HYPERMOBILITY SPECTRUM DISORDERS

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Pathophysiology

Ehlers-Danlos syndrome (EDS) is a genetically heterogeneous disorder of connective tissue that is characterised by articular hypermobility (joints that stretch beyond normal limits), dermal hyperelasticity (skin that can stretch beyond normal limits) and widespread tissue fragility. Individuals with EDS demonstrate altered elasticity, integrity, tissue healing properties, and cognitive function. These alterations often lead to chronic musculoskeletal pain and fatigue and greatly affect the individual's ability to complete activities of daily living and subsequent quality of life. People with EDS appear to present with central sensitisation which causes a lower tolerance to stimuli, which would not normally cause pain. Many individuals with EDS also suffer extraarticular disorders such as anxiety, chronic fatigue syndrome, chronic regional pain syndrome, fibromyalgia, orthostatic intolerance, persistent headaches, incontinence etc. Due to the complex nature of EDS, a multidisciplinary approach with symptom specific management is currently considered best practice. Utilising pharmaceutical management, cognitive behavioural therapy, support from braces, and exercise therapy have shown the greatest improvements in symptoms reduction and quality of life.

Impairments

Impairments associated with hypermobility spectrum disorders include:

- Joint trauma (recurrent dislocations, subluxation, soft tissue damage) which can cause early joint degeneration and persistent pain
- Altered proprioception
- Gastrointestinal dysfunction
- Chronic pain due to hyperalgesia (increased sensitisation)
- Anxiety and mental health disorders
- Orthostatic tachycardia
- Bladder and pelvic floor dysfunction

Joint hypermobility and recurrent injury/dislocation are common impairments associated with hypermobility spectrum disorders. Pain, fatigue, and fear are common barriers to exercise for patients. A recent study showed that 45% of patients with hypermobility disorders also suffer mental health disorders, 41% super cardiovascular dysautonomia, and 27% suffer gastrointestinal dysfunction.

BENEFITS OF EXERCISE

- Significant improvements in mediolateral postural stability
- Increase stamina and functional capacity
- Improved quality of life
- Improved mental health status
- Increased participation: school/work/socially

Exercise Prescription

Exercise prescription is determined by symptom burden. Patients who suffer orthostatic intolerance display limited ability to exercise upright and exercising in supine is often indicated. It is essential to avoid stretching due to the tendency to increase joint laxity, instability and associated subluxations, pain, and dysfunction. Exercise prescription aims to increase strength in a progressive manner using resistance-based exercise. It is essential to teach control within hypermobile joint ranges to reduce injury risk, pain, and fatigue. Research has shown that non-weight bearing exercise prescribed at high repetitions and low load have the greatest outcomes in patients. Other considerations include the implementation of proprioceptive exercise using bio-feedback and balance boards, realistic goal setting, and pacing to improve energy conservation for important tasks.

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GOT A QUESTION?

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