REHAB MANAGEMENT OF JOINT HYPERMOBILITY

Anne Keil PT, DPT
In Motion Therapy, Montrose
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OBJECTIVES

Participants will be able to:

-Identify joint hypermobility syndromes via the Beighton Scale

-Learn when to refer non-benign joint hypermobile individuals for further testing

-Develop a home program consisting of specific exercises, patient education related to awareness of posture & activity modification

-Review and modify if appropriate, an existing exercise program

HYPERMOBILITY DEFINITION

- An increase in range of movement due to looseness in connective tissues, especially ligaments (Keer, 2011)

- **Benign joint hypermobility syndrome: generalized joint laxity with musculoskeletal complaints in the absence of genetic, musculoskeletal, or rheumatic disorders.**

- Genetic form: Inherited genetic trait determined by a person's connective tissue protein matrix genes (Grahame, 2008) and can exist by itself or be a part of a more complex diagnosis.

Beighton score

Give yourself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points

Ehlers-Danlos Support UK

Registered Charity 1157027

Can you bend your thumb back onto the front of your forearm?

left thumb 1 point

right thumb 1 point

Can you bend your knee backwards?

left knee 1 point

right knee 1 point

Can you put your hands flat on the floor with your knees straight?

1 point

left hand 1 point

right hand 1 point

Can you bend your little finger up at 90º (right angles) to the back of your hand?

Can you bend your elbow backwards?

right arm 1 point

left arm 1 point

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Beighton Scale

-Hypermobility is clinically and objectively determined by 4 of 9 points (1 point per maneuver per right or left side)

-This scale is NOT used to rate genetic disorders such as Marfan’s or Ehlers-Danlos Syndromes (EDS has 13 subtypes)

SYMPTOMS OF HYPERMOBILITY

- Patients report discomfort from stiffness and positively like stretching
- Can be symptomless apart from hypermobility.
- Tip of the iceberg?: there is a series of other symptoms (cardiovascular, skin, mucosal, fascial, nervous system, dysmorphism) that result from hypermobility, these should be evaluated for a diagnosis of hypermobility spectrum disorders if complaints are more than just musculoskeletal.
- The tendency for hypermobile individuals to rest at end range and regularly use their joints in a locked position is thought to be an attempt to improve stability (Simmonds, 2008)
- Dislike sustained postures (sitting, standing) or too much activity

DIFFERENTIAL DIAGNOSIS

- Acquired forms of hypermobility exist (training such as dance, martial arts, yoga, etc.).

- Rheumatologic disease (in kids, JRA and hip dysplasia), inflammatory or degenerative diseases, CRPS, fibromyalgia, hypothyroid and other endocrine or collagen diseases must be ruled out.

- **Opportunity for PT/OT/ATCs to help screen for non-benign hypermobility which require referral to appropriate physician (PCP, rheumatologist, genetics clinic).

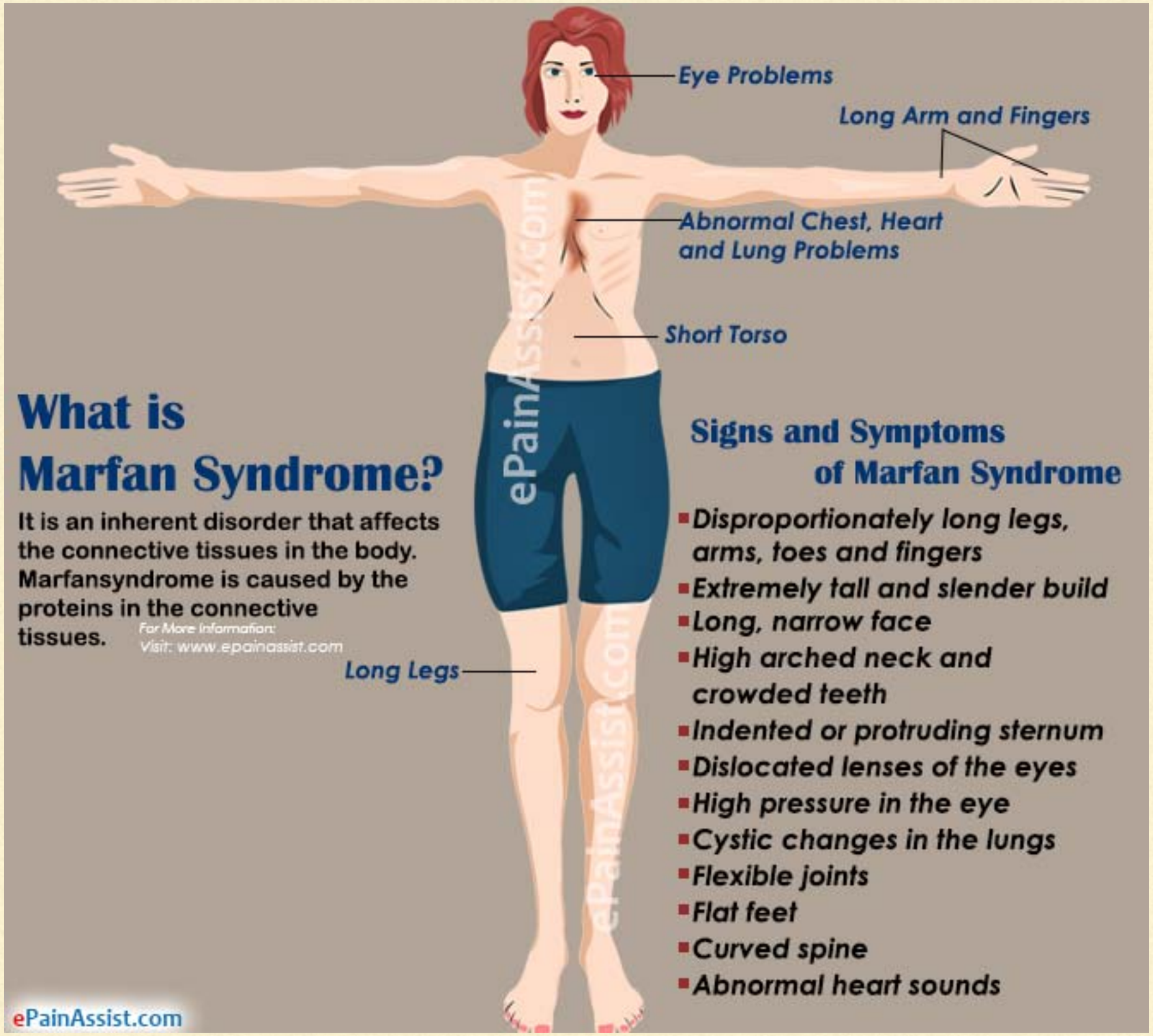
DIFFERENTIAL DIAGNOSIS

In a survey of patients with hypermobility, “Participants highlighted a range of impacts of JHS, incorporating physical, social and psychological domains. **Respondents described difficult journeys to diagnosis, and feeling unsupported and misunderstood by their peers and healthcare professionals**.” (Palmer et al, 2019)

CONNECTIVE TISSUE DISORDERS (IN CHILDREN)

- Ehlers Danlos syndrome: A clinically and genetically heterogenous group of connective tissue disorders caused by mutations in genes associated with collagen synthesis or caused by a mutation in the collagen gene or protein that processes collagen. Prevalence in US 1:10,000.

- Marfan's syndrome: A genetic connective tissue disease resulting in musculoskeletal, cardiovascular, respiratory, ophthalmologic and integumentary compromise, dx via Ghent criteria, with or without family hx. Prevalence in US 1-2:10,000. Physical characteristics: long limbs, face.


Eye Problems
Long Arm and Fingers
Abnormal Chest, Heart and Lung Problems
Short Torso
What is Marfan Syndrome?
It is an inherent disorder that affects the connective tissues in the body. Marfansyndrome is caused by the proteins in the connective tissues.
For More Information:
Visit: www.epainassist.com
Long Legs
Signs and Symptoms of Marfan Syndrome
Disproportionately long legs, arms, toes and fingers
Extremely tall and slender build
Long, narrow face
High arched neck and crowded teeth
Indented or protruding sternum
Dislocated lenses of the eyes
High pressure in the eye
Cystic changes in the lungs
Flexible joints
Flat feet
Curved spine
Abnormal heart sounds
ePainAssist.com

NEED FOR REFERRAL IN MARFAN SYNDROME!

Underdiagnosed by orthopedists and pediatricians.

Can decrease life expectancy if left untreated!

Requires specialist and additional diagnostics to r/o:

-Aortic dilatation

-Aortic dissection

-Aortic valve prolapse

-Ocular lens dislocation

Current Classification	Clinical Presentation	Prior Classification
Classic	Joint hypermobility, Skin hyperextensibility, Atrophic scars Smooth velvety texture	EDS I and EDS II
Hypermobile	Joint hypermobility Mild skin hyperextensibility +/- smooth velvety texture	EDS III
Vascular	Thin skin, Easily bruised Pinched nose, Appearance of premature aging, Rupture of medium and large arteries within uterus and bowel	EDS IV
Kyphoscoliotic	Joint hypermobility, Progressive scoliosis, Scleral fragility with rupture, Tissue fragility, Aortic dilation, Mitral valve prolapse	EDS VI

Current Classification	Prior classification	Clinical Presentation
Arthrochalasia	EDS VII A EDS VII B	Severe joint hypermobility Joint Subluxations, Congenital hip dislocation, Skin hyperextensibility, Tissue fragility
Dermatosparaxis	EDS VII C	Severe skin fragility, Decreased skin elasticity, Easily bruised Hernias Premature rupture of fetal membranes
Unclassified	EDS V EDS VIII EDS X, EDS XI, EDS IX EDS, progerioid form	Classic features? Classic features? and periodontic disease Mild classic features, MVP, joint instability Classic features, occipital horns,

Resource:

-Education on hypermobility spectrum disorders and their management
-free webinars for patients, providers and families
-helpline, support groups
-intended audience is for individuals with hypermobility of a genetic cause, but general principles can apply to acquired hypermobility as well

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HYPERMOBILITY, IF SYMPTOMATIC, CAN BE DUE TO:

- Trauma
- Pain hypersensitivity (chronic pain state)
- Altered proprioception
- Muscle weakness/imbalance
- Malalignment, flat feet, scoliosis, etc., resulting some or all of the above

SYMPTOMS DUE TO TRAUMA

- Macrotrauma: dislocations, subluxations, and surrounding soft tissue damage (ligaments, tendons, muscles). Typical history is of multiple sprains, strains, tendinitis, etc.

- Microtrauma: leads to persistent pain and possibly early joint degeneration. Examples: overuse syndromes, chondromalacia, low bone mineral density

SYMPTOMS DUE TO CHRONIC PAIN

- Occasionally, pain is due to trauma (subluxation, dislocation)

- Hyperalgesia (pain hypersensitivity). Pain science: David Butler, Lorimer Moseley's research.

- Deconditioned state due to reduced physical activity due to longstanding pain (Simmonds, 2010).

PAIN DUE TO ALTERED PROPRIOCEPTION

- Inaccurate sense of where the body is in space and how much effort is needed for movement
- Muscle imbalances
- Tendency to “hang out” at end ranges: genu recurvatum, lumbar lordosis, etc.
- Deconditioning

REHAB EVAL FOCUS

- <u>Subjective exam</u>:
- hx multiple sprains, strains, tendonitis, overuse, subluxations?
- <u>Objective exam</u>:
- plumb line posture (do they “hang” on their ligaments), ROM (calves and HS typically tight)
- strength testing especially core, balance, proprioception
- Passive and accessory joint play testing

TREATMENT:

- Pain science education (train/change your brain). Physical activities without fear, activity modification and pacing.
- Identify and correct aberrant movement patterns especially with sports
- Core strengthening is key to improved extremity function (From a golf swing to cleaning the kitchen)
- Joint protection education, taping, bracing, orthotics, other adaptive equipment (OT) if indicated

TREATMENT GUIDELINES

- Early stage: Home program including nature of their condition, pain management plan, spine/proximal joint stabilization, awareness of joint position/ postures, breathing and relaxation exercises, weight control, avoid high impact activities, low environmental temps. Promote regular aerobic fitness.
- Middle stage: global and peripheral muscle strength, endurance, controlled flexibility training, water exercise, low level cardiovascular training (walking land or water).
- Late stage: increase functional CV exercise, progress muscle strength/ endurance (weight, resistance bands)
- Final stage: long term fitness management (pilates, yoga, tai chi, feldenkrais, qi gong, etc).

TREATMENT GUIDELINES

- Typically a minimum of 3 rehab sessions (initial eval, follow up to the eval, then follow up after patient has been able to implement their program) as hypermobility is more of a lifestyle modification

- May require short term rehab to assist with pain/tightness complaints, or overuse syndromes that may occur. Consider home TENS, regular massages, etc for long term self-maintenance of symptoms

TREATMENT:

- Posture education: avoid “hanging” on ligaments, recurvatum, no “party tricks”, correct pes planus (orthotics)
- Stretching: Varus heel calf stretch, isolated hamstring stretch any tight muscles, watch neutral joint positioning to restore normal ROM
- Dry needling: effective in early rehab to decrease “weak muscle tightness” until muscles get stronger. Not a long term solution.
- Proprioception exercises and balance training is usually more effective than high force plyometric exercises like running. Example: half kneel ball toss
- Pelvic/bladder dysfunction (incontinence?) treatment if present.

STRENGTHENING: TREATMENT

- Water exercise is a great choice! Avoid over-stretching
- Targeted strengthening especially in midranges
  - lats, back extensors, lower abs, obliques, rhomboid, serratus, mid/low traps
  - squats, lunges, proprioception, balance
  - elastic bands, weights, Swiss ball

PRECAUTIONS

- Excessive fatigue: promote rest intervals, hydration, do not eliminate activity
- Sports: no high contact sports (football, lacrosse, hockey).
- Sports requiring flexibility may be limited?
- Swimming: if hypermobile shoulder symptoms, breast stroke may be best choice

OTHER CONSIDERATIONS

Pregnancy, breast feeding:

Relaxin can exacerbate hypermobility symptoms so activity may need to be more modified

CE QUESTIONS:

- 1. What is the Beighton score cutoff that identifies hypermobility? (answer: 4/9)

- 2. Name at least 2 conditions whose symptoms are joint hypermobility and require further MD referral? (answer: Marfans, EDS, RA, and more)

- 3. T or F: Patient education and lifestyle modification are an important initial discussion in individuals with hypermobility.

QUESTIONS?

- Thank you!

REFERENCES

- Carlson M. BJHS, course notes 2018.
- Castori M, Tinkle B, Levy H, Grahame R, Malfait F, Hakim A. A framework for the classification of joint hypermobility and related conditions, Am J Med Genet Part C Sem Med Genet 175C:148-157 2010.
- ehlers-danlos.com
- Grahame, R. Hypermobility: an important but often neglected area within rheumatology. Nature Clinical Practice Rheumatology, Vol 4 P 522-524, 2008.
- Grahame, R. The multi systemic nature and natural history of joint hypermobility syndrome and Ehlers-Danlos syndrome in children: New research date conflict with widely held views. Rheumatology. Vol 56, #12, December 2017, p 2048-2049.
- Keer, R., Simmonds J. Joint protection and physical rehabilitation of the adult with hypermobility syndrome. Current Opinion in Rheumatology. March 2011 Vol 23 #2 P 131-136.
- Liebrand A., DiIennio M., Connective Tissue Disorders in Children: Evaluation and Treatment on land and in the Water, course notes CSM 2011.
- Palmer S et al. The views of people with joint hypermobility syndrome on its impact, management and the use of patient-reported outcome measures. A thematic analysis of open ended questionnaire responses. Musculoskeletal Care. Vol 17 #2, June 2019 p. 183-193.
- Remvig L., Jensen D., Ward R. Epidemiology of general joint hypermobility and basis for the proposed criteria for benign joint hypermobility syndrome: review of the literature. Journal of Rheumatology. April 2007, 34 (4) 804-809
- Simmonds J, Keer R. Hypermobility and the hypermobility syndrome, Part 2: Assessment and management of hypermobility syndrome: Illustrated via case studies. Manual Therapy. Vol 13 #2, P e1-e11 2008.