Chapter 9

Physiotherapy and occupational therapy in the hypermobile adult

Rosemary Keer
Katherine Butler

INTRODUCTION

Joint hypermobility syndrome (JHS) is under-recognized, poorly understood and, sadly, often poorly managed (Beighton 1999). The syndrome does not necessarily cause problems and for many individuals it can be an asset, amply illustrated in the world of music, dance and sport. For others it can be a liability, producing significant problems which are often ignored by the medical profession (Phipps 2003, HMSA 2006). Early recognition and appropriate management can decrease suffering, reduce the need for unnecessary tests and invasive procedures, help to avoid surgery, reduce chronic pain and fear (Cherpel & Marks 1999) and prevent the destructive spiral into depression and physical deconditioning. It is the aim of this chapter to discuss the presentation of the syndrome to aid recognition and diagnosis by primary care practitioners and to explore assessment and management strategies to decrease suffering and disability, increase function and enable the hypermobile individual to effectively self-manage the condition.

PRESENTATION

ASSESSMENT

Pain is the most common complaint, often being widely distributed, involving several joints and areas of the body, and varying in duration from acute (15 days) to as long as 45 years (El-Shahaly & El-Sherif 1991). A typical presentation is illustrated in Keer & Grahame (2003), and Simmonds & Keer (2007). The onset frequently occurs in childhood or adolescence, particularly in girls between the ages of 13–19 years, with 75% of hypermobile adolescents developing symptoms by the age of 15 (Kirk et al 1967) (Chapters 2, 10 and 11). Anecdotally, onset is also often associated with trauma, pregnancy, childbirth, a change in physical activity, such as a marked decrease contributing to deconditioning, or an unresolved joint problem which leads to compensatory strategies producing other joint problems. In addition, the patient will often report past episodes of medical or therapy intervention which were unhelpful and may even have exacerbated their symptoms. Exploring their history in this respect not only helps to identify potential generalized hypermobility, but will also give valuable information regarding what has not helped in the past, so that mistakes are not repeated.

It is unusual for pain to be the only symptom, so asking for details about other symptoms can give clues to tissues or systems which may be potential causes of the problem and may require examination (Table 9.1). Stiffness is a very common complaint and may seem at odds with the range of movement on testing. This is a subjective feeling of stiffness and although the range of movement may appear normal it may not be normal for that particular patient. Asking 'have you ever been able to touch the floor with your palms flat' (Oliver 2000) can be a useful indicator of previous hypermobility, a factor that is acknowledged in both the 5-point questionnaire (Hakim & Grahame 2003) and the Brighton Criteria (Grahame et al 2000) (Chapter 1). The reports of stiffness may also be a reflection of decreasing flexibility with

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Table 9.1 Common symptoms reported and possible tissue or system at fault

SYMPTOM REPORTED	SYSTEM TO BE EXAMINED
Stiffness	Muscle system (muscular tension)
'Feeling like a 90 year old'	Joints Hormonal
Audible clicking, popping, clunking, sensations of subluxation, vulnerability, instability or frank dislocation	Motor control Stability system
Sensations of parasthesiae, tingling, numbness, deadness	Neural system
Tiredness, fatigue, faintness, feeling unwell, flu-like symptoms	Autonomic nervous system Physical deconditioning

age, or due to injury or muscle stiffness/spasm. Paraesthesiaes are frequently non-dermatomal and there may be an increased disposition to development of a neuropathy (Francis et al 1987, March et al 1988).

Aggravating factors to determine the Severity, Irritability & Nature (SIN) (Maitland 1986) are not clear cut. Latency is frequently a problem, with symptoms developing several hours or even a day later, making it very difficult to identify the aggravating activity. This makes it difficult to judge the response to a physical examination and treatment. Careful questioning about the individual's response to physical activity will help, although caution is advised with regard to the amount and vigour of testing, particularly initially.

Hypermobile individuals frequently dislike static postures and report difficulty with standing still, such as when queuing, shopping or attending exhibitions, and sitting for prolonged periods of time. Hypermobility of the spine was found to increase the prevalence of back pain in industrial workers working in standing/sitting postures (Larsson et al 1995) and hypermobility of the spine and knees produced symptoms in musicians due to sustained standing postures (Larsson et al 1993).

In addition, hypermobile individuals dislike repetitive activities, being more likely to develop musculoskeletal lesions (Acasuso-Diaz et al 1993) and reporting an increased frequency of previous episodes and more recurrent episodes of overuse soft tissue lesions such as bursitis, tendonitis and fasciitis at a single site (Hudson et al 1995, 1998) as well as fibromyalgia (Goldman 1991, Acasuso-Diaz & Collantes-Estevez 1998) (Chapter 5). It is often the case that pain or injury has occurred with minimal provocation during everyday activities, such that the patient cannot say what has brought the pain on. This can be very frustrating for the therapist and patient alike, but it is worth viewing this as a valuable piece of the jigsaw, leading to more accurate recognition and diagnosis of generalized hypermobility.

Certain aspects of the patient's history can be valuable in providing clues to hypermobility. Back and knee pain are reported most commonly as a source of pain in childhood (Biro et al 1983) (Chapter 11). Individuals who no longer demonstrate hypermobility due to age, injury or stiffness secondary to pain, may be able to confirm that they were more flexible when younger, or that there is a family history of increased flexibility. Studies have shown that 27–65% of patients with symptomatic hypermobility seen in clinic had relatives with hypermobility (Finsterbush & Pogrund 1982, Biro et al 1983, Bridges et al 1992). There may also be evidence of involvement of other connective tissue, because patients report that they bruise easily (Al-Rawi et al 1985, Kaplinsky et al 1998) and there is frequently a history of varicose veins, hernia, or prolapses (Wynne-Davies 1971, Al-Rawi et al 1985, El-Shahaly & El-Sherif 1991).

Hypermobile individuals frequently recount a long history of soft tissue injuries, strains and sprains (Acasuso-Diaz et al 1993), which can be suggestive of vulnerable tissues or areas of vulnerability due to increased tissue laxity, poor control and muscular weakness. Dislocations and subluxations are more common (Finsterbush & Pogrund 1982). This should prompt a holistic assessment looking at the functioning of the whole body, to identify reasons why a particular area is causing a problem. How well the individual recovered from previous injury/problems will have a bearing on subsequent treatment, expectations and prognosis, particularly as there is anecdotal evidence of poor healing and a slower recovery in this patient group (Russek 2000).

In addition, questions relating to gender, age, ethnicity, weight, details of work or daily activities, leisure and exercise or sport activities will help to give a full picture of the impact of JHS on the individual. It is also important to ask special questions regarding general health and medical history in terms of surgery or illnesses, related or not, such

as fatigue, chronic fatigue syndrome and fibromyalgia, as well as details of medication, so that the physical examination can be tailored to the individual's presentation without risking a flare-up. Discussing medication may also highlight the possibility of JHS as a diagnosis, because there is often a poor response to non-steroidal anti-inflammatory drugs (NSAIDs), and local anaesthetics have been reported as less effective in EDS type III patients (Arendt-Neilson 1990, Hakim et al 2005).

EXAMINATION

Observation of the patient's mannerisms and postures during history-taking may give an indication of hypermobility. Contact of the hand to the face may reveal hyperextension of the digits, constantly changing position or fidgeting and/or sitting in an unusual posture (legs wound round each other, slumped to the side or resting on the lateral border of the feet) are familiar sightings (Fig. 9.1).

Skin is a very important feature and helps to confirm a diagnosis of hypermobility. To the touch it is silky, soft, extensible and thin. Elasticity can be tested by picking up a section from the back of the hand between the thumb and first finger (Chapter 2). Wounds may show poor healing with a thinner or papery scar. There may be stretch marks (striae atrophicae) not associated with weight loss or pregnancy, which appear during the growth phase in certain areas such as across the lumbar spine and pelvis, around the hips, thighs and shoulders.

The Beighton scale (Beighton et al 1973) gives an indication of hypermobility in certain joints (JHM), and while it is quick and easy to use in the clinic, it is limited in its application as only a few joints are tested. Other scales are discussed in more detail in Chapter 1. A diagnosis of JHS can be confirmed using the Brighton Criteria (Grahame et al 2000) which assesses the effect of weaker connective tissue on the body as a whole, but its use is primarily as a research tool. In an out-patient setting, making a diagnosis of JHS will depend on a thorough history and physical examination of the whole body.

It may be appropriate or indeed necessary to refer a patient, whom one suspects of being hypermobile, to a rheumatologist for a definitive diagnosis or further investigations when:

- the patient's symptoms do not fit the examination findings (except with regard to the range of movement as mentioned below)
- a clear diagnosis regarding the nature of the connective tissue disease is required and it is necessary to exclude other pathology

Fig. 9.1 Typical sitting posture: (a) legs wound round each other, (b) resting on the lateral border of the feet

- a complex presentation involving many body areas and systems requires access to other health care professionals
- further investigations are required to provide more information regarding pathology, such as in the spine or peripheral joints to identify degeneration or instability (Grahame 2003).

Pain is also thought to arise because the connective tissue (ligament, tendon, muscle, capsule) in hypermobile individuals is more lax and less resilient and suggests a predisposition to the effects of trauma (acute and chronic) from overuse and misuse. Microtrauma occurs more frequently and with less provocation (Acasuso-Diaz et al 1993, Hudson et al 1998), meaning that even everyday activities can be the cause of tissue trauma and pain. For some, there is the added problem of subluxation, dislocation or instability which can produce, or be the result of, poor movement control. The hypermobile individual has an increased susceptibility to instability due to increased extensibility in the connective tissue restraints (capsule, ligament, muscle, tendon), affecting the passive control system, decreased muscle tone and strength, affecting the active control system, and deficient proprioceptive acuity (Mallik et al 1994, Hall et al 1995), affecting the neural and feedback control system, which is further outlined in Panjabi's model of stability (1992) (Chapter 12.8).

Poor movement control has the potential to lead to further joint problems, setting off a chain reaction or 'domino' effect whereby a problem in one joint can develop into widespread pain affecting many joints over a period of time. Increased healing time may lead to more well-developed compensatory strategies which affect other areas, progressing onto chronic pain and physical deconditioning (Fig. 9.2).

It is helpful to think in terms of compensatory relative flexibility, a concept proposed by Sahrmann (2002). This states that movement occurs more readily at a joint with less stiffness than at a joint with relatively more stiffness. For example, hamstring muscle tension or stiffness either as a result of habitually sitting for long periods, or as a result of injury, will inhibit movement at the hip into flexion during forward bending, potentially leading to overflexion in the spine. It is common to see decreased hip flexion and increased spinal flexion in hypermobile individuals and this may be contributing to low-grade irritation and strain to lumbar spine tissues, producing pain. Figure 9.3a is an example of this and is in contrast to Figure 9.3b which shows well-balanced full range flexion in a hypermobile individual.

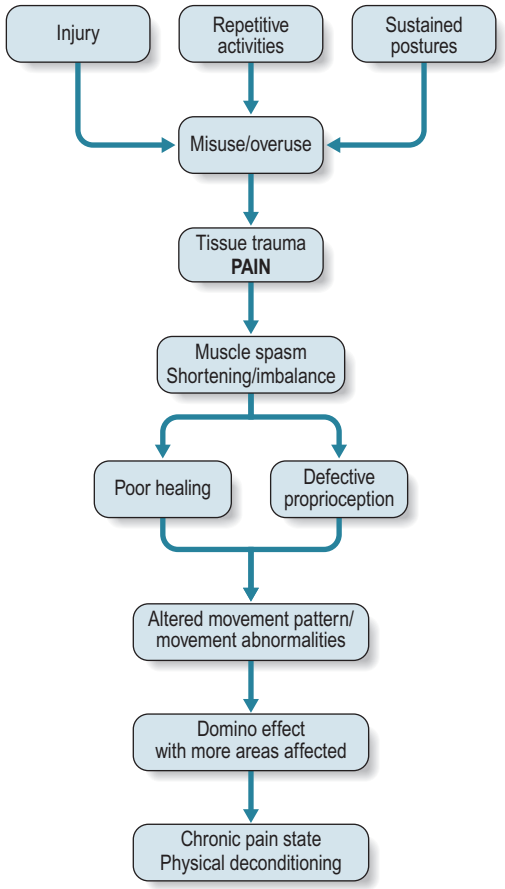

Fig. 9.2 Basis of symptoms

There is evidence of neurophysiological defects such as pain enhancement and autonomic dysfunction (Gazit et al 2003) (Chapter 6.1), which will have the effect of increasing disability, leading to less activity and a vicious spiral of chronic pain and physical deconditioning (Rose 1985). Psychological distress is common, with concerns and difficulties occurring around work, home life and raising children, as well as fear of pain, serious illness or permanent disability. Individuals have often had to withstand years of having their complaints dismissed, being misunderstood or not being believed by the medical profession at large (Phipps 2003). This has, understandably in some cases, led to anger, confusion and depression.

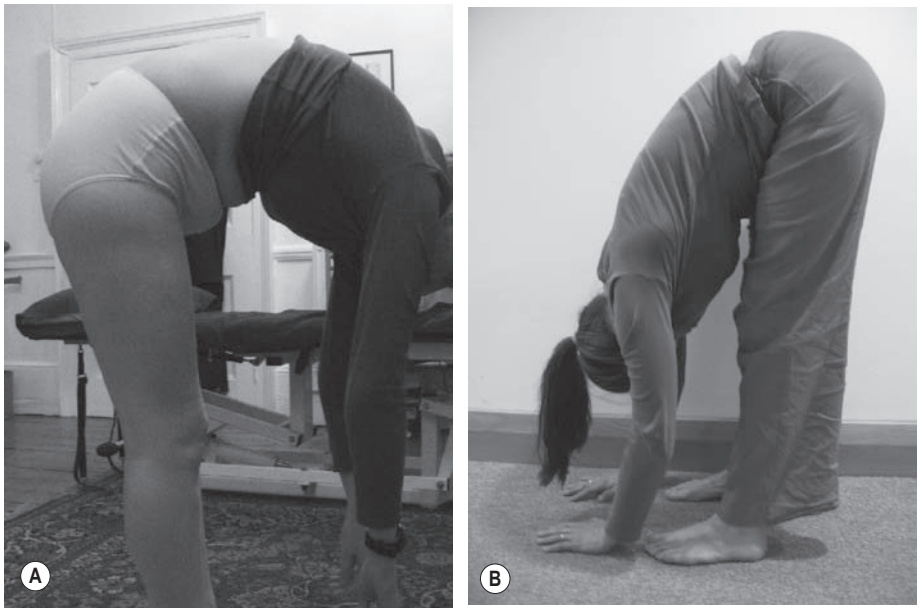

Fig. 9.3 Compensatory relative flexibility: (a) forward flexion with decreased hip flexion relative to spinal flexion, (b) forward flexion with balanced movement throughout the spine and hips

The physical examination starts with observation. The patient, undressed to their underclothes and standing in their normal stance, is assessed from all four directions (front, back and each side) and compared with what is considered the ideal skeletal alignment. This is a standard based on sound scientific principles which involves minimal stress and strain on the body tissues and allows maximal efficiency (Kendall et al 1993). Details of a thorough postural examination are given in Kendall et al (1993) and should include postural alignment, symmetry and weight distribution from head to toe.

Hypermobile individuals show a tendency to rest at the end of joint range and have poor postural alignment. Even if they are initially able to stand or sit in a good position, it is difficult to sustain the position for long periods. It is easier for their bodies to take the path of least resistance and settle into a position which requires decreased muscle work, resting on tension in their soft tissues. Familiar postures are slumped sitting, standing with hyperextended knees and hips (Fig. 9.4) and hanging on the hip. This leads to stress and strain in the collagenous tissues. If these postures are sustained, fluid is forced out and tissue nutrition is adversely affected. The tissue undergoes creep, which leads to deformation,

Fig. 9.4 Standing with hyperextending knees

stretching and weakening over time. There is also a weakening of surrounding muscles through disuse and changes in how and where load is transferred through the kinetic chain. Hyperextension at the knee provides a good illustration (Kendall

et al 1993), with the patella displaced caudally and contact of the femoral condyles on the tibial plateau shifted more anteriorally. This has the potential to weaken and damage tissue, disrupt nutrition of joint tissue and produce pain. There are also effects along the kinetic chain at a distance from the knee, with the ankle joint held in plantarflexion, producing changes in the calf muscles, and also into the hip and lumbar spine.

ACTIVE MOVEMENT

Hypermobile individuals tend to look and move well, even if they have pain. Thus during active range of movement testing their reports of symptoms (subjective) are often at odds with the physical examination (objective). It is possible to see how misunderstandings may arise; the person who complains of widespread debilitating pain affecting several joints can, when asked, bend forward to touch their toes. This tends to go against medical wisdom and may lead to the individual being dismissed or considered a hypochondriac. The explanation may be that although the movement looks to be full range, it may not be 'normal' or full range for that particular person. Additionally, it may be that some areas are moving into hypermobile range (such as lumbar spine) and masking reduced range in an adjacent area (such as the hip joint). It is important to assess the quality of movement and critically appraise where, when and how much movement is occurring throughout the body or kinetic chain, while keeping the biomechanics in mind. Hypermobility may be masked when range of movement is decreased due to fear, pain or stiffness following injury or inactivity or in the case of an older person who may have lost their normal degree of flexibility with age. Clinical reasoning will lead to examination of other areas (skin, other joints, historical flexibility) for confirmation of hypermobility.

It is helpful to examine the effect of moving one area in the context of the whole body. This may mean examining ease and range of movement in all areas of the spine as well as some peripheral joints, if symptomatic. For instance, full flexion at the shoulders may only be possible with lumbar spine extension, because latissimus dorsi is shortened or overactive (Sahrmann 2002), producing an area of relative stiffness. This has the potential to lead to overuse injuries at the shoulder through repetition of an overhead activity, particularly relevant in the sports person. It would therefore be necessary to assess the shoulder and whole spine. For the less active, it may be as simple as increased pronation in the midfoot as a result of stiffness at the ankle into dorsiflexion, either through talocrural stiffness (from injury) and/or soft tissue shortness/tightness in the calves or higher in the kinetic chain. Poor movement patterns may also be due to muscle deconditioning and weakness. A case highlighted in Simmonds and Keer (2008) involved an individual lifting with poor muscle control around the wrist which led to the wrist being pushed into ulnar deviation under load, causing strain to the ligament and tendon on the radial side. Close questioning and detailed examination of everyday habitual activities can highlight significant problems, which can be easily rectified once identified.

Functional activities, assessed as relevant to the individual, need to be examined. These include walking (over a set distance), climbing stairs, sitting to standing, bending, mini squat to full squat, one-leg stand, heel raise and any specific activity known to cause problems. In addition to pain, it is also helpful to look for even weight bearing, balance, coordination, symmetry and load transfer (Lee & Lee 2007). Pain is not always reproduced on testing unless the problem area is in the acute stage. This appears to be due to the tendency for pain to be aggravated by sustained postures or repeated activities. If repeating an activity or sustaining a posture does not reproduce pain, the examination must rely on an analysis of the quality and patterns of movement throughout the kinetic chain. Forward spinal bend is useful for assessing the relative contribution of lumbar, thoracic and hip joints into flexion as well as highlighting segmental hinging or translation. Mini squat is helpful in assessing lower limb alignment during a functional activity. One-leg stand assesses the ability to transfer load effectively through the pelvis and hip. Heel raise can be helpful in assessing control around the foot and ankle.

Hypermobiles frequently use a 'bracing' pattern with breath holding in an attempt to improve stability and produce more force. As the global muscles of the trunk are used, the ribs can become fixed so that chest expansion, and therefore efficient respiration, is affected. The individual either breathes apically or into the abdomen, with the risk of increasing abdominal pressure downwards which can have a deleterious effect on the pelvic floor. It is useful to assess the breathing pattern at rest and with activity.

Many hypermobiles suffer from feelings of instability and 'giving way', subluxations and dislocations, and due to the lax connective tissue, joints may become unstable through injury. Treatment is therefore focused on control of movement and it is important to assess the local postural muscles associated with trunk stability, such as transversus abdominus, deep multifidus and the pelvic floor muscles in terms of timing, atrophy, loss of tonic function, loss of co-ordination and asymmetry. There is also support for the use of real time ultrasound imaging to assess performance of the deep stability muscles, which can be particularly useful for identifying 'cues' which the patient can use to activate the correct muscle (Fig. 9.5).

Examination continues with detailed inspection of individual structures that have been identified as possible contributors to the patient's symptoms.

1. Articular movement testing, both passive and accessory movement, to identify hypermobility, hypomobility and symptom reproduction.
2. Muscle testing, assessing strength, tone, length, timing and tenderness of trigger points.
3. Neural testing, both in terms of function (sensation, reflexes and power) and mobility (Butler 2000). This may involve testing straight leg raise (SLR), upper limb tension tests (ULTTs), slump and thoracic slump, particularly if involvement of the autonomic nervous system is suspected.
4. Specific tests to identify pathology such as impingement (hip and shoulder) and active straight leg raise (ASLR).

PROPRIOCEPTION

It is known that proprioceptive acuity is diminished in individuals with JHS (Mallik et al 1994, Hall et al 1995) and this can have a significant effect on the way an individual moves. The research shows that both joint position sense and threshold to detection of movement are impaired such that they have less awareness of approaching the end of joint range (Chapter 6.4). The reason for this impairment is not known but may be related to increased laxity and elasticity in the tissues, which may explain why the application of tape has been found to enhance proprioception in those with a deficit (Callaghan et al 2002, 2008) and also why supportive, tight garments have been reported as helpful in improving proprioceptive feedback (Simmonds & Keer 2007).

In the laboratory, threshold to detection of passive movement has been found to be more reliable than joint position sense (Juul-Kristensen et al 2008), but detection is affected by the position in range of movement when testing (Ageberg et al 2007). In addition, the effect of pain and injury should be taken into consideration as Juul-Kristensen et al (2008) found that proprioception was poorer in elbows with epicondylitis than in the controls' elbows.

It can be difficult to measure proprioception accurately in the clinic and tests are generally linked to function. The ability to stand on two legs with eyes closed, or on one leg with eyes open and closed as in the Romberg test, will reveal exaggerated swaying and/or loss of balance once the eyes are closed. As in the study above, pain

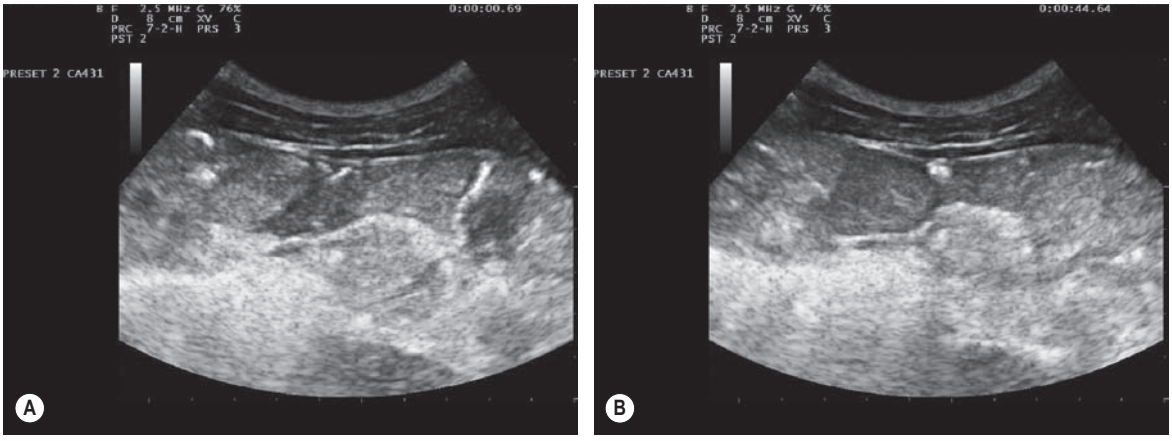

Fig. 9.5 Real-time ultrasound scans of transversus abdominus (TrA): (a) TrA at rest (b) TrA contracting, note slight thickening and sliding of third layer

may contribute to the increased sway observed due to poor stability/control around the ankle, knee, hip and/or sacro-iliac joint. Standing on one leg and observing the strategies employed in order to remain balanced can highlight poor load transference through the lower limb, but particularly through the pelvis and hip. Hip hitch, or a pseudo-Trendelenburg, is frequently observed.

MANAGEMENT

A management plan is formulated following discussion between the patient and the therapist. It is based on the findings of the assessment in terms of what is causing the problem and where treatment should be directed, and an in-depth discussion with the patient about their expectations. Many patients are hoping for a miracle cure, others want to help themselves but have received conflicting advice, while others are fearful of getting worse and 'ending up in a wheelchair'. Recent research has shown greater benefit and cost-effectiveness in treatment for musculoskeletal disorders when patients, who expressed a preference, received their preferred treatment, compared to those who expressed no preference (Preference Collaborative Review Group 2008). In addition, a qualitative study (Cooper et al 2008) exploring perceptions of physiotherapy in a group of chronic low back pain patients highlighted the importance that patients place on communication. It underpinned the other key themes which included individual care, good explanation regarding diagnosis and treatment and involvement in decision making and treatment time. Amanda Sperritt (Gawthrop et al 2007) echoes these findings in an article in the *BMJ* about her experiences of EDS. She comments that 'the most helpful professionals treat me as a partner in the management of the condition' and states that listening and support from the therapist help 'to make life that bit easier'. Amanda also reports that physiotherapy was valuable because it allowed her to play an active part in her treatment, as well as teaching her to use her muscles to protect her joints. It is only by listening to the patient's story that a prioritized problem list and achievable and appropriate goals, which are the key to successful client care, can be agreed (Simmonds & Keer 2007). Gaining a clear understanding of the patient's problem through a comprehensive assessment helps to ensure appropriate and effective intervention.

However, there are reports of physical therapy being unhelpful or even exacerbating symptoms. There may be several reasons for this and most are underpinned by a failure to recognize that the patient has JHS and, as such, has more lax, less resilient connective tissue (Acasuso-Diaz et al 1993). Extra care should be taken with manual therapy. Recovery and healing is slower than in the non-hypermobile, so if this factor is not appreciated there is a temptation to progress more quickly or with more vigour in order to produce a change.

Functional restoration is the main aim of treatment since this enables the individual to self-manage the condition. It is made up of different components.

1. Restoration of a normal range of movement for that particular individual, even if it is hypermobile (Keer et al 2003, Maillard & Murray 2003).
2. Restoration of efficient and effective movement patterns throughout full range of movement, including the hypermobile range. This will include correcting and preventing movement dysfunction and regaining joint stability.
3. Education, reassurance, advice and problem-solving (Rose 1985, Russek 1999).
4. Improving general fitness (Simmonds & Keer 2007) (Chapter 13).

Three patterns of presentation have been proposed (Finsterbush & Pogrund 1982), which require different therapeutic approaches.

1. Episodes of ***acute*** musculoskeletal pain, dislocation and subluxation, initially respond well to the usual therapeutic modalities of electrotherapy, support, ice, movement and advice.
2. Recurrent episodes or a series of episodes of pain at different sites with some physical deconditioning characterize the ***intermediate*** stage and require therapeutic modalities which are adapted and modified to provide functional restoration.
3. ***Chronic***, long-standing, severe and unremitting pain with profound physical deconditioning may require a multidisciplinary pain management programme using cognitive behavioural skills (Harding 2003) (Chapter 8).

It is important to try to give treatment that has meaning for the patient. This may be a treatment

modality or exercise which instantly takes the pain away, or addresses an area of particular concern. This gives the patient a sense of control or more comfort and as such promotes improved movement patterns and muscle recruitment but also has the power to motivate compliance to exercises or advice.

Acute symptom strategies

In the acute stages the primary aim is relief of pain. To this end the usual therapeutic modalities listed above are utilized. Rest is usually recommended for up to five days while the inflammatory stage takes place (Mattacola & Dwyer 2002), with movement to prevent the deconditioning effects of immobilization thereafter. Supporting the injured part can be very important because it allows movement of the body without strain to the injured tissue. Support can vary from a tight garment to a specific splint or brace. If the skin can tolerate it, tape is extremely useful as it moves more readily with the body. Support can help rest an area and put less stress on the body as a whole, but also, more importantly, it can allow pain-free movement. This has the real potential to prevent development of chronic pain by helping to reduce afferent stimulation of peripheral nociceptors. Vierck (2006) suggests that if tissue injury healing (particularly injury to deep tissues) and pain do not settle within 3 months there is a risk of developing chronic widespread pain and fibromyalgia.

'Injury' in a hypermobile individual may be a small everyday incident, such as picking up a bag, which provokes instantaneous pain (Simmonds & Keer 2007) or a sustained posture with joints at the end of their range of movement. In both these cases, if hypermobile connective tissue is not recognized it may be hard to analyse what caused the problem and easy to dismiss the patient's complaints. Inflammation is not necessarily a dominant feature.

There is an increased risk of deconditioning (Rose 1985) and an increased healing time (Russek 2000) in the hypermobile individual, so, while being mindful of the normal healing times, it is wise to be cautious and proceed slowly. Pacing of activities and specific exercises which do not provoke pain are important to prevent an exacerbation. Care should be taken during the proliferative stage up to 21 days after injury and it may be necessary to continue using support. During the remodelling stage controlled stress needs to be applied to produce a robust repair, but again care is needed in this patient group with particular attention to a gradual controlled progression of stress.

Taping has several uses. It can provide support for injured tissue, such as a medial ligament strain at the knee and reduce pain (Macgregor et al 2005, Aminaka and Gribble 2008). There is also evidence that it can facilitate better muscle activation and patterning. Macgregor et al (2005) found that patella tape which produced a lateral stretch across the patella increased VMO activity in people with patellofemoral pain. Christou (2004) found similar effects with both medial tape and placebo tape and suggested the mechanism is via cutaneous stimulation. Interestingly, they also found that tape had the opposite effect in healthy individuals. There is evidence for tape enhancing proprioception, but only in those whose proprioception is poor (Callaghan et al 2002, 2008). Individuals with normal proprioception were either shown to have no effect or a worsening of joint position sense when tape was applied to the knee. This was true whether the subject had a healthy knee (Callaghan 2002) or suffered from patellofemoral pain syndrome (Callaghan 2008). There is also evidence of patella taping having a beneficial effect on dynamic postural control in patients with patellofemoral pain syndrome compared to healthy individuals (Aminaka & Gribble 2008).

Intermediate symptom strategies

This group may have progressed from the acute stage without good healing or resolution and find themselves with a 'domino effect', whereby one initial injury sets off a chain reaction. This may be the result of resting too much and becoming deconditioned, or through developing poor compensation strategies, or being encouraged to push through the pain and irritating more tissues. At this stage it is very important to assess the whole person and all areas of symptoms in order to be able to identify what is continuing to 'drive' the problem and thereby apply appropriate treatment.

The treatment is now based on rehabilitation and functional restoration with the aim of returning the individual back to their former self. Once pain is under control, treatment is concerned with restoring a full range of movement and ensuring good motor control throughout the movement,

even if it is hypermobile. Manual therapy is often used at this stage to restore normal movement and can be applied to the affected tissue(s) as indicated by the assessment.

1. Joints. Gentle, rhythmic, large-amplitude oscillations (Maitland 1986) have been found anecdotally to be most effective at reducing pain and improving joint range. However, high-velocity thrust techniques (HVT) are generally thought to be contraindicated, particularly in the cervical spine, although they can be successfully applied to a stiff thoracic spine in skilled hands.
2. Neural tissue. Restriction to normal neural mobility can be treated effectively with gentle mobilizing techniques as described by Butler (2000). Sustained end of range mobilization is not recommended in the hypermobile individual and overzealous 'stretching' can result in muscle spasm and guarding.
3. Muscle and fascia. Myofascial release, a massage therapy aimed at releasing tension in muscles and fascia (Gordon & Gueth 2006) uses long, stretching, gentle sustained pressure. It is thought to improve relaxation and circulation, and decrease pain, anxiety and lactic acid build up. It can be a useful technique to release muscles that have developed an overactive and dominant pattern, but for best effect is used with patient participation and awareness (Lee & Lee 2007). This helps to highlight to the patient where they may be overworking their muscles and how to release them in the future. It may be necessary to 'switch off' and relax an overactive muscle before an inhibited or 'switched off' muscle can be voluntarily contracted.

Re-educating good movement patterns and motor control often starts with an appreciation and practice in attaining and maintaining good postural alignment, firstly in static postures and progressing onto dynamic postures and specific tasks. Maintaining a good standing and sitting posture for more than a few seconds can be challenging in the beginning for some hypermobiles, because their postural muscles are unused to holding joints (the knee, pelvis and spine in particular) in a more neutral position, so their endurance capabilities need to be built up slowly and gradually.

It is usually necessary to begin this process in non-weightbearing positions to decrease the load through the joint(s). Effective and efficient joint control begins with gaining control of the pelvis and trunk before moving to the peripheral joints. Assessment will have identified which of the deep stability muscles (transversus abdominus, pelvic floor multifidus) need facilitation to recruit effectively both in isolation and in a co-ordinated fashion. Activation is then practised while maintaining a neutral pelvic/lumbar spine position as well as a relaxed breathing pattern to prevent a 'bracing' or co-contraction rigidity strategy. This is often started in lying (supine, prone and side lying) and progressed to sitting and standing. The emphasis is on a low-level (20–30% of maximal voluntary contraction (Richardson et al 2004)) isometric hold repeated several times with relaxation in between.

The ability to maintain good control during weight-bearing is challenged by standing, initially on both legs, and progressed to one leg. All the time attention is paid to ensuring good joint alignment and relaxed but accurate and balanced muscle activation. Once control with static postures is being achieved, movement can be added in the form of knee bends or practising weight transference in stride stand in preparation for gait training. In addition, proprioception and balance can be challenged by introducing an unstable base, in the form of a gym ball, balance board (Fig. 9.6) or foam roller.

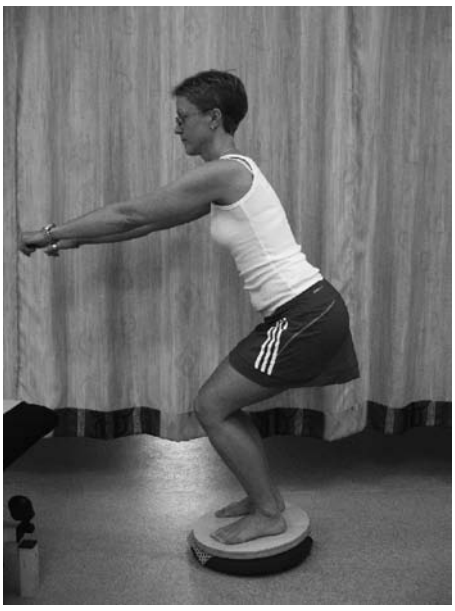

Fig. 9.6 Challenging balance and proprioception through knee bending on a balance board

There has been little research into effective treatment for symptoms as a result of JHS, although there is now evidence in the medical literature that low back pain, function and recurrence can be significantly improved through re-educating spinal stability strategies (Hides et al 1996, O'Sullivan 2000, Richardson et al 2004) and it seems logical that these recommendations can effectively be applied to the hypermobile population. Furthermore it seems that it is not only weakness of the stability muscles that is the problem but also delayed activation which allows unprotected displacement of the trunk to occur with movement of the limbs (Hodges & Richardson 1996, 1998). Specific, low-threshold training of transversus abdominus (TrA) (O'Sullivan 1997) and multifidus (Hides et al 1996) has been shown to be effective in treating chronic low back pain and preventing recurrences, and more recently Tsao and Hodges (2007) have shown this specific training can correct the timing delay in a relatively short period of time. However, there is conflicting evidence on the effectiveness of specific stabilization exercises in chronic back pain, with some considering them no better than other active physiotherapeutic interventions (May & Johnson 2008). Clinical experience provides anecdotal evidence of the ineffectiveness and harm of non-specific exercises, so the decision of what form of exercises to use will depend on clinical reasoning.

For peripheral joints there is evidence that utilizing close chained exercises in the lower limbs over an 8-week period is effective in reducing pain, improving proprioception, muscle strength and quality of life in JHS (Ferrell et al 2004). A study by Kerr et al (2000) showed similar findings in children with JHS. Gaining control of knee extension or hyperextension is vitally important for hypermobile individuals. Not only does it relieve pressure on the knees and prevent ligament damage, it also has a beneficial effect on the rest of the kinetic chain. Exercises can be started in a non-weightbearing position, but close chained to improve proprioceptive feedback. Knee flexion and extension without allowing the knee to fall into a 'lock' position can be practised by bending and straightening the knees through contact with the heels on a gym ball (Fig. 9.7). This can be advanced by performing controlled knee bends in standing on both legs and then on a single leg and progressed further by using an unstable base such as a foam pad or balance board. Exercising is always cognitive with attention on maintaining good alignment through the leg, effective pelvic control, relaxed breathing and low effort.

Similar principles can be applied in the upper limb with low load, close chained exercises being used to improve shoulder and elbow control. These often start by learning how to achieve a good shoulder position, or 'setting' the scapula (Mottram 1997). This is progressed to maintaining good scapula control during movement, such as moving a ball on a work surface and then into

Fig. 9.7 Re-educating knee control using a gym ball

more range on a wall. Further progression can be achieved by increasing the resistance and moving into weightbearing in four-point kneeling.

Once good effective joint control has been established and exercising has begun to work on maintaining control throughout movement, attention progresses to improving the endurance and strength capabilities of the body in order to withstand high load activities without losing control. Muscle endurance training is thought to be important, because type I muscle fibres (slow twitch) atrophy at a faster rate than type II fibres (Harrelson 1998). The emphasis is therefore on increased repetitions and hold time. It is important to ensure that there is good relaxation between muscle contractions, to delay the onset of fatigue. In addition, eccentric muscle work appears to be particularly difficult for the hypermobile individual. Patients frequently use gravity to return the limb to the starting position rather than using slow controlled eccentric muscle activity. Right from the early stages of the rehabilitation programme control through the full range of movement should be emphasized and worked on.

In order to build strength in a muscle, the load is progressively increased in terms of force and speed. Resistance can be applied using free weights or exercise bands. There is some evidence that muscle hypertrophy (through strength training) can lead to increased muscle/joint stiffness (Ocarino et al 2008), which may be a useful additional mechanism to gain control of the hypermobile joint.

Ramsay and Riddoch (2001) found that ballet dancers were far superior to physiotherapy students in proprioceptive accuracy and suggested that rehabilitation should encourage repetition of movement with verbal and visual feedback to improve sensory motor performances. Biofeedback in the form of mirrors and photography (stills and video) can be used to increase awareness of posture and joint control, both in static positions and during performance of specific exercises and tasks. More specialized biofeedback, such as surface electromyographic (EMG) biofeedback, can help detect the presence of excessive muscular activity during the performance of a functional task (Polnauer & Marks 1964, Philipson et al 1990) (Chapter 12.1.). Video is particularly useful when assessing and treating musicians, because the patient can face a video monitor and play their instrument while postural cues are given by the therapist (Dommerholt & Norris 1997). The video camera is placed at various angles, allowing the musician to see his or her posture from several perspectives and the session can be recorded for further study and review. This assessment technique can easily be utilized to review writing, sporting activities, or performance of daily living tasks.

Chronic symptom strategies

Individuals who fall into this group have often had symptoms for a long time, with severe deconditioning, kinesiophobia and psychological effects. They may have become dependent on using aids (collars, braces, sticks, crutches or even wheelchairs), be unable to work and claiming incapacity benefit or registered disabled. Like many chronic pain patients they will very often hold unhelpful beliefs about their pain and disability. But being hypermobile may add additional concerns regarding the weakness or vulnerability of their tissues, which increases the fear and belief that pain equates to damage. Understanding and then challenging these beliefs through interactive informative discussion and paced exercising and activity can encourage a more positive attitude and improve confidence.

Pacing is vital to those with hypermobility (Harding 2003) and fibromyalgia. Cycles of overactivity/underactivity are common and result in a flare-up of pain which requires a period of rest to recover. Repetition of this cycle leads to a downward spiral resulting in deconditioning and a confirmation (in the patient's mind) of weakness and inadequacy. Teaching pacing skills allows the patient to work at their own pace but within recognized guidelines (Harding 2003). The emphasis is on maximizing tissue health and endurance with a range of exercises and activities. Variety is key, with frequent changes between more active and more sedentary tasks, different postures (flexed, extended), smaller movements and larger movements, more intense and less intense muscle work as well as balancing flexibility with strength and endurance (Harding 2003). As has been seen with exercising in general, individuals with JHS need to increase activity levels more slowly to reduce the risk of flare-ups.

SPECIFIC EXERCISE

It is important that the exercises prescribed are specific and target the intended muscle, movement or task. The form of the exercise or movement is of

paramount importance. Remedial exercise becomes as much cognitive as it is physical. For the patient to learn the skills necessary to gain effective control of movement it is necessary for them to engage with their body and develop greater body awareness. Education regarding anatomy, biomechanics and how this particular exercise will help their problem, along with imagery and biofeedback, are powerful tools. Mental practice of motor imagery (mental representation of movement without any body movement) has been shown to improve motor and task performance with the greatest improvement occurring when interventions combine physical and mental practice (Dickstein & Deutsch 2007). In addition, studies have shown that motor imagery can increase muscle strength (Zijdewind et al 2003, Sidaway & Trzaska 2005). This is thought to occur as a result of neural adaptations such as degree of motor unit activation, improved coordination and decreased co-contraction of antagonist muscles (Sidaway & Trzaska 2005). Although the effects of motor imagery in JHS have not been studied clinical experience suggests it is a valuable adjunct to treatment.

While exercising they can be taught techniques to check that their postural alignment remains good, that a certain muscle is not dominant, that a weaker or more inhibited muscle is working. This can be achieved through the use of video, mirrors, pressure biofeedback and the therapist's input. However, if the patient is ultimately to be able to make use of these skills to prevent and correct problems in the future, they have to develop the ability to correct themselves.

Generic exercises do not work without good instruction and practice, because invariably an individual will 'take the path of least resistance'. Patients usually take the easy option, or if they have been in pain, long-standing compensatory strategies may inhibit weaker, less dominant or 'forgotten' muscles. The inhibition of the vastus medialis obliquus (VMO) following knee surgery is well known to physiotherapists. Facilitation is needed to 'switch' the correct muscle on, as is ensuring the reduction of swelling and pain. It is important that an assessment highlights an individual's muscle imbalance and corrects it. It may be appropriate to use manual therapy to inhibit overactive muscles and facilitate underactive ones. This then forms the exercise and is practised in a simple easy way and gradually made more challenging. Although hypermobile individuals do not appear to suffer inflammation in the same way, the principles are similar. Without EMG devices (Chapter 12.1) it is often difficult to tell which muscle is working, but palpation (therapists or patient) and feel (patient feedback) can be used successfully, as well as being critical of the form the exercise is taking from the perspective of the whole body.

In the fibromyalgia literature it is thought that 'exercises that are too vigorous might trigger immune activation with release of pro-inflammatory cytokines provoking a sickness response' (Maier & Watkins 1998), which may be a factor in the exacerbation of symptoms that JHS individuals often report. Exercise should be pain free, although a distinction is made between training pain and an aggravation of symptoms. If an exercise is painful it is either not being performed correctly or is not 'right' for that patient at that particular moment in time. In the author's experience this is particularly true for individuals with fibromyalgia and JHS and time spent modifying an exercise until it is pain-free but still achieving its aim pays dividends in terms of patient confidence and progression.

For those individuals who do not achieve a good response with the treatment approach outlined above, a multi-disciplinary pain management programme (PMP) may be required. There is, however, some evidence to suggest that patients with chronic pain, either caused by hypermobility or in the presence of hypermobility, do not respond as well as expected in a PMP. Hakim & Ashton (2005), on analysing the characteristics of those who did poorly in a PMP, found hypermobility to be a common finding. This does not appear to be an isolated case and a programme modified to hypermobile individuals is going some way to address this problem (Chapter 8).

STRETCHING

Many hypermobiles like to stretch, but have been advised not to by health professionals for fear of damaging their tissues or joints through overstretching. An unpublished audit at Guy's Hospital in 1986 reported by Harding (2003) revealed that patients with joint hypermobility found stretching helpful. This came as a surprise to the audit's authors, but has been borne out repeatedly in clinical experience. Stiffness is a common complaint, with many hypermobiles saying they 'feel like a 90-year-old', but it is not clear what produces this

feeling. We know that hypermobile joints, like non-hypermobile joints, stiffen through disuse in response to pain, but it is also possible that hypermobile individuals are more likely to use their global muscles for stability and are therefore subject to more increased muscle tension and spasm. Stretching can be a helpful antidote to this, but it is important that the hypermobile individual does not overstretch, which is potentially easy to do.

It becomes necessary to differentiate between stretching performed in order to regain and maintain muscle length, relieve muscle tension, or restore and maintain joint range, and stretching to increase an already hypermobile range of motion. It is good to stretch, but care is required. Educating an individual about how they can stretch safely without overstretching into their hypermobile or more vulnerable areas will help develop better body awareness, a skill which can be used in the future to ensure safe exercising.

EDUCATION AND ADVICE

The power of finally receiving a diagnosis after years of searching, suffering and frustration can not be overemphasized. Knowledge is empowering. It allows the patient to move from negative to more positive emotions and start to focus on ways of managing their condition, safe in the knowledge that it is not a life-threatening disorder, and that with the correct input they can make a significant change, not only to their own life, but also to the lives of family and friends.

Understanding the implications of the disorder is thought to help an individual cope more effectively with pain (Russek 1999). Education about the condition is one of the most important aspects of medical intervention and forms a major part of physiotherapy and occupational therapy input. It will generally consist of advice regarding joint care, lifestyle modifications, the judicious use of aids and supports, help with associated problems, exercise and fitness. As therapists we are usually able to give our patients more time, to listen to their individual anxieties or concerns. This helps to gain a full understanding of how the condition is impacting on their life and reassures the patient that someone is finally taking them seriously. Detailed discussion about their daily activities can identify specific problems, which, with a problem-solving approach, can be resolved, better managed and prevented from recurring.

JOINT CARE

Hypermobile individuals tend to rest at the end of their range of movements, particularly at the knees, hips and lumbar spine. Educating patients to avoid unhelpful postures such as 'W' sitting (Fig. 9.8), sitting with the leg tucked under the buttock, sitting cross-legged 'Indian style' and kneeling on plantarflexed feet or resting on the lateral borders of the feet (Fig. 9.1b) for prolonged periods will help to prevent soft tissue strain and subsequent symptoms. Further advice on joint care is given in Chapter 12.2.

Joint care for therapists

Work-related thumb pain is an occupational hazard for manual therapists around the world (Glover et al 2005, McMahon et al 2006, Campo et al 2008), and the presence of hypermobility may be a significant factor in its development (Fig. 9.9). The lifetime prevalence of thumb problems in Australian physiotherapists was 65% with a current prevalence of 41% in the observational study by McMahon et al (2006). Factors found to be significantly associated with thumb problems included thumb joint hypermobility or an inability to stabilize the joints of the thumb. Hypermobility of the carpometacarpal (CMC) joint (see Chapter 12.2) of the thumb and decreased thumb strength were significant factors

Fig. 9.8 Unhelpful postures. Sitting in a 'W'

Fig. 9.9 Hypermobility in the thumb and fingers of a therapist while performing manual therapy techniques on the cervical spine

contributing to thumb pain in another study of Australian physiotherapists (Snodgrass et al 2003) and generalized joint hypermobility (considered with a score of 3/9 on the Beighton scale) was more prevalent (although not statistically significant) in the pain group compared to the non-pain group. Interestingly, although there was no difference between the prevalence of osteoarthritis (visible on radiographs) in the pain and non-pain groups, the prevalence of osteoarthritis in females in this study was higher than in a normal population (19% compared to 2% respectively). This suggests that female physiotherapists are more at risk of developing osteoarthritis at the CMC joint than the normal population and that hypermobility (both at the joint and in general) in addition to occupation may be a contributing factor.

The type and duration of therapy is also a contributing factor. Campo et al (2008) found that manual techniques of soft tissue work and joint mobilizations increased the risk of work-related musculoskeletal disorders in the wrist and hand by 14 and 8 times respectively in those Physical Therapists in the USA who treated ten or more patients a day. It is clearly important that therapists engaged in manual therapy are made aware of the risks and encouraged to take appropriate measures. These will include modifying techniques, using equipment (massage tools, mobilization wedge), reducing hours or patients and wearing protective splints (Chapter 12.2).

Other areas of the body have the potential to cause problems in an occupation as physically active as physiotherapy. Patient transfers, patient repositioning and bent or twisted postures have been given as factors in the development of back pain (Campo et al 2008) and the hypermobile physiotherapist would be well advised to take extra care to avoid problems. As yet, hypermobility is not screened for in the student physiotherapy population, but should possibly be considered in order to highlight those at risk of problems from performing or having performed on them repeated mobilization/manipulation techniques during their under- or particularly post-graduate studies.

LIFESTYLE MODIFICATIONS

There can be specific areas of concern regarding work, home and family life. Helpful advice on activities of daily living is given in Keer et al (2003) and the issues of hypermobility and work-related musculoskeletal disorders are discussed in Mangharam (2003).

Computer use is ubiquitous, often ergonomically unsound, and frequently intense. Optimal ergonomic positioning while at the computer should be encouraged for all patients and keyboard short cuts should be used wherever possible in order to decrease mouse usage during a session at the computer.

Some general concepts that are readily accepted when using a computer keyboard and mouse are:

- keep wrists neutral
- do not rest wrists while typing
- move the whole arm while keying
- avoid stretching the fingers to reach keys that are far away
- keep fingers curved and relaxed
- use a light touch
- keep fingernails short (Stegink-Jansen et al 2000)
- avoid double clicking as much as possible when using the mouse
- position mouse within easy reach to avoid overstretching.

Specific advice regarding pregnancy is discussed in Chapter 12.6 and sport and performance in Chapter 13.

AIDS AND SUPPORTS

In general, the use of aids and supports is discouraged because it can encourage muscle weakness and dependency. However, there is a

place for supporting an area during the recovery phase of an injury with a gradual decrease in use, as discussed above. Furthermore, if there is a risk of continued strain to a joint, despite attempts to improve stability and muscle strength, there is an argument for judicious use of an aid or support. This may involve wearing a collar for travelling, to help prevent a neck strain as a result of sudden stops or humps in the road, or wearing a support on the knee to be able to take part in a particular sport. Aids and supports can be particularly helpful in the hand to enable individuals to continue to work and these are described in more detail in Chapter 12.2.

ASSOCIATED PROBLEMS

There may be other areas in the body producing symptoms as a consequence of lax connective tissue. Flat feet are common and it may be sensible to enlist the help of a podiatrist to perform a gait analysis and possibly fit orthotics. Urogenital problems, such as prolapse and urinary incontinence, may require referral to a specialist physiotherapist for a pelvic examination and advice regarding pelvic floor exercises (Chapter 12.6). Many hypermobiles suffer symptoms of irritable bowel and in cases advice on diet and reduction in constipation-inducing medication can be sufficient while others benefit from more extensive investigations and treatment as described in Chapters 6.2 and 6.3. A poor response to local anaesthetics, if present (Arendt-Niesen et al 1990, Hakim et al 2005), can be distressing for some hypermobiles when attending the dentist or obstetrician and liaison and education with other health professionals can reduce anxiety. The tendency towards low blood pressure and (pre-) syncope may be relieved by an increase in lower limb muscle work and taking in a little more water and salt, but if these measures fail, further investigation and treatment of the autonomic nervous system may be required (Chapter 6.1).

GENERAL EXERCISE AND FITNESS

Once the patient has regained a good level of function and has control of their pain, attention can move to developing a fitness programme and helping the patient return to their chosen activities and sport (Chapter 13). In general, unsupervised sessions in the gym are not advisable for the hypermobile patient, because they may over-do the exercises or do them incorrectly, or indeed exercises may take the patient into their hypermobile range and thus sessions with a trained therapist are advised initially. The therapist can teach, observe and correct the patient to ensure they can perform the exercises/tasks in an accurate and controlled way. We only retain a little of what we are taught and this lends weight to the argument of a review with the patient to check performance at a later date. Some unexpected adaptations can often be seen once patients have been left to exercise on their own. Exercising in the gym is not for everyone, so discussion with the patient may identify sports, activities or other exercise which will enable maintenance of fitness. Particularly good suggestions include Pilates, Alexander Technique and Tai Chi where the emphasis is on slow, controlled movement. Yoga is not contraindicated but care should be taken to avoid overstretching. Good instruction and supervision in the initial stages is essential. It is helpful to find an activity which is enjoyable and achievable so the patient is motivated to develop a life-long habit of exercise (Keer 2003). This subject is discussed in more detail in Chapter 13.

SUMMARY

Generalized joint hypermobility and JHS are frequently overlooked in musculoskeletal conditions, but they are not difficult to identify when looked for and should be a consideration in all musculoskeletal pain presentations. JHS requires a holistic approach in terms of assessment and management, which may involve a multidisciplinary team.

An individualized, modified, therapeutic programme is recommended, based on functional restoration with the emphasis on movement control, joint stability and self-management. Early, accurate diagnosis and effective intervention will help to prevent the downward spiral into chronic pain and deconditioning and thereby reduce suffering.

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