Joint Hypermobility: Normal Variation or Cause for Concern?

By Deborah Quanbeck, M.D., Kristine Greer, D.P.T., and Katie Wilkins, D.P.T.

Joint hypermobility is a condition in which a person’s joints can be flexed beyond the normal range of motion. It affects approximately 10 to 15 percent of children. The diagnosis often appears straightforward—excessive laxity in finger (Fig. 1), elbow, hip and knee joints—and often it is that simple. If there are no other associated symptoms, joint hypermobility is usually considered benign.

Although joint hypermobility alone may not be cause for concern, the condition can be accompanied by joint pain and more frequent injuries (e.g., dislocations, sprains). It also can contribute to arthritis. In some cases, the symptoms signal a more serious condition, such as Ehlers-Danlos syndrome.

When examining a child or adolescent who is unusually flexible or more inclined to have sprains than most children, joint hypermobility may be the cause, and further evaluation is recommended.

About Joint Hypermobility

Joint hypermobility varies with age, gender (it is more common in females than males) and race. Often, it is familial. The condition is much more common in infants and young children than in adults (people tend to become less flexible as they grow older), and it occurs in a high percentage of Asian children. Randomly ascertained data on school-age children found that 50 percent of Chinese-Asian subjects had joint hypermobility as opposed to 7 percent of English-Caucasians. The average age of onset of symptoms is 6 years, but the average age at diagnosis is 9 to 10 years, indicating a two to three delay in diagnosis.

Fig. 1: A classic example of hyperjoint hypermobility.

KEY INSIGHTS

- Typically, clinicians discover joint hypermobility when a child is being evaluated for injuries, such as joint pain or fatigue, flat feet or bunions, and hip dislocations or subluxations.
- The Brights scale for assessing joint hypermobility is considered the gold standard for diagnosis, because it is quick, it is easy to use, and it has high intrarater reliability.
- Nonmusculoskeletal signs and symptoms, such as skin hyperelasticity, excessive bleeding or bruising, prominent veins, or heart murmurs, may point to more serious conditions such as Ehlers-Danlos syndrome.
- Treatment for joint hypermobility depends on the extent of the condition. Physical therapy may suffice for some patients, while others with more involved conditions should be referred to a pediatric orthopaedic surgeon or pediatric rehabilitation specialist.

Gillette’s Maple Grove Clinic Expands Services

Our Maple Grove Clinic has expanded our offering of pediatric specialty services. Patients living in the northwestern metropolitan area now have convenient access to these specialties:

Orthopaedics – Deborah Quanbeck, M.D., Angela Drummond, PMP

Pediatric rehabilitation medicine – Supreet Wills, M.D.

Plastic surgery – Paul Kim, M.D.

Conorrosis care – Martha McGreavy, PNP

Neurotrauma care – Leslie Larson, PNP, Amanda Sasse, FNP

Sleep medicine – Laurel Wells, M.D.

Additionally, the Maple Grove Clinic provides comprehensive rehabilitation and assistive technology services.

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About Deborah Quanbeck

Deborah Quanbeck, M.D., is a board-certified pediatric orthopaedic surgeon. She has extensive experience in treating children with acute pediatric orthopaedic conditions and a wide variety of musculoskeletal disorders.

She received her medical degree from the University of Minnesota Medical School. Following her orthopaedic surgery residency at the University of Minnesota, she completed a fellowship in pediatric orthopaedic surgery at Gillette Children’s Specialty Healthcare and Shriners Hospitals for Children. She is a member of the Pediatric Orthopaedic Society of North America and the American Academy of Orthopaedic Surgeons. She has published journal articles and made numerous professional presentations.
Diagnosing Joint Hypermobility

Typically, clinicians discover joint hypermobility when a child is evaluated for something else, such as: Joint pain and/or fatigue Deformities such as flat feet, bunions (See Fig 2.) clubfoot, hip dislocation or subluxation, and development of the hip (DDH) as many as half of children with DDH have signs of hypermobility Joint effusions Injuries such as recurrent sprains or dislocations Developmental delay or disabilities Gast abnormalities

At Gillette Children’s Specialty Healthcare, we use the Brighton scale to screen for joint hypermobility. The scale assesses only a few joints and is based on four passive maneuvers and one active maneuver. Instead of focusing on the degree of hypermobility at a particular joint, the tool helps clinicians establish how widespread the hypermobility is in that individual. A score of five or more defines generalized joint hypermobility. Not only is the tool quick and easy to use, but it also provides an objective measure based on a single assessment, and it has high intra-rater reliability. As a result, the Brighton scale is considered the gold standard for joint hypermobility assessment.

Once the extent of joint hypermobility is established, clinicians should assess nonsymmetrical skeletal factors, including:

- Skin hyperelasticity, which results from defective collagen molecules in the skin
- Dourgy or fragile skin, which tears easily or shows excessive scarring
- Excessive swelling or bruising
- Physical education difficulties
- Headaches
- Gingival recession
- Prominent veins, hemarthrosis, or easy-ovar venous varices
- Blue sclera
- Heart murmurs
- Family history

Evidence of the nonsymmetrical signs and symptoms mentioned at left is seen in those with more than six points to more serious conditions such as Ehlers-Danlos syndrome, osteogenesis imperfecta, Marfan syndrome and Larsen syndrome.

About Ehlers-Danlos Syndrome

Joint hypermobility is one of the hallmarks of Ehlers-Danlos syndrome (EDS), an inherited connective tissue disorder. Hypermobile skin and fragile tissues are also associated with EDS. According to the Villefranche syndrome, the syndrome is classified into six main types: classic, hypermobile, vascular, kyphoscoliosis, arthralgias and dermatosparaxis.

• Doughy or fragile skin, which tears easily or shows scale, provides helps on maneuvers

• Deformities such as flat feet, bunions (See Fig. 2.), clubfeet, excessive the only dislocation is the DDH individual.

• According to Brito, 2005: "Patients might have postural orthostatic tachycardia syndrome, orthostatic hypotension or orthostatic intolerance.

• Orthopedic concern and musculoskeletal injuries – Patients might experience: Flex - too plantus Ankles - subtle sprains Knees - Patellofemoral pain and patellar instability (subluxation/ dislocation)

• Hips - Possible hip subluxation (tars to have actual hip dislocation); frequent hip subluxations can result in lateral tears

• Shoulders - Multidirectional shoulder instability, shoulder impingement

• Spine - Spondylolisthesis, spondylitic degenerative disc disease, thinning of disc, stenosis and herniated discs

• Delays in gross motor skills

Depending on a patient’s needs, our physical therapists will develop a plan that incorporates the following principles and includes some or all of these treatments.

Strengthening Exercises

Patients who have joint hypermobility inherently have low muscular stiffness and joint instability. Our physical therapists focus on strengthening proximal muscle groups (spinal stabilizers and trunk/core muscles) to help improve patient’s overall function. Additional core stabilization exercises also help children learn to keep their joints in the neutral and protected range.

Proprioception and Balance Training

Closed chain exercises enhance proprioceptive feedback and assist with control of the joint. Open chain multi-directional exercises may be added in later stages of rehabilitation, if the joints are stable. Static and dynamic balance training help improve joint stability during functional and recreational activities.

Physical Therapy Can Address Joint Hypermobility Symptoms

Joint hypermobility syndrome may have these clinical implications:

- Pain - Patients might experience intermittent or activity-related pain, especially among adolescents who are involved in sports and other activities.
- Increased fatigue
- Fibromyalgia and complex regional pain syndrome

Patients with hypermobile joints may report pain. However, we focus on strengthening exercises to improve joint stability, which will eventually relieve patients’ symptoms.

Footwear and Bracing

Excessive ankle joint laxity and pes planus are more common in patients who have joint hypermobility. We may recommend orthoses, footbeds, ankle-stabilizing braces and patellar stabilizing braces.

Gross Motor Skills

Children who have joint hypermobility may have difficulties with coordination or gross motor skills, and physical therapists will evaluate them for delays in gross motor development.

Lifelong Physical Activity

Maintaining an active lifestyle with strengthening exercises can help provide stability to a joint and reduce painful symptoms. Physical therapists can provide instruction on joint protection principles that can help minimize joint overuse during physical activity.
Diagnosing Joint Hypermobility

Typically, clinicians discover joint hypermobility when a child is evaluated for something else, such as:

- Joint pain and/or fatigue
- Deformities such as flat feet, bunions (See Fig. 2.), clubfoot, hip dislocation or subluxation, and developmental dysplasia of the hip (DDH) as many as half of children with DDH have signs of hypermobility
- Joint effusions
- Injuries such as recurrent sprains or dislocations
- Developmental delay
- Hypertension
- Gait abnormalities

Fig 5: - Bunions and flat feet in a nine-year-old boy

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Not only is the tool quick and easy to use, but it also provides an objective measure based on a nine-point scale, and it has high intra-rater reliability. As a result, the Brighton scale is considered the gold standard for joint hypermobility assessment.1

Once the extent of joint hypermobility is established, clinicians should assess nonmusculoskeletal factors, including:

- Skin hyperelasticity, which results from defective collagen molecules in the skin
- Dourghy or fragile skin, which tears easily or shows excessive scarring
- Excessive blushing or bruising
- Physical education difficulties
- Fainting
- Gingival recession
- Prominent veins, hemorrhoids, or early-onset varicose veins
- Blue sclera
- Heart murmurs
- Family history

Evidence of the nonmusculoskeletal signs and symptoms mentioned in the list is so extensive that it points to more serious conditions such as Ehlers-Danlos syndrome, osteogenesis imperfecta, Marfan syndrome and Larsen syndrome.

About Ehlers-Danlos Syndrome

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Kypheoscoliosis, arthrogryposis and dermatosparaxis are rare types of EDS; classic, hyperelastic and vascularity EDS are considered to be the major types. While classic and hyperelastic EDS primarily affect the skin and musculoskeletal system, vascularity EDS is of particular concern because of the potential for arterial, bowel and uterine rupture. Consequently, any patient who has joint hypermobility requires a more thorough assessment to establish or rule out EDS.

Managing Joint Hypermobility

When a serious underlying genetic diagnosis such as Ehlers-Danlos syndrome is obtained, the first step is to refer the patient for genetic testing, and potentially for assessment of bleeding disorders, minimal coagulates or urticarial. For concerns about chewing habits, refer the patient to a pediatric rheumatologist.

For less involved cases, in which symptoms are primarily musculoskeletal, refer the patient to a pediatric orthopedic surgeon, a pediatric rehabilitation medicine specialist or a physical therapist—depending on the extent of involvement.

At Gillette, we take an interdisciplinary approach to care and do so on the appropriate specialists to address each patient’s chief complaints. For example, a patient who has hip dysplasia will be assessed and may require orthopedic surgery. However, another less involved patient may be referred for physical therapy to address pain, strengthening proprioception, balance and delays in gross motor skills.

Conclusion

Patients who present with symptoms of joint hypermobility should have a thorough assessment to determine the extent of the condition and establish whether joint hypermobility is benign or symptomatic of a serious disorder such as EDS.


Physical Therapy Can Address Joint Hypermobility Symptoms

Joint hypermobility syndrome may have these clinical implications:

Pain - Patients might experience intermittent or activity-related pain, especially among adolescents who are involved in sports and other activities.

Increased fatigue

Fibromyalgia and complex regional pain syndrome

Balance - Patients who have excessive joint hypermobility often have difficulty with balance and may have increased awareness of joint disorganization.

Abnormal articulation responses - Patients might have postural orthostatic tachycardia syndrome, orthostatic hypotension or orthostatic intolerance.

Orthopedic concerns and musculoskeletal injuries - Patients might experience:

- Foot - Plantar- 

- Addles - Juxta- 

- Knees - Patellar hyperextension (subluxation/ dislocation)

- Hips - Possible hip subluxation (fears to have actual hip dislocation); frequent hip subluxations can result in labral tears.

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Delays in gross motor skills

Depending on a patient’s needs, our physical therapists will develop a plan that incorporates the following principles and includes some or all of these treatments.

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Proprioception and Balance Training

Closed chain exercises enhance proprioceptive feedback and assist with control of the joint. Open chain multi-directional exercises may be added in later stages of rehabilitation, if the joints are stable. Static and dynamic balance training helps improve joint stability during functional and recreational activities.
Doughy or fragile skin, which tears easily or shows hypermobility in that joint, helps clinicians assess the degree of hypermobility at a particular joint, the tool helps clinicians establish how widespread the hypermobility is in that individual. A score of five or more defines generalised joint hypermobility.

Not only is the tool quick and easy to use, but it also provides an objective measure based on a nine-point scale, and it has high intra-rater reliability. As a result, the Beighton scale is considered the gold standard for joint hypermobility assessment.

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- Gingival recession
- Prominent veins, hemangiomas, or early onsite varicose veins
- Blue sclerae
- Heart murmurs
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EVIDENCE OF THE NONMUSCULOSKELETAL SIGNS AND SYMPTOMS MENTIONED AT LEFT IN YOUNG CHILDREN IS OF LESS SIGNIFICANCE THAN IN ADULTS.

Kypelos / arthrochalasis and dermatoplasia are rare types of EDS; classic hypermobile and vascular EDS are considered to be the major types. While classical and hypermobile EDS primarily affect the skin and musculoskeletal system, vascular EDS is of particular concern because of the potential for arterial, bowel, and uterine rupture. Consequently, any patient who has joint hypermobility requires a more thorough assessment to establish or rule out EDS.

Managing Joint Hypermobility
When a serious underlying genetic diagnosis such as Ehlers-Danlos syndrome is suspected or is certain, the patient is referred for genetic testing, and potentially for assessment of bleeding disorders, minimal Ehlers-Danlos or articular subtype. For concerns about atraumatic events, refer the patient to a pediatric rheumatologist.

For less involved cases, in which symptoms are musculoskeletal, refer the patient to a pediatric orthopedic surgeon, a pediatric rehabilitation medicine specialist or a physical therapist—depending on the extent of impact. At Gillette, we take an interdisciplinary approach to care and draw on the appropriate specialists to address each patient's chief complaints. For example, a patient who has hip dysplasia will be assessed and may require orthopedic surgery. However, another less involved patient may be referred for physical therapy to address pain, strengthening preceptiveness, balance and delays in gross motor skills.

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Patients who present with symptoms of joint hypermobility should have a thorough assessment to determine the extent of the condition and establish whether joint hypermobility is benign or symptomatic of a serious disease such as EDS.

7. screening tool: "Joint hypermobility syndrome: a 2008 update on the criteria for diagnosis and management.4"

Hypermobility is one of the hallmarks of Ehlers-Danlos syndrome (EDS), an inherited connective tissue disorder. Hypermobility and fragile tissues are also associated with EDS. According to the Villevalde snoology, the syndrome is classified into six main types: classic, hypermobile, vascular, kyphoscoliosis, arthrochalasis, and dermatoplasia.

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Although joint hypermobility alone may not be cause for concern, the condition can be accompanied by joint pain and more frequent injuries (e.g. dislocations, sprains). It also can contribute to arthritis. In some cases, the symptoms signal a more serious condition, such as Ehlers-Danlos syndrome.

When examining a child or adolescent who is unusually flexible or more inclined to have sprains than most children, joint hypermobility may be the cause, and further evaluation is recommended.

About Joint Hypermobility

Joint hypermobility varies with age, gender (it is more common in females than males) and race. Often, it is familial. The condition is much more common in infants and young children than in adults (people tend to become less flexible as they grow older), and it occurs in a high percentage of Asian children. Randomly ascertained data on school-age children found that 50 percent of Chinese-Asian subjects had joint hypermobility as opposed to 7 percent of English-Caucasians.1 The average age of onset of symptoms is 6-12 years, but the average age at diagnosis is 9-10 years, indicating a two-to-three-year delay in diagnosis.2

Katie Wilkins, D.P.T.

Katie Wilkins earned a doctorate in physical therapy from the University of Minnesota. She gained her first year of experience in sports medicine and orthopedics at the Sports Medicine and Wellness Center in River Falls, Wis. She works at Gillette Children’s Specialty Healthcare’s Minnesota Clinic.

Kristine Greer, D.P.T.

Kristine Greer earned a doctorate in physical therapy from the University of Minnesota. She began working with adult and pediatric acute care patients at Mercy Hospital in Coon Rapids, Minn. Shortly thereafter, Kristine transitioned into full-time pediatrics at Gillette Children’s Specialty Healthcare. She has worked at Gillette’s Minnesota and Maple Grove clinics.
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**Fig. 1** A classic example of finger joint hypermobility.