please do not voluntarily dislocate the muscle.
• Improve postural awareness and correction
  – Sitting on a balance Swiss ball
  – Holding back flat against the wall
  – Balance board
  – Balance on one leg
• Core strengthening (also improves posture and abnormal muscle tone)
Shoulders in EDS – muscles to work on

- Pectoralis (‘ pecs ’)
- Anterior deltoid
- Infraspinatus
Compression garments, taping

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Shoulders in EDS

• Avoid swimming or exercises that involve moving shoulders to extreme positions (ballet)
• Do not do many exercises at once. Do one or two at once.
• Keep them simple, so they can be done throughout the day.
Upper back pain

• Posture, posture, posture

• Correct the ‘chin poking forward’ position.
  (finger to the chin and push back)
Posture correction with Kinesio tape

Upper back pain in women with EDS

- Sports bra with:
  - racer back (cross straps).
  - Wide straps.
  - Front closure
- Proper fitting – recommend getting it done professionally.
- May have to consider reduction mammoplasty in severe intractable upper back pain

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Thoracic Outlet Syndrome

- Fairly common in EDS.
- Pain in shoulder, and arm
- It maybe because of the collar bone subluxing
- It may be because of muscle spasms
- Nerves (commonly) and blood vessels get pinched at the 1\textsuperscript{st} rib.
Thoracic Outlet Syndrome

Scalenes
This muscle connects your neck to your ribs.

Neurovascular Bundle
If your Scalenes and/or Pec Minor muscles are tight, it may create pressure onto this Neurovascular Bundle, causing pain that leads down to the arm or hand.

Pec Minor
This muscle connects your shoulder blade to your ribs.

www.sgergo.com
Pain patterns in Thoracic Outlet syndrome

http://mathewhawkesphysiotherapy.blogspot.com/2013/05/thoracic-outlet-syndrome.html
Thoracic Outlet Syndrome

- Physical therapy
- Trigger point injections / dry needling with PT
- Kinesio taping
- Surgical correction
Prevention
Prevention

- EDS’ers are more prone to tissue damage even with a minor injury
- Poor wound healing contributes to long standing issues
- Even when it does heal the injuries (especially to joints and ligaments) are not strong enough
Preventing pain

- Do not stand on one hip or sway at hips
- Do not stand on the outside of your feet
- Do not sleep on your stomach, head turned to one side for a prolonged period
Preventing pain

• Do not sit with legs outstretched, or with legs tucked under the buttocks ‘W’ position
Preventing pain

- Do not sit kneeling with buttocks resting on ankles
Preventing Pain – working at home

• Avoid repetitive activities – vacuuming, raking, filling dish washer, stirring. Change activity frequently.

• Work surface (kitchen counter, sink etc.) should be at appropriate height. Use blocks to raise your work surface
Preventing pain - shopping

• Do not carry a heavy bag hanging from a shoulder
• Do not carry grocery bags in hand, especially heavy ones. Keep them light.
• Use shopping trolley. Use wheels wherever you can
• Keep handbags light
Preventing pain – going to the dentist

- EDS’ers wind up making more trips to the dentist
- Talk to dentist about EDS
- Numbing medicine may not work or you maybe too sensitive to them
- TMJ – not to keep mouth open too long
- Position of neck – prefer being in a more flat position with neck supported

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POTS – Postural Orthostatic Tachycardia syndrome

Dysautonomia
POTS - Postural Orthostatic Tachycardia syndrome

- Increase in heart rate by 30 beats/min within 10 minutes of standing
- Standing heart rate of 120 beats/min
- No significant change in blood pressure
- Syncope or almost syncope (fainting)
POTS - Postural Orthostatic Tachycardia syndrome

- Brain fog
- Digestive problems – nausea, gastroparesis (slowing of the intestines)
- Chronic fatigue
- Headaches
- Blurred vision
POTS - tests

• Orthostatics - preferred
• Tilt table test - not a fan of this test
Treatment of POTS

– Increase oral salts for Postural Orthostatic Tachycardia Syndrome (POTS).
– Increase oral electrolyte fluids
– Compression tights up to thighs.
– Abdominal binder.
– Cardiology or Neurology consult for Dysautonomia/POTS.
– Consider a beta-blocker such as Betaxolol or propranolol at night for Postural Orthostatic Tachycardia Syndrome and non-restorative sleep.
POTS - Postural Orthostatic Tachycardia syndrome

Consult Dysautonomia International for more information and recipes.

http://www.dysautonomiainternational.org/
Starting treatment - medicines and exercise

Start low, go slow
Mast Cell Activation Syndrome

MCAS
Mast cells

- Cells in blood
- Normally present in blood
- Contain histamine
- Involved in allergy, wound healing and protection against infection
Mast Cell Activation Syndrome (MCAS)

- Flushing
- Itching
- Unexplained gastrointestinal disturbance especially gastroparesis (slowing of the intestines)
- Unexplained fluctuations in blood pressure
Mast Cell Activation Syndrome (MCAS)

- Nausea, vomiting,
- stomach pains,
- slowing of the gastrointestinal tract (gastroparesis)
- Low blood pressure
- Syncope or near syncope
- Chronic fatigue, headaches
Mast Cell Activation Syndrome (MCAS)

- Temperature instability – hot / cold
- Multiple chemical sensitivities – food, drugs,
- Sensitivities to multiple drugs maybe due to fillers – changing to a different brand may help
- Dry eyes, difficulty focusing,
- Hair loss
- Bladder pain: Interstitial cystitis – inflammation of bladder
Mast Cell Activation Disorder (MCAD)

- In EDS, mast cells are normal in count (not increased)
- Hyper-reactive
- May not have a specific trigger that makes the mast cells release histamine
- Brain fog
- Sleep apnea
Management of MCAS

• Anti-histamine blockers:
  – Most cold medicines
  – Ranitidine, famotidine

• Cromolyn

• Low histamine diet

• avoid: sudden change in temperature, certain seasonings (except olive oil and salt)
NC10 rule

Expectations from different therapies
NC10 rule
NC10 rule
NC10 rule
NC10 rule
NC10 rule
Muscle pain

Myofascial pain
Muscle pain

• Often more painful than the original pain

• Muscles may tighten reflexively, guarding of a painful area, nerve irritation or generalized tension
Treatment of muscle pain

• Treat the cause – joint pain, repetitive use

• Trigger point injections / Dry Needling

• Kinesio taping

• Stretching – very, really very gentle stretches.
Trigger Points

• When muscles are in chronic spasm they stay taut – ‘knots’.

• They develop trigger points which trigger the muscle in to a constant state of spasm

• The trigger points can be painful when touched.

• The pain can spread throughout the affected muscle and other parts
Trigger Points
Trigger Point injection
Dry Needling
Manual Techniques

• Myofascial release: Gentle techniques to improve the mobility of the muscles by mobilizing the tissue around the muscle

• Muscle energy technique: Utilizes positioning of the body and using active muscle contraction to relax muscles and align joints
Muscle Relaxants

- Try to avoid using them. Use only if having a flare up
- Cyclobenzaprine (Flexeril)
- Carisoprodol (Soma)
- Tizanidine (Zanaflex)
- Baclofen
- Benzodiazepine (Valium)
Nerve pain

Neuropathic pain
Nerve pain in Connective tissue disorder / EDS

- Nerve connective tissue is more fragile
- Fragile nerves get over stretched when crossing or associated with hypermobile joints
- Neuropathic pain, Complex Regional Pain Syndrome (CRPS) or Reflex Sympathetic Dystrophy (RSD)
What is CRPS / RSD

- Complex Regional Pain Syndrome
  formerly Reflex Sympathetic Dystrophy
- Syndrome characterized by a continuing pain that is disproportionate to the usual course of any trauma or lesion.
- Usually starts after a trauma, immobilization.
Pain

- The pain is far more than one would expect for the type of injury
- Pain to soft touch
- It may spread to other parts
- One limb may have a different color or temperature
Management of CRPS /RSD

• Much too elaborate for this talk

• For a presentation on CRPS, please visit www.rsdso.org
Caution
Orthopedic surgery

• Surgery for stabilization of joints, tendon relocation, arthroplasty etc – caution, caution
• High risk of poor healing, recurrence, adhesions, Complex regional Pain Syndrome
• Must plan carefully. Consider a surgeon who has experience with Ehlers Danlos Syndrome
Surgery in EDS - suggestions

• Surgical wounds should be closed without tension, preferably in two layers.
• Deep stitches should be applied generously and closely.
• Cutaneous stitches should be left in place twice as long and additional fixation of adjacent skin with adhesive tape can help prevent stretching of the scar.
Caution

• Bed rest, periods of inactivity, avoiding exercise. Ehlers Danlos Syndrome patients decondition rapidly
Low Dose Naltrexone (LDN)

• Naltrexone is an old drug that was used to treat addiction
• When taken in a very very low dose, has been shown to help with chronic neuropathic pain, which is part of the pain in EDS.
LDN and EDS

• Good experience with using Low dose Naltrexone in EDS
• Helps pain
• Helps fatigue
• Helps most symptoms of EDS
Opioids

• Mild doses for a short term are good for acute pain
• Not very effective in Ehlers Danlos Syndrome
• Risk of increasing Mast Cell Activation Syndrome
Cannabis – good option for EDS

- Good news – it works better than narcotics in Ehlers Danlos Syndrome
- Bad news – not FDA approved, regulations
- Vaporize, topical, tincture
- Pain – low THC content, higher CBD content
- Low addiction potential.
- Helps with pain, nausea and gastroparesis
- Does not affect Mast Cell Activation Syndrome.
Avoid

- Avoid high impact sports/activities
- Avoid low environmental temperatures
- Avoid prolonged sitting positions and prolonged recumbency
- Avoid sudden head-up postural change
- Avoid excessive weight lifting/carrying

Gluten free diet

• Gluten as a protein can cause an inflammatory response in the body.
• Migraines, chronic body ache, abdominal pain, hypermobility syndromes (EDS), multiple joint pains
• Hold off on gluten foods for 8 weeks to see if it makes a difference.
Starting treatment - medicines and exercise

Start low, go slow
Service Dogs

• Trained to each person’s physical impairments
• help with functioning and independence
• Constant companion, will often sense its owners pain and will comfort them both physically and emotionally
• Can sense distress and call for help
• Service dogs give patients a feeling of security allowing them to be more active physically and socially
• Provide stability while walking, open and close doors, switch on and off lights
Service Dogs

• 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.
Starting treatment - medicines and exercise

Start very very low, go slow
Thank you

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