

Annotations

Hypermobility of joints

Individuals who have very loose joints have attracted medical curiosity since the time of Hippocrates, and those 'India rubber people' who also have stretchy skin now earn the appellation of the Ehlers-Danlos syndrome. It is only in the last three decades that more subtle patterns of hypermobility have been associated with common disorders of paediatric importance. Carter and Wilkinson devised clinical procedures to show that children who have congenital hip dysplasia and their first degree relatives tend towards generalised hypermobility.¹ Subsequent studies have shown other musculo-skeletal associations of articular hypermobility and have drawn attention to the relatively common benign hypermobility syndrome.²

Diarthrodial joints are constrained from excessive motion by their surrounding soft tissues, primarily the joint capsule, as well as ligaments, tendons, adjacent muscles that move the joints, overlying subcutaneous tissues, and skin. At the biochemical level, the most important constituent of these tissues is type I collagen, a rope like macromolecule that is the most ubiquitous of body structural proteins. Collagen stiffens with aging as a result of increased diameter of assembled bundles of fibrils, as well as increase in the number of intermolecular crosslinks. Stiffness of collagen in ligaments, as assessed indirectly by measurement of joint mobility, increases throughout childhood and early adult life.^{3,4} Clinical consequences of hypermobility are seen when the tensile resistance of articular constraining tissues differs from the normal for that age and also when inappropriate mechanical strain is imposed on those tissues by muscular activity. For example, intrauterine hypermobility predisposes to congenital hip dysplasia^{1,5} and adolescent overindulgence in athleticism may precipitate the hypermobility syndrome.^{2,6}

Assessment of hypermobility

Routine clinical assessment of hypermobility does not require special equipment. Mechanical devices reported for research purposes^{3,7,8} have not supplanted the simple Carter-Wilkinson type of clinical manoeuvres.¹ In modified form,⁴ the procedures are (1) passive apposition of the thumbs to the flexor aspect of the forearm, (2) passive dorsiflexion of the

fifth fingers beyond 90°, (3) hyperextension of the elbows beyond 10°, (4) hyperextension of the knees beyond 10°, and (5) ability to place the palms of both hands flat on the floor with the knees held in extension. By this method a score can be assigned, with a maximum of nine, one point for each of the paired limb procedures and one point for spinal hypermobility; a mobility score of four or greater may be considered hypermobile. The spinal flexion test is the least useful in that performance is influenced by age, limb length, and even the repetitive hamstring stretching of ballet training.⁹ Of course, hypermobility may be demonstrable in other joints such as the wrists or finger joints, and the latter may be particularly impressive in 'collagenopathies' such as Ehlers-Danlos syndrome. The frequency of abnormal mobility varies according to joint, with the knees and elbows remaining hypermobile later in life than the wrists or ankles. The expectation that symptoms occur only in children who are generally hypermobile may lead to under-recognition of localised hypermobility associated symptoms.⁶

Hypermobility of joints is a common clinical finding and is not symptomatic in the majority of children. In general, girls have greater mobility of joints than boys of the same age, ranges are usually greater in the non-dominant limb, and Asians are more mobile than whites. Carter and Wilkinson reported hypermobility in 7% of 285 healthy children aged 6 to 11 years.¹ A more recent American study found generalised hypermobility in 12% of normal schoolchildren and 66% of 32 children with 'juvenile episodic arthralgia'.¹⁰

Hypermobility syndrome

Kirk, Ansell, and Bywaters drew attention to a syndrome of recurring arthralgia, joint swelling, and muscle cramps in hypermobile persons, 75% of their patients being aged 16 years or younger.² Patients with features of Ehlers-Danlos syndrome or Marfan's syndrome were excluded. This type of benign hypermobility probably represents the upper end of the normal range of joint mobility. The hypermobility syndrome is a more common cause of musculo-skeletal symptoms in teenagers, especially girls, than any of the varieties of inflammatory juvenile

arthritis. The typical patient has recurrent pain and sometimes intermittent swelling in one or other knee. Polyarticular symptomatology is unusual and the differential diagnosis is most often between hypermobility and inflammatory joint disease. Although joint effusion occurs, presumably from mild traumatic synovitis, signs of active inflammation will be absent. Other common complaints include bruising and muscle cramps after physical activity. Often there is a history of a growth spurt preceding the onset of symptoms, and diagnostic clues in the family history may include congenital hip dislocation, scoliosis, and past occurrence of similar symptoms or 'growing pains' in a parent.

Associated disorders

As articular hypermobility is so common it may coexist with inflammatory joint disease. For this reason the hypermobility syndrome is a diagnosis of exclusion and simple laboratory tests for inflammatory rheumatic disease may be necessary to reassure both the doctor and the family.

Hypermobility accompanies a multitude of monogenic syndromes, such as osteogenesis imperfecta and other heritable disorders of connective tissue, and multifactorial syndromes, such as congenital hip dysplasia.⁷ Most of these are readily recognisable by their concomitant features, and hypermobility will usually be an incidental or contributory physical finding rather than the primary mode of presentation. Occasionally, a hypermobile tall adolescent with recurring pain or swelling of a knee will also have the long limbs, arachnoidactyly, and anterior chest deformity of Marfan's syndrome; this warrants special attention because of the late life threatening risk of aortic dilatation.

Management

Bracing of very lax and unstable joints may be necessary in patients who have gross hypermobility, usually associated with one of the collagenopathies such as Ehlers-Danlos syndrome type I. The same approach may help to preserve function when instability is due to the hypotonia of muscular or neuromuscular disease rather than ligamentous laxity.

In the benign hypermobility syndrome explanation of the basis of symptoms has particular therapeutic value. Recognition and demonstration of articular hypermobility in a child with previously mysterious aches and pains is often a source of satisfaction to both the patient and doctor, especially when there has been delay in diagnosis.⁶ There may be anxiety that the child has potentially disabling arthritis, and this worry may have been

fuelled by overinvestigation or overuse of drugs.⁶ The fundamental problem is not inflammation, and most of these patients do not need anti-inflammatory medication. Symptoms generally resolve late in adolescence when periarticular tissues develop mature tensile resistance. The strength of joint ligaments cannot be augmented by drugs, and the best that can be done is to strengthen the surrounding musculature. Usually this will mean having a physiotherapist instruct the child in quadriceps exercises, and regular swimming in the neighbourhood pool is a good form of physical treatment. Physical activity should not be restricted unless the child is clearly overindulging in gymnastics, competitive sports, or ballet. The children should be reassessed during adolescence because of the increased risk of scoliosis.

Later in life there is an increased liability to minor injuries,⁹ hernias, varicose veins, pes planus, uterine prolapse, and other consequences of tissue laxity.⁷ Fifty or 60 years later some who were hypermobile in childhood may be more susceptible to certain patterns of osteoarthritis,² but the evidence is no more than preliminary¹¹ and this should not alter the optimism of prognosis.

References

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