Hypermobility 101: An Introduction to Having Hypermobility or Ehlers-Danlos Syndrome

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Slide handouts are available at: https://webspace.clarkson.edu/~lussek/hsd.html
Who Am I?

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• Facilitator of the North America Allied Health Professionals ECHO
• Member of:
  • The Allied Health Working Group of the International Consortium of Ehlers-Danlos Syndromes and Hypermobility Spectrum Disorders
  • Past member of The National Academy of Sciences, Engineering and Medicine Committee on Selected Heritable Connective Tissue Disorders and Disability.
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I do not have any conflicts of interest to report
DISCLAIMER

• I cannot provide individual medical advice in this presentation: I cannot diagnose or provide individual treatment recommendations.

• The information provided here is generally applicable to HSD/hEDS, but your personal situation may be different.

• You should discuss options with your healthcare provider before starting a new management approach.
Learning Objectives

At the end of this session, participants will be able to:

1. List common signs and symptoms of hypermobility
2. Describe common comorbidities, such as Postural Orthostatic Tachycardia Syndrome (POTS) and Mast Cell Activation Syndrome (MCAS)
3. Explain reasons for common pain complaints, and approaches to managing these problems
4. Identify several strategies for managing pain and instability
Hypermobility Lecture Series Schedule

- HSD 101: Intro to HSD/hEDS and self-care
- HSD 102: POTS and POTS self-care, basics of MCAS
- HSD 103: Pain management in HSD/hEDS
- HSD 104: Safe exercise selection and progression with HSD/hEDS
- HSD 105: Posture and joint protection
- HSD 106: Gut issues in HSD/hEDS, POTS, MCAS
- HSD 107: Fatigue in HSD/hEDS and POTS
- HSD 108: Headaches, migraines, & TMJ pain associated with HSD, POTS and MCAS
- HSD 109: Breathing disorders in HSD
- HSD 110: Lumbar instability
- HSD 111: Conservative management of cervical instability
- HSD 112: The vagus nerve
- HSD 113: The role of fascia

I will refer to these if you want more info.
Relevant Handouts Available

https://webspaces.clarkson.edu/~lrussek/research.html

• About HSD/hEDS
  • "Overview of Hypermobility Spectrum Disorder": Overview of HSD/hEDS signs, symptoms, diagnosis & PT management.
  • "Why Zebras are not Horses": Why people with HSD cannot be treated the same as everyone else. Share with your PT.
  • HSD in children and adolescents: Common problems affecting kids and teens.
  • Checklist of physical therapy treatment approaches for HSD/hEDS. What a PT might be able to help with.
  • Surgical precautions for people with HSD/hEDS. Orthopedic surgeries and anesthesia need special consideration in HSD

• Self-Care Strategies
  • Breathing. Breathing incorrectly can increase pain sensitivity, headaches, jaw pain, and more.
  • Posture. Good posture decreases strain on muscles and joints, and can prevent many problems.
  • Joint Protection Strategies Protect your joints and muscles as the first step towards healthier and stronger joints
  • Sleep Hygiene and Positioning. Sleep posture and sleep hygiene strategies.
  • Starting to Exercise. This worksheet helps people identify and overcome roadblocks to being more active.

• Pain Management
  • Pain self-care plan. Create a pain self-care plan to improve your pain management.
  • Pain flare management plan. Create a flare management plan so you know what works when you have a flare.
So, What is EDS?

EHLERS-DANLOS SYNDROME

FAULTY COLLAGEN SYNTHESIS

NOT THE POTS!

POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

INCREASED RISK OF AORTIC ROOT DILATION AND DISSECTING AORTIC ANEURYSM

CHRONIC PAIN

ABNORMAL WOUND HEALING

EASY BRUISING

JOINT HYPERMOBILITY AND HYPEREXTENSIBLE SKIN

Overview of HSD
What Is Connective Tissue?
13+ Types of EDS

- **Hypermobile** (old name Type III): Loose joints, joint pain, connective tissue problems. 90% of all EDS is hypermobile.
  - Previously called Joint Hypermobility Syndrome (JHS or HMS), Benign JHS, or EDS-hypermobility type.
  - At this time, genetic testing is not helpful unless you suspect a form of EDS other than the hypermobile form (HSD/hEDS)
- **Classical** (old names Type I & II): Velvety, stretchy, fragile skin. 2\textsuperscript{nd} most common.
- **Vascular** (old name Type IV): Possible arterial/organ rupture. Rare, but most serious.
- 10 Other sub-types, quite rare.
Classical EDS
Vascular EDS

Common Appearance?
- Translucent thin skin with visible veins especially on face/chest/abdomen*
- Early onset varicose veins*
- Unusual bruising without cause*
- Attached earlobes
- Narrow palate
- Deep-set or almond shaped eyes

*Note, these signs may be present in hEDS, usually less dramatic

www.ehlersdanlosnetwork.org/vascular.html
https://www.pinterest.cl/pin/558868634993169756/
https://www.annabelleschallenge.org/vascular-eds
GJH vs. HSD vs. hEDS

- **GJH** is mostly asymptomatic
- **hEDS** if all 2017 checklist criteria are met
- **HSD** for “all individuals who present with complaints and/or life quality limitations because of joint hypermobility”

- Pain, symptoms, comorbidities & disability are similar for both groups; **hEDS is not worse than HSD**
- Increased severity associated with increased dysautonomia symptoms

Castori, 2017

Aubry-Rozier, 2021; Copetti, 2019
How Common is HSD/EDS?

- We don’t really know, because there isn’t much population research and it is underdiagnosed.
- HSD/hEDS is the most common systemic inherited connective tissue disorder in humans
- Prevalence of hypermobility (including asymptomatic) is reported to be 10-20% in adults and 34% in children hEDS
  - Not all these people have muscle or joint pain
  - Some people present with other problems, such as fatigue, GI pain, etc. first
- Adults in healthcare:
  - Primary care - 30%; pain management - 39%; rheumatology - 37%

Symptoms Through the Life Span

1. **Hypermobile phase**
   - Hypermobile joints
   - Clumsiness/motor delay
   - Constipation/diarrhea
   - Abdominal hernias

2. **Pain phase**
   - Chronic fatigue
   - Unrefreshing sleep
   - Chronic back pain
   - Chronic muscle pain/cramps
   - Strains, sprains
   - Dislocations
   - Anxiety

3. **Pain + phase**
   - Memory/cognitive problems
   - Gastric reflux
   - Recurrent abdominal pain
   - Numbness & tingling
   - Racing heart
   - Incontinence/UTI

4. **Stiffness phase**
   - Tendonosis/tendon rupture
   - Chronic gastritis
   - Stiffness
     - Castori et al, 2011
     - Tinkle et al, 2017
2017 hEDS Diagnostic Criteria

hEDS Must meet all 3 criteria:
1. Generalized joint hypermobility
2. Features of inherited connective tissue disorder
   • Meet ≥2 of 3 categories, A-C
3. Absence of exclusion criteria
   (there isn’t another good explanation)

• Hypermobility Spectrum Disorder exists when people are hypermobile and have some of the hEDS diagnostic criteria
• Pain and symptoms are similar for both groups – one is not worse than the other
• If you are hypermobile and you have problems related to your hypermobility, you have HSD/hEDS

Diagnostic Criteria for Hypermobile Ehlers-Danlos Syndrome (hEDS)

This diagnostic checklist is for doctors across all disciplines to be able to diagnose EDS

The clinical diagnosis of hypermobile EDS needs the simultaneous presence of all criteria, 1 and 2 and 3.

CRITERION 1 – Generalized Joint Hypermobility

One of the following selected:
- Generalized joint hypermobility
- Features of inherited connective tissue disorder
- Absence of exclusion criteria

If Beighton Score is one point below age- and sex-specific cut off, two or more of the following must also be selected to meet criterion
- Can you move (or could you ever) place your hands flat on the floor without bending your knees?
- Can you move (or could you ever) bend your thumb to touch your forearm?
- As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
- Do you consider yourself “double jointed”?

CRITERION 2 – Two or more of the following features (A, B, or C) must be present

Feature A must be present:
- Unusually soft or velvety skin
- Mild skin hyperextensibility
- Unexplained tissue distension or rubes on the back, groins, thighs, breasts and/or abdomen in adolescents, men or pre-pubertal women without a history of significant gain or loss of body fat or weight
- Bilateral piezogenic papules of the heel
- Recurrent or multiple abdominal hernia(s)
- Atrophic scarring involving at least two sites and without the formation of truly papery/hyaline scars as seen in classical EDS
- Pelvic, floor, rectal, and/or uterine prolapse in children, men or nulliparous women without a history of morbid obesity or other known predisposing medical condition
- Dental crowding and high or narrow palate

Feature B:
- Positive family history; one or more first-degree relatives independently meeting the current criteria for hEDS
- Aortic root dilatation with Z-score >+2
- Mitral valve prolapse (MVP) mild or greater based on strict echocardiographic criteria
- Aortic root dilation with Z-score >+2
- feature C total: /2

Feature C:
- Unexplained striae distensae or rubae at the back, breasts, abdomen and/or chest
- Mitral valve prolapse (MVP) mild or greater based on strict echocardiographic criteria
- Aortic root dilation with Z-score >+2

CRITERION 3 – All of the following prerequisites MUST be met

1. Absence of unusual skin fragility, which should prompt consideration of other types of EDS
2. Exclusion of other heritable and acquired connective tissue disorders, including autoimmune rheumatologic conditions. In patients with an acquired CTD (e.g. Lupus, Rheumatoid Arthritis, etc.), additional diagnosis of hEDS requires meeting both Feature A and Feature B of Criterion 2
3. Exclusion of alternative diagnoses that may also include joint hypermobility by means of history, physical examination, and/or molecular genetic testing, as indicated.

Patient name: ___________________________ DOB: __________ DOV: __________ Evaluator: ___________________________

The International Consortium on Ehlers-Danlos Syndrome & Related Disorders

https://www.ehlers-danlos.com/heds-diagnostic-checklist/
THE BEIGHTON SCORING SYSTEM
Measuring joint hypermobility

Overview of HSD

Gain 1 point if ≥2/5 on the 5-Item Questionnaire:
1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
4. As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
5. Do you consider yourself “double-jointed”?

- Officially: prepubescent ≥6; proposed:
  - 3–7 years (≥5 for males, ≥6 for females);
  - 8–18 years (≥4 for males, ≥5 for females)

Singh, 2017; Nicholson, 2022

A. 5th FINGER / ‘PINKIES’
Test both sides: Rest palm of the hand and forearm flat on the surface with palm side down and fingers out straight.
Can the fifth finger be bent/lifted upwards at the knuckle to go back beyond 90 degrees?
If yes, add one point for each hand.

B. THUMBS
Test both sides: With the arm out straight, the palm facing down, and the wrist then fully bent downward, can the thumb be pushed back to touch the forearm?
If yes, add one point for each thumb.

C. ELBOWS
Test both sides: With arms outstretched and palms facing upwards, does the elbow extend (bend too far) upwards more than an extra 10 degrees beyond a normal outstretched position?
If yes, add one point for each arm.

D. KNEES
Test both sides: With knees lockedwards as far as possible, the lower part of the extended leg more than forward?
If yes, add one point for each side.

Used with permission from the Ehlers-Danlos Society, https://www.ehlers-danlos.com/assessing-joint-hypemobility/
Those are not the only important joints

- Beighton Score joints are only special for making an official diagnosis.
  - Other hypermobility assessment tools may be helpful
- Patients may be hypermobile in other joints, such as the shoulder, neck, hip, lumbar spine, etc.
- You should assess any joint that is problematic

Other GJH assessment tools:

- **Upper Limb Hypermobility Assessment Tool:**
  - ≥ 7/12 for GJH (Nicholson, 2018)
- **Lower Limb Assessment Score:**
  - ≥ 7/12 for GJH (Meyer, 2017)
- Older scales:
  - Carter Wilkinson Scoring System
  - Rotes-Querol Scale/criteria
  - Hospital del Mar Scale/criterion

(Nicholson, 2022)
The Terrible Trifecta: HSD, POTS, MCAS

HSD:
- Joint hyperextensibility, tissue fragility
- Subluxations/dislocations
- Easy bruising, CNS issues

MCAS:
- Flushing, pruritis, urticaria
- Dermatographia, respiratory congestion
- Wheezing, throat swelling

POTS:
- Light headedness, vertigo, syncope
- Presyncope, dumping syndrome

GERD, heartburn
- Pain, weakness, fatigue, chest pain
- Abdominal pain, nausea, vomiting, diarrhea
- Bloating, constipation
- Hypotension, exercise intolerance, brain fog
- Palpitations, sleep disturbance

Orthostatic intolerance, dizziness

Seneviratne, 2017; Hakim, 2017
Symptoms of POTS

POSTURAL ORTHOSTATIC TACHYCARDIA SYNDROME

SYMPTOMS

- Dizziness
- Heart palpitations
- Sweating
- Chest pain
- Fatigue
- Shortness of breath

& HOW TO TREAT IT

- Increase Salt Intake
- Increase Fluid Intake
- Avoid Caffeine
- Eat Smaller Meals & Fewer Carbohydrates
- Avoid Prolonged Standing

Great POTS info at: www.potsuk.org and http://www.dysautonomiainternational.org

https://zovon.com/latest-health-news/pots/

Russek: Hypermobility 101

HSD 102: POTS & MCAS
Symptoms of MCAD/S

Some common symptoms of Mast Cell Disease that are caused by mast cell mediator release include:

**Neurological**
- Headache, brain fog, cognitive dysfunction, anxiety, depression

**Cutaneous (Skin)**
- Flushing of the face/neck/chest
- Hives, skin rashes
- Itching with or without rash

**Ear/Nose/Throat/Respiratory**
- Nasal itching and congestion
- Throat itching and swelling
- Wheezing
- Shortness of breath

**Gastrointestinal**
- Diarrhea, nausea, vomiting, abdominal pain, bloating, gastroesophageal reflux disease (GERD)

**Skeletal**
- Bone/muscle pain, osteopenia, osteoporosis

**Cardiovascular**
- Light-headedness
- Syncope (fainting)
- Rapid heart rate, chest pain
- Low blood pressure
- High blood pressure at the start of a reaction
- Blood pressure instability

**Gynecological**
- Uterine cramps
- Bleeding

**Urinary**
- Bladder irritability
- Frequent voiding

**Systemic and/or Organ Specific**
- Anaphylaxis
- Angioedema (swelling)

Symptoms can be sudden and unpredictable in onset.

Learn more at tmsforacure.org

HSD 102: POTS & MCAS
Questions?
Common Complaints and Why They Occur
Musculoskeletal Problems in EDS

- Being hypermobile does not necessarily cause pain
  - There are hypermobile people who are pain free
  - **Hypermobility ➔ Instability**
- Being hypermobile makes people more vulnerable:
  - Poor body awareness allows inappropriate/damaging movement
  - Poor motor control, leads to instability
  - Instability causes stress to the joints and fascia
  - Overworked muscles try to stabilize joints
    - Causing muscle spasm, trigger points and/or tendonopathy
    - Strain from tight muscles, gravity, or poor body mechanics
    - Muscle spasm from sympathetic nervous system activation
- Better understanding can help you become less vulnerable, and have less pain and instability
Examples: Common Musculoskeletal Problems

• Ankle: Traumatic ankle sprain
• Knee: Patellofemoral knee pain
• Hips: Trochanteric pain syndrome (aka “bursitis”)
• Spine: Muscle spasm
• Shoulder: Instability/subluxation/dislocation
• Neck: Trigger points pain
• Hands: Joint instability
• Nerve problems
Ankle Sprain

- Stretchy ligaments don’t control motion
- Poor body awareness & balance allows excessive motion
- Weak muscles don’t control movement well

We cannot change ligaments. Ankle brace or taping to control motion if/when necessary.

Body awareness (proprioception); Balance training.

Compression sleeve and taping may improve proprioception/body awareness.

Muscle strengthening.

Russek: Hypermobility 101
Knee: Patellofemoral Pain

- Loose ligaments allow too much motion of kneecap
- Standing with hyperextended knees allows kneecaps to float
- Tight thigh muscles pull kneecap outward
- Flat feet and hip weakness allow knee to turn inward

Russek: Hypermobility 101
Knee: Patellofemoral Pain

• Loose ligaments allow too much motion of kneecap
• Standing with hyperextended knees allows kneecaps to float
• Tight thigh muscles pull kneecap outward
• Flat feet and hip weakness allow knee to turn inward

Can’t change the ligaments. Train muscles to pull inward. Patellar knee brace; Taping.

Learn to not hyperextend knees.

Carefully stretch tight muscles.

Orthotics can align foot & leg; Strengthen foot, ankle & hip muscles.
Hip: Trochanteric Pain

• Formerly called ‘trochanteric bursitis’ but actually involves tendon

• Dropping pelvis while standing/walking aggravates muscle & tendon
  • Due to weakness and/or poor body awareness

• Flat feet allow knees and hips to turn inward, pulling on hip muscles/tendons
Hip: Trochanteric Pain

• Formerly called ‘trochanteric bursitis’ but actually involves tendon

• Dropping pelvis while standing/walking aggravates muscle & tendon
  • Due to weakness and/or poor body awareness

• Flat feet allow knees and hips to turn inward, pulling on hip muscles/tendons

• May need to rest muscles and tendons to heal. May benefit from a cane to rest.

• Strengthen hip muscles.

• Improve body awareness. Learn to activate muscles properly.

• Orthotics can align leg & hip; Strengthen foot, ankle & hip (rotator) muscles.
Low Back: Spasm

• Muscles tense to brace unstable spine
• Poor body awareness leads to using improper muscles or overusing proper muscles
• Using wrong muscles to breathe increases muscle tension
• Pain and fear of moving increases muscle spasm

HSD110: Lumbar Instability

Picture from https://www.spineuniverse.com/conditions/back-pain/muscle-spasms-leading-cause-back-pain-not-primary-cause

Russek: Hypermobility 101
Low Back: Spasm

- Muscles tense to brace unstable spine
- Poor body awareness leads to using improper muscles or overusing proper muscles
- Using wrong muscles to breathe increases muscle tension
- Pain and fear of moving increases muscle spasm

Train stabilizing muscles to do their job. Learn good body mechanics.

Train body awareness. Learn how to activate stabilizing muscles.

Learn to use diaphragm muscle to breathe, so it can help with spinal stability.

Practice careful/correct movement to learn how to move safely. Gradually gain confidence to move.
Shoulder Instability

- Loose ligaments allow too much movement
- Weak muscles don’t hold shoulder in its proper place
- Poor body awareness allows excess motion
- Poor motor control allows shoulder to slip
- Poor alignment of the shoulder leads to subluxation
Shoulder Instability

- Loose ligaments allow too much movement
- Weak muscles don’t hold shoulder in its proper place
- Poor body awareness allows excess motion
- Poor motor control allows shoulder to slip
- Poor alignment of the shoulder leads to subluxation

Can’t change the ligaments quickly, but realigning shoulder so ligaments are less stretched out allows tissues to snug up. Taping or shoulder brace may decrease excessive movement.

Strengthen proper muscles.

Improve proprioception/body awareness. Compression clothing/taping may help.

Train proper motor control.

Improve posture, muscle balance.
Hand and Finger Instability

• Loose ligaments allow too much movement and movement in wrong directions
• Stress on joints due to use
• Using too much force because of laxity or poor body awareness
• Overuse of finger muscles causes trigger points
Hand and Finger Instability

• Loose ligaments allow too much movement and movement in wrong directions
• Stress on joints due to use
• Using too much force because of laxity or poor body awareness
• Overuse of finger muscles causes trigger points

Can’t make ligaments shorter, but not overstretching may allow tissues to snug up. Splints can limit motion & prevent wrong motion.

Learn joint protection strategies.
Use adaptive tools to decrease joint stress.

Use adaptive tools and splints.
Improve body awareness.

Decrease muscle overuse through adaptive tools, splints, joint protection, & awareness.
Hand and Finger Instability

HSD105: Posture & Joint Protection

Braces & Splints

Russek: Hypermobility Tools
Neck Trigger Points

• Instability of neck joints causes pain and muscle spasm
• Poor posture puts joints in bad alignment, overworks muscles
• Poor proprioception & body awareness increase instability
• Weakness of stabilizing muscles leads to overuse of outer muscles, which develop trigger points
• Stress and anxiety increase muscle tension
Neck Trigger Points

- Instability of neck joints causes pain and muscle spasm
- Poor posture puts joints in bad alignment, overworks muscles
- Poor proprioception & body awareness increase instability
- Weakness of stabilizing muscles leads to overuse of outer muscles, which develop trigger points
- Stress and anxiety increase muscle tension

Strengthen deep stabilizing muscles.

Improve posture and strengthen postural muscles in neck, shoulders and spine.

Train proprioception & body awareness.

Strengthen deep stabilizing muscles. Learn to recruit the correct muscles.

Learn physical and psychological stress management techniques

Russek: Hypermobility 101

Headache Trigger Points
Causes of Nerve Pain

• Stretching nerves due to stretchy tissues and posture
• Compression due to hypermobile bones and bad joint alignment
• Compression due to tight muscles
• Inflammation (e.g., MCAS)
• Stress makes nerves more sensitive
Questions?
Approach to Management of HSD

Assist patient in identifying and managing systemic comorbidities: education, treatment and/or referral

Decrease central, peripheral, and autonomic pain sensitization

Educate for correct posture and joint alignment, body mechanics, joint protection, appropriate use of splints and braces

Proprioceptive and motor control training, with training to relax muscles that are guarding

Stabilization, strengthening, muscle flexibility, aerobic conditioning

Integration of proper alignment & movement into function

Education about flare management
Managing Pain

HSD 103: Managing Pain

Several Pain Handouts

https://steemit.com/employment/@mariebellehelene/fully-flexible

Russek: Hypermobility 101
General Self-Care and Wellness

- Sleep hygiene
- Fatigue management
- POTS self-management
- Diet and gastrointestinal wellness
- Psychological & social wellness
- Body mechanics/ergonomics
  - Braces & splints
- Pacing, prioritization, activity/exercise selection
- Appropriate exercise/activity
Fix the “Issues with your tissues” when you can!
- HSD pain is not ‘all in your head’
- People with HSD are vulnerable to pain/injury

In HSD, stress on tissues is persistent, so tissues often don’t heal.
- HSD has “persistent pain,” which is different from “chronic pain.”

“Chronic pain” is typically associated with a sensitive nervous system that amplifies pain
- Psychological pain management can often help, but won’t fix “issues with your tissues”
FYI: Visceral Referred Pain “Issues”

- Visceral tissues (internal organs) can refer pain
- Pain referral can irritate tissues at the referral site
- This can cause tissue irritation at the referral site
  - Example: intestinal problems can cause trigger points in abdominal muscles, which then hurt
    - Gebhart, 2016
Pain Management: Interventions

• Controlling pain, once you have managed “issues with your tissues”...

• Modalities:
  • Ice for joint inflammation and severe muscle spasm
  • Heat for muscle achiness and mild spasm
  • TENS to decrease pain in muscles or joints

• Topical ointments
  • Mentholated, anti-inflammatory, CBD, capsaicin

• Manual therapy
  • For muscle or fascial pain and restrictions
  • To improve joint alignment

• Manage sensitive nervous system (see HSD103)
Pain Management: Neurosensitivity

• Even though there are real, physical issues with your body, neuroplasticity (nerve learning) can change pain processing in your brain: “Sensitization”

• This is “volume control” for pain

• Stress, anxiety, negative thinking, depression all turn up the “volume control”

• Passive interventions (massage, ultrasound, etc.) often do not work for this type of pain, or only work temporarily (e.g., feel better for a few hours)

• Vagus nerve activation can calm nerves (HSD112)
Benefits of Exercise/Activity

• Regular (appropriate) exercise/activity reduces pain
  • Protects against chronic pain onset
  • Sedentary lifestyle increases risk of chronic pain
• Protects against autonomic dysfunction (POTS)
• Improves function
• Improves sleep quality, decreases fatigue
• Mind-body practice, such as Pilates, yoga, Tai Chi enhance body awareness and relaxation
• Improves mood, decreases anxiety
• Decreases systemic inflammation

Lima et al, 2017
Sabharwal, 2016
Ambrose, 2015
Not All Exercises Are Appropriate

• For exercise to be helpful and not harmful, it must be:
  • The correct exercise (for you, now)
  • Done correctly (proper motor control)
  • At the correct dose (intensity, time/reps)
  • Not overstressing other joints or muscles
• There is no protocol appropriate for everyone with EDS/HSD

• Avoid:
  • Positions that overstretch joints
  • High impact sports/activities
  • Sudden head-up postural change
  • Excessive weight lifting/carrying, joint distraction

• Start low, go slow!
Braces & Splints are popular among people with EDS: [https://www.bauerfeind.com/b2c/](https://www.bauerfeind.com/b2c/)

Silver Ring Splints are popular: [https://www.silverringsplint.com](https://www.silverringsplint.com)

**Braces & Splints**

**EDSers:**

we have a brace/splint for that...

**fb you know you have EDS when**

Russek: Hypermobility 101
Compression Garments for Proprioception

- Thanks to Stephanie Carroll, RN, for suggesting these full body compression garments
  - Bauerfind makes many EDS-appropriate devices: [https://www.bauerfeind.com/b2c/](https://www.bauerfeind.com/b2c/)
  - CWX makes sports compression garments: [https://cw-x.com](https://cw-x.com)
Medications

• Little definitive research evidence for medications
• NSAIDs for true inflammation
  • NSAIDs may slow tissue healing, aggravate GI Sx
• Tricyclics, anti-seizure, SNRI meds for neuropathic pain
• Topical analgesics and anti-inflammatory medications
• Acetaminophen
• Low dose naltrexone (LDN)
• CBD, medical marijuana
• Cautions:
  • Opiates for short term use only
  • Muscle relaxers may aggravate instability

Chopra, 2017
Tinkle, 2017

Russek: Hypermobility 101
HSD: Surgery Precautions

• Surgery is less likely to be successful in people with HSD/hEDS
  • Tissues are more fragile
  • Blood vessel fragility increases bleeding
• Surgery is only effective 34% of the time (Rombaut, 2011);
  • 50% as often as non-hypermobile pts?
• Tissue healing is delayed (Ericson, 2017)
• Special procedures for skin sutures: closer together, leave sutures in longer (Burcharz, 2012)
Summary

• You can learn to manage the “Issues with your tissues” by:
  • Identifying and addressing contributing factors
  • Learning joint protection strategies
  • Doing appropriate exercises, motor control, body awareness training to address those contributing factors
  • Using orthotics, braces, splints, or taping when appropriate

• Pain management strategies also help:
  • General wellness
  • Physical approaches
  • Psychological approaches


Journal Article References


Additional Resources

- **EDS-specific**
  - Ehlers-Danlos Society: [www.ehlers-danlos.com](http://www.ehlers-danlos.com)
  - Hypermobility Syndromes Association: [www.hypermobility.org](http://www.hypermobility.org)
  - My website (with slide handouts) [https://webspace.clarkson.edu/~lrussek/hsd.html](https://webspace.clarkson.edu/~lrussek/hsd.html)

- **POTS and MCAS:**
  - POTS: [www.potsuk.org](http://www.potsuk.org); Dysautonomia International: [http://www.dysautonomiainternational.org](http://www.dysautonomiainternational.org)
  - MCAS: [https://www.tmsforacure.org](https://www.tmsforacure.org)

- **Accommodations**
  - Job Accommodation Network EDS guideline (also has POTS): [https://askjan.org/disabilities/Ehlers-Danlos-Syndrome.cfm](https://askjan.org/disabilities/Ehlers-Danlos-Syndrome.cfm)
  - The School Toolkit: [https://theschooltoolkit.org](https://theschooltoolkit.org)

- **Chronic pain related**
Thank You!
Questions?