Hypermobility Spectrum Disorders (HSD)

Aka “Hypermobility Syndrome,” (HMS) or “Hypermobile Joint Syndrome” (HJS) or “Joint Hypermobility Syndrome” (JHS)

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- Hypermobility Spectrum Disorder (HSD) is the most common systemic inherited connective tissue disorder in humans
  - In the UK prevalence of HSD associated with chronic widespread pain or severely disabling pain (Mulvey, et al, 2013) is almost as high as fibromyalgia (Fayaz et al, 2016) and 100 times higher than rheumatoid arthritis (Humphreys et al, 2013)
  - 80-90% of all EDS is hEDS. HSD affects ~10 million people in the U.S. (Tinkle, et al, 2017)

Common complaints

- **Musculoskeletal**: joint hypermobility, subluxations/dislocations, sprains, muscle spasm, jaw pain, flat feet, finger deformities, joint pain, muscle pain, fractures, widespread pain or hypersensitivity
- **Skin**: stretchy skin, easy bruising, atrophic scarring, poor wound healing, frequent hernias
- **Cardiovascular**: dysautonomia (poor temperature control, poor blood pressure or heart rate control, passing out)
- **Gastrointestinal**: constipation, diarrhea, gas, abdominal pain, gastritis, indigestion, food intolerance
- **Other**: developmental delay, poor coordination, anxiety, trouble sleeping, fatigue, brain fog, frequent infections, allergic reactions, medication sensitivity, incontinence, organ prolapse, sexual dysfunction

- Castori et al, 2011; Columbi et al, 2015; Tinkle et al, 2017

Major Comorbidities (Tinkle et al, 2017)

- Chronic pain: fibromyalgia, myofascial pain, osteoarthritis, temporomandibular pain, chronic headaches
- Developmental delay in children
- Dysautonomia: postural orthostatic tachycardia syndrome (POTS), orthostatic hypotension, chronic fatigue
- Mast Cell Activation Disorder/Syndrome (MCAD/S):
  - GI disorders: gastroparesis (slow digestion), constipation, GERD, IBS, malabsorption syndrome, food intolerance
  - Tethered cord syndrome (Henderson et al, 2017)

hEDS complaints tend to fall in 3 phases, But each individual may have specific problem areas – see diagram below

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

2. **Pain phase**
   - Chronic musculoskeletal pain
   - Strains, sprains, dislocations
   - Unrefreshing sleep
   - Chronic fatigue
   - Memory/cognitive problems
   - Gastric reflux, abdominal pain
   - Paresthesias
   - Tachycardia, anxiety, panic attacks
   - Incontinence/UTI

3. **Stiffness phase**
   - Tendinosis/tendon rupture
   - Chronic gastritis
   - Stiffness

- Castori et al, 2011, Tinkle et al, 2017

**Diagnostic Criteria**

- Until 2017:
  - **Beighton Scale** most often used for GJH
  - **Villafranche Classification** used mostly by geneticists for children: EDS-HT
  - **Brighton Criteria** used mostly by rheumatologists for adults: JHS

- New criteria for hEDS (see Ehlers-Danlos Society web page for details: www.ehlers-danlos.com)
  - They are rather complicated and are very strict because researchers are looking for a genetic defect – many people who had met criteria for JHS or EDS-HT in the past do not meet the current diagnostic criteria for hEDS.

- No clear guidelines regarding HSD
  - A person who is hypermobile, symptomatic, and does not meet criteria for hEDS, if no other diagnoses explain it.
Beighton Score

- 2: Bend 5th finger back >90°
- 2: Touch thumb to forearm
- 2: Elbow hyperextension >10°
- 2: Knee hyperextension >10°
- 1: Palms to floor, knees straight

Before puberty ≥6/9; puberty to 50 years ≥5/9; Over 50 years ≥4/9

- And may earn 1 additional point if you score ≥2/5 Yes responses to:

Add 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”?

Physical Therapy Evaluation

- Focus on primary activity & participation restrictions
  - Start at the biggest complaint & work toward smaller issues
- Identify tissues causing symptoms
- Identify stressors affecting those tissues
  1. Is there an imbalance between lax joints and tight muscles?
  2. Does poor posture or gravity stress joints or muscles?
  3. Are body mechanics stretching or stressing a joint?
  4. Is poor proprioception or motor control leading to instability?
  5. What is causing muscle trigger points?
  6. POTS test, if appropriate

Physical Therapy Treatment Approach must be specific to each patient’s needs. (Engelbert et al, Clinical Guidelines, 2017)

Education

- Educate and empower the patient/family to manage the condition, since it cannot be ‘cured’
- Pain education & self-management: body mechanics, ergonomics, joint protection, TENS, topical medications, self-management of myofascial trigger points
- Body mechanics/ergonomics
- Orthotics, braces & splints if needed; canes, crutches, if condition is very involved
- Appropriate exercise/activity
- Sleep hygiene & fatigue management (also see POTS and Graded Exercise Therapy)
- POTS self-management (for fatigue, exercise intolerance, anxiety, panic, difficulty concentrating, sleep disorder)
- Psychological & social wellness, including relaxation, mindfulness meditation, social contact
- Diet and fluid management – some people benefit from the Heidi Collins diet, FODMAP diet, or low histamine diet
- Other issues: GI dysfunction, MCAD, incontinence, pain with intimacy, etc.
- Refer to other professionals as appropriate

Exercise

- Stabilization, motor control & coordination and body position awareness
- Strengthening, careful not to stress joints; “start low, go slow” as tissues are fragile and strengthen slowly
- Appropriate stretching, careful to stabilize loose joints
- Cardiovascular conditioning using good body mechanics; “start low, go slow”
  - “Graded Exercise Therapy” provides good guidelines for pacing and progressing exercise very slowly
- POTS-specific exercise for patients who have POTS: horizontal rather than upright, lower extremity first, etc.

Pain Management

- Pain management, teaching self-care (see above), TENS, myofascial release,
  - Relaxation and physiological quieting, using mindfulness meditation, biofeedback, Tai Chi, yoga, etc.
- Manual therapy (if/when appropriate to re-align joints): must be careful because joints are loose.
- Address musculoskeletal injuries, such as sprains, trigger points, tendinosis, etc.