Biomechanical Pain and Hypermobility Syndrome in Children and Adolescents

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Differential Diagnosis....

- BIOMECHANICAL
- ARTHRITIS/INFLAMMATION
- MISC
- NON ORGANIC

Prince et al 2010 BMJ
Biomechanical Pain causes...

• **Traumatic** – eg ankle sprain; rectus femoris avulsion fracture; salter-Harris fractures
• **Atraumatic** – eg Anterior knee pain, back pain
• **Overuse** – eg Apophyseal injuries such as Osgood-Schlatters, Severs; osteochondritis dessicans
Risk Factors

- Genetic predisposition
- Growth spurt
- Activity levels
Risk Factors

• Alignment Abnormalities

• Level of conditioning

• Obesity
Risk Factors

• Parental pressure

• Habitual movements/sustained postures

• Hypermobility
Hypermobility

• Thought to have been described first by Hippocrates
• Is common in children
• More marked in females
• Hypermobility diminishes through adult life
Why is Hypermobility relevant to Paediatric Rheumatologists?

• Recent studies have shown hypermobility is a risk factor for musculoskeletal pain in adolescence increasing 2-fold chances of developing pain
• This risk is heightened when obesity also present
• Hypermobility particularly increases risk of MSK pain in shoulder, knee and ankle/foot
Prevalence of Joint Hypermobility in Different Populations of Children

- 7% UK school children
- 8% Greek children
- 12% US school children
- 14% Dutch children
- 40% Brazilian school children
- 43% West African children
- 25% Asian children
Hypermobility

• “Where most of an individual’s joints move beyond the normal range taking into consideration age, gender, and ethnicity”

• Beighton, Grahame & Bird 1999
Differential Diagnoses associated with Hypermobility

• Osteogenesis Imperfecta
• Downs Syndrome
• Stickler Syndrome
• Marfan Syndrome
• Ehlers-Danlos Syndromes
Marfan syndrome

**Major manifestations**
- Skeletal
  - Pectus excavatum or pectus carinatum
  - Arm span:height >1.05
  - Scoliosis
  - Hypermobility
- Ocular
  - Ectopia lentis
- Cardiovascular
  - Aortic dilatation
  - Aortic dissection
- Dura
  - Lumbosacral dural ectasia
- Family
  - First degree family member with Marfan syndrome

**Minor manifestations**
- Skeletal
  - Pectus excavatum
  - Hypermobility
  - High arched palate
- Ocular
  - Myopia
  - Flat cornea
- Cardiovascular
  - MV prolapse
  - PA dilatation
- Pulmonary
  - Pneumothorax
  - Apical blebs
- Skin
  - Striae
  - Recurrent hernias

http://www.marfan.org
Ehlers-Danlos Syndromes
Clinical Presentation

• Persistent joint pains/muscle pain after activity/end of day -
• Fatigue
• Nocturnal Idiopathic pain common
• Subluxation, dislocations, sprains, strains, soft tissue lesions more common
Clinical Presentation

• Mild swelling and stiffness often related to activity
• Delayed motor development and clumsiness, slow to walk, not crawling
• Gait problems – toe walking, in-toeing
• Fidgety and difficulty getting comfortable
Clinical Presentation

• Poor proprioception and coordination

• Reduced muscle tone/Hypotonia

• Poor stamina and general fitness

• Pes planus

• Skin hyperelasticity/easy bruising
Clinical Presentation

- Writing difficulties
- Bowel/bladder problems/Hx herniae: umbilical and inguinal/Gastro oesophageal reflux
- DDH/”clicky hips”
- Cardiovascular autonomic dysfunction
Associated Problems

• Missed school
• Missed PE and sporting hobbies
• Poor sleep
• Pain intolerance – resistance to lignocaine
• High association with Chronic pain syndromes
Causes of Pain

• Muscle spasm – feature of fatigued muscle
• Muscle imbalance – strong get stronger
• Poor muscle control into hypermobile range
• Reduced muscle stamina leading to earlier onset muscle fatigue
Causes of Pain

• Often can present during period of weight gain or rapid growth

• Biomechanical dysfunction eg tight hamstrings

• Overstretch of soft tissues due to proprioceptive impairment
Causes of Pain

• Biomechanical joint inflammation following increased activity

• Pain and swelling inhibits muscle function

• Enhanced pain perception/pain intolerance
Beighton Scoring system for Joint Hypermobility

Figure 1. Beighton’s modification of the Carter and Wilkinson scoring system. Give yourself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points.
1998 Brighton revised diagnostic criteria for Joint Hypermobility syndrome

- REVISED DIAGNOSTIC CRITERIA FOR THE BENIGN JOINT HYPERMOBILITY SYNDROME (BJHS)
  - **Major Criteria**
    - A Beighton score of 4/9 or greater (either currently or historically)
    - Arthralgia for longer than 3 months in 4 or more joints
  - **Minor Criteria**
    - A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
    - Arthralgia (> 3 months) in one to three joints or back pain (> 3 months), spondylosis, spondylolysis/spondylolisthesis.
    - Dislocation/subluxation in more than one joint, or in one joint on more than one occasion.
    - Soft tissue rheumatism. > 3 lesions (e.g. epicondylitis, tenosynovitis, bursitis).
    - Marfanoid habitus (tall, slim, span/height ratio >1.03, upper: lower segment ratio less than 0.89, arachnodactyly [positive Steinberg/wrist signs].
    - Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring.
    - Eye signs: drooping eyelids or myopia or antimongoloid slant.
    - Varicose veins or hernia or uterine/rectal prolapse.
  - **The BJHS is diagnosed in the presence two major criteria, or one major and two minor criteria, or four minor criteria. Two minor criteria will suffice where there is an unequivocally affected first-degree relative.**
  - BJHS is excluded by presence of Marfan or Ehlers-Danlos syndromes (other than the EDS Hypermobility type [formerly EDS III] as defined by the Ghent 1996 (8) and the Villefranche 1998 (9) criteria respectively). Criteria Major 1 and Minor 1 are mutually exclusive as are Major 2 and Minor 2.
Multidisciplinary Management of Benign Joint Hypermobility

• Make diagnosis; onward referral as necessary. (stop unnecessary investigations)

• Educate parents and child – de-medicalise

• Analgesia
Multidisciplinary Management of Benign Joint Hypermobility

- Physiotherapy
- Occupational therapy
- Podiatry – orthotics
- Psychology – if required
Physiotherapy Management

• **Education** of family and child:


• Increase strength of muscles through hypermobile range with specific prescribed graded exercise programme. Use both open and closed chain exercise. Muscles are our only **dynamic control of our joints**

• Can use weights in children – up to 2.5 kg and aim for high repetitions up to 30
Physiotherapy Management

• Improve proprioception and balance
• Improve stamina and endurance of muscles
• Improve general fitness of child
• Address any biomechanical abnormalities/gait
  Improve core stability

• SELF MANAGEMENT – home programme
• School liaison and advice re PE
Best Practice in exercise prescription

• No resistance until 15 reps
• Strength and fitness increase with reps and resistance
• Need to train 20-30 min each session
• Min 2-3 times per week (no increase in benefit of exercising > 4/7)
• Strength gains lost after 6 weeks of deconditioning

Paediatrics June 2001 p1470-1472
Additional benefits.....

- Enhanced motor skills and performance
- Strengthens bones
- Weight management
- Enhance psychosocial well-being
- Cardio-vascular health
- Enhanced resistance to sports injuries
- Habits continued into adult life.....
Pointers....

- **The British Society for Paediatric and Adolescent Rheumatology (BSPAR)**

- **Hypermobility Association HMSA**
  - Phone: 0845 345 4465
  - Email: Web: [http://www.hypermobility.org](http://www.hypermobility.org)

- **The Association of Chartered Paediatric Physiotherapists Website**
  - Left hand column – Click on “information for parents “– click on “Symptomatic Hypermobility” leaflet for parents
  - [http://apcp.csp.org.uk/](http://apcp.csp.org.uk/)