

Paediatric MSK problems: red flags and normal variants

Children are different from adults. When they present with bone, joint or muscle pain, we need to think of a different set of differential diagnoses. We also need to talk to them and examine them differently. If this is an area where you lack confidence, you might find our FREE Paediatric MSK webinar useful. You can sign up here www.gp-update.co.uk/webinars/versusarthritis.

Why is it difficult?

The challenge is that many of the paediatric MSK problems than present in primary care are benign 'normal variants' that time will solve.

But for each of these innocent presentations that will form our 'bread and butter' in general practice, there is an occasional lurking 'evil twin', and we need to have these at the back of our mind so we don't miss them!

A note about 'trauma' and falls:

It isn't always easy to get a clear history, and beware: a history of a simple fall/trauma "a few days ago" may be a red herring, both because these can be common in children and children with joint or muscle problems may be more likely to fall. Children also like to create an explanation for what is happening to them, and linking a symptom with an event is a natural thing to do, even if they are not connected. We also need to consider the possibility of non-accidental injury.

Useful questions for assessing children with MSK problems

acute MSK problems (Paeds and Child Health 2019;29(12)503): **Question?** Why? Tips/caveats and pitfalls How is their general health? Specifically ask about these - they may not be volun-Looking for red flags: teered. Night pain. • Bone pain. Fever. Anorexia/weight loss. Malaise. Did anything happen before Looking for: Falls in children are common and may be a red herthey got these symptoms? ring, and are more common in children with arthritis. Traumatic causes. ٠ Look for inconsistency between the circumstance and Reactive causes. extent of injury – think non-accidental injury. What are they like in the Morning stiffness may show itself as difficulty getting Looking for: morning? dressed or managing stairs. Mechanical vs. inflammatory Child may be particularly miserable in the mornings. features. What can they no longer do Looking for : Ask about: that they could before? • Delay in achieving motor milestones. **Regression of motor milestones** which can be a sign of inflamma- Loss of achieved motor milestones. tory or muscle disease.

The clinical review highlighted some useful questions to ask children and their grown-ups when they present with

Examining children with MSK problems

We all have different levels of skill at this. If you have not come across the pGALS musculoskeletal assessment system before or need a refresher in your paediatric MSK examination skills, we strongly recommend taking a look at this. There is a link to the pGALS site in the useful resources section below. We have also created a one-page summary of this, which can be found in the *pGALs* article in the online handbook.

'Red flag' MSK presentations in children

We should think carefully about any of the following presentations in children:

- Acute painful limp (or a chronic persistent limp).
- Acute joint pain.
- Joint swelling.
- Night pain: persistent or not responsive to simple analgesia, e.g. paracetamol, ibuprofen.
- Deep and throbbing 'bone pain' the child often points to the bone rather than joint.
- Systemic symptom, e.g. loss of appetite, fatigue, weight loss, pallor, lymphadenopathy.
- Delayed, loss or regression of motor milestones.
- Weakness.

Let's look at the 'big 6' serious MSK conditions that relate to these red flag presentations.

Serious MSK conditions not to miss!

Here are the 'big 6' things we don't want to miss:

- Malignancy.
- Infection.
- Non-accidental injury.
- Inflammatory arthritis.
- Progressive muscle disorders.
- Orthopaedic emergencies (non-traumatic).

Malignancy (especially sarcomas and haematological malignancy)

Presentation	Action
Childhood cancer is rare for the individual GP, and often pre-	Consider bone sarcoma (NICE 2015 NG12):
sents non-specifically (BJGP 2012;62(600):e458-e460).	Consider urgent direct-access X-ray (within 48h) for children
Pain may be the first and sometimes only presentation of	and young people with:
childhood malignancy, including cancers of the bone, muscle	Unexplained bone swelling.
and leukaemias.	Unexplained bone pain.
Worrying features include:	Refer using suspected cancer pathway within 48h if X-ray is
Bone pain/night pain waking from sleep.	suggestive of bone sarcoma.
Malaise.Bony lump or swelling.	Consider soft tissue sarcomas (NICE 2015 NG12):
 Bony tenderness. 	Consider an urgent direct-access USS within 48h for children and young people with:
Limb pain is a presenting feature in 43% of children with leu-	 Unexplained soft tissue lump that is enlarging.
kaemia, and joint pain is present in 11% (ADC 2016;101:894).	Make an urgent appointment within 48h for children if:
Note: this may present as a new limp or change in behav- iour/motor function. Also look for signs and symptoms of hae- matological malignancy.	 Ultrasound scan findings are suggestive of soft tissue sarcoma OR Ultrasound scan findings are uncertain and clinical concern persists.
	Consider haematological malignancies (NICE 2015 NG12):
	Offer a FBC within 48h to children and young people with any of:
	• Pallor.
	Persistent fatigue.
	Unexplained fever/persistent infection.
	Generalised lymphadenopathy.
	Persistent or unexplained bone pain.
	• Unexplained bruising or bleeding. In practice, sarcoma may be one of a number of differentials,
	and a paediatric assessment may be more appropriate. A full
	blood count may also be helpful.
	If we are in any doubt whatsoever, we should speak to the pae- diatric team on call.

Infection (septic arthritis and osteomyelitis)

Presentation	Action
 Consider in: Pyrexia of unknown origin, especially in young children when swelling/pain is often late. Acute joint pain, especially involving a single joint. Fever +/- systemic symptoms (these are not always present!). Pain may be severe and the child inconsolable (<i>but the child may not complain of pain and rather be irritable</i>). Unable to weight bear/acute limp. Very rare: consider discitis in the febrile infant who is inconsolable during nappy changes (when the spine is flexed). (<i>Remember: crystal arthropathy is very rare in children and is not a diagnosis to make in primary care.</i>) Have a lower threshold to consider in immunosuppressed children, neonates (especially if premature) and children with sickle cell (BMJ 2014;348:g66). 	Careful examination is important, particularly in young chil- dren; look for crying/distress when particular joints are moved. If you suspect septic arthritis or osteomyelitis, refer immedi- ately, following local pathways. Time is joint: the longer septic arthritis goes untreated, the more permanent joint damage occurs. This is an urgent diagno- sis to make. Do not take or wait for a FBC and inflammatory marker re- sults in primary care. Particularly in young children, raised inflammatory markers may be a late sign.

Non-accidental injury

Presentation	Action
 We should think 'safeguarding' in every single consultation. In MSK presentations, NICE (NG76 2017) reminds us that we should consider if non-accidental injury is a possibility if: An injury or presentation incongruent with history. There is delayed presentation of significant injury or pain. Bruising in the shape of a hand/stick/ligature/fingertip, ANY bruise in a child who is not independently mobile, or bruises on non-bony parts of the face or body. 	Follow usual safeguarding procedures. If you are referring for further assessment, communicate your concerns clearly to the admitting team.

Inflammatory arthritis

Presentation	Action
 Presentation This may be less rare than you think, having a similar prevalence to diabetes and epilepsy in children. It can present differently from in adults. Consider inflammatory arthritis as a possibility in a child with (Paeds and Child Health 2019;29(12)503): Change in behaviour, e.g. reluctance to walk usual distances, wanting to be carried, not wanting to play, new or increasing falls/'clumsiness', parent reports the child "moves like an old person". Painful or swollen joint(s) – in the absence of trauma or infection, juvenile idiopathic arthritis is the most likely cause of a single swollen joint (children very rarely get gout). Pain may not be verbalised, especially by younger children; look for stiffness, swelling, a limp or change in func- 	 Action The initial presentation will determine your action: Acute limp or single swollen joint: will need same-day assessment following local limping child protocol. If the presentation is more subtle or gradual, it may be appropriate to refer direct to paediatric rheumatology (or general paediatrics), depending on your local pathways. Like in adults, no blood test performs well enough to rule inflammatory arthritis in or out, so refer on the basis of a good story.
 tion. May exhibit morning stiffness or gelling, e.g. stiffness after sitting still for a long period (variable behaviour pattern across the day). 	

Progressive muscle disorders, e.g. muscular dystrophies (or inflammatory muscle disorders)

Presentation	Action
MSK problems, e.g. falls, limp, delayed motor milestones,	If a neuromuscular condition is suspected:
may be the first presentation of a primary muscle disorder. Look for:	• Do a creatine kinase but do not be reassured in the pres- ence of a good story/abnormal examination if this is nor-
	mal.

 Progressive proximal weakness, e.g. difficulty with stairs, difficulty jumping from age 2–2.5y, Gower sign (using 	 LDH is more sensitive for inflammatory muscle conditions, e.g. dermatomyositis. Refer to paediatrics (or paediatric neurology, depending on your local pathways).
 hands to walk up legs when rising from floor). Frequent falls. Abnormal gait. Delay or regression of gross motor milestones – regression is particularly concerning as it may suggest acquired neurological or muscle disorder. Myoglobinuria (Coca-Cola urine) – suggests muscle destruction; rare. For a reminder of the signs of muscle weakness, see the useful websites below for a series of videos demonstrating these. 	

Orthopaedic emergencies (non-traumatic)

Presentation	Action
 Here, we are considering Perthes' disease and slipped upper femoral epiphysis (SUFE) – need a reminder? Perthes' disease (BMJ 2010;341:c4250): Idiopathic avascular necrosis of the developing femoral head. Typically presents in boys aged 4–8y. Slipped upper femoral epiphysis (BMJ 2009;339:b4457): Occurs during rapid pubertal growth. More common in obese children. Can be triggered by an episode of trauma or occur more chronically over a period of weeks (don't be put off by an acute-on-chronic picture). Both conditions may present as subacute hip, knee or thigh pain (always examine the joint above and below the reported pain). Examination will show reduced internal rotation and pain at the extremes of movement. 	 If either of these conditions are suspected, call and refer the same day to orthopaedics (sometimes, if SUFE has a more chronic presentation, they may elect to see in outpatients). Diagnosed by plain AP radiograph of the pelvis, though typical changes may be absent in early disease. Management may include bed rest, physiotherapy and, in some cases, surgery. Note that previously undiagnosed developmental dysplasia of the hip may also present this way, and, if suspected, can be referred routinely to orthopaedics (though, in reality, it is likely to be difficult to make this judgement call in primary care!).

The limping child: what to do

This is one of the most common primary care 'red flag' MSK presentations in children. So, who needs referral? The most likely cause(s) of an acute limp varies with age. As we assess the child, we need to keep the differential diagnosis in mind and ask ourselves a number of questions (BJGP 2020;70:467-468):

- Is there a history of trauma? (remembering the caveat above that children may link a pain to a minor incident some days before which is actually a red herring).
- Could this be septic arthritis?

Differential diagnosis of acute limp in children			
In children of all ages, consider:			
Septic arthritis/osteomyelitis.			
Non-accidental injury.			
• Testicular torsion/inguinal hernia/a	ppendicitis/UTI.		
Juvenile idiopathic arthritis.			
• Metabolic conditions, e.g. rickets.			
• Haematological disease, e.g. sickle of	cell.		
Age 1–3	Age 4–10	Age 11–16	
Toddler's fracture.	Transient synovitis.	Slipped upper femoral epiphysis	
• Developmental dysplasia of hip.	• Perthes' disease.	Primary bone tumours	
Neuroblastoma.	• Acute lymphocytic leukaemia.	Osgood-Schlatter disease	

Who should we refer?

Refer children with an acute limp for same-day assessment if they are:

• Aged <3y.

- Unwell: fever or systemically unwell.
- Non-weight bearing.
- Experiencing painful or restricted joints.
- Immunosuppressed.
- Age >9y with painful or restricted hip movements (to exclude slipped upper femoral epiphysis).
- Or if non-accidental injury is suspected.

When can we manage in primary care?

A period of watchful waiting in primary care may be reasonable for **a child aged between 3 and 9y who is well, afebrile, mobile and limping for less than 72 hours** (NICE CKS accessed December 2020).

Why?

• Transient synovitis is the most common diagnosis in this group.

Primary care management

- Recommend rest and simple analgesia.
- Safety-net carefully: advise parents to take the child immediately to A&E if symptoms worsen or the child develops fever or systemic symptoms.
- Reassess after 48–72h: if symptoms are resolving, a diagnosis of transient synovitis can be made without further investigation.
- Offer further follow-up at 1 week: if symptoms have resolved completely, no further action is required. If symptoms have not resolved or any uncertainty about diagnosis remains, refer to paediatrics or paediatric orthopaedics for further assessment.

Having considered red flag presentations and conditions in some detail, let's now look at their more benign twins – the normal variants.

Lower limb 'normal variants'

"Are Jani's legs normal?...They look a bit wonky."

25–50% of new paediatric orthopaedic referrals are for normal physiological variants of the growing child.

Before determining that something is a normal variant/benign, we should ensure that we have considered and ruled out the red-flag conditions detailed above, and that **the following rules are met** (BMJ 2015;351:h3394):

Rules of normal variants

Reassuring features	Worrying features that warrant further assessment
Normal growth	Abnormal growth
Symmetry	Developmental delay
No pain	Asymmetry
No limp	Limping
Normal function	Restricted activity
	Rigid/restricted joints
	Swollen joints/bones
	Abnormal muscle tone
	(Also consider obesity and vitamin D status)

Assessing normal variants

This table summarises the range of what is considered normal variation.

This is pretty impossible to remember but good to know where to look!

Essentially, in the develo	ning child things r	noint out then in	then straighten un
Essentially, in the acvert	ping cinic, cinigs p	point out, then m	inch strughten up.

	Age range for normal vari-	Useful tips	When to refer?
	ant		
Bow legs	Birth to 2y.	With legs held with ankles together, meas-	 If persists beyond the age of
(genu		ure the intercondylar distance at knees;	Зу.
varum)		should be <6cm.	Asymmetry.

Knock knees (genu valgus) Flat feet	Age 3 to 6y. Universal until age 3y when foot arches start to develop. Most flexible flat feet re- solve between 4 and 8y.	Consider vitamin D deficiency if se- vere/persists beyond usual age range, or associated with short stature. Refer if persistent beyond age 3y, asym- metrical or getting worse over time. With legs held with knees together, meas- ure the intermalleolar distance at the an- kles: should be <8cm. Consider vitamin D deficiency if se- vere/persists beyond usual age range, or associated with short stature. Foot should be flexible and painless. Arch should be flexible and painless. No association with pain/functional prob- lems later in life.	 If occurs in child age <2y. Asymmetry. Painful, rigid flat foot.
In-toeing	Birth to 8y.	Custom orthoses are not necessary in asymptomatic children. Three causes:	Rigid foot.
		 Metatarsus adductus (congenital foot deformity in 1/1000 births. Foot has a curved border. If foot is rigid or not improving by 6–9m, casting may be helpful). Internal tibial torsion (feet internally rotated, patellae in neutral position). Femoral anteversion (feet and knees internally rotated, child often W-sits and runs with an odd egg beater pattern). Regardless of cause, vast majority of cases resolve by 8y without intervention – surgical intervention would not be considered before this age. 	 Asymmetry. Persists beyond age 8y with functional impact.
Out-toeing	From first steps. Resolves between 18 and 24m.	If persists beyond this age or starts <i>de</i> <i>novo</i> in older children/adolescents, will need assessment.	Persists beyond age 2y. Occurs new in older children (con- sider SUFE/Perthes').
Tip-toeing	Common from 10–18m as walking develops (helps them to balance). Usually resolves by age 3; can continue until 6 or 7y.	 Assessment is to rule out neurological causes, muscular conditions, inflammatory arthritis and developmental disorders as possible causes. Assess global development. Examine the spine, muscle bulk and tone, ankle extension. Is the gait equal on both sides? Is there pain or stiffness? Is the tiptoe gait a new finding in a child who previously walked normally? 	 Persists beyond age 3y. Unilateral/asymmetrical. Increased muscle tone. Unable to extend at ankles. Spinal abnormality. Developmental delay.
Hypermo- bility	Affects around 30% of chil- dren, with only about 3% having significant sympto- matic disease. The vast ma- jority will have NO underly- ing connective tissue disor- der but we should consider Marfans and EDS if there are other features (see main article on <i>Joint hypermobil- ity</i> in the online handbook).	The Beighton score is not validated for use in children. (See full article on <i>Joint hypermobility</i> in the online handbook.)	 Pain affecting activity. Recurrent injury.

Growing pains

"Petra (age 5) gets these pains in her legs. They happen once or twice a week as she is getting ready for going to bed...they don't seem to stop her doing anything. She never seems to complain of them in the morning."

This is a fairly typical story of growing pains, but this will not be a quick consultation in primary care as growing pains are a diagnosis of exclusion, and a careful assessment is required.

Growing pains are common and may occur in up to one-third of children at some point. Their cause is unknown, but they are a common source of misdiagnosis and delayed diagnosis because both parents and GPs may mislabel symptoms as growing pains when they are actually something else (Sports Health 2017;9(2):132).

Before considering/suggesting that pain could be growing pains, ask whether the story and presentation meet ALL the rules of growing pains?

Rules of 'growing pains'

- Age 3–12y.
- Pain symmetrical in lower limbs and not limited to joints.
- Pains never present at the start of the day after waking.
- Child doesn't limp.
- Physical activity is not limited by the pain.
- Physical examination normal.
- Systemically well.
- Major motor milestones are normal.

Management

If all the rules are met and no other explanation is identified as part of a full assessment (including examination), then management is usually simple measures such as massage, stretching and, if required, simple analgesia with paracetamol or ibuprofen.

We should safety-net carefully about presenting again if symptoms become more severe or if new symptoms develop. If you have remaining uncertainty, reassess in a few weeks and use a test of time to observe what happens.

 Paediatric MSK problems: red flags and normal variants Remember the normal variants and their evil twins. Explicitly look for red flags and consider non-accidental injury. Beware asymmetry, a limp, children who are systemically unwell, children with fever without a focus, loss or regression of motor skills, loss of function, an unwillingness to do things they would normally enjoy. A history of a fall or trauma may be a red herring. If there are no red flags, we can cautiously observe children aged between 3 and 9y who are well, afebrile, mobile but limping for less than 72h; transient synovitis is a common diagnosis in this age group – but we should proactively follow-up and safety-net. Younger children may not report pain, and a change in function may be the first sign of a problem. Normal variants are usually painless, symmetrical and do not impact function.
 Growing pains are a diagnosis of exclusion and must 'follow the rules'. If this is an area you feel less confident in, why not watch our FREE webinar, produced in partnership with Versus Arthritis, which can be found here: www.gp-update.co.uk/webinars/versusarthritis

www	Want to learn more or watch videos of how to examine children? Look at this resource that is free for health professionals (but you have to register): <u>http://www.pmmonline.org</u> Want to brush up on spotting children with muscle weakness and when we might do a CK?: <u>https://childmuscleweakness.org/video-library/#core-signs</u> Need support for children or young people with arthritis? Versus Arthritis can support children, young people and their families: <u>https://www.versusarthritis.org/about-arthritis/young-people/</u>