

## Joint Hypermobility And Fibromyalgia

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During the last decade, the medical condition known as joint hypermobility syndrome (JHS) has captured the interest of a rising number of researchers and clinicians. It is not just JHS alone that is of interest, however, but its overlap and interrelationship with other maladies which fall under the heading of “soft tissue rheumatism”, including epicondylitis, tenosynovitis, bursitis, and fibromyalgia (FM).

As its name suggests, joint hypermobility occurs when large or small joints in the body are able to extend beyond their normal physiological limits.<sup>1</sup> Most of us can probably remember childhood friends or classmates who were able to effortlessly contort their bodies in seemingly impossible ways. We may have seen ballet dancers, athletes, or musicians who are able to perform incredible feats in their crafts. Some of us may even be aware of joints in our own bodies which hyper-extend, pop out of alignment, or become dislocated. Often though, people are unaware of the condition unless it causes them pain or other problems. Because JHS is inherited, several members of a family can have it and just not realize that they are any different from anyone else.<sup>2</sup>

For many years, JHS was considered a minor rheumatologic malady which occurred in 5-15% of the general population, largely females. JHS seemed to cause mainly biomechanical problems which typically occurred following periods of excessive or repetitive physical activity. However, as noted by Rodney Grahame, Emeritus Professor of Clinical Rheumatology at the University College of London Hospitals and a leading authority on joint hypermobility, a lot has changed since then.

Professor Grahame recently addressed a special symposium on JHS at the 2003 Annual Meeting of the American College of Rheumatology (ACR), the first time the condition

has ever been discussed in a full program by the College. In his presentation, he characterized joint hypermobility syndrome as a problem with connective tissue matrix proteins which in turn is caused by genetic defects--some of them identified, others not. He described JHS as a complex and largely neglected condition that “bridges the adult/juvenile divide in a way that no other disease does” and which is now thought to be associated with a range of neurophysiological defects including:<sup>3</sup>

**Skin Characteristics:** One of the more prominent aspects of JHS, the skin is frequently very “stretchy”, soft, fragile, and sometimes also thin or transparent in quality with a tendency to bruise easily. Scar tissue, when it exists, is often thin and papery. Stretch marks may develop on certain areas of the skin, particularly during the teenage years when the body is growing rapidly,

**Proprioceptive Impairment:** Patients have trouble knowing where certain joints are in space, which in turn causes difficulties with musculoskeletal function and stability.

**Pain Perception And Characteristics:** Studies have suggested that as many as half of JHS patients have enhanced pain perception as well as noticeable pain in multiple sites in the body. Many seem to respond poorly to local anesthetics (i.e., at the dentist or during surgery) and require larger dosages for effective pain control.

**Autonomic Dysfunction:** Typical non-joint related, autonomic symptoms include orthostatic hypotension, light-headedness, fainting, tachycardia, fatigue, and heat intolerance, among other symptoms.<sup>4</sup>

**Anxiety:** For reasons that are not yet clearly understood, JHS is also associated with anxiety, post-traumatic stress disorder, and panic attacks which may be explained by over-stimulation of the sympathetic nervous system’s “fight or flight” response.<sup>5</sup>

**Overlap With Other Disorders:** JHS appears to share certain characteristics with three less common connective tissue disorders: Ehlers Danlos Syndrome, Marfan Syndrome,

and osteogenesis imperfecta, but its manifestations are more benign. For this reason, the syndrome is also sometimes known as benign joint hypermobility syndrome, or BJHS.

Joint hypermobility syndrome is more disabling than originally thought, however. Among its symptoms are chronic pain, fatigue, synovitis, spinal problems, osteoarthritis, and dysautonomia.<sup>6</sup>

## **Diagnosing Joint Hypermobility Syndrome**

Since the late 1960's when serious research on JHS first began, a number of classification systems have been used to diagnose the condition. Because there has been no standardized approach to diagnosis, it has been difficult to compare research studies and to move forward in the understanding of JHS.

One of the more widely used early diagnostic systems for JHS was the Beighton Score. A patient simply got a score (up to nine points) based on whether (s)he could perform five maneuvers (See Table 1).<sup>7</sup>

Although the Beighton tests are easy to perform and are therefore very popular, they have several drawbacks according to Professor Grahame and JHS authority Mary Anne Fitzcharles, Associate Professor of Medicine in the Division of Rheumatology at McGill University in Quebec, Canada. Because it only samples five joints, the Beighton Score can easily miss clinically significant hypermobile joints in other parts of the body.<sup>8</sup> For example, having "flat feet" (normal arches which flatten when weight is put on them) can be a tipoff to the existence of JHS,<sup>9</sup> but foot characteristics are not part of the Beighton test. In addition, it is not yet known whether having large joints (i.e., shoulders hips, knees) which are overly mobile causes a different clinical picture in a patient than having hypermobile joints in smaller joints, as in the hands.<sup>10</sup> Also, the scale makes no allowances for changes in joints which result from aging<sup>11</sup> or from other co-existing disorders which affect mobility.

Recently, a new set of diagnostic criteria were adopted by the British Society for Rheumatology. Known as the Brighton Criteria, they incorporate the earlier Beighton Score but also allow a patient to have other extra-articular complaints (See Table 2).<sup>12</sup>

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### **Table 1: The Beighton Score**

- From a standing position and with knees straight, bend over and place both hands flat on the floor without bending the knees (one point)
- Bend the knee backwards (one point for each knee that can do this)
- Bend the elbow backwards (one point for each elbow)
- Bend the thumb backwards so that it touches the forearm (one point for each thumb)
- Stretch the little finger backwards more than 90 degrees (one point for each little finger)

### **Table 2: The Revised Brighton 1998 Criteria For The Diagnosis Of Benign Joint Hypermobility Syndrome (BJHS)**

#### **Major Criteria**

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

BJHS is diagnosed in the presence of two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first-degree relative.

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## Fibromyalgia And Joint Hypermobility

Increasingly, researchers have found that JHS occurs to a significant degree in a subset of FM patients. At the 1995 Annual Meeting of the ACR, Canadian researchers Hudson, Starr, Esdaile, and Fitzcharles presented data on their controlled study of 393 rheumatology patients, reporting that 50 of them (12.7%) had hypermobility.<sup>13</sup> When comparing hypermobile to control patients, they found that the hypermobile ones were more likely to be female (96% vs. 73% of controls) and slightly younger (mean age 45 vs. 53) and were significantly more likely to have soft tissue rheumatism (67% vs. 25%) and fibromyalgia (30% vs. 8%).<sup>14</sup> The hypermobile patients were significantly less likely to have inflammatory arthritis (4% vs. 32%). Among the hypermobile, 44% had widespread pain; 44% had multiple pain sites; 80% had spinal pain; and 54% had symptoms of thoracic outlet syndrome. The research group went on to publish their study in the *British Journal of Rheumatology* in December 1995.

Two studies reported in 1996 by Spanish researchers Acasuso et al.,<sup>15</sup> and Danish researchers Lykkegaard et al.,<sup>16</sup> also found strongly significant relationships between fibromyalgia and joint hypermobility. Both groups of investigators concluded that joint hypermobility may play a part in the pathogenesis of FM in adults.

Two later studies reported by Connecticut researchers Abeles et al. (1997)<sup>17</sup> and by Turkish researchers Karaaslan et al. (2000)<sup>18</sup> failed to find a significant relationship between FM and JHS. However, it should be noted that the Abeles study used subjects between 40 and 60 years old where age might have had an effect on mobility, and the Karaaslan study included healthy controls composed in part of relatives of the patients studied. Since JHS is highly likely to occur in relatives of patients, this sampling procedure could have affected the results of the study.

A 1993 study published by Israeli researchers Gedalia, Press, Klein, and Buskila,<sup>19</sup> suggests that there is also a strong association between joint hypermobility and

fibromyalgia in children. The researchers studied 338 children (179 boys and 159 girls, aged 9-15 years) in an Israeli public school. Of the 338 children, 43 (13%) were found to have joint hypermobility and 21 (6%) fibromyalgia. Of the 21 with fibromyalgia, 17 (81%) had joint hypermobility, and of the 43 with joint hypermobility, 17 (40%) had fibromyalgia. Statistical analysis confirmed that the two were highly associated.

Professor Fitzcharles reminds us that we still do not know why the pain mechanisms of fibromyalgia show up predominantly in the musculoskeletal (MSK) system, and this is why a link between FM and JHS is of interest. She hypothesizes that because tendons and ligaments are important in joint stability, excessively loose ligaments could cause a person to experience repeated microtrauma during the course of even a normal day. This might be exacerbated by the proprioceptive difficulties experienced by people with JHS. She concludes: “It therefore follows that recurrent microtrauma to ligamentous structures in some hypermobile individuals will lead to repeated pain experience and may trigger disordered pain responses”.<sup>20</sup> Dr. Andrew Holman might also remind us that: “evidence of altered autonomic function, whether primary or secondary, in patients with fibromyalgia is building”.<sup>21</sup>

Clearly, a lot more research needs to be done in this area, particularly now that more comprehensive and definitive diagnostic criteria are available for JHS. However, as Professor Fitzcharles concludes in an editorial in the *Journal of Rheumatology* (2000):

*There is thus increasing evidence that at least a subgroup of patients with soft tissue MSK pain, widespread pain, or FM are hypermobile. Clearly, hypermobility is not the only or the major factor in the development of widespread pain or FM, but rather a contributing mechanism in some individuals. Physical conditioning and regular but not excessive exercise is probably protective towards the development of MSK pain.*<sup>22</sup>

Aside from exercise, the treatment of JHS involves a team approach which includes the use of pain medication as necessary, physical therapy, joint stabilization, podiatric care, and patient education.

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