

## Diagnostic Criteria for Hypermobile Ehlers-Danlos Syndrome (hEDS)

This diagnostic checklist is for doctors across all disciplines to be able to diagnose EDS



Patient name:	DOB:	DOV:	Evaluator:	
The clinical diagnosis of hypermobile	EDS needs the simultaneou	s presence of all c	riteria, 1 <b>and</b> 2 <b>and</b> 3.	
CRITERION 1 – Generalized Join	t Hypermobility			
One of the following selected:  □ ≥6 pre-pubertal children and adole □ ≥5 pubertal men and woman to ag □ ≥4 men and women over the age of	je 50 Beighton Sco	re:/9		
If Beighton Score is one point below ag  ☐ Can you now (or could you ever) pl ☐ Can you now (or could you ever) be ☐ As a child, did you amuse your frie ☐ As a child or teenager, did your she ☐ Do you consider yourself "double j	ace your hands flat on the floor end your thumb to touch your fo nds by contorting your body into oulder or kneecap dislocate on m	without bending yo orearm? o strange shapes or	our knees? r could you do the splits?	terion:
CRITERION 2 — Two or more of t	he following features (A,	B, or C) must be	e present	
without a history of significant gai  Bilateral piezogenic papules of the Recurrent or multiple abdominal h  Atrophic scarring involving at least	in or loss of body fat or weight heel hernia(s) two sites and without the formatorolapse in children, men or nulli w palate or more of the following: on both sides, (ii) positive thumler greater based on strict echocal	tion of truly papyrad parous women with b sign (Steinberg sig		in classical EDS
Feature A total:/12  Feature B  Positive family history; one or more	re first-degree relatives indepen	dently meeting the	current criteria for hEDS	
Feature C (must have at least one)  ☐ Musculoskeletal pain in two or mo ☐ Chronic, widespread pain for ≥3 mo ☐ Recurrent joint dislocations or fran	onths			

## CRITERION 3 - All of the following prerequisites MUST be met

- 1. Absence of unusual skin fragility, which should prompt consideration of other types of EDS
- 2. Exclusion of other heritable and acquired connective tissue disorders, including autoimmune rheumatologic conditions. In patients with an acquired CTD (e.g. Lupus, Rheumatoid Arthritis, etc.), additional diagnosis of hEDS requires meeting both Features A and B of Criterion 2. Feature C of Criterion 2 (chronic pain and/or instability) cannot be counted toward a diagnosis of hEDS in this situation.
- 3. Exclusion of alternative diagnoses that may also include joint hypermobility by means of hypotonia and/or connective tissue laxity. Alternative diagnoses and diagnostic categories include, but are not limited to, neuromuscular disorders (e.g. Bethlem myopathy), other hereditary disorders of the connective tissue (e.g. other types of EDS, Loeys-Dietz syndrome, Marfan syndrome), and skeletal dysplasias (e.g. osteogenesis imperfecta). Exclusion of these considerations may be based upon history, physical examination, and/or molecular genetic testing, as indicated.

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