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Masterclass

Hypermobility and the hypermobility syndrome, Part 2: Assessment and management of hypermobility syndrome: Illustrated via case studies

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Abstract

Joint hypermobility syndrome (JHS) is a largely under-recognised and poorly understood multi-systemic hereditary connective tissue disorder which manifests in a variety of different clinical presentations. The assessment and management of patients with the syndrome is often complicated, requiring a comprehensive patient-centred approach and co-ordinated input from a range of medical, health and fitness professionals. The functional rehabilitation process is frequently lengthy, with education of the patient and family, sensitively prescribed and monitored physical therapy interventions and facilitation of lifestyle and behaviour modifications being the mainstay of the plan.

Two typical but very different case studies are presented, each illustrating key aspects of the assessment and highlighting the variety of management strategies and techniques required by therapists to facilitate successful outcomes.

Keywords: Hypermobility syndrome; Assessment; Management; Functional rehabilitation

1. Introduction

Joint hypermobility syndrome (JHS) is an under-recognised and often poorly managed multi-systemic hereditary connective tissue disorder. Because of the ubiquitous nature of connective tissue, JHS may manifest in a variety of different ways. Assessment and management of patients with the syndrome therefore requires a range of strategies and skills. This paper, which is designed to be read in conjunction with the accompanying masterclass article (Simmonds and Keer, 2007), illustrates two typical but different presentations of the syndrome.

Case study one focuses on the management of a 37-year-old woman, who at the time of writing was still under physiotherapy care. The emphasis in this presentation is on the early to middle management over 4 months of physiotherapy care which combined manual therapy and rehabilitation. It also demonstrates the value of a multi-disciplinary approach as input from a psychologist, neurologist and later a fitness instructor was also utilised.

Case study two provides an overview of the management of a 16-year-old male adolescent with JHS and marfanoid habitus. This case describes a range of key assessment techniques and highlights the role of education, goal setting and carefully monitored and prescribed exercises and functional rehabilitation. The case

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also highlights the importance of recognising JHS early on in life when lifestyle behaviours and postural habits can be more easily addressed and modified.

2. Case one: subjective examination

2.1. Current history

A 37-year-old woman, referred to as Mrs. AM, was referred to physiotherapy by a Consultant Rheumatologist for help in managing longstanding wide spread debilitating symptoms associated with JHS. Mrs. AM was married with three children (aged 12, 10 and 2 years old). In addition to caring for her family she made jewellery and hoped to be able to start a business. She had some help with housework each week.

Mrs. AM's complaints/symptoms included:

(a) low back pain, which was aggravated by standing, lying supine, sitting, bending, lifting or carrying and walking. Once she had been in a static posture for 10 min or more (such as lying or sitting) she found it very difficult to get up again;
(b) upper thoracic and neck pain, which was constant and associated with a left-sided headache and also periods of dizziness. She was unable to identify any aggravating factors, although she felt as if her head was too heavy for her neck and needed additional support, particularly when sitting. Resting helped to ease the pain. The neck pain was associated with clicking;
(c) bilateral elbow, wrist and hand (R > L) pain with occasional pins and needles bilaterally in all four fingers, aggravated by jewellery making, cooking, lifting and carrying. Clicking in all the arm joints was associated with the upper limb pain;
(d) left hip, bilateral knee (R > L), shin and left ankle pain aggravated by walking and going downstairs; and
(e) fatigue.

Generally, the pain and dysfunction had been increasing over the last 6 months and also appeared to be worse in the week before her period. She admitted to feel frustrated and depressed and had been taking antidepressants intermittently. Pain medication had been largely ineffective. There were times when she felt she could not cope with demands of family life, which included added pressure of having a son with a disability and also elderly parents in poor health living in another country.

There were also symptoms compatible with autonomic nervous system dysfunction. These included dizziness, light-headedness, lack of concentration, forgetfulness, irritability, palpitations and shortness of breath, which can be indicative of dysautonomia (Gazit et al., 2003). In order to confirm this diagnosis referral to a neurologist was made for further investigation.

Dysautonomia has been shown to be an extraarticular manifestation of the JHS (Gazit et al., 2003). This can take the form of orthostatic hypotension, which is a drop in blood pressure on changing position (sitting to standing or lying to sitting) or postural orthostatic tachycardia syndrome (POTS), which is an increase in heart rate of 30 beats a minute or more on changing position. This can produce palpitations and shortness of breath. However, the most common form of dysautonomia in JHS may be orthostatic intolerance, which occurs after a period of standing. The blood vessels in the legs dilate and blood 'pools' causing blood pressure to drop. The study by Gazit et al. (2003) showed orthostatic hypotension, POTS and uncategorized orthostatic intolerance in 78% of a cohort of patients with JHS compared to 10% of controls.

2.2. Previous history

The problem started in the low back 10 years ago during the pregnancy of her second child. This spread to involve the thoracic and cervical spines over the next few months. Osteopathy gave temporary relief, however the pattern was one of a gradual build up of pain and dysfunction. Four years ago she noticed pain spreading to her elbows, knees and lower limbs. She had been noted to be a 'bendy' child and had performed contortionist tricks and ballet. From the age of 14 she had suffered recurrent ankle and wrist sprains.

In the past, she had exercised regularly by going to the gym up to three times a week; however, she had not been attending gym for the past 6 months because of increasing neck and knee pain.

MRI of her head and neck had found no significant abnormalities, although she was told she had 'wear and tear' in her cervical spine by the rheumatologist.

2.3. Family history

Her second son, aged 10, had recently been diagnosed with JHS and was seeing a specialist physiotherapist for rehabilitation.

2.4. 24-hr pattern

She was woken from sleep by low back pain when she turned over. In the morning, she woke feeling tired and heavy in her body and noticed pain in her feet immediately on standing up. The pain and discomfort were activity or posture related and generally accrued during the day such that she had to take a rest sometime in the day.

Pain intensity was judged to be 8–9/10 on a visual analogue scale (VAS) (where 0 = no pain and 10 = worst pain ever experienced).

2.5. Key subjective features

- widespread symptoms including pain and fatigue
- moderate disability
- physical de-conditioning
- signs of ANS dysfunction
- mild levels of fear avoidance behaviour
- 'yellow flags'; anxiety, depression, stress and family pressures.

3. Objective examination

The aim of the initial examination was a global functional assessment with more detailed examination of the lumbar spine, pelvis, hips and ankle. Examination of the cervical spine, upper limbs and knees took place over subsequent appointments.

3.1. Standing posture

Mrs. AM stood in posterior pelvic tilt, with decreased weightbearing on the left leg, hyperextension of the knees (R > L) and so effectively 'hanging on the right hip'. Her feet flattened on weightbearing and there was increased pronation in the hind and mid-foot. Her skin was soft and showed marked stretchiness in the phase of taking up slack, as tested by picking up the skin on the back of the hand (Simmonds and Keer, 2007). A number of small paper-thin scars and striae atrophicae were noted. These had been present since the age of 12 and not connected with pregnancy or change in weight.

3.2. Lumbar spine movements

Forward flexion restricted by pain in the posterior legs and low back. Mrs. AM failed to reach 90° hip flexion, bending forward by over flexing in the lumbar spine. Return from flexion was achieved by extending the lumbar spine.

Extension showed a good range of motion with pain at the end of range.

Side flexion showed normal range and quality of motion with no pain.

3.3. Thoracic spine movements

Restricted movement into extension and rotation with pain on left rotation.

3.4. Cervical spine movements

Fluctuated between a full range of movement with no pain and moderate restriction, affecting rotation (left > right) and left-side flexion particularly associated with neck pain.

Forward bending of the spine is one of the manoeuvres in the Beighton score, used to identify hypermobility. Mrs. AM could only reach as far as 3 in below her knees at the time of testing, although in the past she could easily touch the floor with both palms. She scored positively for the other manoeuvres in the Beighton score and so overall her score was 8 (historically 9)/9. Outside the Beighton scale, other joints including other fingers, thumb, toe and shoulder joints were also hypermobile. She conformed to the 1998 Brighton Criteria for JHS, which is equivalent to the Ehlers–Danlos syndrome hypermobility type, formerly EDS III (Grahame et al., 2000).

3.5. Sitting posture

Mrs. AM sat in posterior pelvic tilt and slouched into flexion of the whole spine. In an attempt to find stability she crossed the right leg over the left such that the right ankle rested on top of the left knee.

3.6. Sit to stand

She used her hands to push up and started the movement with lumbar spine flexion rather than hip flexion.

Single leg standing highlighted poor control, with the right sacro-iliac joint unlocking on weight bearing in a trendelenberg pattern. Weight bearing on the left leg was achieved with hip hitching on the right. Hip flexion in standing was difficult on the right with spinal flexion occurring and considerable difficulty in lowering of the leg due to poor eccentric control.

3.7. Lying supine

She disliked this position as it produced pain in the back and pelvis, and after 10 min (or more) the whole area felt locked and difficult to move.

Active Straight Leg Raise Test (*ASLR*) (Mens et al., 1997) *supine*: Lifting right leg difficult, leg heavy. Compression (ASIS) lifting leg much easier.

3.8. Peripheral joint testing

Ankle: The left ankle was restricted in all directions except eversion by pain and tightness in the lateral lower leg.

3.9. Muscle testing

There was an inability to activate the pelvic floor or transversus abdominus (assessed by palpation). Attempts at drawing in the abdominal wall and lifting

the pelvic floor were made by bracing and breath holding. Weak gluteal muscles.

3.10. Passive physiological movement testing

Hip: Right hip rotated into flexion late.

3.11. Accessory movement testing and palpation

Spine: Tender, but mobile all levels of the lumbar spine. Joint stiffness in the left costotransverse and intervertebral joints T2–7 and stiffness at left C2/3 joint.

Decreased multifidus bulk, decreased gluteal tone and overdevelopment and increased tension in rectus femoris, tensor fascia latae, adductors and piriformis.

Ankle: Decreased antero-posterior glide of talus at talo-crural joint. Increased tension in lateral calf musculature.

3.12. Neurological assessment

There were no neurological abnormalities detected.

3.13. Key objective findings

- widespread joint hypermobility
- poor movement patterns using end of range positions
- poor load transference through the pelvis
- overuse of mobile lumbar spine
- movement restriction in thoracic and upper cervical spines with overuse of mobile mid cervical spine
- inability to flex right hip to 90°
- decreased weightbearing through left leg due to ankle dysfunction
- ineffective trunk stability strategies.

3.14. Goals

- to reduce pain to a manageable level
- to return to normal life
- to return to the gym to improve fitness and lose weight.

4. Management

Initially, treatment involved a combination of different modalities and focused on the primary area of dysfunction. Manual therapy was performed on the tense overactive muscles around the right hip and sacro-iliac joint to improve/restore normal movement. A pelvic Com-Pressor belt (Lee, 2003) was used with extra support across the ASIS to help support the pelvis while the patient developed an effective trunk stabilising pattern. Trunk stabilisation involved re-education of lateral costal breathing and isolation of low effort pelvic floor, transverses abdominus and multifidus muscle contraction. This was started in side lying and progressed, once it was effectively being performed, to more functional positions such as sitting, standing and during everyday activities.

Manual therapy was also applied to the left ankle. This took the form of joint mobilisations to the talo-crural and subtalar joints in combination with muscle release work to the calf and peronei to restore normal biomechanics and mobility. This was followed-up with exercises to re-educate efficient load transference through the ankle and foot and included heel raises and deep knee bend in standing.

Posture re-education and joint awareness (pelvis, hip, knee and ankle) were practised in different positions using biofeedback (mirror work, still photographs and video) and progressed using a 'sit-fit' to provide an unstable surface initially when sitting and as core stability and postural control improved this was progressed to standing.

Joint mobilisations were also applied to the thoracic and cervical spines in combination with muscle release to the erector spinae. The patient found exercising on a half roll (see Fig. 1) (using effective postural stabilising muscle activation) very effective at releasing muscular tension and improving thoracic mobility. Clinically, it appears that hypermobile individuals frequently overuse the global muscle system (Bergmark, 1989) and have difficulty recruiting the local postural muscle system. It was helpful therefore to inhibit or 'switch off' the dominant global muscle (in this case erector spinae) in order to improve recruitment of the trunk stabilisers, in particular multifidus.

During one session, Mrs. AM complained of acute pain in her right thumb and radial side of the wrist. The pain had started 2 days previously after lifting her handbag from the seat of the car. Examination of the way she lifted her bag identified the lift occurring in ulnar deviation thereby putting strain through the

Fig. 1.

Fig. 2.

Fig. 3.

lateral side of the wrist and thumb (see Fig. 2). The muscles which produce radial deviation and hold the wrist in a more neutral position were inactive. Once this was pointed out to Mrs. AM (with the use of mirrors and video) she was able to correct the movement (see Fig. 3), quickly learning to lift objects maintaining the wrist in neutral and instantly abolishing the pain. This was followed up with advice and guidance on exercises to strengthen her wrists and fingers. Therapeutic putty was used to practice finger joint control, encouraging a good pattern of co-contraction around the finger joints during different finger, thumb and gripping movements with attention to maintenance of a neutral wrist posture.

4.1. Comment

Finding a way that the patient can instantly change or 'switch off' a pain is a great motivator for producing a change in movement patterning and can be utilised in the clinic to facilitate permanent behaviour change. Acute onset of pain through soft tissue strain is often easier and quicker to affect through movement change, whereas long standing chronic pain does not always respond in such a quick and clear cut way.

Sessions were also used to discuss and address Mrs. AM's fears and improve her understanding of the condition (Russek, 2000). Mrs. AM was particularly concerned as her son was also affected. Topics included pacing of activities, general information on joint care and problem solving. An example in this case involved re-education of lifting and carrying in order to reduce pain and strain, as discussed above. Working with the patient to resolve particular problems which they may be experiencing helps to give them confidence to return to activities that may have caused problems before and help them return to normal activities. Encouraging a healthy lifestyle with attention to good posture and joint control, avoiding prolonged periods of static postures or repetitive activities and developing a life-long habit of regular exercise to maintain general fitness are all important to enable the patient to have the confidence to manage the condition themselves.

Mrs. AM also attended regular counselling sessions with a psychologist, which helped to deal with the stress and pressures in her life.

4.2. Status at 4 months

After 4 months of attending physiotherapy (initially twice a week and then once a week reducing to once every 2 weeks), the patient reported that two of the goals had been achieved. There had been a significant reduction in pain (VAS 2–3/10), such that pain was now manageable and she had returned to her normal life activities. She felt ready to return to the gym, which she intended to do after the school holidays. In addition, she reported feeling less tired, not needing to rest everyday and better able to stand, sit and walk for longer periods without symptoms. She no longer used the compressor belt. The back, hip, knee, and ankle pain and the elbow, wrist and hand pain had resolved, although she was still troubled by intermittent neck stiffness and pain on turning to the left. This appeared to be related to the amount of activity she had undertaken. Pain in her feet on standing in the morning had also resolved.

Objectively, Mrs. AM demonstrated improved trunk stability and joint control. She had changed her sitting posture and no longer rested in a posterior pelvic tilt position. She had a full range of forward trunk flexion and was able to touch the floor with her hands. Movement patterns, when returning from flexion to the upright position and on moving from sitting to standing were fluent and efficient. The right sacro-iliac joint was more stable on single leg standing and she was able to flex and extend the right hip without discomfort.

Results from investigations of the autonomic nervous system by the neurologist showed Mrs. AM to have low blood pressure averaging 80/55 on supine testing.

Recommendations were to increase her fluid and salt intake and to improve the muscle pump effect from the lower legs through exercise. Following her holiday it is proposed that she will also start on medication.

Mrs. AM was ready to return to the gym with the aims of maintaining her joint mobility and muscular strength and improving her cardiovascular fitness. Liaison with a fitness instructor at the gym in order to discuss the specific needs of the patient in view of her problem areas and the influence of joint hypermobility was an essential part of the plan. A review appointment, prior to attending the gym, with a further review was suggested 3 months later.

5. Concluding discussion

This case study illustrates a typical presentation of a patient with JHS. People with the condition, may be aware of having been 'bendy' or more flexible in their youth and also may have developed intermittent joint aches and pain which subsequently have become more persistent. In the early stages, they may well have been more able to cope, as in Mrs. AM's case, however, frequently an incident or trauma triggers an exacerbation which progressively gets worse and affects many other areas, rather like a 'domino effect'. In this example, as with many women, it is the effects on the body of pregnancy, childbirth and then looking after young children that bring the patient to the health practitioner desperate for help (Gurley Green, 2001). Some patients unfortunately report that physical therapy has often had little beneficial effect and at worse resulted in deterioration (Keer, 2003). A full understanding of the condition and the presentation is necessary to avoid a poor result, which often leads to the patient 'therapist shopping', getting more frustrated and depressed with the problem becoming progressively more chronic. The approach therefore needs to be holistic, patient centred, specific and aimed at giving the patient the tools to manage the problem themselves.

6. Case two: subjective examination

6.1. Current history

A 16-year-old high school boy, referred to as MH, was referred by a Consultant Rheumatologist for rehabilitation following the diagnosis of JHS. Like many hypermobile people, MH had performed contortionist tricks during his childhood and his parents reported that he had complained of long standing intermittent musculo-skeletal pain from an early age. Of note was the substantial rate of growth of 10 cm over the previous 12–18 months and the potential for this to impact on joint biomechanics.

MH reported a history of increasing low back pain over the previous 2 years with intermittent radiation into the right and left posterior thighs. The back pain had been further exacerbated in a school physical education session, which involved cross-country running with a pack on his back. MH also reported anterior knee pain, ankle instability and low grade neck pain, all common sites for the manifestation of JHS in the adolescent population (Middleditch, 2003). Understandably, MH and his parents were anxious and concerned about the impact of hypermobility on long-term health, fitness and quality of life.

6.2. Social history

MH's parents were not aware of other family members being hypermobile. However, MH's father, who was present at the initial assessment, was observed to have hyperextending 1st metacarpo-phalangeal joints and he also reported longstanding persistent low back problems, indicating aspects of hypermobility and potentially JHS.

6.3. Hobbies and sports

Usual activities included playing competitive club level squash and recreational cycling. MH had reduced his involvement in physical activity as a result of his pain.

6.4. Lumbar spine

MH reported constant low back pain with radiation into the posterior thighs. On the VAS pain ranged from 4/10 on a good day to 7/10 on a bad day. Pain was aggravated by running and sustained period of sitting. Pain sometimes took as long as 2 days to settle. It was generally worse in the morning and eased during the day. Occasionally, the pain increased during the day and this was related to the amount of physical activity undertaken or time spent sitting and studying. Pain did not wake him at night. He slept on a very firm single bed mattress with one large pillow.

6.5. Knees

Knee pain was mildly—moderately irritable, and was described as an intermittent low grade dull ache, occasionally sharp and localised over the anterior, inferior aspect of the patella (VAS 3/10–5/10). The pain was aggravated by stair climbing, squash and sustained sitting. It was relieved by rest, but could take between 2 h and 2 days to settle.

6.6. Neck

Neck pain was also intermittent, superficial and low grade (VAS 2/10–4/10). This pain was aggravated by sustained postures, particularly relating to long periods of studying. Irritability of this pain was considered mild as neck pain was generally relieved with rest overnight.

6.7. Ankles

MH described his ankles as unstable as they frequently "gave way" particularly when fatigued and when running and changing directions in squash. His ankles were not painful.

6.8. Neurological assessment: NAD

6.8.1. Special questions

General health: MH had no recent illnesses or surgery.
Investigations: recent echocardiogram NAD.
Med: nil.

6.8.2. Key subjective features

- widespread pain and/or dysfunction including lumbar spine, cervical spine, knees and ankles
- reduced levels of physical activity—leading to de-conditioning
- significant growth and development leading to potential joint biomechanical changes and imbalances
- anxiety of both MH and his parents regarding the long-term effects of hypermobility syndrome on health, fitness and quality of life.

7. Objective examination

The plan for the objective examination was to assess global functional activity and the extent of the tissue laxity and then to examine the specific presenting complaints.

7.1. Observation

MH was tall and slim with ill-defined musculature. In standing, his chin was protracted and his glenohumeral joints were anteriorly positioned. Furthermore, he had a flattened thoracic kyphosis, a hyperlordosis between L3–S1 and his scapulae were protracted with slight winging on active shoulder elevation. In sitting, MH adopted a slumped position with his pelvis in a posterior pelvic tilt. This position further increased the protracted position of his chin and the anterior position of the humeral head in the glenoid fossa.

7.2. Sit to stand

Recruitment of the vastus medialis muscles was latent on sit to stand, left more so than right. MH quickly adopted a hip hitch, hip hanging posture and locked one knee into hyperextension once in standing.

7.3. Gait analysis

MH had a mild trendelenberg gait pattern and intermittently flicked his knees into hyperextension when walking. He had bilateral pes planus with flattening of the medial arches and excessive pronation of the subtalar joint on weight bearing. These features were corrected when wearing sports footwear which provided good hind and mid foot control.

7.4. Joint and soft tissue assessment

There was evidence of widespread joint laxity and he scored 6/9 on the Beighton scale. Outside of the scale, other joints including the glenohumeral joints, metacarpo phalangeal joints and the interphalangeal joints of the fingers and toes also displayed excessive ranges of movement. He had elongated fingers and toes (arachnodactyly) a feature of the Marfanoid Habitus (Grahame, 2003) confirmed by a positive Steinberg's test which involves instructing the patient to fold his thumb into a closed fist (Staud, 2005). This test is positive if the thumb tip extends beyond the palm of hand. MH had 1 cm of skin laxity when the skin was drawn up above the 3rd metacarpal of the hand. His small pox and BCG scars were thin and papery. Additionally, he had striae atrophicae across the lumbar region of his back and a high arched palate and absence of the lingual frenulum. These findings conformed with the 1998 Brighton Criteria for benign joint hypermobility (equivalent to Ehlers–Danlos Hypermobility Type III with marfanoid habitus), as described by Grahame et al. (2000).

7.5. Lumbar spine

Lumbar spine flexion was restricted by tightness in the hamstrings and pain in the lumbar spine. The tips of fingers reached the mid tibia (usually MH could touch the floor). There was reduced intervertebral movement between L2–S1 on flexion and visible muscle tightness of the paraspinal muscle groups in the lower lumbar region. Lumbar spine extension was restricted by pain to 20° (MH reported usually being able to double that range). Observable hinging and increased intervertebral movement was observed between the L4/5 and L5/S1 motion segments.

Lumbar side flexion testing showed a similar movement pattern with increased movement in the lower lumbar segments and stiffness in the upper lumbar segments.

On palpation, the lower thoracic and upper lumber accessory intervertebral movements were stiff in all directions, while the lower lumber segments were painful and more mobile on palpation with an "empty" end feel. Erector spinae muscles in the region felt "ropey" when palpated and there were a number of active trigger points in these muscles.

7.6. Muscle function and testing

MH had difficulty recruiting and sustaining contractions of his pelvic floor muscles and transversus abdominus when assessed in crook lying and four-point kneeling. Delayed recruitment and poor endurance and strength of the gluteal muscles, particularly gluteus medius was evident when performing external rotation of the hip in side lying. This weakness was highlighted in walking where it had been noted earlier that MH walked with a mild trendelenberg gait pattern.

7.7. Cervical spine

Range of movement was "normal" for MH in all directions except for upper cervical and lower cervical flexion, where the para-vertebral muscles were observably tight and appeared to restrict range of movement. Palpation revealed very mobile, mid cervical intervertebral movements with the upper cervical tissues and intervertebral muscles and tissues feeling "thick and boggy". Sternocleido mastoid (SCM) and the upper fibres of trapezius were tight and there was palpable spasm and active trigger points in these muscles.

7.8. Muscle function

MH had difficulty recruiting the deep neck flexors and his SCM muscles were very over active. Active movements of the neck and shoulders highlighted increased activity of the upper trapezius and SCM muscles with associated shoulder girdle elevation and poor scapulo-humeral rhythm.

7.9. Lower limb

The lower limbs were generally well aligned, although the patellae were positioned slightly laterally and superiorly. Timing of the VMO was latent on sit to stand and he quickly snapped into hyperextension. Repeated one legged squatting and deep knee lunges reproduced anterior knee pain symptoms.

Ankle inversion and plantar flexion movements were excessive on both active and passive movement of both ankles. Medial arches were flattened on weight bearing and restored when unloaded.

7.10. Balance and proprioception

Standing stalk test with eyes open and closed revealed increased perturbation especially when standing on the left foot. MH frequently over shot markers when asked to hop onto designated points on the floor. The ability to reposition the lumbar spine into the neutral position in four-point kneeling was also poor.

7.11. Key objective findings

- widespread joint hypermobility, tissue laxity and pain
- poor posture and altered movement patterns using end of range positions
- overuse of the lower segments of the lumbar spine
- protective spasm and active trigger points in the SCM, trapezius, thoracic and lumbar erector spinae muscles
- poor recruitment, timing, endurance and strength of local postural and stabilising muscles in the neck, trunk and lower quadrant
- Poor lumbar spine and lower quadrant proprioception.

8. Management

The initial management strategy aimed to address the anxiety of MH and his parents with regard to his condition and then to agree to short- and long-term goals.

Discussions were had with MH and his parents at the beginning of the rehabilitation programme and at intervals throughout the process in order to alleviate anxiety and provide reassurance regarding the long-term prognosis. It was particularly important to discuss the nature of the condition in the light of his stage of growth and development and the implications of JHS on his involvement in sport and the impact on study habits and home ergonomics. There were also discussions about the importance of changing postural habits and improving stability, strength and endurance through a functional rehabilitation programme and long-term commitment to exercise and physical activity.

8.1. Short-term goals

- improve posture and body awareness
- reduce low back, thigh and knee pain
- improve upper and lower quadrant muscle strength — endurance and functional stability.

8.2. Long-term goals

- improve cardiovascular fitness and sports specific functional capacity in order to enjoy an active

sporting lifestyle and prevent further pain and injuries
- return to playing regular squash and other recreational physical activities
- adopt a lifelong commitment to exercise and maintain fitness in order to reduce the risk of injury and long-term pain.

8.3. Initial stage

Initial treatment sessions consisted of postural re-education and developing lumbar control through functional activities such as sitting and standing as advocated by Middleditch (2003). This was then progressed to activities such as sitting to standing, walking and bending over with postural control to pick things up. Swiss ball work was also introduced in the early sessions. These activities were complemented by manual therapy techniques such as gentle mobilisations of the upper lumbar and lower thoracic motion segments and soft tissue release of tight thoracic and lumbar para-spinal muscles. MH was also taught the concept of pacing activities, particularly with respect to the long periods of time he spent studying. Ergonomics relating to his desk, position of his computer and chair were also discussed.

Carefully prescribed proprioception exercises and strength-endurance exercises training were then gradually instigated to address trunk and lower quadrant stability. Trunk stability training included: training transversus abdominus, the pelvic floor muscles, the gluteal muscles, the lower and middle fibres of trapezius, multifidus, serratus anterior and deep neck flexors mainly in functional positions, i.e. sitting, standing and four-point kneeling. Furthermore, the poor timing and endurance of vastus medialis and gluteus medius were facilitated in stride standing, mini squats and sit to stand. Like many hypermobile people, MH found isolating and recruiting individual muscle groups difficult and therefore sensory feedback including careful hand placement over specific muscle groups and around joints, PNF patterning, tape and mirrors were all important to help facilitate muscle activity and appropriate motor control and control of movement. Exercises were initially directed towards training in the inner to middle joint range and were gradually progressed into the outer range where the joints were less stable due to muscle weakness and altered proprioception.

8.4. Middle stage

Once the lumbar symptoms had begun to settle and a degree of trunk stability had been achieved, a progressive functional rehabilitation circuit training programme was introduced. Exercises in the circuit were initially mainly closed chain in order to enhance proprioceptive feedback and assist with control (Fig. 4). Later, open chain multi-directional exercises were incorporated in order to train more functionally. Stationary cycling and treadmill walking were also included in the rehabilitation programme in order to address aerobic components of fitness.

During the middle stage of the programme, the main aims were to develop strength-endurance and to improve motor control, proprioception and improve function. Exercise prescription was carefully monitored and progressed on a week-by-week basis and exercises were prescribed at a level where a minimum of two sets of eight exercises could be achieved with good quality movement and without causing pain. Repetitions were also gradually increased to a maximum number of 25 and sets increased to three to target the endurance element. MH attended the clinic on average, once a week for a further 14 weeks. These sessions were used to monitor the quality of the exercise, further progress of the programme and treat any symptoms that arose as a result of the exercises.

8.5. Final stages

During the final stages of the programme, sport specific functional rehabilitation for squash was

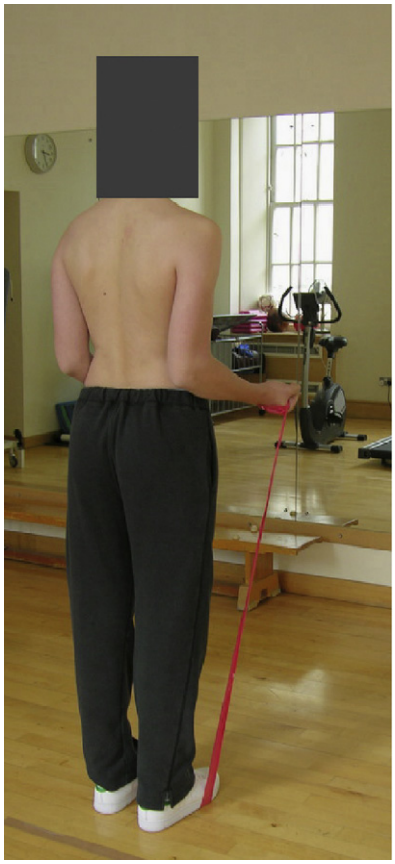

Fig. 4.

addressed by incorporating specific shadowing drills, running and cutting drills, progressive lower limb plyometric exercises and racquet activities on the minitramp to challenge proprioception (Fig. 5).

8.6. Comment

Functional rehabilitation was based on the principles of skill acquisition and motor learning and included concepts of attention focus, self-control/self-efficacy and training in dyads as described by McNevin et al (2000). Also underpinning the rehabilitation were the physiological principles of training (Wilmore and Costill, 2004; Arnold and Gentry, 2005). Improving proprioception is considered to be a key element for the amelioration of symptoms in this patient group (Ferrell et al., 2004) and therefore was a key component in the programme. MH was encouraged to use a training log with personal targets to assist with motivation. Exercises during the final phase was aimed at improving higher level function, therefore it was important to monitor quality of movement and to apply the overload principle in order to stimulate training gains and mimic the demands of squash and other higher level activities.

Fig. 5.

8.7. Discharge and discharge planning

MH was discharged from regular physiotherapy rehabilitation sessions after 17 weeks. He described himself as 90–95% better and he felt generally stronger and more physically stable and able. His back pain had almost completely resolved (VAS 0–2) and his knee pain was negligible. The quality and control of his spinal and lower limb movements were much improved, although it was noted that when he was fatigued, old patterns re-emerged. He had returned to playing squash on a recreational basis with plans to compete in the following month. He was also cycling recreationally with no ill effects.

MH was advised to continue with a maintenance programme of exercises. The discharge plan included a 1 and 4 month review. With consent from MH and his parents, the school physical education teacher and squash coach will be informed in order to discuss the key issues associated with hypermobility, particularly in relation to reducing the risk of injury and prudence with regard to sudden increases in physical workload.

9. Concluding discussions

This case study, reports the assessment and physiotherapy management of an adolescent young man with hypermobility syndrome and marfanoid habitus. Following a thorough examination, discussions with parents, medical consultant and MH, a shared management plan was developed and implemented. It should be highlighted that there were set backs in MH's progress and that this is a common pattern when managing people with this syndrome. Of particular note were transient joint and soft tissue reactions in the knees and elbows, particularly in the middle stage of rehabilitation when increased load was applied to the joints and soft tissues. These symptoms were addressed by altering the mechanics of the exercise, i.e. avoiding hyperextension of the joints and reducing load either by reducing the lever arm or reducing the resistance and applying tape to assist with proprioception. This case report provides an example of a progressive functional rehabilitation programme implemented at an important developmental stage. Manual therapy in conjunction with clinically reasoned functional rehabilitation and implementation of appropriate behavioural strategies will hopefully lead to long-term amelioration of symptoms and effective self management.

10. Summary

Treating and managing patients with JHS can often be slow and very challenging. A patient centred holistic

approach is recommended. The process frequently involves communicating and co-ordinating input from the multi-disciplinary health and fitness team. Education regarding the nature of the condition and the implications for long-term health, quality of life, sport and physical activity involvement, ergonomics and work routines is an important aspect of the process. Facilitating lifestyle and behaviour changes and carefully administering a range of therapeutic interventions, including manual therapy and functional exercise therapy are central to the management plan.

These two case studies illustrate common, but very different clinical presentations of JHS. They highlight the application of key diagnostic criteria and assessment procedures used in the patient examination. They also provide examples of some of the strategies and skills required by therapists to successfully rehabilitate people with the syndrome.

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