

# Hypermobility Spectrum Disorders (HSD)

Aka “Hypermobility Syndrome,” (HMS) or “Hypermobile Joint Syndrome” (HJS) or “Joint Hypermobility Syndrome” (JHS)  
 Information compiled by Leslie Russek, PT, DPT, PhD, OCS, Clarkson University and Canton-Potsdam Hospital.

- Hypermobility Spectrum Disorder (HSD) is the most common systemic inherited connective tissue disorder in humans
  - 80-90% of all EDS is hEDS . HSD affects ~10 million people in the U.S. (Tinkle, et al, 2017)
  - Many musicians are hypermobile, and flexibility may be beneficial for playing.

## 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, anxiety, 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 – other conditions that may also be present (Tinkle et al, 2017)

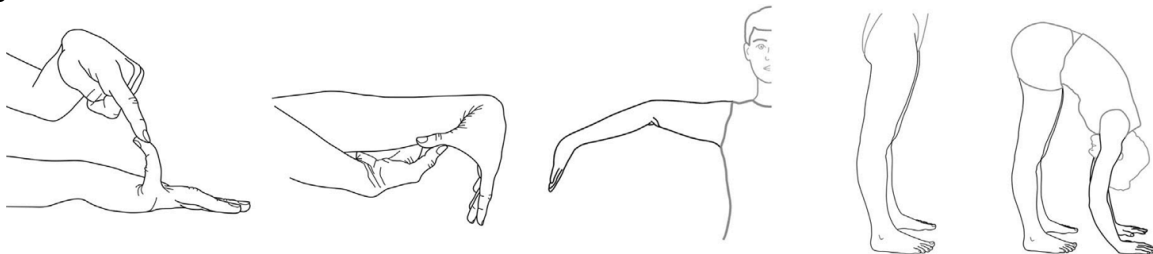
- Chronic pain: fibromyalgia, myofascial pain, osteoarthritis, temporomandibular pain, chronic headaches
- Developmental delay in children
- Dysautonomia: postural orthostatic tachycardia syndrome (POTS), orthostatic hypotension, chronic fatigue, anxiety
- Mast Cell Activation Disorder/Syndrome (MCAD/S): overactive immune system, frequent infections
- GI disorders: gastroparesis (slow digestion), constipation, GERD, IBS, malabsorption syndrome, food intolerance
- Neurological problems such as tethered cord syndrome or Chiari (Henderson et al, 2017)

HSD complaints tend to fall in 3 phases, but each individual may have specific problem areas, or problems at any age/phase

| Hypermobile phase  | Pain phase   | Stiffness phase  |
|--|--|--|
| Hypermobile joints<br>Clumsiness/motor delay<br>Constipation/diarrhea<br>Abdominal hernias | Chronic musculoskeletal pain<br>Strains, sprains, dislocations<br>Unrefreshing sleep and chronic fatigue<br>Memory/cognitive problems<br>Gastric reflux, abdominal pain<br>Paresthesias, numbness, tingling<br>Tachycardia (racing heart), anxiety, panic attacks<br>Incontinence/urinary tract infections | Tendinitis/tendon rupture<br>Chronic gastritis<br>Stiffness<br><br>• Castori et al, 2011, Tinkle et al, 2017 |

## Diagnostic Criteria

- 2017 criteria for **hEDS** (see Ehlers-Danlos Society web page for details: [www.ehlers-danlos.com](http://www.ehlers-danlos.com))
  - The hEDS criteria 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.
  - Before 2017, we used the Brighton Criteria and Villafranche Classification, so you may still see these.
- **hEDS vs. HSD**
  - HSD is diagnosed in someone has generalized hypermobility (Beighton score, shown below) and has some of the characteristics of hEDS but not enough to have hEDS, and no other diagnoses explains the complaints.
- **Beighton Score**



**Beighton Score (see pictures, above) Cutoff: Before puberty  $\geq 6/9$ ; puberty to 50 years  $\geq 5/9$ ; Over 50 years  $\geq 4/9$**

- 2: Bend 5<sup>th</sup> finger back  $>90^\circ$
- 2: Touch thumb to forearm
- 2: Elbow hyperextension  $>10^\circ$
- 2: Knee hyperextension  $>10^\circ$
- 1: Palms to floor, knees straight

If you miss the cut-off by 1 point, you can add 1 point by scoring  $\geq 2/5$  on the “5-Item Questionnaire” below:

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”?

## **Other Joints May Also Be Hypermobile**

The neck, fingers and shoulders may also be hypermobile.  
See pictures, right.



CORRECT RIGHT THUMB POSITION: Place the thumb under the clarinet thumbrest just above the first knuckle of the thumb. Notice the backwards C of the right hand around the clarinet body.

## **Understanding the Problem**

- When people have many complaints, focus on the ones that have the greatest impact on their lives
  - Start with the biggest complaint & work toward smaller issues
- Identify tissues causing symptoms
- Identify stressors affecting those tissues – this is critical! You need to fix the cause of symptoms, not just symptoms
  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 or placing too high a demand on muscles?
  4. Is poor coordination or motor control leading to instability?
  5. If there are muscle trigger points, what is causing them?
  6. Are you fatigued, anxious, trouble sleeping? Consider POTS

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

### **Education**

- **Learn to manage the condition, since it cannot be 'cured'**
- Pain education & self-management: body mechanics, ergonomics, joint protection, TENS (trans-cutaneous electrical stimulation, aka 'electronic massage'), topical rubs, self-management of myofascial trigger points
- Body mechanics/ergonomics: using your body wisely, good posture, not stretching joints or straining muscles
- Orthotics, braces & splints if needed; canes, crutches, if condition is very involved, or if involving fingers
- Appropriate exercise/activity (in general, low impact, controlled activity; aquatic exercise, Pilates, Tai Chi etc. are ideal)
- 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: occupational therapist, nutritionist, psychologist, gastroenterologist, etc.

### **Exercise**

- Stabilization, motor control & coordination and body position awareness. Always start by moving correctly!
- Strengthening, careful not to stress joints; "start low, go slow" as tissues are fragile and strengthen slowly
- Appropriate stretching of tight muscles, careful to stabilize loose joints
- Cardiovascular conditioning using good body mechanics; "start low, go slow." Follow POTS guidelines if necessary
- "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: learning about pain, cognitive behavioral strategies such as: pacing, stress management, avoiding negative thinking
- Relaxation and physiological quieting, using mindfulness meditation, biofeedback, Tai Chi, yoga, etc.
- Self-care using exercise, TENS ('electrical massage'), braces/splints, topical pain relievers, etc.
- Manual therapy (if/when appropriate): myofascial release, gentle joint realignment, etc.
- Address musculoskeletal injuries, such as subluxations, sprains, trigger points, tendinosis, impingement, etc.

### **Resources**

- The Ehlers-Danlos Society web site: [www.ehlers-danlos.com](http://www.ehlers-danlos.com) has excellent information on EDS for both patients and providers, guidance for school accommodations, and full text of the 18 professional publications on EDS from 2017.
- The Hypermobility Syndrome Association (HMSA): [www.hypermobility.com](http://www.hypermobility.com) has excellent information on hypermobility for both patients and providers, including an excellent section for children and adolescents.
- POTS UK, Postural (Orthostatic) Tachycardia Syndrome: [www.potsuk.com](http://www.potsuk.com) has excellent information on POTS for both patients and providers.
- Chronic pain self-management resources:
  1. The American Chronic Pain Association: [www.theacpa.org](http://www.theacpa.org);
  2. "Live plan be" [www.liveplanbe.ca](http://www.liveplanbe.ca);
  3. PainACTION guide: <http://painaction.wpengine.com/wp-content/uploads/2017/08/Your-Guide-to-Pain-Management.pdf>
- Additional links and resources are available at my EDS web page: <https://webpace.clarkson.edu/~lrussek/hsd.html>