Investigating the child with arthritis

Each month we present authoritative advice on the investigation of a common clinical condition, specially written for family doctors by the Board of Continuing Medical Education of the Royal Australasian College of Physicians.

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Before tackling the investigation of a child with arthritis one needs to be certain that the child does in fact have arthritis. Many children are referred to a rheumatologist with a 'positive arthritis test' (either antinuclear antibody or rheumatoid factor) performed on a questionable clinical background of nonspecific aches and pains without any objective signs of arthritis. Children and their parents are often caused unnecessary anxiety by these diagnoses of possible juvenile arthritis. The parents may have obtained a mound of literature on the disease from the internet, often from unorthodox sources, and may have tried a range of expensive unproven remedies.

This article describes the clinical approaches to some of the more frequently encountered conditions in children presenting with joint swelling. Arthralgia, nonspecific limb aches and soft tissue pains are not covered here.

Definition of arthritis

Arthritis is defined as objective swelling of a joint (as evidenced by a clinician able to examine joints) and the presence of two or more of the following:

- limitation in the range of movement
- tenderness or pain on motion
- increased heat in one or more joints.

Acute arthritis involving a single joint should be suspected as being septic arthritis until proven otherwise.

Juvenile idiopathic arthritis (juvenile rheumatoid arthritis, juvenile chronic arthritis or juvenile arthritis) by definition needs to have onset at an age less than 16 years and be persistent for at least six weeks (and preferably longer than three months) before the label is appropriate.

Evaluation of arthritis

Paediatric rheumatology relies largely on disease pattern recognition for the classification,

IN SUMMARY

- **Be sure the child has arthritis before investigating further.**
- **Disease pattern recognition is important in the classification of arthritis as imaging and laboratory findings may be relatively nonspecific.**
- **An acute swollen joint in a febrile child should be investigated for septic arthritis using joint aspiration and culture.**
- **In the physical examination of patients with arthritis, systemic features such as fever, rash or uveitis should be looked for in addition to the number and distribution of swollen joints.**
- **For chronic arthritis, the investigative tests performed depend on the presenting clinical picture. Appropriate tests include antinuclear antibody, anti-double stranded DNA antibodies, HLA–B27, rheumatoid factor, complement, full blood count, erythrocyte sedimentation rate, C-reactive protein, urinalysis and renal function tests.**

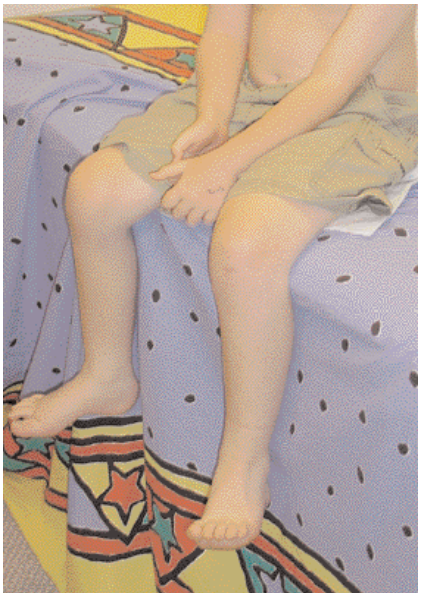


Figure 1. Reactive arthritis following *Salmonella* infection – the child presented with a swollen knee and ankle.

management and prognosis of arthritis as the laboratory findings may be relatively nonspecific.

Juvenile arthritis

Juvenile chronic arthritis has a prevalence of approximately 1 per 1000 and an incidence of about 1 in 10,000. This contrasts with the prevalence of soft tissue aches and pains (including benign nocturnal limb pain) in children of somewhere between 5 and 10%.

Transient synovitis

Transient synovitis of the hip, a disorder occurring most commonly in boys in the 3 to 10 years age group, is often preceded by a respiratory tract infection, pharyngitis or otitis media. An episode of transient synovitis may occur in up to 3% of children: 60 to 75% will have complete resolution of pain within two weeks. Although single episodes are more common, recurrences are reported in 5 to 15% of children.

Reactive arthritis

Reactive arthritis is arthritis occurring after an infectious disease where the joint fluid remains sterile.

Rheumatic fever

The prototype paediatric rheumatic condition of acute rheumatic fever, described by Thomas Sydenham in 1848, is now rare in the Australian Caucasian population although it is still prevalent among the Aboriginal population.

Poststreptococcal reactive arthritis

Poststreptococcal reactive arthritis (PSRA) is a condition following streptococcal pharyngitis or skin infection resulting in a reactive swelling of small and large joints. There is no universal agreement between clinicians on the need for long term antibiotic prophylaxis as carditis is an uncommon long term sequela in these children.

Other reactive arthritis

Infectious micro-organisms such as *Mycoplasma, Salmonella, Shigella, Yersinia* and *Campylobacter* can all cause reactive arthritis (Figure 1). Ross River fever virus and parvovirus are two of the more common viruses causing this type of arthritis.

Other arthritis

Other causes of arthritis and diseases associated with the condition include:

- sepsis in a joint (septic arthritis)
- Henoch–Schoenlein purpura (arthritis of the large joints)
- Kawasaki's disease
- systemic lupus erythematosus
- psoriasis.

Appropriate investigations

Clinically detectable arthritis – that is, objective swelling of a joint which also shows pain on movement and limited range of movement – may be divided into acute and chronic forms.

Acute arthritis

Septic arthritis

An acute swollen joint in a child who is febrile should be investigated using joint aspiration and culture. A plain x-ray of the region and blood tests (full blood count, blood film and erythrocyte sedimentation rate) may be part of the initial work-up. Nuclear scans are unlikely to be helpful in the management of

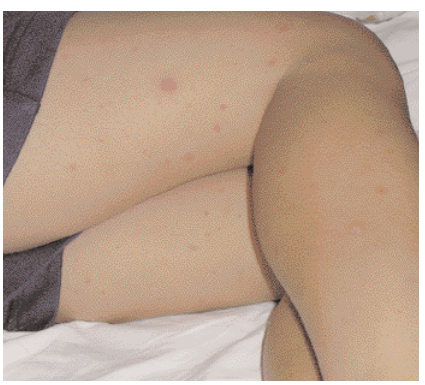
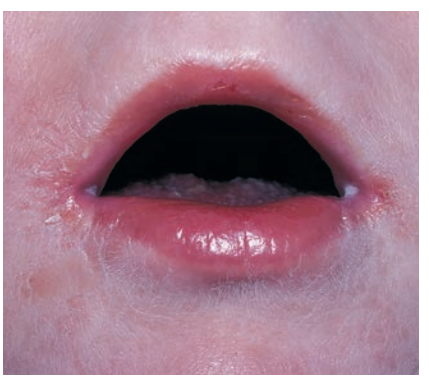
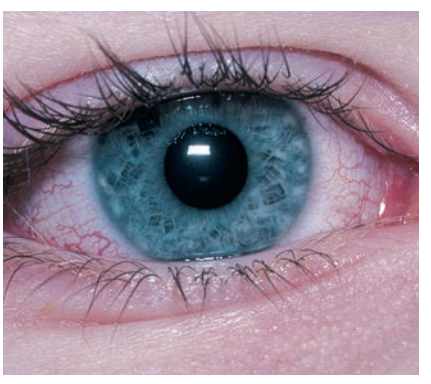


Figure 2. The typical rash of Henoch–Schoenlein purpura.





Figures 3a and b. Kawasaki's disease. a (left) Cracked red lips. b (right). Conjunctival hyperaemia, without exudate.

septic arthritis unless associated osteomyelitis is suspected because of local bony tenderness.

Henoch-Schoenlein purpura

Arthritis of the large joints (ankles, knees, wrists) associated with the typical palpable purpura and buttock/lower limb distribution of Henoch–Schoenlein purpura suggests this diagnosis (Figure 2).

Kawasaki's disease

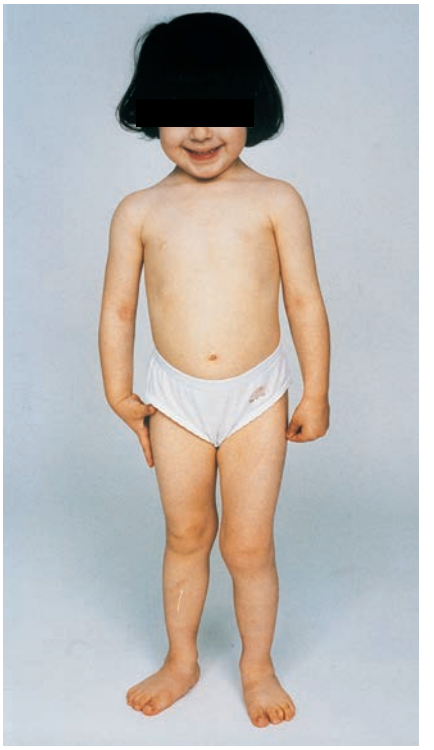
Kawasaki's disease should be considered if there is arthritis associated with the preceding fever and the lymphadenopathy, mucosal and cutaneous findings of this disease (Figures 3a and b).

Chronic arthritis and persisting effusion

Investigation of chronic arthritis is influenced by the presenting clinical picture.

Oligoarticular juvenile arthritis

Oligoarticular juvenile arthritis is the most common form of chronic arthritis in childhood. It typically affects young girls (aged 1 to 3 years at onset). They appear well except for having swollen and stiff knee, ankle or wrist joints (Figure 4). Stiffness is worse in the morning and there may be a limp. These patients are often antinuclear antibody-positive so antinuclear antibody is an essential investigation. Often full blood count and erythrocyte sedimentation rate are normal.



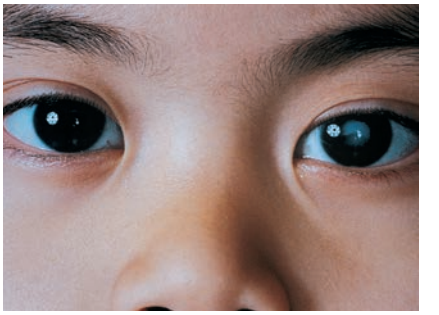


Figure 4 (left). Oligoarticular juvenile arthritis typically affects young girls aged 1 to 3 years at onset, who appear well except for having a swollen stiff joint and, sometimes, a limp.

Figure 5 (above). Asymptomatic occult smouldering iritis. This young girl has severely impaired vision with cataract formation. She presented with a swollen knee, which was only diagnosed as oligoarticular arthritis many months later, having been incorrectly attributed to trauma.

Asymptomatic chronic uveitis is a very serious feature of the disease and patients should be referred to a paediatric ophthalmologist to check for this initially and then on a three-monthly basis for five years (Figure 5).

Onset of oligoarticular juvenile arthritis at an older age often occurs in human leucocyte antigen (HLA) B27-positive adolescent boys, who present with swollen peripheral large joints or enthesitis (Figures 6 and 7). Occasionally these swollen joints require drainage for relief. The patient's family history should be checked for other HLA B27-related diseases, such as ankylosing spondylitis, acute inflammatory iritis and inflammatory bowel disease. This group has recently been classified as enthesitis-related arthritis (ERA).

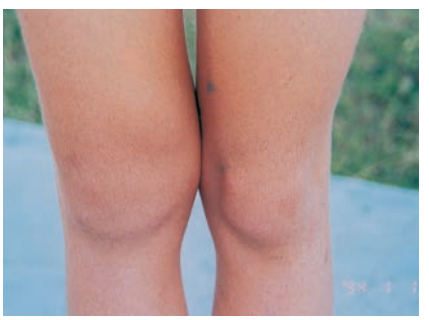


Figure 6. Oligoarticular juvenile arthritis (enthesitis-related arthritis) in an adolescent boy, showing the typical asymmetrical large joint swelling.

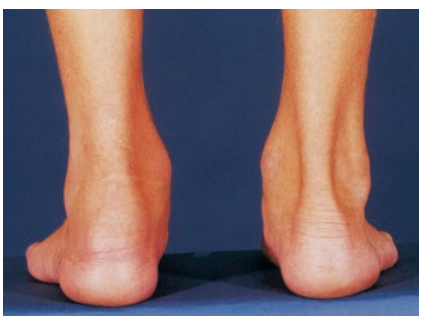


Figure 7. Enthesitis in an adolescent boy with oglioarticular juvenile arthritis (ERA), showing marked swelling at the insertion of the Achilles tendon.

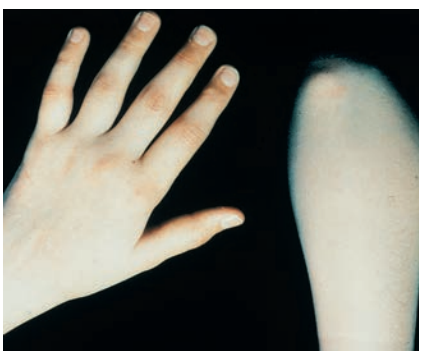


Figure 8. Symmetrical small joint involvement and a rheumatoid nodule in true juvenile rheumatoid arthritis.

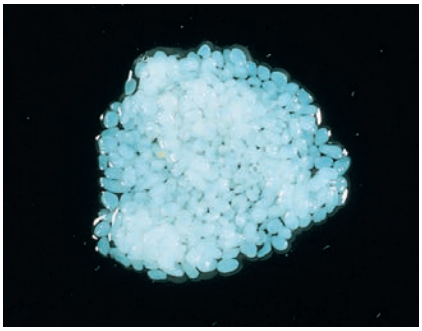


Figure 9. 'Rice bodies' obtained from aspiration of a knee joint affected by systemic onset juvenile chronic arthritis.

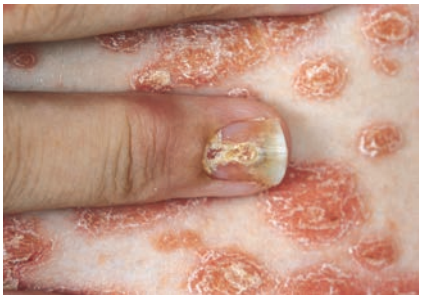


Figure 10. The pitting and typical psoriatic rash of psoriatic arthritis in a child who presented with a knee effusion and a swollen 'sausage' toe.

Polyarticular juvenile arthritis

Arthritis with symmetrical involvement of five or more small or large joints typically occurring in adolescent girls is characteristic of true juvenile rheumatoid arthritis – that is, adult type rheumatoid arthritis in a patient aged less than 16 years (Figure 8). Investigations include rheumatoid factor, antinuclear antibody, full blood count and erythrocyte sedimentation rate.

Systemic onset juvenile arthritis

Marked joint swelling associated with a persistent intermittent high fever, rash and serositis suggests systemic onset juvenile chronic arthritis (JCA), also known as Still's disease. Markedly raised inflammatory parameters support this diagnosis: these include erythrocyte sedimentation rate, C-reactive protein and platelets.

Aspiration of the joint (using a 12G cannula) will reveal very turbid inflammatory fluid with a high white cell count, or even 'rice bodies' from the active proliferative synovitis (Figure 9).

Psoriatic arthritis

Psoriatic arthritis typically presents as swollen joints with a psoriatic skin rash and pitting of the nails (Figure 10). A child with an asymmetrical inflammatory arthritis and a first degree relative with psoriasis may also suggest this diagnosis.

Systemic lupus erythematosus

If a patient presenting with symmetrical polyarticular arthritis also has a vasculitic rash and fever, weight loss, lethargy, oral ulceration, alopecia and an active urinary sediment, then systemic lupus erythematosus should be considered. Investigative tests include antinuclear antibody, anti-double stranded (ds) DNA antibodies, complement levels, full blood count, erythrocyte sedimentation rate, urinalysis and renal function tests. The arthritis in systemic lupus erythematosus responds readily to anti-inflammatory medication and often disappears when corticosteroids are used to treat the more severe renal or central nervous system manifestations of the disease.

Conclusion

Children with chronic arthritis have painful, swollen, stiff joints with reduced ranges of movement. They must be differentiated from the much larger group of children with transient limb pains who do not require expensive invasive investigation.

Knowledge of common diseases patterns helps characterise juvenile arthritis and allows for tailored therapy. MT