Clarkson University Performing Arts Injury Prevention

# Hypermobility Syndrome

Joint Hypermobility Syndrome (JHS) and Hypermobility Syndrome (HMS) are old names for a condition now called hypermobile Ehlers-Danlos Syndrome, or hEDS. hEDS is a condition in which people have pain as a result of joints that are too flexible, as well as other symptoms. Being hypermobile can be an advantage in some occupations, such as dance and music. However, people with hEDS are vulnerable to developing many symptoms related to joints and other body systems.

***Diagnosis***. Although hEDS is an inherited connective tissue disorder, there is currently no imaging or lab test to diagnose it. Instead, we use a set of criteria that have to be met. To make things more confusing, the diagnostic criteria are changing in 2017. JHS used to be diagnosed using the 'Brighton Criteria' and EDS-hypermobile was diagnosed using the Villafranche Criteria; you will still hear about these criteria until the new ones gain traction.

All the criteria require that multiple joints be hypermobile. The set of joints tested is the Beighton (BAY-ton) Score, which assigns 1 point for each of 9 joints that are tested as shown in the figure, here. A score of 5 or more is required (people who have lost flexibility can count joints that used to be hypermobile).

**Figure: the Beighton Score tests**

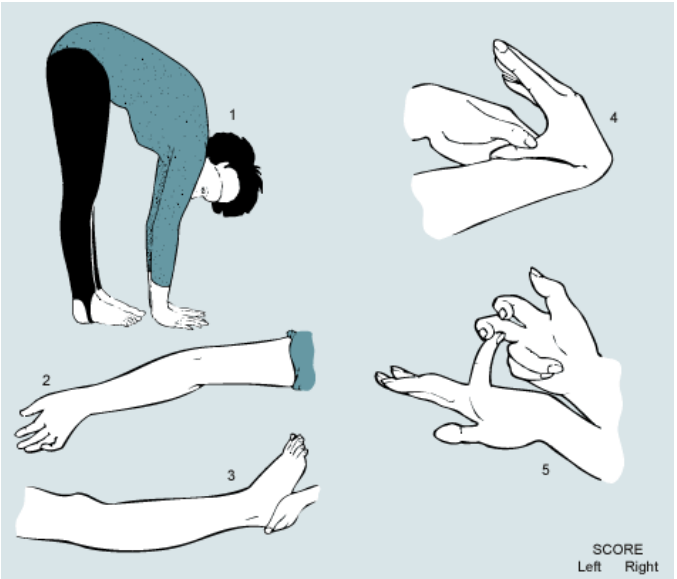


**Figure 1. Beighton's modification of the Carter and Wilkinson scoring system. Give youself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points.**

(figure from http://www.arc.org.uk/arthinfo/patpubs/6019/6019.asp)

We currently use the Brighton Criteria to decide whether a person has JHS. The Brighton Criteria have a number of major and minor criteria, as shown below. Note that new diagnostic criteria will be published in 2017, but those criteria are rather complicated.

*The following is a simplified version of the new criteria*

For a diagnosis of hEDS, you need 1, 2 & 3

1. Beighton Score of ≥5/9 (now or in the past)
2. 2 of 3 secondary characteristics*
3. No other condition to explain the findings

2. *Secondary characteristics (need 2 of 3 categories)
   - A. ≥ 5 of the following:
     - Unusually soft, velvety skin
     - Stretchy skin (>1.5 cm on forearm)
     - Unexplained stretch marks
     - Recurrent hernias
     - Thin papery scars at ≥ 2 locations
     - Pelvic, rectal or uterine prolapse
     - Arachnodactyly (very long fingers)
     - Very long arms (1.05x your height)
     - Mitral valve prolapse
     - Aortic root dilation
   - B. 1st degree relative diagnosed with hEDS
   - C. Any one of the following:
     - Pain in ≥ 2 limbs, daily ≥3 months
     - Chronic widespread pain ≥3 months
     - Recurrent non-traumatic dislocations

hEDS is considered an 'autosomal dominant' condition – meaning that it is inherited; it does not appear to have a single genetic abnormality, but is due to a variety of factors. hEDS is believed to be present in 1-3% of people, but it is much more common among women and certain ethnic groups (Arabic and Spanish heritage). Dancers and musicians also tend to have high levels of hEDS as increased flexibility is an advantage in these arts.

Although joint flexibility is often the most obvious problem in JHS, the abnormality actually affects any tissue that contains type I collagen. As a result, hEDS affects most body systems and can result in a wide range of symptoms, some of which can be quite surprising for a condition known primarily for causing joint flexibility. Signs and symptoms tend to change over the life span, and each person has a different combination.

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Some type of pain is typical of symptomatic hEDS. Common complaints include the following:

- Bone and joint problems:
  - Frequent sprains, subluxations and dislocations
  - Chronic joint pain, including low back pain and temporomandibular joint disorders
  - Osteoarthritis
  - Scoliosis
  - Decreased bone density, increased fracture rate
- Soft tissue problems:
  - Chronic soft tissue 'rheumatism' (tendinitis, bursitis, synovitis, tenosynovitis, fasciitis, or tendon ruptures)
  - Frequent muscle strains
  - Trigger points and chronic muscle pain
- Neurological problems:
  - Motor delay (in children)
  - Clumsiness, frequent falls, trips, or bumping into things
  - Nerve compression disorders (e.g., carpal tunnel), tingling and prickling sensations
  - Joint position sense and motor control deficits
  - Dizziness
  - Restless leg syndrome
  - Raynaud syndrome (very poor circulation into hands)
  - Fibromyalgia
  - Headaches, migraines
  - Poor response to local anesthetics, such as Novocain
- Psychological changes
  - Insomnia, unrefreshing sleep
  - Chronic fatigue
  - Memory or concentration problems
  - Anxiety and panic disorder
  - Depression
- Gastrointestinal problems:
  - Irritable bowel syndrome, constipation or diarrhea, bloating, abdominal pain
  - Gastroesophogeal reflux, chronic gastritis, heartburn
  - Nausea
  - Hernias (umbilical, inguinal, crural)
- Cardiovascular problems
  - Easy bruising
  - Postural Orthostatic Tachycardia Syndrome (POTS) (heart racing when suddenly standing up)
  - Racing heartbeat and palpitations
  - Varicose veins
  - Mitral valve prolapse
- Dermatologic problems
  - Hyperextensible skin
  - Slow or poor healing or scarring
- Urogenital problems
  - Urinary incontinence
  - Prolapsed bladder or uterus
  - Dysmenorrhea, endometriosis, pelvic pain

***Managing hEDS***. This is a chronic condition that cannot be 'cured' but it can often be managed to minimize pain and discomfort. The first step is to learn about the condition to better understand your symptoms. You can also learn how to protect your joints through better posture and body mechanics. Sometimes devices such as jar openers or ergonomic aids (e.g., modifications to musical instruments) can decrease stress on vulnerable joints, especially in the hands. Splints and braces can help stabilize fingers and wrists in musicians. Orthotics or taping can be helpful for dancers. When too much strenuous activity causes pain, pacing with appropriate rest breaks can help. Good posture and body mechanics are essential: because the joints are not stable, poor posture places excessive stress on both joints and muscles.

Appropriate exercise is important, as it keeps muscles strong and maintains coordination to protect joints. Exercises should be done using good body mechanics and a stable core. Stabilization, balance and joint control exercises (for proprioception, or the ability of joints to know where they are in space) can also help.

Joint instability makes some muscles work too hard to provide stability; these muscles go into spasm and develop trigger points, which can refer pain to other areas of the body. You may need to first retrain and strengthen muscles that are supposed to provide stability before you stretch or massage tight muscles. Whenever stretching muscles, use good body mechanics so you do not stretch nearby joints, as the body tends to stretch where it is already most flexible.

Standard medications are often not helpful for hEDS pain. The joint may hurt because the mechanoreceptors (nerve endings sensitive to mechanical stress) have been stimulated. Anti-inflammatory medications might not help if there is not inflammation.

If you think you may have h-EDS, discuss this with your doctor or your physical therapist. h-EDS is not well recognized or understood by many health care providers. That's why the logo is a zebra!

For more information about h-EDS

- **The Hypermobility Syndrome Association** at: www.hypermobility.org/.
- For musicians: http://www.foundations-for-excellence.org/file_storage/infosheet83_1.pdf
- For dancers: http://www.foundations-for-excellence.org/file_storage/infosheet74_1.pdf
- McCormack M. Teaching the Hypermobile Dancer. IADMS Bulletin for Teachers. 2010;2(1):5-8. Available at https://c.ymcdn.com/sites/www.iadms.org/resource/resmgr/Public/Bull_2-1_pp5-8_McCormack.pdf summarized at:
- http://www.4dancers.org/2014/07/the-hypermobile-dancer/