Red Whale

In association with VERSUS ARTHRITIS

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis (JIA) is more common than we might realise, affecting the same proportion of children as type 1 diabetes or childhood epilepsy.

It is often diagnosed relatively late; this is due to a combination of factors, including delayed recognition in primary care and general paediatric services. Where the diagnosis is suspected, all children should see a paediatric rheumatologist; this may be a direct or indirect referral via general paediatrics, depending on local pathways.

In this article, we look at when we should consider the diagnosis, what we should do and how we can support children with this condition and their families.

The main reference for this article is a helpful review article from Paediatrics and Child Health (Paediatrics and Child Health 2019;29(12):503). You may also find the article on *Paediatric MSK: red flags and normal variants* helpful.

What is juvenile idiopathic arthritis?

- The most common chronic inflammatory arthritis of childhood, with a prevalence of 1 in 1000 children.
- Diagnosed with the presence of inflamed swollen joints for >6w duration in children and young people aged <16y.
- There are 2 'peak times' for it to occur – preschool and adolescence.
- Not a single condition but made up of a heterogenous group of subtypes; the most likely subtype will be diagnosed by secondary care and this will influence management.
- **It is associated with anterior uveitis which is often 'silent' in children but, if undetected, can lead to irreversible visual loss; all children with JIA will attend regular eye appointments.**
- **Like inflammatory arthritis in adults, early diagnosis and intervention significantly improve the prognosis and outcomes of children with JIA, with less joint damage, less disability and better quality of life.**

Presentation

It can present differently from inflammatory arthritis in adults, so we need an even greater index of suspicion to consider it, and need to ask the right questions.

As well as the usual symptoms and signs we may see in adults, beware of and look for:

- Change in behaviour, e.g. reluctance to walk usual distances, wanting to be carried or dressed, not wanting to play, new or increasing falls/'clumsiness', parent reports that 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**. Other causes of atraumatic single swollen joints are very rare in children, though in teenagers we should also consider osteosarcoma.
- Pain may not be verbalised, especially by younger children; look for stiffness, swelling, a limp or change in function.
- May exhibit morning stiffness or gelling, e.g. stiffness after sitting still for a long period – *may struggle after long journeys in the car, a double lesson sat still at school or 'carpet time'.*
- Systemic features of fatigue, lethargy and poor appetite may be present.

To consider a diagnosis of JIA, symptoms must be present for >6 weeks.

A note about 'trauma' and falls:

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 be vigilant and consider the possibility of non-accidental injury.

Examination

MSK

- Use the pGALS system to examine the child (see the link below to the pGALS site if you are not familiar with this – it is a free resource for healthcare professionals). A 'copy me' approach can be a useful way of getting children to actively move their joints. *You may also find the separate pGALS article helpful.*
- Look particularly for asymmetry by comparing each joint in turn. This can be an effective way of detecting swelling and stiffness/restricted movement, which can be more difficult in children, who are inherently more flexible and wriggly!

General

Multisystem involvement should also be considered, particularly if any red flag symptoms are present.

- Check for lymph nodes, hepatosplenomegaly.
- Look for rashes/nail signs, including psoriasis, and signs of anaemia.
- Look at eyes for signs of uveitis, though remember that this may be silent in children; if a form of JIA is diagnosed, all will need an eye assessment.
- Dip urine for blood and protein; check BP.

Reactive and post-viral arthritis

This can mimic symptoms of JIA.

- It usually occurs 7–10 days after an acute illness, e.g. varicella, EBV, rubella, GI infection. Also consider STIs in sexually active young people, which may need treating.
- Remits spontaneously within 2–3 weeks. Management is symptomatic with NSAID analgesia if needed.

Subtypes of JIA and their clinical features

From a primary care perspective, we need to recognise that this could be an inflammatory arthritis and refer. Identifying subtypes is something that will take place in secondary care. However, this table may be useful for deciphering discharge letters.

Subtype of JIA	Features
Oligoarthritis	1–4 joints affected in the first 6 months. Then, after 6 months, it may be subclassified as: • Persistent: continues for >6m but never affects more than 4 joints. • Extended: affecting >4 joints after the first 6m. This is the most common subtype (70%) and carries the best prognosis (many cases will resolve before adulthood). Often presents in preschool girls with knee or ankle involvement.
Polyarticular	Affects 5 or more joints in the first 6 months. May be subclassified as: • Rheumatoid factor positive (rare, often in teenage girls, more likely to mimic RA). • Rheumatoid factor negative (most common).
Enthesitis-related arthritis	Arthritis and enthesitis *(inflammation at the insertion of a ligament, tendon or capsule to bone, particularly around the foot or knee)* or Arthritis or enthesitis and at least 2 of: • Sacroiliac joint tenderness. • Inflammatory back pain. • HLA-B27 positive. • Family history of HLA-B27-positive disease.
Psoriatic arthritis	Arthritis and psoriasis or Arthritis and at least 2 of: • Dactylitis. • Nail changes. • Family history of arthritis.
Systemic onset	Arthritis, fever (daily recurrent) and rash (often maculopapular and worse with fever). • Least common subtype (10% of cases); may present as a 'fever of unknown origin'.

	• One-third of children develop severe polyarthritis. • Systemic features can pre-date arthritis by some weeks.
Undifferentiated	Unknown cause or not fulfilling any of the above.

Referral

If you suspect JIA as a possible diagnosis, the initial presentation will determine your action:

- In this context, an acute limp or single swollen joint will need same-day assessment following the limping child protocol, either via acute paediatrics or ED. Do not do bloods in primary care and wait for the results. For information on ALL limping children, see the article on *Red flag MSK presentations in children*.
- 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 we should refer on the basis of a good story and not delay the referral for blood tests – these will be done in secondary care. *We may consider an urgent FBC if one of our differentials is malignancy (see article on* Red Flag MSK presentations in children*).*
- We can offer NSAIDs for symptom relief (if no contraindications) while waiting for an appointment.

Management

This will be determined by the paediatric rheumatology team, and may include:

- NSAIDs for symptomatic relief.
- Corticosteroids: usually intra-articular or pulsed IV if needed. Oral are generally avoided, though may be needed for more severe cases.
- Methotrexate is main DMARD used in JIA.
- Biologics as per NICE guidance (2015 TA373).

Vaccinations

- It is important that ALL children and young people follow the immunisations schedules.
- Live vaccines are *usually* contraindicated in immunosuppressed patients; this will include JIA patients on steroids, DMARDs (usually methotrexate) and biologics.
- However, advice is constantly changing and we should consult the green book (e.g. children on methotrexate can now receive MMR and varicella vaccine).
- Flu vaccination should be offered in the form of inactivated flu vaccine NOT the nasal spray.
- Varicella vaccination may be indicated if there has not been previous exposure, prior to starting immunosuppression.

	**Juvenile idiopathic arthritis** • As common in children as type 1 diabetes or epilepsy. • Early diagnosis and treatment improves prognosis. • Most common atraumatic cause of a swollen joint lasting >6w. • Ask about changes in behaviour and function. • Compare both sides to look carefully for swelling and stiffness. • Refer on the basis of a good story; no blood tests we do in primary care can rule out JIA.
	**PGALS and more information can be found here:** http://www.pmmonline.org/ **Information for children, young people and their families:** Versus Arthritis has extensive information designed to support children, their families and their teachers: https://www.versusarthritis.org/about-arthritis/young-people/ There is also a Versus Arthritis symptom tracker app for teens and young adults which is free to download: https://www.versusarthritis.org/about-arthritis/young-people/arthritis-tracker/ Versus Arthritis Young People and Families Services: As primary care clinicians, we can refer young people with long-term MSK conditions to VA for support by simple email: • 12–18 years old and their families in England: YPF@versusarthritis.org

	• 0–25 years old and their families in Wales: YPFSWales@versusarthritis.org • 10–25 years old and their families in Scotland: YPFScotland@versusarthritis.org • 0–25 years old and their families in Northern Ireland: ypfsNI@versusarthritis.org