Chronic arthritis in children and young people

Amy Rowan
Ruth Wyllie
Helen Foster

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 verbalize 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 teams facilitates prompt treatment and prevention of complications. The emergence of novel and biologic agents, as well as earlier and more aggressive treatment, has helped to optimize clinical outcomes and dramatically changed the way that JIA has been managed over the last 10 years. The approach to management is multidisciplinary, and includes close liaison with other specialists and primary healthcare teams, as well as education and support for the family. Adolescence is a time of physical, psychological and emotional change, and the multidisciplinary team is fundamental to helping adolescents cope with the implications of a chronic disease, often complex treatment regimens, and transitional care into the adult world.

Keywords Juvenile arthritis; management; multidisciplinary team; transitional care

Juvenile idiopathic arthritis (JIA) is the most common cause of chronic arthritis in children (incidence 1 in 10,000 per year, prevalence 1 in 1000) and encompasses a heterogeneous group of diseases of unknown aetiology.[1,2] The term JIA replaces previous terminologies and is classified by predominantly clinical criteria (Table 1).[3]

Establishing a diagnosis of JIA

History

Making a diagnosis of JIA relies on clinical skills. Investigations help to exclude other pathology including malignancy and infection, although it must be remembered that no investigations are diagnostic. Careful clinical assessment can differentiate between inflammatory joint conditions such as JIA, mechanical causes of joint pains, and other conditions causing joint pain and swelling. The differential diagnosis for JIA is extensive (Table 2), with conditions ranging from the benign (e.g. hypermobility) to the life threatening (e.g. malignancy, such as leukaemia and solid tumours, infection, non-accidental injury).

The clinical assessment of a child is not the same as that of an adult,[4,5] and it is essential that the assessor is aware of the differences in normal ranges of movement in children compared to adults, as well as the normal changes in gait, development and motor developmental milestones in children. The history may be primarily from the parents or carers, and may initially be of vague complaints, such as 'my child is not right' or 'she no longer wants to do x'; consequently it is often difficult to localize the site of joint pathology from the history alone and assessment must include, as a minimum, a basic joint examination (such as pGALS – see below) in the context of other systems. The young child may not be able to verbalize pain, and a change in behaviour may be the presenting feature – such as being more irritable, clingy, or reluctant to play. Assessment of the child's daily activities is important; avoidance of activities previously enjoyed (e.g. in play or sport) is worrying, as is regression of achieved motor milestones (such as walking or handwriting); this may be observed by parents or others, such as teachers, and may be suggestive of inflammatory joint disease.

Other features in the history that suggest inflammatory joint disease include morning joint stiffness and pain; parents may notice that their child is 'slow to get going in the morning' or experiences stiffness after periods of rest, such as after long car rides – this is known as 'gelling'. They may also notice joints that appear swollen but this can be subtle and easily be overlooked. Mechanical joint pain typically worsens with physical activity, and swelling is uncommon and often transient. The presence of any 'red flags', such as weight loss, fever, night pain and bone tenderness, suggests infection or malignancy and warrants urgent assessment in secondary care.

Examination

The paediatric Gait Arms Legs and Spine (pGALS) musculoskeletal examination (Figure 1) is quick and easy to perform with simple manoeuvres often used in clinical practice, and has been validated in the school-aged child with excellent sensitivity and specificity (see The rheumatological examination in *MEDICINE* 2014; **42**(4): 197–201).[6] Free educational resources to demonstrate pGALS are available (http://www.arthritisresearchuk.org/health-professionals-and-students/video-resources/pgals.aspx). pGALS is acceptable in acute paediatric settings,[7] and is an effective way to assess all joints. It is useful in the context of vague presentations such as leg pains or limp. Abnormalities on pGALS examination can be followed with more detailed joint examination such as pREMS (paediatric Regional Examination of the Musculoskeletal System) which is based on the 'look, feel, move, function, measure' approach.[8] Joint swelling caused by effusion or synovitis is the most reliable physical sign of JIA, but may be subtle in very young children, and difficult to assess in joints such as the hip, shoulder and ankle, especially where changes are symmetrical.

Amy Rowan *MBBS MRes DRCOG is a GP Registrar, currently working in Paediatric Rheumatology at Great North Children's Hospital, Newcastle Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK. Competing interests: none declared.*

Ruth Wyllie *BSc RGN RSCN is Paediatric Rheumatology Lead Nurse at Great North Children's Hospital, Newcastle Hospitals NHS Trust, Newcastle upon Tyne, UK. Competing interests: none declared.*

Helen Foster *MD FRCP FRCPCH DCH Cert Med Ed is Professor of Paediatric Rheumatology at Newcastle University, UK and Honorary Consultant at Great North Children's Hospital, Newcastle Hospitals NHS Trust, Newcastle upon Tyne, UK. Competing interests: none declared.*

The classification of juvenile idiopathic arthritis, with clinical criteria

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

Differential diagnosis of joint pain in children

Life-threatening conditions
- Malignancy (leukaemia, lymphoma, neuroblastoma, bone tumour)
- Sepsis (septic arthritis, osteomyelitis)
- Non-accidental injury

Joint pain with no joint swelling
- Hypermobility syndromes (transient swelling is sometimes reported by patients)
- Complex regional pain syndromes (localized or widespread)
- Orthopaedic syndromes (e.g. slipped upper femoral epiphysis, Perthes' disease)
- Metabolic (e.g. hypothyroidism, lysosomal storage diseases)

Joint pain with joint swelling
- Juvenile idiopathic arthritis
- Trauma
- Infection
  - Septic arthritis and osteomyelitis (viral, bacterial [including Lyme disease], mycobacterial)
  - Reactive arthritis (post-enteric, sexually acquired)
  - Infection related (rheumatic fever, vaccination related)
- Inflammatory bowel disease (Crohn's disease or ulcerative colitis)
- Autoimmune rheumatic disease (systemic lupus erythematosus, scleroderma, dermatomyositis)
- Sarcoidosis
- Metabolic (e.g. osteomalacia [rickets], cystic fibrosis)
- Haematological (haemophilia, haemoglobinopathy)
- Tumour (benign/malignant)
- Developmental/congenital (e.g. spondylo-epiphyseal dysplasia)

Table 2

Investigation

Laboratory tests are seldom diagnostic but may help to exclude other diagnoses and be used by specialist teams to monitor disease activity and adverse effects of immunosuppressive drugs. However, blood tests and radiographs are initially often normal in JIA, which can provide false reassurance at the time of presentation. If there is clinical concern, then referral to a paediatric rheumatology team for specialist assessment should NOT be delayed.

Raised acute-phase reactants (erythrocyte sedimentation rate, serum C-reactive protein) and a high serum ferritin concentration may be present in active systemic-onset JIA; however, all these can be entirely normal in other subtypes of JIA, especially in oligoarthritis. Auto-antibodies are not diagnostic but often used to inform prognosis. In the presence of JIA, positive antinuclear antibodies (ANA) indicates a high risk of chronic anterior uveitis; this complication affects 30% of children with JIA, is invariably asymptomatic but potentially blinding, and can be detected only by a slit-lamp examination by an experienced ophthalmologist. However, a negative ANA does not exclude JIA, whereas a positive ANA can also occur in autoimmune rheumatic conditions (such as systemic lupus erythematosus), non-rheumatic conditions and healthy children. Rheumatoid factor (RF) is a poor diagnostic test, being present only in a minority of children with JIA; however, if positive in a child with polyarticular disease, it predicts a more aggressive disease course and worse prognosis. Synovial fluid examination to exclude sepsis is mandatory in the assessment of a child with a single hot, swollen joint. Plain radiographs in early JIA are often normal. Investigations such as ultrasound scanning, magnetic resonance imaging (MRI), computer-assisted tomography (CT) and bone scans may be arranged as part of specialist assessment. MRI is sensitive to early changes of inflammatory arthritis but availability may be limited and, in young children, invariably requires sedation or general anaesthetic. Ultrasonography is also sensitive to early changes, is well tolerated by young children, and is increasingly used to define normal changes in growing healthy children, as well as the extent and severity of arthritis.

Principles of management

The multidisciplinary approach

There is considerable potential for morbidity associated with JIA (Table 3), although complications are now seen less frequently,