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Clinical management of benign joint hypermobility syndrome: a case series

Patricia Adele Boudreau, BSc, DC[1]
Igor Steiman, BSc, DC, MSc, FCCS(C)[2]
Silvano Mior, DC, FCCS(C), PhD[3]

Background: *Benign Joint Hypermobility Syndrome (BJHS) is a relatively prevalent condition of the spectrum of heritable connective tissue disorders, with musculoskeletal, visceral and psychological manifestations. The conservative management of the musculoskeletal symptomatology must be modified for optimal effectiveness and minimal sequelae.*

Purpose: *To provide an overview of the presentation, assessment, chiropractic management, and outcomes of patients with BJHS.*

Study Design: *Case series*

Discussion: *Recognizing joint hypermobility as a significant contributing factor in patients presenting with musculoskeletal complaints is often challenging. The lack of awareness of BJHS may delay the diagnosis as well as effective management. Manual therapy should be used judiciously; active exercise is an essential element of care. We provide an overview of the presentations,*

Contexte : *Le syndrome d'hypermobilité articulaire bénigne (SHAB) est une affection relativement répandue faisant partie des maladies héréditaires du tissu conjonctif, qui se caractérise par des manifestations musculosquelettiques, viscérales et psychologiques. Il faut changer le traitement conservateur des troubles musculosquelettiques pour optimiser son efficacité et réduire le plus possible les séquelles.*

Objectif : *Donner un aperçu des manifestations du SHAB, de l'évaluation des symptômes, des traitements chiropratiques et des résultats obtenus chez des patients atteints du SHAB.*

Méthodologie : *Série de cas*

Discussion *: Il est souvent difficile de déterminer si l'hypermobilité articulaire est un important facteur contributif chez les patients atteints de troubles musculosquelettiques. Le SHAB étant une maladie méconnue, le diagnostic et une prise en charge efficace risquent d'être retardés. La thérapie manuelle doit être utilisée judicieusement; l'exercice actif est une composante essentielle du traitement. Nous présentons un aperçu des manifestations, de l'évaluation des symptômes, des traitements chiropratiques et des*

[1] Private practice
[2] Department of Clinical Education, Canadian Memorial Chiropractic College
[3] Department of Research and Innovation, Canadian Memorial Chiropractic College

Corresponding author: Adele Boudreau, 189 Barrieau Rd. Moncton NB, E1G 1J9
E-mail: aboudreau@pthealth.ca

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assessment, chiropractic management, and outcomes of three patients with BJHS. Future clinical trials are necessary to determine effective clinical management strategies for patients with BJHS.

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KEY WORDS: joint hypermobility, connective tissue disorder, manipulation, chiropractic

résultats obtenus chez trois patients atteints du SHAB. Il faudrait effectuer des essais cliniques pour trouver des stratégies de prise en charge efficaces chez les patients atteints du SHAB.

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MOTS CLÉS : hypermobilité articulaire, trouble du tissu conjonctif, manipulation, chiropratique

Introduction

Hypermobile joints can be a consequence of a number of heritable connective tissue disorders. One such disorder is benign joint hypermobility syndrome (BJHS). BJHS is characterized by generalized ligamentous laxity and the presence of musculoskeletal pain without signs of systemic rheumatologic disease.[1-5] Many experts suggest that BJHS be considered part of a hypermobility spectrum as a milder form of the hypermobility type Hypermobile Ehlers-Danlos Syndrome (hEDS), consisting of hypermobility and arthralgia in several joints.[6] The main distinguishing factors between these two conditions are the scores on the Brighton Criteria as well as laboratory tests.[5,7] The diagnostic Brighton Criteria characterize hypermobility findings into "major" and "minor" categories (Table 1).[7] Within these criteria is the Beighton Score, a scoring system utilized in the diagnosis of a hypermobility syndrome, to quantify the extensiveness

Table 1.
Revised diagnostic criteria for benign joint hypermobility syndrome (BJHS).[7]
BJHS is diagnosed in the presence of two major criteria, or one major and two minor criteria, or four minor criteria.

Major Criteria
1. A Beighton Score of 4/9 or greater (currently or historically)
2. Arthralgia for 3 months in 4 or more joints
Minor Criteria
1. A Beighton score of 1, 2 or 3/9 (0, 1, 2, or 3 if aged 50+)
2. Arthralgia (≥3 months) in 1-3 joints, or back pain ≥3 months, spondylosis, spondylolysis/ spondylolisthesis
3. Dislocation/subluxation in one or more joints or in one joint on more than one occasion
4. Soft tissue rheumatism ≥3 lesions (e.g., epicondylitis, tenosynovitis, bursitis)
5. Marfanoid habitus (tall, slim, span/height ratio >1.03, upper:lower segment ratio <0.89, arachnodactyly [+Steinberg/wrist signs])
6. Abnormal skin: striae, hyperextensibility, thin skin, papyraceous, or scarring
7. Eye signs: drooping eyelids or myopia or antimongoloid slant
8. Varicose veins, hernia/rectal prolapse

of hypermobility in several predetermined articulations (Table 2).[5]

BJHS is a relatively common phenomenon with a prevalence ranging from 5% to 18% in Caucasian populations[8,9] and up to 43% in non-Caucasian populations[10]. Younger individuals express a greater degree of joint laxity, which decreases with age.[10,11] There is an approximately 2:1 female to male relative prevalence of BJHS.[12] Therefore, a small percentage of the population presenting to a chiropractor's office may present with findings suggestive of BJHS. This is important, considering that the majority of chiropractors use spinal manipulation in management of joint pain and function.

The restoration of joint mobility and function are typical goals of chiropractors and other manual therapists. While spinal manipulative therapy is a common therapeutic approach, it may not be appropriate for all patients presenting with hypermobile joints which, in theory, should not require treatments intended to impart increased mobility to articulations. The lack of obvious extra-articular signs can present a diagnostic challenge to a manual therapist, complicating the management of what otherwise may appear to be a straightforward case of mechanical pain. Despite a significant amount of research on BJHS, it remains insufficiently identified, inadequately understood and poorly managed by health practitioners.[13] Therefore, the purpose of our paper is to provide an overview of the presentation, assessment, management and outcomes of three cases of patients presenting with BJHS who sought chiropractic care. The case presentations are discussed in light of the current literature about BJHS.

Table 2.
Nine-point Beighton score for joint hypermobility.[5]

Description	Bilateral Testing	Scoring (maximum points)
Passive dorsiflexion of the fifth metacarpophalangeal joint to ≥ 90 degrees	Yes	2
Passive hyperextension of the elbow ≥ 10 degrees	Yes	2
Passive hyperextension of the knee ≥ 10 degrees	Yes	2
Passive apposition of the thumb to the flexor side of the forearm, while shoulder is flexed 90 degrees, elbow is extended, and hand is pronated	Yes	2
Forward flexion of the trunk, with the knees straight, so that the hand palms rest easily on the floor	No	1
Total		9

Case Presentations

Case 1

A 26-year-old Caucasian female chiropractic student was evaluated for chronic, intermittent low back and left lower extremity pains. She attributed the onset to a fall off a swing during childhood and subsequent aggravation by a motor vehicle collision five years prior to presentation. The progressive constant, dull, aching pain was localized to the left sacroiliac region and radiated distally to the posterolateral aspect of the left thigh; she denied radiation past the knee. The intensity of the pain ranged from 3–10/10 in intensity on a verbal pain rating score. It was aggravated by prolonged sitting, cycling, crossing her legs, fatigue, and positioning for side-posture lumbar manipulations in chiropractic technique class. She obtained some relief by exercising, walking and resting. Three months of chiropractic care had given inconsistent results; she would typically feel sore for hours after manipulation for her low back pain, followed by a short period of relative improvement before the pain would return.

She reported a number of prior musculoskeletal complaints, particularly in her knees, feet and shoulders. Otherwise, she felt she was in good health. She had been a high calibre athlete but since attending chiropractic college her conditioning had decreased significantly.

On examination, no obvious postural deviations were noted. Her lumbar range of motion was mildly painful in forward flexion, reproducing the left thigh pain. All other back ranges of motion were pain-free and appeared significantly greater than normal. External rotation of the left hip was limited to 50% of normal by pain. Straight leg raising was pain-free at 110 - 120 degrees, bilaterally. No neurological deficits were noted. Palpation revealed ten-

derness and decreased mobility at the right C4-5, T5-6, T12-L1, L4-5 and left sacroiliac articulations. Tenderness was palpated in the erector spinae and quadratus lumborum musculature bilaterally, and the left gluteal and tensor fasciae latae musculature. Joint flexibility using the Beighton Score was scored by the chiropractor at 9/9, including findings of mild genu recurvatum, marked hyperextension of both elbows and the ability to passively appose each thumb to the anterior forearm. Lumbar spine radiographs, including AP, lateral and lumbosacral spot views, revealed mild postural changes with a mild left lateral list of the lumbar spine, but were otherwise unremarkable.

The patient was diagnosed with BJHS and left sacroiliac joint dysfunction. The plan of management included ergonomic modification when sitting in class, restriction from involvement as a training partner in technique class, soft tissue therapy to relieve the myofascial component of her complaint, an exercise program directed at improving strength, flexibility and endurance, and limited spinal manipulation. The focus was to limit the passive treatment component and encourage the active component of care. The importance of compliance with the plan of management was impressed upon the patient.

Approximately two months later, the patient reported she was much improved. Seven years later, she reported that she was experiencing occasional low back pain as a consequence of clinical practice, but these episodes were relieved by specific manipulation. She also reported that her symptoms would be aggravated by a lack of physical activity.

Case 2

A 23-year-old Caucasian female presented with left-sided spinal pain, extending from the base of the skull to approximately T8, that had been present for roughly two months. She denied any precipitating event but reported an extensive history of similar complaints. She rated the dull, aching pain as 5/10 in intensity, worse in the evenings, and occasionally present in the mornings. If the pain began in the morning, it would typically persist for the rest of the day. She reported aggravation by sitting and lying supine, and described no relieving factors.

The patient also reported "clicking" in the hips, eye pain with prolonged reading, left jaw pain, and dry skin on the backs of her legs. She reported a past history of a fall when skiing four years prior. No other significant medical history was elicited. She described her life as stressful due to her schooling, which involved a great deal of desk and computer work. She was attempting to improve her diet and had started an aerobic exercise program (three days weekly). She had received no treatment of any kind for her current or previous episodes.

Physical examination revealed a young woman in no significant distress. She was neurologically intact. No cervical bruits were present. Gross range of motion of the cervical spine was decreased in extension-rotation bilaterally, provoking the neck pain of chief complaint. Thoracic range of motion was restricted in extension-rotation and left rotation by pain at the T6 - T9 region. Spinous palpation was extremely tender at T5-6. Tenderness was present in the trapezius and levator scapulae musculature, primarily on the left. Deep inspiration provoked mid-back pain.

The patient scored 8/9 on the Beighton Score. During mobility testing, both elbow joints gave a popping sound upon gentle hyperextension. The patient reported that this was normal for her. No remarkable skin extensibility was observed. No radiographs were taken.

The patient was diagnosed with BJHS, and thoracic facet irritation with myofascial strain secondary to postural strain. Treatment included manipulation directed to the mid-thoracic spine and soft tissue therapy directed at the upper thoracic and cervical musculature. The patient was given a strengthening routine using weight machines and was encouraged to continue aerobic exercise.

She was treated four times over the course of nine days and reported good resolution of her complaint. She had started the prescribed exercise program and reported some mild muscular stiffness. She declined to attend a two-week follow-up appointment because she felt well.

Case 3

A 23-year-old Caucasian female was evaluated for complaint of neck and back pain that started insidiously approximately three years prior. She described the pain as diffuse throughout the upper and lower back. She also reported frequent "cracking" of her joints. Her neck felt stiff and she experienced sharp pain with movements such as rotation and extension. Aggravating factors included carrying bags and lifting heavy objects. She rated the pain in the upper cervical spine at 5-6/10, lower cervical spine

at 5-7/10 and thoracic spine at 8-8.5/10 in intensity. She had no previous chiropractic treatment for this condition. She used muscle relaxants, massage and frequent "self-adjusting" for relief. The patient scored a 21/45 (moderate perceived disability) on the Neck Disability Index.

Past medical history included panic attacks, anxiety attacks and depression. She typically slept a few hours without sleep aid medication and approximately five hours when taking a sleep aid. She rated her current stress level, which disrupted her sleep, as "severe". Her current medications were clonazepam, Adderall XR, Ativan and Wellbutrin.

Physical examination revealed a patient who was 5'11" tall and weighed 120 lbs (i.e., a tall ectomorph), with arachnodactyly. The patient's skin appeared to be slightly more extensible than other patients her age, but did not have a velvety texture. She scored 3/9 on the Beighton Score. However, she actively demonstrated an ability to subluxate her glenohumeral joints bilaterally and bring her heel to her hip posteriorly while in extreme hip internal rotation. Her finger extension and elbow extension were within normal limits; genu recurvatum was not evident. No skin lesions, muscular atrophy or scoliosis were observed. Postural examination revealed the right shoulder was slightly lower than the left.

Cervical spine flexion was mildly increased and caused a pulling sensation in the thoracic spine. Extension was mildly limited initially, recreating the patient's pain, but she was able to proceed to full extension, causing pain in the interscapular region. Bilateral lateral flexion was moderately increased and did not elicit pain. Similarly, right rotation was mildly increased and did not cause pain. Left rotation was within normal limits and recreated the thoracic spine pain of chief complaint. Thoracic active range of motion was within normal limits but extension caused interscapular pain and bilateral rotation caused a pulling sensation. Flexion and bilateral lateral flexion were unremarkable.

Orthopaedic testing[14] revealed left cervical Kemp's test caused a pressure sensation in the cervicothoracic junction and lumbar Kemp's test bilaterally causing low back pain. Left-sided Jackson's test caused the patient's pain of chief complaint in the cervicothoracic junction. Left cervical doorbell test caused ipsilateral interscapular referral but only caused local pain when performed on the right side. The following orthopaedic tests were unremarkable: right-sided cervical spine Kemp's, cervical spine neutral compression, Spurling's, and right-sided Jackson's. Auscultation of the heart did not reveal any abnormal rhythm or sounds.

The patient was diagnosed with a cervicothoracic strain, as well as BJHS according to the revised diagnostic criteria for BJHS (Table 1) given that she had one major criterion (arthralgia for three months in four or more joints) and two minor criteria (Marfanoid habitus; skin hyperextensibility). The plan of management included soft tissue therapy to affected muscles, spinal manipulative therapy to hypomobile segments, and strengthening exercises. The proposed frequency of care was two to three times per week for six weeks; however, due to other health issues the patient only attended 12 of the 16 recommended visits. The patient's presenting complaint had improved but psychological issues had increased. She also suffered from numerous viral illnesses, which limited her ability to attend treatment. Both of these factors limited the success of her treatment.

Discussion

The above case presentations can be encountered by chiropractors and other manual therapists in clinical practice. Each of these female Caucasian patients, all in their twenties, had histories of a variety of musculoskeletal complaints over a number of years. They had orthopaedic signs of mechanical joint pain with no apparent neurological deficits nor overt features of an arthritic or other systemic pathology. In Case 1, the history of lack of long-term benefit from chiropractic manipulation cued the clinician to re-evaluate the patient and treatment plan, resulting in a more specific diagnosis and an effective treatment plan that yielded excellent results. In both Cases 2 and 3, the lack of an apparent etiology and the long history of similar complaints led the clinician to evaluate for hypermobility in the physical examination. However, all patients demonstrated joint laxity according to the revised Brighton Criteria.[7]

The Brighton Criteria categorize hypermobility findings into "major" and "minor" categories (Table 1). The Beighton Score is a scoring system commonly utilized in the diagnosis of a hypermobility syndrome to quantify the extensiveness of hypermobility in several predetermined articulations (Table 2).[5] It is a measure of articular laxity[11] that incorporates a composite score based on

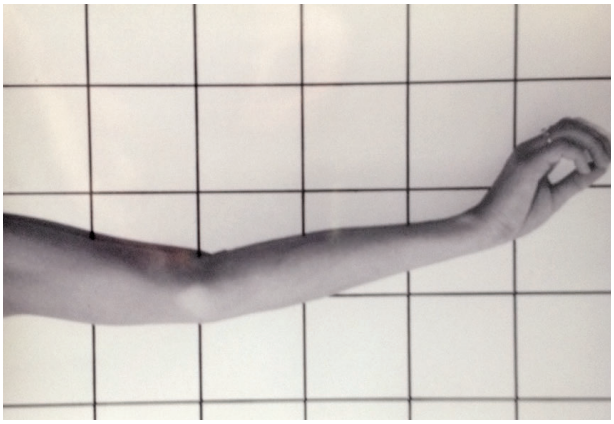

Figure 1. *Elbow hyperextension*

Figure 2. *Finger hyperextension*

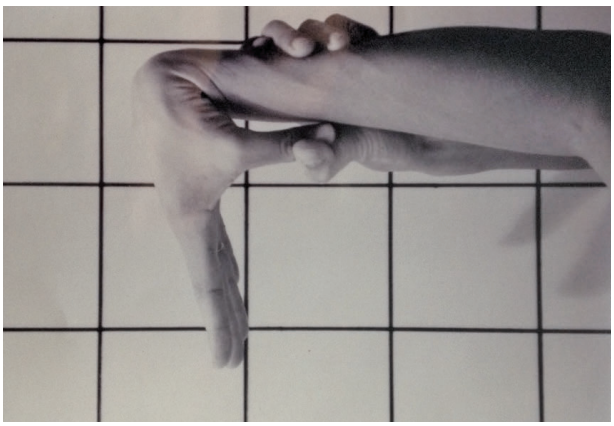

Figure 3. *Thumb hyperextension*

passive elbow hyperextension beyond 10 degrees (Figure 1), passive finger hyperextension (Figure 2), passive thumb apposition to the anterior surface of the forearm (Figure 3), passive knee hyperextension (Figure 4), and the ability to place both palms on the floor while standing (Figure 5) to rate generalized joint laxity. Each of these criteria receives a score of one with a total possible score of nine. A patient is considered to be hypermobile if the score is four or greater. The cut-off of 4/9 is arbitrary but is commonly used in the literature.[11] Some authors have advocated measuring only the non-dominant side (giving a maximum score of five) to avoid joints that may be lax due to an exercise training effect.[15]

Cases 1 and 2 scored 9/9 and 8/9 on the Beighton Score, respectively. Case 3 scored a 3/9 on the Beighton Score, but the patient demonstrated joint laxity in her glenohumeral and femoro-acetabular joints, which are not included as part of the Beighton Score. Case 3 demonstrated one major criterion (arthralgia for three months in four or more joints) and two minor criteria (Marfanoid habitus; skin hyperextensibility), which therefore meets the Brighton Revised Criteria[7] for BJHS (Table 1).

Both the Beighton Score and Brighton Criteria for BJHS have been examined and found to demonstrate good-to-excellent inter-examiner reproducibility.[16] Another measure of joint hypermobility, not applied in this case series, is a

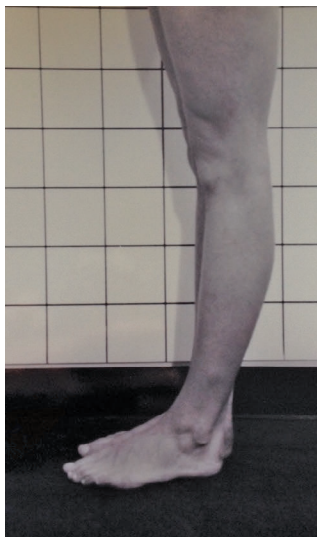

Figure 4.
Knee hyperextension

Figure 5.
Trunk forward flexion

Table 3.
*Clinical signs of Marfan syndrome and Ehlers-Danlos syndrome.**

Syndrome	Clinical signs	
Marfan Syndrome	Arachnodactyly Aortic root dilatation Positive family history	Ectopia lentis Dolichosternomelic habitus Scoliosis, anterior chest deformity Mitral valve prolapse
Classical Ehlers-Danlos Syndrome (cEDS)	Joint hypermobility Bruising/tissue friability	Skin hyperextensibility (velvety texture) "Cigarette paper" scars
Hypermobile EDS (hEDS)	Joint hypermobility	Mild skin hyperextensibility
Vascular EDS (vEDS)	Translucent skin Joint laxity in the hand Bowel and uterine rupture	Prominent bruising Relatively normal large joint mobility Mildly extensible skin
Kyphoscoliotic EDS (kEDS-PLOD1/kEDS-FKBP14)	Ocular fragility	Soft velvety hyperextensible skin
Arthrochalasia EDS (aEDS)	Marked joint laxity	Soft skin Congenital hip dislocation
Periodontal EDS (pEDS)	Easily bruised fragile skin Abundant scarring	Progressive periodontal disease (loss of teeth in second or third decade)

Adapted from: Malfait F, Francomano C, Byers P, Belmont J, et al. The 2017 International classification of the Ehlers-Danlos syndromes. Amer J Med Genetics 2017; 175c (1): 8-26.

five-item questionnaire by Hakim and Grahame (Table 4). This questionnaire can be useful for the clinician to incorporate in their initial history to screen for BJHS. Answering "yes" to two or more of these questions suggests hypermobility with sensitivity of 85% and specificity of 90%.[17]

As in these cases, when generalized hypermobility is combined with myalgia of over three months' duration, the criteria for BJHS have been met (Table 1).[18] If BJHS is considered, then alternative causes of generalized joint laxity should first be ruled out: heritable connective tissue disorders such as Marfan syndrome, Ehlers-Danlos syndrome, Stickler's syndrome, Larsen syndrome and osteogenesis imperfecta.[19] Generalized laxity is a prominent finding in such patients but, unlike those with BJHS, they present with significant cardiovascular, skin, bone and eye abnormalities.[20] Common features of Marfan and Ehlers-Danlos syndromes are listed in Table 3. The skin may be stretchy in BJHS similarly to Ehlers-Danlos syndrome, and can manifest as eyelid laxity or drooping eyelids. However, the skin in patients with BJHS lacks the velvety texture and reduced thickness that is seen in Ehlers-Danlos syndrome.[20]

A recent study[21] reviewed the controversy regarding the association of joint hypermobility and osteoarthritis, and added new data pointing away from such an associ-

Table 4. *Five-point hypermobility questionnaire.*[18]

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?
3. As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
4. As a child or teenager, did your kneecap or shoulder dislocate on more than one occasion?
5. Do you consider yourself "double-jointed"?

Answering yes to 2 or more of these questions suggests hypermobility with sensitivity of 85% and specificity of 90%.

ation. Significant extra-articular manifestations of BJHS[22] include autonomic disturbances[23], skin fragility[24], easy bruising[25,26], ocular ptosis, varicose veins[20], Raynaud's phenomenon[27], urogenital prolapses[28,29], developmental motor co-ordination delay (DCD)[30], alterations in neuromuscular reflex action[31], fibromyalgia[32], carpal and tarsal tunnel syndromes and neuropathies[33,34], lower bone density[20,35], depression[36], anxiety and panic attacks[37], lumbar disc herniations[38], and gastrointestinal symptoms[39]. Case 3 included common extra-articular features of BJHS in the clinical history, reporting anxiety, panic attacks and depression. There appears to be a relationship between joint hypermobility and a higher risk of developing an anxiety disorder.[40] Recent investigations found structural differences between individuals with and without joint hypermobility: joint hypermobility has been associated with a greater bilateral amygdala volume - the key emotion-processing region of the brain.[41] The association likely reflects the genetic basis for BJHS.[42]

BJHS has been demonstrated to be a genetically inherited disorder containing a strong genetic component with an autosomal dominant pattern, believed to affect the encoding of the connective tissue collagen's protein.[24] Nevertheless, there are currently no laboratory tests to diagnose BJHS, as in other systemic disorders[43], only the "Revised diagnostic criteria for BJHS" (Table 1). Many experts now consider the commonalities in symptoms between BJHS and the hypermobility type Hypermobile Ehlers-Danlos Syndrome (hEDS) to be indistinguishable and suggest that BJHS is part of a hypermobility spectrum, as the mildest form of the hypermobility type Ehlers-Danlos.[6]

The primary underlying cause of joint hypermobility is the ligament laxity, which mainly determines the maximum range of motion.[44] Early studies utilizing electron microscopic evaluation of ligaments of patients with BJHS demonstrated reduced thickness of collagen fibrils.[45] It has been also proposed that individuals with BJHS have an abnormal ratio of type III to type I collagen.[2] Type I collagen, containing a high tensile property, is the most common collagen in the body. Type III collagen, located mainly in organs such as the gut, skin and blood vessels, is much more extensible.[46] Furthermore, patients with BJHS demonstrate significantly lower activity of prolidase, an enzyme influential in the collagen structure of ligaments, suggesting an altered collagen metabolism causing joint hypermobility.[47]

Research has also evaluated the symptomatic overlap of fibromyalgia (FM) and BJHS[32,48] in both adults and children[49]. While the underlying mechanism for pain hypersensitivity in FM has been extensively evaluated, there is little knowledge regarding the enhanced sensitization to pain in BJHS.[50] Individuals with FM have an overall lower threshold to pain, attributed to a form of central sensitization or small-fibre polyneuropathy in response to repeated noxious stimulation.[51,52] Conversely, in BJHS, it is hypothesized that pain is a result of repeated microtrauma from abnormal joint hypermobility that contributes to chronic arthralgia.[36] It is also theorized that related structural differences in emotion-processing systems may cause individuals with hypermobility to have a heightened susceptibility to (threat of) pain and/or a perturbation of autonomic control.[41] Interestingly, variations in emotion-processing systems also occur in other pain disorders including FM, irritable bowel syndrome and complex regional pain syndrome.[53] Moreover, women with FM are 44% more likely to be hypermobile.[54] In keeping with the female predominance reported in the literature, all three of our cases were female.

In addition to sex, ethnicity and age are significant factors in the occurrence of BJHS, with increased prevalence among individuals of Asian and African descent.[55,56] In the adolescent and child populations, BJHS tends to occur with juvenile fibromyalgia (JFM).[57-59] Exploring the patient's childhood history of joint hypermobility and repeated injuries may be clinically beneficial in identifying adults with BJHS, as seen in the Hakim and Grahame[17] five-item questionnaire (Table 4). Similarly, if a female patient has a clinical history of FM, evaluating joint hypermobility may be beneficial with respect to clinical management.

The sports injury literature demonstrates no clear relationship between generalized joint laxity and injury.[60,61] It remains unclear if individuals with BJHS are at increased risk of injury compared to their non-hypermobile counterparts. Studies have demonstrated a higher risk of injury in military recruits and ballet dancers who were identified as lax or very lax.[62,63] In contrast, Krivickas and Feinberg[64] found that hypermobile male athletes (Beighton 4-6/9) had a 66% lesser chance of injury than their less mobile colleagues. However, no difference in overall injury rates was observed in NCAA lacrosse players, though hypermobile athletes showed an increased rate of

ankle injuries.[65] More recent reports have found increased injury rates in hypermobile (vs. "tight") rugby players[66], netball players (ankle, knee and finger injuries)[67], and soccer players[68]. Thus, the consensus is building about risk from hypermobility in the athletic population.

The athletic population, in any case, may not be an ideal comparison for the general population for two reasons. One is that strengthening of musculature around the joint may aid the dynamic stability of joints in the trained individual.[64,69] The second is that exercise may improve joint proprioception[70], which appears decreased at specific joint angles in hypermobile individuals when compared to controls[70,71]. Vigorous exercise in relatively deconditioned individuals may lead to injury[72] but may have little effect on injury rates in highly trained individuals, as athletes have enhanced proprioceptive abilities when compared to non-athletes[73].

Consideration of the role of exercise and the potential risks of high impact activity in deconditioned individuals has implications for exercise prescription in patients with BJHS. A graded increase in activity was beneficial in the presented cases and seems appropriate for deconditioned patients with BJHS. Recently, a study by Celenay and Kaya[74] demonstrated that a spinal stabilization program can decrease pain complaints, and improve postural stability and muscle endurance in women with BJHS. An interesting perspective on the role of exercise in the management of hypermobile patients is our first case, in which a highly trained athlete became progressively deconditioned and had an increase in symptomatology. Vigorous activity may be relatively contraindicated in hypermobile individuals[58,62] but this restriction might be lifted once a sufficient training effect has been achieved. Overall, maintenance of physical fitness is imperative for managing symptoms of BJHS, especially activities that are focused on neuromusculoskeletal control; e.g., swimming, Tai Chi, pilates, yoga and dance.[75]

The management of patients with BJHS can be challenging for the patient as well as the practitioner. As described by Simmonds and Keer[75], "patience, coupled with good communication and sensitive handling skills are required as physical problems are often longstanding and include secondary complications and psycho-social issues." There is no conclusive evidence in the literature regarding best practices for patients with BJHS. However, one report highlights the importance of patient education, therapeutic exercise, and modification of work and lifestyle in the management of BJHS[76], reflected in our cases. Patients may also be advised specifically on rest and pacing activities, and have benefited from treatment with modalities including ultrasound and transcutaneous nerve stimulation, taping and splinting, or wearing firm fitting clothing to improve perceived joint stability.[75] Although a recent review suggests that such passive treatment modalities may be ineffective in the management of neck pain and associated disorders[76], it is unclear if such findings are also pertinent to hypermobile patients based on the existing science.

Patients should be provided realistic expectations since their recovery and healing is often slower than in their non-hypermobile counterparts[36] by the time required to improve joint proprioception and strength. In the three cases presented above, the goal of management shifted to the protection of the joints by emphasizing an active exercise program to increase endurance and strength. After seven years, the patient in Case 1 maintained her excellent results as she continued her exercise regime, being able to participate in relatively high impact activities, such as hiking and backpacking.

It may seem paradoxical to apply manipulation, a treatment intended to impart mobility to articulations[77], in patients with BJHS. However, joint dysfunctions were detected and treated with manipulation in the cases described above. The judicial application of high velocity, low amplitude (HVLA) spinal manipulation appears to have benefited these three patients. Currently, only one other study describes a similar successful treatment of a patient with BJHS using a multimodal approach including HVLA spinal manipulations.[76] However, the potential for concomitant decreased pain thresholds in individuals with ligament laxity[41,76] supports our clinical experience that hypermobile individuals seem to report soreness after physical therapies, such as massage or manipulation, more often than non-hypermobile individuals. Modified techniques, including modification of the application of pressure and force, should be considered when treating hypermobile patients after informing them of this possibility.

The management of BJHS is complex and pain management can be difficult in most cases. Prolotherapy is an alternative therapy, not explored in the three cases presented, which can be considered if conservative therapy

has been exhausted. Prolotherapy consists of the injection of growth factors or growth factor stimulators that cause a brief inflammatory response, thus causing increased cellular activity that generates new collagen and extra-cellular matrix.[78] This process increases connective tissue strength and has demonstrated potential to aid symptoms from BJHS.[79] Nonetheless, current research has not fully demonstrated the effectiveness of prolotherapy in BJHS.[80]

A case series cannot draw conclusions about efficacy nor effectiveness of the treatment interventions as presented herein. However, a case series can be hypothesis generating, and future research may illuminate best practice in the management of BJHS patients.

Summary

Recognizing joint hypermobility as a significant contributing factor in patients presenting with musculoskeletal complaints is often challenging for chiropractors who rely on clinical judgement, best evidence and knowledge of physiology to provide effective care. The lack of awareness of BJHS may delay the diagnosis as well as delay effective care, thus exacerbating symptoms.[13] Manual therapy should be used cautiously, but may afford unique benefits in managing pain due to spinal motion segment dysfunction in hypermobile patients. As demonstrated by the cases presented, active exercise is an essential element of care, especially in maintaining joint proprioception. Considering differential diagnoses and quickly reconsidering management in the absence of expected improvement in these cases allowed for a beneficial shift from passive to active care. Simple clinical tests, such as the Brighton Criteria and Beighton Score (Tables 1 and 2) as well as the Hakim and Grahame questionnaire (Table 4), are valid tools that enable the clinician to identify patients with BJHS who will benefit from such a clinical approach. Future research should determine effective clinical management strategies for patients with BJHS, as well as elucidating provocative activities and occupations.

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