Hypermobility 101

What is hypermobility?
Joint hypermobility refers to increased active or passive movement of a joint beyond its normal range.

Can you have hypermobility without having Hypermobility Spectrum Disorder?
Yes, the two can be independent from each other (depending on symptoms).

Why is there pain with hypermobility?
Current theories emphasize the importance of localized biomechanical overloading and chronic soft tissue injury due to joint laxity and instability. Repetitive microtrauma may lead to altered kinematics, which, in turn, cause overload on other joints and further soft tissue injury manifesting as arthralgias and diffuse musculoskeletal pain

What is the difference between joint hypermobility and joint laxity?
Laxity indicates instability, whereas hypermobility denotes an increase in the joint's range of motion beyond normal

Do you have Generalized Joint Laxity?

- Can place hands flat on the floor without bending the knees
- Can passively dorsiflex the fifth metacarpophalangeal joint to 90° or greater
- Can oppose the thumb to volar aspect of forearm
- Can hyperextend the elbow 10° or more
- Can hyperextend the knee 10° or more

1 *point for first statemenmt and 1 point per statement per hand for the rest*

Beighton score ___/9 —> Does your score meet the criteria below for your age?

≥ 6 for prepubertal children and adolescents
≥ 5 for pubertal men and woman to age 50
≥ 4 for men and women over age 50

If Beighton score is 1 point below age-specific cutoff, *yes* answer to 2 of the following questions will meet criterion:

1. Can you now (or 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 teen did your shoulder or kneecap dislocate more than once?
5. Do you consider yourself "double jointed"

Do you meet the hypermobile Ehlers-Danlos Syndrome criteria?

Do you have 2 or more of features A, B, C, and ALL from Criteria F?

Feature A (5 must be present)

- Unusually soft or velvety skin
- Mild skin hyperextensibility
- Unexplained striae distensae or rubae at the back, groins, thighs, breasts, and/or abdomen in adolescence, men or prepubertal women without a significant gain or loss of body fat or weight
- Bilateral piezogenic papules of the heel
- Recurrent or multiple abdominal hernia(s)
- Atrophic scarring involving 2+ sites and without the formation of truly papyraceous and/or hemosideric scars as seen in classical EDS· Pelvic floor, rectal, and/or uterine prolapse in children, men, or nulliparous women w/out a history of morbid obesity or other known predisposing condition
- Dental crowding and high or narrow palate
- Arachnodactyly, as defined in one or more of the following: (1) positive wrist sign (Walker sign)
- on both sides, (2) positive thumb sign (Steinberg sign) on both sides
- Arm span-to-height ratio >/= 1.05
- Mitral valve prolapse mild or greater based on strict echocardiographic criteria
- Aortic root dilation with Z-score > 12

Feature B: Positive family history; 1 or more first-degree relatives independently meeting the current criteria for hEDS

Feature C (must have at least 1)

- Musculoskeletal pain in 2 or more limbs, recurring daily for at least 3 months
- Chronic, widespread pain for >/= 3 months
- Recurrent joint dislocations or frank joint instability, in the absence of trauma

Criteria F

- Absence of unusual skin fragility, which should prompt consideration of other types of EDS
- Exclusion of other heritable and acquired connective tissue disorders (CTDs), including autoimmune rheumatologic conditions. In patients with an acquired CTD (eg, lupus, rheumatoid arthritis, etc), additional diagnosis of hEDS requires meeting both features A and B (chronic and/or instability) cannot be counted toward a diagnosis of hEDS in this situation.
- 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 disorder (eg, Bethlem myopathy), other hereditary disorders of the connective tissue (eg, other types of EDS, Loeys-Dietz syndrome, Marfan syndrome), and skeletal dysplasias (eg, osteogenesis imperfecta). Exclusion of these considerations may be based upon history, physical examination, and/or molecular genetics testing, as indicated.

If joint hypermobility is found, but not the above criteria for hEDS, refer to specialist if concern for other conditions, otherwise diagnose with one of the four subtypes of Hypermobility Spectrum Disorder
(as long as there is one secondary musculoskeletal manifestations)

- General
- Peripheral for hands and feet only
- Localized for 1 joint or joint group
- Historial for past joint hypermobility that has decreased with age