

# Hypermobile Ehlers-Danlos Syndrome



## FAST FACTS

### 6 years

of age, minimum age for assessment of hEDS

### 30 minutes, 5 days/week

of low-impact exercise such as swimming or riding a recumbent bike recommended for management of hEDS

### 10%

of the population may be considered hypermobile

### No known genetic cause, but it typically runs in families

Hypermobile Ehlers-Danlos Syndrome (hEDS) is the most common form of a group of inherited connective tissue disorders. hEDS is generally characterized by joint hypermobility, skin hyperextensibility, and tissue fragility. The diagnosis of hEDS is clinical; there is no identifiable genetic cause yet, so no test is available. Hypermobility presents across a spectrum ranging from asymptomatic joint hypermobility to hEDS. There is no cure for hEDS.

## ASSESSMENT

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

## HPE RED FLAGS

Prior to referring for 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 orthotic tachycardia and sleep disturbances.

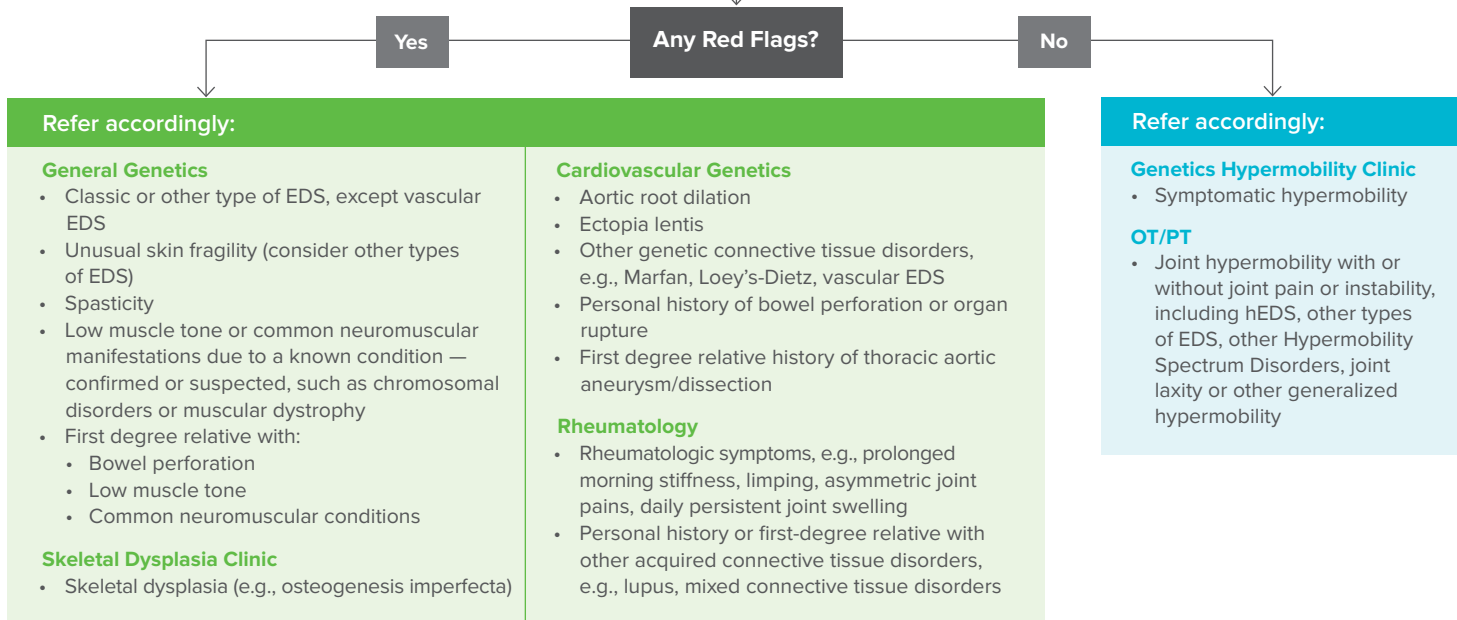
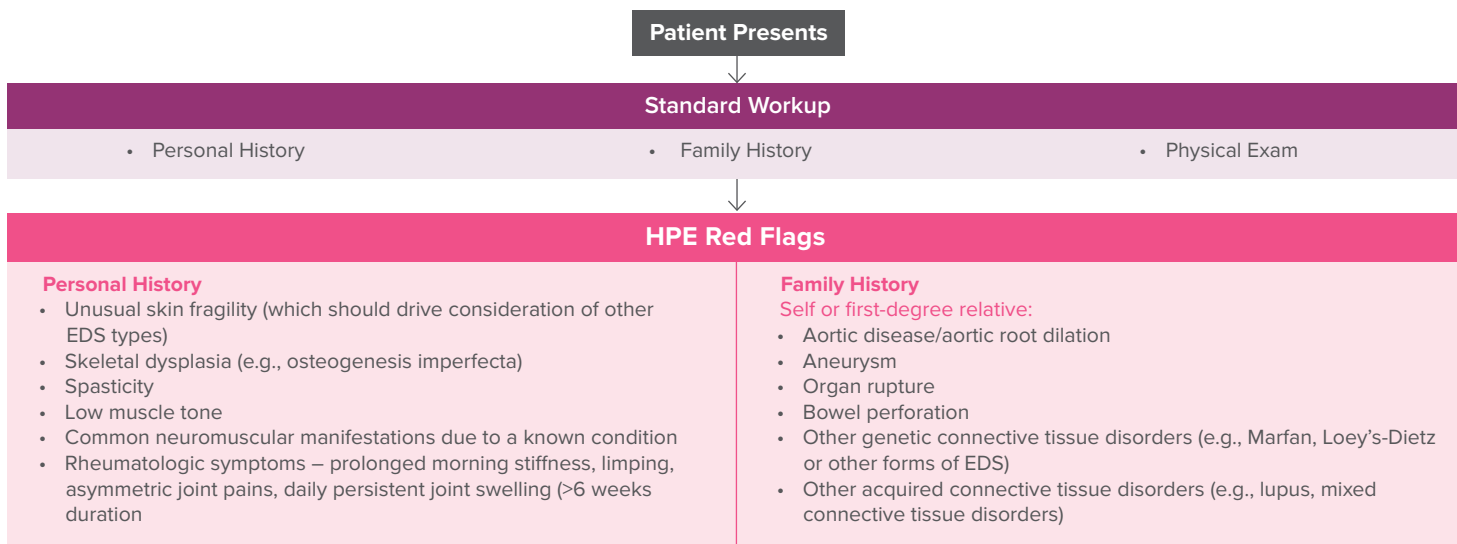
## WHEN TO REFER

1. Consider other disorders or conditions before referring for hEDS.
2. Refer based on 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.

If you have clinical questions about patients with Hypermobile Ehlers-Danlos Syndrome, email [EDSforDocs@cchmc.org](mailto:EDSforDocs@cchmc.org)

# Hypermobile Ehlers-Danlos Syndrome



Consider diagnosis based on 2017 diagnostic criteria found at [www.ehlers-danlos.com](http://www.ehlers-danlos.com).

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

<p><b>1. Generalized joint hypermobility (as defined by the Beighton score)</b></p> <p>a. ≥6 for pre-pubertal children and adolescents</p> <p>b. ≥5 for pubertal adolescents and young adults</p>	<p><b>2. Two or more of the following must be present</b></p> <p>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>)</p> <p>b. One or more first-degree relatives independently meeting current diagnostic criteria for hEDS</p> <p>c. Musculoskeletal complications of joint instability</p>	<p><b>3. Meet ALL of these prerequisites</b></p> <p>a. Absence of unusual skin fragility</p> <p>b. Other heritable and acquired connective tissue disorders including autoimmune rheumatologic conditions have been considered</p> <p>c. Alternative diagnoses that may include joint hypermobility by means of hypotonia/connective tissue laxity have been ruled out</p>
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For urgent issues, or to speak with the specialist on call 24/7, call the Physician Priority Link at 1-888-636-7997.