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The Evidence-Based Rationale for Physical Therapy Treatment of Children, Adolescents, and Adults Diagnosed With Joint Hypermobility Syndrome/Hypermobile Ehlers-Danlos Syndrome

FOR NON-EXPERTS

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Introduction

Physical therapy/physiotherapy is key to managing Joint Hypermobility Syndrome/Hypermobile Ehlers-Danlos syndrome (JHS/hEDS). However, many clinicians (including physical therapists) are not familiar with how this condition is diagnosed, the common signs and symptoms, or best treatment approaches. This document summarizes what is known about the physical therapy approaches best supported by current best quality research and expert opinion.

Generalized joint hypermobility (GJH) is measured using a set of nine tests called the Beighton score, but the exact cut-off for considering a person hypermobile has been disputed for several decades. The diagnostic criteria for JHS/hEDS, which include the Beighton score, are presented in a companion article (see the summary of Malfait, *et al*. 2017).

The presentation of JHS/hEDS in children and adults is fairly similar. Pain is the most common problem and is present due to joint instability, overload of musculoskeletal structures (muscles, tendons, ligaments, joints), or abnormal movement patterns; pain may be localized or widespread. People with JHS/hEDS also appear to be hypersensitive to painful stimuli, that is, they feel more pain than a non-hypermobile individual in response to the same input; this may be due to their sensitive central nervous system processing pain differently.

People with JHS/hEDS have decreased tolerance to exercise, in part because exercise frequently causes increased pain. People are therefore less active and, as a result, have decreased strength and aerobic fitness. Hypermobility in children can also present as "floppy infant" syndrome. Both children and adults with JHS/hEDS tend to be less coordinated, have reduced body awareness and balance and tend to report feeling clumsy and falling more often than healthy individuals. In children, decreased gross motor control can lead to developmental delay, including learning to walk at a later age. Similarly, decreased fine motor control can result in difficulty with handwriting.

Pain is not the only source of complaints in JHS/hEDS. Fatigue is also common, and is sometimes more disabling than the pain. Psychological symptoms, such as depression, anxiety, and panic disorder are also common. Problems with the autonomic ("fight or flight") nervous system can lead to low blood pressure or an excessively fast heart rate. Gastrointestinal problems include constipation or diarrhea, reflux, and abdominal pain. Urinary incontinence is common in both children and adults. Skin tends to be overly stretchy and more vulnerable to bruising and slower to heal.

The combination of pain, fatigue, poor coordination, and other systemic symptoms can ultimately lead to decreased ability to perform normal, daily tasks at home, school, or work. This, in turn, can result in significantly decreased quality of life, self-image, and social function for people with JHS/hEDS.

Assessment and Treatment Principles

It is important that physical therapists perform a thorough initial assessment to rule out other more severe disorders of connective tissue where hypermobile joints may be present such as *osteogenesis imperfecta* ("brittle bone" disease), Marfan, Loeys-Dietz, or other forms of Ehlers-Danlos syndrome. If any of these other conditions is suspected, a referral to a rheumatologist should be made. Referral to an appropriate specialist should also be made where multisystem problems such as a racing heart rate, low blood pressure, gastrointestinal, and bladder issues impact significantly on a person's life. Readers can refer to other guideline papers for more information regarding these related issues in individuals with JHS/hEDS. Physical therapy is often helpful for those other problems, especially to help manage blood pressure and a racing heart.

Physical therapists should educate people with JHS/hEDS about how to protect joints and manage symptoms. Exercise is the cornerstone of treatment and there are several high quality research trials in both children and adults which provide the evidence for this. Although research to date has mainly focused on knee strengthening, body awareness, and balance training, other studies have demonstrated that core stability and endurance exercises are also effective for reducing pain and improving function. Interestingly, research in children with knee pain and JHS/ hEDS found that parents reported improved psychological and social function, including self-esteem, when children exercised through their entire range of movement, including the hypermobile range, when compared to children exercising only through the "normal" range. Research suggests that combining education and exercise with cognitive behavioral approaches (how people think about pain or functional limitations) may be particularly effective for improving pain and reducing disability in adolescents and adults.

Some expert clinicians recommend manual therapy (a variety of hands-on techniques), taping for support or body awareness feedback, pool exercise, and relaxation techniques to help manage symptoms. However, research has not proved that these treatments are effective for people with JHS/hEDS. Experts also recommend that treatment should be individualized and focus on learning to control movements. The cardiovascular and muscle strengthening exercises should be carefully taught and graded based on guidelines from the American College of Sports Medicine to avoid a flare-up of symptoms. Interviews with parents of children who took part in a treatment research trial found that children kept up with their exercises better when supervised by parents and when it became part of a family activity. Returning to sport and performing arts after an injury or illness should also be carefully progressed to avoid re-injury.

The clinical guidelines recommend that children with flexible flat feet, pain, or trouble with weight-bearing activities, should use orthotics and/or wear sensible footwear. Sensible footwear means that shoes and sandals should be comfortable and supportive, especially around the heel cup, and the soles of the shoes should be well-cushioned. Preliminary findings of a small research trial suggest that use of orthotics may improve the walking mechanics of children with GJH and developmental coordination problems.

Splints and braces are sometimes used by people to help protect their joints and may be helpful after a flare up of symptoms and during early rehabilitation. There is currently limited research evidence to support the long term use of splints for the hands and wrists. Mobility aids, such as canes/walking sticks, crutches, or wheelchairs, should be prescribed judiciously after careful discussion with the multidisciplinary team and education with regard to their use. People with JHS/hEDS should avoid overusing these mobility aids because the aim of rehabilitation is to become as independently fit and able as possible through exercise and lifestyle choices.

Much more research is needed to help physical therapists determine the most effective types of treatments. Also, better education of physical therapists in management of JHS/hEDS will help ensure that the most up to date care programs are provided for people with this condition.

Original source papers

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