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

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Slide handouts are available at: https://webspace.clarkson.edu/~lrussek/hsd.html

Who Am I?

- Professor Emeritus, Physical Therapy Department, Clarkson University
- Staff PT, St. Lawrence Health System, Potsdam NY
  - Clinical specialties: hypermobility, fibromyalgia, headaches, temporomandibular disorders
- Frequent presenter to professional and patient groups at national conferences
- Author of multiple review and research articles on hypermobility
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I do not have any conflicts of interest to report.
DISCLAIMER

• I cannot provide individual medical advice in this presentation
• 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. Describe common signs and symptoms of hypermobility
2. Identify common comorbidities, such as Postural Orthostatic Tachycardia Syndrome (POTS) and Mast Cell Activation Syndrome (MCAS)
3. List reasons for common pain complaints, and approaches to managing these problems
4. Identify several strategies for managing pain, once “issues with the tissues” have been addressed
“Hypermobility 101” Lecture Series

- 6/18/21 HSD 101: Basics of HSD/hEDS and self-care
- 6/25/21 HSD 102: POTS and POTS self-care, basics of MCAS
- 7/9/21: HSD 103: Pain management in HSD/hEDS
- HSD 104: Safe exercise selection and progression with HSD/hEDS
- (HSD 105: HSD/hEDS in children)
- HSD 106: Gut issues in HSD/hEDS, POTS, MCAS
- HSD 107: Fatigue in HSD/hEDS and POTS
- HSD 108: Headaches, migraines, and TMJ pain associated with HSD, POTS and MCAS
- HSD 109: Breathing dysfunctions in HSD

So, What is EDS?

I will refer to these if you want more info

https://medcomic.com/medcomic/ehlers-danlos-syndrome/
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.

• **Classical (old names Type I & II):** Velvety, stretchy, fragile skin. 2nd most common.

• **Vascular (old name Type IV):** Possible arterial/organ rupture. Rare, but most serious.

• 10 Other sub-types, quite rare.

Classical 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
The Hypermobility Spectrum

- hEDS if all checklist criteria are met
- HSD for “all individuals who present with complaints and/or life quality limitations because of joint hypermobility” (Castori, 2017)
- Pain, symptoms, & disability are similar for both groups – one is not worse than the other
  - Patients who are hypermobile and have problems related to their hypermobility, have HSD/hEDS
- At this time, genetic testing is not helpful unless you suspect a form of EDS other than the hypermobile form (HSD/hEDS)
  - Reports of a genetic marker for hEDS are in the public, but not yet reviewed/published in the scientific literature

A Confusion of Names

- Generalized joint hypermobility (GJH): asymptomatic
- Old terminology
  - Joint hypermobility syndrome (JHS) & hypermobility syndrome (HMS): terms often used by rheumatologists
  - Ehlers-Danlos Syndrome – hypermobility type (EDS – HT or type III) terminology used by geneticists
- Newly proposed terminology: hypermobile Ehlers-Danlos Syndrome (hEDS) and Hypermobility Spectrum Disorder (HSD)
- I will refer to it as ‘hypermobility’ or ‘HSD’
How Common is HSD/EDS?

- Overall prevalence in US: 1-3% (we think)
- 80-90% of all EDS is hEDS
- hEDS in musculoskeletal healthcare: 30-55%\(^1\)
  - In Omani women, perhaps not this high in the US
- hEDS is the most common systemic inherited connective tissue disorder in humans\(^2\)
- Affects ~10 million people in the U.S.\(^2\)

1. Clark & Simmonds, 2011
2. Tinkle, et al, 2017

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

### 1: Generalized Joint Hypermobility

**Beighton Score ≥5/9**

- ≥2/9 pre-puberty, ≥4/9 over 50 yrs old
- 2: Bend pinky back >90°
- 2: Touch thumb to forearm
- 2: Elbow hyperextension >10°
- 2: Knee hyperextension >10°
- 1: Touch palms to floor, knees straight

Can 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 teenagers, did your shoulder or kneecap dislocate on more than one occasion?
5. Do you consider yourself “double jointed”?

**Russek: Hypermobility 101**

Those are not the only important joints...

• But these particular joints are only important for making an official diagnosis.
• You may be hypermobile in other joints, such as the shoulder, neck, fingers, etc.

Common Comorbidities

• Developmental delay in children (including ADHD)
• Chronic pain: fibromyalgia, myofascial pain, arthritis
  • Pain sensitivity (Scheper et al, 2017)
• Dysautonomia: Postural Orthostatic Tachycardia Syndrome (POTS), anxiety, gut problems, sexual dysfunction, poor temperature control
• Mast Cell Activation Disorder/Syndrome (MCAD/S): systemic inflammation, gut problems, skin problems
• Gastrointestinal disorders: constipation, GERD, IBS, malabsorption syndrome
  • Tinkle et al, 2017
Symptoms of POTS

[Diagram of POTS symptoms]

Great POTS info at: [www.potsuk.org](http://www.potsuk.org) and [http://www.dysautonomiainternational.org](http://www.dysautonomiainternational.org)

Symptoms of MCAD/S

[Diagram of MCAD/S symptoms]

HSD 102: POTS & MCAS

Leslie Russek; Clarkson
Why HSD, POTS, MCAS?

• These 3 conditions frequent co-exist
  • “The Terrible Trifecta”
  
  Seneviratne, 2017; Hakim, 2017

• Together, they look a lot like fibromyalgia
  

• “Hypermobility 102” will discuss diagnosing and managing POTS and MCAS

Questions?
Common Complaints and Why They Occur

Musculoskeletal Problems in EDS

- Being hypermobile, alone, does not necessarily cause pain
  - There are many people who are hypermobile and pain free
- HOWEVER... being hypermobile makes you more vulnerable to musculoskeletal imbalances that cause pain & instability
  - Ligaments are loose
    - Joints slip out of alignment, or gravity pulls them out
    - Tight muscles pull loose joints out of alignment
  - Body awareness (proprioception) is decreased
  - Muscles have often become weak
- Better understanding can help you become less vulnerable, and have less pain and instability
Examples: Common 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.
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

- Muscles/tendons are overstretched because weak hip muscles don’t stabilize hip
- Dropping pelvis while standing/walking aggravates muscle & tendon
  - Due to weakness and/or poor body awareness
- Loose hips may ‘slip’ out of alignment
- Flat feet allow knees and hips to turn inward, pulling on hip muscles/tendons

May need to rest muscles and tendons to heal.
Strengthen hip muscles.

Avoid stress while tissues heal.
Using a cane may help.

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

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

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

Russek: Hypermobility 101

![Normal and Unstable Shoulder](https://orthop.washington.edu/patient-care/articles/shoulder/atraumatic-shoulder-instability.html)

Can’t change the ligaments quickly, but re-aligning 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.

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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.

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

- 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

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
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

Pain Management: Tissues

- Fix the “Issues with your tissues”!
  - EDS pain is not ‘all in your head’
  - Many types of chronic pain are known to continue after tissues have healed
  - HOWEVER, in EDS, stress on tissues is persistent, so tissues often don’t heal.
    - EDS has “persistent pain,” which is different from “chronic pain.”
  - Your neural processing of pain may be amplified (typical of chronic pain), but that is not the only problem.
    - Psychological pain management can sometimes 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, (capsaicin)
- Manual therapy
  - For muscle or fascial pain and restrictions
  - To improve joint alignment
- Managing neuroplasticity...
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

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

- Bauerfeind braces are popular among people with EDS: https://www.bauerfeind.com/b2c/
- Silver Ring Splints are popular: https://www.silverringsplint.com

Compression Garments for Proprioception

- Thanks to Stephanie Carroll, RN, for suggesting these full body compression garments
  - Bauerfeind makes many EDS-appropriate devices: https://www.bauerfeind.com/b2c/
  - CRC makes sports compression garments: https://crxc.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
• Cautions:
  • Opiates for short term use only
  • Muscle relaxers may aggravate instability

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 (Burcharth, 2012)
Pain Management: Neuroplasticity

• 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.

Managing Neuroplasticity

• Relaxation training:
  • Diaphragmatic or slow breathing
  • Meditation, “Tapping,” etc.
  • Stress management
  • Biofeedback, e.g., HRV, breathing (can now be done with smart-phone apps)
• Hope, gratitude, positive attitude
• Find help with this approach on-line
  • Curable app: www.curablehealth.com
    • Curable is now available free for 6 weeks through a health care provider
Additional Resources

- EDS-specific
  - Ehlers-Danlos Society: [www.ehlers-danlos.com](http://www.ehlers-danlos.com)
  - Hypermobility Syndromes Association: [www.hypemobility.org](http://www.hypemobility.org)
  - My website (with slide handouts) [https://webspace.clarkson.edu/~russek/hsd.html](https://webspace.clarkson.edu/~russek/hsd.html)
  - Job Accommodation Network EDS guideline: [https://askjan.org/disabilities/Ehlers-Danlos-Syndrome.cfm](https://askjan.org/disabilities/Ehlers-Danlos-Syndrome.cfm)
  - POTS: [www.potsuk.org](http://www.potsuk.org)
  - MCAS: [https://www.tmsforacure.org](https://www.tmsforacure.org)
- Potsdam Fibro/EDS Support Group: [https://webspace.clarkson.edu/~russek/pfsg.html](https://webspace.clarkson.edu/~russek/pfsg.html)
- Chronic pain related

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
Questions?

Journal Article References

Thank You!