Connecting the dots with EDS, POTS, MCAS, GI, Neurological and Physiotherapy in managing pain in EDS

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Disclosure and disclaimer

- I have no actual or potential conflict of interest in relation to this presentation or program.
- I have no financial interest in any of the products mentioned in this presentation

- This presentation will discuss “off-label” uses of medications

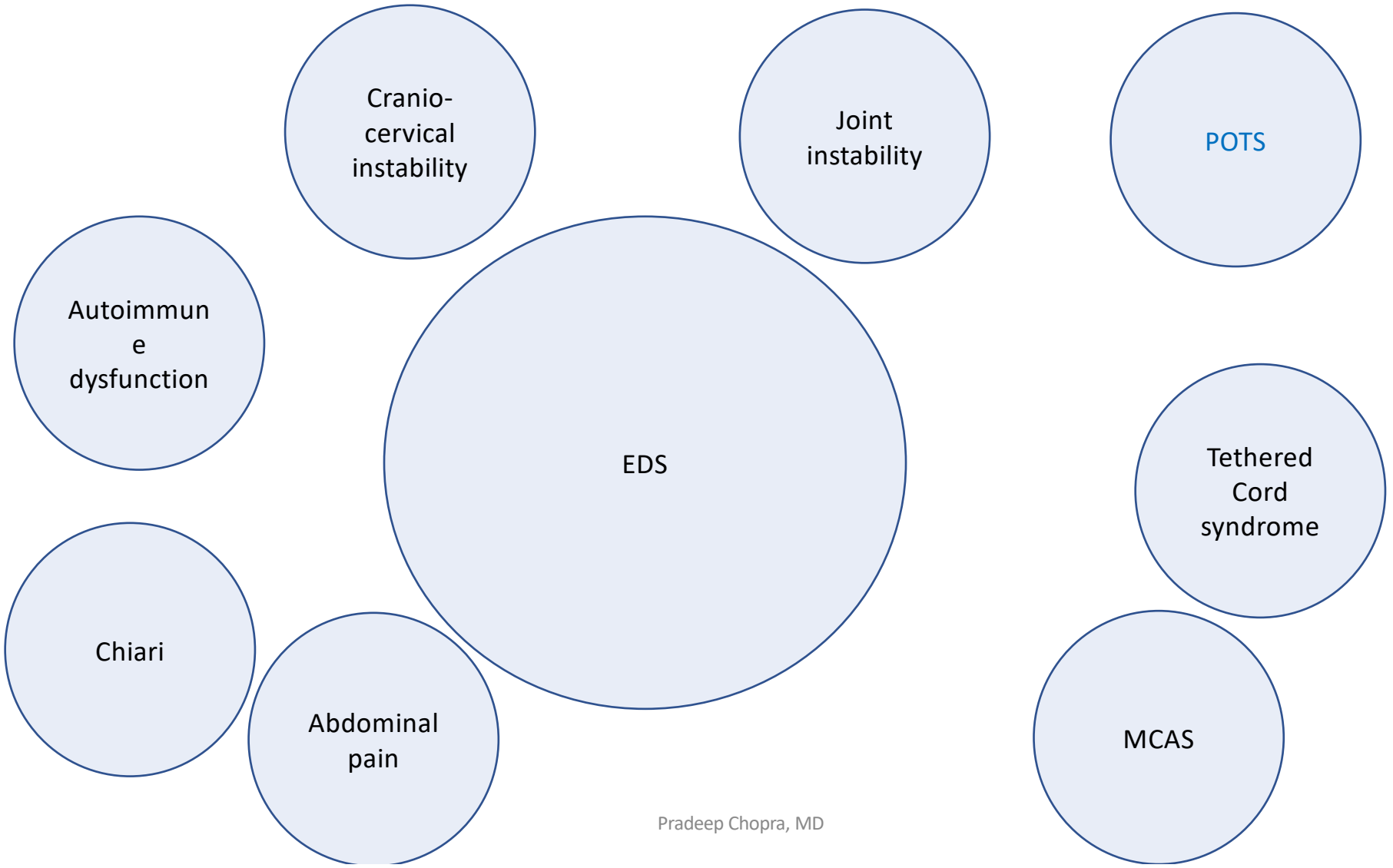
- Discussions in this presentation are for a general information purposes only. Please discuss with your physician your own particular treatment. This presentation or discussion is NOT meant to take the place of your doctor.

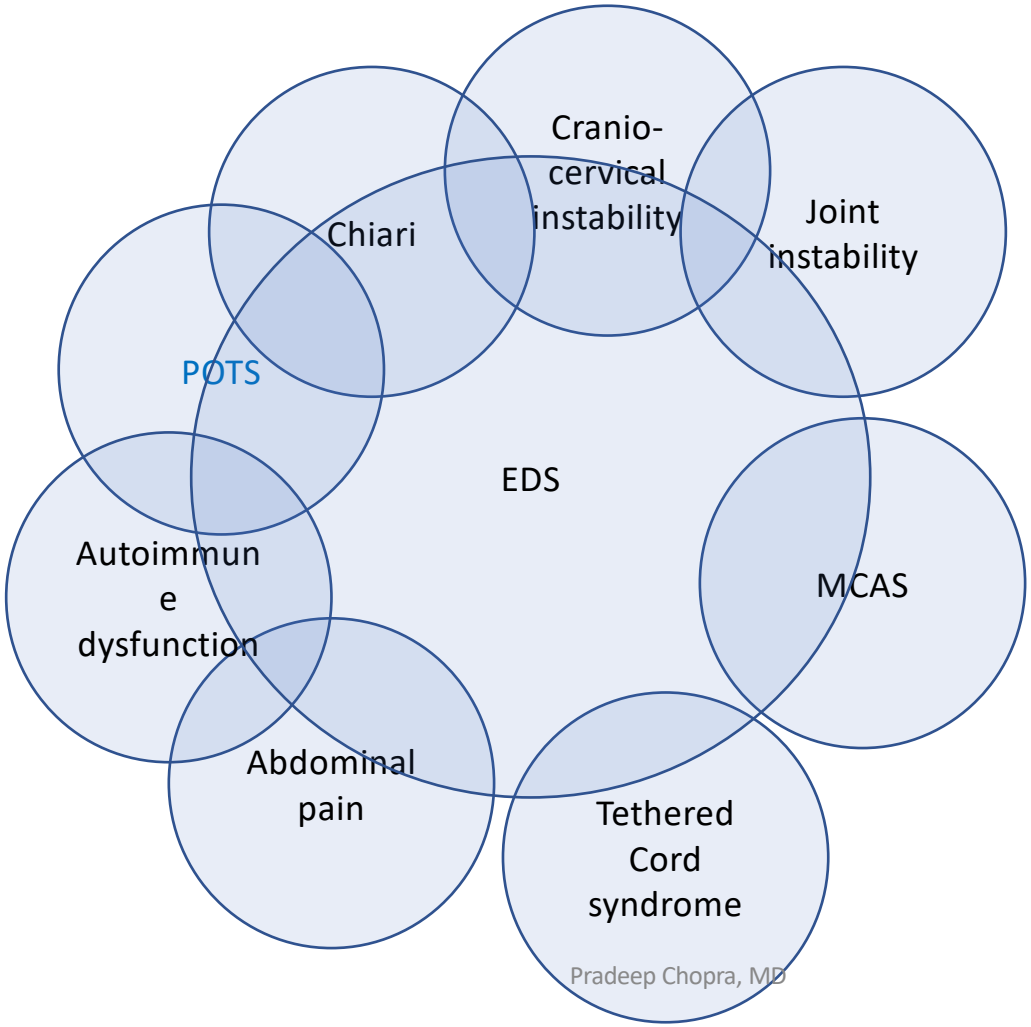
Disclosure

- Training and Fellowship, Harvard Medical school
- Weird Pain Medicine specialist
- Assistant Professor – Brown Medical School, Rhode Island, USA
- Special interest in complex pain conditions

Connecting the dots

- EDS is a complex condition
- This presentation is about connecting the dots between all the symptoms that come with having EDS.


Cranio-cervical instability
Joint instability
POTS
Autoimmune dysfunction
EDS
Tethered Cord syndrome
Chiari
Abdominal pain
MCAS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Tethered
Cord
syndrome

Three things to know in EDS

1. EDS is not a disease. It is a form of the human body.
2. Soft connective tissue
3. Poor joint position sense (proprioception)
4. Co-existing conditions (Mast cell activation, POTS, Chiari etc.,)

Pain in EDS by body regions

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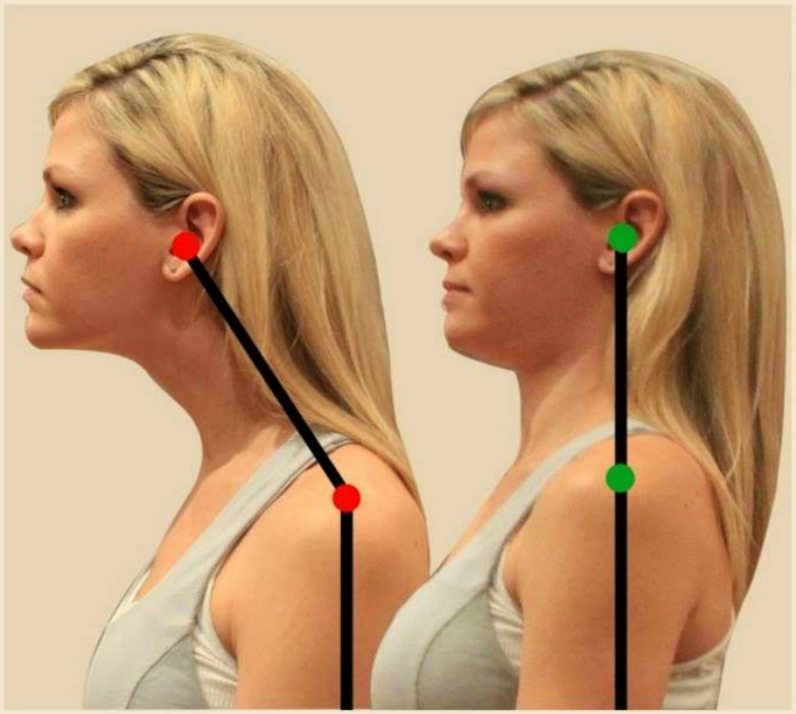
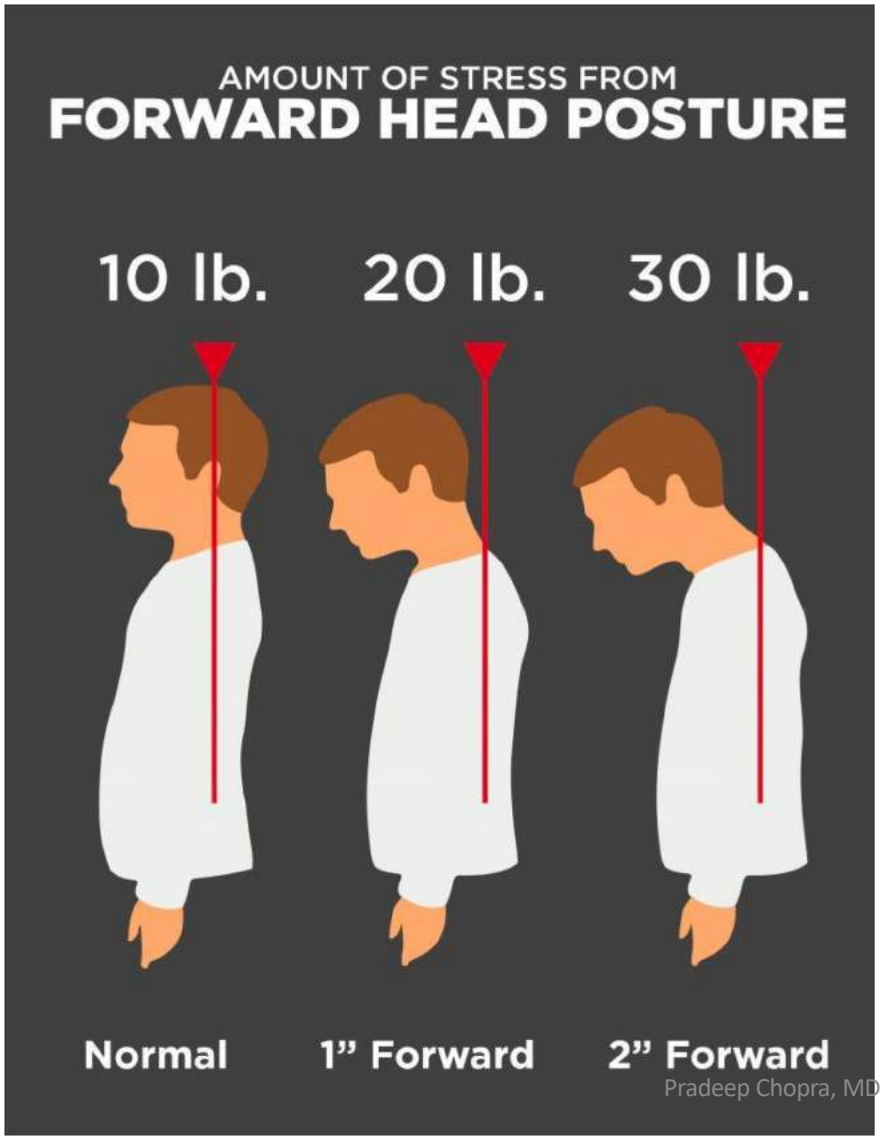
Common causes of headaches in EDS

1. Migraines
2. Chiari malformation
3. Cervicogenic Headaches – from muscles
4. Temporo Mandibular joint dysfunction (TMJ)
5. PoTS / Dysautonomia
7. Spontaneous CSF (Cerebrospinal) leak – low pressure inside the head
8. Cranio Cervical Instability (Instability of the neck and head)
9. Idiopathic Intracranial Hypertension (raised pressure inside the head)

“My head feels too heavy to hold up”

Chin poking forward position



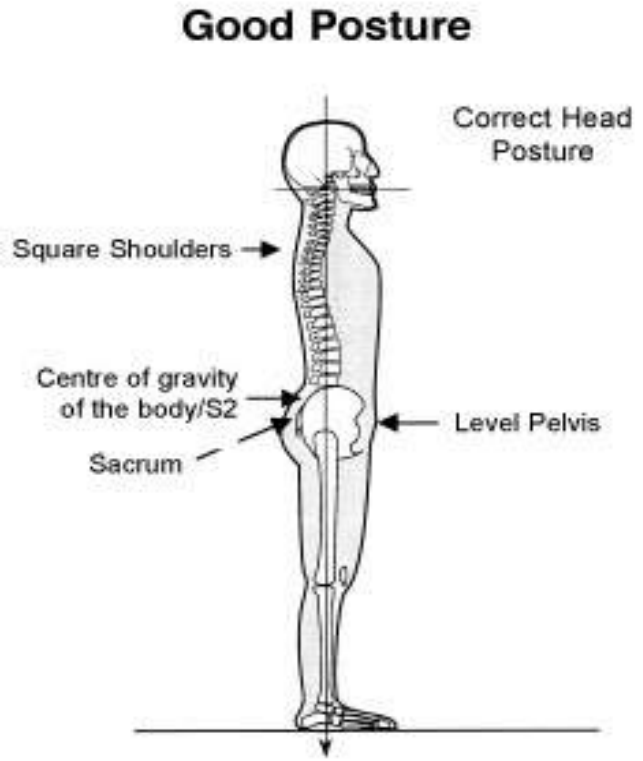


https://www.braceability.com/blogs/articles/text-neck-how-smartphones-hurt-your-spine

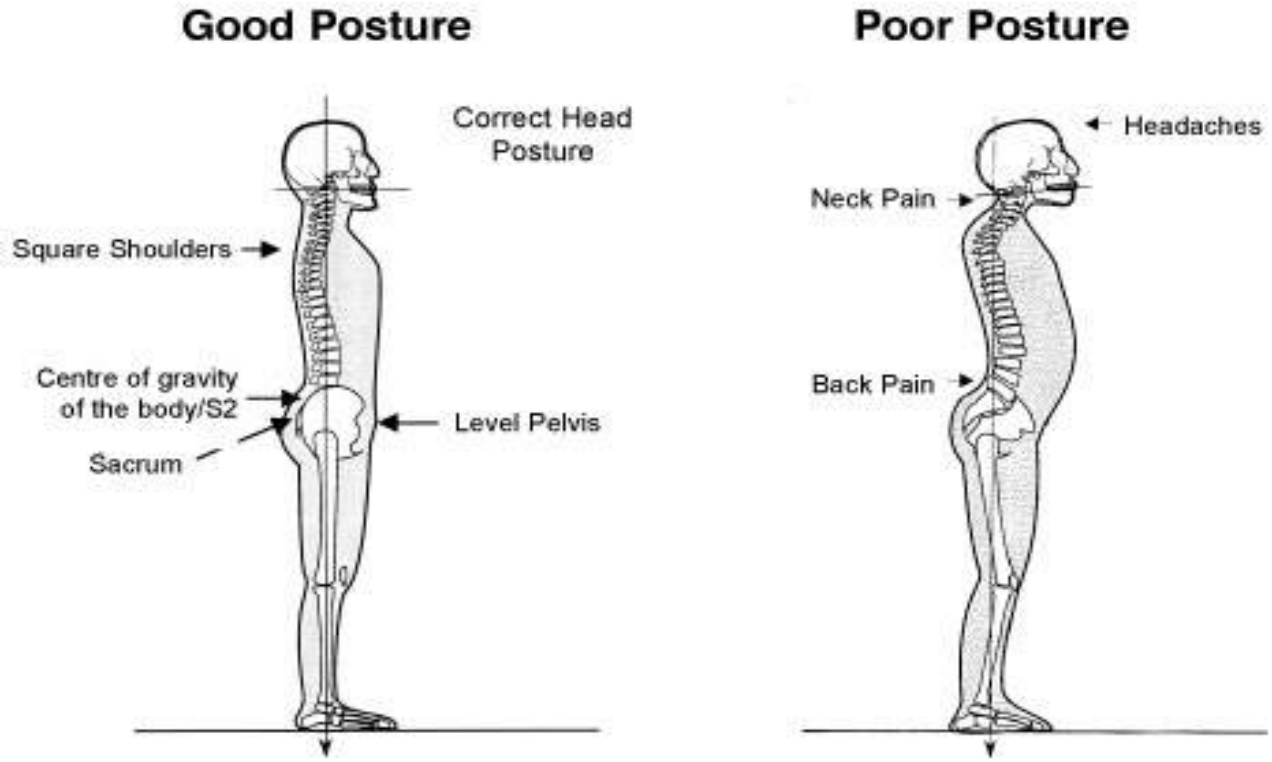
Neck pain and headaches

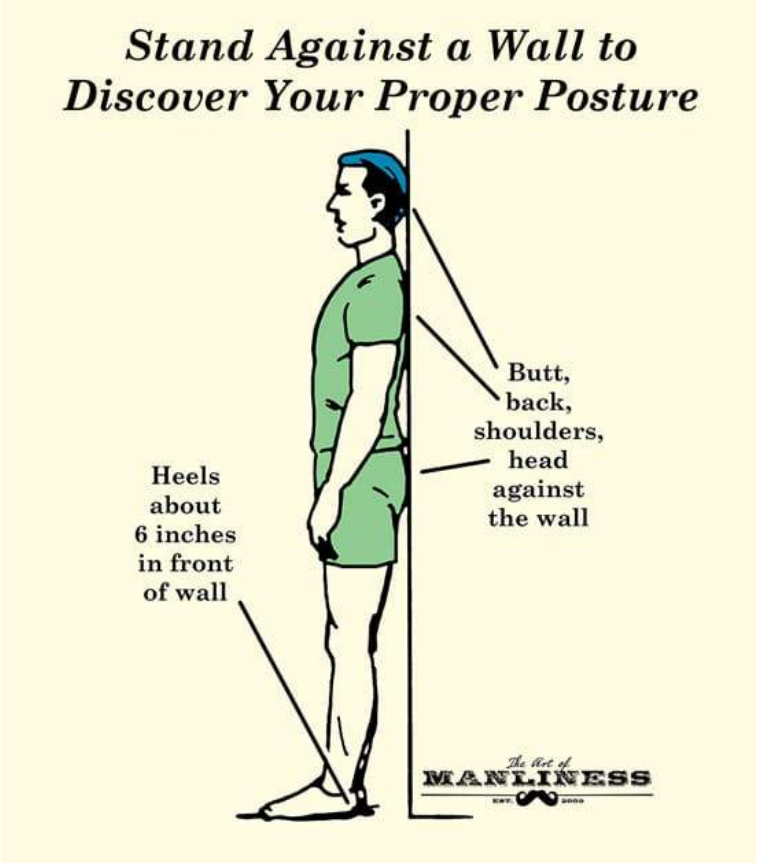
- A common cause of neck pain is posture, using a smart phone (“Texting neck”)
- 'Chin poking forward position'
- Before looking at other reasons, correct this first
- If there are other reasons like cervical instability, Chiari malformation etc – these need to be addressed

Pain from a poor posture



Pain from a poor posture




Stand Against a Wall to Discover Your Proper Posture
Butt, back, shoulders, head against the wall
Heels about 6 inches in front of wall
The Art of MANLINESS

Common reasons for poor posture in EDS

- Vision – Blurry vision. Usually intermittent
- Postural Orthostatic Intolerance (POTS)
- Laxity of spinal ligaments
- 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
- Manage POTS

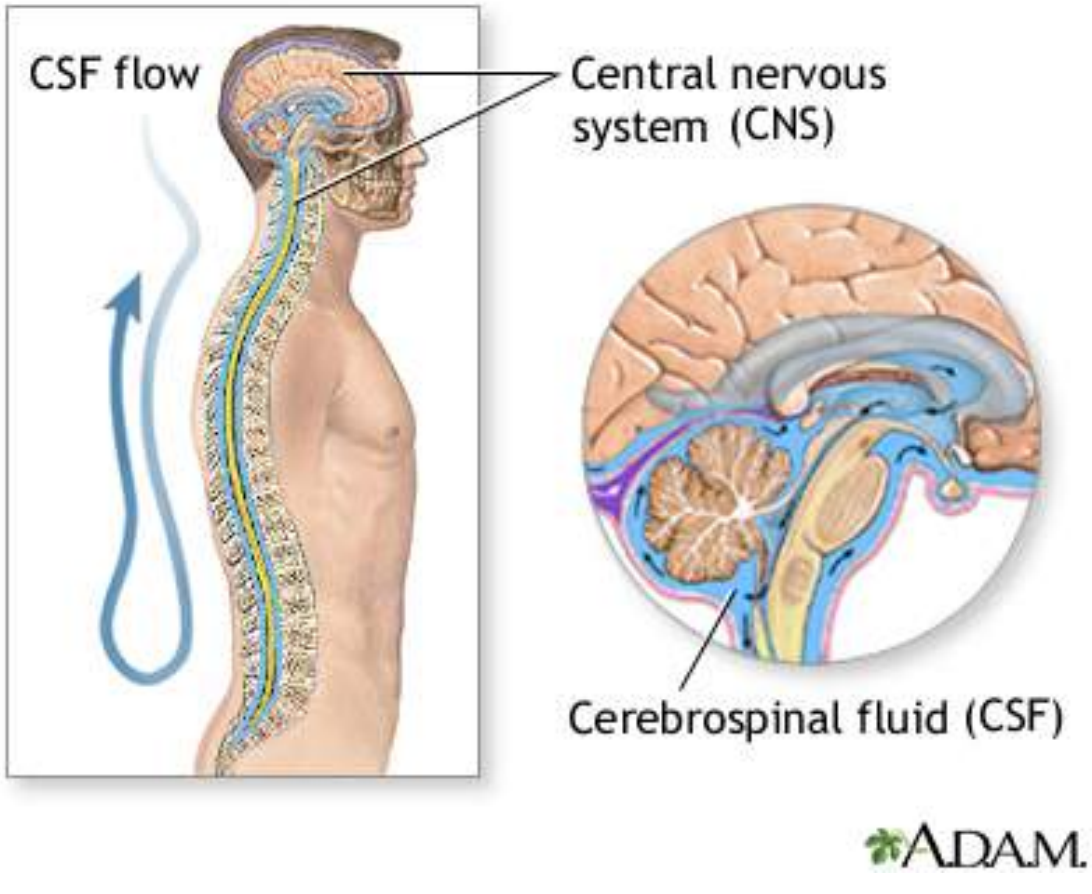
“My whole head hurts,
I see double,
I can feel my heart beat in my ears, ”

Headaches – Idiopathic Intracranial Hypertension (IIH)

- Raised pressure inside the head
- Vision problems – double vision, sensitivity to light
- Ringing in the ears which pulses (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



Spontaneous CSF leak

- Headaches get worse being upright
- Almost resolve with lying supine
- Headaches worse by the end of the day
- Severe headache all day in long standing cases
- Nausea and vomiting
- Tinnitus – change in CSF pressure can cause change in inner ear pressure
- Neck pain

Differential diagnosis

POTS: Postural Orthostatic Tachycardia Syndrome

	CSF leak	POTS
Headache	+	+
Headache worse upright	+	+
Headache better supine	+	± Not significantly better
Neck pain	+	+
Fatigue	+	+
Nausea	+	+
Orthostatics	-	+ Heart rate greater than 30BPM with no change in BP
Dizziness	-	+

CSF leak - managements

- Increase PO fluids and caffeine
- Abdominal binder
- Epidural blood patch – high volume. May need multiple patches
- Directed epidural fibrin glue
- Surgical repair

Schievink WI, Gordon OK, Tourje J: Connective Tissue Disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension: A prospective study. Neurosurgery 54:65-71, 2004

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

Spontaneous CSF leak

“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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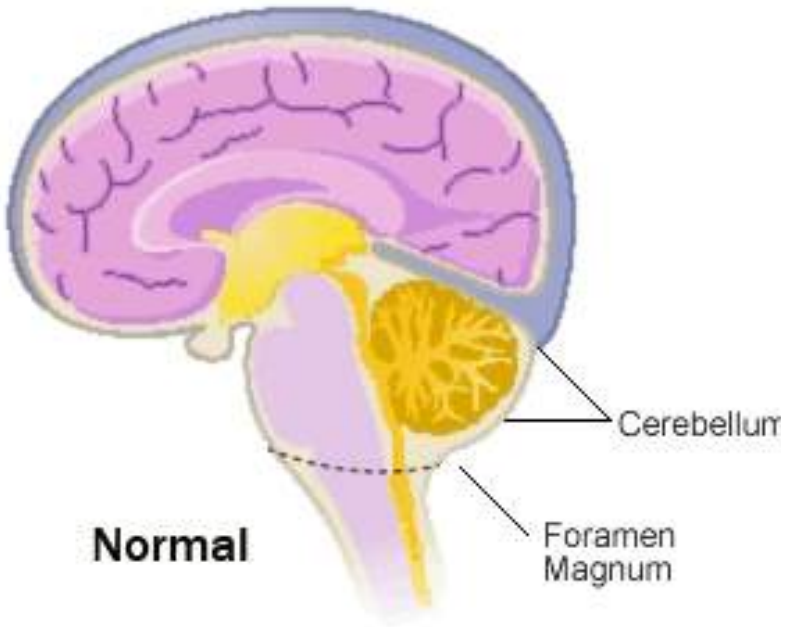
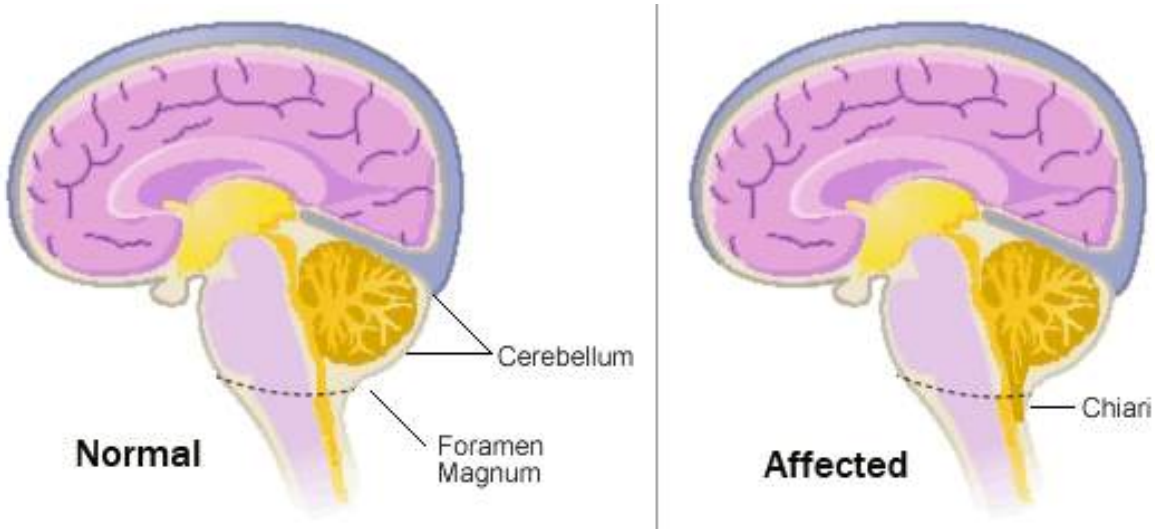
Chiari malformation



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

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**
- Pressure headaches in the back of the head (suboccipital)
- Neck pain
- Balance problems
- Numbness or paresthesia's to arms or legs
- Dizziness
- Difficulty swallowing
- Poor Hand co-ordination
- Headaches made worse by coughing or straining

- Ringing or buzzing in the ears
- Hearing loss
- Nausea, vomiting
- Muscle weakness
- Vision problems

Chiari Malformation and EDS

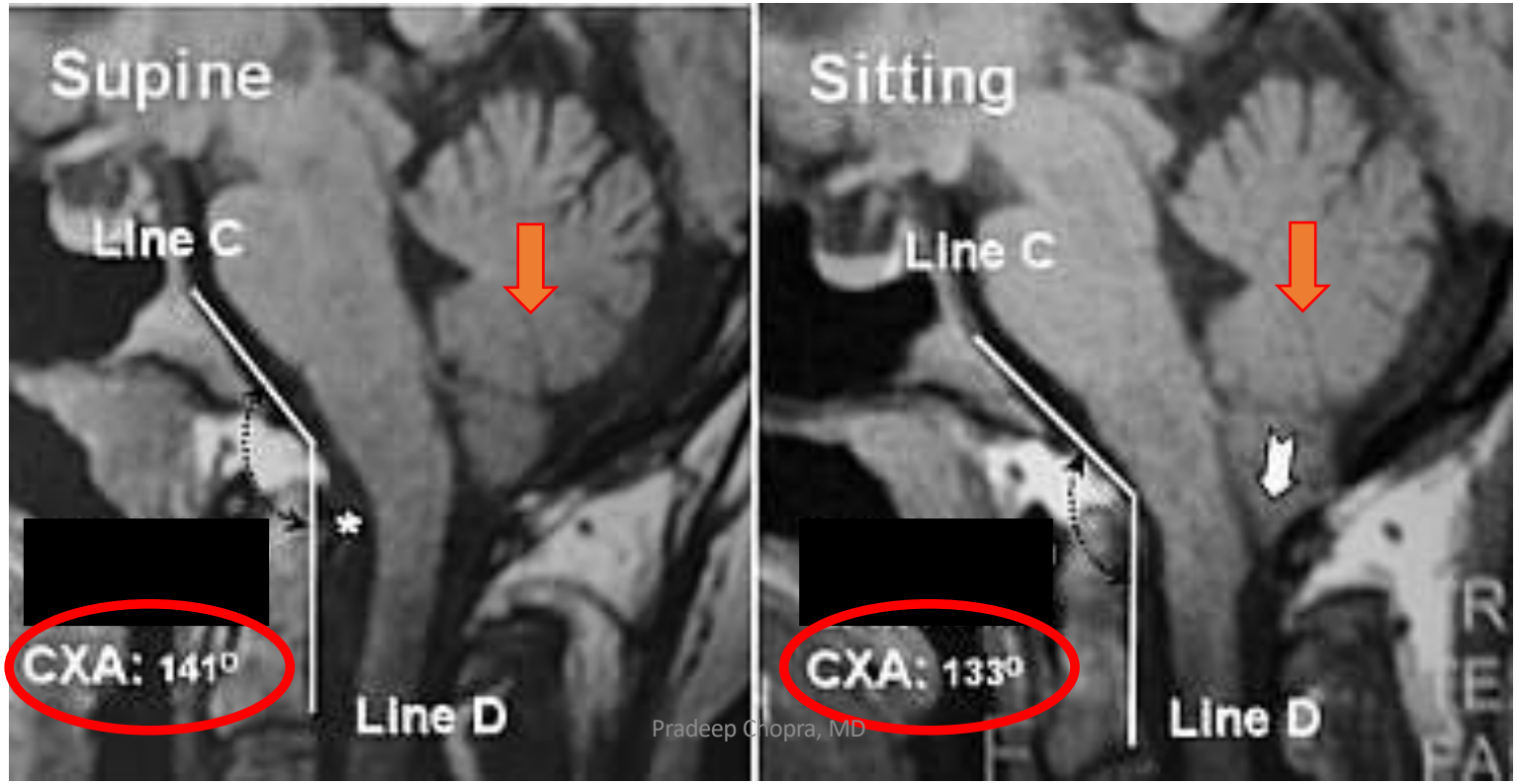
A diagnosis of Chiari Malformation depends on:

1. Good history and physical examination

2. Upright MRI of the neck (cranial settling in the upright position)

Cranial Settling in EDS- Deformative stresses on the brain stem, lower cranial nerves, spinal cord

Clivo-axial angle normal 140^0



Chiari malformation

- The herniation causes obstruction to CSF flow.
- Empty Sella syndrome – flattening of the pituitary gland and resulting hormonal changes.

- There may be an association between Chiari, Craniocervical instability and Tethered cord syndrome in EDS

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

Chiari Malformation

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
- 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

- Need functional imaging technology
- Static pictures are not helpful
- 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 fusion

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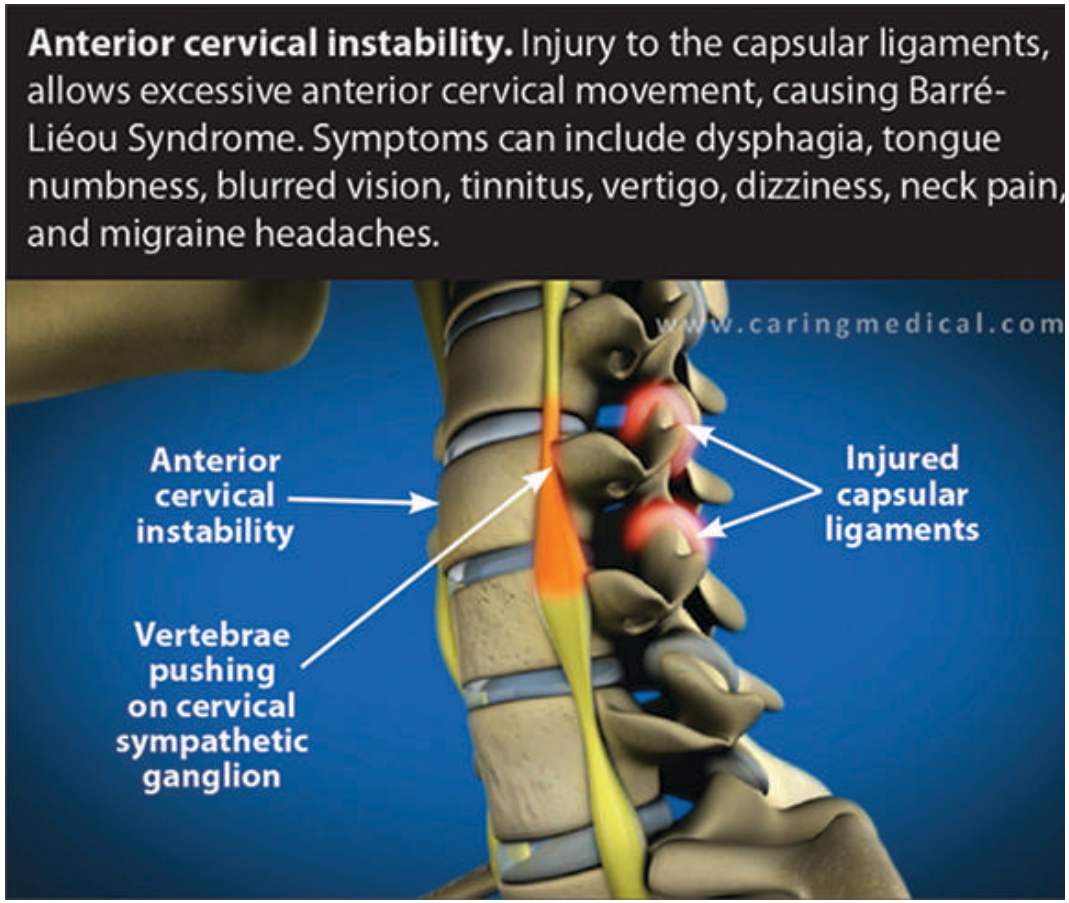
Vista® MultiPost Therapy Collar – an improved design

Barrie – Lieou syndrome

- Cervical instability pushing on the autonomic nerves
- Usually triggered by a whiplash type injury
- Difficulty swallowing
- Tongue numbness
- Blurred vision
- Tinnitus
- Dizziness
- Neck pain
- Headaches



Ross Hauser, MD, Caring Medical

Soft neck collars are useless

“neck warmers”

TMJ Pain

Temporo Mandibular Joint Dysfunction

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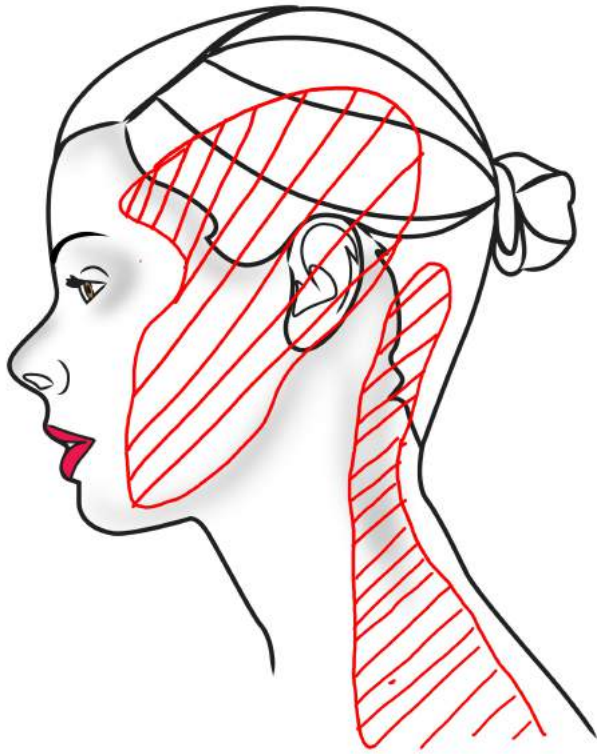
Temporo-Mandibular joint pain



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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

Dental issues in Ehlers Danlos Syndromes

- Teeth: weak and thin enamel, prone to cavities
- Gums: periodontal gum weakness, gingivitis, easy bleeding, delayed healing after surgery, tissue breakdown after surgery (tooth extraction), gum recession and pocketing
- Poor tooth stability, crowding of teeth
- Very high incidence of gingivitis – this increases inflammatory reactions in the body (higher diabetes, higher arthritis etc)

Livionex® - an excellent dental gel for EDS and MCAS

- No salicyclates
- Free of gluten, sugar, chemicals, color dyes
- No harsh abrasives or detergents
- Helps rebuild enamel and disrupts plaque formation
- Helps repair connective tissue to close pockets

Numbing medicine injections

- Commonly used numbing medicine injections (Lidocaine) may be ineffective

- Carbocaine (Mepivicaine) works well in EDS

Upper back pain

Upper back pain in EDS

- Usually poor posture
- 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

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

High Racer back – Posture bra

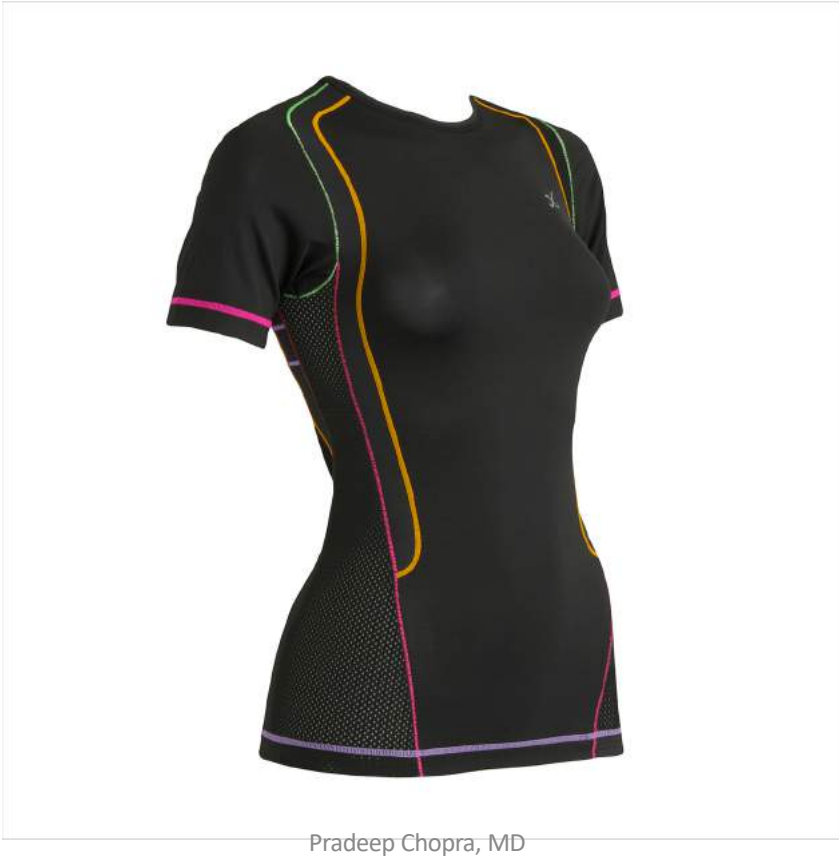
Compression garments

www.dmorthotics.com or

Pam Cowans -

p.cowans@dmorthotics.com

Joint support compression. cw-x (check Zappos.com)



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

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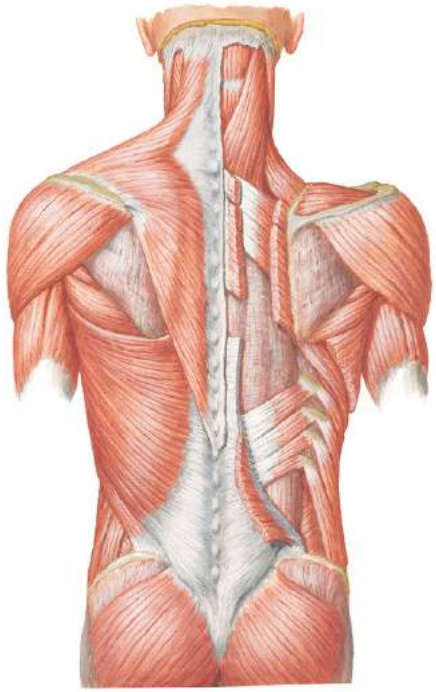
Spinal Instability

- 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

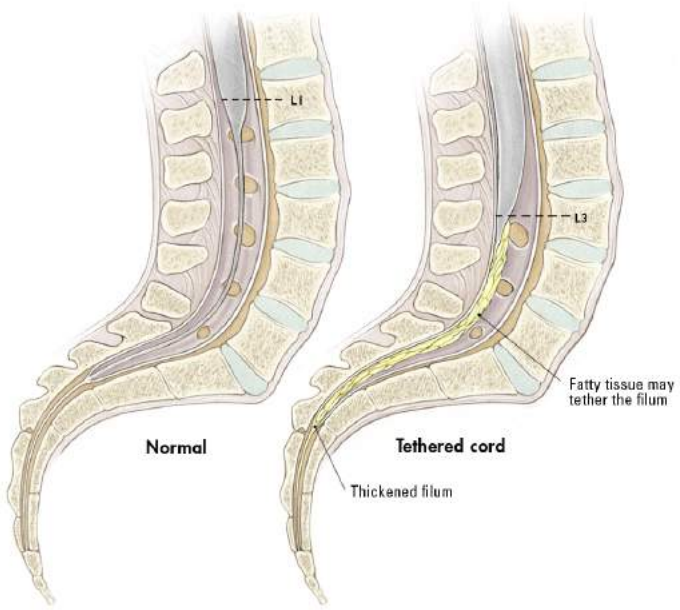
- Steroid injections not very helpful but can be used in very select cases

Brace for lumbar and Sacroiliac joint – Lumbo Lady

Good support for the SI joint and lower lumbar region

Tethered Cord Syndrome

Tethered Cord Syndrome (TCS)



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

Clinical symptoms

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

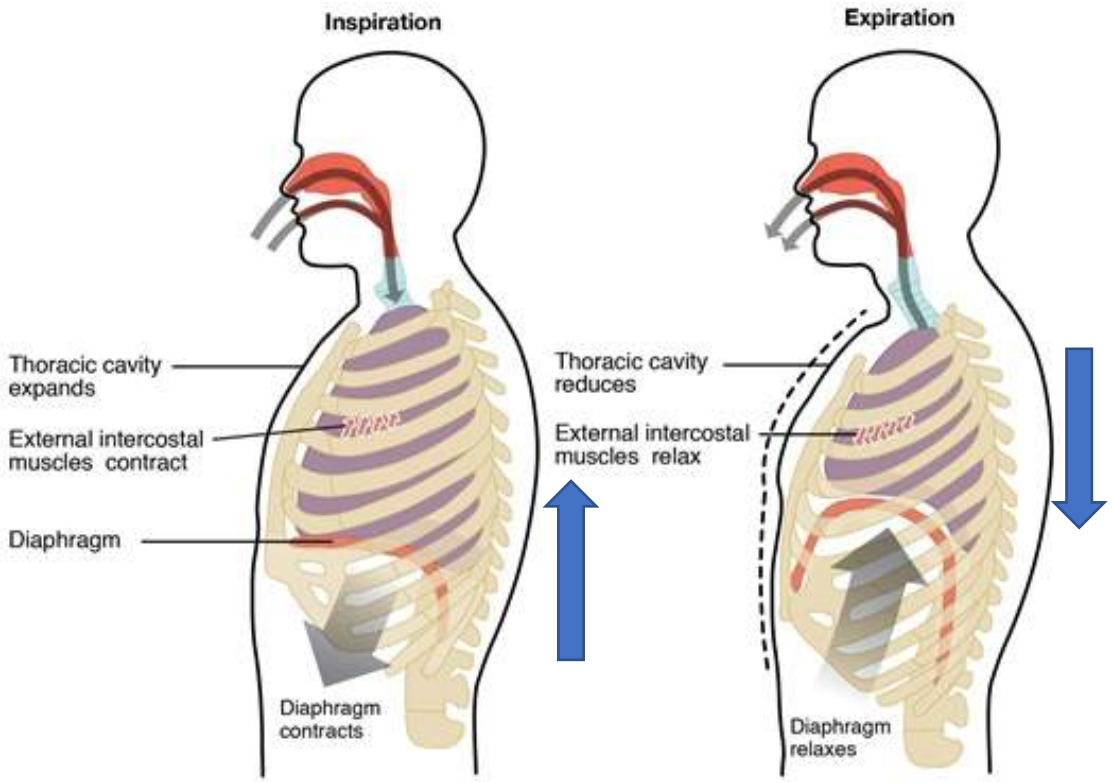
Neurogenic bladder

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

Pain from Ribs in EDS

Breathing and rib pain

- Patients with EDS often have “Air Hunger” – a feeling of having forgotten to take a breath
- Pain from ribs
- All the tests for heart and lungs are normal.
- Each rib has 3 joints in the back


Inspiration
Expiration
Thoracic cavity expands
External intercostal muscles contract
Diaphragm
Diaphragm contracts
Thoracic cavity reduces
External intercostal muscles relax
Diaphragm relaxes

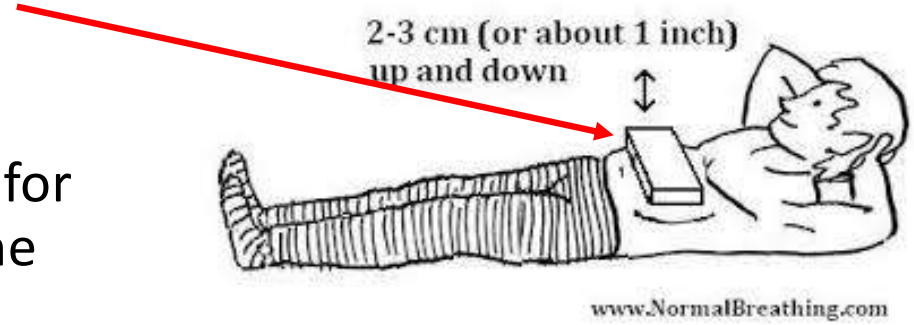
Breathing and rib pain

- 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)

- Book or sand bag should be over the belly button

- Lie on your back
- Breathe in and breathe out for 20 minutes a day – lifting the book up



http://www.normalbreathing.com/

Diaphragm release exercise

- Lie on your stomach on the floor
- Place a 1.5 inch rubber ball under your belly button
- Just lie comfortably in this position for 20 minutes.
- If you get uncomfortable before a minute, use a smaller ball.

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 and strengthening lower back
- Rib subluxations happen with uncoordinated breathing (remember poor proprioception), and poor posture

Pain in the arms in EDS

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Pain in arms

- Shoulder joint subluxations, dislocation
- Thoracic outlet syndrome
- Elbows: Tendonitis, bursitis, hyperextension
- Wrist and fingers: subluxations, muscle pain, tendonitis

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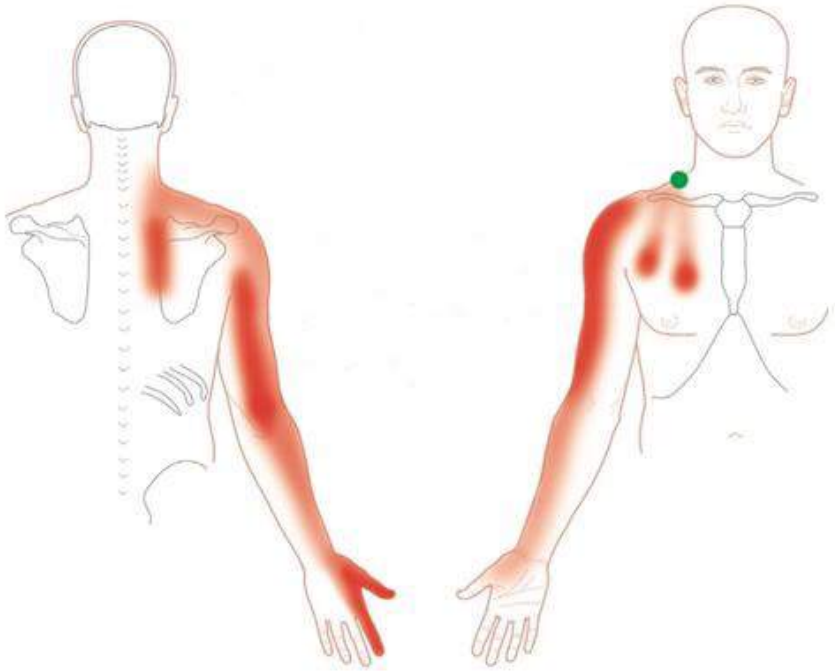
Shoulder pain

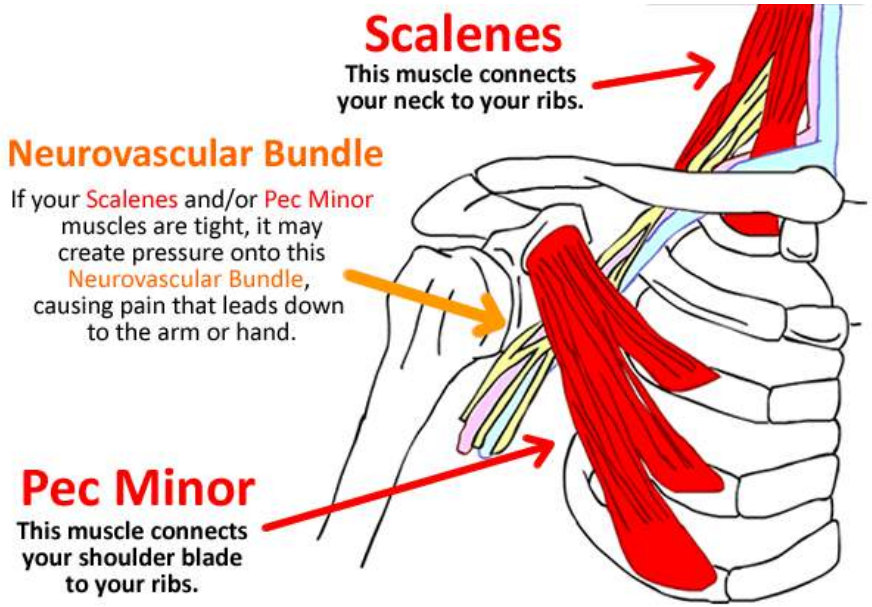
- Laxity of the shoulder joint causes the muscles (rotator cuff) around the shoulder to spasm

- Thoracic Outlet syndrome.

Pain patterns in Thoracic Outlet syndrome



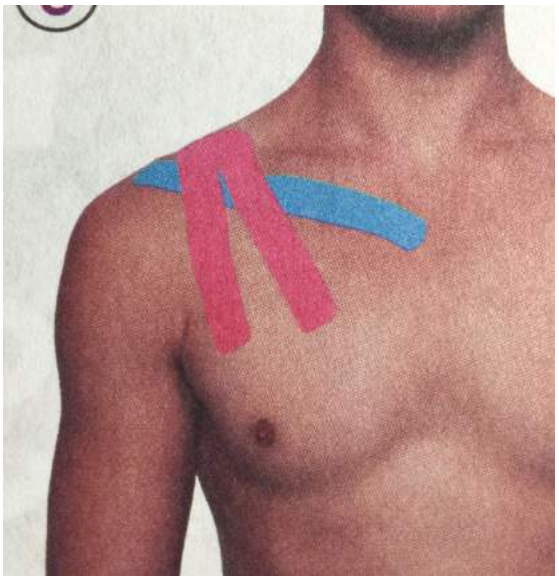
Thoracic Outlet Syndrome



Thoracic Outlet Syndrome

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

Kineseo Taping for shoulder joint pain



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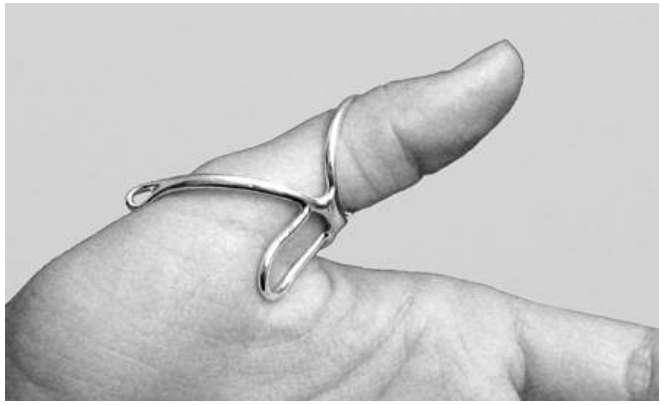
The EDS way of holding a pen

- 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

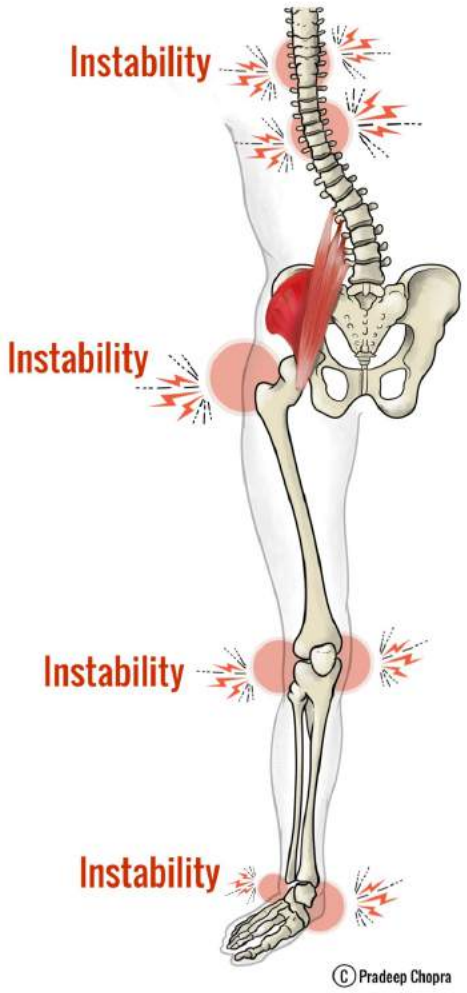
Do braces make your muscles weaker?

- No, they do not cause muscle atrophy
- 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.

Legs

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


Instability
Instability
Instability
Instability
© Pradeep Chopra

Flexible flat feet – predominantly in the forefoot

Pronation

http://www.integrativepersonaltraining.com/blog/run-flat-feet-knee-pain/

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Weight evenly distributed
Weight shifts to the inside of the sole
Left Foot
Normal Foot
Flat Foot
(excessive pronation)
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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 !!

Pedag
www.pedagusa.com

Ankle brace to stabilize the ankle joint

BAUERFEIND®

Knees

Patella is stabilized muscles of the thigh

Hyperextension at the knees

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

Treatment options for knee pain in EDS

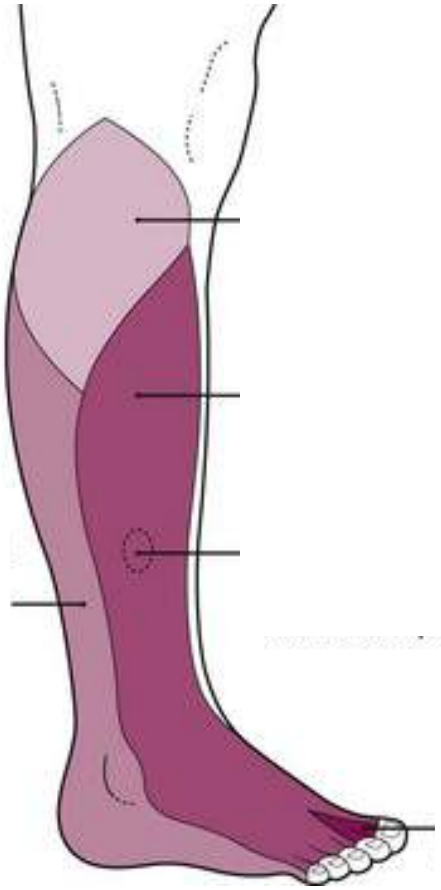
- Stabilize the feet and ankles, first
- Strengthen muscles around the knee
- Knee brace
  - Two straps above knee
  - Two straps below knee
  - Patella stabilizer
  - Metal strut to prevent hyperextension

Wrap around knee brace - Gripper

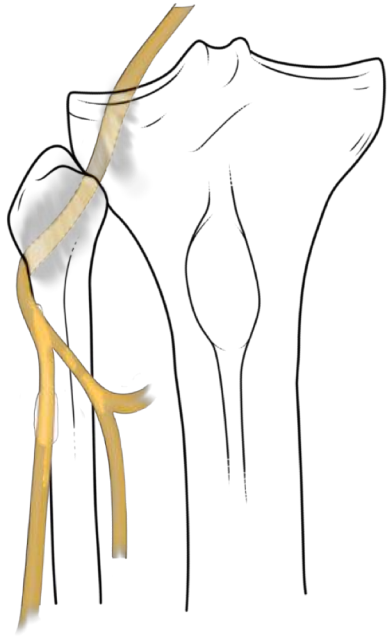
A missed cause of leg pain – the proximal tibio fibular joint

Knee pain – often missed cause

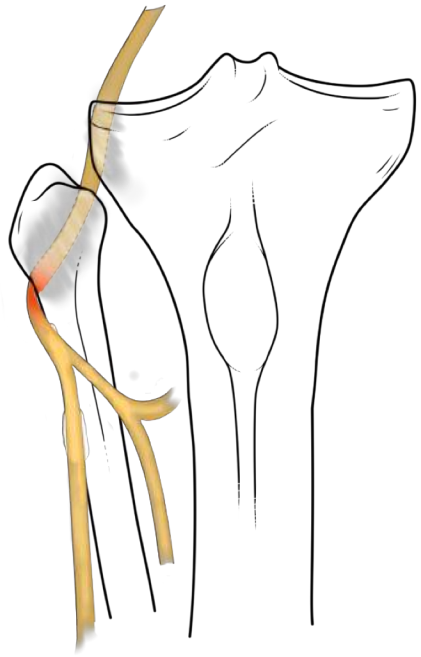
- Site of pain from the proximal Tibiofibular joint
- It can inflame the peroneal nerve which causes pain down the side of the leg and even foot drop



Proximal Tibio-Fibular joint

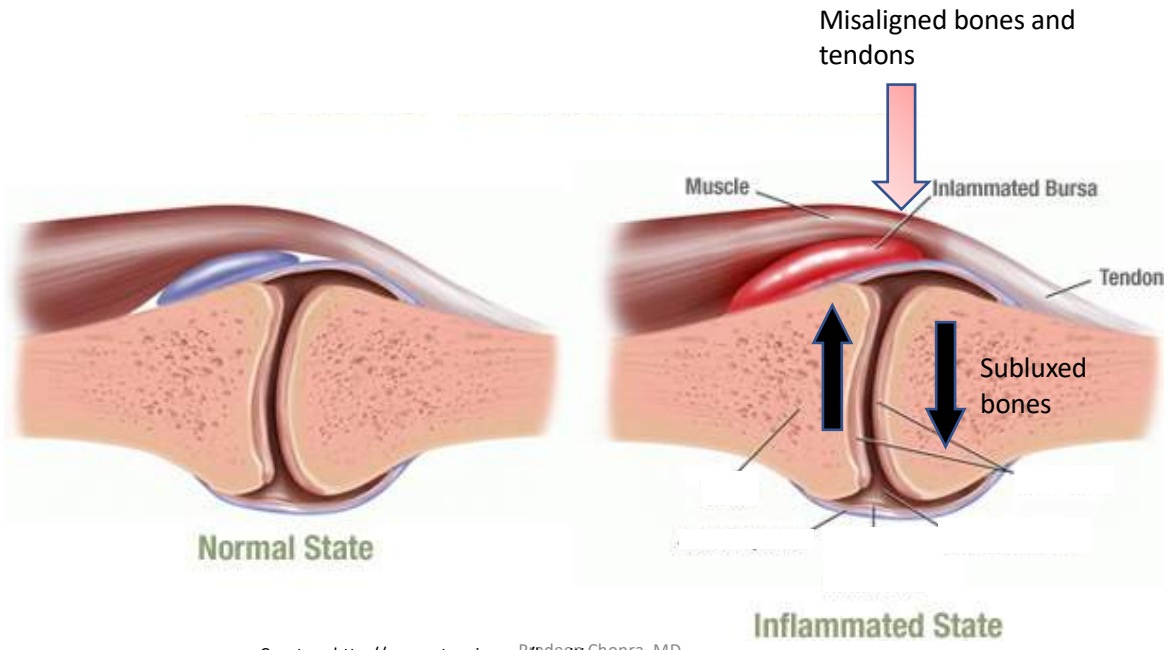


Proximal Tibio-Fibular joint



Tendonitis and bursitis

Tendonitis and bursitis in EDS



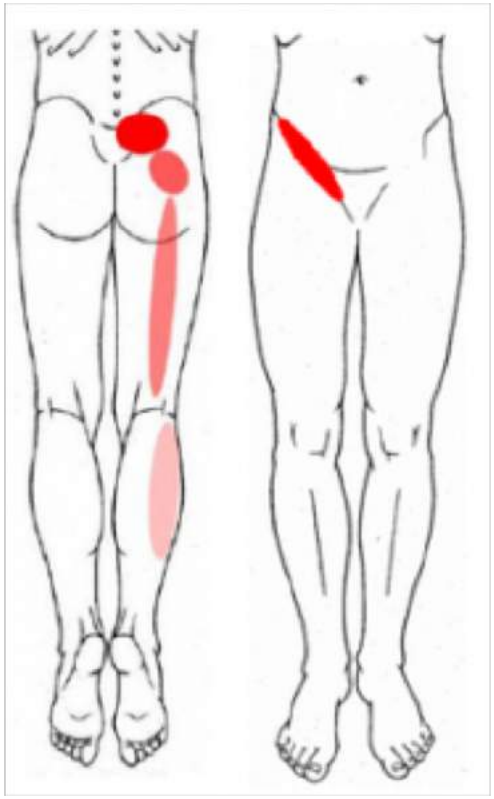
Courtesy http://www.ateevia.com/bursitis.aspx

Tendonitis and bursitis in EDS

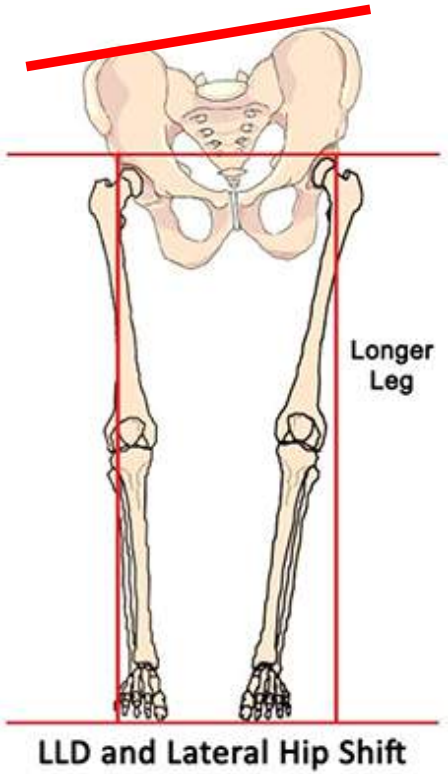
- Treatment lies in correcting the underlying problem –
  - correct bracing to align the joints
  - Correct posture (especially standing)
  - Avoid repetitive use of joint
  - Maintain proper balance

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



- Leg length discrepancy



LLD and Lateral Hip Shift

LumboTrain® Lady – a good brace for SIJ

BAUERFEIND®

Dysautonomia / POTS (Postural Orthostatic Tachycardia Syndrome)

Dysautonomia

- This is a fancy name for a bunch of medical conditions that affect the nerves that control automatic functions in our body.
- “Dysfunction of Autonomic Nervous System”
- PoTS – Postural Orthostatic Tachycardia Syndrome
- Its one of the most common types seen in EDS

Postural Orthostatic Tachycardia syndrome (PoTS) - Symptoms

- Fainting, dizziness
- Heart racing (Palpitations)
- Fatigue
- Headaches
- Difficulty maintaining body temperature
- Poor concentration “brain fog”
- Feeling of constant anxiety
- Stomach pain
- Wake up feeling tired

PoTS

- Patients with EDS pool excessive blood to their lower extremities,
- This triggers an increased activity in the same nervous system that controls ‘fight or flight’

POTS - Postural Orthostatic Tachycardia syndrome

Dysautonomia International for more information and high salt diet recipes.

http://www.dysautonomiainternational.org/

- EDS Awareness

https://www.chronicpainpartners.com/

PoTS

- The constant feeling of dizziness makes patients feel unstable
- The laxity of the joints makes the muscles tighten reflexly
- This constant imbalance of muscles worsens their pain and fatigue

Anxiety in EDS

- 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 PoTS– its because of excessive activity of the flight or fight nerves to control blood pressure.

POTS - Postural Orthostatic Tachycardia syndrome

- 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
- Syncope or almost syncope (fainting)
- In children an increase of 40 beats/minute

POTS - tests

- Orthostatics – Measure blood pressure and heart rate:
  - after lying supine for 5 minutes
  - standing up - immediately
  - Standing for 10 minutes
- Q-sweat test
- Tilt Table Test

Sample readings in POTS

Lying flat for 5 minutes	**110/66**	**82**
Standing, immediately	100/68	115
Standing for 10 minutes	112/65	108

PoTS - treatment

- Increase salt by mouth (salt pills: http://saltstick.com/vitassiumpage/)
- Increase electrolyte oral fluids
- Trial of 'Banana bag oral rehydration fluid https://www.bananabagdrink.com or Gatorade
- Compression tights up to thighs.
- Abdominal binder or trial of a one size smaller swimsuit.
- Cardiology or Neurology consult for Dysautonomia/POTS.
- Consider a beta-blocker such as Metoprolol

Treatment of POTS

- Exercise is the key here.
- Exercise lying down.
- Dallas protocol
- Exercise in pool – simple walking, treading water
- https://youtu.be/jbGNfuXVHwc
- https://youtu.be/oXnNL_furCw
- https://youtu.be/hB8yr1e823Q

Courtesy: Dysautonomia International

Treatment of POTS

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Courtesy: Dysautonomia International and RSDSA

Exercise in pool for EDS

- Exercises in pool:

- https://youtu.be/g7df_ek50Jk
- https://youtu.be/mVvzsFk6rPo?list=PLPS8D21t0eO-X68fTNKk7UEkegxDqzQ3U
- This one is also good for CRPS/RSD
- https://youtu.be/wUnwbNsIk1c

POTS – one last thing

- POTS can be because of other conditions such as Sjogrens syndrome, Antiphospholipid syndrome, celiac disease, anykylosing spondylitis. Myasthenia gravis, multiple sclerosis
- Can be seen in Autoimmune conditions where patients develop antibodies to alpha 1, Beta 1, Beta 2 adrenergic receptors, muscarinic 3 receptors, (12)g-Acetylcholine receptors, cardiac lipid raft proteins.
- These patients may respond well to IVIg infusions.

Postural Orthostatic Tachycardia syndrome (PoTS) - Symptoms

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- Fatigue
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- Difficulty maintaining body temperature
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- Stomach pain
- Wake up feeling tired

Mast Cell Activation Syndrome

MCAS

Mast Cell Activation Syndrome (MCAS)

- Mast cells are a type of cell found in the blood.
- They are part of our body defense mechanism (immune system)
- When they detect an attack (infection or something that is not good for the body) they get activated
- Once mast cells get activated they release a bunch of chemicals such as histamine, cytokines etc

Mast Cell Activation Syndrome (MCAS)

- In MCAS, mast cells are inappropriately triggered and release excessive amount of these chemical mediators (histamine, cytokines etc.)
- It is NOT mastocytosis (whole different condition)

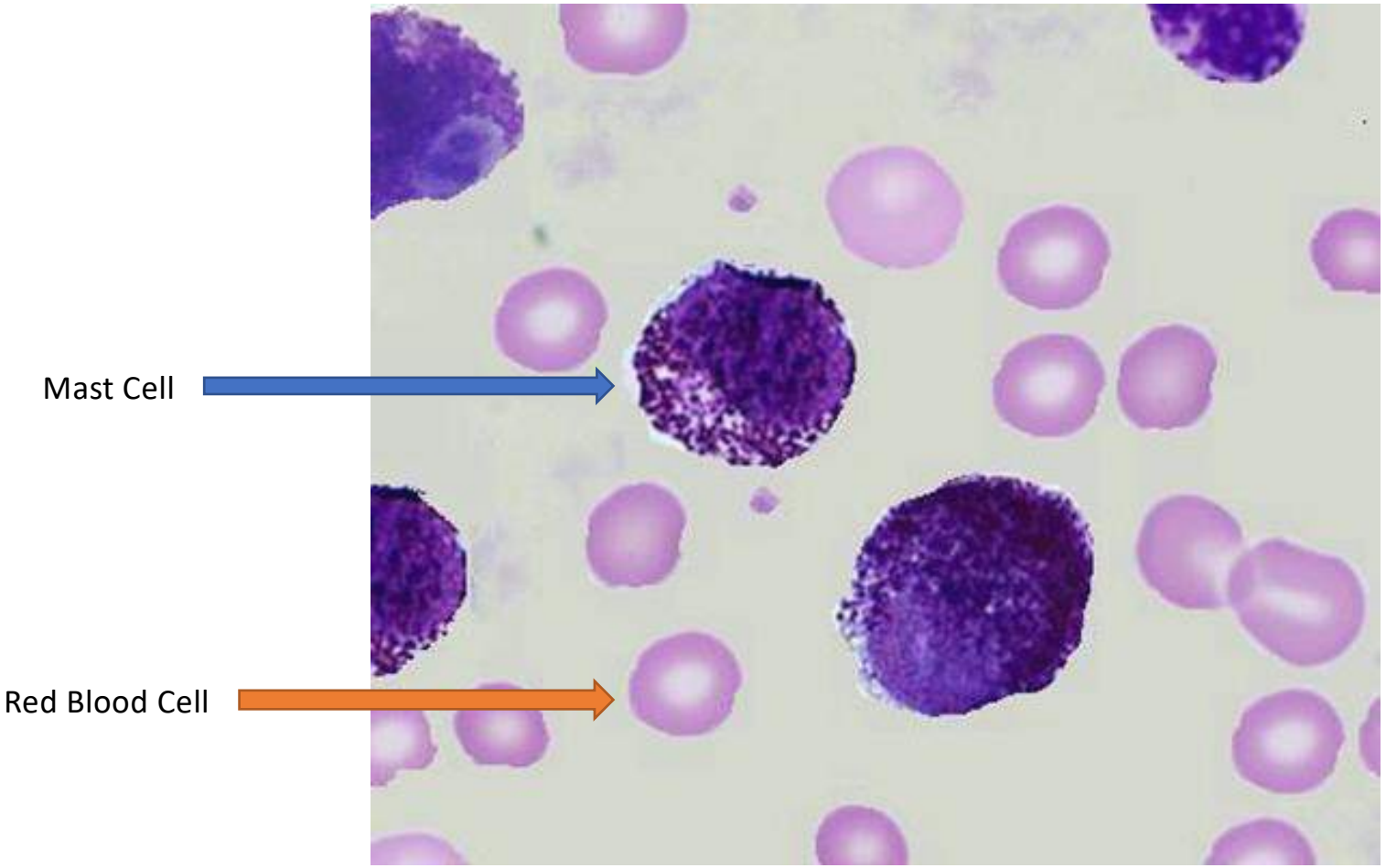
Mast Cell Activation Syndrome (MCAS)

- Rashes, hives, itchy
- Fatigue, tiredness
- Muscle pain
- Bone and joint pain
- Abdominal pain, bloating, nausea
- Palpitations, pre-syncope
- Flushing especially after a hot shower
- Constant feeling of having flu

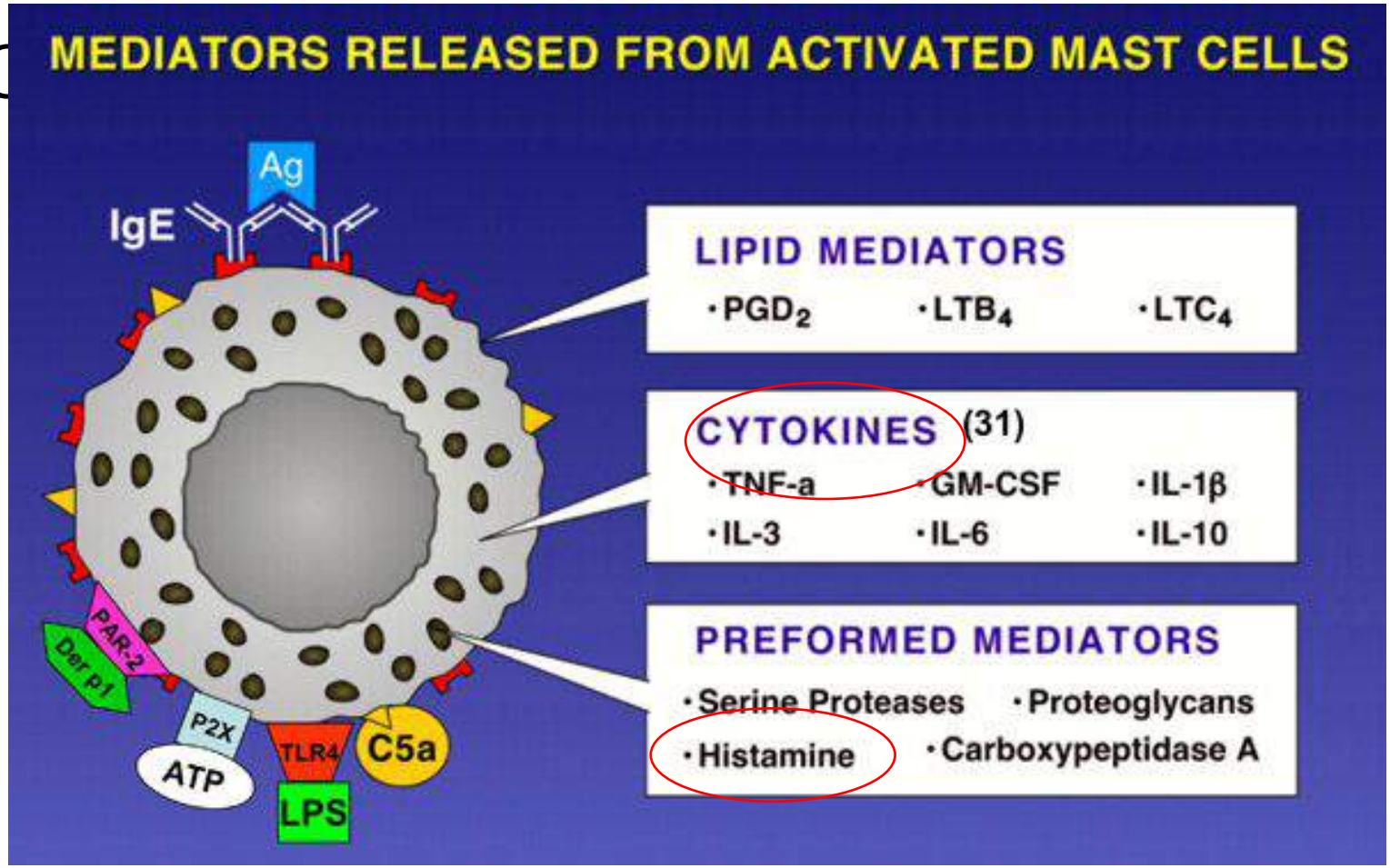
Dermatographism (writing on skin)

Urticaria Multiforme - Scientific Figure on ResearchGate. Available from: https://www.researchgate.net/Dermatographism-Linear-wheals-after-writing-on-the-skin_fig4_236106893 [accessed 3 Oct, 2018]

Mast Cells



MC



Courtesy Ann Maitland, MD

Mast Cell Activation Syndrome (MCAS)

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- Fatigue, tiredness
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- Flushing especially after a hot shower
- Constant feeling of having flu

MCAS – Testing

- Tests for MCAS are NOT reliable. However, these test can be done:
- Urine 24 hour collection for N-methylhistamine.
- Same 24 hour collection Prostaglandin D2/F2 or 11 Beta prostaglandin F, whichever is available.
- Serum tryptase.
- Serum Chromogranin A level

MCAS - tests

- The diagnosis of MCAS depends largely on a physician who knows MCAS and is clinical
- Laboratory tests are very difficult and unreliable.
- If a biopsy sample is taken from endoscopy then:

  Please request Pathologist for the following:
- Number of Mast Cells per high power field.
- Stain for Mast Cells with CD117.
- Comment on the morphology and histology.
- Request KIT-D816V mutation analysis by PCR

MCAS – treatment

- Ranitidine (Zantac®) 150mg twice a day.
- Loratidine (Claritin®) one tablet twice a day.
- Cetrizine (Zyrtec®) 10mg one tablet twice a day.
- Clemastine 2mg p.o. b.i.d.
- Rupatadine 10mg p.o. b.i.d.
- Quercetin 1000 mg / day
- Consider Ketotifen.
- Consider Cromolyn oral 200mg four times a day.
- Cromolyn Nebulized.

MCAS – treatment

- Avoid triggers for Mast cell activation such as NSAIDs, alcohol, opioids and sudden temperature changes.
- Inactive ingredients (binding agents, preservatives) in pills may contribute to MCAS.
- Consider a leukotriene inhibitor such as Singulair.
- Low histamine diet.

MCAS – treatment

- Review and change possible hidden food sources and the possibility of accidental exposure to such foods.
- Some of the usual suspects for MCAS to avoid are:
    - Seasonings (except olive oil, salt).
    - All dairy.
    - Eggs.
    - Grains except quinoa and rice.
    - Avoid beef products but okay to take lamb, venison and poultry.
- Food and drug reactions are a common cause of unexplained hives and rashes. Some of the other causes of hives may be exposure to change in temperature (hot to cold), rubbing skin, physical exertion, pressure and direct exposure to sunlight.

MCAS – treatment

- Other methods to avoid exacerbation maybe:
    - Use pollen guards in windows.
    - HEPA air filters, A/C.
    - Use a mask (e.g. VOG mask) while dusting or vacuuming.
    - Avoid long baths or showers, rapid changes in temperature, wool, dust and cigarette smoke.
- May use nasal saline during flare ups

Abdominal Pain

Abdominal pain

- Very common in EDS
- Slowing down of the intestines
- Alternating diarrhea and constipation
- PoTS, MCAS –causes nausea, acid reflux, bloating and constipation
- Prolapse
- Acid reflux from MCAS

- Low FODMAP diet – for bloating, pain and diarrhea
- Treating PoTS and Mast Cell Activation Syndrome helps
- In case you are wondering:
- FODMAP - **F**ermentable **O**ligo-, **D**i-, **M**ono-saccharides **A**nd **P**olyols

Low FODMAP diet

- https://www.kcl.ac.uk/lsm/research/divisions/dns/projects/fodmaps/faq.aspx
- There are 3 stages to the low FODMAP diet:

- Stage 1: **Restriction**: In this stage, you reduce your FODMAP intake by avoiding foods that are high in FODMAPs for four to eight weeks as this period is considered long enough to identify if symptoms will respond to a low FODMAP diet.

- Stage 2: **Reintroduction:** If your symptoms have improved following FODMAP restriction, it is important to reintroduce some high FODMAP foods. This will enable you to identify which FODMAPs you are most sensitive to, as well as how much of a high FODMAP food triggers your symptoms.

- Stage 3: **Personalization**: The long term aim of a low FODMAP diet is to personalize your diet so you only avoid foods that trigger your symptoms and you return to as normal a diet as possible.

Dysfunction of movement of intestines

- Can lead to changes in the gut microbiome (the helpful bacteria that live in our intestines)
- Problems with movement of intestines may lead to Small Intestinal Bacterial Overgrowth (SIBO)

Small Intestinal Bacterial Overgrowth - symptoms

- Abdominal pain/discomfort
- Bloating
- Diarrhea
- Weakness
- gassy

SIBO – problems

- Weight loss
- Vitamin deficiency – A,D,E,K, B12,
- Vitamin excess – Folate
- Iron deficiency
- Low protein

SIBO – testing

- Organisms in the intestine produce hydrogen and methane.
- Breath test can detect levels of hydrogen and methane
- Elevated methane levels can be treated with statins (lovastatin – red rice yeast)
- Treat any dysautonomia
- Treat any MCAS
- Treat constipation
- Drugs for improving GI motility (Resolor, Iberogast, Mestinon, erythromycin)

SIBO – treatment

- Elevated methane levels can be treated with statins (lovastatin – red rice yeast)
- Specific antibiotics to treat SIBO.
- Be cautious of repeating antibiotics
- Treat any dysautonomia
- Treat any MCAS
- Treat constipation
- Drugs for improving GI motility (Resolor, Iberogast, Mestinon)

SIBO – treatment

- Avoid simple carbohydrates
- Think of pureed foods.

Median Arcuate Ligament Syndrome

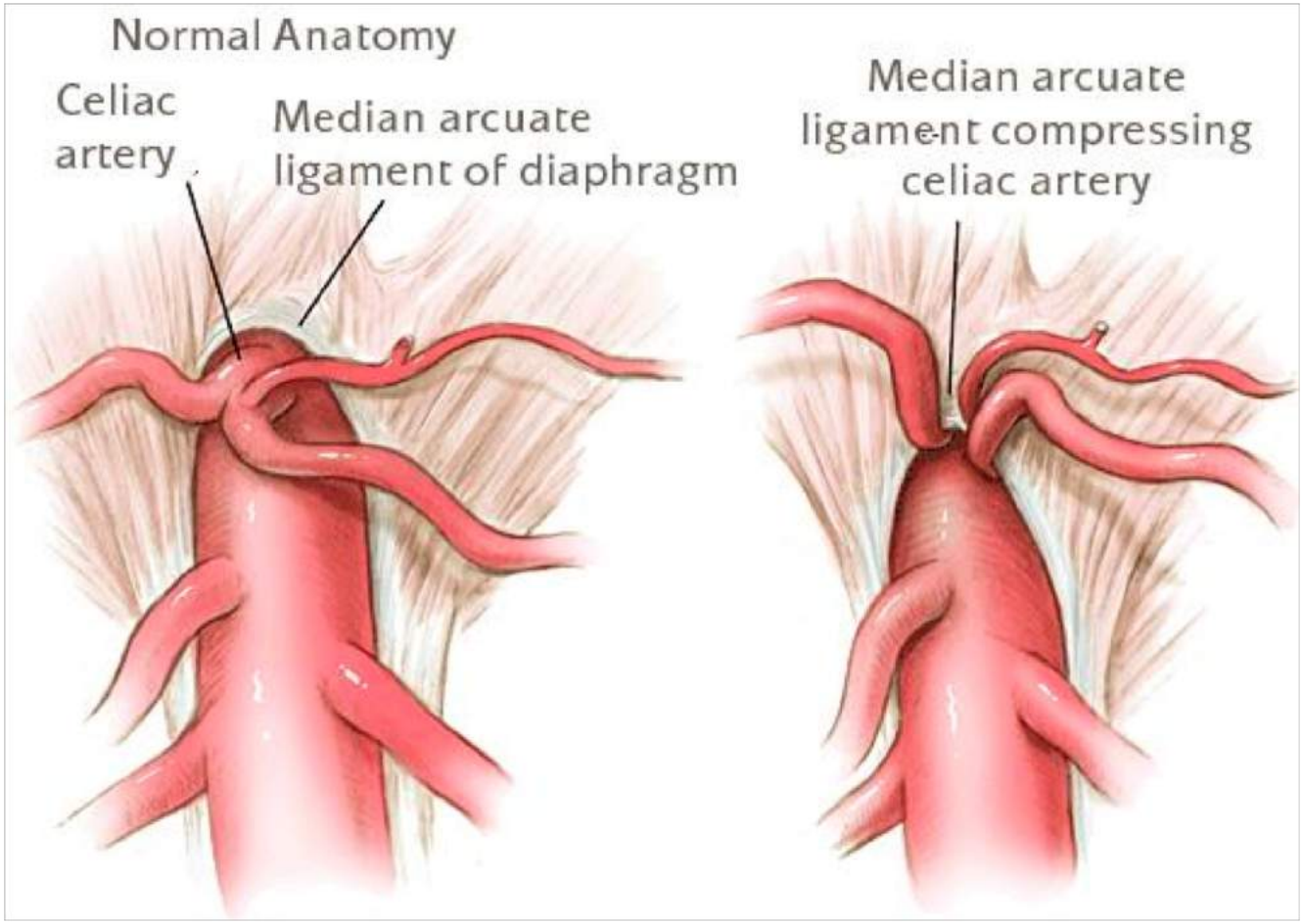
MALS

Abdominal pain – MALS

- Median Arcuate Ligament Syndrome (MALS).
- Belly pain after eating food.
- Can trigger PoTS

Abdominal pain – MALS

- There is an artery that supplies blood to part of the intestine
- It is called Celiac Artery
- This artery pokes out from under a ligament called Median Arcuate ligament
- There are 2 theories about why MALS causes pain
- The median arcuate ligament is tight causing compression of the celiac artery
- There is a nerve in that area called Celiac ganglion which may be getting compressed or inflamed.


Normal Anatomy
Celiac artery
Median arcuate ligament of diaphragm
Median arcuate ligament compressing celiac artery

Abdominal pain – MALS

- Treatment is surgical decompression
- If treated early, symptoms resolve
- If treated late, may need IVIg infusion

Pelvic pain

Pelvic pain in EDS - Dysmenorrhea (painful menstrual periods)

- Birth control to stop periods – may help with joint laxity also
- NSAID's (ibuprofen, naproxyn)
- Control Mast Cell Activation Syndrome

Pelvic pain in EDS - Interstitial cystitis (bladder pain)

- Maybe part of Mast Cell Activation syndrome
- Cromolyn or Ketotifen
- Palmitoyl ethanolamide (PEA)

Pelvic pain in EDS – Vulvodynia (vaginal pain)

- Maybe part of Mast Cell Activation syndrome
- Benadryl douche or cream
- Cromolyn douche or cream

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

- 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.
- Parents – please research the facility carefully before taking your child there.
- The facility should have trained physicians in EDS and its complications
- There are some hospitals that have hired “Medical Child Abuse” specialists.

Behavioral

- To diagnose a child with Conversion disorder or Munchausen by Proxy without a dedicated multidisciplinary team approach and without concrete evidence is extremely harmful to the patient.

- To label an adult with pain as being psychological is severely harmful. It closes all doors to correct treatment.

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 a 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.

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://freedomservicedogs.org

To manage pain first find out what's broken

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

- For example, pain in the shoulder joint can be from:
  - Dislocated shoulder joint,
  - Muscle spasms
  - Nerve or blood vessel impingement or damage
  - All of the above

- The treatment of each of these is different

Types of tissue injury in EDS

- **Micro trauma** – small repetitive traumas resulting in constant tissue breakdown. Invisible tissue breakdown. This is far more common.

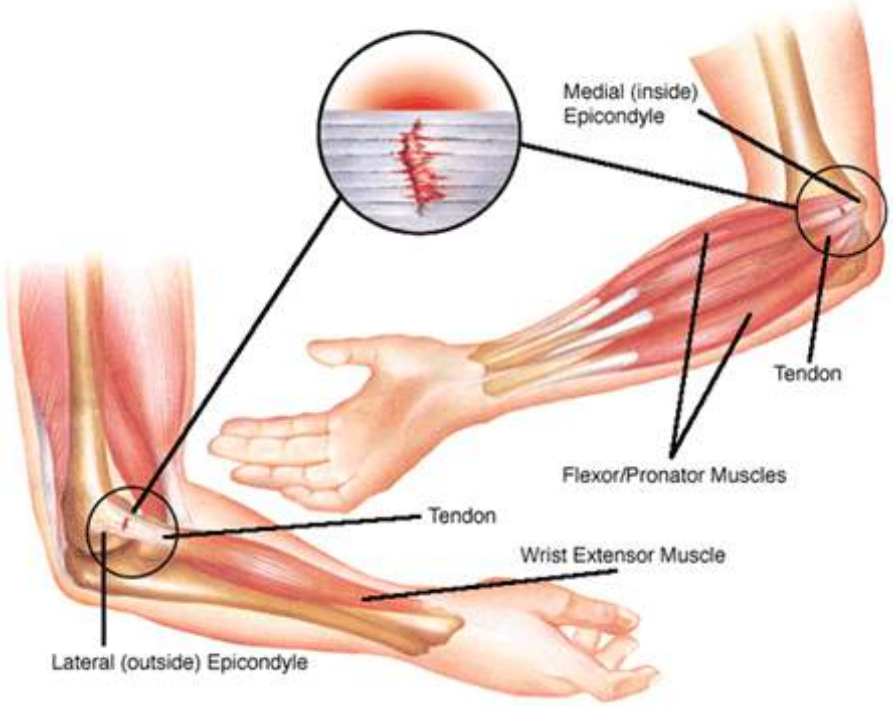
For example – overstretching muscles and ligaments. Vacuuming, chopping food, dusting

- Macro trauma – a large event or trauma resulting in injury.

For example - Dislocation or fracture

Macro trauma

Micro trauma



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.

  - Follow the NC10 rule (if you can get at least 10% from one treatment, five such treatments will give you 50% relief)

Joint Position sense

Proprioception

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

Proprioception (Joint Position Sense)

- Proprioception, also known as Joint Position Sense, is the ability of a joint to

1. determine its position,
2. detect movement, and
3. sense of resistance to force

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.

Proprioception – Joint sense

- The brain uses sensors in the skin as a way of figuring out the position of different parts of the body.
- Wearing skin tight clothes over joints and limbs will help the brain understand where the body is
- One of the reasons that exercising in water helps is because your skin senses contact with water

Proprioception exercises

- 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 avoid swimming

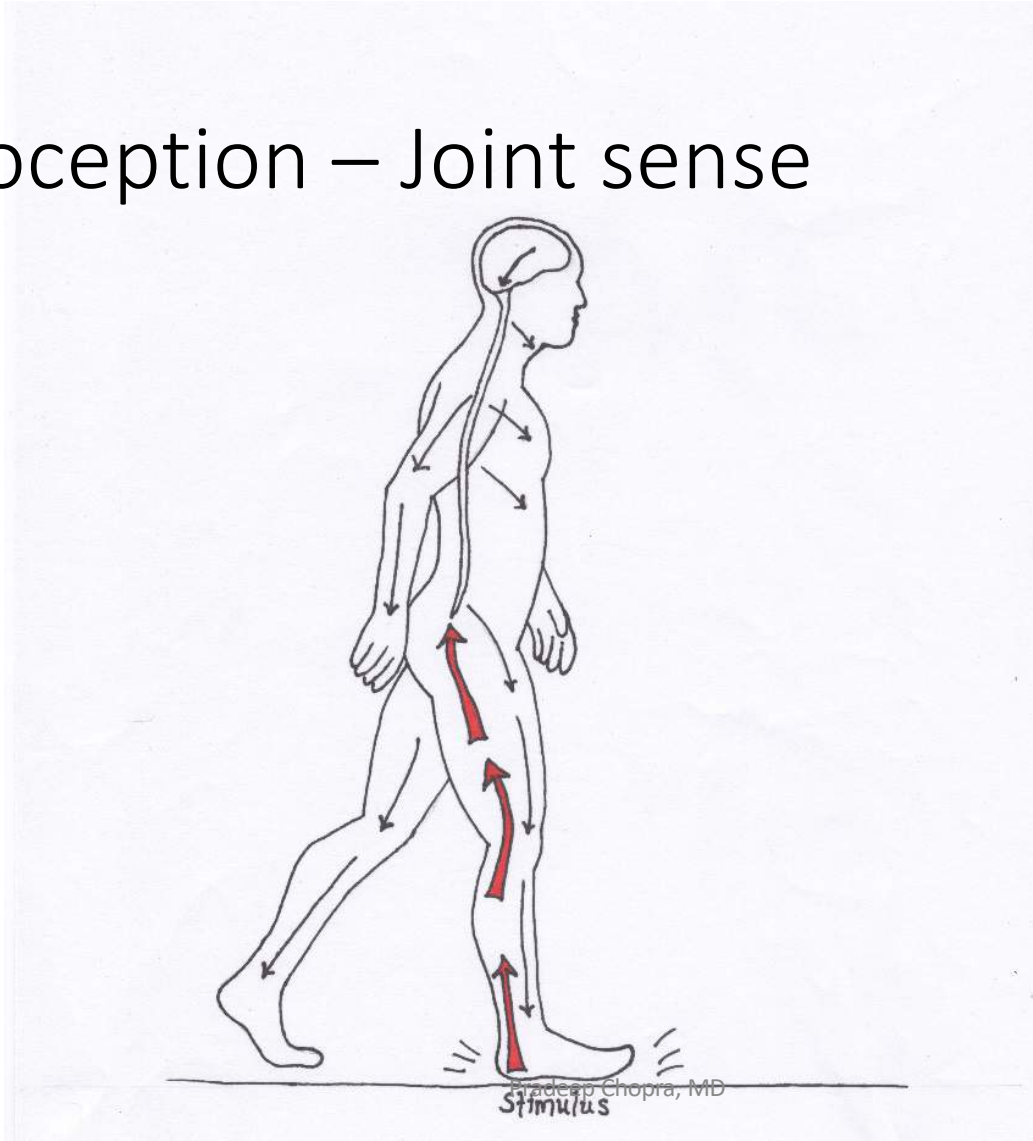
Stand up paddle board

Compression garments

Proprioception

Key concept

Proprioception – Joint sense



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

Exercise in water - walking

Exercises for EDS

Exercises for EDS

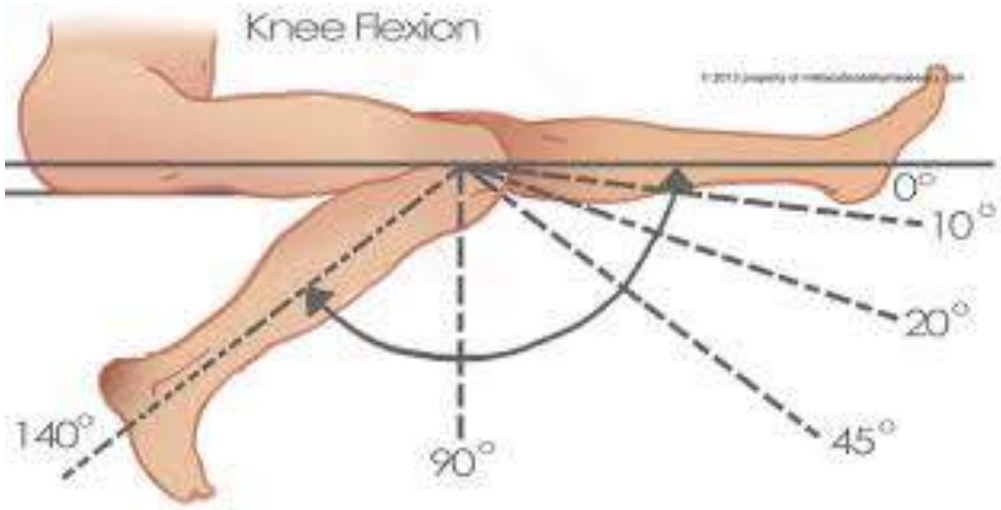
- Muscles strengthening without stressing the joints – for stabilizing joints
- Building on proprioception – for joint protection and efficient use of muscles.
- Exercising with fatigue
- It should be daily, even if it's a little bit

Muscle strengthening exercises

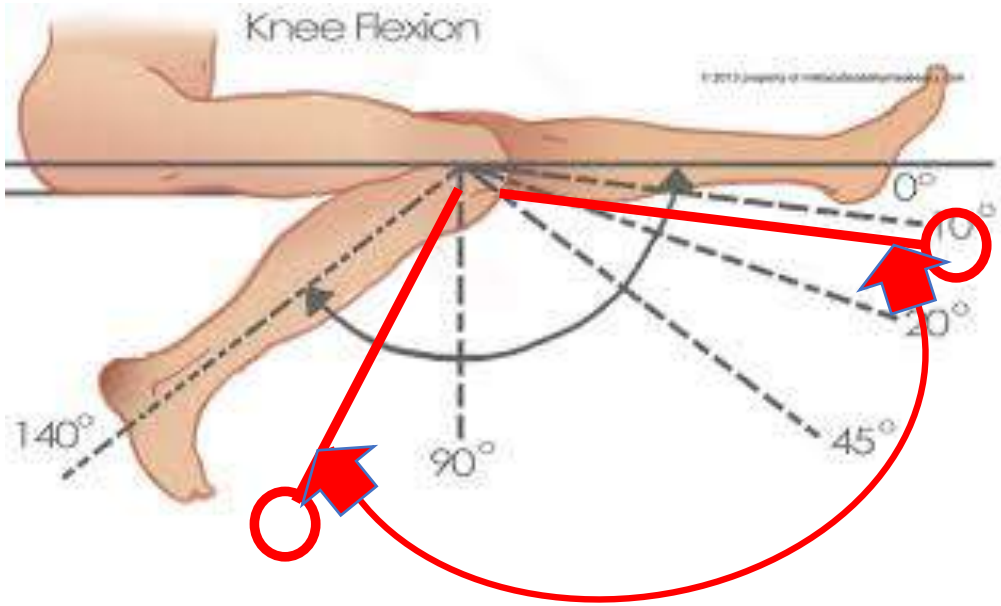
- Avoid joint loading or joint stressing
- Focus on muscle strengthening
- Avoid extreme stretching

- Start very low, really very low
- Progress slowly, really slow

Keep limb movements within extreme range of motion



Keep limb movements within extreme range of motion



Aquatic therapy

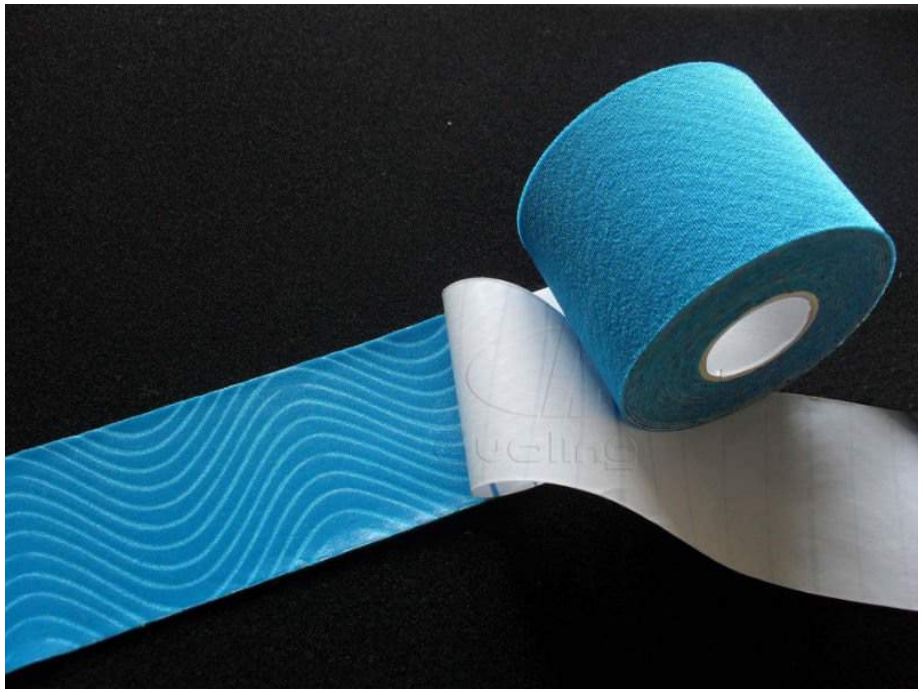
- Best form of exercise in EDS
- The contact of water with the skin helps the brain move your muscles more efficiently
- The water makes us weigh less which takes the load off the joints allowing us to exercise freely
- Avoid swimming – it strains the joints of the neck and shoulders.

Avoid swimming – injury to shoulder and neck

Neuromuscular taping (Kinesio™/ Rock Tape ™)

Pradeep Chopra, MD

Finger print pattern



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.

Pradeep Chopra, MD

Kinesio™ taping - mechanism

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

- Helps reduce swelling

Pradeep Chopra, MD

Kinesio™ taping - uses

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

Pradeep Chopra, MD

How to prevent skin reactions to K-tape

- Cavilon cream® by 3M
- Leukotape® K
- Milk of magnesia

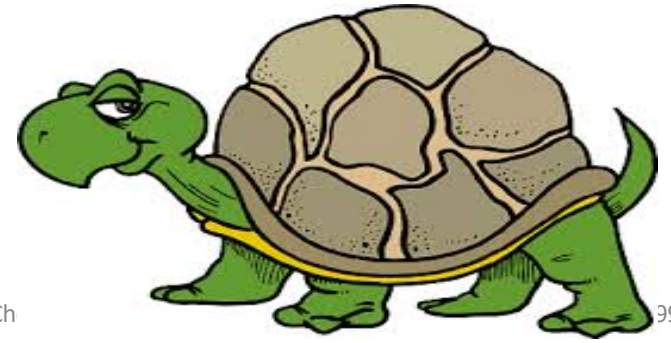
The Feldenkrais Method

The Feldenkrais Method

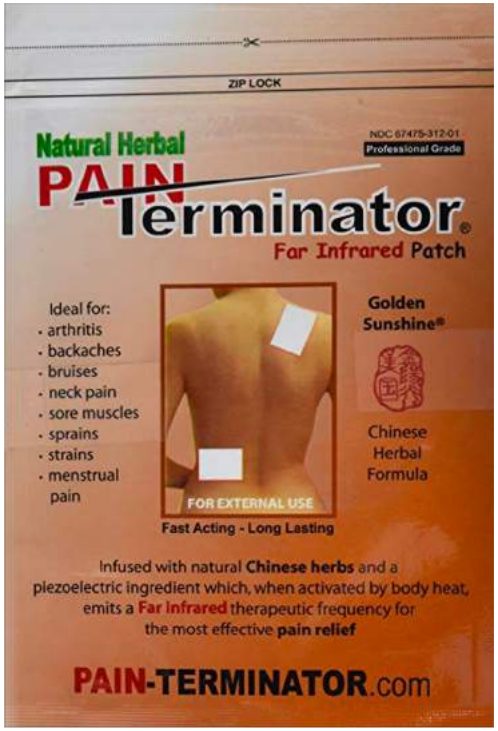
- It is a type of physiotherapy that helps repair impaired connections between the brain and the body
- Patients with EDS and chronic joint conditions develop inefficient or strained habitual movement patterns
- **It uses slow repetition to teach correct and safe movements in EDS**
- The movements are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

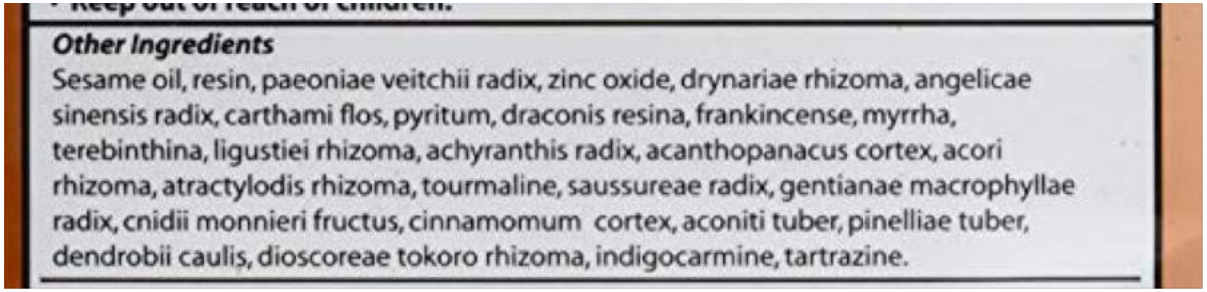
Starting treatment - medicines and exercise

Start low, go slow



Miscellaneous - Amazon





Miscellaneous – Palmitoylethanolamide(PEA) topical cream 2%

Sold as Soothamide.
Moderate relief

CGRP antibodies

- CGRP – Calcitonin Gene Related Peptide
- It is a protein in the body involved in transmission of pain
- CGRP antibodies are medicines to block the effect of CGRP and decrease pain
- In the last few months there have been 2 such CGRP antibodies on the market and a few to be released soon.
- Currently, approved for treatment of migraines only.
- They may be effective for other pain conditions.

Poor concept of exercising in pain

- No pain, no gain

Poor concept of exercising in pain - baloney

- No pain, 

Fatigue in EDS

Fatigue

- EDS
- PoTS / Dysautonomia
- Mast Cell Activation Syndrome (MCAS)
- Drugs
- Pain
- Poor sleep
- Secondary Mitochondrial dysfunction

Fatigue - causes

- Gastrointestinal: Median Arcuate Ligament Syndrome (MALS)
- Obstructive sleep – tracheomalacia (EDS), sleep apnea
- Hormonal - Hypothalamus - Pituitary-adrenal axis suffering from hypoperfusion or Sheehan's syndrome.
- Sleep apnea: Obstructive, central apnea, non-restorative sleep
- Non-restorative sleep secondary to Dysautonomia

Symptoms of EDS, POTS, MCAS

Headaches, brain fog, dizzy

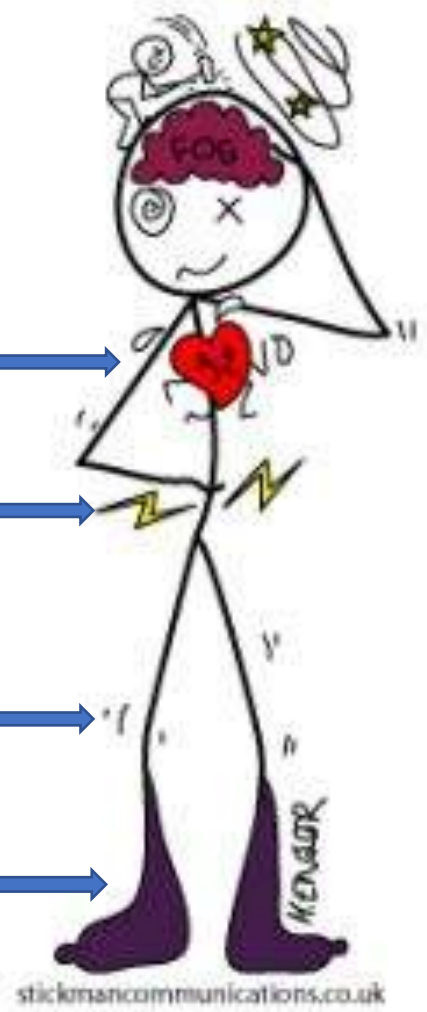
Blurry vision

Racing heart

Belly pain

Tremulousness

Dark discoloration of feet



www.potsuk.org

Fatigue in EDS

Non Ehlers Danlos Syndrome

- Muscles – relaxed at rest and contract with activity
- Ligaments – tensed at rest, they support and stabilize the body

Ehlers Danlos Syndrome

- Muscles – They are tensed and constantly attempt to stabilize even at rest
- Ligaments – provide no tension and stability

Medicinal Marijuana

Medicinal marijuana

- The human body has two types of receptors – CB1 and CB2
- CB1 receptors are found in the brain
- CB2 receptors are found in the rest of the body, immune cells and glia cells in the Central Nervous System
- Chemicals that cause inflammation in the peripheral parts of the body are modulated by cannabinoids. Hence, cannabis applied topically are helpful

Medicinal Marijuana

- Reasonable choice to try.
- Anecdotally – works well in patients with EDS
- Higher CBD levels and lower THC levels
- Vaporizing, edibles
- Topical CBD over joints, tendons, ligaments – very helpful
- Does not affect Mast Cell Activation Syndrome (MCAS) as much as NSAID's and opioids

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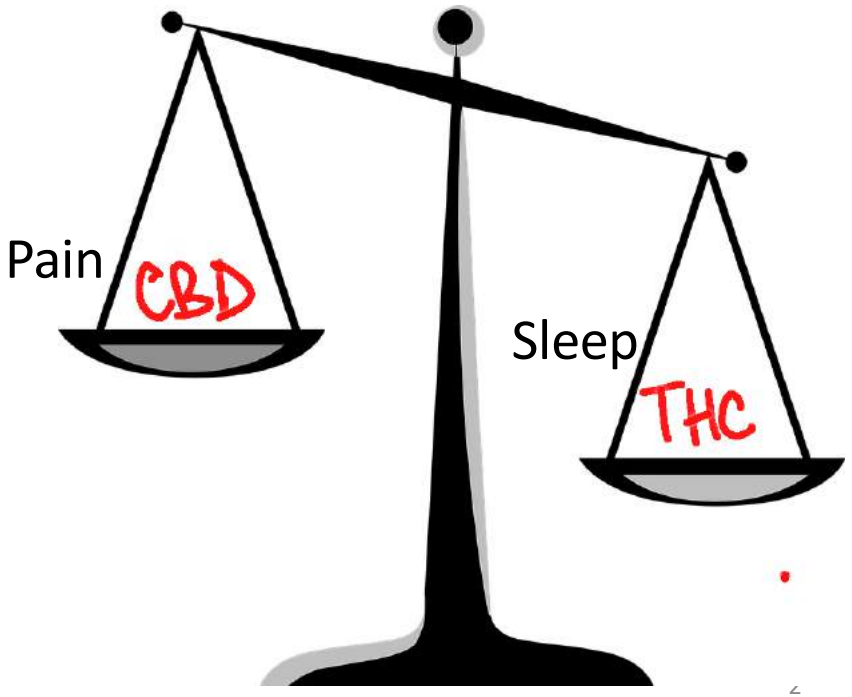
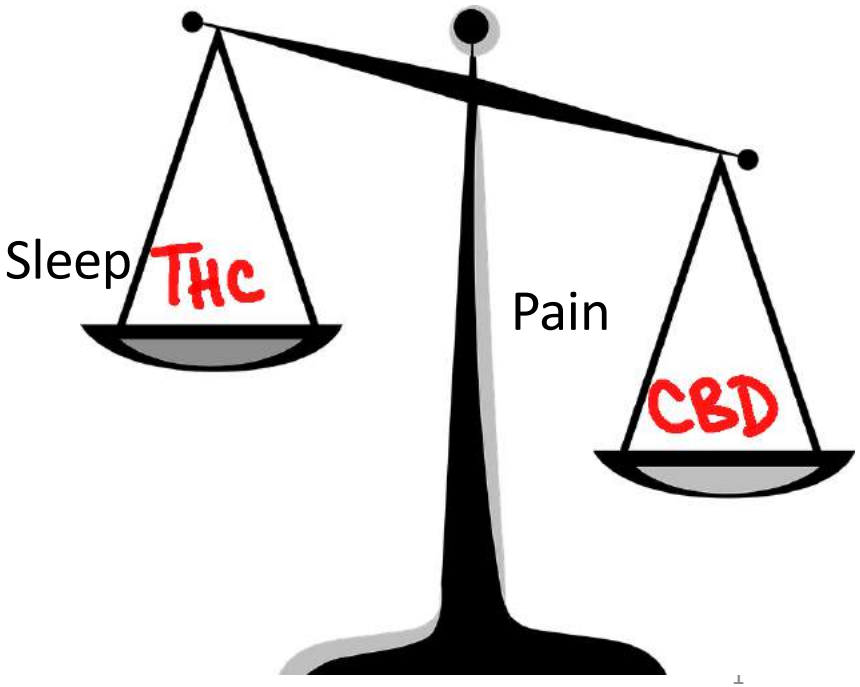
Daytime use



For sleep



Low Dose Naltrexone

LDN

Low Dose Naltrexone (LDN)

- Competitive antagonist of opioid receptors
- Clinically used for 30 years for addiction
- Suppressive effects on the CNS glia, whch helps with Central Sensitization

3-66

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.

Medications in EDS

- Patients with EDS may have an abnormal response to medications
- Maybe very sensitive,
- May have a poor response
- May need higher or lower doses than usual

Muscle Relaxants

- Limited value in EDS
- Cyclobenzaprine (Flexeril)
- Carisoprodol (Soma)
- Tizanidine (Zanaflex)
- Baclofen
- Benzodiazipine (Valium)

Acetaminophen/ Paracetamol

- One of the safest analgesic in EDS
- By itself, not a strong analgesic
- Potentiates the effects of other analgesics (NSAID's, opioids)

Non steroidal Anti-inflammatory (NSAID)

- Risks with taking it orally
- May combine with acetaminophen / paracetamol to lower dose
- Helpful with inflammatory pain
- Good value to using it topically

Opioids / Narcotics

- Good analgesic
- Short term use to break the cycle of pain or muscle spasm
- In general, most patients with EDS do not respond well to opioids
- Long term use – risk of opioid hyperalgesia

Adjuvants

- Anti-seizure, anti-depressents
- May use only if there is a neuropathic component to the pain

Anti-seizure medicines

- Gabapentin
- Pregabalin

Anti-depressants

- Tricyclic antidepressants – amitriptyline, nortriptyline, doxepin
- SNRI – duloxetine, milnacipran
- SSRI – not very helpful for pain.

Suggested treatments (future)

- Oxygen (Prof Hamonet)
- Hormonal modulation (Dr. Gompel)
- Levodopa/carbidopa for muscle spasms
- EPSOM (Magnesium) salt bath
- Topical analgesics – predictable absorption

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.

Mineral supplement for fatigue

- Anecdotal experience
- Combination of CoQ10, L-carnitine, creatinine
- Good experience with fatigue

Levodopa-carbidopa

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

Thank you

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