Research

Clinical Brief: Recognition of Benign Joint Hypermobility Syndrome (BJHS)

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Topics in Integrative Health Care 2011, Vol. 3(1) ID: 3.1005

Published on March 31, 2012 | Link to Document on the Web

Epidemiology

The first known reference to joint laxity which leads to joint hypermobility is attributed to Hippocrates who, in the 4th century BC, described the Scythians as being "so-loose-limbed that they were unable to draw a bow-string or hurl a javelin."¹ The prevalence of generalized joint hypermobility varies from 10 to 30 percent.²⁻⁵ This was illustrated in a study of 123 healthy medical students where 22 (18%) had at least one lax joint, and 14 others (11%) had three or more lax joints.⁶ Joint laxity is more common in the right limb, females, blacks, and in children from families with higher socioeconomic status.⁷⁻⁹

The term "hypermobility syndrome" was first used by Kirk, Ansell, and Bywaters in 1967 to describe a disorder in which musculoskeletal pain and generalized joint hypermobility occurred together.¹⁰ It has been used to distinguish this symptomatic, but not life threatening disorder from diseases such as Marfan syndrome and specific types of Ehlers-Danlos syndrome in which aortic or other arterial dissections and/or aneurysms develop.¹¹ The prevalence of Benign Joint Hypermobility Syndrome (BJHS) is less certain. One study of New Zealanders of indigenous or European ancestry found symptoms and joint hypermobility in 0.72 percent of the study sample.⁴ In one Canadian rheumatology clinic, hypermobility was noted in 50 of 378 (13%) consecutive patients.¹² Physicians may not be cognizant of the implications of a missed diagnosis and may not routinely recognize BJHS during the clinical examination.

Clinical Assessment

The signs and symptoms of BJHS are variable. Most commonly, the initial complaint in a hypermobile patient is joint pain, which may affect one or multiple joints and may be generalized or symmetric.¹² BJHS is diagnosed according to the Revised 1998 Brighton criteria, a major criterion of which is the presence of demonstrable joint hypermobility. (Figure 1)
Diagnosis is entirely clinical using the previously listed criteria and patient history. There are currently no known biological markers for BJHS, though it is possible to rule out other genetically influenced collagen disorders such as Marfan’s, Ehlers-Danlos, and osteogenesis imperfecta through laboratory testing if necessary for differential diagnosis.

**Figure 1. Brighton criteria for diagnosis of BJHS**

BJHS is diagnosed in the presence of two major criteria; one major criterion plus two minor criteria; or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first degree relative in family history. The syndrome is excluded by the presence of Marfan’s or Ehlers-Danlos syndromes (other than the hypermobility type of Ehlers-Danlos syndrome).

**Major Criteria**

- Beighton score of =4 (Figure 2)
- Arthralgia for longer than 3 months in 4 or more joints

**Minor Criteria**

- Beighton score of 1, 2, or 3 (Figure 2)
- Arthralgia (>3-month duration) in one to three joints or back pain (>3-month duration) or spondylosis, spondylolysis/spondylolisthesis.
- Dislocation or subluxation in more than one joint, or in one joint on more than one occasion.
- Three or more soft tissue lesions (eg, epicondylitis, tenosynovitis, bursitis)
- Marfanoid habitus (tall, slim, span greater than height, upper segment less than lower segment arachnodactyly)
- Skin striae, hyperextensibility, thin skin, or abnormal scarring
- Ocular signs: drooping eyelids, myopia, antimongoloid slant
- Varicose veins, hernia, or uterine or rectal prolapse
- Mitral valve prolapse

The Beighton hypermobility score is widely used in assessing for hypermobility of peripheral joints and the spine. One point is awarded for the ability to perform each of four maneuvers (bilaterally) and one point for having an unusually flexible spine (Figure 2) with a total of nine possible points, with a score of four or more, in adults indicating joint hypermobility. The Beighton score has excellent validity and high inter-observer reproducibility.
Figure 2. Nine-point Beighton score for joint hypermobility

One point is gained for each side of the body for the first four maneuvers listed below, such that the hypermobility score is a maximum of 9 if all are positive.

- Passive dorsiflexion of the fifth metacarpophalangeal joint to at least 90 degrees. (1 point for left; 1 point for right)
- Passive opposition of the thumb to the volar aspect of the ipsilateral forearm (1 point for left; 1 point for right)
- Active hyperextension of the elbow to at least 10 degrees (1 point for left; 1 point for right)
- Active hyperextension of the knee to at least 10 degrees (1 point for left; 1 point for right)
- Placement of hands flat on the floor without bending the knees (1 point)

Early screening and treatment among healthcare providers

The primary clinical manifestations of hypermobility and pain in multiple joints must raise a red flag for early screening procedures. Physicians who use manipulative techniques must recognize that joint hypermobility syndrome can be a contraindication to high-velocity, low amplitude spinal and joint manipulation. Overzealous physical manipulation may cause damage, such as: (a) precipitating dislocation of intervertebral or peripheral joints, (b) inflicting rupture on ligaments, joint capsules, muscles, or tendons, or (c) precipitating pathological fractures in fragile bone. The treatment of BJHS can include non-steroidal anti-inflammatory drugs for pain control, a strength training program to provide muscular stability and stabilization to the joint, focusing on improving proprioception to the joint with supportive splints or joint taping and finally, the use of mobilizing techniques to restore natural mobility to joints or spinal segments where these have been lost as a result of deconditioning and kinesiophobia. The incidence of BJHS can be misdiagnosed or under-diagnosed and physicians should be aware of this clinical presentation to enhance their diagnostic acumen and improve clinical outcomes.

References


