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 flared beyond the normal range of motion. It affects approximately 10 to 15 percent of children. The diagnosis often appears straightforward—excessive laxity in fingers (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 can also 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. It is more common in young children and young and 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-aged children found that 50 percent of Chinese-Asian subjects had joint hypermobility as opposed to 7 percent of English-Caucasian subjects.1

*Fig. 1 - A classic example of flares joint hypermobility*

KEY INSIGHTS

- Typically, clinicians diagnose 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 Beighton scale for assessing joint hypermobility is considered the gold standard for diagnosis, because it is quick, is easy to use, and it has high interrater 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 orthopedic surgeon or pediatric rehabilitation specialist.
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 developmentally 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
-Difficulty with fine and gross motor skills.
-Gast abnormalities

At Gillette Children’s Specialty Healthcare, we use the Beighton 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 nine-point scale, and it has high intra-rater reliability. As a result, it provides an objective measure based on a nine-point scale, helping clinicians establish how widespread the hypermobility is in that individual. A score of five or more defines generalized joint hypermobility.

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
- Doulbery or fragile skin, which tears easily or shows excessive scarting
- Excessive blushing or bruising
- Physical education difficulties
- Hemmas
- Gingival recession
- Prominent veins, hemangiomas, or early onset varicose veins
- Blue sclera
- Heart murmurs
- Family history

Evidence of the nonmusculoskeletal signs and symptoms mentioned in left to right order points to more serious conditions such as Ehlers-Danlos syndrome, osteogenesis imperfecta, Marfan syndrome and Larsen syndrome.

About Ehlers-Danlos Syndrome

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

Kypelosynthesis, arthrochalsia and dermatosparaxis are rare types of EDS: classic, hypermobile and vascular EDS are considered to be the major types. While classic and hypermobile EDS primarily affect the skin and musculoskeletal system, vascular EDS is of particular concern because of the potential for arterial, bowel, or 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 identified, the physician will refer the patient for genetic testing, and potentially for assessment of bleeding disorders, mineral imbalance or arrhythmia. For concerns about rheumatologic diseases, 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 impairment. 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 preconception, 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 disease such as EDS.

References

9. 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 decreased awareness of a joint’s position during movement (impaired proprioception).
- Abnormal autonomic responses: Patients might have postural orthostatic tachycardia syndrome, orthostatic hypotension or orthostatic intolerance.
- Orthopedic concerns and musculoskeletal injuries: Patients might experience:
  - Foot - Pes planus
  - Ankles - Achilles tendinitis
  - Knees - Patellofemoral pain and patellar instability
  - Hips - Possible hip subluxation (due to causes that have actual hip dislocation), frequent hip subluxations can result in labral tears
  - Shoulders - Multidirectional shoulder instability, shoulder impingement
  - Spines - Spondylolisthesis, spondylolysis, degenerative disc disease, thinning of disc space 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 muscle strength and joint stability. Physical therapists’ physical therapies can provide focus on strengthening proximal muscle groups (proprio stabilizers and trunk/core muscles) to help improve function. In addition, 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 helps in joint stability during functional and recreational activities.
### Beighton Scale

The Beighton scale is considered the gold standard for joint hypermobility assessment. It involves at least two 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.

<table>
<thead>
<tr>
<th>Joint</th>
<th>1 point for each elbow</th>
<th>1 point for each hand</th>
<th>1 point for each knee</th>
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<tbody>
<tr>
<td>Palms of the hands rest flat on the floor</td>
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<tr>
<td>Forward flexion of the trunk with the knees fully extended so that the palms of the hands rest flat on the floor</td>
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<tr>
<td>Hyperextension of the elbows beyond 10 degrees</td>
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<tr>
<td>Hyperextension of the knees beyond 10 degrees</td>
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<tr>
<td>Passive dorsiflexion of the metacarpophalangeal joint beyond 90 degrees</td>
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<td>Passive apposition of the thumb to the flat aspect of the other hand</td>
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### Managing Joint Hypermobility

When a serious underlying genetic diagnosis such as Ehlers-Danlos syndrome is a possibility, the Beighton scale is considered the gold standard for joint hypermobility assessment. It is in that individual. A score of five or more defines generalized joint hypermobility.

### Joint Hypermobility Syndrome: Clinical Manifestations

- **Hypermobile joints**, which are more common in patients who have joint hypermobility. We may recommend orthoses, footwear, ankle stabilizing braces and patellar stabilizing braces.

### Managing Joint Hypermobility

- Patients who present with symptoms of joint hypermobility should have an 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.

### Conclusion

Patients who present with symptoms of joint hypermobility should have an 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.

### Evidence of the nonmusculoskeletal signs and symptoms monitored in left to see if it is part of a larger picture:

- Skin hyperelasticity, which results from defective collagen metabolism.
- Dullness or flabby skin, which tears easily or shows excessive scarring.
- Excessive blistering or blushing.
- Physical education difficulties.
- Excessive bleeding or bruising.
- Doughy or fragile skin, which tears easily or shows excessive scarring.
- Skin hyperelasticity, which results from defective collagen metabolism.
- Joint pain and/or fatigue.
- Joint effusions.
- Injuries such as recurrent sprains or dislocations.
- Developmental delay.
- Rash.
- Gast abnormalities.

### 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 decreased awareness of a joint's position during movement (referred as proprioception).

**Abnormal autonomic responses**

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

**Orthopedic concern and musculoskeletal injuries**

- Patients might experience:
  - Foot - pes planus
  - Ankle - ankle sprains
  - Knees - Patellar dislocation and patellar instability (subluxation/dislacation)
  - Hips - Possible hip subluxations (ties to have actual hip dislocation); frequent hip subluxations can result in lateral laxity.
  - Shoulders - Multidirectional shoulder instability, shoulder impingement.
  - Spine - Spondylolysis, spondylolisthesis, degenerative disc disease, thinning of disc space, 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 (trunk stabilizers and trunk/core muscles) to help improve overall function. We may recommend 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 seen stable. Static and dynamic balance training help improve joint stability during functional and recreational activities.

### Postural Re-Education and Joint Protection Principles

We educate patients about neutral sitting postures, avoid knee and elbow hyperextension while standing or in quadruped positions, lifting mechanics, and the proper way to carry a backpack.

### Education on Stretching

We recommend that patients avoid stretching hypermobile joints to relieve pain. Instead, we focus on strengthening exercises to improve joint stability, which will eventually relieve patients' symptoms.

### Footwear and Bracing

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

### Gross Motor Skills

Children who have joint hypermobility may have difficulties with coordination or gross motor skills. Physical therapists can provide instruction on joint protection principles that can help minimize joint overuse during physical activity.

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- **Increased fatigue**
- **Fibromyalgia and complex regional pain syndrome**
- **Balance**
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Diagnosing Joint Hypermobility
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- 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
- Clumsiness
- Skin hyperelasticity, which results from defective collagen

The Beighton scale is considered the gold standard for joint hypermobility assessment.5 It is in that individual. A score of five or more defines generalised hypermobility.

Managing Joint Hypermobility
When a serious underlying genetic diagnosis such as Ehlers-Danlos syndrome is suspected, refer the patient for genetic testing, and potentially for assessment of bleeding disorders, mineral atrophy or sarcoma risk. For concerns about rheumatoid arthritis, refer the patient to a pediatric rheumatologist.

For less involved, reactive, symptoms as is primarily musculoskeletal, where the patient to a pediatric orthopedic surgeon, a pediatric rehabilitation medicine specialist or a physical therapist—depending on the extent of impairment.

At Gillette, we take an interdisciplinary approach to care and testing, and potentially 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.

1. Beighton Scale

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Evidence of the nonmusculoskeletal signs and symptoms mentioned is left to secondary care to point 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.

Hypertrophic skin and fragile tissue are also associated with EDS. According to the Villefranche nosology, the syndrome is classified into six main types: classic, hypermobile, vascular and dermatosparaxis.

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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.

- Strengthening Exercises

**Postural Re-Education and Joint Protection Principles**

**Education on Stretching**

**Lifelong Physical Activity**

**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 safe. Static and dynamic balance training helps improve joint stability during functional and recreational activities.**
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.1 The condition is more common in females than males and is more common in Asian children than in other children.2,3

Variation or Cause for Concern?

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.4

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 can also 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.5

The average age of onset of symptoms is 6 years, but the average age at diagnosis is 9 years, indicating a two to three year delay in diagnosis.6

Joint hypermobility may be caused by a variety of conditions, including Ehlers-Danlos syndrome. Common findings include joint laxity, skin hyperextensibility, and large joints. 

Typically, clinicians discover joint hypermobility when a child is being evaluated for other things, such as joint pain or fatigue, flat feet or bruising, and hip dislocations or subluxations.

The Beighton score, a method for assessing joint hypermobility, is considered the gold standard for diagnosis, because it is quick, is easy to use, and it has high intrarater reliability.

Nonmusculoskeletal signs and symptoms, such as skin hyperextensibility, 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 orthopedic surgeon or pediatric rehabilitation specialist.

KEY INSIGHTS

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NEWS & NOTES

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 specialists:

- Orthopedics – Deborah Quanbeck, M.D., Angela Drummond, PNP
- Pediatric rehabilitation medicine – Supreet Deshpande, M.D., Angela Sinner, D.O.
- Plastic surgery – Paul Kim, M.D.
- Craniofacial care – Martha McGreery, PNP
- Neurotrauma care – Leslie Larson, PNP, Amanda Slesser, PNP
- Sleep medicine – Laurel Wells, M.D.

Additionally, the Maple Grove Clinic provides comprehensive rehabilitation therapies and assistive technology services.
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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.1 The exact age of onset of symptoms is 6.2 years, but the average age at diagnosis is 9.0 years, indicating a two to three year delay in diagnosis.4

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REFERENCE