Hypermobility Syndrome
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Hypermobility Syndrome

Hypermobility syndrome was recognized as a distinct pathology by Kirk et al[1] in 1967. Since then, the syndrome has been identified by a variety of names: "hypermobility syndrome (HMS),"[2–9] "joint hypermobility syndrome,"[10–13] "hypermobile joint syndrome,"[14] and "benign hypermobile joint syndrome."[15,16] Other reports do not recognize this disorder as a syndrome, but refer to the manifestations of joint hyperlaxity, joint hypermobility, or articular hypermobility. In the *International Nosology of Heritable Disorders of Connective Tissue*, Beighton et al[17] identified this syndrome as "familial articular hypermobility syndrome." Beighton et al excluded genetic diseases that include joint hypermobility as an associated finding, such as Ehlers-Danlos syndrome, osteogenesis imperfecta, and Marfan syndrome.

Despite the proliferation of names, HMS has been given relatively little attention in the literature. Most reports are in the rheumatology literature, with virtually none in the orthopedic or physical therapy literature. This lack of reports may be due to several reasons. First, individuals with HMS are often seen by orthopedic physicians and physical therapists as a result of an acute or chronic disorder, which may be treated without the health care provider acknowledging the underlying HMS. Second, the diagnostic criteria for HMS are not well-defined and have not been consistent among research reports.[8,18–20] In particular, patients with HMS lack laboratory or radiological findings that could identify HMS, unlike many other rheumatologic or orthopedic conditions. The diagnosis, therefore, is frequently made through exclusion of other disorders. Third, individuals with HMS often do not have the decreased mobility seen with many chronic conditions,[4] nor do they always have the inflammation seen with many acute conditions. Finally, because HMS lacks a definitive pharmacological or surgical treatment, physicians may have perceived little benefit in its diagnosis.

Key Words: *Connective tissue disorders, Hypermobility, Joint instability, Physical therapy.*

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Table 1.
Possible Neuromusculoskeletal Signs for Individuals With Hypermobile Joint Syndrome

Acute or traumatic
Sprains[5]
Recurrent ankle sprains[3]
Meniscus tears[8]
Acute or recurrent dislocations/subluxations of the:
Shoulder[3]
Patella[2,3,58,59]
Metacarpophalangeal joint
Temporomandibular joint[14]
Traumatic arthritis
Bruising[10,52]
Fractures[5]
Chronic or nontraumatic
Soft tissue rheumatism[1,5,6,38,46,68]
Tendinitis
Epicondylitis
Rotator cuff syndrome
Synovitis
Juvenile episodic synovitis
Bursitis
Chondromalacia[23,56]
Back pain[6,23,31,32,56,84]
Scoliosis[3,6,8,a]
Fibromyalgia[6,21,46,51,68]
Temporomandibular joint dysfunction[14,60–62]
Nerve compression disorders
Carpal tunnel[6]
Tarsal tunnel[6]
Acroparesthesia
Thoracic outlet syndrome[46]
Raynaud syndrome[5,36]
Flat feet and sequellae[3,8,56]
Unspecified arthralgia or effusion of foot, ankle, knee, hip, back, neck, shoulder, elbow, wrist, or fingers[1–3,6,23,46,57]
Osteoarthritis[1,6,51–53]
Delayed motor development[43,44]
Congenital hip dislocation[19]

[a] Binns M. Joint laxity in idiopathic adolescent scoliosis. *J Bone Joint Surg Br.* 1988;70:420–422.

Patients with HMS often have complaints that are frequently diffuse, chronic, and inconsistent with observed pathology. These individuals may be improperly identified as having hypochondria, as malingering, or as having nonspecific chronic pain, without further investigation into the source of their complaints.[11,21] Individuals with HMS may not get a diagnosis, or they might be misdiagnosed.[22] Failure to recognize the underlying HMS may lead to unnecessary or inappropriate diagnostic studies, surgical procedures, and patient management,[23] especially for children.[24]

Reports of the prevalence of hypermobility syndrome should be viewed cautiously because of the variability in diagnostic criteria used.

Some reports of HMS describe it as "benign" when compared with the serious connective tissue diseases that have hypermobility as one of their signs.[15,16,25,26] Hypermobility syndrome has also been described as representing the upper end of the normal distribution for ligamentous laxity,[27,28] with no greater incidence of pain or injury.[29,30] Some authors[25,31–34] have even proposed that hypermobility may be an asset in certain sports or professions. The mobility present in people with HMS, however, is considered beyond the normal range by most researchers,[6] with repeated reports describing increased incidence of pain and associated disorders (Tab. 1).

A goal of this update is to increase awareness, understanding, and discussion of HMS through examination of the prevalence, diagnosis, clinical presentation, and pathophysiology. Although physical therapy for people with HMS has been recommended by many authors,[2,3,11,26,35] there are no published reports regarding the efficacy of physical therapy or any other treatment for individuals with HMS. Hopefully, increased recognition will lead to increased research about this disorder.

Prevalence

Reports of the prevalence of HMS must be viewed cautiously because of the variability in the diagnostic criteria used. Hypermobility syndrome has been reported in 0.6%[18] to 31.5%[36] of adults without joint pain, depending on age, ethnicity, and criteria for assessing hypermobility (Tab. 2).[37] This syndrome is more prevalent among females than among males.[15,18,36–41] Reports indicate that HMS may be from 1.1 times[42] to 5.5 times[18] more prevalent among females than among males. Hypermobility syndrome is also more prevalent among Asians than among Africans, and it is more prevalent among Africans than among Caucasians.[4,39] Children without symptoms of HMS tend to have rates of hypermobility that are higher than those of

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Table 2.
Prevalence of Hypermobility Syndrome Reported in the Literature for the General Population[a]

Male Subjects		Female Subjects		Total Subjects					
%	N	%	N	%	N	Criteria Used	Age (y)	Population	Reference
0.6	168	3.3	334	2.4	502	Beighton 6/9	20–82	Africans	Beighton et al[18]
1.0	104	2.9	104	1.9	208	Beighton 5/9	21–70	Caucasians	Wordsworth et al[39]
2.8	422	8.9	214	4.9	636	Modified Carter-Wilkinson[b] 2/3		US adults	Jessee et al[15]
6.2	145	7.1	140	6.7	285	Carter-Wilkinson 4/5	6–11	British school children	Carter and Wilkinson[19]
		8.0	50			Beighton 3/5	50+	Patients without arthritis	Scott et al[53]
6.7	134	18.3	126	12.3	260	Modified Beighton[c]	5–17	US school children	Gedalia et al[38]
6.0	150	21.9	114	12.9	264	Beighton 5/9	15.5 avg	US adolescent athletes	Decoster et al[41]
				16.2	606	Beighton 3/5	38.5 ± 11	Swedish factory workers	Larsson et al[45]
6.9	360	33.7	300	19.1	660	Beighton 3/5	14–68	US music students	Larsson et al[37]
23.6	1,187	31.5	587	29.8	1,774	Beighton (4–6)/9	20–24	Iraqi students	Al-Rawi et al[36]
				31.7	416	Carter-Wilkinson 5/9	5–17	Non-Caucasian Brazilian school children	Forleo et al[42]
33.7	445	38.4	560	36.3	1,005	Carter-Wilkinson 5/9	5–17	Brazilian school children	Forleo et al[42]
				39.6	589	Carter-Wilkinson 5/9	5–17	Caucasian Brazilian school children	Forleo et al[42]

[a] "General population" refers to samples not selected because of joint pain or other medical conditions.
[b] Excluding dorsiflexion and knee hyperextension.
[c] Criteria as in Beighton et al,[18] except for hyperextension of fingers to lie parallel to forearm (as in Carter and Wilkinson[19]) rather than hyperextension of fifth metacarpophalangeal joint to 90 degrees.

adults (between 6.7%[19] and 39.6%[42]), again depending on the population and criteria used (Tab. 2). Hypermobility appears to decrease with age.[18,43–45]

In children with fibromyalgia, the prevalence of hypermobility may be as high as 81.0% (Tab. 3).[21] Not all groups with joint pain described in Table 3, however, had high rates of HMS. For example, only 2.9% of children with juvenile rheumatoid arthritis (RA) had HMS versus 65.6% of those with juvenile episodic arthritis or arthralgia.[38] In a study of adults,[46] 32.0% of patients without hypermobility who were seen in a rheumatology clinic had inflammatory arthritis, compared to only 4.3% of patients with hypermobility.

Diagnosis

Carter and Wilkinson[19] proposed the first scale for rating hypermobility syndrome, in which one point was given if the patient could do each of the following indicators of HMS: passive apposition of the thumb to the forearm, passive hyperextension of the fingers and wrist so that the fingers lie parallel to the forearm, hyperextension of the elbow past 10 degrees, hyperextension of the knee past 10 degrees, and excessive dorsiflexion and eversion of the foot. They proposed that a score of 3/5 or higher indicated HMS.

Beighton et al[18] modified these criteria, providing the diagnostic criteria most commonly used today and still considered the yardstick for proposed scales.[4] The Beighton scale gives a patient one point for each of the following characteristics: passive extension of the fifth metacarpophalangeal (MCP) joint past 90 degrees, passive apposition of the thumb to the forearm, hyperextension of the elbow past 10 degrees, hyperextension of the knee past 10 degrees, and trunk flexion allowing the palms to be placed flat on the floor. Beighton et al scored each limb separately for the first 4 items, generating a possible score of 9. Subsequent researchers have sometimes combined right and left sides, generating a possible score of 5. There is no universal agreement on a threshold for HMS; some researchers use a Beighton scale score of 5/9, other researchers use a Beighton scale score of 6/9, and still other researchers use a modified Beighton score of 3/5 (Tab. 2).

Bulbena et al[8] compared the hypermobility scale proposed by Carter and Wilkinson,[19] the modified scale of Beighton et al,[18] and an 11-point scale proposed by Rotés.[20] Along with other criteria, Bulbena et al determined which criteria most clearly distinguished individuals with HMS from a control group of patients without HMS who were seen in a rheumatology clinic. Bulbena et al found each of the scores to be highly correlated to the other scores. By doing a cluster analysis, they were able to identify the 10 criteria most highly predictive of HMS. The frequency of positive criteria seen in patients identified as having HMS is shown in Table 4. Several,

Table 3.
Prevalence of Hypermobility Syndrome Reported in the Literature for Populations Reporting Joint or Muscle Pain

Male Subjects		Female Subjects		Total Subjects					
%	N	%	N	%	N	Criteria Used	Age (y)	Population	Reference
				2.9	34	Modified Beighton 3/5[b]	5–17	Israeli school children with juvenile rheumatoid arthritis	Gedalia et al[21]
				5.7	262[a]	Beighton 3/5	NA	Patients seen in pediatric arthritis clinic	Biro et al[2]
				8.7	1,311	Beighton 3/5	3–70	Patients seen in rheumatology and rehabilitation clinics	el-Shahaly and el-Sherif[6]
0.0	33	20.6	97	15.4	130	Beighton 5/9	18–83	Patients with rheumatologic disorders	Bridges et al[51]
		24.0	50			Beighton 3/5	50+	Patients with osteoarthritis	Scott et al[53]
				50.0	70	Beighton 5/9[c]	NA	Patients with temporomandibular joint disease	Buckingham et al[14]
56.8	37	68.7	67	64.4	104	Modified[d] Carter-Wilkinson 3/3	12–47	Patients with patellar dislocation	Grahame[4]
30.0	10	66.5	200	64.8	210	Beighton 3/5[e]	21–78	US patients with fibromyalgia	Goldman[68]
				65.6	32	Modified Beighton 3/5[b]	5–17	US school children with juvenile episodic arthritis/arthralgia[f]	Gedalia et al[38]
				81.0	21	Modified Beighton 3/5[b]	9–15	Israeli school children with fibromyalgia	Gedalia et al[21]

[a] 3/15 subjects were diagnosed with juvenile arthritis, leaving 12/262 or 4.6% with primary hypermobility syndrome.
[b] Criteria as in Beighton et al,[18] except for hyperextension of fingers to lie parallel to forearm (as in Carter and Wilkinson[19]) rather than hyperextension of fifth metacarpophalangeal joint to 90 degrees.
[c] Original report used Beighton 4/9 as the cutoff. To allow comparison with other reports using Beighton 5/9 criteria, percentages were recomputed using raw data reported.
[d] Including only tests of hyperextension of elbows and knees, apposition of thumb to forearm.
[e] Specified that hypermobility needed to be present in bilateral elbows or knees to score on either criterion.
[f] Excluded children with juvenile rheumatoid arthritis.

but not all, of the common findings are included in Carter and Wilkinson's criteria and in Beighton and colleagues' criteria. Bulbena et al also proposed that there be different cutoff criteria for women and men, as women tend to have more positive signs than men have. This discussion shows that there are neither agreed on criteria nor agreed on scores for the diagnosis of HMS.

Attempts to more precisely quantify hypermobility have been proposed, including controlled hyperextension of the fifth finger using a hyperextensometer[47] (a mechanical device that hyperextends the MCP joint using a controlled torque) and other measuring devices that use a controlled torque to apply passive overpressure.[28] Global measures, however, appear to have greater sensitivity for identifying people with HMS than do isolated hyperextensometric measures.[33] Symptoms do not appear to be directly correlated to the number of joints involved.[23] That is, individuals with marginal scores on these tests may have more symptoms than do individuals with high scores.

Laboratory tests may be done to rule out related but more serious disorders (eg, RA and other inflammatory polyarthritic conditions,[1] Ehler-Danlon and Marfan syndromes). The increased collagenase and protease breakdown of articular cartilage that is seen in people with RA is not seen in people with HMS. If the patient has Ehlers-Danlos or Marfan syndrome, both of which are hereditary connective tissue disorders with associated joint hypermobility,[39,48] the diagnosis of HMS is precluded.[5,26,49] Findings of hyperelastic skin, hernias, lenticular abnormalities,[6] and abnormal body proportions[1] are seen in patients with Ehlers-Danlos or Marfan syndrome but not in patients with HMS. Easy bruising and poor wound healing may be seen in patients with HMS as well as in patients with Ehlers-Danlos or Marfan syndrome. Osteogenesis imperfecta is another collagen disorder that may need to be ruled out, even though patients with this disorder often demonstrate joint hypermobility.[50] Systemic lupus erythematosus,[51] poliomyelitis, tabes dorsalis, myotonia congenita, and some neurological conditions[1] are also excluded.

Clinical Presentation

People with HMS may have complaints that have lasted from 15 days to 45 years (average time=6.5 years), and the onset of symptoms may occur at any age from 3 to 70 years.[6] These individuals may go to an orthopedic physician, rheumatologist, pediatrician, or physical therapist with any of a wide range of traumatic or nontraumatic pain complaints (Tab. 1). Frequently, these individuals report multiple complaints over a prolonged period. They typically lack the positive laboratory findings found in rheumatologic disorders and, in the absence of acute trauma, lack the radiologic changes, inflammation,

swelling, and decreased mobility typical of orthopedic pathology. Patients with HMS often have a poor response to oral analgesic and anti-inflammatory medications, unless they have inflammation due to recent trauma.[11]

People with HMS may have an increased incidence of nerve compression disorders, although data are scant. El-Shahaly and el-Sherif[6] reported acroparesthesia (excessive paresthesia in multiple limbs) in 57.9% of the 114 patients with HMS they tested. They found carpal tunnel syndrome in 31.6% of the patients and tarsal tunnel syndrome in 14.0% of the patients. They also reported a greater incidence of paresthesia among women than among men. Hudson et al,[46] however, reported no difference in nerve entrapment neuropathies between patients with HMS and a control group referred to a rheumatology clinic (data were not presented). In that study, which included patients with thoracic outlet syndrome and musculoskeletal complaints, the authors found that 54.3% of the patients with HMS had thoracic outlet symptoms (they did not compare patients with HMS and patients without HMS in their study).

Patients may also have sequellae of HMS.[8,49,52] Some authors[6,51] reported that up to 60% of individuals with HMS developed osteoarthritis (OA) as opposed to 30% of patients without HMS who were seen in a rheumatology clinic. Other researchers[53] found that 24% of patients with OA had HMS, as opposed to only 8% of patients seen for general medical conditions. An increased incidence of OA may be secondary to chronic or traumatic biomechanical abnormalities or proprioceptive deficits. Mobile joints such as the patellofemoral joint and the mid-cervical spine seem to be affected most.[54,55] Hypermobility syndrome was more common in patients with chondromalacia patellae than in matched controls.[56] Articular cartilage is made up of primarily type II cartilage, and it is suspected that HMS may include an abnormality of type II collagen.[54]

Although sprains, subluxations, and dislocations are more common in people with HMS,[3,57–59] the amount of tissue damage occurring with these acute injuries may actually be decreased due to the increased laxity of joint structures. For example, patients with HMS and acute patellar dislocation had an incidence of chondral injury and avulsion fracture of only 33%, whereas patients without hypermobility had an 80% incidence.[59]

Many researchers[60–62] have found a correlation between temporomandibular dysfunction (TMD) and HMS, with up to 54.3% of patients with TMD demonstrating HMS. One study[14] also showed that individuals with HMS had abnormal disk position and excessive anterior mandibular movement. Not all researchers found this association between generalized hypermobility and TMD.[63]

Table 4.
Ten Musculoskeletal Characteristics Most Common in Hypermobility Syndrome (HMS)[8]

Characteristic	Incidence in 114 Subjects With HMS
Excessive ankle dorsiflexion and foot eversion[a]	94%
Finger metacarpophalangeal joint extension past 90°[a,b]	93%
Thumb abduction to the forearm[a,b]	92%
Patellar hypermobility	89%
Excessive shoulder lateral rotation	84%
Excessive hip abduction	78%
Knee hyperextension past 10°[a,b]	77%
Elbow hyperextension past 10°[a,b]	75%
Ecchymosis	63%
First metatarsophalangeal joint extension past 90°	61%

[a] Those characteristics included in Carter and Wilkinson's diagnostic test[19] (except that Carter and Wilkinson tested fifth finger and wrist extension, so fingers are parallel to the forearm instead of metacarpophalangeal joint extension past 90°).
[b] Those characteristics included in Beighton and colleagues' diagnostic test,[18] with the addition of excessive trunk flexion.

Infants with HMS have higher incidences of motor delay, even in the absence of an identified neurological deficit. Among infants between 8 and 14 months of age, 30.2% of those with HMS had motor delay versus 10.9% of infants without HMS. Six months later, 75.9% of the infants with HMS no longer had hypermobility; 83.3% of these infants caught up in motor development, whereas only 54.5% of the infants who remained hypermobile caught up.[43] At 5 years of age, children who had hypermobility and motor delay at age 18 months were 3 times as likely as other children to have motor delay.[44] Other reports have shown no motor delay in elementary school children with HMS.[64]

People with HMS are more likely (69.3%) to have anxiety disorders than are comparison groups with rheumatological conditions (22.0%)[10] or groups with other chronic medical conditions (21.3%).[65] The incidence of anxiety disorders among individuals with HMS was 3 times greater when mitral valve prolapse (a common finding with HMS) was present.[10] Anxiety may also be due to the perception of joint instability and frequent pain and injury without understandable antecedent.

The high incidence of psychological disorders in people with HMS is similar to that seen with fibromyalgia.[66,67] Ninety percent of individuals with HMS and fibromyalgia reported sleep disturbances.[68] Because HMS and fibromyalgia appear to be related, the causes of psycho-

logical disorders in people with HMS may be similar to those proposed for fibromyalgia: abnormalities in serotonin metabolism, stage IV sleep patterns, and levels of substance P,[22,69] or perceptual hypervigilance.[70]

Other systemic findings are highly correlated with HMS. Mitral valve prolapse is 3 times more prevalent in patients with HMS than in other patients and may be present in up to one third of all individuals with HMS.[5,10,12,51,71] Uterine prolapse is also more common,[6,72,73] as are rectal prolapse[74] and abdominal hernias.[75] An increased incidence of varicose veins has been reported by some authors[6] as has increased bruising.[10,71] Increased skin elasticity and decreased skin thickness in patients with HMS has been documented by some authors[16] but not by others.[71] One surgeon reported wide, thin scar formation after surgery in patients with HMS.[58]

Some authors[12] have reported morphological differences in subjects with HMS: arm span greater than height, wide-set eyes, beaked nose, slim fingers. Studies, however, that specifically exclude individuals with Marfan or Ehlers-Danlos syndrome (as specified in the diagnosis[17,26]) typically do not show morphological differences.

Pathophysiology

Hypermobility syndrome appears to be inherited as a gender-influenced dominant trait.[11,13,17,19,34,76] It appears to be an abnormality of type I collagen,[11] although studies to identify a single gene abnormality have not been successful.[15] Other collagen diseases have also been related to multiple collagen or genetic abnormalities.[77]

Type I collagen is the most common collagen in the human body. With a high tensile strength, type I collagen is normally abundant in connective tissues such as tendon, ligament, joint capsule, skin, demineralized bone, and nerve receptors. Type II collagen is found primarily in hyaline cartilage. Type III collagen is found in the same tissues as type I collagen, but usually in lesser amounts. Thin and elastic compared with type I collagen, type III collagen is found in greater relative amounts in extensible connective tissues, such as the vascular system, skin, and lung.[78] In patients with HMS, the ratio of type III collagen to type III+type I collagen is increased.[11,12] Normally, this ratio is 18%:21%, whereas in patients with HMS, it is 28%:46%. Electron microscopy of skin biopsies showed that individuals with HMS had a decreased number of thick collagen fibers and an increased prevalence of fine disorganized fibers when compared with age-matched controls.[12] The abnormal ratio of type III to type I collagen is thought to cause the decreased tissue stiffness seen in patients with HMS. Decreased stiffness of joint structures produces the joint hypermobility most obvious in patients with HMS; decreased stiffness of other tissues may result in the prolapse seen in other organs. For example, aortic compliance is increased in patients with HMS.[12] Mitral valve prolapse is thus caused by decreased stiffness of the chordae tendineae that normally limit valve movement.

Nerve tissue also appears to be affected in patients with HMS. The increased incidence of acroparesthesia reported in individuals with HMS[6] may be due to abnormalities in the nerve tissue as well as surrounding connective tissues. Individuals with HMS are less accurate than individuals without HMS in reproducing a proximal interphalangeal joint angle.[7] Studies have shown that position sense at the knee is also decreased in patients with HMS. In particular, subjects with HMS did not have the ability to locate their joint at end-range extension found in subjects without symptoms of HMS.[9] Increased mobility and decreased joint position sense could make the joints of people with HMS more vulnerable to damage from what would be minor trauma in a person without symptoms of HMS.

Treatment of Patients With Hypermobility Syndrome

Education

Education is probably the most important treatment that physical therapists can provide to individuals with HMS. Because people with spinal hypermobility are reported to have a higher incidence of back pain in sedentary jobs,[32] it seems likely that education regarding ergonomics and body mechanics may decrease the incidence of back pain among people with HMS. As education about joint protection has been shown to increase function and decrease pain in subjects with RA, education regarding joint protection in HMS may similarly decrease pain, traumatic injury, and subsequent degenerative disease in vulnerable joints.[79] It seems appropriate to advise individuals with HMS in the selection of jobs, sports, or recreational activities that will not exacerbate their condition. In theory, patients with HMS may be able to continue certain activities at a decreased frequency or intensity or be able to modify biomechanical stresses through change in technique or use of protective splints for vulnerable joints. Splints, braces, and taping may also be used judiciously to protect other vulnerable joints.[2]

Helping patients with HMS understand their disorder may help them cope with the pain they experience. Goldman[68] found that for individuals with both fibromyalgia and HMS, the presence of HMS was associated with increased participation in a treatment regimen. He attributed increased participation to the improved understanding and acceptance patients had for their disorder.

Exercise

Although exercise will not increase stiffness of the lax ligaments seen in patients with HMS, strengthening and proprioceptive exercises are recommended for musculature surrounding affected joints.[2,3,9,35,80–83] Individuals with HMS and fibromyalgia who exercised reported greater improvement in symptoms than did those who did not exercise.[68] Indiscriminant exercise, however, could be harmful. For example, lightweight women rowers who had excessive spinal mobility were more likely to have back problems if they participated in a stretching program for their backs.[84] It appears reasonable, therefore, to advise individuals with HMS to use stretching exercises cautiously, distinguishing between stretching muscles and stretching joints, as the former may be beneficial but the latter may be harmful.

Medication

Chronic pain in people with HMS is not always associated with inflammation. Some authors[2,38] recommend the use of nonsteroidal anti-inflammatory medications, whereas other authors[11,80] report that the use of these drugs is neither practical nor effective.

Directions for Future Research

Many unanswered questions remain regarding HMS. Prospective studies of physical therapist patients are needed to determine the percentage of patients with HMS. Such studies could determine, for example, how many patients with HMS have musculoskeletal problems versus nerve compression disorders.

Although many authors recommend exercise for these patients, few have any data on which to base that recommendation. Moderate- and low-impact strengthening exercises, cardiovascular exercise for weight control, and stretching of muscles rather than joints seem theoretically sound recommendations, but the assumptions on which these recommendations are based must be tested. Given the predisposition of patients with HMS for cumulative trauma injuries, studies are needed to determine how much exercise is appropriate for these individuals. The use of orthotic devices, braces, and taping for patients with HMS also warrants further research.

References

1 Kirk JA, Ansell BM, Bywaters EGL. The hypermobility syndrome: musculoskeletal complaints associated with generalized joint hypermobility. *Ann Rheum Dis.* 1967;26:419–425.

2 Biro F, Gewanter HL, Baum J. The hypermobility syndrome. *Pediatrics.* 1983;72:701–706.

3 Finsterbush A, Pogrund H. The hypermobility syndrome: musculoskeletal complaints in 100 consecutive cases of generalized joint hypermobility. *Clin Orthop.* 1982;168:124–127.

4 Grahame R. "The hypermobility syndrome." *Ann Rheum Dis.* 1990; 49:190–200.

5 Grahame R, Edwards JC, Pitcher D, et al. A clinical and echocardiographic study of patients with the hypermobility syndrome. *Ann Rheum Dis.* 1981;40:541–546.

6 el-Shahaly HA, el-Sherif AK. Is the benign joint hypermobility syndrome benign? *Clin Rheumatol.* 1991;10:302–307.

7 Mallik AK, Ferrell WR, McDonald AG, Sturrock RD. Impaired proprioceptive acuity at the proximal interphalangeal joint in patients with the hypermobility syndrome. *Br J Rheumatol.* 1994;33:631–637.

8 Bulbena A, Duro JC, Porta M, et al. Clinical assessment of hypermobility of joints: assembling criteria. *J Rheumatol.* 1992;19:115–122.

9 Hall MG, Ferrell WR, Sturrock RD, et al. The effect of the hypermobility syndrome on knee joint proprioception. *Br J Rheumatol.* 1995;34: 121–125.

10 Bulbena A, Duro JC, Porta M, et al. Anxiety disorders in the joint hypermobility syndrome. *Psychiatry Res.* 1993;46:59–68.

11 Child AH. Joint hypermobility syndrome: inherited disorder of collagen synthesis. *J Rheumatol.* 1986;13:239–243.

12 Handler CE, Child A, Light ND, Dorrance DE. Mitral valve prolapse, aortic compliance, and skin collagen in joint hypermobility syndrome. *Br Heart J.* 1985;54:501–508.

13 Henney AM, Brotherton DH, Child AH, et al. Segregation analysis of collagen genes in two families with joint hypermobility syndrome. *Br J Rheumatol.* 1992;31:169–174.

14 Buckingham RB, Braun T, Harinstein DA, et al. Temporomandibular joint dysfunction syndrome: a close association with systemic joint laxity (the hypermobile joint syndrome). *Oral Surg Oral Med Oral Pathol.* 1991;72:514–519.

15 Jessee EF, Owen DS Jr, Sagar KB. The benign hypermobile joint syndrome. *Arthritis Rheum.* 1980;23:1053–1056.

16 Mishra MB, Ryan P, Atkinson P, et al. Extra-articular features of benign joint hypermobility syndrome. *Br J Rheumatol.* 1996;35:861–866.

17 Beighton P, de Paepe A, Danks D, et al. International Nosology of Heritable Disorders of Connective Tissue, Berlin, 1986. *Am J Med Genet.* 1988;29:581–594.

18 Beighton P, Solomon L, Soskolne CL. Articular mobility in an African population. *Ann Rheum Dis.* 1973;32:413–418.

19 Carter C, Wilkinson J. Persistent joint laxity and congenital dislocation of the hip. *J Bone Joint Surg Br.* 1964;46:40–45.

20 Rotés J. *Rheumatologia Clinica.* Barcelona, Spain: Espaxs; 1983.

21 Gedalia A, Press J, Klein M, Buskila D. Joint hypermobility and fibromyalgia in schoolchildren. *Ann Rheum Dis.* 1993;52:494–496.

22 Krsnich-Shriwise S. Fibromyalgia syndrome: an overview. *Phys Ther.* 1997;77:68–75.

23 Lewkonia RM, Ansell BM. Articular hypermobility simulating chroninc rheumatic disease. *Arch Dis Child.* 1983;58:988–992.

24 Southwood TR. Recent developments in the understanding of paediatric musculoskeletal pain syndromes. *Ann Rheum Dis.* 1993;52: 490–492.

25 Grahame R, Jenkins JM. Joint hypermobility: asset or liability? A study of joint mobility in ballet dancers. *Ann Rheum Dis.* 1972;31: 109–111.

26 Sheon RP, Kirsner AB, Farber SJ, Finkel RI. The hypermobility syndrome. *Postgrad Med.* 1982;71:199–209.

27 Wood PHN. Is hypermobility a discrete entity? *Proc R Soc Med.* 1971;64:690–692.

28 Silman AJ, Haskard D, Day S. Distribution of joint mobility in a normal population: results of the use of fixed torque measuring devices. *Ann Rheum Dis.* 1986;45:27–30.

29 Mikkelsson M, Salminen JJ, Kautiainen H. Joint hypermobility is not a contributing factor to musculoskeletal pain in pre-adolescents. *J Rheumatol.* 1996;23:1963–1967.

30 Grana WA, Moretz JA. Ligamentous laxity in secondary school athletes. *JAMA.* 1978;240:1975–1976.

31 Larsson LG, Baum J, Mudholkar GS, Kollia GD. Benefits and disadvantages of joint hypermobility among musicians. *N Engl J Med.* 1993;329:1079–1082.

32 Larsson LG, Mudholkar GS, Baum J, Srivastava DK. Benefits and liabilities of hypermobility in the back pain disorders of industrial workers. *J Intern Med.* 1995;238:461–467.

33 Brodie DA, Bird HA, Wright V. Joint laxity in selected athletic populations. *Med Sci Sports Exerc.* 1982;14:190–193.

34 Beighton P, Horan FT. Dominant inheritance in familial generalised articular hypermobility. *J Bone Joint Surg Br.* 1970;52:145–147.

35 Bird HA, Barton L. Joint hyperlaxity and its long-term effects on joints. *J R Soc Health.* 1993;113:327–329.

36 Al-Rawi ZS, Al-Aszawi AJ, Al-Chalabi T. Joint mobility among university students in Iraq. *Br J Rheumatol.* 1985;24:326–331.

37 Larsson LG, Baum J, Mudholkar GS. Hypermobility: features and differential incidence between the sexes. *Arthritis Rheum.* 1987;30: 1426–1430.

38 Gedalia A, Person DA, Brewer EJ Jr, Giannini EH. Hypermobility of the joints in juvenile episodic arthritis/arthralgia. *J Pediatr.* 1985;107: 873–876.

39 Wordsworth P, Ogilvie D, Smith R, Sykes B. Joint mobility with particular reference to racial variation and inherited connective tissue disorders. *Br J Rheumatol.* 1987;26:9–12.

40 Rikken-Bultman DG, Wellink L, van Dongen PW. Hypermobility in two Dutch school populations. *Eur J Obstet Gynecol Reprod Biol.* 1997;73: 189–192.

41 Decoster LC, Vailas JC, Lindsay RH, Williams GR. Prevalence and features of joint hypermobility among adolescent athletes. *Arch Pediatr Adolesc Med.* 1997;151:989–992.

42 Forleo LH, Hilario MO, Peixoto AL, et al. Articular hypermobility in school children in Sao Paulo, Brazil. *J Rheumatol.* 1993;20:916–917.

43 Jaffe M, Tirosh E, Cohen A, Taub Y. Joint mobility and motor development. *Arch Dis Child.* 1988;63:159–161.

44 Tirosh E, Jaffe M, Marmur R, et al. Prognosis of motor development and joint hypermobility. *Arch Dis Child.* 1991;66:931–933.

45 Larsson LG, Baum J, Mudholkar GS, Srivastava DK. Hypermobility: prevalence and features in a Swedish population. *Br J Rheumatol.* 1993;32:116–119.

46 Hudson N, Starr MR, Esdaile JM, Fitzcharles MA. Diagnostic associations with hypermobility in rheumatology patients. *Br J Rheumatol.* 1995;34:1157–1161.

47 Bird HA, Brodie DA, Wright V. Quantification of joint laxity. *Rheumatol Rehabil.* 1979;18:161–166.

48 Beighton P, Horan FT. Orthopaedic aspects of the Ehlers-Danlos syndrome. *J Bone Joint Surg Br.* 1969;51:444–453.

49 Lewkonia RM. The bioloby and clinical consequences of articular hypermobility. *J Rheumatol.* 1993;20:220–222.

50 McKusick VA. *Heritable Disorders of Connective Tissue.* 3rd ed. St Louis, Mo: CV Mosby Co; 1966:47.

51 Bridges AJ, Smith E, Reid J. Joint hypermobility in adults referred to rheumatology clinics. *Ann Rheum Dis.* 1992;51:793–796.

52 Bird HA, Tribe CR, Bacon PA. Joint hypermobility leading to osteoarthrosis and chondrocalcinosis. *Ann Rheum Dis.* 1978;37: 203–211.

53 Scott D, Bird H, Wright V. Joint laxity leading to osteoarthrosis. *Rheumatol Rehabil.* 1979;18:167–169.

54 Grahame R. Clinical conundrum. *Br J Rheumatol.* 1989;28:320.

55 Lewkonia RM. Does generalized articular hypermobility predispose to generalized osteoarthritis? *Clin Exp Rheumatol.* 1986;4:115–119.

56 Al-Rawi Z, Nessan AH. Joint hypermobility in patients with chondromalacia patellae. *Br J Rheumatol.* 1997;36:1324–1327.

57 Sutro CJ. Hypermobility of bones due to "overlengthened" capsular and ligamentous tissues. *Surgery.* 1947;21:67–76.

58 Runow A. The dislocating patella: etiology and prognosis in relation to generalized joint laxity and anatomy of the patellar articulation. *Acta Orthop Scand.* 1983;201(suppl):1–53.

59 Stanitski CL. Articular hypermobility and chondral injury in patients with acute patellar dislocation. *Am J Sports Med.* 1995;23:146–150.

60 Adair SM, Hecht C. Association of generalized joint hypermobility with history, signs, and symptoms of temporomandibular joint dysfunction in children. *Pediatr Dent.* 1993;15:323–326.

61 Westling L, Holm S, Wallentin I. Temporomandibular joint dysfunction: connective tissue variations in skin biopsy and mitral valve function. *Oral Surg Oral Med Oral Pathol.* 1992;74:709–718.

62 Westling L, Mattiasson A. General joint hypermobility and temporomandibular joint derangement in adolescents. *Ann Rheum Dis.* 1992; 51:87–90.

63 Dijkstra PU, de Bont LG, Stegenga B, Boering G. Temporomandibular joint osteoarthrosis and generalized joint hypermobility. *Cranio.* 1992;10:221–227.

64 Davidovitch M, Tirosh E, Tal Y. The relationship between joint hypermobility and neurodevelopmental attributes in elementary school children. *J Child Neurol.* 1994;9:417–419.

65 Wells KB, Golding JM, Burnam MA. Psychiatric disorder in a sample of the general population with and without chronic medical conditions. *Am J Psychiatry.* 1988;145:976–981.

66 Robbins JM, Kirmayer LJ, Kapusta MA. Illness worry and disability in fibromyalgia syndrome. *Int J Psychiatry Med.* 1990;20:49–63.

67 Krag NJ, Norregaard J, Larsen JK, Dunneskiold-Samsoe B. A blinded, controlled evaluation of anxiety and depressive symptoms in patients with fibromyalgia, as measured by standardized psychometric interview scales. *Acta Psychiatr Scand.* 1994;89:370–375.

68 Goldman JA. Hypermobility and deconditioning: important links to fibromyalgia/fibrositis. *South Med J.* 1991;84:1192–1196.

69 Smythe HA. Studies of sleep in fibromyalgia; techniques, clinical significance, and future directions. *Br J Rheumatol.* 1995:34:897–899.

70 McDermid AJ, Rollman GB, McCain GA. Generalized hypervigilance in fibromyalgia: evidence of perceptual amplification. *Pain.* 1996;66:133–144.

71 Pitcher D, Grahame R. Mitral valve prolapse and joint hypermobility: evidence for a systemic connective tissue abnormality? *Ann Rheum Dis.* 1982;41:352–354.

72 Al-Rawi ZS, Al-Rawi ZT. Joint hypermobility in women with genital prolapse. *Lancet.* 1982;I:1439–1441.

73 Norton PA, Baker JE, Sharp HC, Warenski JC. Genitourinary prolapse and joint hypermobility in women. *Obstet Gynecol.* 1995;85: 225–228.

74 Marshman D, Percy J, Fielding I, Pelbridge L. Rectal prolapse: relationship with joint mobility. *Aust N Z J Surg.* 1987;57:827–829.

75 Wynne-Davis R. Familial joint laxity. *Proc R Soc Med.* 1971;64:689–690.

76 Horton WA, Collins DL, DeSmet AA, et al. Familial joint instability syndrome. *Am J Med Genet.* 1980;6:221–228.

77 Kuivaniemi H, Tromp G, Prockop DJ. Mutations in collagen genes: causes of rare and some common diseases in humans. *FASEB J.* 1991;5:2052–2060.

78 Prockop DJ, Kivirikko KI. Collagens: molecular biology, diseases, and potentials for therapy. *Annu Rev Biochem.* 1995;64:403–434.

79 Nordenskiold U. Evaluation of assistive devices after a course in joint protection. *Int J Technol Assess Health Care.* 1994;10:293–304.

80 Gedalia A, Brewer EJ Jr. Joint hypermobility in pediatric practice: a review. *J Rheumatol.* 1993;20:371–374.

81 Barrack RL, Skinner HB, Cook SD. Proprioception of the knee joint: paradoxical effect of training. *Am J Phys Med.* 1984;63:175–181.

82 Perlau R, Frank C, Fick G. The effect of elastic bandages on human knee proprioception in the uninjured population. *Am J Sports Med.* 1995;23:251–255.

83 Barrett DS, Cobb AG, Bentley G. Joint proprioception in normal, osteoarthritic, and replaced knees. *J Bone Joint Surg Br.* 1991;73:53–56.

84 Howell DW. Musculoskeletal profile and incidence of musculoskeletal injuries in lightweight women rowers. *Am J Sports Med.* 1984;12: 278–282.

Hypermobility Syndrome
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