Guidance for Management of Symptomatic Hypermobility in Children and Young People – A Guide for Professionals managing Children and Young People with this condition

This guidance document has been compiled by the Allied Health Professionals Group working within BSPAR Section Council and has been designed to help and support therapists working with children and young people (CYP) presenting with symptomatic hypermobility and musculoskeletal pain.

The WHO definitions: Childhood 0-10 years, Adolescence 10-19 years and Young Adulthood 20-25 years.

This guidance replaces the former guidance from Jan 2013, ratified by the BSPAR Executive Committee in June 2013. In order to cover the complexities of this condition in some CYP, some aspects of management have been divided into different professions; however, there will be significant overlap as to who provides the intervention depending upon local teams.

Objectives

- To develop expert consensus opinion on the holistic management for CYP with symptomatic hypermobility, using a biopsychosocial model which acknowledges the impact of the condition
- To enable children, young people and their parents/carers to be active participants in the management of the condition
- To optimise the standard of care of CYP with symptomatic hypermobility by empowering them, their carers, and other health professionals through the provision of education, information, support and treatment if required
- To ensure efficient, cost effective and evidence-based therapy management for CYP with this condition

Introduction

Recent criteria (2017 EDS International classification) and Scottish Paediatrics and Adolescent Rheumatology Network (SPARN) have acknowledged that hypermobility lies within a spectrum. Hypermobile joints are common in the general population and many individuals with hypermobility do not experience any significant difficulties. In fact, in many situations such as sportspeople and dancers, as examples, hypermobility can have positive advantages.

However, some people experience difficulties and symptoms which are understood to be related to being hypermobile. Hypermobility should be regarded as a “normal variant”: difficulties occur mainly when the body has become weak and deconditioned.

It is not always helpful for a young person to have the label of Ehlers Danlos Syndromes (EDS), and new criteria has been published (2017 EDS international classification) to clarify this. The significant risks associated with the other forms of EDS can mistakenly be assumed to also apply to this group, causing unnecessary distress.

Assessing and managing this condition in CYP requires knowledge of potential extra-articular symptoms including abdominal involvement, headaches, fatigue etc. although there is a normal variance of these symptoms in the paediatric and adolescent population.
generally. Care must be taken to ensure that appropriate importance is placed upon each symptom and the condition is NOT over-medicalised.

**Aims**

This guidance aims to provide further information for health professionals involved in the management of CYP with this condition and to outline the main issues that may need to be considered when developing an individualised management plan:

- To listen to the CYP and family’s concerns and deal with the expectations of CYP and parents. The aims of treatment are to empower the CYP and families to understand the management of this condition and develop confidence, skills and knowledge in self-management strategies

- To improve current symptoms, and to learn to avoid future complications

- To work together with CYP and parents to develop age appropriate and developmentally appropriate individual management plans and to work towards clearly defined and shared goals

- To recognise that the longer-term outlook for young people with this condition is very positive and with the correct management, all CYP should be able to participate in all activities they want to without ongoing professional support

**Subjective Assessment**

Could include the following as appropriate:

- Presenting problems
- Past medical history
- Birth and developmental history
- Drug history
- Social and family history
- Mental health and well-being
- Ethnic and cultural concerns
- Financial benefits claimed/PIP payments
- Participation in ADL including self-care, leisure, sleep and fatigue
- Impact on learning and participation in school activities, school attendance % and hobbies
- Career development/planning
- HEEADSSS 3.0 Psychosocial assessment for Adolescence (see https://app.appinstitute.com/heeadss for trigger questions and training video)
- What previous information has been given, and by whom and the level of understanding of management
- Other professional involvement
Presenting Problems and Common Symptoms:

- **Joint and/or muscle aches and pains** often occurring after activity, end of day or during the night

- **Lower limb** more frequently affected due to weight bearing nature of joints

- **Muscle and joint stiffness**, usually after exercise or increased activity, may occur for few days after (Delayed Onset Muscle Soreness or DOMS)

- **Fatigue** – often associated with reduced walking distance and reduced exercise tolerance. There may be difficulties attending school full-time due to tiredness. Fatigue is commonly linked to the level of deconditioning of the child or young person. Poor sleep will also impact on fatigue levels

- **Difficulty sitting still** and poor organisation of movement

- **Postural problems**

- **Easy bruising** – this is benign and not of concern

- **Clicking joints** – joints can click spontaneously or be clicked deliberately and can be performed safely many times a day. This only becomes a concern if it becomes habitual and obsessive impacting on quality of life.

- **Reduced coordination and balance** – poor proprioception leading to clumsiness and reduced balance, poor core stability leading to difficulties with fine motor control such as handwriting. However, hand function and control develop with age and should be compared with age appropriate norms. If these symptoms do not improve with improving strength and gross motor skills and have a significant impact upon ADLs, below what would be expected for chronological age and intelligence level, an assessment for a Developmental Coordination Disorder (DCD) may be advisable by an appropriate professional with relevant experience.

- **Abdominal pain** – this is common in childhood and adolescence generally and if persistent further advice and management should be sought from the GP

Additional but rare symptoms

**Gastrointestinal and urinary tract symptoms** – non-specific symptoms such as nausea, stomach ache, diarrhoea, constipation, fecal and urinary incontinence can be a feature of the condition however they are also very common symptoms in children and young people generally and may not be related to their hypermobility.

- Routine treatment of paediatric constipation effective with these symptoms

- Advice about toilet habits can be useful such as having a stool to rest their feet on in front of the toilet to help optimize the position of the pelvis and enable effective bowel opening

- Onward referral for medical management of significant bowel symptoms may be required

- Onward referral for medical management of significant urological symptoms may be required
**Cardiovascular autonomic dysfunction** – patients with symptomatic hypermobility may describe symptoms of autonomic dysfunction, which affects heart rate, blood pressure and blood flow, such as:

- Light-headedness and dizziness, particularly on standing
- Fainting
- Heart palpitations or a racing heartbeat
- Non-specific fatigue

It is important to differentiate from anxiety symptoms. Those with severe symptoms of cardiovascular autonomic dysfunction should be referred to a specialist Centre with experience in the assessment and management of this in CYP.

**Other Cardiac features**

Increased aortic root size and mitral valve prolapse had been reported to be more common in patients with symptomatic hypermobility, but current evidence suggests they are usually of little clinical consequence and is extremely rare.

It is important not to alarm the child or adolescent and their family and so assessment of this should be very carefully considered and explained to the family, as increased anxiety about this condition is not helpful. Consideration of cardiology referral may be useful for a child or young person who scores highly on the hEDS criteria.

**Objective Assessment & Suggested Outcome Measures**

This list is not prescriptive and suggested guidance only.

**Observation and general wellbeing**

- Dysmorphic features
- Skin elasticity (see EDS classification 2017)
- Marfanoid features (EDS classification 2017)
- Observe for signs of self-harm (upper thighs/non-dominant arm)
- Patient VAS for perceived level of general wellbeing, completed if the child is old enough
- PedQL 4.0 – (Paediatric Quality of Life Inventory, Varni et al)
- MSK HQ (over 18) – Hill et al, 2016

**Baseline measurements**

- Height/weight/centiles/BP

**Pain**

- Age-appropriate pain VAS can be used

**Fatigue**

- Age-appropriate fatigue VAS can be used
Joint range of movement (ROM)

- Baseline joint ROM useful, looking at all joints
- Look for focal hypermobility
- Beighton Scale may be useful but does not indicate severity of symptoms. International Consortium on EDS proposes Hypermobility score of 6/9 and over, for pre-pubertal children and adolescents

Muscle length

- Awareness that muscles that move over two joints may become tight despite generalised hypermobility

Muscle strength

- Baseline assessment of muscle strength very important and research suggests is related to development of symptoms if submaximal. Kendall Muscle Strength Scale (0-10) provides a score that monitors change over time
- Specific muscles to consider depending on the area affected:
  o Inner-range quadriceps (SLR without a quadriceps lag)
  o Hip abductors, specifically gluteus medius
  o Hip extensors, specifically gluteus maximus
  o Ankle plantar-flexors
  o Central core stability
  o Consider shoulder stability and strength for hand function/strength
- Consider how muscles function

Balance

- Standardised SLS – eyes open, eyes shut, timed – standardised protocol without shoes

Posture

- Should be assessed particularly in sitting and standing dependent on exercise activities and lifestyle and hobbies

Gait

- Gait analysis important and consideration of the need for orthotics

Stamina

- Six minute walk test (6MWT) may be useful. A two minute walk test shows good correlation with 6MWT
- Timed functional test e.g. step test or sit to stand in one minute can be useful

Function
Timed functional tests as above can be useful.

CHAQ (Childhood Health Assessment Questionnaire – Nugent 2001) can demonstrate a measurement of function however is not formally validated for symptomatic hypermobility.

Objective assessment of performance including self-care, dressing, leisure, school attendance (%), handwriting, dexterous manipulation, activity organisation can be considered and various specific outcome measures are available.

**Management of Symptomatic Hypermobility**

All professionals involved with CYP and their carers should work collaboratively to formulate a cohesive management programme to help to improve symptoms, establish realistic expectations, encourage full function and help to develop the skills and knowledge in order to **self-manage** the symptoms and participate in usual activities. A thorough MDT assessment can be helpful to determine the most appropriate plan.

**Self-management**

- It is helpful for families to understand that this is not a life-threatening condition and in many instances, hypermobility can be advantageous. Care should be taken to avoid over-medicalising CYPs symptoms.
- Motivational interviewing techniques can be useful.
- Successful management involves full involvement of the young person, adolescent and carers in the management plan.
- Age and developmentally appropriate language should be used.
- Encouragement to maintain a healthy lifestyle, maintain hydration, eat a well-balanced diet, participate in regular gentle exercise, participate fully in school, activities and social events, and to get quality sleep are important.
- Children and young people may benefit from joint protection advice and energy conservation principles for carrying out everyday tasks, to avoid placing unnecessary strain on their joints and minimising pain. Often encouragement is needed to include appropriate exercise into their daily routines. It is well recognised that significant periods of inactivity such as illness, or during growth periods, may exacerbate symptoms.
- A good understanding of pacing activities should be developed by young people, avoiding the “boom and bust” cycle where over-exertion leads to pain and fatigue; however, prolonged rest can be counter-productive. A baseline level of activity should be established and maintained and gradually increased.

**Pain management and education**

We usually understand pain to be a warning about damage to the body. When this is the case a reasonable response to the pain would be to withdraw or avoid activity. However, it is understood that symptoms in hypermobility are an indication that the body is not strong enough to do the task and that it requires strengthening.

This means that the young person should continue to engage in everyday activities and exercise, to avoid de-conditioning, not do less. Recognition and management of chronic pain...
is an important part of management for children and young people with symptomatic hypermobility.

Pain can affect concentration, memory, mood, sleeping and also impact on quality of life. If appropriate, referral to a unit experienced with the management of chronic pain in young people can be helpful if they fail to progress with local management, as chronic pain is most effectively managed with a combination of physical and psychological techniques.

Pain medications are often ineffective, and the side effects should be carefully considered. Pain management interventions can be provided by physiotherapists, occupational therapists and psychologists depending on local service provision. The aim of pain management is to increase quality of life. Therefore, CYP-led goals will be an important part of boosting motivation to engage in the self-management programme.

Fear of the unknown can be a big source of anxiety for both the young person and their carers, so education focused on addressing these personal beliefs can be helpful in reducing such fears, as fear can lead to avoidance. Challenging unhelpful beliefs and behaviours, such as excessive rest, catastrophising or avoidance are helpful to understand their role in the fear and pain cycle.

It is helpful to establish realistic expectations. There will be times of increased pain, after a lot of sport or injury, and a management plan for dealing with these times should be discussed. It is really important to discuss a relapse plan to ensure that during these times, long periods of rest are not adopted.

For children and young people suffering with symptoms, it is helpful to consider their relationships with others. Parents can be hugely disempowered if their child is experiencing pain and it is therefore important to help parents develop strategies that they can use to ensure they promote positive rather than unhelpful patterns of behaviour for their child.

Similarly, fatigue is not necessarily an indication that more rest is required. Rather, it may signal that the body requires more fitness training, pacing of activities and better sleep hygiene. It is important to recognise that normal sleep patterns change throughout childhood, adolescence and then into adulthood.

It is important to consider this when assessing fatigue levels and abnormal sleeping patterns. Establishing baseline activity levels and gradually pacing up activity and exercise is the most effective cure for fatigue during the day and will help to improve exercise tolerance over time.

Promotion of a Comfort Tool Box, into which the young person and family/carers put their own collection of activities and interventions which can be used to reduce the pain experience, can be a useful adjunct.

These can include items to encourage distraction, positive coping statements, relaxation scripts or apps, sleep hygiene, mindfulness, aromatherapy oils, exercises, comforting objects. These may be real or a virtual box that has been created by the young person and family/carers (https://www.swft.nhs.uk/application/files/7714/6012/5294/chronic_pain_management_for_teenagers.pdf)

Physiotherapy

Promoting lifestyle change and enjoyable activity is needed for young people with symptomatic hypermobility. Physiotherapy can help to promote this from a young age,
encouraging activities that can be incorporated into daily life. Specific exercise programmes to target specific areas of the body can be useful to facilitate normal function and participation in age-appropriate activities.

Goals of physiotherapy intervention:

- Restore and maintain full muscle strength and function throughout the full range of movement
- Restore effective and efficient movement patterns
- Improve general fitness, exercise tolerance and stamina
- Provide education, reassurance advice, pain management and help develop problem solving skills
- Promotion of self-management and reduce fear of movement and increased confidence

Patients with symptomatic hypermobility may have a poor tolerance of static, repetitive or excessive activity, often resulting in pain and/or fatigue later that day or the following day.

This may be due to muscle imbalances resulting in “delayed onset muscle soreness” (DOMS). It is important to give an explanation re DOMS and this may help to reduce the worry about activities aggravating symptoms and thus a reluctance to exercise. DOMS is a completely normal response to an increased level of exercise and is not an indication of damage.

The physiotherapy will be most effective if targeted to the specific areas of difficulty the young person is experiencing. It is important to establish correct technique/position for the activity/exercise being carried out.

Physiotherapy may include:

- **Normal activities**, including PE and a graded return to sport, providing individual advice on specific elements of activities as appropriate.

- **Specific exercises**, such as a progressive resisted exercise programme to target the specific muscles that are weak and that are required to control the joints into the hypermobile range. A variety of techniques can be used ranging from open to closed chain exercise, lower impact to start with, and including functional exercises graded individually for each patient

- **Advice on postural alignment**, avoiding extremes at end range positions and prolonged static loading can help to protect against injury and enables muscles to function most efficiently

- **Core strengthening** – exercises to build core strength, muscle strength and endurance improve stability, balance and coordination. Every day, age-appropriate, functional activities may be incorporated. Wobble boards, mats, cushions and gym balls may be helpful

- **Stretching**, which can be used to maintain muscle length, joint range and to stretch out old injuries or muscle spasm. Stretching can be useful before bedtime to help with nighttime symptoms. Stretching is not advised to increase an already hypermobile range
• **Advice** – to avoid static loading and avoid repeated self-subluxation. It is not thought that ‘unusual’ resting positions are harmful if non-symptomatic

• **Goal setting** – should be realistic. Both short and long-term goals can help patients make progress towards full function

• **Proprioception training** – research shows that proprioception programmes have shown good results and improve patient outcomes along with strengthening programmes

• **Sport specific assessment** – an assessment of specific sports techniques along with coaches if appropriate may be helpful to enable the young person to participate fully

**Occupational Therapy**

Occupational Therapy (OT) assessment and intervention can be an essential component of the CYP’s life.

OTs holistically assess all activities of daily living both personal activities of daily living (PADLs e.g. washing, dressing, toileting) and domestic activities of daily living (DADLs e.g. chores, making a drink or snack). OTs also assess:

• Functional mobility
• Hand function and handwriting
• Sleep hygiene and fatigue
• Leisure activities
• Education and work
• The impact of relationships
• Mood and behaviour

Comprehensive assessment will allow the OT to identify barriers to occupational performance and design the most appropriate intervention to maximise function. The main aim of OT intervention is to promote self-management where possible and this can be achieved through a variety of means. Assessment must be age and developmentally appropriate and tailored to the CYP’s individual needs.

**Activities of daily living**

Advice, education, strategies, functional goals, hand and shoulder exercise and small aids can improve CYP independence in all ADLs. A biopsychosocial approach and motivational interviewing skills are helpful for engaging CYP in being able to increase their function in these areas. Liaison with parents and carers, educational environment and significant others is considered helpful for facilitating this. Short-term small aids may be used as a compensator approach to increase function, however longer term use of these should be discouraged, as their use may contribute to increased muscle weakness.

**Pain and Fatigue Management**

CYP with hypermobility generally have deconditioning of their muscles which affects their stamina and endurance. This can result in pain and fatigue with prolonged activity and can lead to avoidance behaviors, which in turn exacerbate pain and fatigue.
Educating a CYP to look after their joints and posture can greatly improve their ability to sustain activities. Energy conservation principles which include pacing, prioritising, planning and education on avoiding the boom and bust cycle can empower a CYP’s ability to manage day-to-day activities more effectively.

Evaluation of sleep patterns to establish difficulties is necessary. It is important to acknowledge that sleep routines change during normal adolescence and are not the same as in adulthood. See sleep diaries on [https://sleepcouncil.org.uk/sleep-diary/](https://sleepcouncil.org.uk/sleep-diary/)

Education on sleep hygiene and use of appropriate apps and podcasts in addition to relaxation techniques can have a very positive effect on quality and quantity of sleep.

Education and advice on the cycle of pain that can be associated with hypermobile joints and its effects on function are essential to promote self-management. The CYP can be equipped with a ‘toolkit’ which helps with understanding chronic pain, mood, motivation and function.

A pacing approach can be helpful in systematically increasing activity over time with the aim for the CYP to be able to manage all their daily activities.

**Hand function**

A thorough hand assessment in sitting and standing, and in conjunction with a shoulder assessment, can identify problems with function usually related to pain and fatigue in muscles with prolonged grip and pinch. The CYP can be advised on a SMART goal orientated exercise program functionally related which will improve endurance for all hand related activities.

The DASH writing assessment is useful for educational establishments and there are many types of pens, pen grips, computer accessories available which can all improve a CYP function and reduce pain with sustained activity. Support for reasonable adjustments at educational environments and work can enable the CYP to feel validated and promote confidence to self-manage in these aspects of life.

**Splinting**

The use of splinting is generally avoided due to risk of deconditioning, however splinting can be considered in the following circumstances in conjunction with a home exercise program of strengthening:

- DIP hyperextension whilst writing can be improved using an oval 8 if pen grips are not sufficient
- Splinting of the thumb may be provided if the MCP or CMC subluxes during functional activities – a neoprene splint is preferable

**The Young Person and behaviour**

There is considerable evidence about the development of the adolescent brain and the necessity for experimentation to aid normal development and promote resilience. Communication is the key to help the YP and the psychosocial assessment, HEEADSSS 3.0 is useful alongside other assessments with this age group. OTs can use this tool to educate YP to keep them safe and self-manage whilst they are in this important stage of development.

**Podiatry**
Podiatrists provide advice on suitable footwear. Supportive footwear is beneficial, especially if CYP have flat feet. Supportive footwear includes:

- Shoes which are stiff around the heel
- A sturdy sole to act as a shock absorber
- Soft uppers, preferably with laces or buckles, that support the whole foot
- Boots with laces or high top trainers appropriate and comfortable

This does not mean that other shoes cannot be worn for short periods of time or special occasions. Orthotics may be prescribed for those CYP with flat feet where the positioning and function of the feet may be contributing to symptoms.

Clinical Psychology

Psychology input may be required for some CYP to ensure that symptomatic hypermobility does not prevent the young person fully participating in life.

Psychology input may focus upon pain management skills and techniques or on thoughts, feelings and behaviors in response to symptoms. Cognitive behavior therapy (CBT) may be useful in modifying unhelpful beliefs and behaviors, though many other approaches may be utilised.

If the CYP is experiencing low mood and/or anxiety these may negatively impact upon their symptoms and their ability to self-manage. Lack of activity and participation, in turn, can further exacerbate symptoms and soon CYP can be stuck in a negative cycle and may become fearful of pain and thus avoid activity and more pain and fatigue ensues.

A clinical psychologist, as part of an MDT approach, would be able to work with the CYP and their family in order to help modify these feelings so that they are able to regain control of their symptoms and their life. Referral onward to a specialist team with appropriate expertise is recommended.

General Advice

- Activity and sport: physical activity should be actively encouraged for all CYP with symptomatic hypermobility, however it is extremely important that the CYP is fit enough to participate in the sport they wish to do. If this is not the case, a CYP may need to focus on building suitable strength and stamina before engaging in the new activity. School and college liaison is important to facilitate graded participation as appropriate and suitable advice offered. Within education, suitable sporting activities should be encouraged, with advice on the suitability of high impact activities and allowances made for reduced stamina. Some activities, such as contact sport, may need more physical preparation than others. Current evidence suggests during childhood it may be better to enjoy a variety of sports rather than focus on one. Current guidance suggests that “age” should be an approximate guide to the number of hours of high level sport participation per week. (Jayanthi et al 2015)

- Weight management: maintaining optimum body weight is important. Extra weight can significantly strain lower limb joints and increase symptoms of pain and fatigue. Healthy diet advice may be helpful in weight management and also the management of constipation. Becoming underweight is also not ideal as this can make maintaining adequate muscle strength difficult, and if this is the case, it may not be safe to
engage in an intensive exercise programme until weight stabilises. Weight issues may reflect body image issues which in turn may reflect mental health issues

- **Equipment**: the general philosophy that applies to the provision of equipment is that the most effective solution to a physical challenge is to find a way to get stronger and fitter so that it can be done with minimum adaptations. Helpful suggestions such as buggy boards or scooter can help to facilitate activity in the young. Special equipment is not required. Provision of a wheelchair for a CYP with symptomatic hypermobility is not recommended and is counter-productive to the CYP ability to maintain muscle strength, function and endurance. Long-term use of crutches can be equally unhelpful and should also be avoided in the management of hypermobility. Adaptive equipment e.g. raised toilet seat and bath-boards should not be routinely prescribed, instead the focus should be on increasing strength, proprioception and function.

- **Concerning symptoms**: for some families, the philosophy of self-management and maintaining strength and full independent function is, unfortunately, very challenging and the fear of pain and symptoms may be such that they find it very difficult to follow the advice of the professionals. These families may need more time and interventions in order to help facilitate understanding of the pain messages and these families may require specialist psychological input, in order to help them modify their beliefs. These families would benefit from referral to a MDT team which includes a level 3 child protection trained paediatric rheumatologist or paediatrician.
Helpful Resources

- APCP leaflet Symptomatic Hypermobility
- APCP leaflet Choosing footwear for children
- APCP leaflet Flat feet in young children
- APCP leaflet Choosing the right school bag
- Ergonomics4kids.co.uk
- http://hypermobility.org/

Apps

- Happy not perfect
- Headspace
- Sleep cycle
- Deep sleep hypnosis – mindfulness
- Stress relief hypnosis
- Sleepio- itunes
- Smiling minds
- www.scarleteen.com/

Podcasts

- http://www.mentalhealth.org.uk/help-information/podcasts
- Progressive relaxation for better sleep
- Wellbeing and sleep: quick fix exercise
- Wellbeing and sleep: the full works
Further Academic Reading


*BMJ Open* 2016;6:e012331. doi: 10.1136/bmjopen-2016-012331


Scottish Paediatric & Adolescent Rheumatology network (SPARN) Referral pathway for children with Joint Hypermobility


Time lapse imaging tracks brain maturation from 5-20 Paul Thompson, Ph.D. UCLA Laboratory of Neuroimaging

Acknowledgments

This guidance has been reviewed and produced after a review of literature by AHP MDT members of BSPAR Section Council and where there is a lack of evidence, an agreement of consensus expert opinion has been sought.

Issue number 2.1, May 2018

This guidance document was ratified by the Council of the paediatric and adolescent rheumatology section of the British Society for Rheumatology and is designed to support the delivery of paediatric and adolescent rheumatology.

Ratified by BSPAR Section Council: June 2019

To be reviewed in 2022