

JOINT HYPERMOBILITY

Generalized joint laxity (loose joints throughout the body) is fairly common, present in between 4% and 13% of the population. This hypermobility can occur in the absence of a systemic disease that affects connective tissues, like Ehlers-Danlos syndrome or Marfan syndrome.

While joint hypermobility is often labeled "benign," it can lead to and be associated with many health issues, including: joint pain, dislocations, spinal issues, muscle tightness and spasms, thin or stretchy skin, bruising, fatigue, and dizziness. And it's important to know that a broad range of conditions — like anxiety, fibromyalgia, chronic fatigue syndrome, and digestive issues — can occur along with joint hypermobility. This often leads to misdiagnosis, so it's important for people to do their own research and be their own health advocate.

How doctors diagnose joint hypermobility

Doctors may begin the process of diagnosing joint hypermobility by asking the following questions:

- Can you now, or could you ever, place your hands flat on the floor without bending your knees?
- Can you now, or could you ever, bend your thumb to touch your forearm?
- As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- As a child or teenager, did your shoulder or kneecap dislocate on more than one occasion?
- Do you consider yourself double-jointed?

If the answers to these questions indicate possible joint hypermobility, the next step will be to assess Beighton score. (Note: the Brighton criteria are used to determine the Beighton score; don't be confused by the similar spelling!)

Beighton score is between 0 and 9 depending on the ability to do the following things. Typically, a Beighton score of 4 or more indicates generalized joint laxity.

- Bend the pinky finger backward at the knuckle beyond 90 degrees (1 point each pinky)
- Flex the wrist forward and push the thumb back to touch the forearm (1 point each side)
- Extend the elbow more than 10 degrees beyond a normal straight position (1 point each elbow)

- Extend the knees more than 10 degrees beyond a normal straight position (1 point each knee)
- Bend forward and place the palms of the hands flat on the floor without bending the knees (1 point)

Benign Joint Hypermobility Syndrome (BJHS) and Ehlers-Danlos Syndrome (EDS)

Benign joint hypermobility syndrome (BJHS) is the presence of musculoskeletal complaints in hypermobile individuals who do not have a systemic rheumatologic disease. BJHS is thought to be the result of an abnormality in collagen, and mutations in the fibrillin gene have also been identified. The syndrome is often inherited, as first-degree relatives with the disorder can be identified in around 50% of cases.

The most common symptoms felt in people with BJHS are joint hypermobility and joint pain (arthralgia). When joints are excessively loose, it can cause abnormal wear and tear on joint surfaces, leading to pain and degeneration of joints over time. Some research suggests that joint proprioception (internal sense of joint position and movement) in people with BJHS can be impaired, and insufficient or incorrect sensory feedback can lead to dysfunctional movement patterns, poor posture, and pain.

When joints are very loose, people must rely on their muscles for stability in posture and movement. So, it is common for people with joint hypermobility to develop chronic muscle tension and experience muscle spasms.

Ehlers-Danlos syndrome (EDS) is a group of connective tissue disorders that can be inherited. EDS is currently classified into 13 different subtypes. The most common symptoms of EDS include hypermobile joints, thin or hyperelastic (abnormally stretchy) skin, and skin that bruises or scars easily.

One of the 13 subtypes is hypermobile EDS (hEDS). Benign hypermobility joint syndrome is thought to be a mild version of hEDS; some researchers suggest that BJHS is on a continuum with hEDS.

Since EDS is a problem with the structure of connective tissue, various symptoms can occur throughout the body — from skin, muscles, tendons, and ligaments to blood vessels, organs, gums, and eyes. And while joint hypermobility can lead to increased muscle tension because of the need for stability, some types of EDS involve muscle hypotonia (abnormally loose muscles). EDS can cause serious, even life-threatening health conditions including cardiovascular problems, organ rupture, and prolapse.

Both BJHS and hEDS are associated with a broad range of stress-related, psychological, behavioral, neurological, somatic, and gastrointestinal conditions, which you can see in the tables below. The occurrence of these conditions along with BJHS and hEDS may be caused by many different factors, including altered body perception, differences in emotion processing regions of the brain, and autonomic nervous system dysfunction.

When any of the conditions below are present, patients may be misdiagnosed or their underlying joint hypermobility may go undetected. Due to the broad range of symptoms and conditions related to joint hypermobility, some patients may be seen as being hypochondriacs.

Musculoskeletal and extra-articular features of JHS/hEDS	
Category	Features
Musculoskeletal	Joint: Joint laxity, arthralgia/myalgia, dislocation/subluxation, osteoarthritis, chondromalacia patellae, temporomandibular joint dysfunction, pain
	Soft tissue: Ligament/muscle/meniscus tear, epicondylitis, bursitis, tendinitis, capsulitis, Baker cysts
	Spine: Disc prolapse, loose back syndrome, spondylolysis, spinal abnormalities, spinal stenosis, scoliosis
Extra-articular	Neurologic: Dysautonomia, headache, chronic regional pain syndrome, carpal tunnel syndrome, developmental coordination disorder, fixed dystonia
	Gastrointestinal: Visceroptosis, irritable bowel syndrome, gastroesophageal reflux, hiatus hernia, chronic constipation, rectal evacuatory dysfunction, functional gastrointestinal disorder, Crohn's disease, oropharyngeal dysphagia
	Mucosa: Blue sclera, xerostomia, xerophthalmia, vaginal dryness, agenesis/ absence of the lingual frenulum, mucosal fragility (with subsequent spontaneous bleeding)
	Urologic: Urinary stress incontinence
	Gynecologic: Pelvic organ prolapse, irregular menses, meno/metrorrhagias, dysmenorrhea
	Psychiatric: Anxiety, depression, eating disorders, psychological distress
	Skin: Skin hyperextensibility, hypertrophic scarring, skin fragility, striae, easy bruising (capillary fragility), atopy
	Cardiovascular: Mitral valve prolapse, postural tachycardia syndrome, Chiari malformation, aortic valve regurgitation
	Others: Fibromyalgia, chronic fatigue syndrome, somatosensory amplification, increased interoception and exteroception, decreased proprioception
JHS/hEDS: joint hypermobility syndrome/Ehlers-Danlos type 3-hypermobile type	

Source: References 2,5,6

Images reprinted from <u>"Anxiety and joint hypermobility: An unexpected association" by</u> <u>Andrea Bulbena-Cabré, MD, PhD, MSc(Res) and Antonio Bulbena, MD, MSc(Cantab), PhD.</u> <u>Current Psychiatry. 2018 April;17(4):15-21</u> Figure 1

The Neuroconnective phenotype: 5 Dimensions



somatic symptoms include dysautonomia, astnenic somatotype, blue sciera, "easy bruising," eczemas, dyskinesia, dislocations, prolapses, and hypertrophic scars. Somatic illnesses include irritable bowel syndrome, dysfunctional esophagus, chemical sensitivities, dizziness, fatigue, fibromyalgia, "dynias," hypothyroidism, asthma, migraines, temporomandibular dysfunction, and food intolerance. Psychopathology includes increased interoception, exteroception, decreased proprioception, anticipatory anxiety, phobias, mood symptoms, eating disorders, neurotic personality traits, neurodevelopmental disorders, high sensitivity to loss, and high positive confrontation. Behavioral dimensions are patterns of defense mechanisms that often are identifiable at the extreme of a continuous axis (ie, fight/flight, avoidance/dependency). Somatosensory symptoms include increased olfactory sensitivity, eye-contact difficulty, selective photophobia, dyspnea, dysphagia, choking, palpitations, joint pain, and enhanced sensitivity to weather and chemicals. Joint hypermobility can be present in other conditions, including but not limited to:

- **Marfan syndrome:** a connective tissue disorder resulting from a mutation in FBN1, one of the genes that makes fibrillin
- Osteogenesis imperfecta: a group of genetic disorders that mainly affect the bones
- **Down syndrome:** a genetic disorder typically associated with a characteristic facial appearance and developmental and cognitive delays
- **Homocystinuria:** an inherited disorder in which the body is unable to process certain building blocks of proteins properly
- **Hyperlysinemia:** an inherited condition caused by a shortage of the enzyme that breaks down lysine, a building block of most proteins

Recommended treatment for Benign Joint Hypermobility Syndrome, and how Clinical Somatics can help

Since BJHS is nonprogressive and noninflammatory, treatment is generally supportive in nature. People with BJHS can often effectively manage their condition by modifying their activities, protecting their joints, reducing excess muscle tension, and learning how to improve their body mechanics.

Modification of activities: Overtraining and excessive joint movement are known to induce symptoms like joint pain in people with BJHS. People with BJHS should notice what activities increase symptoms, and reduce or avoid them. Types of exercise that involve intense stretching should be avoided, and the focus of workouts should be on increasing strength and stability.

Joint protection: People with BJHS are advised to not sit cross-legged with their knees bent, and to stop doing unusual joint movements to entertain their friends. They should also stand with their knees slightly bent if they have a tendency to hyperextend their knees. Bracing, splinting, or taping can provide beneficial joint support.

Reducing excess muscle tension while not stretching connective tissues: People with joint hypermobility can develop chronic muscle tension and experience muscle spasms because their muscles have to work extra hard to stabilize their joints. So while it is important to avoid building up excess muscle tension, it is also critical for people with BJHS to avoid static stretching, which stretches tendons and ligaments. Clinical Somatics exercises are an ideal and highly effective way for people with BJHS to release tight muscles without stretching their connective tissues.

Improve body mechanics: Exercises that focus on proprioception (internal sense of body position) allow people with BJHS to retrain their posture and movement, and improve their ability to instinctively avoid positions and movements that could be painful or damaging.

The slow, conscious nature of all Clinical Somatics exercises, along with the specific proprioceptive exercises that are taught, make Clinical Somatics an ideal therapeutic modality for people with BJHS.