Diagnosing arthritis in children

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Abstract

Arthritis in children is common, and a major cause of potential morbidity, with significant long-term consequences, joint damage and disability if left untreated. Diagnosing Juvenile Idiopathic Arthritis (JIA) can be challenging, and relies heavily on clinical assessment; investigations are helpful to exclude other conditions, but are often normal in JIA at presentation. The history may be vague, and the child may be too young to verbalise symptoms; detailed probing for inflammatory symptoms and a comprehensive examination of the child's joints are therefore essential. If JIA is suspected, early referral to specialist multidisciplinary teams facilitates prompt treatment and prevention of complications. The emergence of novel and biologic agents, as well as earlier and more aggressive approaches to treatment, have helped to significantly improve clinical outcomes.

Keywords joint swelling; juvenile idiopathic arthritis; limping child; pGALS

Juvenile Idiopathic Arthritis (JIA) is the most common, chronic inflammatory arthritis of childhood, encompassing a heterogeneous group of conditions of unknown aetiology with the same prevalence as childhood epilepsy and diabetes (1 in 1000); see Table 1 for a description of the individual JIA subtypes. The emergence of novel biologic agents, as well as a move towards earlier and more aggressive treatment regimens, has dramatically improved the outcome for children and young people (CYP) with JIA. Furthermore, early diagnosis and access to specialist care facilitates prevention of complications such as joint damage and visual loss due to uveitis, which cause disability and adverse quality of life. There is, however, a recognised delay in diagnosis and referral for CYP with JIA to paediatric rheumatology services; the explanation is multifactorial, including complex pathways of care for CYP presenting to healthcare professionals in primary care and hospital services rather than paediatric rheumatology directly, reduced awareness about arthritis, and many clinicians who first encounter these children report low self-confidence in their musculoskeletal (MSK) clinical skills.

This article aims to increase awareness and knowledge about how to identify CYP with possible JIA, with three illustrative clinical case scenarios.

Case 1: Chloe's story

Chloe is a 3-year-old girl who presented to the emergency department (ED) with her Mum due to a 2-week history of intermittent limp. Up until this point she had been walking confidently from around the age of 14 months, but now asked to be carried downstairs in the morning. Mum also mentioned that she was "grumpy" after car journeys and "just not right". That said, Chloe was not complaining much in the way of pain, despite falling over a few days before the onset of her symptoms. A recent coryzal illness had settled in the last week.

In the department, Chloe appeared well and was apyrexial; general assessment (cardiovascular, respiratory and abdominal examination) was normal. A paediatric Gait, Arms, Legs and Spine (pGALS) assessment confirmed a limp, with the left knee held in flexion, along with an effusion and thigh muscle wasting (see Figure 1). She had normal movement in both hips, however there was restriction and possible swelling in her right ankle. Investigations revealed normal full blood count (FBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and sterile blood cultures; an X-ray of her knee and ankle did not show any bone pathology. She was discharged with a prescription for ibuprofen and referred to the local paediatric rheumatology team as an outpatient.

Case 2: Lily's story

Lily is a 12-year-old girl referred urgently to paediatric rheumatology by her GP with a 2-month history of painful, swollen metacarpophalangeal (MCP) joints, bilateral knee and ankle pain, and associated lethargy. She described feeling stiff in the morning for around 90 minutes, and also after sitting for a prolonged period of time (e.g. after a double lesson of maths). Lily was a keen runner but had stopped this activity due to pain. Her appetite was poor and her parents reported that she had become more withdrawn in recent weeks.

In clinic, Lily looked pale but did not have any palpable lymph nodes or hepatosplenomegaly; cardiovascular, respiratory and abdominal examination was unremarkable. She did however have obvious swelling of her knees, ankles, elbows, wrists, and the small joints of her hands with restricted passive movement (see Figure 2).

Case 3: Connor's story

Connor is a 3-year-old boy who presented to the ED with a 5-day history of temperatures and leg pain. This was on the background of coryzal illness, sticky eyes, sore throat, intermittent abdominal pain, reduced appetite and a reluctance to walk. His parents described an erythematous rash over his trunk, legs and arms. On examination, he was pyrexial, reluctant to weight bear, and had a mildly inflamed pharynx but no visible exudate on his tonsils. No other focus of infection was found.

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Classification of juvenile idiopathic arthritis

Characteristic	Clinical features
Age at onset	<16 years
Minimum duration	6 weeks
Subtypes	
Systemic	Arthritis Fever Rash
Oligoarthritis	1–4 joints affected during the first 6-months Persistent – affects no more than four joints throughout course Extended – affects more than four joints after first 6-months
Polyarthritis	Rheumatoid factor +ve – affects five or more joints in first 6 months Rheumatoid factor –ve – affects five or more joints in first 6 months
Enthesitis-related[a] arthritis	**Arthritis and enthesitis, or arthritis or enthesitis with at least two of the following:** Sacroiliac joint tenderness Inflammatory back pain HLA-B27+ Family history of HLA-B27+ related disease
Psoriatic arthritis	**Arthritis and psoriasis or arthritis and at least two of:** Dactylitis Nail changes Family history of psoriasis
Undifferentiated	Arthritis of unknown cause or not fulfilling above categories

[a] Enthesitis is the term for inflammation of the insertion of ligament, tendon, capsule or fascia to bone, particularly around the foot and knee.

Table 1

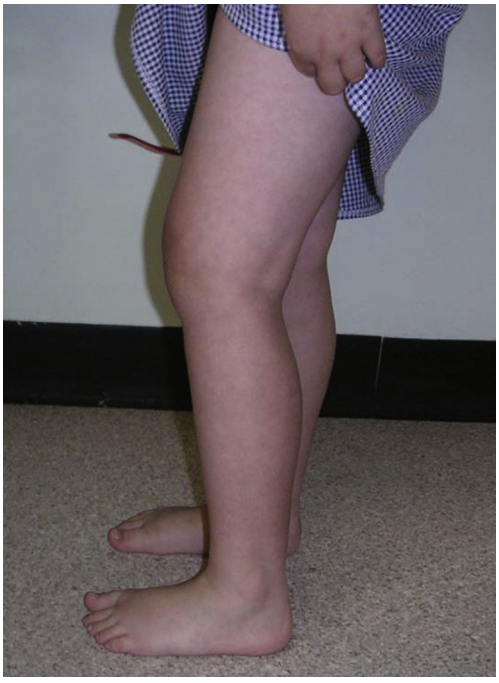

Figure 1 Flexion contracture left knee.

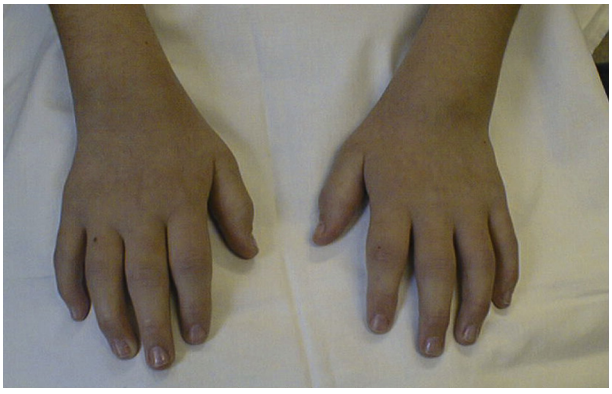

Figure 2 Polyarthritis affecting the small joints of the hands and wrists.

Initial blood work showed a white cell count (WCC) of 32 (10*9/L; normal range 5.5–15.5), neutrophil count of 29 (10*9/L; 1.5–8.5), platelet count of 884 (10*9/L; 150–450), haemoglobin of 95 g/L (115–155), ESR of 104 mm/hour (1–10), CRP of 178 mg/L (0–5) and ferritin of over 15,008 μg/L. His blood film was reported as showing reactive changes only. During his inpatient stay for further investigation, he was noted to have a rise in temperature at similar times each day; this was associated with a salmon-pink rash which improved as the temperature normalised (see Figure 3).

The limping child – thinking about Chloe and Connor's story

The differential diagnosis for the limping child – that is, a child presenting with an asymmetrical gait – is extensive, and comprises a spectrum of aetiologies from benign, self-limiting conditions through to life-threatening diseases such as septic arthritis and acute lymphoblastic leukaemia. Persistent limp and daytime symptoms are exclusion criteria for growing pains, a common label to describe children with aches and pains of unclear cause (see www.pmmonline.org for the 'rules of growing pains'). Differentiating the potential diagnosis of limp can be facilitated by knowledge of the common and significant causes according to age, as shown in Table 2. For the limping child presenting to primary care, indications for same day acute paediatric assessment include:

- The very young (under 3 years of age)
- The ill and febrile
- The non-weight bearing
- Children with painful, restricted joints
- The child who is immunosuppressed

Multiple joint swelling – Lily's story

In Lily's case of polyarthritis the likely diagnosis is JIA, although leukaemia should be considered. Other conditions to consider in a case of multiple joint swelling include: