Living with Joint Hypermobility Syndrome (JHS)

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Upon reflection I should have realised that I was either a little bit different or I had some unusual powers of flexibility. It is perhaps unusual to be able to put ones legs behind one's neck, or to manage to put both hands flat on the floor without warming up first. I remember as a teenager being told that I had "swayback knees" (see Figure 1), but I just got on with my life at the time which was then (and remains) very ballet orientated. Not perhaps a wholly unexpected choice of activity for someone with an above average level of joint mobility; indeed there is a prevalence of up to 70% of ballet dancers who are hypermobile[1].

Figure 1: Hypermobile or 'swayback' knees

Whilst as a dancer I benefit from more aesthetically pleasing extensions[2,3,4] there has been a price to pay for my increased flexibility in the form of the Joint Hypermobility Syndrome (JHS). It is certainly possible to be 'Generalised Hypermobile (GH) in a range of joints and not experience symptoms. The way in which hypermobility is clinically determined involves using The Beighton Score[5] (See Table 1) and hypermobility is confirmed in those who are able to do four or more of the nine movement assessments in the Beighton Score.

Table 1: The Beighton Score

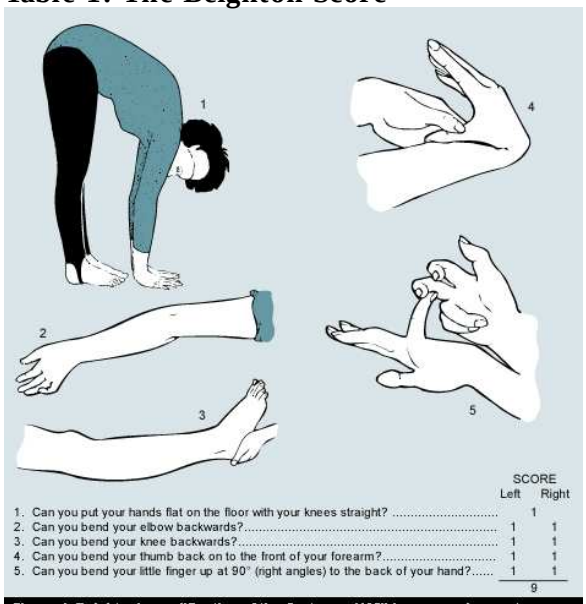
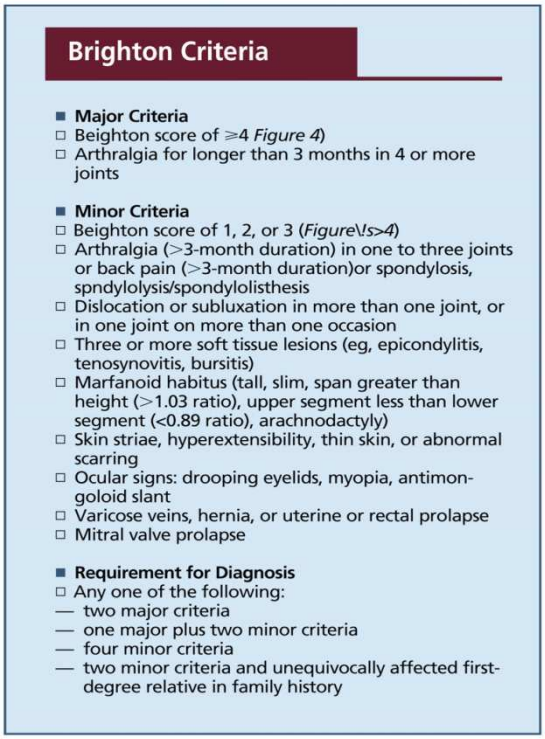


Figure 1. Beighton's modification of the Carter and Wilkinson scoring system. Give youself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points.

Table 2: Brighton Criteria



Brighton Criteria

- **Major Criteria**
  - Beighton score of ⩾4 *Figure 4*)
  - Arthralgia for longer than 3 months in 4 or more joints
- **Minor Criteria**
  - Beighton score of 1, 2, or 3 (*Figure\!s>4*)
  - Arthralgia (>3-month duration) in one to three joints or back pain (>3-month duration)or spondylosis, spndylolysis/spondylolisthesis
  - Dislocation or subluxation in more than one joint, or in one joint on more than one occasion
  - Three or more soft tissue lesions (eg, epicondylitis, tenosynovitis, bursitis)
  - Marfanoid habitus (tall, slim, span greater than height (>1.03 ratio), upper segment less than lower segment (<0.89 ratio), arachnodactyly)
  - Skin striae, hyperextensibility, thin skin, or abnormal scarring
  - Ocular signs: drooping eyelids, myopia, antimongoloid slant
  - Varicose veins, hernia, or uterine or rectal prolapse
  - Mitral valve prolapse
- **Requirement for Diagnosis**
  - Any one of the following:
    - two major criteria
    - one major plus two minor criteria
    - four minor criteria
    - two minor criteria and unequivocally affected first-degree relative in family history

JHS is a multisystemic and symptomatic condition[6] and so additional measures are required in order to captivate the diversity of the condition using the Brighton Criteria, (see Table 2)[7,8,9] Those who ultimately have JHS will score positively on the Brighton Criteria, whilst those who are only GH are asymptomatic. It is important to note that a diagnosis of JHS is ultimately made by a Consultant

Rheumatologist, although an experienced physiotherapist or GP might recognise the signs and symptoms. However, according to recent research only one in 20 are correctly diagnosed[10] and this might be because of the deceptive appearance of general good health that is exhibited by most patients with JHS – most don't look unwell[11].

The overriding symptom and complaint from apparently 'healthy' patients with JHS is pain[12]. My own experience of pain is reflected in years of chronic lower back pain that started when I was 18 years old. There was no particular trauma that initiated the pain, but eventually the pain became chronic and remained constantly. Even though an MRI scan showed 'degenerative disc changes and a posterior disc bulge' Experts are more certain that my pain has been caused by hinging at the lumbar spine and overuse of a particularly hypermobile section of my spine. In addition, and like so many other JHS patients I get pain and trauma in other joint areas and muscle pain where muscles fatigue unnecessarily quickly because they cannot cope with protecting an abnormal range of movement[13] at the various joints I have which are hypermobile.

In order to understand why pain is so often the resulting symptom in patients with JHS it is important to understand what is going on with the tissue fibres of the body which cause hypermobility. The body is made up of different tissue fibres of which one type of protein is called collagen. The body is covered in collagen fibres and it is thought that it is these collagen fibres which might be faulty or abnormal in those with JHS[14]. Hence JHS might be called a Connective Tissue Disorder which is related to two other more serious connective tissue disorders called Ehler-Danlos Syndrome and Marfan Syndrome[15]. In patients with those more serious conditions the heart can also be affected and expert clinical management is required.

If it is the collagen fibres which are causing problems, it is crucial to understand that these are not just affecting the joint areas, but the whole body systemically including the lungs and gastrointestinal tract, for example. Once one understands this it becomes easier to explain why JHS can indeed be related to other conditions. Research has now shown links with JHS asthma and JHS and gastrointestinal conditions such as IBS[16]. This is because of the way in which the collagens in the lungs and gut might be 'overstretched' in those with JHS[17].

JHS has been shown to be a heritable connective tissue disorder and so it is extremely likely that other first degree relatives will also be hypermobile. Rheumatologists are very interested in this aspect of it and will ask about family history and medical history. The genetic research about JHS is also beginning to show links with other medical conditions in addition to musculoskeletal problems. For example, research in Spain has shown a link with JHS and anxiety with JHS patients up to 16 times more likely to experience anxiety and panic disorders[18],[19].[20] It is interesting to speculate why this might be the case but from personal experience I would suggest that it is perhaps because the body in someone with JHS often feels out of control and is in a chaotic range of motion because of systemic joint instability. Other experts also suggest that the anxiety might come from a response to fear from the pain that these unstable joints cause[21]. Either way the evidence is building in terms of JHS and its genetic links to other conditions.

Recent research has also begun to show overlaps with JHS and learning disorders, in particular with Developmental Coordination Disorder (formerly known as Dyspraxia). Kirby and Davies (2007) have begun to show an overlap with JHS and DCD and research in progress[22] involving dancers is showing how concentration and focus might be impaired in hypermobile dancers – perhaps linked with DCD[23]. Again, in my personal experience, I am

aware of coordination problems, the fact I was late learning to walk and had great difficulties in sport at school all certainly relate to DCD and symptoms caused by the JHS. I still experience problems in memorising sequences in dance classes and in learning and coordinating new movements. Perhaps the emergent research by Kirby and Davies might explain why this is the case. Another reason why patients with JHS might experience some movement difficulties might also relate to difficulties with proprioception.

Proprioception is about knowing, understanding and sensing exactly where a joint is in space. It is governed by the Golgi tendons and joint proprioceptor senses and in patients with JHS proprioception is frequently impaired. One way in which to test proprioception and balance involves the 'Stalk Test' (Figure 2) or balancing on one leg with the eyes shut. The amount of wobble experienced informs both balance and how good the person's proprioceptive system is. Those with JHS are often more unstable and have a poorer sense of the location of their joints in space which is why they often stand in poses at the end range of their movement[24] (Figure 3). Other sensory feedback can also be affected- for example I often have little or no sensation in my right hip and hamstrings and emergent research suggests this might be related to reflex arcs and the nervous system and the way the body interprets sensation and pain[25].

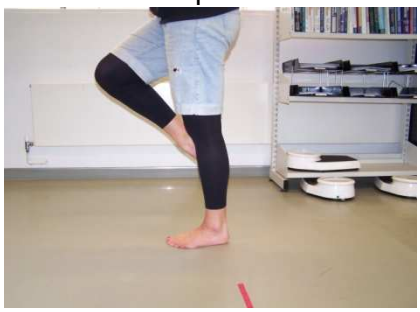


Figure 2: The Stalk Test

Figure 3: Standing in hyperextension

There is plenty of evidence to suggest the relationship with JHS and chronic pain[26],[27]. Pain becomes chronic when (in simple terms) the pain receptors remain on and constantly alerting the body that there is pain, although there is no actual danger associated with this pain, hence the nervous system develops a fault. Pain might become chronic in JHS patients owing to the fact the same areas are repeatedly overused. I know that my back pain becomes worsened when I am hinging into the joint and not using postural muscles to protect the area. However it might not be due to personal laziness that my muscles do not show great endurance. Fatigue and poor muscle endurance are another aspect of JHS[28].

There are times when I feel completely exhausted, disproportionate to activity levels, or that I feel "run-over". Part of the reason for this overwhelming fatigue that is often a symptom in JHS patients might be owing to the fact that the muscles in a person with JHS are working at least twice as hard as in a non-hypermobile person because they have to work to sustain an abnormal range of movement (ROM). Patients with JHS are often observed fidgeting[29] and this might also relate to poor muscular endurance and weakened deep postural muscles and poor core stability.

Part of the medical management of JHS involves working on strengthening and developing postural control in order to manage this extra ROM. I personally have to do daily strengthening exercises in order to manage my joint instability. I have been working for 18 months with a JHS expert physiotherapist who has supported me with an exercise regime which has for the first time in my life given me some

core stability and helped me to manage my joint laxity owing to the JHS. In addition I have been attending pilates sessions on an almost one to one basis and pilates has particularly revolutionised how I manage my spinal hypermobility and activate some of the deeper postural muscles. As I now see it, my health and wellbeing is now dependent upon my continuing with regular pilates and physiotherapy sessions.

Part of the problem with many JHS patients is that they de-condition and lose muscle tone rapidly if they stop any form of exercise. Most often patients with JHS stop exercising because of pain. In 1999 I gave up ballet and all exercise completely because of back pain and ended up making my JHS symptoms far worse. I gained over 20 kilos in weight, I had more pain, so I moved less and then I stopped working for a while, I then became severely depressed. Finally, I attended a pain management course and found out that pain does not equal damage, I was given permission to re-start ballet again. It was very hard to start ballet at such a low point remembering what my body could do in the past. Not long after I restarted ballet I partially tore my right calf muscle. Soft tissue trauma is another adjunct of JHS[30]. In this instance the injury actually did me a favour. I found a physiotherapist who treated me seriously – I had been dismissed by many in the past as they said my back "looked horrendous". The physiotherapist who I found (and is still working with me, to her credit) not only made a diagnosis of JHS, later confirmed by a consultant rheumatologist and leading expert in JHS, and has continued to help me to rebuild my body. The process has been difficult, a great challenge and hard work. However as those who know me intimately, giving up is not in my nature and my perseverance is now paying off. I have shed those extra 20 kilos, I have increasingly functional gluteals, hamstrings and adductor muscles and developing core stability and deep abdominal strength. My back hurts much less and the ROM is now much better managed. Although I still hang into my knees, I am developing an improved sense of proprioception and if I concentrate very hard and maintain my hamstrings (which incidentally I am unable to feel), I avoid going into the extremities of my hyperextension.

So what does the future hold? Apparently as one ages, the symptoms relating to JHS gradually diminish[31], but I will retain my flexibility which will make me appear fitter compared to my non-hypermobile contemporaries. JHS is a condition that is predominant in females and more common in the African and Asian communities compared to Caucasians. In addition some females with JHS also suffer complications at the hands of their chemistry and the female hormone progesterone can also increase tissue laxity making some of the JHS pain and symptoms worsen just before and during menstruation[32]. For some women changing their contraceptive medication towards an oestrogen based contraception can improve their symptoms sufficiently. The hormone relaxin responsible for relaxing the pelvic ligaments during childbirth can similarly cause difficulties for women with JHS.

JHS was originally described as a Benign condition[33], and although the condition may not kill me, there is no cure to transform the faulty collagen fibres that myself and other JHS patients have inherited. JHS can also affect the bony sockets and some JHS patients have more shallow hip and shoulder sockets leading to a greater ROM[34]. The key to JHS as I see it is in terms of self-management and having a team of medical professionals who can offer support when symptoms flare up or become problematic. Sometimes I find my 30-minute session with my physiotherapist to be similar to workplace supervision and it is a chance for me to discuss symptoms, or to reflect upon progress gleaned through my exercise regime. There is no doubt that a good sense of humour helps with this condition as it is just too easy sometimes to collapse into one's joints and other interesting postures (see Figure 4).

Figure 4: JHS patient in an interesting pose

Finding medical professionals with experience of the condition will also help. The Hypermobility Syndrome Association (HMSA) is also invaluable and has a very helpful website and online Forum community. As far as I am concerned I hope to manage ballet well into my dotage and if I keep matching my flexibility to strength ratio via physiotherapy and pilates and pace myself during more difficult times, I hope to manage this.

I cannot conclude this article without mentioning how invaluable The Bowen Technique has been in helping with my back pain and general aches and pains and poor energy levels (see Figure 5). The Bowen Technique is a gentle soft-tissue therapy founded in Australia by the Late Tom Bowen. It involves a series of gentle rolling type moves across muscle and tendon fibres which generate an integrated healing response. Moves are done in a series with short breaks in between each set in order to give the body a chance to rest and to respond to the work. Bowen therapy had such an impact upon me and my pain that in 2002 I trained as a Bowen Therapist. I am now personally interested in conducting research into its efficacy in treating patients with JHS. Further information can be found on my website www.bowenworks.org

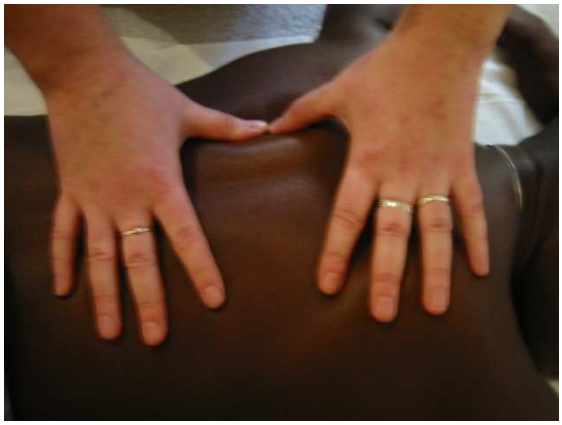


Figure 5: The Bowen Technique (Upper back)

For further information about hypermobility please visit the HMSA website www.hypermobility.org and my blog which has been documenting my progress in physiotherapy, pilates and ballet over the past 18 months and also mentions Bowen Technique.
http://danceinjuryrecovery.blogspot.com/

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