Hypermobility spectrum disorders (HSD) are a group of conditions related to symptomatic joint hypermobility (JH). HSD refer to symptomatic hypermobility after excluding other forms of Ehlers-Danlos Syndrome (EDS), including hypermobile EDS (hEDS). The diagnosis of HSD or hEDS is clinical; there is no identifiable genetic cause, so no test is available. HSD and hEDS can be equal in severity, and need similar management, validation, and care. There is no cure for HSD or hEDS.

**ASSESSMENT**

Perform a standard health history and physical exam, with probing history questions for both patient and family.

**HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS**

Prior to referring for HSD or hEDS, consider other conditions which require additional workup, referral or testing.

**Personal History**
- Unusual skin fragility (which should drive consideration of other EDS types)
- Skeletal dysplasia (e.g., osteogenesis imperfecta)
- Spasticity
- Low muscle tone
- Common neuromuscular manifestations due to a known condition
- Rheumatologic symptoms

**Family History**
- Self or first-degree relative:
  - Aortic disease/aortic root dilation
  - Aneurysm
  - Organ rupture
  - Bowel perforation
  - Other genetic or acquired connective tissue disorders

**MANAGEMENT/TREATMENT**

Treat based on symptoms, and refer to specialists for help with associated complications/ issues and further education:

- Do low-impact exercise for 30 minutes 5 days a week.
- Hydrate and keep track of daily water intake.
- Manage and prevent injury over the long-term through strengthening, proprioceptive training, joint protection and endurance training (as guided by OT/PT). Consider nontraditional interventions like massage, yoga, meditation and acupuncture. Do not recommend joint/ spinal manipulation therapy due to joint laxity/instability.
- Recommend NSAIDS, heating pads and cold packs as needed for pain. Other medications can help for certain types of pain in specific situations which often need expert input to ensure the balance between benefit and risk is maintained.
- Treat associated symptoms, which may be more debilitating and have more impact on daily living than the joint symptoms—including anxiety, depression, dysautonomia, fatigue, functional GI disorders, headaches, postural orthostatic tachycardia and sleep disturbances.

**WHEN TO REFER**

1. Consider other disorders or conditions before referring for HSD or hEDS.
2. Refer based upon the patient’s age or the presence or absence of Red Flags. See algorithm on reverse side for more specific information.

If you would like additional copies of this tool, or would like more information, please contact the Physician Outreach and Engagement team at Cincinnati Children’s.
Hypermobility Spectrum Disorders
including Hypermobile Ehlers-Danlos Syndrome

<table>
<thead>
<tr>
<th>Patient Presents</th>
<th>Standard Workup</th>
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<tbody>
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<td>• Personal History • Family History • Physical Exam</td>
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</table>

**HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS**

See page 1 for list of HPE Red Flags

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**Refer accordingly:**

**General Genetics**
- Classic or other type of EDS, except vascular EDS
- Unusual skin fragility (consider other types of EDS)
- Spasticity
- Low muscle tone or common neuromuscular manifestations due to a known condition—confirmed or suspected, such as chromosomal disorders or muscular dystrophy
- First degree relative with:
  - Bowel perforation
  - Low muscle tone
  - Common neuromuscular conditions

**Cardiovascular Genetics**
- Aortic root dilation
- Ectopia lentis
- Other genetic connective tissue disorders, e.g., Marfan, Loey’s-Dietz, vascular EDS
- Personal history of bowel perforation or organ rupture
- First degree relative history of thoracic aortic aneurysm/dissection

**Rheumatology**
- Rheumatologic symptoms, e.g., prolonged morning stiffness, limping, asymmetric joint pains, daily persistent joint swelling
- Personal history of bowel perforation or organ rupture
- First degree relative with:
  - Other acquired connective tissue disorders, e.g., lupus, mixed connective tissue disorders

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**Skeletal Dysplasia Clinic**
- Skeletal dysplasia (e.g., osteogenesis imperfecta)

**Refer accordingly:**

**OT/PT**
- Joint hypermobility with or without joint pain or instability in any age
- State in referral: Evaluate and treat for hypermobility, joint protection, core strengthening and orthotics if needed

**Hypermobility Clinic**
- Symptomatic hypermobility in patients who have entered puberty or older
- Patients with hypermobility without red flags can also be managed by PCP (management guidelines at www.ehlers-danlos.com)

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**For children who have not turned 18 years of age or reached skeletal maturity, consider diagnosis based on the Diagnostic Criteria for Paediatric Joint Hypermobility found at www.ehlers-danlos.com**

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<tr>
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<tbody>
<tr>
<td>Must be minimum of 6</td>
<td>Must be minimum of 3</td>
<td>Must be minimum of 2</td>
<td>Any number causing distress or disability? Y/N</td>
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</tbody>
</table>

**For children who have reached skeletal maturity and adults ≥18, consider diagnosis based on the 2017 Diagnostic Criteria for Hypermobile Ehlers-Danlos Syndrome (hEDS) found at www.ehlers-danlos.com**

To make an hEDS diagnosis, all three criteria (1, 2, 3) must be met.

<table>
<thead>
<tr>
<th>1. Generalized joint hypermobility (as defined by the Beighton score)</th>
<th>2. Two or more of the following must be present</th>
<th>3. Meet ALL of these prerequisites</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. ≥6 for pre-pubertal children and adolescents</td>
<td>a. Minimum 5 of 12 systemic manifestations of a more generalized connective tissue disorder (<a href="http://www.ehlers-danlos.com/heds-diagnostic-checklist/">www.ehlers-danlos.com/heds-diagnostic-checklist/</a>)</td>
<td>a. Absence of unusual skin fragility</td>
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<tr>
<td>b. ≥5 for pubertal adolescents and young adults</td>
<td>b. One or more first-degree relatives independently meeting current diagnostic criteria for hEDS</td>
<td>b. Other heritable and acquired connective tissue disorders including autoimmune rheumatologic conditions have been considered</td>
</tr>
<tr>
<td></td>
<td>c. Musculoskeletal complications of joint instability</td>
<td>c. Alternative diagnoses that may include joint hypermobility by means of hypotonia/connective tissue laxity have been ruled out</td>
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For urgent issues, or to speak with the specialist on call 24/7, call the Physician Priority Link® at 1-888-987-7997.