Hypermobility syndrome

What is hypermobility syndrome?

Joint hypermobility syndrome, also known as benign hypermobility syndrome, is a connective tissue disease characterised by joint instability, chronic pain, and minor skin changes. It shares many clinical features of Ehlers-Danlos syndrome, hypermobility type and so many authorities consider them as one disease process [1] .

However, many people do not fully meet the diagnostic criteria for hypermobile Ehlers-Danlos syndrome (hEDS) but their hypermobility still causes problems, including pain and dislocations, poor sense of where their joints are without looking, a tendency to injury, curvature of the spine, and degenerative joint and bone disease [2] .

Other commonly associated features of joint hypermobility syndrome include marked fatiguability, tiredness, back pain, joint subluxation and soft tissue injuries.

Many with joint hypermobility syndrome have pain and fatiguability when writing by hand and experience posture-related muscular pains, generalised tiredness and altered sleep patterns and concentration - all of which can affect school performance [3] . Extra-articular symptoms may include abdominal pain, headaches and mood disturbance.

Prevalence

Joint hypermobility is common and is found in up to 30% of children [4] .

However, approximately 3% of the general population are believed to have joint hypermobility syndrome [1] .

Hypermobility syndrome symptoms[3]

The main presenting features are joint hypermobility with exercise-related muscle and joint pains and some level of fatigue. However, there is enormous variability in symptom severity. Commonly the history is of:

- Joint/muscle pains after activity and at night - usually more lower-limb than upper- limb - most typically, calf and thigh muscles and knees. Younger children tend to report more pain, possibly because teenagers are developing stronger, tighter muscles.
- Swelling, heat or redness are not usually present, unless there is associated injury.
- Muscle and joint stiffness occur the day after activity, often for several days.
- Fatigue, often with reduced exercise tolerance. This relates to deconditioning through exercise avoidance and may impact on all aspects of life, affecting energy levels and concentration.
- Fidgetiness, probably from deconditioned muscles.
- Headaches, often relating to trapezius muscle spasm and poor sitting posture.
- Easy bruising.
- Clicking of joints, both spontaneously and deliberately. Clicking the joints will not harm them unless it becomes obsessive.
- Reduced co-ordination and balance, probably secondary to reduced core strength, leading to clumsiness.
- Handwriting may fatigue easily and fine motor control may be affected.
- A small proportion may have problems with abdominal pain with or without bladder and bowel dysfunction. It is not always clear whether this relates directly. Abdominal pain is common in childhood and frequently relates to constipation. Constipation may increase in incidence when there is reduction in physical activity and muscle tone.

- Rare associations are:
    - Postural orthostatic tachycardia syndrome (PoTS).
    - Hernia: studies of children with hernia have shown that they show an increased prevalence of Beighton score of 4 or more compared to the general population [5] .
    - Uterine or rectal prolapse.
    - Joint dislocation (although subluxation is slightly more common).

A 2005 review of a UK population of 125 children diagnosed with benign joint hypermobility syndrome (BJHS) found that [6] :

- Average age at onset of symptoms was 6.2 years and age at diagnosis 9.0 years.
- The major presenting complaint was arthralgia in 74%, abnormal gait in 10%, apparent joint deformity in 10% and back pain in 6%.
- Mean age at first walking was 15.0 months; 48% were considered 'clumsy' and 36% as having poor co-ordination in early childhood.
- 12% had 'clicky' hips at birth and 4% actual congenital dislocatable hip.
- Urinary tract infections were present in 13% and 6% of the female and male cases, respectively. 14% had speech and learning difficulties diagnosed.
- History of recurrent joint sprains was seen in 20%.
- Actual subluxation/dislocation of joints was seen in 10%.
- 40% had problems with handwriting tasks.
- 48% had major limitations of school-based physical education activities.
- 67% had major limitations of other physical activities.
- 41% had missed significant periods of schooling because of symptoms.

- 43% described easy bruising.
- 94% scored > or =4/9 on the Beighton scale for generalised hypermobility, with knees (92%), elbows (87%), wrists (82%), hand metacarpophalangeal joints (79%) and ankles (75%) being most frequently involved.

Diagnosis[7]

Diagnosis is clinical, using the widely accepted Brighton criteria. These combine the Beighton hypermobility score (first developed to quantify joint laxity in Ehlers-Danlos syndrome) with symptoms. There are no confirming genetic or biochemical tests.

Beighton hypermobility score[8]

The Beighton hypermobility score is a 9-point scoring system to quantify joint laxity and hypermobility. A higher score equates to higher joint laxity. The threshold for joint laxity in a young adult is 4-6, with scores above 4 correlating well with pain levels in patients diagnosed with benign joint hypermobility syndrome.

Joint	Finding	Pts
Left little finger	Passive dorsiflexion beyond 90°	1
	Passive dorsiflexion < =90°	0
Right little finger	Passive dorsiflexion beyond 90°	1
	Passive dorsiflexion < =90°	0
Left thumb	Passive dorsiflexion to flexor aspect of forearm	1
	Cannot passively dorsiflex thumb to flexor aspect of forearm	0
Right thumb	Passive dorsiflexion to flexor aspect of forearm	1
	Cannot passively dorsiflex thumb to flexor aspect of forearm	0
Left elbow	Hyperextends beyond 10°	1
	Extends < =10°	0
Right elbow	hyperextends beyond 10 °o	1
	Extends < =10°	0
Left knee	Hyperextends beyond 10°	1
	Extends < = 10°	0
Right knee	Hyperextends beyond 10°	1

	Extends < =10°	0
Trunk flexion with knees fully extended	Palms and hands can rest flat on the floor	1
	Palms and hands cannot rest flat on the floor	0

In children high scores correlate with increased range of motion and with exercise-related pain. However, many children with high scores are not symptomatic[9] [10] [11] .

Brighton diagnostic criteria[8]

These accepted diagnostic criteria incorporate the Beighton scoring system, in combination with the presence of persistent symptoms.

Diagnosis of benign joint hypermobility syndrome requires two major criteria or one major and two minor, in the absence of diagnosed Ehlers-Danlos or Marfan's syndromes. Two minor criteria are considered enough for diagnosis if there is a clearly affected first-degree relative.

Major criteria

- Beighton score of 4 or more.
- Arthralgia for >3 months in four or more joints.

Minor criteria

- Beighton score of 1-3.
- Arthralgia for >3 months in one to three joints.
- Back pain for >3 months.
- Spondylosis/spondylolysis/spondylolisthesis.
- Dislocation/subluxation of more than one joint, or in one joint more than once.
- More than three soft tissue inflammatory conditions (eg, tenosynovitis, epicondylitis).

- Marfanoid habitus (tall, slim, span/height ratio >1.03, arachnodactyly, upper/lower segment ratio <0.89.
- Skin striae, thin skin, hyperextensible skin, papyraceous scars.
- Drooping eyelids, myopia or antimongoloid slant.
- Varicose veins, hernia or uterine/rectal prolapse.

Differential diagnosis

The main differential diagnoses are the hereditary connective tissue disorders and inflammatory joint conditions:

- Ehlers-Danlos syndrome (type 3; hypermobility type): Ehlers-Danlos syndrome (EDS) is an umbrella term for a group of heritable soft connective tissue disorders characterised by generalised joint hypermobility, skin texture abnormalities, and visceral and vascular fragility or dysfunctions [4] .
- Marfan's syndrome.
- Juvenile idiopathic arthritis.
- Rheumatoid arthritis.
- Ankylosing spondylitis.
- Fibromyalgia.

Diagnosis is not straightforward, as benign joint hypermobility syndrome and Ehlers-Danlos syndrome type 3 are difficult to distinguish. One school of thought suggests that Ehlers-Danlos syndrome type 3 (hypermobility type) - which is the most common variant - and benign joint hypermobility syndrome are the same condition. There are no genetic or biochemical tests that can distinguish between the two; diagnosis is made on physical examination and medical history.

Which of the two is diagnosed is a clinical judgement as there is no absolute medical consensus. Ehlers-Danlos syndrome is more likely to be the diagnosis where there is a pattern of autosomal dominant inheritance, or where there are associated non-benign medical conditions such as mitral valve prolapse, uterine, rectal or bladder prolapse and (in particular) recurrent dislocations. Benign joint hypermobility syndrome may be the diagnosis where the main symptoms are pain and joint hypermobility with little in the way of associated conditions. However, There there is a clinical overlap between joint hypermobility syndrome and Ehlers-Danlos syndrome [4] .

It is essential to recognise hypermobility syndromes with potentially life threatening complications. Genetic testing is only available for some syndromes, but is indicated if there is a reasonable probability regarding a specific syndrome, especially if this syndrome can have life-threatening complications [12] .

There is some overlap in symptoms with fibromyalgia; both conditions may share heightened central pain sensitivity [13] .

Hypermobility syndrome assessment

Due to lack of awareness, variable clinical presentation, and reliance on physical examination for diagnosis, joint hypermobility syndrome is largely overlooked, leading to delayed or missed opportunities for diagnosis, and inappropriate interventions [1] .

Management begins with the assessment, which should include a full history, symptom scoring and discussion of the child's activity levels, including hobbies, physical activity and sleep, the impact of symptoms on learning and participation, and the child's level of understanding of the problem.

It is useful to perform a Beighton score and a baseline assessment of muscle strength. This is done using the Kendal scale, which scores the strength of each muscle from 0-10 [14] . The muscles to be assessed should include quadriceps, hip abductors, hip extensors, plantar flexors and core stability muscles. Posture and gait should be assessed with a view to shoe orthotics; also, a stamina test (eg, a six-minute walk) may be useful.

Hypermobility syndrome treatment and management[15]

The principle of treatment is that strengthening of specific muscle groups will help to support the hypermobile joints, improve posture and increase strength, tone and fitness. Treatment includes patient education, activity modification, stretching exercises, osteopathic manipulation and physiotherapy. The goal is full participation in all activities.

Self-management

The aim is for young people to manage their own exercise; however, this may take time. Working in partnership with the child will be crucial to improving outcomes.

The most important factor in understanding of the condition, for patients, is that the pain in benign joint hypermobility syndrome is not a sign of muscle damage or an indication that activity is harmful. It means that the muscles are not strong enough and need strengthening. Increasing, rather than decreasing, physical activity is therefore the solution. Similarly, fatigue is an indicator that the body requires better-paced activity and increased fitness. Gradually increasing activity levels is the best treatment for daytime fatigue; when there is a daytime slump in energy levels, a snack and a walk are a better solution than a rest.

Encouragement will be needed to begin or increase daily exercise. Significant periods of inactivity greatly exacerbate symptoms of BJHS. Encourage normal activity and a return to sport. The most effective exercise programme is a progressive resisted exercise programme targeting weakened muscles which are needed to control the joints in their hypermobile range. If these muscles are completely effective then posture and function will improve.

Return to exercise may need to be gradual. A good understanding of pacing activities needs to be developed by children with BJHS to avoid overexertion leading to injury or discouragement.

Pain management

Pain can affect concentration, memory, mood and sleeping. Reassurance is needed that the pain is not harmful. It is also important to help parents to develop strategies to promote helpful patterns of behaviour for their child. There will be times of increased pain - for example, after a lot of sport or injury. It is really important that at such times rest is not promoted for long periods of time.

In general, medical approaches to pain management are not beneficial. Instead, it is useful for the child and family to develop their own interventions to reduce the pain. These can be distractors such as activities, music and TV, as well as positive coping statements, relaxation scripts, aromatherapy oils, exercises, comforting objects and pictures.

Some patients will have already developed a chronic pain syndrome at the time of presentation. These patients benefit from a comprehensive pain management programme, incorporating cognitive behavioural therapy techniques.

Physiotherapy

A commitment to exercise is needed. Physiotherapy can help to promote this using specific exercises that can be incorporated into daily life. The aim of physiotherapy is:

- To restore full muscle strength and function throughout the full range of movement.
- To restore effective movement patterns.
- To improve general fitness.
- To restore normal range of movement.
- To provide education, reassurance, advice and pain management and to develop problem solving.
- To provide advice on activities which are vital to maintain strength and stamina.
- To provide advice on and help with postural alignment which will help ensure the exercise programme is effective.
- To provide advice on core strengthening exercises which are helpful for strength, stability, balance and posture.

- To provide advice on stretches which are used to maintain muscle length and joint range and to iron out old injuries.
- To incorporate programmes based on enhancing proprioception, using mirrors for feedback. Such programmes have achieved good results.

Patients with benign joint hypermobility syndrome tend to have poor tolerance of repetitive activity, experiencing pain and fatigue the following day. They also tend to have increased pain hypersensitivity. This is a normal response to an increased level of exercise and is not an indication of damage. Nevertheless, exercises using high repetitions and low weights are very effective.

Patients may also have reduced proprioception, making it harder for them to carry out exercises correctly alone. Supervision of exercises is important, particularly early on.

Occupational therapy (OT)

OT may be helpful for participation in activities of daily living and helping with appropriate interventions. This may include building skills, suggesting strategies, helping with pacing activity and educating the young person and others in order to enable independence in daily activities. An OT assessment in school may also be helpful. Assistance with writing instruments, seating, desks and bags may all be helpful interventions.

Podiatry

Footwear should be supportive if ankles are weak. Orthotics may be prescribed where the positioning and function of the feet may contribute to symptoms.

Choosing sport

Sport will be greatly beneficial in the long term but it is important to be sure that the child is fit enough to participate at the level chosen, in the chosen sport. It may be necessary to build up strength and stamina first to avoid being set up to fail or, worse still, being injured. Particular care needs to be taken around trampolining and bouncy castles, which put particular strain on hypermobile joints. Children with hyperextensible joints typically do well in activities in which flexibility is desirable, such as gymnastics and ballet; however, if repeated dislocations occur, these activities may need to be replaced with those which place less strain on the joints.

Weight management

This is important, since obesity correlates with increasing pain scores due to the added strain on the joints. Being underweight - with weaker, unsupportive muscles - also has a negative impact. Dietary advice should aim to address both weight and any constipation.

Mood

If mood is low then engagement and motivation are less likely to be positive and this may need to be separately addressed. Sleep management should also be discussed.

Equipment

With the exception of orthotics in selected cases, none should be required. Wheelchairs and crutches are particularly unhelpful in the long term, encouraging illness behaviour.

Support

Ongoing support may be necessary for some time. See below for contact details.

Guidance for schools

Schools may benefit from advice on helping pupils with benign joint hypermobility syndrome. This includes all of the above. In addition:

- Sporting activity should be encouraged, making allowances for reduced stamina.
- Altering seating may help with posture
- Try to avoid the child staying still for extended periods of time. Walking about for brief periods at intervals may prevent discomfort.
- Pupils may need extra time to move between classes and travel around the school.
- If OT assessment suggests handwriting is fatiguing it may be appropriate to recommend extra time during examinations.
- Urinary urgency and bowel problems may be part of BJHS so access to toilets should not be restricted.

- Storage may be helpful for books so they don't have to be carried around all day.

Hypermobility syndrome prognosis

Benign joint hypermobility syndrome is generally a self-limiting condition which improves as the child develops increased muscle size and strength, reducing joint looseness. A full return to normal activities is the usual outcome. The speed with which this occurs depends on the severity of the condition, the commitment and effort of the child and family to the management programme and on the level of support they require and receive. Improvements can be surprisingly fast. However, some individuals continue to have problems into adulthood, usually of joint laxity resulting in easy injury.

BJHS rarely leads to arthritis, although problems with the kneecap may develop in those in whom it has recurrently dislocated. Adults who retain their benign joint hypermobility syndrome are at slightly greater risk of developing osteoarthritis of affected joints. A small review of young adult soldiers with the condition found significant associated morbidities, including tenosynovitis, degenerative joint changes and synovitis [16] .

Benign joint hypermobility syndrome may be misnamed. Whilst not a life-threatening illness, if it is left unchecked the consequences of exercise avoidance and reduced school performance may be lifelong [6] .

Dr Mary Lowth is an author or the original author of this leaflet.

Further reading

- Hypermobility Syndromes Association
- Malfait F, Francomano C, Byers P, et al; The 2017 international classification of the Ehlers-Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):8-26. doi: 10.1002/ajmg.c.31552.

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