JAFA Juvenile Arthritis Foundation Australia

DIAGNOSING JUVENILE IDIOPATHIC ARTHRITIS (JIA)

A Case Study prepared for JAFA by Dr Roger Allen



It's 4pm on a Friday...

A couple have brought their 4-year-old daughter in to see you. They are worried because for the past few weeks she's appeared stiff in the mornings and less active in her normal play. Today they have noted a swollen left knee. They've looked up the internet and are particularly concerned after reading about **'juvenile idiopathic arthritis (JIA)'.** They ask, "but doctor, arthritis is surely just a disease in older people". As an experienced GP you are aware children can have arthritis, but you also see many children with various aches and pains – **so where do you start?**

1. Making a diagnosis of JIA

The diagnosis of JIA typifies the need for a thorough history and physical examination. Investigations are generally of secondary importance and can be confusing as no specific tests are diagnostic even if positive results are found e.g. antinuclear antibody and HLA B27 can be found in healthy children. Similarly, x-rays are not helpful in the early stages of disease. Ultrasound may confirm synovitis.

A strong suspicion of JIA in its early stages is indicated by:

- Pain and swelling of single or multiple joints
- Stiffness in the early morning typically improving with activity
- Reluctance to move the affected joint(s) through the normal range of movement
- Lack of confounding features on history that might otherwise explain the problem such as recent injury, intercurrent viral illness.

JIA is an umbrella term for a group of disorders. Current classification criteria list seven subtypes based on various criteria such as the number of affected joints, extra-articular features e.g. psoriasis or enthesitis (inflammation at tendon insertion such as the Achilles tendon attachment to the calcaneus), or the small group behaving as an adult rheumatoid patient with a +ve rheumatoid factor who are mainly teenage girls.

It is important to emphasize that confirming if JIA is the correct diagnosis may sometimes only become clearer over time. Don't be reluctant to ask for advice or to refer if uncertain whether a child's signs/symptoms could indicate JIA. Paediatric rheumatology teams are keen to assist.

2. Can JIA be confused with other significant conditions

Even if the working diagnosis is leaning toward JIA there are differential diagnoses to consider – look for the "red flags" that might redirect the evaluation. Some examples are:

An acute onset with an inflammatory appearance

- Painful single joint swelling with fever – consider septic arthritis
- Localised bone tenderness with fever – consider acute osteomyelitis
- Hip pain with recent viral URTI – consider transient synovitis of the hip i.e. "irritable hip"
- Polyarthritis with vasculitic rash (typically buttocks/lower limb) – consider Henoch-Schonlein purpura (HSP)

An acute onset with no systemic features

- Acute hip pain - consider slipped upper femoral epiphysis (SUFE)
- Trauma - if inconsistent findings to given history need to consider non-accidental injury

An insidious musculoskeletal onset with systemic features

- Nocturnal pain, pallor, weight loss – consider malignancy such as acute lymphoblastic leukaemia
- Abdominal pain, weight loss, mouth ulcers, tender nodular rash on legs – consider Crohn's disease

Recurrent musculoskeletal onset with no systemic features

- Activity related discomfort – consider hypermobility / Osgood-Schlatter disease (patellar tendon)
- Localised hip discomfort – consider Perthes' disease
- Nocturnal discomfort with otherwise normal function/examination – consider benign nocturnal limb pain i.e. "growing pains"

3. Successful management of JIA epitomizes the rheumatologic concepts of the "window of opportunity" for initiating therapy and following a "treat to target" approach

Studies have convincingly shown that there is a time limited "window" during which optimal therapeutic intervention produces better disease control, and importantly, an improved prognosis for children with JIA in the longer term. Advances in therapy have been dramatic over the past two decades with each JIA subtype warranting its own specific "targeted" approach. Without underplaying the significance, a diagnosis of JIA will bring for a child and family, the expectation for a child to be participating in normal age-appropriate activities is real and even anticipating the potential for achieving disease remission.

JIA is one of the more common chronic diseases of childhood affecting 1-4 per 1000 children under 16 years of age. Typical of many chronic disorders in children a team approach to care is required to achieve best practice. Paediatric rheumatology teams include a mix of paediatric rheumatologists, allied health (physiotherapy and occupational therapy), rheumatology nurses and other support staff such as psychologists, social workers. Due to the relatively common associated problem of uveitis, ophthalmology services are also relevant. **It is important that the GP also see themselves as part of the team.**

4. What role can/should the GP potentially expect to play as part of the team?

- General support for the family, including siblings, who may struggle particularly in the early period following diagnosis
- Encourage compliance with therapy
- Ensure the child is up to date with standard vaccinations but be aware, if child is on immunosuppressive therapy, some live vaccine protocols may be modified
- May assist with monitoring blood tests for children on some types of medication
- Ensure the family realise that the GP remains responsible for the normal day to day health requirements of the child/family and not to assume the specialist team becomes the "go to" alternative

Back to our Friday afternoon 4-year-old child….

In these times of health facilities being stretched and long waiting periods to be seen, the paediatric rheumatology teams know the importance of expediting assessment and initiating appropriate management for any child with JIA. If a GP considers their patient may have JIA, they are encouraged to call or write URGENT on a referral letter or use whatever method they find most expeditious in getting the child seen asap!