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.

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If you have clinical questions about patients with HSD or hEDS, email EDSforDocs@cchmc.org.

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**FAST FACTS**

- **10%** of the population may be considered hypermobile but do not require referral or further workup if no red flags in history or PE

- **30 minutes, 5 days/week** of low-impact exercise such as swimming or riding a recumbent bike recommended for management of HSD or hEDS

- **no known genetic cause**, but it typically runs in families

- **pre-pubertal children with hypermobility and no red flags** can be evaluated by a pediatric Physical Therapist who can focus on joint protection, core strengthening and orthotics if needed

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Tool developed by Cincinnati Children’s physician-hospital organization (known as Tri-State Child Health Services, Inc.) and staff in the James M. Anderson Center for Health Systems Excellence. Developed using expert consensus and informed by Best Evidence Statements, Care Practice Guidelines, and other evidence-based documents as available. For Evidence-Based Care Guidelines and references, see www.cincinnatichildrens.org/evidence.
Hypermobility Spectrum Disorders
including Hypermobile Ehlers-Danlos Syndrome

HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS

**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—prolonged morning stiffness, limping, asymmetric joint pains, daily persistent joint swelling (>6 weeks duration)

**Family History**
- Self or first-degree relative:
  - Aortic disease/aortic root dilation
  - Aneurysm
  - Organ rupture
  - Bowel perforation
  - Other genetic connective tissue disorders (e.g., Marfan, Loey's-Dietz or other forms of EDS)

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
  - 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

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

- **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 or first-degree relative with other acquired connective tissue disorders, e.g., lupus, mixed connective tissue disorders

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
- Limited capacity as of October 2022
- Patients with hypermobility without red flags can also be managed by PCP (management guidelines at www.ehlers-danlos.com)

Consider diagnosis based on 2017 diagnostic criteria found at www.ehlers-danlos.com

1. Generalized joint hypermobility (as defined by the Beighton score)
   a. ≥6 for pre-pubertal children and adolescents
   b. ≥5 for pubertal adolescents and young adults

2. Two or more of the following must be present
   a. Minimum 5 of 12 systemic manifestations of a more generalized connective tissue disorder (www.ehlers-danlos.com/heds-diagnostic-checklist/)
   b. One or more first-degree relatives independently meeting current diagnostic criteria for hEDS
   c. Musculoskeletal complications of joint instability

3. Meet ALL of these prerequisites
   a. Absence of unusual skin fragility
   b. Other heritable and acquired connective tissue disorders including autoimmune rheumatologic conditions have been considered
   c. Alternative diagnoses that may include joint hypermobility by means of hypotonia/connective tissue laxity have been ruled out

For urgent issues, or to speak with the specialist on call 24/7, call the Physician Priority Link® at 1-888-987-7997.