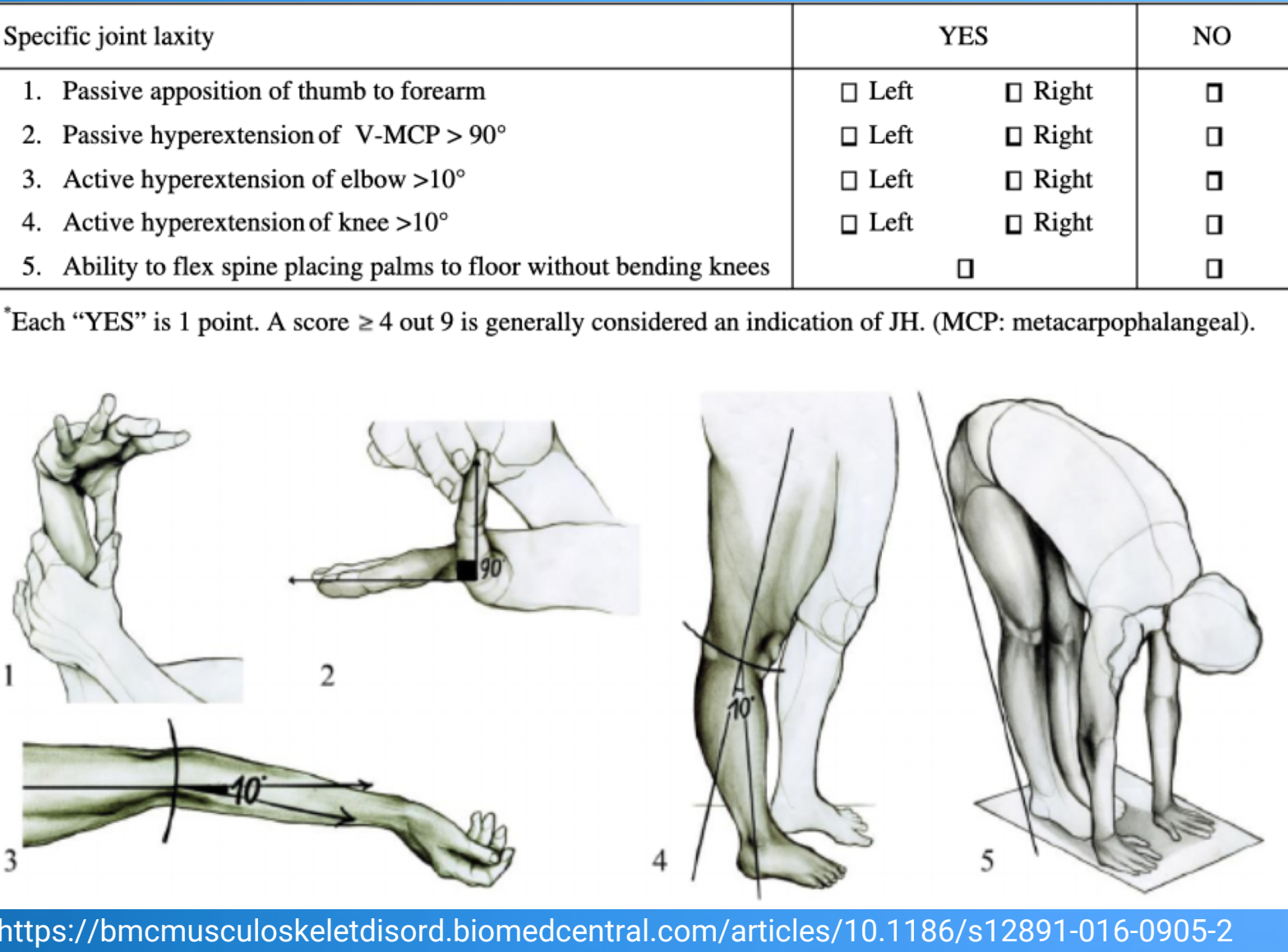


Exercise Prescription for Patients with Ehlers-Danlos Syndrome



Introduction

EDS is a genetic connective tissue disorder which can be classified 13 different ways. Some key features which distinguish this disease are joint hypermobility, skin fragility, joint pain without arthritis, and rupture of blood vessels and internal organs. This pathology can present itself at any age, and no particular type is more common than another. Ethnicity, race, and gender do not seem to determine prevalence of this disease. 1 in every 20,000 people are diagnosed with classical EDS (Ghali et al., 2019). The process of diagnosing a patient includes examination of medical history, skin and musculoskeletal system, and other factors through primary and secondary care. Pharmacological strategies are targeted at symptom management. For example, benzodiazepines such as Valium or Xanax can be used to treat restless leg syndrome. Benzodiazepines act as a tranquilizer to reduce the muscle spasms in restless leg syndrome (Castori et al., 2012).



Each "YES" is 1 point. A score ≥ 4 out 9 is generally considered an indication of JH. (MCP: metacarpophalangeal).

The Beighton scoring system is primarily used to diagnose joint hypermobility, which is a key indicator of the presence of EDS. Other criteria for diagnosing EDS include: unusual skin characteristics, positive family history for EDS, musculoskeletal pain, recurrent joint dislocations/instability, and skin fragility (Ghali et al., 2019).

Exercise Testing

Goniometer	•It is used to measure range of motion •Clinicians should expect most patients to be hypermobile •The goal is to limit hypermobility (Liaghat et al., 2020)
Visual Analog Scale (VAS)	•It is used to measure pain not only in the pre- and post- assessments, but also during training sessions (Schepter et al., 2017)
Checklist Individual Strength (CIS)	•It is used to measure fatigue in the pre- and post- assessments •It includes questions regarding perception, motivation, activity, and concentration (Schepter et al., 2017)
Six-Minute Walk Test	•It is used to measure activity limitations (Schepter et al., 2017) •It is also used to measure cardiorespiratory fitness
30-sec Chair Rise Test	•It is used to measure activity limitations (Schepter et al., 2017) •It is also used to measure muscular endurance
Handgrip Dynamometer	•It is used to measure muscular strength (Schepter et al., 2017) •Specifically, it can also measure isometric shoulder strength by assessing the maximum isometric voluntary contraction of shoulder scaption, internal rotation and external rotation (Liaghat et al., 2020)

Exercise Prescription

In general, the focus needs to be on symptom management and limiting the progression of the disease. Compiled in Table 1 are various prevention and intervention strategies for different symptoms. Inspiratory muscle strength training is useful not only for patients with respiratory insufficiency, but also for hypermobile patients to improve lung function (Castori et al., 2012; Reychler et al., 2018). It is also important to note that the strengthening exercises are not important for hypertrophy or exponential gains, but to reduce activity limitations (Castori et al., 2012; Schepter et al., 2017). Low-impact activities are very important to limit progression (Castori et al., 2012). Some great examples of low-impact sports/activities are swimming, pilates, and walking (Simmonds et al., 2015).

Table 1								
Prevention and Intervention Strategies to Limit Progression and Manage Symptoms/Pain								
Symptoms/Pain	Aerobic Fitness	Strengthening Exercises	Low-Impact Sports/Activities	General Relaxation	Weight Control	Stretching Exercises	Proprioception Exercises	Respiratory/Diaphragmatic Exercises
Acute Articular Limb Pain	x	x	x					
Chronic Articular Limb Pain	x	x	x		x			
Localized Muscular Limb Pain						x		
Generalized Muscular Limb Pain	x							
Back/Neck Pain	x		x		x	x		
Headache		x						
Abdominal Pain				x				
Visceral Pelvic Pain				x				
Dysmenorrhea				x				
Musculoskeletal/Neuropathic Pelvic Pain	x				x			
Muscle Weakness	x	x					x	
Respiratory Insufficiency								x
Castori et al., 2012; Reychler et al., 2018; Schepter et al., 2017; Simmonds et al., 2015								

Conclusion

The anticipated outcomes of exercise for individuals with this disease is to be able to strengthen areas around joints so that they are less prone to over-extension injuries. Exercise for this population can help to add stability around the joints and reduce the allowed degrees of hypermobility. Exercise can allow for the affected population to complete activities of daily living (ADLs) with less injury possibilities. Contra- indications include avoiding reaching fatigue during exercise (use CIS), avoiding high impact activities, and utilizing limited range of motion exercises. The overall goal in training this population is symptom management and slowing/reducing the disease progression. Exercise programming for each session is going to change based on the severity of the client's symptoms that day. The goal is increased quality of life and longevity, so progress is likely to be small.

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Special Considerations

