Physical Therapy for Hypermobility Spectrum Disorders

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Learning Objectives

• After completing this session, participants will be able to:
  1. Interpret common patient signs and symptoms with respect to intervention needs
  2. Identify appropriate physical tests and measures for patients with HSD
  3. Propose appropriate physical therapy interventions including lifestyle, physical, functional and activity modification with HSD
  4. Formulate exercises guidelines that will not flare patients with HSD

Patient Variability

• Just like no two zebras have exactly the same stripes...
• No two patients have exactly the same issues
• There are no ‘recipes’ for HSD
• It is not just about joint flexibility

Major Comorbidities

• Chronic pain: fibromyalgia, myofascial pain, OA, TMD
• Hyperalgesia (Scheper et al, 2017)
• Developmental delay in children
• Dysautonomia: POTS, thermoregulation, gut, sexual dysfunction
• Mast Cell Activation Disorder: systemic inflammation
• Gastrointestinal disorders: GERD, IBS, malabsorption syndrome
• Tethered cord syndrome (Henderson et al, 2017)

HSD 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
3. Pain + phase
   • Memory/cognitive problems
   • Gastric reflux
   • Recurrent abdominal pain
   • Parasthesias
   • Tachycardia
   • Incontinence/UTI
4. Stiffness phase
   • Tendonosis/tendon rupture
   • Chronic gastritis
   • Stiffness
   • Castori et al, 2011
   • Tinkle et al, 2017

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Where To Start?

- Focus on patient's deficits & goals
- Look for the key structures/problems that are causing the patient's deficits
- Start at the biggest complaint & work toward smaller issues
- Each patient is unique

Which symptoms bother them the most?

- Fatigue
- Autonomic
- Psychological/cognitive
- Other: GI, UG, immune, skin, cardio

Managing Pain

Pain is often the first and most common complaint in HSD

1. Figure out the type(s) of pain
2. Identify the source(s) of pain
3. Fix the cause, whenever possible
4. Alter pain processing when necessary

Biopsychosocial Model of Pain

Types of Pain

- CENTRAL SENSITIZATION: Central nervous system becomes hyper-responsive
- DEEP NOCICEPTIVE: "normal" pain sensing in response to threats in the environment; sharp
- INFLAMMATORY: increased pain sensing that raises sensitivity to normal levels of pressure and activity; aching
- "Sterile inflammation"
- NEUROPATHIC: abnormal pain in response to things that normally 'do' and 'don’t' hurt; burning, tingling, tingling.
Nociceptive Pain

- Mechanical pain is generally proportional to tissue stress or damage.
- Inflammatory pain remains after tissue damage has occurred.
- ‘Sterile inflammation’ exists without injury; occurs due to sensitive nerves.

Deep Nociceptive Pain

- Due to deep musculoskeletal tissues (e.g., bone, ligament, bursae, etc.)
- Vague, cramping, aching pain
- Sometimes referred
- Changes based on movement or stress to those deep tissues

Visceral Nociceptive Pain

- Vague, often referred to a different location
- Changes based on organ activity (e.g., digestion, increased heart rate, etc.)

Muscle Trigger Point Pain

- Trigger points (TrP) are local, hyper-irritable muscle fibers
- They cause pain in typical referral patterns
  - Some TrP cause other types of symptoms
- In people with HSD, unstable joints often provoke TrP in surrounding muscles that are overworked trying to provide stability

Neuropathic Pain

- Occurs when there is damage to nervous system
  - Peripheral nerves
  - Spinal cord
  - Brain
  - Pain is caused by the faulty nervous system
  - Pain might be felt in locations other than where the damage is
  - The symptoms shown here may be present as well

Central Sensitization

- The central nervous system can become oversensitive
  - It is like “turning the volume up” on pain
- Often aggravated by psychological and social factors such as stress, anxiety, etc.
Physical Exam

- Tests to determine
- Type of pain
- What tissues are causing symptoms?
- What imbalance or deficit is leading to those problems?
- Patient’s baseline function
- POTS: poor-man’s tilt table test

Identify the Source of Pain

- Identify symptom-causing tissues
- What structure is it?
- The problem might not be where the pain is

Trochanteric Pain Syndrome

- Often (mis)diagnosed as trochanteric bursitis
- Most likely a problem in the gluteus medius muscle
- Trigger point
- Muscle tear
- Tendinitis/tendinosis
  
  (Bird et al, 2001)

Trigger Point

- Sternocleidomastoid (SCM)
- Symptoms
  - Headache, migraine, jaw pain
  - Ringing/fullness in the ear
  - Dizziness or vertigo
  - Blurry/double vision, drooping eyelid
  - Sore throat, trouble swallowing
  
  Travell & Simons

Example:
- Trigger Point
  - Gastrocnemius
  - Inflammation where ribs connect to the sternum.
  - Pain may be sharp, aching, or pressing.
  - Pain increases when you take a deep breath or cough.
  - Might feel like a heart attack.

Example:
- Costochondritis

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Find the **Cause**

- Find the biological problem if you can
  1. Is there an imbalance between lax joints and tight muscles?
  2. Does poor posture or gravity put stress on joints or muscles?
  3. Are body mechanics stretching or stressing a joint?
  4. Is poor motor control leading to instability?
  5. What is causing muscle trigger points?

1. Mechanical Imbalances

   - Example: Upper Crossed Syndrome:
     - Imbalance between weak and tight muscles
     - Pulls spine out of alignment and aggravates spinal instability
     - Causes neck, shoulder & upper back pain, headaches, pain & numbness into arms and hands

2. Poor Posture

   Poor neck posture puts a huge strain on the joints and muscles of the neck.

   Hansraj KK. Assessment of stresses in the cervical spine caused by posture and position. *Surg Techn Int.* 2014

2. Posture: What’s Different in HSD?

   - Patients present with unusual positional faults, especially where gravity is the problem
   - Hanging on ligaments

3. Body Mechanics

   - Ergonomics at school/work/home/car
   - Assistive devices (e.g., pens, tools, etc)
   - Sleeping posture, surface, support

3. Overstretched Joints

   Example: The weight of blankets pulls ankles down, causing ankle pain or trigger points in the shin and calf muscles

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4. Poor Motor Control

- Common locations: neck, low back, shoulder, knee-cap

5. Trigger Points

- What is causing the TrP?
- Piriformis TrP causes:
  - Sacroiliac malalignment
  - Leg length difference, flat foot
  - Prolonged driving or sitting
  - Hip joint replacement
  - Impact trauma
  - Running
  - Sexual activity/positioning

POTS:
Poor Man’s Tilt Table Test

- Pt lies supine 5 minutes, asses HR, BP, Sx
- Pt stands stationary (does not move legs)
- Assess HR, BP Sx at <1 min, 5 min, 10 min
- HR increase ≥ 30 (adults), ≥ 40 (children) without orthostatic hypotension
- E.g.

<table>
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<th></th>
<th>Supine</th>
<th>1 min</th>
<th>5 min</th>
<th>10 min</th>
</tr>
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<tr>
<td>HR</td>
<td>73</td>
<td>106</td>
<td>122</td>
<td>NA</td>
</tr>
<tr>
<td>BP</td>
<td>103/72</td>
<td>109/80</td>
<td>118/75</td>
<td>NA</td>
</tr>
</tbody>
</table>

Physical Therapy Intervention

Key Components of PT Program

- Patient education
- Assistive devices
- Pain management
- Manual therapy (if/when appropriate)
- Exercise
  - Engelbert et al, Clinical Guidelines, 2017
  - And clinical pearls from experience

Patient Education

Educate and empower the patient/family

- Pain education & self-management
- Body mechanics/ergonomics
  - Braces & splints
- Appropriate exercise/activity
- Sleep hygiene
- POTS self-management
- Diet and fluid management
- Psychological & social wellness

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Hypermobile Ehlers-Danlos Syndrome

Posture and Gravity
- Muscles unable to overcome gravity may need support
- Pillows, Kinesiotape, posture brace, etc.

Bracing and Taping
- Multidirectional instability common
- Hips, shoulders, SI, knees, ribs, fingers, feet...
- Learn to use muscles to brace for the activity
- Patients should self-manage painful areas as they arise – use braces only as needed
  - Don’t over-use braces
  - Benefit may be from proprioceptive feedback

Manage Trigger Points
1. Teach pt to identify the TrP
2. Figure out what causes the TrP and fix it
3. Self-management of TrP with TrP pressure release, myofascial release, ice, topical ointments (e.g. Biofreeze)
4. Teach proper muscle stretching (careful not to stretch lax joints)
5. Strengthen the muscle only after resolving TrP

Trigger Point Pressure
- TheraCane
- Tennis Balls

Trigger Point Resources
- Pain Relief with Trigger Point Self-Help, by Valerie DeLaune
- http://www.triggerpoints.net
- TrP devices: Thera Cane Massager, Pressure Pointer, Backnobber, Reflexball.

Pain Self-Management
- Techniques to decrease pain:
  - Self-care with heat, ice, TENS
  - Relaxation, meditation, biofeedback
  - General exercise
  - Topical rubs
    - Mentholated (cold) – best for TrP
    - Capsaicin (hot) – good for joints, neuropathic pain. Needs to be used regularly
    - Medicated (e.g., trolamine salicylate) – good for superficial structures, e.g., fingers, ribs, kneecap

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Self-Care with Exercise

- Regular exercise:
  - Modulates/inhibits pain processing
  - Protects against autonomic dysfunction (e.g., POTS)
  - Improves function
  - Improves sleep quality, decreases fatigue
  - Improves mood, decreases anxiety
  - Improves immune function
  - Decreases systemic inflammation

Pain Management: TENS

- Transcutaneous electroneural stimulation (TENS) using evidence-based settings
  - Conventional: 80-150 Hz
  - Acupuncture-like: 2-8 Hz
  - Increase until painful, then decrease slightly
  - 'don't set as 'strong but comfortable'' because this under-doses
  - Most effective when used during activity
  - Can enhance central pain modulation

Neuro-Cognitive Approaches

- Decrease sensitization of the nervous system:
  - Relaxation,
  - Meditation
  - Biofeedback
  - Cognitive behavioral approaches
  - Pain neuroscience education: "Explain Pain"

Relaxation Training

- Yoga, Tai Chi, Ai Chi, Qigong, aquatic exercise
- Diaphragmatic breathing
- Meditation (e.g., mindfulness, guided relaxation, visualization)
- Biofeedback (e.g., heart-rate variability)
- Physiological quieting

Pain: Patient Resources

- UC Davis pamphlet: search "UC Davis pain self-management plan"
- American Chronic Pain Association: [https://www.theacpa.org](https://www.theacpa.org)
- On-line pain self-management: [https://www.liveplanbe.ca](https://www.liveplanbe.ca)
- Pain Toolkit, on-line videos and modules: [https://www.paintoolkit.org/tools](https://www.paintoolkit.org/tools)
- For adolescents: [http://www.growingpains.org](http://www.growingpains.org)

Fatigue & Sleep Disorder

- The 2nd most common complaint
- Sometimes the primary complaint
  - (Oleim et al, 2017; Engelbert et al, 2017)
- Good patient resource:
  - [https://sleepfoundation.org](https://sleepfoundation.org) for sleep hygiene
  - "The Insomnia Answer: A Personalized Program for Identifying and Overcoming the Three Types of Insomnia" by Glovinsky & Spielman
Managing Fatigue

- Patient education to:
  - Address causes
  - Improve sleep hygiene
  - Apply cognitive behavioral approaches
- Employ graded exercise therapy
  - Excellent on-line resource: “Graded Exercise Therapy: A self-help guide for those with chronic fatigue syndrome/myalgic encephalomyelitis”

POTS Self-Management

- Education & reassurance
  - Explain link to panic and anxiety (Tinkle et al, 2017)
  - http://www.dysautonomiainternational.org
- Increase isotonic fluid intake (H₂O + salt)
- Compression stockings
- Decrease orthostatic intolerance
  - Change positions slowly; isometric exercises before movement, ankle pumps
  - Avoid hot environment, large meals, alcohol, vasodilator meds, sympathomimetic meds
  - When fatigued or panicking, lie down with feet elevated

GI Self-Management

- Dietary advice
  - Identify and treat abdominal muscle TrP, which can cause constipation, diarrhea, bloating, gas, cramps, indigestion
  - Rectus abdominus
  - Abdominal obliques
  - Transversus abdominus
  - Physiological quieting (relaxation)

Pain Management (in Clinic)

- Manual therapy
- Dry needling (if legal in your state)
- Physiological quieting
- Biofeedback
- TENS (trial)
- Avoid modalities in the clinic

Manual Therapy

- Relieve mm spasm, TrP, & fascial adhesions
- Decrease pain & autonomic tone
  - Physiological quieting
- Realign joints carefully
  - “Mobilization with Movement,” muscle energy techniques, neurodynamics
  - Stabilize nearby structures
  - Abdominal propulsive massage

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### Manual Therapy
- **DO NO HARM**
- Do not over-mobilize
- Especially upper cervical instability
- Treat cautiously as you would a hypermobile pregnant patient
- Be localized, specific and controlled
- Or refer to manual therapy specialist

### Exercise Prescription
- Proprioception, stabilization, motor control & coordination
- Strengthening
- Appropriate stretching
- Cardiovascular conditioning
  - “Graded Exercise Therapy”
  - (POTS-specific exercise)

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### Exercise: Do No Harm!
- Research shows that many patients with HSD have negative past physical therapy experiences due to:
  - Iatrogenic joint injuries
  - Unmet rehabilitation needs
    - Bovet et al, 2016

### Exercise: Do No Harm!
- Avoid mechanical pain
  - Stabilization & motor control are critical
    - Think before moving
    - External stabilization may be necessary
  - Tensile strength of tissues varies with activity, menses, inflammatory state, age etc.
  - Slow progression to allow histological changes

### Exercise: Do No Harm!
- Caution with overuse: nerve entrapment, trigger points, tendinosis
- Caution with neural tensioning (e.g., tethered cord)
- Caution with changing forces
  - e.g., exercise bands increase resistance
- Allow tissue recovery between exercise bouts
  - 2-hour rule: discomfort should return to baseline within 2 hours

### Appropriate Exercise/Activity
- Avoid:
  - High impact sports/activities
  - Sudden head-up postural change
  - Excessive weight lifting/carrying, joint distraction
  - Try Qigung, Tai Chi, Pilates, yoga, etc. as multipurpose exercise
    - These provide strengthening, proprioception, balance, and calm the nervous system

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Instability & Coordination/ Balance Deficits

- People with HSD have decreased sense of where they are in space – i.e., proprioception
- Weakness due to deconditioning increases instability and balance problems
- If people become fearful and limit their activity, weakness and proprioception get worse
  - Engelbert et al, 2017

Exercise: Proprioception

- External focus exercises – put a laser on it and draw on the wall, wobble boards
- Biofeedback (e.g., Stabilizer™)
- Alternating isometrics, dynamic stabilization
- Emphasize good motor control
  - https://www.motionguidance.com

Exercise: Balance

- External focus exercises – put a laser on it and draw on the wall, wobble boards
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- Alternating isometrics, dynamic stabilization
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Progressing Exercises

**EASIER**
- More support, stable surface
- Isolate individual joints
- Focus and visualize
- Slow, mid-range motions
- Low impact
- Low effort (RPE ~4/10)
- Progress slowly

**HARDER**
- Less support, unstable surface
- Integrate multiple joints
- Add distractions
- Faster, full-range motions
- Higher impact
- Higher effort (RPE ~7.5/10)
- Follow 2-hour rule

Graded Exercise Therapy

- If POTS is present, address that
- Must be carefully done to avoid aggravating the fatigue or the HSD
  - “Start low, go slow!”
  - It may take months to years to recover fully

POTS Exercise

- Progress exercises gradually
  - Start horizontal, progress to vertical
  - Start with compression garments
  - Gradual progression of exertion
  - Allow for recovery days
  - Focus on aerobics and lower extremity resistance. Pool exercise good.
- Children’s Hospital of Philadelphia (CHOP) protocol http://standinguptopots.org/fnuwhf285hy9hjewf/misc30-chop-modified-dallas-pots-exercise-program
- Graded Exercise Therapy approach (see prior resource)
In Summary: There is a lot PT can do to minimize Patient problems! But it takes some detective work.

Books

For health professionals
- Keir & Grahame. Hypermobility Syndrome: Diagnosis and Management (for Physiotherapists), 2003
- Hakim & Ker. Hypermobility, Fibromyalgia and Chronic Pain. 2010

For patients

Resources

EDS information
- www.ehlers-danlos.com
- Has Am J Med Gen EDS articles from March, 2017
- Suggested adaptations for children in school
- http://hypermobility.org (UK)
- Excellent support pages for children & teens
- http://medicalzebras.com
- http://ehlersdanlosnetwork.org
- www.oneEDSvoice.com
- www.TheZebraNetwork.com

Dysautonomia information

For Questions?
- We have been seeing these patients for years
- We just didn’t know it
- Now we know

THANK YOU!

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