Management of complex pain in children and adults with Ehlers Danlos Syndrome

Pradeep Chopra, MD
Assistant Professor (Clinical)
Brown Medical School.
USA

EDS Awareness 2017
Introduction

• Pain Medicine specialist with a special interest in complex pains in adults and children

• Training and Fellowship, Harvard Medical school in Pain Medicine

• Assistant Professor (Clinical) – Brown Medical School, Rhode Island, USA
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.
# New Terminology

<table>
<thead>
<tr>
<th>Old</th>
<th>New</th>
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<tbody>
<tr>
<td>Ehlers-Danlos syndrome</td>
<td>the Ehlers-Danlos syndromes</td>
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<tr>
<td>Hypermobility EDS</td>
<td>Hypermobile EDS</td>
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<tr>
<td>HEDS / VEDS / CEDS</td>
<td>hEDS / vEDS / cEDS</td>
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<tr>
<td>Joint Hypermobility Syndrome</td>
<td>Hypermobility Spectrum Disorders</td>
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<td>Type 3, III, 4, 2, etc.</td>
<td>Please do not use numeral descriptors. It will now be Hypermobile, Classical, Vascular, etc. or hEDS, cEDS, vEDS, etc.</td>
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</tbody>
</table>
• Ehlers Danlos Syndrome (EDS) is not a disease

• It is a form of the human body
Connective tissue

• The human body is made up of different parts

• They are all connected to each other by connective tissue

• Joints, tendons, ligaments, bones, skin, internal organs are all made up of connective tissue
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
Ehlers Danlos Syndromes

• Two important things to remember:

1. Weak connective tissue
2. Poor joint position sense (proprioception)
Connective tissue – non - Ehlers Danlos Syndromes

• Connective tissue is made of collagen
• The connective tissue is strong
• Does not stretch as much
• Does not break easily
• Heals well when broken
Connective tissue – Ehlers Danlos Syndromes

• Connective tissue is made of collagen
• The connective tissue is weak
• It stretches easily
• It breaks easily
• It does not heal well
Weak scar in EDS
Types of tissue injury in EDS

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

- Micro trauma – small repetitive traumas resulting in tissue breakdown. For example – overstretching muscles and ligaments.
Macro trauma

Micro trauma
Joint Position 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. That is what makes them uncoordinated
Proprioception – Joint sense
Proprioception – Joint position sense

- With repetitive injury, wear and tear of joints, we start to lose proprioception
- This is seen with almost any condition that affects joints EDS, arthritis, athletics
- Patients tend to lose their balance easily
- When they lose balance, the body tends to counteract by straining other muscles.
Proprioception – Joint position sense

• With poor Joint Position sense (Proprioception) we do not use our muscles efficiently.
• This causes fatigue, tiredness and pain
Poor proprioception
Compression clothes

• The brain uses signals from the skin to understand the position of the joints and muscles.

• Wearing compression clothes helps the brain understand the position of the body parts, muscles
Compression garments
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
Proprioception exercises - Wobble board
Proprioception exercises - Stand up paddle board
Muscle strengthening exercises

• Avoid joint loading or joint stressing
• Start very low
• Progress slowly
• Focus on muscle strengthening
• Avoid extreme stretching
Keep limb movements within extreme range of motion
Exercise in water - walking
Avoid swimming – injury to shoulder and neck
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.
The Feldenkrais Method

• It is a type of physiotherapy that helps repair impaired connections between the brain and the body
• Patients with EDS develop inefficient or strained habitual movement patterns
• The Feldenkrais Method teaches new patterns using gentle, slow, repeated movements.
• It uses slow repetition to teach correct and safe movements in EDS
The Feldenkrais Method

• It is based on principles of physics, biomechanics and an understanding of learning and human development.

• This method of exercise is excellent for improving proprioception in Ehlers Danlos Syndrome.

• Can be done sitting or lying down

• Each session consists of comfortable, easy movements within the limits of safety
The Feldenkrais Method

• http://www.feldenkrais.com
Tendonitis and bursitis
Tendonitis and bursitis in EDS

Misaligned bones and tendons

Subluxed bones

Normal State

Inflamed State

Muscle

Inflamed Bursa

Tendon

Pradeep Chopra, MD

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
Subluxations and Dislocations
Pain in subluxation and Dislocations

• When a joint subluxes or dislocates, the pain is usually from muscle spasms around the joint.
• Pain from capsular stretch
• Not as much from the bones
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 Proprioception
- 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.
Head and neck
Common causes of headaches

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

• A common cause of neck pain is posture
• Chin poking forward position
• Correction is easy.
• Before looking at other reasons, correct this first
• If there are other reasons like cervical instability, Chiari malformation etc – these need to be addressed
How Heavy is Your Head?

5.4 Kg / 12 lbs.
How Heavy is Your Head?

5.4 Kg / 12 lbs.  14.5 Kg / 32 lbs.

NORMAL POSTURE  2 INCHES FORWARD

5 cm forward
5.4 Kg / 12 lbs.  14.5 Kg / 32 lbs.  19 Kg / 42 lbs.

NORMAL POSTURE  2 INCHES FORWARD  3 INCHES FORWARD

5 cm forward  7.6 cm forward
Pain from a poor 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
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)
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
Lift the phone, don’t drop your head!
Migraines

• Common in EDS
• Connection between EDS, MCAS, POTS, Dysautonomia
• Patent foramen ovale (a kind of hole in the heart)
• Strong link between diet and migraines.
• Good treatments for migraine.
TYPES OF HEADACHES

Sinus: pain is usually behind the forehead and/or cheekbones

Cluster: pain is in and around one eye

Tension: pain is like a hand squeezing the head

Migraine: pain, nausea & visual changes are typical of classic form

http://www.drtomaroexceptionaldentistry.com/headaches.html
Headaches – Idiopathic Intracranial Hypertension (IIH)

- Also known as pseudotumor cerebri
- Raised pressure inside the head
- Visual disturbances
- Photophobia (sensitivity to light)
- Ringing in the ears (tinnitus)
- Maybe because of narrowing of blood flow (venous sinus stenosis)
- Diagnosis: spinal tap, eye exam, MR venography
- Treatment: weight loss, medicines to decrease CSF, VP shunt, stent
Spontaneous CSF leak

Cerebrum

Ventricular system

Spinal cord

CSF flow
Upper back pain
Upper back pain in EDS

• Usually poor posture
• Shoulder instability
• Rib subluxation
• Repetitive Strain Injury – doing a repetitive task with hands or arms (typing, vacuuming)
• 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 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 (Breast reduction) in severe intractable upper back pain
High Racer back – Posture bra
Compression garments to improve proprioception
Neck
Cervical spine (Neck) issues in EDS

• Cranio Cervical Instability
• C1-C2 instability
• Lower Cervical kyphosis
• Cervical disc degeneration (commonly at C4-C5 and C5-C6)
• Chiari malformation
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) – Upper cervical (neck) – CO to C2

• Neck pain / stiffness
• Headaches
• Dizziness
• Paresthesia to face
• Fatigue
• Poor sleep
• Tinnitus (ringing in ears)
• Nausea
• Poor vision
• Anxiety
• Lightheaded
• Poor balance
• Difficulty swallowing
Atlanto Axial instability

https://www.youtube.com/watch?v=4swxqxpwW0Oc
Atlanto Axial instability (AAI)

• 50% of patients with Rheumatoid arthritis have AAI
• Also common in Marfan’s syndrome
• It is one of the most mobile joints in the body (allow for the head to move in all sorts of angles)
• In EDS – ligament laxity (transverse and alar ligaments)
• As the C1 rotates over C2 to 35 degrees – the vertebral artery on the opposite side gets kinked
• At 45 degrees the opposite vertebral artery is completely occluded
Vertebral artery compression with rotation

- Tinnitus (38%)
- Fainting (24%)
- Blurred vision (19%)
- Headaches
- Difficulty swallowing
- Symptoms improve with neck brace
- Rotational MRI of neck
Atlanto Axial instability

• Neck brace
• Physical therapy to strengthen neck muscles
• Avoid activities that provoke exacerbation
• Surgical fusion if all else fails.
Cranio Cervical Instability

- The ligaments from the skull to the spine are incompetent
- Laxity of the ligaments causes nerve injury
- There is deformation of the brain stem (lower end of the brain) and the spinal cord
- Blood flow is altered
- CSF flow is altered
- Basilar invagination or ventral brainstem compression
Cranio Cervical instability (CCI) – Lower cervical (neck) – C3 to C7

• Muscle spasms
• Crepitation (cracking sensation)
• Neck pain
• Tingling and numbness in fingers
Barrie – Lieou syndrome

- Cervical instability pushing on the autonomic nerves
- Difficulty swallowing
- Tongue numbness
- Blurred vision
- Tinnitus
- Dizziness
- Neck pain
- Headaches

Anterior cervical instability. Injury to the capsular ligaments, allows excessive anterior cervical movement, causing Barré-Liéou Syndrome. Symptoms can include dysphagia, tongue numbness, blurred vision, tinnitus, vertigo, dizziness, neck pain, and migraine headaches.
Basilar Invagination

- C2 pressing against the brain stem causing a narrowing of the foramen magnum

http://www.neuroradiologycases.com/2012/01/basilar-impression-bi.html
Basilar Invagination

• Symptoms are more apparent when the head is bent backwards
• Weakness of arms and legs
• Tingling in arms and legs
• Altered vision, hearing
• Neck pain
• Confusion
• Difficulty swallowing or talking
• Breathing problems
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.
• Functional MRI (fMRI)
• Digital motion x-ray (DMX)
Cranio Cervical instability in EDS – MRI scan findings

• These measurements have to specifically asked for when getting an MRI.

1. Clivo-axial angle (normal 145 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)
  • Prolotherapy – Hackett-Hemwell prolotherapy

• Severe Instability:
  • Surgical fusion
TMJ Pain

Temporo Mandibular Joint Dysfunction
Temporo-Mandibular joint pain
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
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
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
• Local anesthetic may not work or onset will be delayed
Chiari Malformation
Symptoms of Chiari Malformation

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

- Sleep apnea
- Dysautonomia /POTS
- Associated with cranio-cervical instability or basilar invagination
Chiari malformation

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

http://www.craniofacial.vcu.edu/conditions/chiari.html
Chiari Malformation and EDS

• Higher incidence of Chiari in EDS
• Cranial settling (loose ligaments)
• Posterior gliding of condyles
• Reduction of the clivus-axis angles, clivus atlas angle, atlas-axis angle.
Cranial Settling in EDS- Deformative stresses on the brain stem, lower cranial nerves, spinal cord

Clivo-axial angle normal $140^\circ$
MRI for Chiari Malformation in EDS

• MRI in upright position – Important
  • These measurements have to specifically asked for when getting an MRI.
  1. Clivo-axial angle (normal 145 to 160 degrees)
  2. Harris Measurement (instability if > 12mm)
  3. Grabb, Mapstone and Oakes measurement (> 9mm suggests high risk of ventral brainstem compression)
Pain in the back
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
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
Tethered Cord Syndrome
Clinical symptoms

- Low back pain
- Neurogenic bladder
- Leg weakness and sensory loss
- Musculoskeletal abnormalities
Tethered Cord syndrome and EDS

• Tethered cord syndrome present in 14% of patients with Chiari Malformation

• Tethered cord syndrome present in 63% of patients with low lying cerebellar tonsils (i.e. these people did not quiet meet the criteria for Chiari)

• That means 77% of patients with EDS and Chiari symptoms may have Tethered Cord.
Tethered Cord Syndrome (TCS)

Tethered Cord syndrome

- Aching, burning pain in low back, legs and feet.
- Weakness in legs
- Legs – heaviness, stiffness, tightness, cramps
- Tingling or numbness in pelvic area or legs
- Decreased sensation in the area between the legs
- History of toe walking
Tethered Cord Syndrome and EDS

• Check for Tethered cord if diagnosed with Chiari Malformation and vice versa
• Bladder problems:
  • Urinary hesitancy
  • Increased (decreased) frequency,
  • Urgency
  • Sense of incomplete emptying of the bladder
• Nocturia – going often at night
• Irregular stream
• More than 3 urinary tract infections in a year
• Incontinence
Tethered Cord Syndrome and EDS

• Weakness to legs with normal arm strength
• Scoliosis, kyphosis
• Ankle and foot deformities
• Ankle pronation
• Flat feet
Tethered Cord Syndrome and EDS

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

- The spinal cord hangs freely in the spine, it slides up and down in a sheath (much like a sword in a sheath)
- In TCS the lower end is tethered to the bottom of the spine
- As we grow taller, the spinal cord gets stretched.
Neurogenic bladder

• Increased frequency
• Urgency
• Sense of incomplete evacuation of bladder
• Incontinence to urine
• More than 3 urinary tract infections in a year
Tethered Cord Syndrome and EDS

- MRI is **NOT** a useful tool for diagnosing TCS
- Diagnosis is based on clinical history and examination
- A urodynamic study maybe helpful
Breathing 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
• All the tests for heart and lungs are normal.
• Each rib has 3 joints in the back
• The diaphragm is an important muscle for breathing
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
• Book or sand bag should be over the belly button

• Lie on your back

• Breathe in and breathe out for 20 minutes a day

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

- Ribs form joints in the back
- Rib subluxations happen with uncoordinated breathing (remember poor proprioception), and poor posture
- Singing (high and low notes), wind instrument like a flute or recorder
- May help with chest muscle and rib pain and strengthening lower back
Pelvic pain
Pelvic pain in EDS

- Dysmenorrhea (painful menstrual periods)
- Dyspareunia (painful intercourse)
- Sacroiliac joint dysfunction
Pelvic pain in EDS - Dysmenorrhea (painful menstrual periods)

• Birth control to stop periods – may help with joint laxity also
• NSAID’s (ibuprofen, naproxyn)
Pelvic pain in EDS - Interstitial cystitis (bladder pain)

- Maybe part of Mast Cell Activation syndrome
- Cromolyn or Ketotifen
- Palmitoyl ethanolamide (PEA) (www.vitalitus.com)
Pelvic pain in EDS - Dyspareunia (painful intercourse)

- Maybe part of Mast Cell Activation syndrome or EDS
- Hyaluronic acid locally (try Amazon)
Pain in the arms in EDS
Shoulder and arms
Pain in arms

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

**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
Thoracic Outlet Syndrome

- Physical therapy
- Kinesio taping
- Botox injections
- Surgical correction
Kineseo Taping for shoulder joint pain
Hand and wrists
Muscles of the hand

• In EDS, patients tend to grip objects tightly to compensate for poor proprioception

• Small intrinsic muscles of the hand fatigue easily
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
- Kinesio taping is a good option
Do braces make your muscles weaker?

• **NO**
  • It’s a common misconception
  • No brace is tight enough to stop muscles from moving
  • In fact, braces stabilize joints so your muscles can move the joints more efficiently.
Legs and knees
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
Ankles and feet
Flexible flat feet – predominantly in the forefoot
Over-Pronation ankles usually associated with flat feet

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

Weight shifts to the inside of the sole

Left Foot

Normal Foot

Flat Foot (excessive pronation)
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

• 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

Deep heel cup
Firm material
Supportive arch

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Ankle brace to stabilize the ankle joint
Knees
Patella is stabilized muscles of the thigh
• A hypermobile patella can make the knee unstable
• It causes pain in the muscles that support the patella

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

Copyright Pradeep Chopra
Knee pain – often missed cause of pain in the leg

• The proximal tibio-fibular (PTF) joint is on the outside of the knee.
• Like all joints it is prone to subluxations or arthritis.
• A subluxing PTF joint affects the Peroneal nerve, which affects the side of the leg and causes pain in the leg and foot drop.
Proximal Tibio-Fibular joint
Ilio Tibia band (Fascia Lata Fascitis)

- Pain on the side of the thigh up to the knee
- It is usually because of an unstable knee or hip joint

- The IT band is tightened in subluxation of the PTF and hypermobile patella

- Treat the knee or hip problem

- Stretching the IT band may not help
Trochanteric Bursitis

- Pain on the side of the hip
- Worse with lying on that side
- Getting up from a chair after being seated for a while.
- The problem may be because of an unstable hip or knee
Tendonitis and bursitis in EDS

Misaligned bones and tendons

Subluxed bones

Normal State

Inflamed Bursa

Muscle

Tendon

Inflamed State

Courtesy http://www.ateevia.com/bursitis.aspx
Abdominal Pain in EDS
Abdominal Pain in EDS

• Very high incidence
• Gastroparesis
• Laxity of the intestines
• Prolapse of internal organs
• Hernias
Abdominal pain

- Mast Cell Activation syndrome is a very common cause of GI motility issues.
- The intestines are controlled by the same autonomic nervous system that is responsible for Dysautonomia.
- GI motility may be either slow or fast
- Acid reflux – part of EDS and also because of MCAS
Constipation

- May be ‘rectal evacuatory dysfunction’.
- Squatty potty
- Patients with EDS having gastrointestinal issues are often labelled as having “Functional GI disorder”.
- In the true sense Functional GI disorders are not psychological BUT very often gastroenterologists who don’t quite understand EDS use the term to imply the abdominal pain is psychological
Gastrointestinal system (GI system) and Chronic pain

• In chronic pain the diversity of bacteria is less (normally, approximately 1000 different types of bacteria)

• This causes GI inflammation, the lining of the intestines is damaged, and increased production of pro-inflammatory cytokines

• TLR4 receptor activity is increased. This has been associated with inflammation.

• TLR4 is one of the receptors where LDN works.
Gastrointestinal system (GI system) and CRPS

• Are we destroying our ‘friendly’ bacteria with our artificial foods, preservatives, chemicals, antibiotics?
• SIBO – Small Intestine Bacterial Overgrowth – talk to Gastroenterologist
• The test for SIBO is a breath test and can be done at home
Identification and Treatment of New Inflammatory Triggers for Complex Regional Pain Syndrome: Small Intestinal Bacterial Overgrowth and Obstructive Sleep Apnea

Leonard B. Weinstock, MD, FACG,†‡‖ Trisha L. Myers, PA-C,§ Arthur S. Walters, MD,‡ Oscar A. Schwartz, MD,∗‖ Jarred W. Younger, PhD,‡#‡ Pradeep J. Chopra, MD,‡‡‡‡ and Anthony H. Guarino, MD‡‡‡

Complex regional pain syndrome (CRPS) is evoked by conditions that may be associated with local and/or systemic inflammation. We present a case of long-standing CRPS in a patient with Ehlers-Danlos syndrome in which prolonged remission was attained by directing therapy toward concomitant small intestinal bacterial overgrowth, obstructive sleep apnea, and potential increased intestinal mobility. We theorize that cytokine production produced by small intestinal bacterial overgrowth and obstructive sleep apnea may act as stimuli for ongoing CRPS symptoms. CRPS may also benefit from the properties of low-dose naltrexone that blocks microglial Tol-like receptors and induces production of endorphins that regulate and reduce inflammation.

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From the Departments of *Anesthesiology and Internal Medicine, Washington University School of Medicine, St. Louis, Missouri, †Specialist in Gastroenterology, St. Louis, Missouri, ‡Department of Neurology, Vanderbilt University, Nashville, Tennessee, §Department of Medicine, West County Medical Group Center for Sleep Medicine, St. Louis, Missouri, ‖Department of Psychology, Washington University, and ‡‖Department of Neuroscience, Pain and Fatigue Lab, University of Alabama, Birmingham, Georgia, US; #Department of Anesthesiology, Emory University School of Medicine, Atlanta, Georgia, and ‡‡Department of Anesthesiology, Washington University School of Medicine, St. Louis, Missouri, USA. Accepted for publication October 28, 2015.

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Leonard B. Weinstock has served on the Speakers bureau for Salix, Rollevirol (commercial), and Xilons (commercial) for irritable bowel syndrome. He is a member for a management and war PI on a study of naltrexone for CRPS. He is a board member for the National Institute of Arthritis, Diabetes, Digestive, and Kidney Diseases and is a member of the CRPS Treatment Committee of the United Kingdom. Pradeep Chopra, MD, has served as a consultant for DePuy Synthes, and has received grant funding from Abbott and Amgen.
Diet

• Low FODMAP diet – Fructose, Oligosaccharides, Disaccharides, Momoamines, Polyols
• Gluten free
• Cromolyn oral
Caution

• Antibiotics of the fluoroquinolone such as cipro may cause tendonitis and tendon rupture.
Surgery in EDS done to help with pain, actually makes the pain worse.
Behavioral

• Conversion disorder (CD) – less than 1% \(^{(1)}\)

• Munchausen by Proxy (MBP) – 0.5/100,000 \(^{(2)}\)

• EDS – 1% to 10%

• The diagnosis of CD or MBP are often made by providers with little training in Psychiatry and vice versa most psychiatrists have no training in pain conditions.

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.
Dysautonomia / POTS (Postural Orthostatic Tachycardia Syndrome)
Postural Orthostatic Tachycardia syndrome (POTS) - Symptoms

- Fainting, dizziness
- Heart racing (Palpitations)
- Fatigue
- Headaches
- Cold hands and feet
- Poor concentration “brain fog”
- Feeling of constant anxiety
POTS - Postural Orthostatic Tachycardia syndrome - diagnosis

- 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 while lying down, standing up for 10 minutes - preferred

• Tilt table test
Diagnosis of POTS

Increase in heart rate by 40 beats/minute with very little change in blood pressure

5 mins

Standing

Immediately

120/80

118/78

Standing

for 10 minutes

122/80

70

110

105
Treatment of POTS

• Increase oral salts
• Increase oral electrolyte fluids
• Compression tights up to thighs.
• Abdominal binder (wear swim suit 1 size smaller)
• Cardiology consult for Dysautonomia/POTS.
POTS - Postural Orthostatic Tachycardia syndrome

Consult Dysautonomia International for more information and high salt diet recipes.

http://www.dysautonomiainternational.org/
• The constant feeling of dizziness makes patients feel unstable
• The laxity of the joints makes the muscles tighten reflexly
• This constant use 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 simply Dysautonomia – malfunction of the autonomic system.
Mast Cell Activation Syndrome
MCAS
Mast cells

- Cells in blood
- Normally present in blood
- Contain histamine, cytokines and a bunch of other chemicals
- Involved in allergy, wound healing and protection against infection
MEDIATORS RELEASED FROM ACTIVATED MAST CELLS

LIPID MEDIATORS
- PGD$_2$
- LTB$_4$
- LTC$_4$

CYTOKINES (31)
- TNF-$\alpha$
- GM-CSF
- IL-1$\beta$
- IL-3
- IL-6
- IL-10

PREFORMED MEDIATORS
- Serine Proteases
- Proteoglycans
- Histamine
- Carboxypeptidase A
EDS, POTS and MCAS

A New Disease Cluster: Mast Cell Activation Syndrome, Postural Orthostatic Tachycardia Syndrome, and Ehlers-Danlos Syndrome

Cheung, Ingrid, Vadas, Peter

Journal of Allergy and Clinical Immunology, Volume 135, Issue 2, AB65, February 2015
Mast Cell Activation Syndrome (MCAS)

- MCAS is a large, prevalent collection of illnesses resulting from mast cells which are inappropriately activated but are NOT significantly proliferated (different from Mastocytosis).
Never Bet Against Occam

Mast Cell Activation Disease and the Modern Epidemics of Chronic Illness and Medical Complexity

LAWRENCE B. AFRIN, M.D.
Mast Cell Activation Syndrome (MCAS)

- Unexplained skin flushing, unexplained hives
- Headaches
- Sweating
- Flushing after a hot shower
- Mental Fog
- Abdominal discomfort
- Diarrhea, constipation - alternating
- Dermatographism (write on skin)
Signs and Symptoms of Mast Cell Activation Disorders

**Skin (80-90% reactions)**
- Hives
- Itch
- Rashes

**Brain (> 20% reactions)**
- Mental fog
- Headache
- Dizziness
- Confusion

**Heart, Blood Pressure (10-45% reactions)**
- Fast Heart Rate
- Palpitations
- Dizziness
- Fainting

**Airway (70% reactions)**
- Lungs = chest tightness, wheeze, can’t take a deep breath

**Gastrointestinal tract (30-45% reactions)**
- Nausea
- Cramping
- Abdominal Pain
- Vomiting
- Diarrhea

**Genito-Urinary tract (>10% reactions)**
- Uterine Cramping
- Frequent urination
Mast cells

- Inappropriately activated
- MCAS may present as waxing-waning or persistent inflammatory condition
- Activated Mast cells release mediators including histamine and cytokines
Mast Cell Activation Syndrome - Labs

Evidence of an increase in a validated urinary or serum marker of mast cell activation:

1. Tryptase:
   1. levels are persistently >15 ng,
   2. an increase of tryptase greater than the patient’s baseline during a symptomatic period on >2 occasions

2. Less specific but helpful markers are
   1. 24-hour urine histamine metabolites
   2. Prostaglandin D2 or its metabolite 11-b-prostaglandin F2.

3. Bone marrow biopsy
Diagnosis of MCAS

• Think of MCAS if you feel that the presentations is “unusual”, “odd”, “weird”, “bizarre”

• “allergies” to innocuous medications or “allergies” to unusually high number of things
Management of Mast Cell Activation Syndrome (MCAS)

• Anti-histamine:
  • Diphenhydramine, cetirizine (H1 blockers)
  • Ranitidine, famotidine (H2 blockers)
• Cromolyn
• Ketotifene
• Montelukast
• Dietary changes
Management of Mast Cell Activation Syndrome (MCAS)

- 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.
- Low histamine diet.
- 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.
Management of Mast Cell Activation Syndrome (MCAS)

• 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.

• Other methods to avoid exacerbation maybe:
  • Use pollen guards in windows.
  • HEPA air filters, A/C.
  • Use a mask while dusting or vacuuming.
  • Avoid long baths or showers, rapid changes in temperature, wool, dust and cigarette smoke.

• May use nasal saline.
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
Symptoms of EDS, POTS, MCAS

- Headaches, brain fog, dizzy
- Blurry vision
- Racing heart
- Belly pain
- Tremulousness
- Dark discoloration of feet
Nerve pain
Complex Regional Pain Syndrome (CRPS)
Neuropathic pain in EDS

Reflex Sympathetic Dystrophy (RSD)


Family of Neuropathic pain such as CRPS and EDS

• Chronic widespread pain is common in EDS
• Part of this pain is neuropathic and Complex Regional Pain Syndrome (CRPS)


Signs and Symptoms of CRPS

• Pain starts in one limb
• Constant pain, even at rest with intermittent exacerbations. Unexplained and diffuse
• Severe pain
• Temperature change, color change.
• Edema
• Area of pain larger than the primary injury
• Limited range of motion
Signs and Symptoms of CRPS

• Allodynia - pain on light touch
• Creepy, crawly sensation to touch - dysesthesia
• Nail growth changes (faster, distorted), hair growth changes (coarser, darker, rapid growth, hair falling), skin changes (atrophy of skin), skin lesions
What Complex Regional Pain Syndrome is not.....

- There is no such thing as Amplified Pain Syndrome.
- It has been classified as the worst pain known to mankind – worse than amputation of a digit (McGill University)
Color, temperature and swelling

94°

88°

Swelling
Fatigue in EDS
Fatigue

• Plenty of reasons for fatigue
• EDS
• POTS
• MCAS
• Drugs
• Pain
• Poor sleep
Fatigue in EDS

- Myopathy and axonal sensorimotor polyneuropathy (EMG with mixed myopathic and neurogenic) (1)
- They depend largely on their muscle tone to maintain posture at rest because of ligament laxity
- Non-restorative sleep
- Postural Orthostatic Tachycardia Syndrome
- MCAS
- ?secondary mitochondrial dysfunction

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
Fatigue in EDS

- Stimulating the nervous system with amphetamine may not be the best choice
- Correct the underlying cause.
- Combination of ubiquinone and L-Carnitine
- Frequent breaks, do not push through fatigue
- Adequate hydration
Sample mixture of supplements for fatigue

- Ubiquinone: 300mg PO QD
- Riboflavin (B₂): 100mg PO QD
- Acetyl-L-Carnitine: 416 mg PO QD
- Thiamine (B₁): 300mg PO QD
- Pyridoxine (B₆): 50mg PO QD
- Cobalamin (B₁₂): 50 mcg PO QD
- Creatine: 2g PO QD
- Mannitol (filler): 1.834 g

Pradeep Chopra, MD
Other treatments

- Oxygen
- Hormonal modulation
- 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, 20 minutes per day up to twice a day.
• May administer as needed for severe symptoms of fatigue or dizziness.
Levodopa-carbidopa

- Anecdotal experience
- Significant relief for muscle spasms and dystonia
- Low dose
- Sinemet®
Poor concept of exercising in pain

• No pain, no gain
Poor concept of exercising in pain

- No pain, no gain

Pradeep Chopra, MD
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,
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 may be helpful
Medicinal Marijuana

• MM basically contains 2 substances – THC and CBD
• THC works on CB1 and is responsible for the cognitive effects
• CBD works on CB2 and is responsible for pain relief, helps autoimmune dysfunction.
• For MM to work, both THC and CBD have to be together, separating them is not as effective. This is called the Entourage effect.
• One can take MM with a higher concentration of CBD and lower concentration of THC – for pain
• Higher THC and lower CBD for sleep
Cannabis

• Cannabis can reduce the migration of inflammatory chemicals to the site of injury and into the brain.

• This is especially important because in immune dysfunction, migration of inflammatory cells into tissues and nervous system contributes to neuropathic pain
Medicinal Marijuana

• Reasonable choice to try.
• Anecdotally – works well in patients with EDS
• Higher CBD levels and lower THC levels
• Vaporizing, edibles
• Topical over joints and muscles.
• Does not affect Mast Cell Activation Syndrome (MCAS) as much as NSAID’s and opioids
NC10 rule

Expectations from different therapies
NC 10 rule
NC 10 rule
NC 10 rule
NC 10 rule
NC 10 rule

50% relief

10% relief
10% relief
10% Relief
10% Relief
10% Relief

NC10 rule

Pradeep Chopra, MD
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, which....

• Attenuates production of pro-inflammatory cytokines and neurotoxic superoxides (chemicals that cause inflammation)
Low Dose Naltrexone (LDN)

• There are several theories as to how LDN may work.
1. Transiently blocks opioid receptor leading to positive feedback production of endorphins (Zagnon)
2. LDN increases production of OGF (opioid growth factor) as well as number of and density of OGF receptors by intermittently blocking the opiate receptor. Increased in OGF repairs tissue and healing.
3. Naltrexone blocks the effect of TLR4 (Toll Like receptors) which decreases glial cell activation
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.
Treatment of Complex Regional Pain Syndrome (CRPS) Using Low Dose Naltrexone (LDN)

Pradeep Chopra · Mark S. Cooper

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Abstract Complex Regional Pain Syndrome (CRPS) is a neuropathic pain syndrome, which involves glial activation and central sensitization in the central nervous system. Here, we describe positive outcomes of two CRPS patients, after they were treated with low-dose naltrexone (a glial attenuator), in combination with other CRPS therapies. Prominent CRPS symptoms remitted in these two patients, including dystonic spasms and fixed dystonia (respectively), following treatment with low-dose naltrexone (LDN), LDN, which is known to antagonize the Toll-like Receptor 4 pathway and attenuate activated microglia, was utilized in these patients after conventional CRPS pharmacotherapy failed to suppress their recalcitrant CRPS symptoms.

Keywords Chronic pain · Complex regional pain syndrome · CRPS · Reflex sympathetic dystrophy · RSD · Neuropathic pain · Naltrexone · Fixed dystonia · Allodynia · Vasomotor · Ulceraion · Dystonic spasms · Conversion disorder · Functional movement disorder · LDN dysfunctions. One of the characteristic symptoms of this condition is that the pain is out of proportion to the initial injury. Diagnoses of CRPS are often delayed because it is under recognized (Hinkley 2012). If effective treatments are given early enough in progression of the disease, there is reduced chance for the spread of regional pain, autonomic dysfunction, motor changes, and negative sensory symptoms, such as hypoalgesia (Marinus et al. 2011). As CRPS progresses, it becomes refractory to sympathetic nerve blocks, conventional analgesics, anticonvulsants and antidepressants.

During neuroimmune activation, TLR4 (Toll-Like Receptor 4) is upregulated in microglia, resident immune cells of the central nervous system (Watkins et al. 2009). After transection of the L5 spinal nerve in the rat, TLR4 expression is increased in spinal microglia. This correlates with the rodent developing neuropathic pain (Tanga et al. 2005). From a post-mortem analysis of a CRPS patient, activated microglia and astroglia in the central nervous system (CNS) have been implicated in the generation of CRPS symptoms (Del Valle et al. 2009). Activation of TLR4 in both microglia and CNS neurons augments the production of pro-inflammatory cytokines via the NF-kB pathway (Milligan and Watkins 2009; Leow-Deke et al. 2017). NF-kB is a multi-functional transcription factor.
CENTRAL SENSITIZATION

Key concept to understanding all chronic pain
Central Sensitization

• A normal sensation (e.g. touch) produces an abnormal response (like pain) because the brain and spinal cord are sensitized

• Definition: Increase in the excitability of neurons within the central nervous system (CNS) so that normal inputs produce abnormal responses
Central Nervous System

• The Central Nervous system (CNS) is made of 2 parts:

1. Brain
2. Spinal cord
Normal pain

• Normally, an injury will cause pain and the signals are sent to the brain

• In the brain, the signal gets an emotional component and we sense pain
Normal pain

• Once the injury heals, the signals stop and everything returns to normal
CRPS

- In CRPS, the pain signals continue even after the injury heals.
- The brain also tries to send signals down to suppress the pain signals.
- In CRPS, there is a constant barrage of pain signals travelling up and down.
Central Sensitization

• The constant barrage of signals travelling up and down the brain and spinal cord makes the nervous system sensitive
• This is called Central Sensitization
• Hence, normal touch or a minor injury anywhere in the body, magnifies the pain greatly
Central Sensitization in CRPS

• In CRPS (and other chronic pains), the problem lies in the Central Nervous system

• Any treatment for CRPS, should be to treat it at the level of the Central Nervous system

• Treating the pain at the periphery may not help and may even make the pain worse
What really happens in CRPS /Central Sensitization
Central Sensitization

• Two things happen in Central Sensitization:
  1. Glial cells get activated
  2. NMDA receptors are activated
Central Sensitization: Activated Glial Cells

- Glial cells make up 70% of all the cells in our Central Nervous System
- Under normal circumstances, they remain dormant and are part of the nervous system's immune function

Central Sensitization: Activated Glial Cells

- Glial cells make up 70% of all the cells in our Central Nervous System
- Under normal circumstances, they remain dormant and are part of the nervous system's immune function
This is what glial cells look like

Courtesy Jarred Younger, PhD

Sonja Paetau, University of Helsinki

Pradeep Chopra, MD
Central Sensitization: Activated Glial Cells

- In CRPS these glial cells are activated.
- Activated glia release certain chemicals (Cytokines) that cause nerves to become inflamed.
- Glial cells are an important link between the nervous system and the immune system and inflammation and pain.

Glia and nerves under normal conditions
Activated Glia

Nerve

Glia
Chemicals released by activated Glia
Nerve inflammation
The problem is with the glia cells
Graded Motor Imagery
Stage 1: Left/Right discrimination – Graded Motor Imagery

• In CRPS, people often lose the ability to identify left or right images of their painful body parts.
• This ability is important for normal recovery from pain
• The good news is that the brain is plastic and changeable.
• The ‘Recognise’ app helps regain this ability
Stage 2: Explicit Motor Imagery - Graded Motor Imagery

• The process of thinking about moving without actually moving
• Imagined movement can actually be hard work if you are in pain.
• 25% of our brain is made of ‘mirror neurons’ – they start firing when you think of moving or even watch someone else move
• Imagining movements before actually moving you use the same neurons that you would use when you actually move
Stage 3: Mirror therapy- Graded Motor Imagery

• By hiding the affected limb behind a mirror, you can trick the brain into believing that the reflection of the normal hand is the affected limb.

• In your brain you are exercising the affected limb as you move the normal limb.
Three stages of Graded Motor Imagery delivered sequentially

- Left / right discrimination
- Explicit Motor imagery
- Mirror therapy

www.gradedmotorimagery.com

Pradeep Chopra, MD
Pain receptor behavior

• When we take a drug for pain for a long time there is downregulation of the receptors, which means....
• The body’s response to the drug is not as good.
• If we stop the drug for sometime, the receptors are upregulated, which means....
• Restarting the drug gets a better response at a lower dose.
Pain receptor behavior - drug rotation

• Ideally, a person could switch between drugs of a different class.

• For example, a patient on opioids for some months can take a ‘drug holiday’ for a few weeks to months.

• During this time, they can try medicinal marijuana (if legal) or ketamine (sublingual) or NSAID’s

• After some time restart opioids at a lower dose.
Sleep and EDS
Sleep and EDS

• Pain keeps them awake
• If they fall asleep they continue to produce adrenaline (because of Dysautonomia/POTS) they have light, dream-filled sleep
• Increased number of sleep disrupting ‘arousals’
• Wake up unrefreshed – Non-restorative sleep.
Brain activity during sleep – non-restorative sleep pre-treatment
Brain activity during sleep – after beta blocker
Non-restorative sleep

Courtesy Alan Pocinki, MD
Non-restorative sleep

• Good sleep hygiene – comfortable mattress, dark and quite room, no digital lights
• Beta blockers – propranolol
• Alpha blockers – clonidine, guanfacine
• Pain medicines
Connecting the dots.....

• Fatigue (EDS, POTS, MCAS)
• Diffuse pain (EDS, POTS, MCAS)
• Headaches (EDS, POTS, MCAS, TCS, CM)
• Joint pain (EDS, TCS)
• Dizziness (POTS, CM)
• Poor balance (EDS, POTS, TCS, CM)
• GI issues (MCAS, EDS)
• Feeling anxious (POTS, MCAS)
Thank you

Pradeep Chopra, MD
painri@yahoo.com