Case Report

Examination and Treatment of a Patient With Hypermobility Syndrome

Background and Purpose. The purpose of this case report is to present the examination, evaluation/diagnosis/prognosis, intervention, and outcome of a patient with hypermobility syndrome (HMS). Hypermobility syndrome has been widely recognized in the rheumatology literature, but it has seldom been discussed in the orthopedic literature and has only recently been described in the physical therapy literature. The signs and symptoms of HMS are common among patients seen in orthopedic physical therapy clinics; however, the HMS may be overlooked while treating individual joints or tissues causing pain. Case Description. The patient was a 28-year-old woman with complaints of chronic, multiple-joint pain. After years without a diagnosis, a rheumatologist had recently diagnosed her as having HMS. Outcomes. Following intervention that emphasized patient education and activity modification, the patient's complaints decreased. Discussion. Recognition of HMS underlying common orthopedic problems may facilitate appropriate patient education and management. [Russek LN. Examination and treatment of a patient with hypermobility syndrome. *Phys Ther.* 2000;80:386–398.]

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

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Hypermobility syndrome (HMS) is a dominant inherited connective tissue disorder described as "generalized articular hypermobility, with or without subluxation or dislocation."[1(p586)] The primary manifestation is excessive laxity of multiple joints. Hypermobility syndrome is different from localized joint hypermobility and other disorders that have generalized joint hypermobility, such as Ehlers-Danlos syndrome,[1] rheumatoid arthritis,[2] lupus,[3] and Marfan syndrome.[4] Laboratory tests are used to rule out these other systemic disorders when HMS is suspected. Fibromyalgia syndrome (FMS) often coexists with HMS and is 3.8 times more common in adults with HMS than in those without HMS.[5] Up to 81% of children with FMS have HMS.[6]

Although the pathophysiology in HMS is not yet understood, the disorder appears to be a systemic collagen abnormality. The ratio of type I to type III collagen is decreased in skin.[7] Abnormality in collagen ratios is associated with joint hypermobility and laxity of other tissues.[7] Although the diagnostic criteria for HMS involve joint abnormalities, HMS also affects cardiac tissue and smooth muscle in the gastrointestinal system and in the female genital system.[3,8] Individuals with HMS also have deficits in joint position sense.[9,10] Readers are referred to review articles[11–13] for further information about the pathology and diagnosis of HMS.

Hypermobility syndrome is diagnosed through clinical examination and laboratory tests used to rule out other disorders that may cause multiple-joint hypermobility. The most commonly reported diagnostic criteria were described by Beighton et al,[14] based on a modification of a scale proposed by Carter and Wilkinson.[15] Bulbena et al[13] compared these criteria, along with hypermobility at additional joints and other characteristics such as easy bruising. They assessed the ability of each criterion to be used to predict the presence of HMS. Table 1 shows the criteria for each of these 3 scales. Researchers and clinicians have not only failed to agree on a single scale, they have also failed to agree on a specific cut-off criterion for HMS in these scales.[11] Bulbena et al[13] found very good concurrent and predictive validity for diagnosis of HMS using 5 of the 9 characteristics in the Beighton scale, 3 of the 5 characteristics in the Carter and Wilkinson scale, and 5 of the 10 characteristics for women and 4 of the 10 characteristics for men in the Bulbena scale (a Beighton scale score of 5/9 was used as the gold standard). The Bulbena scale score provided the best ability to distinguish individuals with HMS from those who did not have HMS.[13]

Individuals with hypermobility syndrome can be taught how to decrease pain to manageable levels through joint protection and activity modification.

In this case report, I follow the guidelines in the *Guide to Physical Therapist Practice*[16] (the Guide). The examination includes patient history, systems screening, and tests and measures. Although not all of the tests and measures performed were necessary for the diagnosis of HMS and assignment using the Guide criteria, these tests and

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Table 1.
Criteria for Hypermobility Syndrome (HMS) as Defined by Beighton et al,[14] Carter and Wilkinson,[15] and Bulbena et al[13]

	Criterion	Beighton	Carter and Wilkinson	Bulbena et al
Thumb	Apposition to forearm	X[a]	X[a]	X[b]
Metacarpophalangeal joint	Hyperextension	X[c]	X[d]	X[c]
Elbow	Hyperextension ≥10°	X	X	X
Knee hyperextension	Hyperextension ≥10°	X	X	
Trunk	Flexion to place palms flat on floor while standing	X		
Ankle/foot	Excessive dorsiflexion and eversion[e]		X	X
Shoulder	Lateral rotation ≥85° from neutral (elbow at side)			X
Hip	Abduction ≥85°			X
Patella	Easily moved to the sides			X
Metatarsophalangeal joint	Dorsiflexion ≥90°			X
Knee flexion	Heel to contact buttocks			X
Ecchymoses	Eccymoses after minimal trauma			X
	Total possible points	9[f]	5	10
	Minimum score for HMS[g]	5/9	3/5	5/10 female 4/10 male

[a] Apposition of thumb to touch forearm.
[b] Apposition of thumb to within 21 mm of forearm.
[c] Hyperextension of fifth metacarpophalangeal joint to 90°.
[d] Hyperextension of fingers and wrist so fingers are parallel to forearm.
[e] No specific range identified.
[f] Right and left sides are counted separately for thumb, metacarpophalangeal joint, elbow, and knee, giving a possible total of 9 points.
[g] From Bulbena et al.[13]

measures are reported to provide readers with an example of patient presentation. The examination is followed by evaluation, diagnosis, and prognosis. Intervention comprises the treatment provided. Outcomes were assessed at 1-month and 1-year follow-ups.

Case Description

Examination

General demographics. The patient was a 28-year-old Caucasian woman who was referred for physical therapy "evaluation and patient education" by her rheumatologist, who had recently diagnosed her condition to be HMS.

Social history. There were no relevant findings for social history.

Occupation. She was employed as a physical therapist in an outpatient practice.

Growth and development. The patient was right-hand dominant. There were no relevant findings regarding her developmental history.

Living environment. She lived independently.

History of current condition. The patient reported an approximately 5-year history of recurrent, multiple-joint pain. The specific joints involved had varied and included her feet, ankles, knees, hips, shoulders, wrists, and fingers. She reported that pain usually developed without known cause, persisted for several weeks to several months, and then subsided. She described a single episode of temporomandibular joint locking when she woke in the morning. Her most frequent and debilitating pain was generally related to her wrists, which she said she injured yearly during martial arts. After approximately 5 years of recurrent pain, she saw a rheumatologist because she was unable to maintain her accustomed level of activity and was concerned that her worsening condition and multiple-joint involvement might indicate rheumatoid arthritis or another progressive disorder.

At the time of the examination, the patient had chronic, multiple-joint pain in the bilateral first metatarsophalangeals, left anterior ankle, bilateral anteromedial knees, left hip, both shoulders, right wrist, bilateral second metacarpophalangeal (MCP) joints, and right first carpometacarpal joint. She reported pain with use of these joints, particularly with movements at the end-range.

She described some activities that produced or increased the pain. Running increased the ankle, knee, and hip pain. Shoulder pain was increased when she was removing sweaters overhead or lying on her side. Wrist pain increased when she turned doorknobs, placed weight on either extended wrist, or did manual therapy such as massage or mobilizations. Twisting during martial arts and forceful gripping also led to increased pain. Thumb

metacarpophalangeal pain was increased when she did small joint mobilizations or trigger point massage.

She reported other problems of easy bruising and frequent skin lacerations with slow healing and said she had difficulty sleeping due to what she described as pain from where her body contacted the bed. She said she slept on a thick feather comforter, which decreased nighttime pain. After seeing the rheumatologist, she stated that she was no longer worried about having a progressive disease and that inability to sleep was her primary concern.

Functional status and activity level. The patient stated that she was able to perform all activities of daily living; however, pain with functional activities often required compensatory movements. For example, she was unable to lift heavy items such as frying pans or full cartons of milk with one hand. She described having an active lifestyle until approximately 1 month prior to being seen for physical therapy, when pain limited her activity. She had been jogging approximately 4.8 km (3 miles) daily while wearing 6.8-kg (1½-lb) wrist weights on each wrist, mountain biking 3 days per week, doing calisthenics 2 days per week, and participating in martial arts 5 days per week. The patient stated that she was unable to maintain her previous activity level due to pain.

Medications. She reported minimal benefit from antiinflammatory medications such as ibuprofin (600 mg twice daily, used for 2 weeks), heat, or ice. She was taking no medications at the time of the examination.

Other tests and measures. The referring rheumatologist had ruled out rheumatoid arthritis and related systemic disorders through clinical examination and blood tests. The physician had also ruled out mitral valve prolapse, which is seen with increased incidence in HMS.[7,17]

Past history of current condition. The patient reported having previously seen several orthopedic and sports medicine physicians for what she described as "repeated wrist sprains," but they gave no definitive diagnosis. The patient stated that she had had multiple wrist radiographs and a bone scan of her wrists approximately 5 years previously, without positive findings except for a bilateral slightly increased scapholunate space. After several episodes of seeing physicians without receiving diagnoses or recommended treatments, she discontinued seeking medical attention. She had attempted strengthening her wrists with wrist curl and gripping exercises, but she reported increased pain with the attempt. She ultimately self-treated her wrists with custom-made splints for 2 to 3 months and her other joints with rest as needed.

Past medical/surgical history. The patient said she had had all of the usual childhood illnesses, including tonsillitis (with tonsillectomy), chicken pox, measles, scarlet fever, and mononucleosis. She reported having had recurrent childhood right ear infections, necessitating 4 surgeries to repair the eardrum and inner ear bones. She said she had a gastric ulcer 8 years previously, with chronic low-level gastrointestinal irritability since that time. She described having an episode, approximately 1 year previous to seeing the therapist, of chronic fatigue that severely limited function outside of work. She received no definitive diagnosis for the chronic fatigue; however, use of isoniazid medication (300 mg a day for 9 months)*—after receiving a positive routine purified protein derivative (PPD) test for tuberculosis—coincided with restoration of prior levels of energy and function.

Family history. Her family history included nothing that seemed relevant to her current symptoms except that her mother had diffuse chronic joint and muscle pain. The patient recalled that her mother often complained about leg, knee, and neck pain that radiated into both upper extremities. Her mother had seen multiple physicians, and she had been diagnosed with cervical osteoarthritis and had been given various diagnoses, including osteoarthritis of the knee, sacroiliac dysfunction, and lumbar disk herniation, for her lower-extremity pain. Her mother had received medical treatment, physical therapy, or massage for various painful conditions yearly for almost 10 years. An uncle on her mother's side had died in his twenties of muscular dystrophy (type undiagnosed at the time of his death).

Health status. The patient referred to herself, half jokingly, as "a hypochondriac without a cause." She acknowledged chronic and recurrent pain and illness that physicians were often unable to diagnose. She was discouraged by the nearly constant joint pain and resulting limitation in function. She recognized that she was able to perform social roles adequately, but she was discouraged because she felt that her function was not up to her desired standards.

Social habits. The patient did not smoke, drink, or use drugs. She maintained an active lifestyle, as described earlier.

Systems Review

The patient appeared to be an energetic and fit individual. A screening review of physiologic and anatomic status was not performed at the time of the physical therapy examination because the rheumatologist

* Isoniazid is a medication indicated for actively growing tubercle bacilli; 300 mg a day for 6 to 12 months is a standard course of treatment.

Table 2.
Range of Motion and Joint Mobility Found in This Patient

Joint Motion	Range	
	Right	Left
5th finger metacarpophalangeal joint extension	90°	90°
Thumb apposition to forearm	Full	Full
Wrist extension	105°	110°
Wrist flexion	110°	110°
Elbow extension	0°	0°
Shoulder lateral rotation (measured at 90° of abduction)	125°	130°
Shoulder lateral rotation (measured at neutral)	100°	90°
Trunk flexion (standing hands to floor)	Flat	Flat
Hip medial rotation[a]	90°	85°
Hip lateral rotation	80°	85°
Straight leg raise	110°	105°
Knee extension	0°	0°
Patellar mobility: total medial to lateral excursion[b]	3.5 cm	3.5 cm
Hip abduction	55°	55°
Ankle dorsiflexion	35°	35°
Flat foot[c]	Third degree	Third degree
First metatarsophalangeal joint extension	95°	95°

[a] Joint range of motion measured according to methods described by Norkin and White.[18]
[b] Measured from medial border in maximal medial deviation to medial border in maximal lateral deviation.
[c] Using Feiss line as described in Magee DJ. *Orthopedic Physical Assessment.* 2nd ed. Philadelphia, Pa: WB Saunders Co; 1992:489.

Table 3.
Range of Motion Testing to Indicate Muscle Length in This Patient[a]

Muscle and Position	Range	
	Right	Left
Latissimus: shoulder flexion in lateral rotation (humerus relative to table)	25°	25°
Pectoralis major, sternal portion: shoulder flexion at 135° of abduction (humerus relative to table)	20°	15°
Pectoralis major, clavicular portion: shoulder horizontal abduction (humerus relative to table)	0°	0°
Pectoralis minor: posterior acromial border (height from table)	10.4 cm	9.1 cm
Psoas: Thomas test, knee held straight (thigh relative to table)	15°	20°
Rectus femoris: Thomas test, knee flexion (flexion from 0°)	40°	40°

[a] Testing performed using positions as described in Kendall and McCreary,[25] with the amount of motion indicating length.

reported doing a comprehensive review of cardiopulmonary, musculoskeletal, and neuromuscular function 2 weeks earlier. Communication ability, affect, cognition, and learning style did not appear to present problems.

Tests and Measures

Pain. The patient rated her pain as 3–5/10 (0 to 10 scale, with 0=no pain and 10=worst pain) at rest and 5–8/10 with aggravating activities. She reported that pain never decreased below 3/10 throughout the previous 3 months.

Range of motion. Passive joint ranges of motion, measured according to Norkin and White,[18] are reported in Table 2. Reliability of range of motion measurements varies depending on the joint. Intratester reliability is generally high (intraclass correlation coefficient≥.80) for wrist,[1] elbow,[2] shoulder,[3] and ankle[4] passive range of motion measurements. Straight-leg-raising measurements have been shown to have high intertester reliability (intraclass correlation coefficients=.87 and .94) for patients with chronic low back pain. No evidence was found to indicate the degree of reliability for passive range of motion measurements of the hip, patellofemoral, metacarpophalangeal, and metatarsophalangeal joints. No reliability data were found for measurements of thumb apposition to forearm. Evaluation of her non–weight-bearing rear-foot alignment (measured prone in subtalar joint neutral) indicated what I believed was associated with the measurement of bilateral rear-foot varus deformity of approximately 4 degrees and forefoot varus deformity of approximately 2 to 4 degrees, although these values are relatively small given the error (lack of reliability). Measurements of rear-foot alignment have been shown to be reliable,[23] whereas measurements of forefoot alignment are thought to be influenced by examiner experience.[24]

Despite the generalized increased range of motion, she appeared to have moderate tightness bilaterally of the pectoralis major (clavicular portion), latissimus, rectus femoris, and iliopsoas muscles (measured as described by Kendall and McCreary[25]). Again, the reliability of these clinical measurements is not known. Table 3 shows the results of range of motion testing used to indicate muscle length.

I used what is called "neurodynamic" (previously called "neural tension") testing according to procedures described by Butler.[26] Reliability and validity have not been documented for these measures, and little about their use has appeared in the peer-reviewed literature; nevertheless, I chose to use this technique. According to Butler and Gifford,[27] in some circumstances there may be abnormal physiology, causing decreased nerve gliding or stretch, but they have not provided evidence for this assertion. The patient reported pain and paresthesias along the distal median nerve distribution (forearm

and hand, bilaterally) with upper-limb tension testing 1 for the median nerve (ULTT1=shoulder abduction, scapular depression, supination, shoulder lateral [external] rotation, and elbow extension). She reported pain and paresthesias along the ulnar nerve distribution (proximal to the elbow into the hand on the left and into the forearm and hand on the right) with upper-limb tension testing 3 for the ulnar nerve (ULTT3=shoulder abduction, wrist extension, supination, scapular depression, and shoulder lateral rotation and full abduction). I could feel increased resistance, and the patient reported pain and paresthesias before obtaining the full range of motion during either of the "neurodynamic tests" proposed by Butler.[26]

Joint integrity and mobility. End-feel, although a category of questionable reliability, was assessed during passive range of motion testing, according to the characteristics described by Cyriax.[28] The end-feel at the elbows and knees was hard, but end-feel at other joints was neither firm nor empty. Although I felt some resistance, the joints felt like they might go further; motion was discontinued because of the patient's complaint of discomfort. Axial compression of her thumbs (as if doing small joint mobilizations) also caused thumb interphalangeal joint extension and MCP joint hyperflexion that the patient could not voluntarily correct. Although there is no standardized measure or norms for patellar mobility, the patient had side-to-side motion of more than one half the patellar width bilaterally. I deemed this to represent excessive patellar mobility bilaterally (Tab. 2).

The Neer impingement test (forceful shoulder elevation in medial [internal] rotation) is often used to identify rotator cuff tendon impingement,[29] although data on the reliability and validity of these measures are not known. The patient had positive Neer impingement tests in both shoulders. Acromioclavicular compression (active flexion to 90°, adduction to 15°, full medial rotation, with the subject resisting downward force) is used by some clinicians to test for acromioclavicular sprains.[30] This patient had positive acromioclavicular compression tests in both shoulders. A FABER test (flexion, abduction, and lateral rotation) may be used to screen for hip pathology.[31] The FABER test of the left hip was positive, with pain and limited mobility, compared with the right side. A Watson test[32] (radial deviation and flexion of the wrist while applying dorsal pressure on the scaphoid) for scapholunate dissociation was also positive. Although these "special tests" lack documented reliability and validity, I felt they were helpful in forming my clinical impression.

There was no redness, warmth, or other signs of inflammation at any of the involved joints. Although the patient had pain with movement of several joints, she had tenderness to palpation only at the acromioclavicular joints and the left talocrural joint.

Muscle performance. Manual muscle testing was performed according to the method of Kendall and McCreary.[25] In my opinion force was within normal limits and pain-free throughout the upper and lower extremities. Although reliability of manual muscle test grades above Fair have been shown to be low,[33] I believe my findings indicate force was not a problem for this patient.

Hypermobility testing was done using all 3 of the most common scales (see Tab. 4 for descriptions of specific tests). The patient had a Beighton scale score of 5/9, a Bulbena scale score of 8/10, and a Carter and Wilkinson scale score of 2/5. Very good interrater reliability (kappa >0.7) has been demonstrated for these indicators of the presence of HMS.[13]

Evaluation, Diagnosis, and Prognosis

Evaluation. Even when HMS is suspected, a patient with HMS may have localized pathology that should be treated. I therefore believed a full orthopedic examination was necessary. This patient had multiple-joint pain without any single traumatic episode. In her case, I believed overuse was likely a contributing factor in her chronic and recurrent pain. She had findings that I believe are consistent with active involvement of the rotator cuff (positive Neer impingement test[29,34]) acromioclavicular joint (positive acromioclavicular compression test), left hip (positive FABER test), and median and ulnar nerves (positive neurodynamic tests).[26] Most of the pain did not appear to be associated with inflammation (her joints lacked tenderness to palpation, and there was no redness or warmth). Although each joint could be evaluated and treated individually, the presence of widespread chronic, recurrent, and variable symptoms suggested to me a common underlying pathology.

Diagnosis. The examination findings were consistent with a diagnosis of HMS. The patient's Beighton scale score met the minimum criterion of 5/9, and her Bulbena scale score exceeded the minimum criterion of 5/10 for women. The inconsistency among the HMS scores was demonstrated by her Carter and Wilkinson scale score, which did not meet the minimum criterion of 3/5. In my view, it is important to identify and address the underlying hypermobility rather than treat the individual symptomatic joints. This is because I believed that her symptoms were caused by stresses that exceeded the hypermobile tissues' ability to resist. The goal of her treatment was to either decrease the stresses or increase the tissues' ability to resist.

Table 4.
Criteria for Hypermobility Syndrome (HMS) Met by This Patient (Marked as "X") as Defined by Beighton et al,[14] Carter and Wilkinson,[15] and Bulbena et al[13]

	Criterion	Beighton et al	Carter and Wilkinson	Bulbena et al
Thumb	Apposition to forearm	X[a]	X[a]	X[b]
Metacarpophalangeal joint	Hyperextension	X[c]	Not met[d]	X[c]
Elbow	Hyperextension ≥10°	Not met	Not met	Not met
Knee hyperextension	Hyperextension ≥10°	Not met	Not met	
Trunk	Flexion to place palms flat on floor while standing	X		
Ankle/foot	Excessive dorsiflexion and eversion[e]		X	X
Shoulder	Lateral rotation ≥85° from neutral (elbow at side)			X
Hip	Abduction ≥85°			Not met
Patella	Easily moved to the sides			X
Metatarsophalangeal joint	Dorsiflexion ≥90°			X
Knee flexion	Heel to contact buttocks			X
Ecchymoses	Eccymoses after minimal trauma			X
	Total possible points	9[f]	5	10
	HMS score for this patient	5/9	2/5	8/10

[a] Apposition of thumb to touch forearm.
[b] Apposition of thumb to within 21 mm of forearm.
[c] Hyperextension of fifth metacarpophalangeal joint to 90°.
[d] Hyperextension of fingers and wrist so fingers are parallel to forearm.
[e] No specific range identified.
[f] Right and left sides are counted separately for thumb, metacarpophalangeal joint, elbow, and knee, giving a possible total of 9 points.

According to the *Guide to Physical Therapist Practice*,[16] her condition was best described by Musculoskeletal Pattern E: "Impaired Joint Mobility, Muscle Performance, and Range of Motion Associated With Ligament or Other Connective Tissue Disorders." Although she may have had an inflammatory disorder at several joints, inflammation did not appear to be a major component of her current problem. The ICD-9 code for this patient was 728.5: "Hypermobility syndrome." Among the ICD-9 codes listed in the Guide, the most appropriate was 718.8: "Other joint derangement, not elsewhere classified, *Instability of joint*."

Prognosis. Prognosis for HMS is mixed. On the one hand, there is no cure for the disorder. The goal for treatment, therefore, is not return to "normal" (ie, not hypermobile) joint mobility but restoration of relatively pain-free function. That is, treatment does not eliminate the underlying impairment of excessive mobility. However, physicians specializing in HMS propose that treatment improves function and decreases disability.[6,35]

Some authors [2,12,36–39] assert that HMS is not progressive and does not necessarily lead to progressive deformity or disability in the way that rheumatoid arthritis, for example, might. From this point of view, the prognosis is good. Individuals with HMS, however, have a greater incidence of many acute and chronic musculoskeletal disorders[5,11] and tend to develop more osteoarthritis than individuals without hypermobility.[3,40,41] Hypermobility syndrome also is associated with some other systemic disorders, such as mitral valve prolapse.[6] Overall, therefore, prognosis is fair to good. In the opinion of some physicians and in my clinical experience, patients with HMS can function and their quality of life often can be improved with treatment but they will usually have chronic or recurrent problems.

Intervention

Coordination, communication, and documentation. I communicated with the physician to obtain the medical diagnosis and results of the physician's examination. I developed plans for patient education and documented how I conducted the examination, evaluation, and intervention.

Patient/client-related instruction. The primary emphasis of intervention with this patient was education about the syndrome, about body mechanics and joint protection, and about lifestyle modification. I described the disorder to the patient as a noninflammatory, nonprogressive connective tissue disorder. This description reassured the patient that she did not have a progressive rheumatoid-type disorder that would lead to worsening disability or deformity. In this case, the rheumatologist had also explained the disorder.

Functional training in self-care and home management/ functional training in community and work integration. I told the patient that her joints were vulnerable to stress at end-range and that passive stretches and positions that would not cause problems for an individual without HMS could cause chronic or recurrent problems for her.

Although research on joint protection has not been done on HMS, the joint instability in HMS is similar to that seen in the active phase of rheumatoid arthritis. In rheumatoid arthritis, research has shown that some forms of education regarding joint protection can increase function and decrease pain.[42] She was instructed to modify her body mechanics and ergonomics to avoid stretching her joints past end-range during work, daily activities, and exercise. She was advised not to move her joints into end-ranges. For example, I advised her to modify the techniques she used at work to protect her joints and maintain them at or near midrange and to avoid techniques such as doing joint mobilizations with an extended wrist or a hyperflexed thumb MCP joint. During exercise and recreational activities, she was to maintain joints in midrange.

Therapeutic exercise. Although individuals with HMS are typically given a strengthening program in an effort to provide muscular stability to involved joints, I felt that this patient's high levels of exercise were excessive. She was instead advised to discontinue use of wrist weights while running and to eliminate or limit participation in calisthenics and martial arts. Patients with HMS may be given exercises such as balance and coordination exercises (eg, use of a wobbleboard) to improve their joint position sense.[9,10] Because this patient was athletic and active, I did not believe that additional exercises were appropriate at this time. Although stretching of tight muscles was not recommended, the patient was advised that if she chose to stretch tight muscles, stretching should be done selectively to those muscles with documented tightness and stretching techniques needed to isolate tight muscles and not impose stress on surrounding joints. For example, hip flexor stretches should not allow excessive lumbar lordosis.

Prescription, application, and, as appropriate, fabrication of devices and equipment (protective and supportive). The patient was advised to use protective and supportive splints as needed. When doing small joint mobilizations, she could wear a thumb spica splint. When bicycling more than 30 minutes (the amount of time for her symptoms to typically appear), she could wear wrist splints to prevent prolonged stretching of the wrists into extension. Because martial arts were a contact activity, precluding use of rigid wrist splints, she was encouraged to tape her wrists to limit motion. To protect finger joints during manual therapy at work, she could use products designed to assist trigger point massage. She was advised to select footwear with adequate arch and calcaneal support or to use orthoses to provide support for her excessive pronation.

The patient stated that she understood the explanation of the disorder and my instructions. The patient stated that she was comfortable making the recommended activity modifications. I told her to contact me in 1 month, or sooner, if she had questions.

Outcomes: 1-Month Follow-up

Approximately 1 month following the physical therapy consultation, I contacted the patient by telephone to ask about her status. The patient estimated that her pain had decreased by approximately 30%. She stated that she consciously avoided end-range and passive joint stretches during both vocational and avocational activities. She rated her pain as 0–3/10 on average and 3–5/10 at worst. She estimated that 30% of her waking time was pain-free.

Functional limitation/disability. The patient reported decreased pain at work and during activities such as massage and joint mobilizations when she monitored body mechanics and minimized stress to joints. She was able to perform all activities of daily living and all work-related activities with some compensations but pain below 3/10. She had not returned to her desired recreational activity level, but, by discontinuing calisthenics and the use of wrist weights while jogging, she was able to resume approximately 70% of her prior level in activities that were most important to her (ie, martial arts and jogging with her dog).

Secondary prevention. The patient stated that, through understanding her disorder, she had been able to identify certain activities that appeared to be responsible for pain in certain joints. For example, when she discontinued wearing wrist weights while running, she no longer had acromioclavicular joint pain. When she discontinued hyperextending her index finger MCP joint by using her index finger as a shoe horn to don shoes and to doff socks, her index finger MCP joint pain disappeared. The patient also recognized that during martial arts she sometimes kneeled with ankles dorsiflexed and toes hyperextended, supporting her body weight. When she altered her toe position, her first metatarsophalangeal joint pain was eliminated. When she used arch supports and 4-mm medial rear-foot posting in her running shoes, patellofemoral pain decreased considerably. She reported decreased wrist and thumb pain during manual therapy techniques and martial arts by maintaining joints at midrange during those activities.

Although the patient was pleased with her improvement, she reported continued pain at multiple, though fewer, joints. Involved joints varied depending on activity. The most common complaints involved the metatarsal arches, left ankle, knees, low back, left medial elbow/forearm, and both wrists. She also reported continued difficulty sleeping due to diffuse nighttime discomfort. The patient felt she was managing these complaints,

other than the sleep disturbance, adequately with self-care. The patient and I agreed that additional physical therapy intervention was not necessary at this time.

Outcomes: 1-Year Follow-up

One year following the initial physical therapy evaluation, I again contacted the patient by telephone. She reported a decrease in frequency of joint pain (50% of the time she was pain-free). She rated her pain, when present, as 3/10 at rest and 5/10 with aggravating activities. The specific joints involved at any one time continued to vary, but included the same areas that had been painful at the 3-month follow-up.

Functional limitation/disability. She reported that activity modification protecting one joint sometimes transferred stress to other joints, which subsequently became symptomatic. Therefore, she was unable to avoid stress to all joints all of the time. Protecting her joints limited her function somewhat, but did not seriously compromise her ability to work, maintain a household, and participate in recreational activities. She stated that she had decreased her running to 3 times per week and had eliminated calisthenics to "save" her joints for activities such as work, gardening, and martial arts.

Secondary prevention. She reported involvement of additional joints over the year. She reported 2 episodes of acute low back pain with radiation into the left leg caused by bending over while digging in her garden. She reported that she was able to decrease radiation of pain into the leg with lumbar extension exercises and by maintaining lumbar extension during all functional activities. She also reported symptoms of left buttock and anterior hip pain. She reported acute bilateral metatarsal head pain that consistently prevented her from walking barefoot on hard surfaces and sometimes prevented her from walking her dog. She reported intermittent acute left anterior shin and ankle pain.

The patient also reported increased pain and paresthesias over the medial elbow and ulnar border of the forearm and hand. She attributed these symptoms to increased use of the telephone (prolonged elbow flexion) at work.

She reported having had continued difficulty sleeping due to multiple-joint pain. She believed that inability to sleep had begun to cause recurrent headaches. She had returned to her rheumatologist (also her internist), who prescribed low doses (25 mg) of nortriptyline (a tricyclic antidepressant that acts as a serotonin reuptake inhibitor). The patient reported a dramatic decrease in nighttime discomfort, with resultant improvement in sleep and remission of headaches, with use of the medication.

Table 5.
Ten Musculoskeletal Characteristics Most Common in People With Hypermobility Syndrome (HMS)[13]

Characteristic	Incidence in 114 Subjects With HMS
Excess ankle dorsiflexion and foot eversion	94%
Finger metacarpophalangeal joint extension past 90°	93%
Thumb abduction to the forearm	92%
Patellar hypermobility	89%
Shoulder lateral rotation	84%
Hip abduction	78%
Knee hyperextension past 10°	77%
Elbow hyperextension past 10°	75%
Ecchymosis	63%
Metatarsophalangeal joint extension past 90°	61%

Overall, the patient reported that she was able to manage her ongoing joint pain and was moderately content with her modified lifestyle. She stated that she was able to recognize the onset of both acute and overuse injuries sooner and was sometimes able to intervene to decrease severity.

Discussion

Examination: History and Tests and Measures

This case report presents a patient with HMS. Hypermobility syndrome is 1.1 times[43] to 5.5 times[14] more prevalent in women than in men. Her 5-year history of multiple-joint pain is typical, as individuals with HMS often have complaints that have lasted from 15 days to 45 years (average time=6.5 years).[40] The absence of acute trauma, inflammation, and swelling is common in patients with HMS and may confound diagnosis. Because the patient had normal force production and no decrease in mobility and lacked clear radiologic changes, prior medical evaluations had not identified a pathology. Her history of seeking medical assistance multiple times without diagnosis or beneficial treatment is also common among patients with HMS.[12]

This patient had a history and physical findings typical of HMS. Table 5 shows the 10 joints most likely to be hypermobile.[13] This patient had all of the characteristics of hypermobility listed in Table 5 except abduction of the hips and hyperextension of the elbows and knees. Patients with HMS may have had an acute injury, in which case examination may identify a sprain, subluxation, tendinitis, nerve compression, or other pathology that may need to be addressed in the intervention. This patient's symptoms are also typical of those of patients with HMS. Table 6 shows common complaints among patients with HMS; this patient shared at least 8 of the 15 complaints. Paresthesias, although not caused by a joint

Table 6.
Distribution of Complaints in This Patient Compared With Typical Complaints for Individuals With Hypermobility Syndrome (HMS)

Presenting Complaint	Incidence in 114 Subjects With HMS[a]		Complaints in This Patient
	Number	Percentage	
Soft tissue rheumatism (tenosynovitis and tendinitis)	84	73.7	
Paresthesia (hands and/or feet)	66	57.8	X
Knee pain	37	32.4	X
Low back pain	28	24.6	X
Shoulder pain (and/or stiffness)	25	21.9	X
Heel pain and/or plantar fasciitis	24	21	
Elbow epicondyle pain	24	21	
Polyarthritis/polyarthralgia (more than 4 joints)	15	13.1	X
Hand and wrist pain (in small joints)	14	12.3	X
Cervical region pain	11	9.7	
Fibromyalgia (myalgic spots)	11	9.7	
Dorsal region pain	10	8.8	
Ankle pain and/or swelling	8	7	X
Forefoot pain (metatarsalgia)	7	6.1	X
Heel cord and calf pain	5	4.3	

[a] From el-Shahaly and el-Sherif.[40]

disorder, are common in patients with HMS and were seen in this patient. The reason for the prevalence of nerve compression disorders is not clear.

This report describes the results of tests that a physical therapist is likely to do with a patient having these complaints. It does not attempt to validate the specific tests and measures chosen for the examination. A review of the literature[11] did not reveal any published reports of physical therapy examination findings other than the range of motion tests used to identify HMS. In addition, no reports were found describing the muscle tightness observed in this otherwise hypermobile patient.

Sleep disturbance, although seldom studied in patients with HMS, may be common: 90% of individuals with both HMS and fibromyalgia reported sleep disturbances,[6] and the incidence of HMS appears to be increased among people with fibromyalgia.[5,6] The relationship between HMS and fibromyalgia suggests that HMS might share some of the physiological abnormalities, such as the decreased cerebrospinal fluid seratonin levels, seen in fibromyalgia.[44] Low doses of tricyclic medications, such as that prescribed in this case, are often effective in treating the sleep disturbance seen in both fibromyalgia[45] and headaches,[46] but these effects have not been previously described in patients with HMS.

The family history of diffuse chronic pain is also consistent with HMS as a dominant inherited disorder.[1,12] Osteoarthritis, particularly of the cervical spine, is a common sequela of HMS, raising suspicion that the patient's mother may also have HMS.[3,40,47] At this stage, however, sequelae such as osteoarthritis may limit the mother's mobility. In older individuals, therefore, failure to meet the HMS criteria according to Bulbena et al[13] or Beighton et al[14] might not rule out the presence of the underlying connective tissue disorder found in HMS.

Evaluation, Diagnosis, Prognosis

In general, the correlation among the 3 HMS scales is good.[13] This patient, however, scored high according to the Bulbena scale, achieved the minimum required score according to the Beighton scale, and did not meet the criteria for HMS according to the Carter-Wilkinson scale. One of the limitations in the Beighton scale is the limited number of joints tested. In this individual, 4 of the 5 scored joints were in the hand; she might have had localized hypermobility but not generalized hypermobility, even though she met the Beighton scale criteria. The Beighton scale score is not correlated with the severity of symptoms.[2] Although this patient had a Beighton scale score of 5/9, her symptoms were widespread and chronic. The Bulbena scale includes joints throughout the body and theoretically should provide a better assessment of generalized hypermobility. This patient's score of 8/10 on the Bulbena scale appears to reflect her widespread, chronic pain. Therefore, the Bulbena scale scoring criteria, in my opinion, should be recommended as the standard test for HMS.

This patient failed to meet the criteria for HMS according to the Carter-Wilkinson scale, in part because that scale requires simultaneous MCP joint and wrist extension to lay the fingers parallel to the forearm. This maneuver may stretch the extrinsic finger flexor muscles more than the MCP joint. Because this patient appeared to have several shortened muscles, despite her joint

laxity, it is not surprising that she failed to meet this criterion.

In this case, the diagnosis of HMS was first made by a rheumatologist who had already performed laboratory and clinical tests to rule out related and potentially more serious disorders. The physician had tested for associated systemic disorders such as mitral valve prolapse and had conducted laboratory tests to rule out rheumatoid and other inflammatory polyarthritic conditions.[38] Ehlers-Danlos and Marfan syndromes are other hereditary connective tissue disorders with associated joint hypermobility[48] that must be excluded before a diagnosis of HMS can be made.[17,39,49] Clinical findings of hyperelastic skin,[50] hernias, lenticular abnormalities,[40] and abnormal body proportions[38] are seen in people with Ehlers-Danlos and Marfan syndromes but not in people with HMS. Easy bruising[13,51] and poor wound healing may be seen in patients with HMS as well as in people with Ehlers-Danlos and Marfan syndromes. Osteogenesis imperfecta is another collagen disorder that might need to be ruled out, as patients with this disorder often demonstrate joint hypermobility.[52] Systemic lupus erythematosis,[3] poliomyelitis, tabes dorsalis, myotonia congenita, and neurological flaccid conditions[38] are also excluded from the diagnosis of HMS. If I had been the first health care professional to recognize possible HMS, I would have advised the patient to see a physician to rule out other serious disorders that have multiple-joint hypermobility as a finding to consider HMS as a diagnosis.

The prognosis for this patient was typical for individuals with HMS, who often continue to have pain, but they can be taught to decrease pain to manageable levels through activity modification. Some of the individual impairments, such as the underlying hypermobility, will not change with intervention. Inflammation or pain may resolve at specific sites, but they are likely to be replaced with complaints elsewhere, as seen in this patient. I believe it is important, therefore, to distinguish prognosis for impairments from prognosis for function and disability.

Intervention

Review of the literature showed no research regarding efficacy of treatment for HMS.[11] Consequently, intervention was directed by my clinical experience in treating patients with HMS. Although most patients with HMS are given strengthening exercises, this patient had an extremely high level of activity prior to diagnosis. The patient, therefore, was advised to decrease activity, particularly those activities that I believed stressed her joints at the end-range. The decrease in pain with activity modification supported my hypothesis that joint stresses were causing some of her chronic problems. For example, the wrist weights appeared to have created distraction of the acromioclavicular joints sufficient to result in symptoms of a mild acromioclavicular sprain. Excessive forceful gripping, twisting, and compression through the extended wrist may have contributed to her wrist sprain, and running may have contributed to her patellofemoral syndrome. Instability of the thumb MCP joint, aggravated by frequent use to do joint mobilizations, could have produced chronic thumb MCP pain.

This patient did not participate in exercise in the clinic, although in my experience patients with HMS often benefit from guided, progressive exercise programs emphasizing joint stabilization and joint position sense. Furthermore, this patient was a physical therapist, so the treatment involved less intervention than might be appropriate for another patient with HMS. Once she was made aware of the increased vulnerability of her joints, this patient was able to evaluate her body mechanics and apply principles of joint protection independently. Patients with less knowledge about body mechanics and exercise might require greater amounts of guidance and training in ergonomics and body mechanics.

Other than addressing body mechanics at each involved joint, intervention was not directed at treating each of the patient's impairments. If preventing joint stresses had not been adequate to relieve symptoms, a brief period of direct intervention such as those suggested for pattern E in the Guide[16] (eg, physical agents or manual therapy) might have been appropriate. I believe the focus of intervention for patients with HMS must ultimately lie with function and disability rather than impairment. Patient education is likely to be the most important intervention for all patients with HMS.

Outcome

This patient's outcome, in my opinion, was typical for patients with HMS. She was able to decrease impairment and functional limitation, but she was not able to prevent them entirely. The 1-year follow-up demonstrated that although this patient had continued diffuse pain, and she reported a greater sense of control over her disorder as a result of accurate diagnosis and education. Because she now knew the limitations of what the medical community could do for her disorder, she decreased use of health care services that she knew would be of no benefit. Although she had modified some of her goals to accommodate her physical limitations, overall she felt that she was able to satisfy role expectations of work, home, and recreation adequately and up to her desired standards. The patient and therapist agreed that these were optimal outcomes for this patient at this time.

Conclusion

This case report describes a patient with diffuse, chronic, and recurrent pain due to HMS. Diagnostic criteria proposed by Bulbena et al,[13] although not currently the most commonly used criteria, appear to be the most effective at identifying generalized hypermobility. Although sensitivity and specificity have not been computed for the Bulbena scale criteria, these criteria have demonstrated reliability, validity, and internal consistency.[13] The patient examination, evaluation, diagnosis, prognosis, intervention, and outcome were discussed to aid physical therapists in recognizing and treating patients with this disorder. Recognition is particularly important when patients have chronic or recurrent pain or a nonspecific diagnosis or when they have had extensive medical testing without a definitive diagnosis. Patients, such as the patient described in this case report, may be seen within the medical system multiple times over a period of years without recognition of the underlying HMS. Education about the nature and course of HMS frequently reassures patients that they have a real disorder, but one that is not inherently progressive. There is no published literature on the efficacy of medical or physical therapy management of HMS. Research is needed to support or refute the recommendations proposed here.

Education and activity modification provide the core of intervention for HMS. Strengthening and proprioception exercises may be helpful to improve muscular stability at specific joints. Use of protective splints may also be beneficial. Treatment of specific joint disorders may be appropriate, especially in the presence of acute trauma or inflammation. Physical therapists also should recognize and address the underlying hypermobility. Intervention should emphasize joint protection and injury prevention, as both traumatic injuries and chronic pain are likely to be recurrent.

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