

Masterclass

Hypermobility and the hypermobility syndrome

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Received 5 March 2007; received in revised form 6 March 2007; accepted 12 May 2007

Abstract

Hypermobility joints by definition display a range of movement that is considered excessive, taking into consideration the age, gender and ethnic background of the individual. Joint hypermobility, when associated with symptoms is termed the joint hypermobility syndrome or hypermobility syndrome (JHS). JHS is an under recognised and poorly managed multi-systemic, hereditary connective tissue disorder, often resulting in a great deal of pain and suffering. The condition is more prevalent in females, with symptoms frequently commencing in childhood and continuing on into adult life.

This paper provides an overview of JHS and suggested clinical guidelines for both the identification and management of the condition, based on research evidence and clinical experience. The Brighton Criteria and a simple 5-point questionnaire developed by Hakim and Grahame, are both valid tools that can be used clinically and for research to identify the condition. Management of JHS frequently includes; education and lifestyle advice, behaviour modification, manual therapy, taping and bracing, electrotherapy, exercise prescription, functional rehabilitation and collaborative working with a range of medical, health and fitness professionals. Progress is often slow and hampered by physical and emotional setbacks. However with a carefully considered management strategy, amelioration of symptoms and independent functional fitness can be achieved.

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Keywords: Hypermobility; Hypermobility Syndrome; Brighton Criteria; Assessment; Management

1. Introduction

Joint hypermobility is defined as a condition in which most of an individual's synovial joints move beyond the normal limits taking into consideration the age, gender and ethnic background of the individual (Grahame, 2003a). Hypermobility may be inherited (Child, 1986; Beighton et al., 1989a), or acquired through years of training and stretching, as seen in ballet dancers and gymnasts (Grahame, 2003a). Furthermore, hypermobility may also develop as a result of changes in connective

tissue in a number of other diseases (Beighton et al., 1989b). Where once it was common for health and exercise professionals to view hypermobility as 'the upper end of a Gaussian distribution of the normal joint range of movement'; we now understand that, in fact, it represents a departure from normality (Grahame, 1999). Hypermobility may pose no problems, but in some individuals it predisposes to a wide variety of soft tissue injuries and internal joint derangements, arthritis, arthralgias or myalgias, which lead sufferers to seek medical attention (Grahame, 1990; Cherpel and Marks, 1999; Dolan et al., 2003). Joint hypermobility, when associated with symptoms is termed the joint hypermobility syndrome or hypermobility syndrome (JHS), (Grahame, 2003a). JHS is one of the well-defined polygenic heritable connective tissue disorders (Zweers et al., 2004) which presents with a recognisable

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Fig. 1. “Saint Cyriaque” Matthias Grunewald (1460–1528): demonstrates hyperextension of the metacarpal phalangeal and interphalangeal joints.

phenotype, described and discussed in more detail later in this paper.

Hypermobility is not new in the arts and medical humanities. Hippocrates in the 4th century BC is said to have speculated that the Scythians were defeated in India because the hyperlaxity of their shoulder and elbow joints prevented them from drawing a bow or hurling a javelin effectively (Beighton et al., 1989c; Larsson et al., 1993a). Notably, artists Matthias Grunewald (1460–1528) observed hypermobility in “Saint Cyriaque” in the Heller Retable (Fig. 1), and later, Peter Rubens observed hyperextension of the metacarpal joints, flat footedness and hyperlordosis in “The Three Graces” (1638–1640), Prado, Madrid (Dequecker, 2001). The musical successes of Paganini were attributed to his extreme hand mobility in the 18th century (Larsson et al., 1993b; Cherpel and Marks, 1999). However, by the end of the 19th century, rather than being characterised as an oddity, impediment or asset, joint hypermobility was recognised as having considerable clinical significance (Grahame, 1971). Into the 20th century, clinical observation and research led to the specific recognition of the JHS by Kirk et al., 1967, who described the occurrence of ‘musculo-skeletal symptoms in the presence of generalised joint laxity in otherwise normal subjects’.

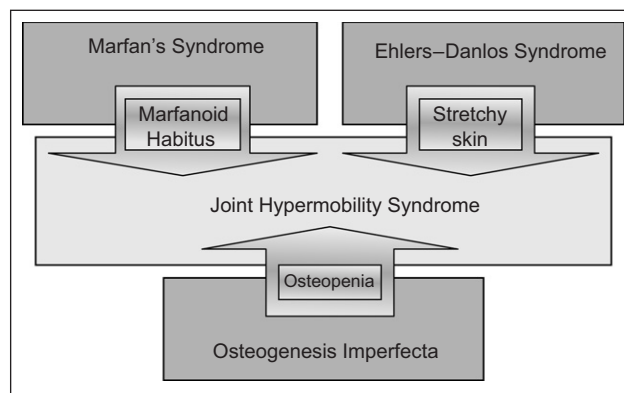


Fig. 2. Inter-relationship between the heritable connective tissue disorders. Adapted from Grahame (2003b).

Now in the 21st century, we know much more about JHS. Far from individuals being ‘otherwise normal or healthy subjects’, the condition is beginning to be taken more seriously. JHS has now been classified as a hereditary connective tissue disorder (HCTD) sharing the less severe features with its’ more serious counterparts; Marfan’s Syndrome, Ehlers–Danlos Syndrome and Osteogenesis Imperfecta, as illustrated by Grahame (2003a) (Fig. 2). JHS is the commonest of all the HCTDs and one that is seen most frequently in clinical practice (Grahame, 2003a). Many authorities consider JHS to be synonymous with Ehlers–Danlos hypermobility type, formerly known as Ehlers–Danlos type III (Grahame, 2001).

Despite the substantial volume of published literature JHS continues to be under-recognised, poorly understood and inadequately managed by the medical and physiotherapy professions (Gurley-Greene, 2001; Keer, 2003). This paper aims to provide the reader with an overview of JHS including the epidemiology, pathogenesis, presentation and suggests guidelines for assessment and management.

1.1. Epidemiology and demographics

The reported prevalence and incidence of hypermobility and JHS varies in the literature reviewed. This is largely due to the use of varying screening and diagnostic criteria. We know that gender, ethnicity and age are important factors, with hypermobility being more prevalent in females and those of African or Asian descent when compared with their Caucasian counterparts (Cherpel and Marks, 1999; Russek, 1999). We also know that it decreases with age (Bridges, 1992; Larsson et al., 1993a; Russek, 1999). The prevalence of hypermobility in children has been estimated to be 10–25% (Biro et al., 1983; Ümit et al., 2005) with a higher incidence in girls than boys (Larsson et al., 1987; Qvindelnd and Jonsson, 1999). The prevalence of



hypermobility in adults also varies, from 5% in the USA (Jessee et al., 1980) to between 25% and 38% in Iraq (Al-Rawi et al., 1985) and 43% being recorded in the Noruba tribe in Nigeria (Birrell et al., 1994).

Hakim and Grahame (personal communication, 2004) investigated referral data from a West London Rheumatology clinic over a six-month period in 2003, showing JHS to be more prevalent amongst non-Caucasian females in this discrete population. Within the female, non-Caucasian group, the JHS phenotype was present in 58% of the sample and in the same male cohort, 29% of individuals exhibited the JHS phenotype.

Similarly, in a cross-sectional, matched, controlled study of female patients aged between 18 and 50 years in multicultural Oman, 55% of female patients attending the rehabilitation outpatient department exhibited the JHS phenotype (Clarke and Simmonds, 2007).

1.2. Pathogenesis

The JHS is a genetically inherited disorder, presenting with an autosomal dominant pattern, thought to affect the encoding of the connective tissue proteins collagen (Grahame, 2003b). It has been proposed that individuals with JHS display an abnormal ratio of type III to type I collagen (Child, 1986). Type I collagen has a high tensile strength and is the most common collagen in the body, abundant in tendon, joint capsule, skin, demineralised bone and nerve receptors. Type II collagen is found in cartilage, and designed to withstand compressive stress, whereas type III collagen is much more extensible and disorganised, occurring in organs such as the gut, skin and blood vessels (Beighton et al., 1989d) which may explain the inherent laxity or 'reduced tissue stiffness' (Russek, 1999). Mutations in genes encoding collagen type V have also recently been implicated (Malfait et al., 2005), with type V collagen under normal control interacting with type I collagen during fibrillogenesis and having a role in regulation of fibril diameter. An alteration in this process may potentially lead to thinner, fine and more disorganised collagen fibres. Skin fibroblast biopsy analysis, has allowed researchers to further investigate the microscopic structural discrepancies that may define HCTDs. Malfait et al. (2005) hypothesise that it is the interference with the processing of the N-propeptide of either α -chain ($\alpha 1$ or $\alpha 2$) of type I collagen that is responsible for Ehlers–Danlos-like symptoms of skin laxity, joint subluxation and dislocation.

The nervous system is affected in individuals with JHS. Lack of efficacy of local anaesthetics when injected or topically applied, has been reported by Arendt-Nielsen et al. (1990). The mechanism for this is unknown. Studies have also shown that individuals with JHS are less accurate than individuals without the

condition at reproducing proximal interphalangeal joint angles (Mallik et al., 1994). Research also shows that position sense at the knee is decreased, particularly the ability to locate end-range extension (Hall et al., 1995). Laxity and fragility of connective tissue coupled with a decreased proprioceptive acuity and altered neuromuscular reflexes are the possible causes of the predisposition of individuals with JHS to damage and injury (Johansson et al., 2000; Stillman et al., 2002).

1.3. Clinical presentation

Hypermobility does not necessarily result in problems and may sometimes be considered an asset (Grahame, 2003a). However, for those less fortunate, hypermobility and tissue laxity can be the cause of a variety of debilitating symptoms.

Symptoms frequently commence in childhood with the potential to continue into adult life (Grahame, 2001). One study (Kirk et al., 1967) reported three quarters of hypermobile adolescents developing symptoms by the age of 15 and Lewkonja and Ansell (1983) and Murray and Woo (2001) recognise JHS as one of the most frequent causes of musculo-skeletal symptoms in children and adolescents, particularly girls, aged between 13 and 19 years of age.

The predominant presenting complaint is pain, which is often widespread and longstanding, with patients reporting pain ranging from 15 days to 45 years (El-Shahaly and El-Sherif, 1991). In addition there are many other symptoms reported by patients associated with the joints, such as, stiffness, 'feeling like a 90 year old', clicking, clunking, popping, subluxations, dislocations, instability, feeling that joints are 'vulnerable' as well as symptoms affecting other tissues such as paraesthesiae, tiredness, faintness, feeling unwell and suffering flu-like symptoms (Keer, 2003). Fig. 3 illustrates a typical patient pain chart. Complaints are sometimes difficult to match with the way the patient looks or moves (Russek, 2000) as individuals frequently look well and move well. This infrequently leads to the patient being misunderstood and at worst the patient is made to feel like a hypochondriac and may be labelled as having psychological problems (Child, 1986).

Extra articular manifestations of the syndrome may include skin fragility and laxity (Grahame, 1999, 2003a), autonomic disturbances (Gazit et al., 2003; Hakim and Grahame, 2004), ocular ptosis, varicose veins (Mishra et al., 1996), bruising (Bridges, 1992; Kaplinsky et al., 1998), urogenital prolapses (Al-Rawi and Al-Rawi, 1982; El-Shahaly and El-Sherif, 1991), Raynaud's phenomenon (El-Garf et al., 1998), development motor co-ordination delay (DCD) (Kirby and Sugden, 2007), alterations in neuromuscular reflex action (Johansson et al., 2000; Stillman et al., 2002), neuropathies, tarsal



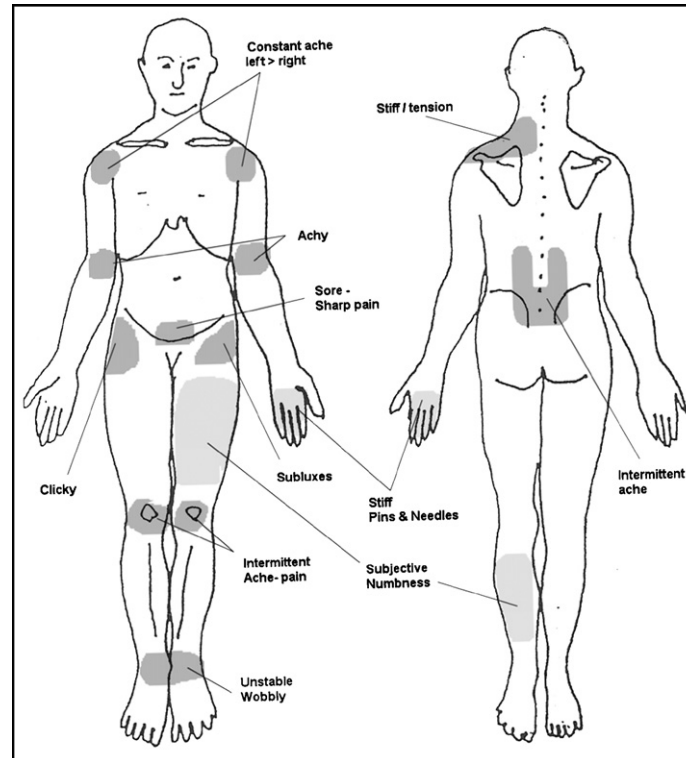


Fig. 3. Typical JHS body chart presentation.

and carpal tunnel syndrome (Francis et al., 1987; March et al., 1988), fibromyalgia (Acasuso-Diaz and Collantes-Estevez, 1998), low bone density (Mishra et al., 1996, Gulbahar et al., 2006), anxiety and panic states (Bulbena et al., 1993) and depression (Grahame, 2000).

2. Patient assessment

Recognising generalised hypermobility as a contributory factor to musculo-skeletal complaints is often difficult, frequently overlooked, or not considered. This occurs for a variety of reasons; lack of knowledge or experience, focusing exclusively on the problem area rather than looking at the patient as a whole and failing to recognise that a 'normal' range of movement may not be 'normal' for the hypermobile patient. Additionally, it may also be difficult to identify JHS as a contributory factor to a patient's complaints of pain and dysfunction in older patients, and in those who have stiffened significantly in response to aging, disuse and pain, as they may have lost their previous flexibility.

The following discussion of characteristics may assist the therapist to recognise and diagnose JHS when examining a patient.

2.1. Subjective examination

The onset of symptoms is frequently associated with trauma, pregnancy, childbirth, unresolved previous joint

problem (s), or de-conditioning related to a sedentary lifestyle. Problems in childhood can be a useful clue to the presence of hypermobility with many hypermobile individuals reporting joint pains, particularly in the back and knees in childhood and there is often a history of growing pains or benign paroxysmal nocturnal leg pain (Maillard and Murray, 2003). A history of participation in activities such as ballet and gymnastics where inherent flexibility is considered an asset may be an indication of hypermobility.

Past history of soft tissue injuries, joint pain, fractures, dislocation and subluxations, particularly if they occur with minimal provocation and have been slow to resolve, may be a good indicator of hypermobility and may also provide valuable information about tissue healing rate, which has been reported as slower in hypermobile individuals (Russek, 2000).

As JHS is an inherited disorder, exploring the family history may also assist with its recognition. Several authors (Biro et al., 1983, Finsterbush and Pogrud, 1982) have reported that between 27% and 65% of their patients had relatives with a history of joint hypermobility. Even if it is not known that relatives were hypermobile, other complaints such as arthritis, multiple joint problems and dislocations may provide further clues and point to a possible case of inherited hypermobility.

Exploratory questions about other areas of the body and body systems may reveal a host of other problems



Box 1**5 point hypermobility questionnaire (Hakim and Grahame 2003).**

- o Can you now [or could you ever] place your hands flat on the floor without bending your knees?
- o Can you now [or could you ever] bend your thumb to touch your forearm?
- o As a child, did you amuse your friends by contorting your body into strange shapes or could you do the splits?
- o As a child or teenager, did your kneecap or shoulder dislocate on more than one occasion?
- o Do you consider yourself “double-jointed”?

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

for the individual with JHS. Patients are often reluctant to reveal this information, as they may not consider these issues to be related to their presenting musculoskeletal complaint(s). These systemic signs and symptoms may include urogenital problems (prolapse, incontinence), vascular problems (bruising, varicose veins, low blood pressure), neural problems (clumsiness, unsteadiness, paraesthesiae, neuropathies). Furthermore, poor response to local anaesthetics has also been associated with JHS (Arendt-Nielsen et al., 1990) and may result in significant distress for individuals when not recognised or believed, for example, by a dentist when carrying out dental work or an obstetrician during childbirth.

Finally, the inclusion of a simple five-part questionnaire devised by Hakim and Grahame (2003) can easily be incorporated into the assessment of most patients. The questionnaire has been shown to have good sensitivity and specificity and correctly identified hypermobility in 84% of a group of subjects. The questionnaire is outlined in Box 1.

2.2. Objective examination

2.2.1. Observation

Observing sitting position during the subjective part of the examination may give a clue to the presence of hypermobility. Individuals with JHS frequently fidget and adopt end of range postures such as entwining their legs, sitting rotated and twisting in the seat, or side sitting (Oliver, 2000). When sitting unsupported, hypermobile individuals frequently slouch, and rest in posterior pelvic tilt position. We speculate that these postures are an attempt to find some stability through tightening of the ligaments. Observation of the hands while talking can be a very helpful indicator of the syndrome as often individuals with JHS show hyperextension at the metacarpal-phalangeal and/or interphalangeal joints.

It is important to observe the whole body and in the majority of cases, this will mean undressing to underwear



Fig. 4. Papyraceous (paper) scarring.

and observing the individual from all directions. Papyraceous scarring as can be seen in Fig. 4, is a documented feature of JHS (Grahame, 2003b) and therefore inspection of surgical and injury scars should be observed and noted. An interesting clinical observation is that muscle definition often appears poor with low resting tone even when the individual has been training and is reasonably fit. Both static postures, including standing and sitting and dynamic activities such as walking, stair climbing, sit to stand, standing on one leg and squatting should be carefully observed. Hypermobile individuals are frequently observed to adopt end of range postures. A typical standing posture shows flat feet, knees and hips hyperextended and lumbar spine in a sway posture with increased compensatory curve higher up the spine. Hip hitch, drop and adduction ‘hip hanging’ is also frequently observed when standing on one leg.

2.2.2. Measuring hypermobility

The Beighton score (Beighton et al., 1973) is an easy to administer 9-point scale where points are given for

Box 2

Brighton criteria—diagnostic criteria for hypermobility syndrome (Grahame et al., 2000)

Major criteria

1. A Beighton score of 4/9 or greater (either currently or historically)
2. Arthralgia for longer than 3 months in four 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 (for 3 months or longer) in one to 3 joints or back pain for (for 3 months or longer), spondylosis, spondylolysis/spondylolisthesis.
3. Dislocation/subluxation in more than one joint, or in one joint on more than one occasion.
4. Soft tissue rheumatism: three or more lesions (e.g. epicondylitis, tenosynovitis, bursitis).
5. Marfanoid habitus (tall, slim, span/height ration > 1.03 upper: lower segment ration less than 0.89, arachnodactyly (positive 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. BJHS is excluded by the presence of Marfan or Ehlers—Danlos syndromes other than the EDS hypermobility type (formerly EDS III).

nb: Criteria Major 1 and Minor 1 are mutually exclusive, as are Major 2 and Minor 2.

the performance of five manoeuvres. It is generally considered that hypermobility is present if 4 out of 9 points are scored. The scale was not designed for clinical use and has been criticised because it only samples a few joints and gives no indication of the degree of hypermobility. Other scales, such as the Comtopasis score (Grahame, 2003a) and the 10-point Hospital del Mar (Barcelona) criteria (Bulbena et al., 1992) have also been used to identify hypermobility, mostly in the context of research, as they are often too time-consuming to perform in the clinical setting.

The Beighton score has been incorporated into a more comprehensive and validated set of criteria used to identify JHS called the Brighton Criteria (Grahame, 2000), Box 2. This set of criteria takes account of not only the presence of joint hypermobility, either currently or historically, but also links this to symptoms and other characteristics of connective tissue laxity. This is an important advance as patients seek help from medical practitioners usually not for hypermobility but rather for the effects of hypermobility. As with other scales, it was initially designed for use in research, but is proving to be a useful clinical diagnostic tool (Grahame, 2003b).

Active movement testing may reveal 'normal' range of movement despite the patient complaining of symptoms such as pain and stiffness. Therapists are generally trained to identify and associate restricted or reduced movement as indicative of a problem. Full range movement is often interpreted, as meaning there is not

a problem. However, the question that therapists needs to ask is, is this 'normal' range of movement being performed by the patient 'normal' for them? A simple question to ask with regard to lumbar flexion is, "could you ever place your hands flat on the floor without bending your knees?" Observation of hypermobility or excessive movement in other joints, not associated with the problem area, may be helpful in confirming the diagnosis of hypermobility and JHS.

It is particularly important to analyse quality and patterns of the movement, rather than quantity of movement in this group of individuals, as information gained through this observation will often provide an indication of the direction treatment should take. The concept of compensatory relative flexibility (Sahrmann, 2002) is an important consideration, and occurs as a result of muscle imbalances, joint stiffness, poor motor control and altered recruitment patterns. A good example of this is, spinal extension, where frequently the majority of movement occurs in the mid lumbar spine with relatively little or no movement in the thoracic spine. The lumbar spine is one of the most mobile sections of the vertebral column and in the hypermobile individual often moves excessively. If this movement pattern is repeatedly re-enforced through activities of daily living, it may lead to pain arising from overuse in the lumbar spine motion segments.

In addition to demonstrating an excessive range of movement in some joints, as described earlier,

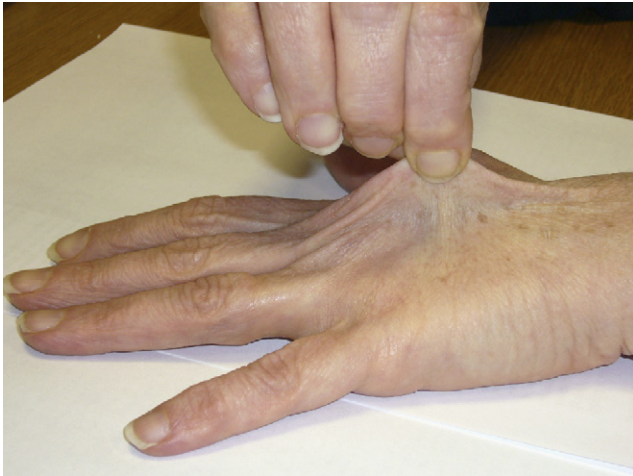


Fig. 5. Testing skin laxity on the dorsum of the hand.

proprioceptive deficits are also frequently observed. In the clinical environment, balance testing is generally used as an indication of lower limb proprioception. Double and one legged Romberg tests, with arms by the side and eyes closed and where the clinician observes the tendency to sway or fall to the side can easily be incorporated into the assessment to give a subjective indication of proprioception (Harrelson and Leaver-Dunn, 1998). More objective and reliable tests of sway may be performed and measured using calibrated standing platforms (Cachupe et al., 2001).

Joint and soft tissue palpation, a valuable and important part of the clinical examination, must be performed with care because of tissue fragility, decreased tissue resistance and increased mobility requiring less force to produce movement. Hypermobile joints frequently have an 'empty', 'boggy' or 'soggy' end-feel which may help to alert the clinician to the diagnosis. In JHS the skin is often observed to be extensible and soft. Skin pliability and elasticity can be assessed by picking up the skin on the back of the hand and assessing the excursion as in Fig. 5.

3. Management

The management of individuals with JHS can be very challenging. 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. Patients frequently arrive in the hope of finding a miracle cure having been treated by an array of allopathic and complementary practitioners. Management, is the operative word, as progress is often slow and hampered by frequent set backs and flare-ups.

Very little has been reported or written about the physiotherapy management and treatment efficacy of

JHS. Of note, however is an article by Cherpel and Marks (1999), which provides a good review including well-reasoned, evidence based suggestions for management. Further to this, a seminal physiotherapy case report by Russek (2000) gives a thoughtful overview of the assessment, prognosis and management of an athletic 28 year old chronic JHS sufferer. Russek's report highlights the importance and need for education, therapeutic exercise, adaptation and modification of work and lifestyle activities. Detailed, evidence based and clinically reasoned strategies with good inclusion of case scenarios are also reported in the text produced by Keer and Grahame (2003).

As with most patients, developing a prioritised problem list along with agreed short, medium and long-term goals is the key to successful client care. Initial management will often involve modulation of an acute episode of pain or injury (Keer, 2003). This may be achieved through advice and discussion regarding rest, pacing activities, joint care and use of a range of modalities including ultrasound and transcutaneous nerve stimulation, tape and splinting, heat and ice, gentle mobilisations of associated hypomobile areas, massage, muscle energy techniques and acupuncture. In some cases of extreme joint hypermobility and laxity, we have found advising patients to wear firm fitting underwear, lycra cycling shorts and upper body clothing helpful for improving perceived joint stability and reducing pain.

Patients usually respond well to these modalities, although recovery and healing is often slow (Grahame, 2000). Extra care should be taken with manual therapy, as pain is often latent and easily aggravated. The increased vulnerability and fragility of the connective tissue has to be recognised and considered when deciding on dosage. It is generally considered that high velocity thrust techniques (HVT) or Grade V's are contra-indicated in the hypermobile patient, although in skilled hands gentle precise HVT's or Grade V's can be beneficially applied to a stiff thoracic spine. The therapist should be mindful of the effect a mobilisation applied to a stiff area can unintentionally have at an adjacent hypermobile section, as vigorous treatment can often cause an exacerbation or flare-up with deleterious effects. At its worst, tissue damage occurs. There may also be other effects, which the therapist may not be aware of as the patient may not return for further sessions due to a loss of confidence in the therapist or therapy, leading to another downturn for the patient and further searching for help. This can be an important factor in the downward spiral, which frequently occurs in hypermobile individuals. Pain can become a debilitating unremitting symptom, leading to kinesiphobia and de-conditioning. If an individual reaches this stage, referral to a pain management programme is advised where the patient will receive both psychological and physiological input.

Once the acute injury and pain has been attended to, consideration needs to be given to the underlying condition and chronic management issues. This may include additional podiatric assessment with a view to provision of orthotics to improve foot biomechanics and support. The management will almost certainly require a degree of behaviour modification including pacing techniques, coping strategies and addressing ergonomics, work and lifestyle issues. Furthermore dietary advice with regard to irritable bowel symptoms, supplements, nutrition and weight management may also be required. JHS is more common in females and therefore attention may need to be given to issues such as incontinence, pregnancy and caring for young children. Readers are directed to chapters by Harding, Mangharan and Keer, Edwards-Fowler and Mansi, in Keer and Grahame (2003), where these concepts are explored in greater detail.

As stated earlier, patients with JHS often present in a de-conditioned state due to fear avoidance and reduced activity, making them a high-risk group for other disorders. This is of key importance to the JHS patient group, where a multitude of conditions have been associated with the syndrome (Grahame, 2003b), some of which may be averted or managed by regular exercise and improved levels of fitness.

Therefore, developing and identifying ways to encourage physical activity is an important part of the management of JHS, as reduced physical activity is known to be a major modifiable risk factor for numerous systemic diseases and complex disorders (DoH, 2004).

3.1. Principles of rehabilitation

Application of the principles of exercise physiology and motivational strategies are advised when constructing and implementing rehabilitation and physical activity programmes. The primary principle of readiness or preparedness of the individual to undertake or participate in a rehabilitation programme is fundamental to the process (Simmonds, 2003). Other principles, including specificity of training in order to target the appropriate physiological systems, and overload, intensity and frequency of training should be considered and continuously monitored. Strength in particular, is known to be highly specific to training and therefore consideration of the type of muscle activity, number and sets of repetitions and frequency of training is necessary (Wilmore and Costill, 2004; Arnold and Gentry, 2005). Initial strength gains have been attributed to neurological adaptation while gains due to muscle hypertrophy come later (Sale, 1988). It is our observation that strength gains are slower in this group of patients and this may be attributed to alterations in both central and peripheral neuromuscular physiological processes.

Research is required in this area to explore and further evidence this. Muscle endurance training is also an important part of the reconditioning process as the slow twitch type I muscle fibres are purported to atrophy at a faster rate than type II fibres (Harrelson, 1998). This suggestion has significant implications for postural muscles where the endurance capacity is crucial to function and may be part of the explanation as to why individuals with JHS, tend to fidget and appear unable to sustain sitting and standing postures.

3.2. Early rehabilitation

It is recommended that rehabilitation in the very early stage focuses on improving body awareness, proprioception and proximal joint stability. In particular the authors have found Swiss ball and hydrotherapy to be particularly useful, especially when dealing with patients where protective muscle spasm is an issue. It is important, particularly in the early stages, that exercises given to the patient either during a therapy session or as home exercises are *pain free*. A distinction needs to be drawn between training pain and exacerbation of *their* pain. This may mean modifying even the most simple exercise to ensure that it is being performed correctly and is appropriate for the patients' stage of rehabilitation. Where possible recruitment of stability muscles, once learnt, can be encouraged during normal activities, such as walking, sit to stand, stairs and housework. Manual guidance, joint approximation techniques and the use of tape to facilitate proprioception (Callaghan et al., 2002) may be helpful, although prudence is required where skin is fragile and sensitive.

3.3. Middle and late stage rehabilitation

Once a reasonable level of proximal stability has been achieved, individuals should be encouraged to continue to improve their strength, endurance, balance and co-ordination and to engage in more regular physical activity. Graded exercises using theraband, aimed at improving both concentric and eccentric strength and endurance is recommended along with the use of mirrors to enhance proprioception, see Fig. 6. A study by Kerr et al. (2000), demonstrated positive benefits of a stabilising exercise programme and a recent study by Ferrell et al. (2004), showed significant improvement in knee joint proprioception and balance, following an eight week exercise programme employing progressive closed chain kinetic exercises. The study also showed improvements in quality of life, reduction of pain and improved muscle strength.

Achievable goals should continue to be discussed, agreed and monitored using diaries, pedometers and accelerometers. The programme of rehabilitation reconditioning should be integrated and should address



Fig. 6. Closed chain exercises using theraband and mirrors aid proprioceptive feedback.



Fig. 7. Deep water walking and running is an excellent means of increasing and maintaining fitness.

the three primary systems that influence normal movement, the cardiorespiratory, musculo-skeletal and neurological systems. Cardiorespiratory conditioning and weight control can be achieved through a suitably designed low intensity aerobic programme of walking, deep water running (Fig. 7), cross trainer, stationary and

out of door bicycling where target heart rates can be monitored.

3.4. Sport, the performing arts and physical activity

Those wishing to take up athletic activity or return to sport or performance activities should undergo a functional training or rehabilitation approach, whereby both skill acquisition and sport specific training is undertaken. The relationship between generalised hypermobility and increased injury risk is not conclusive, however, there is mounting evidence to suggest that it is an intrinsic injury risk factor in many sports and performance activities including; American football (Nicholas, 1970), gymnastics (Kirby et al., 1981), basketball (Gray et al., 1985), female soccer (Soderman et al., 2001), professional ballet (McCormack et al., 2004), male rugby (Stewart and Burden, 2004) and junior netball (Smith et al., 2005). Therefore a degree of caution is recommended when advising patients with regard to sport and performance participation. A gradual return to training and match play with education regarding joint and tissue protection and care to coaches, parents and players is recommended.

Maintenance of physical fitness through regular safe physical activity is considered paramount for continued self-management of the condition and patients should be encouraged to develop a life-long commitment to physical activity and to remain fit through activities which are focussed on neuro-musculo-skeletal control. Recommended activities therefore include, recreational swimming, Pilates, tai chi, chi gung, some forms of yoga and dance. Whatever form of physical activity is recommended or adopted after the therapy intervention, it should be enjoyable, pain free and relevant to the individual.

3.5. Support groups

When required and as necessary, patients can be directed to the [Hypermobility Syndrome Patient Association, UK \(HMSA, www.hypermobility.org\)](http://www.hypermobility.org), a well-organised charity which has an active interactive website and is supported by medical and allied health practitioners with specialist interests and knowledge.

4. Conclusion

Hypermobility syndrome is a complex, under recognised and poorly managed inherited connective tissue disorder often resulting in a great deal of pain and suffering. Physiotherapists working alongside other members of the multidisciplinary team have an important role in both the identification and management of the condition. The Brighton Criteria and a simple 5-point questionnaire

devised by Hakim and Grahame (2003) are both valid tools, which can be used in the clinical and research setting to identify the condition. Because of the ubiquitous nature of connective tissue proteins (Grahame, 2003c), the possible consequences of tissue trauma are vast and patient presentations are therefore variable. Setting and monitoring carefully considered shared goals along with behaviour modification are important strategies for achieving the ultimate goals of independence and long-term functional fitness. Progress is often slow and hampered by setbacks and exacerbation of pain and psychological distress. However, with persistence and insight into the pathogenesis of the disorder, rewarding outcomes are possible. Future clinical research involving both qualitative and quantitative methods will aid the further development of clinical guidelines.

Part two of this Masterclass will be available online from November 2007.

Acknowledgement

The authors wish to thank Professor Rodney Grahame CBE MD FRCP FACP, Consultant Rheumatologist, University College Hospital and Honorary Professor, University College London, in the Department of Medicine, for his contribution to this paper and ongoing support for our work in this field.

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