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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.