Referral Support Service

Rheumatology

RH03
Hypermobility Syndromes

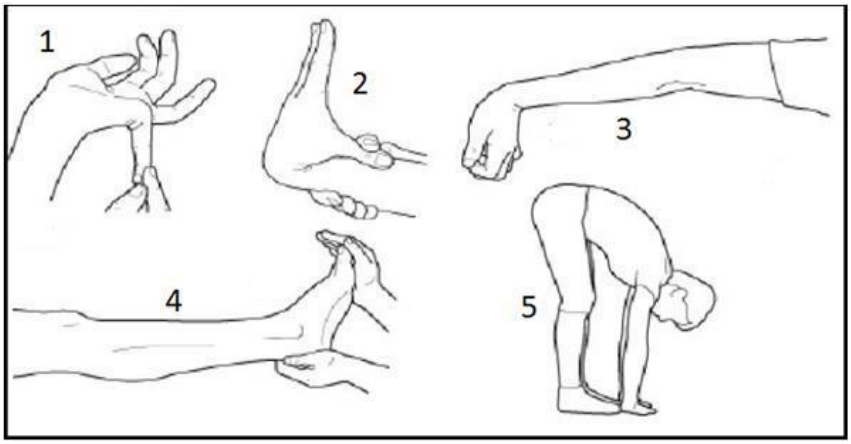
Definition

Joint hypermobility is a common cause of joint pain and stiffness predominantly in young patients, but may continue into later years. Inherited abnormalities of connective tissue disease such as Ehlers Danlos or Marfan's syndrome associated with joint hypermobility are rare. It is important to identify the latter groups as they are at risk of significant, but rare complications.

Assessment

Joint hypermobility can be assessed using a number of methods, but the Beighton score is a simple and reliable assessment tool.

The Beighton Score (**Figure 1.** Manoeuvres in the Beighton Score)



A total of 9 points are collated from 5 manoeuvres comprising:

1. Passive dorsiflexion of the little fingers beyond 90° – 1 point for each hand
2. Passive apposition of the thumbs to the flexor aspects of the forearm – 1 point for each thumb
3. Hyperextension of the elbows beyond 10° – 1 point for each elbow
4. Hyperextension of the knee beyond 10° – 1 point for each knee
5. Forward flexion of the trunk with knees fully extended so that the palms of the hands rest flat on the floor – 1 point

A score of 5 or more indicates generalised hypermobility.

To assess patients for inherited collagen disorders, broader assessment is required. These conditions should be suspected if additional systemic features outlined below are present:

Musculoskeletal:

- **Pain** in 2 or more limbs daily for at least 3 months, widespread pain for ≥3 months
- **Recurrent joint dislocations** in the absence of trauma – 3 or more atraumatic dislocations in the same joint, or atraumatic dislocations in 2 different joints occurring at different times, or medical confirmation of joint instability at 2 or more sites not related to trauma

Family History:

- First or second degree relative with inherited collagen disorder

Skin:

- **Abnormal elasticity**
- **Scarring**- atrophic > 1site or classic tissue paper or hemosiderotic.
- **Striae** - Unexplained.
- **Bilateral piezogenic papules of the heel**
- **Connective tissue/ fascia weakness**:
- Recurrent or multiple abdominal hernia
- Pelvic floor, rectal, and/or uterine prolapse in children, men or nulliparous women without predisposing medical condition

Oral abnormalities:

- Dental crowding and high or narrow palate

Cardiac abnormalities:

- Mitral valve prolapse (MVP)
- Aortic root dilatation.
- Autonomic symptoms, incl. tachycardia, hypotension, syncope

Abnormal body habitus:

- Arm span-to-height ≥1.05 AND/OR upper segment/lower segment ratio <0.89. Arachnodactyly, disproportionately long slender digits.

Eyes:

- History of lens dislocation
- Myopia

Management

The vast majority of patients with joint hypermobility require reassurance, physiotherapy and simple analgesia. There are no specific additional treatments for inherited disorders of connective tissue, however, some patients will require screening for complications e.g. echocardiography and ophthalmic review and also genetic confirmation of some of the disorders is possible e.g. certain Ehlers Danlos syndrome sub-types

Referral Information

Indications for referral

- Referral to secondary care should be considered if the patient has confirmed hypermobility by Beighton score and has more than one systemic feature.
- Seek advice and guidance if patients do not fulfil these criteria, but you have concerns.

Information to include in referral letter

- Beighton Score
- History of recurrent joint dislocation

- Systemic symptoms of concern related to inheritable disorders of connective tissue (see above)

Patient information leaflets/ PDAs

https://www.versusarthritis.org/about-arthritis/conditions/joint-hypermobility/