Physical therapy for hypermobility

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Principles of management

There have been very few treatment intervention studies undertaken to date. Research by Barton and Bird reported improvements in joint stability and body pain with strengthening and stability exercises. Likewise, Kerr et al reported a good response to a progressive six-week exercise programme in a retrospective study of 39 children with joint hypermobility syndrome (JHS). Furthermore, Ferrell et al reported significant improvements in proprioception and pain with an eight-week programme of closed chain and proprioception exercises for individuals with hypermobile Ehlers-Danlos syndrome (hEDS) / JHS aged between 16 and 49 years. Other authors have reported improvements with graduated exercise programmes combined with education, behavioural and lifestyle advice.

Because of the ubiquitous nature of collagen, hEDS will present with a variety of different signs and symptoms. Therefore current best practice management of hEDS is essentially an individualised problem-solving approach. A multidisciplinary approach to rehabilitation is recommended, including occupational therapists, podiatrists, physical therapists, osteopaths, sports therapists, nurses and psychologists depending on the individual's needs. Physical education teachers, sports coaches, school nurses, music and dance and classroom teachers all need to be involved in the wider management plan, especially where children and adolescents are concerned.

Principles of management include:

- Treating the treatable, for example acute soft tissue lesions and injuries.
- Relieving pain where possible through the use of soft tissue work, gentle mobilisations, electrotherapy and support of joints and tissues.
- Education and behaviour modification to enable individuals to manage the condition with minimal reliance on medical input or medication.
- Liaison with family and other associated professionals where appropriate, e.g. occupational health, school teachers, school nurses, occupational therapists etc.
- Improving the endurance and strength capacity of the postural support and joint-stabilising muscles.
- Improving balance and coordination.
- Improving stamina and general fitness.
- Re-educating posture and gait to avoid or correct abnormalities in biomechanics.
- Facilitating a return to normal activities and functioning and promoting an active lifestyle.

These aims work together to ensure that the individual has improved functional capacity, spatial awareness, joint stability and control, which facilitates self-management, independence with a minimum of external support.

Pain relief

It is important for both individual sufferers and the family of sufferers to understand that the pain they are experiencing is due to the hypermobility and associated musculoskeletal insufficiencies and not to any other pathology such as an inflammatory arthritic condition. It is then easier to understand why a rehabilitation programme is the treatment of choice. Individuals can be reassured that the pain will ease, but only when the muscles are strong and fit and are protecting the joints more fully. It is often found that the pain is the last thing to improve and only does so slowly, and this should be emphasized at the start of the programme. It is important to realize that the pain does not signify damage or primary inflammatory arthritis, but indicates soft tissue sprain- and strain-type injury due to poor control of the joints. This is usually benign and self-limiting if managed well.

As an adjunct to exercise, other methods of pain relief include hot packs or cold packs on specific joints. Transcutaneous electrical nerve stimulation (TENS) machines may also have a role to play in this aspect. Frequently hypermobile individuals will present with a mixture of hypomobile (stiff) and hypermobile segments or joints. Manual therapy including mobilisations of stiff hypermobile joints can be helpful, as can soft tissue massage, trigger point work and myofascial release to alleviate pain associated with muscle spasm. Relaxation and visualization techniques can help the individual to manage pain and can be very useful at night-time if they have difficulty sleeping due to pain and discomfort.

Pacing and monitoring of activity is an extremely important part of pain management. It is often the case that individuals will report pain after activities such as vacuuming the house or sitting for prolonged periods at work. This may result in an increase in pain over the next day and not feeling well enough to go to work, or feeling the need to rest. This settles and the individual recovers enough to be able to do many activities again with the same result and the cycle continues. This causes constant ‘peaks and troughs’ of symptoms and causes a major disruption to life. The idea of pacing is to stop this rise and fall of symptoms by levelling out activities and gradually building them up again. The key is to balance out the activities so the amount done on each day is not enough to cause a flare-up. It is better to vary the activities, trying not to spend too long in any one position or activity. If necessary an activity can be repeated a few times a day rather than being done all in one go. The amount of time spent on each activity is then gradually increased weekly until they are able to achieve functional activity goals and resume normal life. For further help with pacing see

http://www.med.umich.edu/painresearch/patients/Pacing.pdf

Posture and motor control

Suboptimal posture, both static and dynamic, is a common clinical finding in hEDS individuals and recent research has suggested the trunk is most affected. An individually tailored posture re-education programme showed good improvement in pain and quality of life.

Improving trunk stability is often the most appropriate starting point for a rehabilitation programme because efficient trunk stability is required for effective peripheral joint control. Patients with recurrent low back pain have been shown to have an altered postural strategy whereby activation of the deep postural stability muscles which support the spine during movement are delayed and may produce pain as tissues are overloaded through lack of muscular support. This change in muscle activation can produce functional changes in the representation of affected muscles on the somatosensory cortex in the brain. But because the brain is 'plastic' these changes, associated with pain, can be reversed by motor-skill training.

Motor-skill training is an important part of the rehabilitation process and Boudreau and colleagues suggest several components to maximise success:

- Exercises should target a specific component of movement which requires greater skill and precision.
- Motor-skill training should be pain-free. Pain rapidly alters the excitability of the motor cortex and contributes to protective strategies and so hinders learning.
- Rehabilitation exercises should be goal-orientated or 'cognitive' to enhance brain changes.
- Quality is preferable to quantity to prevent fatigue and pain interfering with improvements in task performance.

Research has demonstrated that specific, isolated, low level, skilled stabilisation training is preferable to non-isolated functional exercise and can restore the timing of activation of postural muscles to near-normal levels. These changes can occur relatively quickly with instruction and practice, can lead to brain pathway reorganisation and motor learning and be transferred to functional activities.

Muscle strengthening

Individuals often present to clinic in a physically deconditioned state resulting from reduced physical activity due to longstanding pain from recurrent injury or postural misuse. Muscle weakness is a common clinical finding, particularly in the presence of pain, and has been found to occur in the knee extensors and to a lesser degree in the knee flexors of adults with hEDS. In addition, there is some evidence to suggest that hypermobiles need to work harder and use a different strategy to stabilise the knee in quiet standing and with more challenging tasks there is less use of the gluteal muscles.

Joint stability, muscle strength and improving dynamic muscle control to supplement the ligamentous insufficiency should help to support and minimize trauma to joints. It can be useful, especially if the individual is experiencing significant pain, to start with static exercises in the hypermobile range before progressing to dynamic work and then on to resisted work. The exercises may progress from non-weightbearing to weightbearing work and hydrotherapy can be a useful adjunct. Hydrotherapy can also be useful if weightbearing and land based exercises prove difficult or too painful. The buoyancy of the water helps to support the body and the warm water helps to relax muscles making movement easier.

External support

As a rule the use of aids and supports is discouraged as it can exacerbate muscle weakness and promote dependency. However, there is a place for the judicious use of support in certain circumstances. During the acute phase of an injury support in the form of a brace or tape can be helpful to support a vulnerable joint to allow movement and also to help facilitate proprioception, healing and postural control. Aids and supports can be used to allow the wearer to partake in a specific activity such as sport, gardening, or playing a musical instrument, which would not be possible without the support. Equally prudent use of devices such as pen grips can be a good adjunct to a hand-muscle strengthening programme, as they reduce the force required to sustain the gripping of a pen, therefore reducing the pain and fatigue experienced in fingers and wrists during school work for example.

Gait rehabilitation and functional rehabilitation

A combination of hypermobile joints, reduced proprioception, altered motor control, weak muscles and reduced stamina can profoundly affect gait. Individuals with hEDS often adapt to their hypermobility by altering the mechanics of how they function. This can lead to overloading in some areas of the body with increased pain, pain in other locations and fatigue. To correct this, the causes of the abnormalities need to be identified and worked on separately, alongside the hypermobile individual recognizing the abnormalities and trying to correct them. The use of video recording and a mirror can aid this and give the individual helpful visual feedback as they work on improving their gait and posture. It is important to work on specific functional activities and to develop with the individual energy-saving, biomechanically correct, safe and pain-free ways of approaching these actions. Practising these movements can be included in their rehabilitation programme and can be as easy as step-ups and step-downs on a stair and/or repeated sitting to standing from a chair.

Individuals frequently present with pronated, flat feet on weightbearing as a result of their hypermobility and this can contribute to lower limb symptoms and an altered gait pattern. This can be improved through exercise and by choosing sensible footwear which supports the foot and has some shock-absorbing qualities. The characteristics of the ideal shoe are seen in the more solid types of trainer with a strong heel counter, robust fastenings to support the midfoot and a cushioned sole. If these measures are insufficient the hypermobile foot may respond well to functional insoles or orthoses, either over the counter or bespoke. These may take the form of a heel cup or arch support that will support the position of the hindfoot or the medial arch respectively, helping to correct the foot position. It can be argued that this may be encouraging weaker foot muscles, but in practice, the benefits of correcting the biomechanics of the foot (through exercise and/or insoles) can have a positive effect upon the whole gait pattern as it reduces the abnormal forces throughout the foot and the other joints of the kinetic chain, therefore reducing the pain.

Balance and proprioception

Given that proprioception and balance deficits are common in hEDS, techniques related to these problems should be incorporated into fitness and rehabilitation programmes. Landmark work by Proske and Gandevia has emphasised the importance of skin in proprioception and

kinaesthetic sense. Because of the skin laxity, we recommend enhancing sensory input via the skin by the use of 'hands on' movement facilitation by therapists, the wearing of tight fitting clothing and neoprene gloves and the use of tape during specific exercise or functional rehabilitation sessions. 'Rhythmical stabilisations' are a useful method to improve postural stability, both globally and specifically. Balance and proprioception as well as pain, muscle strength and quality of life were significantly improved by an eight week programme of closed chain exercise (weightbearing) combined with practice on a balance board. These simple exercises include squats, plié, bridging, four point kneeling, standing on one leg and progressing on to more dynamic balance activities using a balance (wobble) board, foam roller and Swiss ball to provide a further challenge. Even more striking than the improvement in symptoms, was the finding that the reflex associated with the quadriceps muscle, found to be absent or diminished in 50% of hEDS individuals prior to the programme, was now elicited in all participants following the programme. This is thought to be due to improved motor neurone activation at a spinal level and suggests plasticity in the spinal circuitry.

Psychological support

In some cases where pain and loss of independent function are seriously impacting on quality of life it can be helpful to enlist the help of a clinical psychologist. The reason for this is that pain can change based on what we think, how we feel as well as what we do. There are numerous psychological techniques, which can help individuals to change their relationship with pain helping them to cope better. These include learning effective pain management strategies, learning strategies to reduce stress, anxiety and worry, improving sleep as well as relaxation techniques and mindfulness. Many find this support invaluable and are able to change many unhelpful coping strategies into helpful ones and are then able to leave behind the 'chronic pain cycle' that may have developed in the preceding phase. There is useful information on the internet to help individuals work on some of these strategies themselves at:

www.moodjuice.scot.nhs.uk/chronicpain.asp

www.breathworks-mindfulness.co.uk

www.painsupport.co.uk

www.painconcern.org.uk

General fitness

Individuals with hEDS often become more sedentary due to their pain and muscular weakness ensues, which develops over time resulting in the individual becoming generally de-conditioned and lacking in general fitness. It is therefore important to incorporate some aerobic fitness work into the rehabilitation programme to improve cardiovascular health and improve energy and stamina. Care needs to be taken early on when there is still suboptimal muscle strength to ensure that the fitness aspect of the programme is of low impact to the joints and does not increase symptoms or cause a flare-up. This can happen if the programme is progressed too fast or with too much vigour. Aquatic graded exercises, swimming and deep water running are desirable methods of exercise as there is generally less stress on the joints.

It is preferable that a normal swimming pool is used for ongoing management, as hydrotherapy pools are too hot for distance swimming. Tai Chi and Pilates are also recommended as they facilitate balance and control. Bicycling can be good for aerobic work and again does not over-stress the joints. Nordic pole walking can be particularly effective as it is a functional activity. It is hypothesised that the closed chain nature of the activity assists proprioceptive feedback and engages the trunk sling muscles facilitating trunk (core) stability. The aim is to encourage a lifelong commitment to exercise and maintenance of good general fitness, through normal activities and a return to sport.

Joint hypermobility is a relatively common phenomenon which may be an asset or may predispose to a range of clinical problems. Because of the ubiquitous nature of collagen, hEDS may present in a variety of clinical presentations. Positive recognition and avoidance of unnecessary investigations and drug therapy are among the most important interventions. Most children are well managed with simple advice and reassurance, while adults frequently require a more structured rehabilitation programme. Modification of activities and behaviour may be required to redress the balance between healthy physical activities and high-impact physical pursuits. If untreated or undiagnosed hEDS may result in the development of a 'chronic pain cycle' and a high level of disability. This will require an intensive rehabilitation programme to manage the symptoms effectively and improve functional capacity. It is vital that individuals with hEDS and their families are clear in their understanding of the condition. It is also important to stress that a self-management-led programme with support from health, sport and exercise professionals is the most appropriate long-term treatment approach. The future will no doubt yield more appropriate assessment tools and perhaps genetic analyses for identifying individuals at risk, thus allowing earlier implementation of preventive strategies.

This article has provided a critical review of the pathogenesis, epidemiology and clinical assessment and management strategies associated with hEDS. There is evidence to suggest that participation in sport, dance and performance is a risk factor for some individuals and also a suggestion that healing times and rehabilitation may be slower as a result of altered proprioception and connective tissue synthesis. Clearly exercise and participation in physical activity is an important management strategy and has a health promoting role, therefore it is imperative that professionals who work in this domain are aware of the condition, can assist prevention and work collaboratively within a multidisciplinary team to help manage children and adults with hEDS and their associated problems.

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