15-minute consultation: a structured approach to the management of hypermobility in a child

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Abstract

Objective: To present a structured approach for an outpatient consultation of a child with hypermobility.

Method: Review of literature and description of the approach commonly adopted in a paediatric out-patient setting.

Conclusion: A focused history and examination is key to reaching an appropriate differential diagnosis and planning management for children with hypermobility.

Case: A 10-year-old child is referred to your general paediatric clinic with frequent 'cracking and clicking' of her joints and a history of intermittent aches and pains over the last 6 months.

Background

Musculoskeletal symptoms are a common reason for presenting to primary care and secondary general paediatric care (1). Within subsequent referrals to paediatric rheumatology, a significant amount of hypermobility is found. Joints that are more flexible or move in excess of normal range of motion are considered hypermobile. Children's joints inherently possess a greater range of movement than adults, with a gradual reduction in this range observed over time with age. Typically, girls tend to be more hypermobile as compared to boys (2). In terms of ethnicity, Asians tend to more hypermobile than Sub-Saharan Africans, and Caucasian are the least hypermobile (2, 3). Hypermobility also tends to run in families, but the exact underlying genetic cause is unknown.

Most children with hypermobility are asymptomatic and use their increased range of joint movement to their advantage when participating in sports or hobbies. However, some children experience symptoms secondary to their hypermobility and this is know as benign joint hypermobility syndrome (BJHS). The clinician is therefore faced with the challenge of determining which symptoms are correctly related to hypermobility.

Defining hypermobility

The prevalence of hypermobility as an isolated phenomenon has been reported to vary widely across different populations, between 2.3-64% depending on age, ethnicity and the scoring system used for defining hypermobility (4-9). Despite this, the prevalence of pain amongst children with generalised hypermobility has been shown to range between 30-55% (6, 7). Internationally, the Beighton score is the most widely used scoring system for hypermobility, with a score of ≥ 4/9 usually being used as the cut-off (see Figure 1 A - E). Some authors have therefore suggested using a Beighton score of 5 or 6 in children (2, 8). However, as the Beighton score still lacks validation for use in children, a pragmatic definition of BJHS as **'a child or**

young person with musculoskeletal pain and signs of hypermobility, with no other cause found for their symptoms' has been advocated (10).

Figure 1: Components of the Beighton score (11). A maximum of two points are scored for each of the maneuvers demonstrated (on each side of the body) involving **(1A)** little fingers, **(1B)** thumbs, **(1C)** knees and **(1D)** elbows. One point is scored for spinal flexion **(1E)** when able to place the palms of the hands flat on the floor with knees straight.

What should you cover in the history?

Hypermobility related joint pain usually occurs during or after activity and is typically experienced in the evening after an active day. It is most often localised to the lower limbs but can affect any joint. Symptoms usually improve with rest. This contrasts with inflammatory conditions, where resting leads to 'gelling' or stiffness. Hypermobility has been shown to be associated with growing pains which are a separate entity characterized by bilateral, intermittent non-articular pains involving the lower limbs; occurring during late afternoons, evenings, often waking the child from sleep, with a normal physical examination and normal laboratory parameters (whenever performed) (12). Hypermobility can also be associated with joint dislocation / sprains, back-pain and anterior knee pain. There may be a past history of poor co-ordination in early childhood, 'clicky' hips, congenital hip dislocation, constipation, urinary tract infection and generalized pain (13).

Examination of a child with hypermobility

General examination should reveal a child with normal growth and systemic examination. The cardiovascular, respiratory and ocular features of hereditable connective tissue disorders such as Marfan's syndrome, Ehlers Danlos and Osteogenesis imperfecta should be absent, but there may be some overlapping subtle signs e.g. easy bruising, poor or atrophic scar formation and mild skeletal deformities (see Figure 2) (13, 14). The differential diagnosis of BJHS should be considered during the examination (see Table 1). Musculoskeletal examination will reveal increased range of joint movement, which may be generalized or localised to particular joints.

Figure 2: Clinical features seen in BJHS and hereditable connective tissue disorders (e.g. Marfan's syndrome, Ehlers-Danlos, Osteogenesis Imperfecta). TMJ = temporomandibular joint. UTI = urinary tract infection. Pes planus = mobile flat feet. Calcano valgus = lateral positioning of the heel. Spondylolysis = weakness or stress fracture in one of the bony bridges that connect the upper with the lower facet joints of the vertebra. Spondylolisthesis = anterior or posterior displacement of a vertebra in relation to the vertebrae below.

Heritable connective tissue disorders

- Marfan's
- Ehlers-Danlos

• Ostogenesis Imperfecta
Juvenile Idiopathic Arthritis
Pain syndromes • Diffuse idiopathic pain syndrome • Localized idiopathic pain syndrome • Fibromyalgia
Malignancy • Leukaemia • Ewing's Sarcoma • Osteosarcoma
Congenital syndromes with Hypermobility * • Down's • Williams • Sticklers

Table 1: Differential diagnosis to consider in the hypermobile child
* Usually pre-diagnosed on basis of characteristic features.

Establishing a diagnosis of hypermobility

The majority of cases of hypermobility are mild and easily recognized with good history taking / examination. Consider appropriate investigations when there are features of the differential diagnoses shown in Table 1. Otherwise, a positive diagnosis should be made to help to prevent inappropriate investigations. Explanation of the condition and reassurance that there is no evidence for any serious underlying pathology is very important for children and their families.

Management of the hypermobile patient

The role of the doctor

The child and family should be reassured that their symptoms will improve with time. It is important that the family realises that on-going pain, cracking or clicking of joints does not signify on-going damage to the joints, but that recurrent minor sprain-type injuries may occur due to ligamentous laxity and weakness of the supporting musculature. It should be emphasized that such injuries are usually self-limiting and can be treated with physiotherapy. Long-term / regular analgesic use is often unhelpful.

The role of the physiotherapist
Physiotherapy and exercise is the mainstay of treatment in more severe cases of hypermobility that impacts on daily activities (see Table 2). Hydrotherapy can be useful initially as the heat can provide pain relief and improve muscle spasm. Targeted or generalised dry land physiotherapy approaches are equally efficacious, leading to a significant and sustained reduction in joint pain (15). The rehabilitation programme must build-up at an appropriate pace to maintain the child and families confidence. For a child who has had 12 months of symptoms it should be expected that they might

take a further 12 months to recover. Activities such as swimming and cycling should initially be advocated, and once the patients' strength has improved, they should resume normal sporting activities.

• Increase muscle strength and stamina
• Reduce laxity of the joints by developing the supporting musculature
• Modify the child's gait to correct biomechanical abnormalities
• Improve joint proprioception
• Improve general fitness
• Enable the child and family to return to normal physical functioning and manage symptoms independently with minimal ongoing in-put from doctors or therapists

Table 2: Key aims of physiotherapy in hypermobility

The role of other therapists

Occupational therapy input can be especially useful when there are problems with manual dexterity and specific functional activities due to ligamentous laxity. Children with BJHS often adapt biomechanically when undertaking functional tasks, leading to pain and fatigue in compensatory areas. Pen grips and sloping writing surfaces can be used alongside hand-muscle strengthening exercises to help reduce the force required to sustain a pen grip. Splinting of hypermobile joints is not recommended as it can lead to further weakening of periarticular muscles. Hypermobile children often also have very pronated flat feet leading to abnormal biomechanical forces in the lower limbs. Orthotics can modifying the orientation and movement of the medical arch, ankle, sub-talar and knee joints (16), improving symptoms and gait biomechanics (17).

Behavioral interventions

Children with severe BJHS may get into a pattern of peaks and troughs in symptoms and activity, leading to major disruption to their lives and school refusal. In such situations, it is important to introduce the idea of pacing and 'evening out of activities' to the child and family. Specific tasks should be set as a minimum each day (e.g. school) with additional activities built up week after week. For this to work, the child, family and therapist must all be involved in planning of the pacing programme. Where pain is more generalised, associated with fatigue and loss of function, a clinical psychologist should be involved to help identify potential psychological stressors and coping strategies.

Conclusions

Hypermobility is a common problem, which leads to troublesome clinical symptoms in a minority of patients. Positive recognition of the condition at an early stage and explanation to families is key to altering the trajectory of the

condition. If left unrecognized, hypermobility can lead to the development of chronic pain, which will require intensive musculoskeletal rehabilitation and can have a significant impact on the life of the child and their family.

5 MCQ's

1. **Which of the following are associated with hypermobility? (tick all that apply)**
    a. Female gender
    b. Male gender
    c. Caucasian ethnicity
    d. Asian ethnicity
    e. Age

2. **Which of the following scoring systems have been validated for use in children? (tick all that apply)**
    a. Beighton score
    b. Brighton score
    c. Bulbena score
    d. Carter and Wilkinson score
    e. None of the above

3. **When do hypermobility related symptoms usually occur? (tick all that apply)**
    a. In the morning and after periods of immobility
    b. During daily activities (e.g. walking to school) and sports
    c. In the evening after an active day
    d. During the night

4. **Which of the following clinical symptoms and signs may be associated with benign joint hypermobility syndrome? (tick all that apply)**
    a. Short-lived joint swelling
    b. Skin elasticity
    c. Anterior knee pain
    d. Poor co-ordination
    e. Heart murmur

5. **Which of the following conditions is associated with benign joint hypermobility syndrome? (tick all that apply)**
    a. Congenital hip dislocation
    b. Constipation
    c. Inflammatory arthritis
    d. Chronic pain syndromes
    e. Marfan's syndrome

Answers to MCQ's

1. A, D, E correct

2. E correct
3. B, C, D correct
4. A, B, C, D correct
5. A, B, D correct

Further information for healthcare professionals:

Arthritis Research UK 'Hands On' practical guide to hypermobility

- http://www.arthritisresearchuk.org/health-professionals-and-students/reports/reports-archives.aspx

Detailed review on hypermobility in children and adolescents

- Murray, K. J. Hypermobility disorders in children and adolescents. *Best Pract Res Clin Rheumatol*. 2006;20(2):329-351.

Review of publications relating to differential diagnosis in hypermobility

- Tofts LJ, Elliott EJ, Munns C et al. The differential diagnosis of children with joint hypermobility: a review of the literature. *Pediatr Rheumatol.* 2009, **7**:1 doi: 10.1186/1546-0096-7-1

Patient information on hypermobility:

Arthritis Research UK website

- http://www.arthritisresearchuk.org/arthritis-information/conditions/joint-hypermobility.aspx

Hypermobility Association

- http://hypermobility.org

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