Clinical Reasoning Considerations for the "Flexible" Patient: A Ligamentous Laxity Overview

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ABSTRACT

Background and Purpose: Individuals with excessive joint hypermobility often seek physical therapy care. Despite the common clinical occurrence, a consensus is lacking on how to best conservatively address the specific needs of this poorly understood population. The aim is to explore concepts and clinical reasoning considerations when treating a person with a joint hypermobility syndrome (JHS). **Methods:** A pragmatic outline was established including clinical manifestations, evaluation, prognosis, and clinical reasoning processes to determine intervention. The literature was identified through PubMed and CINAHL. **Clinical Relevance:** Only a subset of individuals with joint hypermobility become symptomatic. Joint hypermobility syndrome includes many ligamentous laxity conditions requiring the clinician to appreciate different disease characteristics. There are questionnaires and objective evaluation tools available to assist with developing individualized treatment. **Conclusion:** The evaluation and construction of a meaningful treatment plan for individuals with JHS can be challenging. Combined clinical knowledge and sound clinical reasoning processes can assist with optimizing outcome.

Key Words: Ehlers-Danlos, generalized hypermobility, joint hypermobility

INTRODUCTION

Joint hypermobility is defined as the ability of a joint to move past the clinically defined normal standards for range of motion (ROM).[1] It can occur at one joint or at multiple joints throughout the body. When excessive motion occurs in multiple joints, it is characterized as generalized joint hypermobility (gJHM). Generalized joint hypermobility is asymptomatic with no functional loss despite having increased ROM. Identifying an individual with gJHM is often made using the Beighton score.[1] The cut-off scores for the Beighton assessment are inconsistent and lack a consensus on how to best identify individuals with gJHM.[1,2] This is further supported by a large prevalence range from 2-57% for individuals with gJHM indicating inclusion criteria remains uncertain.[3] Typically, a Beighton score of 4 out of 9 indicates gJHM in a general adult population.[1] There are some additional studies indicating gJHM is present in women if a score of 5 of 9 is achieved, and a score of at least 6 out of 9 is needed to determine the presence of gJHM in children.[1,2]

Joint hypermobility syndrome (JHS) occurs when joint hypermobility becomes symptomatic. These symptoms were previously believed to be only limited to localized pain, instability, and decreased proprioception. However, a progressive understanding appreciates this condition is much more complex. Due to the nature of the tissues affected, the condition can present in a variety of ways. In addition to the musculoskeletal complaints, such as increased likelihood for joint sprains, meniscal injuries, and stress fractures, other body systems are affected manifesting as disturbances in pain perception, anxiety, fatigue, and gastrointestinal interruptions.[4-7] Congenital conditions that present with ligamentous laxity and subsequent joint hypermobility are Down's syndrome, Marfan syndrome, Loeys-Dietz syndrome, and Osteogenesis imperfecta.[8-10] Joint hypermobility syndrome is considered by some sources to be a mild form of Ehlers-Danlos syndrome (EDS) hypermobility type while other sources indicate JHS is a diagnosis of exclusion and separate from EDS.[8]

Conservative management by a physical therapist is often the preferred first method of treatment for these conditions due to their musculoskeletal nature. Treatment can vary from stability exercises, proprioception training, and patient education. Patient education is focused on modifying movement, lifestyle changes, and addressing persistent pain.[11] Unfortunately, there is little consensus for the best way to manage individuals with a joint hypermobility condition; therefore, an increased awareness and understanding of JHS is important as physical therapists are the best health care provider to appreciate the specific needs in this hypermobile population.

The aim of this article is to present a review of the literature regarding JHS and offer clinical information to conservatively manage individuals with suspected or confirmed joint hypermobility syndromes. Lastly, the article could serve to identify knowledge gaps and areas for future research.

JOINT HYPERMOBILITY CONDITIONS AND CLINICAL MANIFESTATIONS

Individuals with joint hypermobility often present to physical therapy due to joint pain.[6] Physical therapists must recognize the patient's underlying condition and how it is contributing to their current complaint. The physical therapist should appreciate the various characteristics these conditions present with in order to properly address the individual patient's needs.

The common characteristics of EDS, Marfan syndrome, Osteogenesis imperfecta, and Down's syndrome are listed in Table 1. Table 1 identifies the most common condition characteristics that may help construct a differential diagnosis, although it is not a complete list of symptoms related to joint hypermobility. Ehlers-Danlos syndrome presents with many different types. Ehlers-Danlos syndrome, hypermobility type III is the most common and has an almost identical clinical presentation to JHS.[4,12] Unfortunately, JHS can often be considered a diagnosis of exclusion.[8]

In addition to musculoskeletal complaints, many individuals may report high levels of fatigue, depression, and anxiety with any ligamentous laxity condition.[8] Other clinical observations may include a lack of proprioception, generalized hyperalgesia, various neuropathies including tarsal tunnel and carpal tunnel syndrome, ptosis, varicose veins, low bone density, and postural orthostatic tachycardia syndrome. Patients may present with bowel and bladder dysfunction, including pelvic organ prolapse.[8,13]

CLINICAL EVALUATION

Individuals may arrive to physical therapy with an undiagnosed ligamentous laxity condition. It would be prudent for the physical therapist to properly screen for ligamentous laxity and consider referral to the proper medical provider for diagnosis and additional management.

A thorough subjective history is recommended during a clinical evaluation for an individual with suspected JHS. The subjective intake should aim to gain an understanding of the current and past injury and health history, mechanism of injury, and aggravating and alleviating factors. Identifying how these complaints influence functional loss is important.

The objective evaluation should include ROM measurements while noting if these are outside of typical norms. Strength measures and a general neurological screen should be assessed. Blood pressure and heart rate measurements within the initial session is advisable due to the common occurrence of related hypotension. Functional tasks should be observed to understand the individual's movement strategies, motor control, and compensations. Both daily functional tasks and sport specific tasks should also be observed.

Subjective Examination

The Hakim and Grahame questionnaire (Table 2) and musculoskeletal and non-musculoskeletal screening questions (Table 3) can assist with developing a list of differential diagnoses.[8,13,14] It is important to investigate the timeline of symptom development, especially childhood presentations, to determine a progression or long-standing presentation of related injuries or pain. The individual may describe multi-system involvement, including gastrointestinal, vascular, and bowel/bladder issues. They may report clumsiness, unsteadiness, or coordination deficits. After ruling out more serious pathologies, these responses can increase suspicion of a JHS diagnosis. The patient responses to the Hakim and Graham short questionnaire will assist in development of a thorough objective examination and patient centered goals.[14]

Objective Examination

The Beighton score is a widely used measure of gJHM and is helpful in quickly observing if excessive ROM is present in multiple joints. An adult individual is considered positive for gJHM with a score of 4 out of 9 or greater; for children 6 out of 9 or greater (Table 4).[15] Positive responses to questions from Table 2 can lead the clinician to perform movements described in the Beighton score during the examination for additional objective data. This information can then be incorporated into the Brighton score (Table 5) to determine if a JHS diagnosis is suspected.[16] Joint hypermobility syndrome is considered present when the individual presents with one of the following: (1) 2 major criteria, (2) 1 major and 2 minor criteria, and/or (3) 4 minor criteria (Table 5).[16] Recall, symptomatic complaints limiting function is a key characteristic difference between gJHM and JHS.

Additional static and dynamic balance measures may be helpful in developing a complete clinical picture. This is because individuals with JHS frequently have vestibular and somatosensory dysfunction.[17,18] Static measures may include single leg stance with eyes open, eyes open with cervical extension, and eyes closed.[17,18] Dynamic measures may include single leg squat, single leg hop tests, Y-balance test, or star excursion balance test.[1,17] Impairments may be found in some or many of these measures to help with development of the individual's plan of care. Typical outcome forms, such as the Hip Outcome Score, may be used to periodically assess functional progress, or decline, during the plan of care.[19]

This is not an exhaustive list and additional objective measures may be needed to address a specific individual's complaint and goals.

Table 1. Review of Specific Joint Hypermobility Syndromes Along with Common Characteristics to Assist with Recognition

Joint Hypermobility Syndromes	Common Clinical Presentations
Ehlers-Danlos syndrome[8] • Classic Type (I) • Vascular Type (IV) • Kyphoscoliotic Type (VI-A) • Musculocontractural Type (VI-B) • Dermatosparaxis Type	Bilateral clubfoot Developmental delays Dysmorphic facies Extensive and easy bruising Large hernias Marfanoid habitus Muscle weakness Scleral fragility Scoliosis Sensory neural hearing loss Severe muscle dystonia Severe muscle hypotonia Skin hyperextensibility Thin translucent skin Velvety skin texture
Marfan syndrome[8]	Ascending aorta dilation Fingers and toes abnormally long and slender Funnel chest High palate Muscle hypoplasia Scoliosis
Osteogenesis imperfecta[10]	Aortic root dilation Conductive deafness Decreased pulmonary function Heart murmurs Scoliosis Teeth discoloration
Down's syndrome[9]	Brachycephaly Flat nasal bridge Folded ear Gap between 1st and 2nd toes Incurved 5th finger Muscular hypotonia Narrow palate Nystagmus Oblique eye fissure Short neck

Table 2. Five Question Screening Questionnaire to Assist Clinicians to Identify Individuals with Joint Hypermobility[14]

Patient Questions to Ask if Joint Hypermobility is Suspected
- Can you now (or could you ever) place your hands flat on the floor without bending your knees?
- Can you now (or could you ever) bend your thumb to touch your forearm?
- As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- As a child or teenager, did your shoulder or kneecap get dislocated on more than one occasion?
- Do you consider yourself double jointed?
A "Yes" answer to 2 or more of the above questions has 80% sensitivity and 90% specificity for indicating the individual has joint hypermobility.[2]

Table 3. Multi-system Screening Questions for Individuals with JHS (Adapted Questions)[8,13]

Subjective Questions	Common Responses
1. Did you have any injuries or notable periods of pain as a child?	Periods of joint pain commonly occur in the posterior knees. Also, the patient may report a history of benign paroxysmal nocturnal leg pain (growing pains).
2. Did your subluxation/dislocation and/or fracture occur without great provocation?	Minimal impetus is needed for the fracture or subluxation/dislocation to occur.
3. Do injuries take a long time to heal?	Injuries may heal more slowly than standard tissue healing timeline.
4. Do you have a family history of joint hypermobility?	Often times there is a positive family history.
5. Can you describe your pain?	Pain is often reported as dull.
6. When do you have your pain?	Reports baseline pain but symptoms are made worse with activity. Symptoms typically feel the best in the morning and worst at the end of the day. Activities that use the involved joint influences pain.
7. Do you feel fatigue?	Fatigue, sometimes severe, is a common symptom, as well as sleep disturbance.
8. Do you have headaches?	Headaches are a common symptom; these may be migraines or other.
9. Do you ever feel lightheaded?	Reports feeling lightheaded or dizzy at various times. Low blood pressure, a fast heart rate, and increased sympathetic tone are common symptoms.
10. Do you have any stomach discomfort?	Commonly reports bloating, nausea, or vomiting after meals. Often encourages eating less.
11. Do you feel uncoordinated or clumsy?	Balance deficits, unsteadiness and clumsiness are symptoms are often reported.
12. Are you experiencing any symptoms that you feel are unrelated to the incidence bringing you to physical therapy?	Symptoms may include bowel and bladder dysfunction and prolapse of pelvic organs.

PROGNOSIS AND CLINICAL REASONING PROCESS FOR DETERMINING INTERVENTION

The prognosis for JHS is generally considered good since it is a nonprogressive and noninflammatory condition. Joint hypermobility tends to naturally decrease as the individual ages providing a natural "protection" to the joint.[8] Common sense reasoning indicates preserving the joint will ultimately promote and sustain function; however, available data to support this concept is lacking. Longitudinal studies are needed to ultimately determine long-term prognosis associated with the recommended management strategy, but it is recognized there are short-term benefits to conservative management including pain control and functional capacity.

While it is necessary to address the individual's area of primary concern, it is likely the individual will have, or currently has, multiple areas of pain or dysfunction. Areas of pain and dysfunction should be addressed directly while also incorporating general exercise strategies. The hypermobile person may benefit from an individualized exercise program but detailed information on a well-rounded program is not well established.[20,21] Clinicians may incorporate aerobic capacity, strength, coordination, and motor control training that address all systems rather than only the direct areas of pain. This approach may also assist with long-term self-management of symptoms. For those individuals with high irritability or difficulty participating in full weight-bearing activities, low impact training like water aerobics, modified swimming strokes, water treadmill, body weight supported treadmill, or elliptical may be beneficial to begin an exercise program.

Fatigue must be considered when developing an exercise program since it is a very common symptom within the JHS population. Clinicians should ensure proper education on a gradual increase in duration of activity with greater rest times to allow for proper recovery and joint protection. A common complaint can also include disturbance in restful sleep. If there is a disturbance in sleep reported, guidance on proper sleep hygiene and education on sleep positioning may assist to promote successful sleep.

The proprioceptive impairments typically observed in the JHS population can be addressed with closed kinetic chain strengthening and training on dynamic surfaces.[22,23] These individuals will likely need postural education during functional tasks that may include use of tactile cues, taping, and mirror

Table 4. The Beighton Score is a Clinical Objective Test for Joint Hypermobility. Variability exists for cut-off scores.[1]

Beighton Score	Scoring
- Passive flexion of the thumb allows the touch of the volar aspect of the forearm (repeat on both sides)	1 point per side
- Passive hyperextension (>90°) of the fifth finger with the palm and wrist touching a solid surface (repeat on both sides)	1 point per side
- Active hyperextension (>190°) of the elbows with the upper limb extended and the palm turned up (repeat on both sides)	1 point per side
- Active hyperextension (>190°) of the knees while the subject stands up (repeat on both sides)	1 point per side
- Active hyperextension of the lumbar spine by inviting the subject to touch the floor with both palms but without flexing the knees per side	1 point
Generalized joint hypermobility: ≥4 for adults[1,2,15] **Children:** ≥5, 6 or 7 is remarkable for joint hypermobility[15] **Female Adults:** ≥ 5 is remarkable for joint hypermobility[15]	

Table 5. The Brighton Score for Joint Hypermobility Syndrome and Classification Criteria[16]

Brighton Score for Joint Hypermobility Syndrome
Major Criteria
1. Beighton score of 4/9 or greater
2. Arthralgia for more than 3 months in 4 or more joints
Minor Criteria
1. A Beighton score of 1, 2, or 3 out of 9 (0-3 if over age 50)
2. Arthralgia for ≥ 3 months in 1-3 joints, or back pain ≥ 3 months, or spondylosis, spondylolysis, spondylolisthesis
3. Dislocation or subluxation in more than one joint, or in one joint on more than one occasion
4. Soft tissue rheumatism in ≥ 3 locations (eg, epicondylitis, tenosynovitis, bursitis)
5. Marfanoid habitus
6. Abnormal skin (eg, striae, hyperextensible, thin, papyraceous scarring)
7. Eye abnormalities (eg, drooping eyelids, myopia and mongoloid slant)
8. Varicose veins or hernia or uterine/rectal prolapse
Remarkable for Joint Hypermobility Syndrome if: - Two major criteria are present **OR** - One major and two minor criteria are present **OR** - Four minor criteria are present

feedback due to impaired proprioceptive awareness.[13]

Medical management including nonsteroidal anti-inflammatory medication could assist in reducing acute symptoms; however, this is not recommended as a long-term management strategy.[13] The physical therapist should screen for the presence of anxiety and depression as these are frequently observed in this population.[24] Consider a mental health referral if screening is positive and especially if the individual's mental health is promoting fear-avoidance behavior. Cognitive behavior therapy may be recommended to assist with coping strategies and to address any associated fear and anxiety of future injury in these individuals.[8]

Most individuals with JHS can be conservatively managed; however, if there are repeated joint subluxations or dislocations with related pain and functional loss, a surgical referral should be considered.[25] The common goal should be to preserve the longevity of the joint by reducing repeated injury.

Individuals with JHS will likely need long-term follow-up with a physical therapist due to fluctuations in symptoms and potential involvement of multiple joints and body systems. Once the individual's acute symptoms have stabilized and an individualized exercise program has been developed, less frequent visits are recommended with continued monitoring. Deductive clinical reasoning processes must be incorporated to best direct the patient.[26] These individuals may also benefit from use of telehealth services or other remote communication media for ongoing monitoring to eliminate the need for frequent in-clinic visits.

CONCLUSION

It is important to appreciate the difference between asymptomatic gJHM and symptomatic JHS. Only when individuals with joint hypermobility become symptomatic is it important to consider the varying possible diagnosis associated with JHS. Proper conservative management at any stage of the hypermobile condition can be meaningful. Earlier intervention would be optimal as education and intervention could influence the trajectory of the individual's condition to best preserve overall joint health.

It is advisable to subjectively screen individuals with suspected ligamentous laxity issues while considering specific objective tests, such as the Beighton and Brighton score, to quantify the overall joint hypermobility. The combined subjective and objective information will help develop an individualized treatment plan and estimate prognosis. Conservative management recommendations can and should include low impact aerobic exercise, proprioception and balance training, and strength building activities. Addressing any mental health needs may also be necessary, especially if functional progress is impeded. Lastly, stepping away from a "joint only" treatment approach is necessary when working with individuals with JHS. The clinician much appreciate JHS is a multi-system issue in order to optimize both short-term and long-term outcomes. Sound clinical reasoning can assist with development of an effective conservative management strategy to best match the patient's needs. Addressing single joint flare-ups or localized injury associated with a ligamentous laxity syndrome may be necessary in the short-term, but if a person with multi-joint, non-traumatic issues seeks care, then a comprehensive approach should be considered to guide the patient to a long-term optimal outcome. This literature review identified future research could include systematic reviews on

conservative management for individuals with JHS. Recognizing the unique characteristics and special needs of this under-recognized and under-studied population is necessary to best promote optimal care for the "flexible" patient.

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