Pain management in Ehlers Danlos Syndrome

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Disclosure and disclaimer

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

• Training and Fellowship, Harvard Medical school

• Pain Medicine specialist

• Assistant Professor – Brown Medical School, Rhode Island
Pain in EDS by body parts

- Head and neck
- Shoulders
- Jaws
- Chest
- Abdomen
- Hips
- Lower back
- Legs
- Complex Regional Pain Syndrome – CRPS or RSD
Pain in EDS

• From nerves – neuropathic
• From muscles – Myofascial
• From Joints – nociceptive pain
• Headaches
Muscle pain

Myofascial pain
Muscle Pain

• Muscles are held together by fascia – ‘saran wrap’ which is made of collagen

• Muscle spasms or muscle knots develop to compensate for unbalanced forces from the joints
Muscle pain

• Most chronic pain conditions are associated with muscle spasms
• Often more painful than the original pain
• Muscles may tighten reflexively, guarding of a painful area, nerve irritation or generalized tension
Muscle pain

- Myofascial pain syndrome is a chronic form of muscle pain.
- Myofascial pain syndrome centers around sensitive points in your muscles called trigger points.
Treatment of muscle pain

- Treat the cause – joint pain, repetitive use
- Trigger point injections
- Stretching
- Relaxation techniques
Trigger Points

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

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

• The trigger points can be painful when touched.

• The pain can spread throughout the affected muscle and other parts
Manual Techniques

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

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

• Strain-Counterstrain: A positional technique to help relax tight muscles

• Craniosacral Therapy: A gentle technique to help balance the nervous system
Muscle Relaxants

- Cyclobenzaprine (Flexeril)
- Carisoprodol (Soma)
- Tizanidine (Zanaflex)
- Baclofen
- Benzodiazepine (Valium)
Muscle relaxants

• Not true muscle relaxants

• Decrease tone

• Lethargy – good for bedtime dosing to sleep

• Approved for long term use: Baclofen, tizanidine
Risk Factors

- Muscle injury. Stress on your muscles can cause trigger points to form. Repetitive stress also may increase your risk.
- Inactivity. If you've been unable to use a muscle, such as after surgery or after a stroke, you may experience trigger points in your muscle as you start to move it during your recovery.
Nerve pain

Neuropathic pain
Nerve pain in Connective tissue disorder / EDS

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

• Is it Mitochondrial disorder?
• Mitochondria found in cells of the human body
• They produce energy
• Suspect, if the pain is widespread and involves nerves
• Exercise intolerance, muscle weakness, seizures, developmental delays etc.
Neuropathic pain medications

- Gabapentin (Neurontin®)
- Pregabalin (Lyrica®)
- Duloxetine (Cymbalta®)
- Milnacipran (Savella®)
Gabapentin and pregabalin

• Promising results have been shown.

• Gabapentin – go up to 900mg, then 1800mg. Slow acting drug.

• Pregabalin – 150mg to 300mg. Faster acting drug.

• The difference may not be obvious at first but when they come off it, they notice an increase in pain

• Chance of increased dizziness in POTS - usually stops after being on the drug for sometime.

• Start low, go slow
SNRI (selective nor-epinephrine release inhibitors)

- Duloxetine (Cymbalta®)
- Milnacipran (Savella®)
SNRI (selective nor-epinephrine release inhibitors)

• Increase nor-epinephrine levels in the central nervous system
• Increased nor-epinephrine levels help in modulating pain signals
• Milnacipran increases nor-epinephrine by 3 times as compared to duloxetne
• May increase blood pressure and heart rate
Antidepressants

• Tricyclic antidepressants (TCA) well studied in neuropathic pain

• Reuptake blockers of serotonin and noradrenaline (Amitritiyline, nortriptyline) – work well

• SSRI (Prozac, Zoloft) – do not work well for pain
Reflex Sympathetic Dystrophy (RSD)

Complex Regional Pain Syndrome (CRPS)
Common clinical features of RSD

- Continuous burning pain
- Pain disproportionate in intensity to the inciting event
- Pain to touch – Allodynia
- Pain not in any specific nerve distribution or even to the site of injury
- Swelling
- Increased / decreased sweating
Symptoms of CRPS / RSD

• Pain – severe, constant
• Temperature difference
• Hypersensitivity
• Tremor, Involuntary movements, muscle spasms, atrophy (weakening of the muscle)
• Increased sweating
• Color difference
• Bone thinning
• Swelling
• Hair and nail changes
Headaches and Neck pain
Headaches

• Mostly unknown cause
• Cerebrovascular reactivity
• Prone to migraines and Tension Type Headaches
Headaches and neck pain

• Headaches may be caused by neck pain
• Headaches may be from
  – inside the head (Migraines) or
  – outside (chronic daily headaches or tension type headaches)
• Treatment for both is different
Headaches and Neck pain

- C1-C2 instability
- Kyphosis (Bent backwards) of neck spine
- Degeneration of neck disks (especially C4-C5, C5-C6)
- Cranio-cervical instability (junction between head and neck not stable)
- Chiari malformation
Symptoms of neck (C1-C2) instability

- Neck pain
- Headaches in the back of the head (occipital headaches)
- Fainting sensation with rotating head to the side
- Choking sensation
- Treatment – surgical fusion, strengthening of neck muscles, try bracing for flare ups
Migraines

- Treatment depends on the frequency – if they are occasional, do not treat
- Abortives for infrequent migraines
- Prophylactic for frequent migraines
- Triptans (Rizatriptan - Maxalt®, Sumatriptan - Imitrex®, etc – very effective as abortive, not prophylactic)
- Triptans not approved for children under 12 years
- Prophylactic in children – Cyprohetadine - Periactin® Improves appetite also
- Other prophylactics – amitriptyline, nortriptyline, topiramate
Red Flags

• Acute (sudden) abdominal or flank pain may be due to a ruptured uterine, intestine or twisted intestine
• Bleeding in the head may present with sharp pain in head, change in mental status or seizure
• Emergency
TMJ Pain

Temporo Mandibular Joint Dysfunction
TM Joint pain

A: Sternal division

B: Clavicular division
Temporo-mandibular joint dysfunction (TMJ)

- Present in 70%
- Clicking sound
- Grinding at night or clenching teeth
- Joint pain, chewing muscle pain
- Treatment: avoid excessive mouth opening, caution when yawning, orthodontist specializing in TMJ
- Avoid over the counter mouth guards
Arm and leg pain

- Brachial plexus neuropathy
- Lumbosacral neuropathy
- Mechanical pressure due to dislocations and subluxations
- Nerve disorders in EDS make them prone to damage
Spine pain

- Facet joint instability
- Sacroiliac joint pain
- Muscles around the spine
- Degenerative disc disease
Spine pain

- Steroid injections
- Core strengthening exercises
- Temporary brace if flare up
Tendons and Ligaments

- Ligaments and tendons are made of connective tissue
- Ligaments connect bone to bone
- Tendons connect muscle to bone
- Tendons are an extension of the strong connective tissue that surrounds all muscles – the fascia
Tendon and ligament pain

• Bracing or spica
• Topicals
• Preventive measures
  – Avoid overstretches or hyperextending joints
  – Avoid high impact activities
Prevention
Preventing pain

• Do not stand on one hip or sway at hips

• Do not stand on the outside of your feet

• Do not sleep on your stomach, head turned to one side for a prolonged period
Preventing pain

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

• Do not hyperextend knees when standing
Preventing pain

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

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

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

- EDS patients wake up with pain and stiffness
- Use a soft mattress
- Prefer using separate mattresses for partner and yourself
- Feather pillow or memory foam pillows
Preventing pain - shopping

- Do not carry a heavy or light bag hanging from a shoulder
- Do not carry grocery bags in hand, especially heavy one. Keep them light.
- Use shopping trolley. Use wheels wherever you can
- Keep handbags light
Preventing pain – clothes

• Keep clothes loose and light
• Women – bra with wide straps and possibly cross-over straps
• Wear light and soft shoes. Use well balanced orthotics
Preventing pain – going to the dentist

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

- Opioid or narcotics
- NSAID’s (non steroidal anti-inflammatory drugs)
- Acetaminophen / paracetamol
- Anti-depressants
- Anticonvulsants
- Other
LDN

Low Dose Naltrexone
Low Dose Naltrexone (LDN)

- Naltrexone blocks the effect of opioids.
- Approved 30 years ago by the FDA for opioid and alcohol addiction.
- In the late 1980’s it was discovered that at low doses it helped patients with immune disorders.
- Since then, LDN has been used extensively by patients with immune disorders like Multiple sclerosis, Crohn’s disease, Rheumatoid arthritis.

Pradeep Chopra, MD
Low Dose Naltrexone (LDN)

- There are several theories as to how LDN may work
- Low side effect profile
- Inexpensive
- Useful for chronic pain
- Useful for symptoms of EDS
Low Dose Naltrexone (LDN)

• Good experience with using in EDS and chronic pain
• Unsure as to how it helps EDS.
• Most patients with EDS report improvement in function
Acetaminophen / Paracetamol

• One of the safest analgesics
• By itself is good for mild to moderate pain
• Potentiated effect when combined with other analgesics such as NSAIDS (Naproxen, Ibuprofen) or opioids (Codeine, Oxycodone).
• Potentiating effect helps lower the need to take NSAIDs and Opioids.
• Keep an eye on maximum dose – 3000gms/ day (9 regular Tylenols®)
NSAID (Non steroidal anti-inflammatory drugs)

• Naproxen, Ibuprofen etc

• Good analgesics. Better if combined with acetaminophen/paracetamol

• Significant side effects – gastric irritation, kidneys
• Avoid in Irritable Bowel Syndrome, Ulcerative Colitis

• Not useful in neuropathic pain

• May be used in joint pain

• Better off using it as a topical for superficial joints (Shoulder, knee, ankles, hands)
Topicals

• NSAID’s are commonly effective

• Avoids the side effects of oral medicines

• Far more effective locally at the site of pain
Topicals – ointment, lotions, patches

- Compounding pharmacies can make a compound with NSAID’s
- DMSO and Active Max® – use as additive to help better penetration
- Diclofenic acid with DMSO (Pennsaid®)
- Diclofenac acid (Voltaren® gel)
- Flector® patch helpful
- Lidocaine patch not helpful
Opioids

- Counterproductive for chronic pain
- Mild doses for a short term are good for acute pain
- Safer than using NSAID’s (ibuprofen, naproxen)
- Maybe combined with NSAID’s or acetaminophen for better effect and lower dose
Prevalence of Addiction

Addiction: 2% to 5%

Total Pain Population

Gluten free diet

- Gluten is a protein found in wheat, rye, barley and other grains
- One can develop an intolerance at any age.
- Gluten as a protein can cause an inflammatory response in the body.
- Migraines, chronic body ache, abdominal pain, Fibromyalgia, hypermobility syndromes (EDS), multiple joint pains
- Hold off on gluten foods for 8 weeks to see if it makes a difference.
Starting treatment - medicines and exercise

Start low, go slow
Service Dogs

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

- POTS – they can sense when their owner is having an episode of dizziness or seizure
- EDS and pain - they protect the limb from being injured or touched
- Helps boost confidence in their owners.
Watson and Usher
Exercise and Physical Therapy
EDS – Exercise and Physical Therapy Goals

- Restore function
- Learn how to properly adjust limb movements
- Muscle strengthening
- Aqua therapy
Physical Therapy - two types

• Patients who have recently developed pain – PT should focus more on pain

• Patients who have had for a while (Chronic) – PT should be more time based
Physical Therapy – Pain focused

• The level and progression of exercise is determined by the intensity of the pain.
• The intensity of the exercise should be low in cases of (or days of) highly intense pain.
• Similarly, the intensity of exercise should be high if the pain is relatively low.
• Its not harmful if there is an increase in pain for a short duration (approx. 1 to 2 hours) after exercise
Physical Therapy – Pain Focused

• A program is designed based on the patients limitations
• Patient exercise within those limits.
• It builds confidence and increases confidence in using their limbs
• Pain focused PT has been shown to decrease considerably symptoms of recent onset RSD
Physical Therapy – Time focused

• The level of exercise is built over time unrelated to intensity of pain

• Preferred approach in cases of long standing

• Pain focused PT can give way to Time Focused PT and vice versa