Chronic pain in patients with hypermobility

Joint hypermobility syndrome (JHS) is a ‘connective tissue disorder with hypermobility in which musculoskeletal symptoms occur in the absence of systemic rheumatologic disease’ (Simpson 2005). JHS occurs in 4 to 13% of the general population (Seckin et. al. 2005). Women have greater joint laxity than men and up to 5% of healthy women have symptomatic joint hypermobility compared to 0.6% of men (Engelbert et. al. 2004). The Brighton Criteria and is a valid tool that can be used clinically and for research to identify the condition (Simmonds & Keer 2011) (Appendix 1).

Patients with hypermobility can present with a wide variety of conditions, such as traction injuries at tendon or ligament insertions, joint or tendon sheath synovitis, chondromalacia patella, rotator cuff lesions, or back pain due to soft tissue injury or disc prolapse. Others may suffer joint instability (chronic dislocation or subluxation), stress fractures to metatarsal bones or vertebral bodies, or develop chronic arthritis (Grahame 1990).

JHS is characterised by an increase in laxity and fragility of connective tissues. The pathophysiology of hypermobility is not fully understood but appears to be a systemic collagen abnormality (Acasuso-Diaz & Collantez-Esteves 1998). Tissues such as tendon, ligament, bone, cartilage, and skin, which rely on the considerable tensile strength of their collagen component for their physical integrity, are more likely to fail mechanically. Therefore hypermobile persons have a general predisposition to soft tissue injury, coupled with an impairment to healing which may not only be slow but also may be incomplete (Grahame 2009). Pain in a hypermobile patient can occur in any joint but most commonly presents in the load-bearing joints of knees or ankles and consequently pain often presents later in the day and morning stiffness is uncommon (Simpson 2005).

Fibromyalgia commonly coexists with hypermobility and is 3.8 times more likely to occur in adults with hypermobility than adults without (Russek 2000). Acasuso-Diaz & Collantez-Esteves (1998) showed an association between joint hypermobility and fibromyalgia, proposing that excessive muscle stress may lead to an increase in the excitability of nociceptive nerve endings of the muscle and lead to a chronic pain condition. Joint instability in patients with hypermobility may lead to excessive muscle stress and thus cause this pain.

In hypermobile patients, chronic pain is often aggravated by body movement which leads to the patient adopting a strategy of movement avoidance as a means of pain avoidance (kinesiophobia), thus leading to muscle deconditioning which may in turn worsen the condition (Grahame 2009).

Long-term management of hypermobility involves a modification of activities, particularly those that induce symptoms such as excessive joint movement, vigorous and repetitive activities, over training, poor pacing, and a focus on increasing joint flexibility rather than joint stability (Simpson 2005).

Physical rehabilitation is necessary to address factors of joint laxity and instability, muscle weakness, and a lack of proprioception (Grahame 2009). Management strategies include: developing core support, increasing muscular tone, proprioception enhancement, focusing on primary area(s) of dysfunction, joint stabilisation, posture re-education, joint awareness, functional stability and endurance. Functional re-training aims to target control of the neutral joint position, re-training dynamic joint control in the direction of stability dysfunction and rehabilitating global stabilisers (Kemp et al 2010, Simpson 2005, Grahame 2009).  The use of mobilising techniques to restore natural mobility to joints or spinal segments where these have been lost as a result of deconditioning and kinesiophobia may also play a beneficial role in the management of JHS (Simmonds & Keer 2001 cited in Ross 2011).

Treating pain in those who are hypermobile is challenging due to a lack of evidence-based studies on the effectiveness of specific treatments. Physiotherapy is considered the most successful strategy involving strengthening, proprioception exercises, and gentle stretching (Castori et al 2012).

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**Appendix 1. The Brighton score and the Beighton score**

**The Brighton score for JHS**

**Major criteria**

* A Beighton score of 4/9 or more (either current or historic)
* [Arthralgia](http://en.wikipedia.org/wiki/Arthralgia) for more than three months in four or more joints

**Minor criteria**

* A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
* Arthralgia (> 3 months) in one to three joints or back pain (> 3 months), spondylosis, spondylolysis/spondylolisthesis.
* Dislocation/subluxation in more than one joint, or in one joint on more than one occasion.
* Soft tissue [rheumatism](http://en.wikipedia.org/wiki/Rheumatism). > 3 lesions (e.g. epicondylitis, [tenosynovitis](http://en.wikipedia.org/wiki/Tenosynovitis%22%20%5Co%20%22Tenosynovitis), bursitis).
* [Marfanoid](http://en.wikipedia.org/wiki/Marfanoid) habitus (tall, slim, span/height ratio >1.03, upper: lower segment ratio less than 0.89, arachnodactyly (positive Steinberg thumb / Walker wrist signs).
* Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring.
* Eye signs: drooping eyelids, [myopia](http://en.wikipedia.org/wiki/Myopia) or antimongoloid slant (Palpebral slant)
* [Varicose veins](http://en.wikipedia.org/wiki/Varicose_veins) or [hernia](http://en.wikipedia.org/wiki/Hernia) or uterine/[rectal prolapse](http://en.wikipedia.org/wiki/Rectal_prolapse).

**Beighton score for hyperlaxity**

1. Dorsiflexion of the right second finger to 90’
2. Dorsiflexion of the left second finger to 90’
3. Apposition of right thumb to volar aspect of forearm
4. Apposition of left thumb to volar aspect of forearm
5. Hyperextension of the right elbow by 10’
6. Hyperextension of the left elbow by 10’
7. Hyperextension of right knee by 10’
8. Hyperextension of left knee by 10’
9. Hands flat on the floor with knees extended

4+ criteria fulfilled = hyperlaxity

(<http://www.hypermobility.org/diagnosis.php> Date accessed: 20/02/13)