Chronic arthritis in children and young people

Michael Hughes
Ruth Wyllie
Helen Foster

Abstract

Arthritis affects 1 in 1000 children and young people (CYP) and is 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 on clinical assessment. Investigations are helpful to exclude other conditions, but are often normal in JIA at presentation. The history can 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 15 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 Management; MRCP; multidisciplinary team; transitional care

Key points

- In the absence of trauma or sepsis, juvenile idiopathic arthritis (JIA) is the most likely diagnosis of a single swollen joint in a child
- Outcome is optimized by prompt referral to, and management by, an experienced paediatric rheumatology team — referral is recommended as soon as arthritis is suspected even if joint swelling is not confirmed on examination
- Careful history-taking and physical examination usually provide the diagnosis; laboratory tests and imaging are seldom diagnostic but help to rule out differential diagnoses. Skill in the paediatric Gait Arms Legs and Spine examination (pGALS) is useful to identify abnormal joints that may not be apparent from the history alone
- Prompt referral to an ophthalmologist and slit-lamp examinations to screen for asymptomatic uveitis are mandatory in all cases of suspected JIA
- There is increasing evidence to support earlier and more aggressive intervention with the potent immunosuppressive medicines that are now available

Introduction

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

Michael Hughes *RGN BSc Nursing (Child) is Paediatric Rheumatology Nurse Specialist at Great North Children's Hospital, Newcastle Hospitals NHS Trust, Newcastle upon Tyne, UK. Competing interests: none declared.*

Ruth Wyllie *RGN BSc Nursing (Child) 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 and Honorary Consultant at Great North Children's Hospital, Newcastle Hospitals NHS Trust, Newcastle upon Tyne, UK. Competing interests: none declared.*

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 of 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, and it is essential that assessors are aware of the differences in normal ranges of movement in children compared with adults, as well as the normal changes in gait, development and motor developmental milestones.

The history can be primarily from the parents or carers, and can 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 the presenting feature can be a change in behaviour, such as being more irritable, clingy or reluctant to play. Assessment of

The classification of JIA, 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-positive – affects five or more joints in first 6 months • Rheumatoid factor-negative – affects five or more joints in first 6 months
Enthesitis-related[a] arthritis	Arthritis and enthesitis, or arthritis with at least two of the following: • Sacroiliac joint tenderness • Inflammatory back pain • HLA-B27-positive • Family history of HLA-B27-positive 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 (patients sometimes report transient swelling)
- 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

- JIA
- 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, ulcerative colitis)
- Autoimmune rheumatic disease (systemic lupus erythematosus, scleroderma, dermatomyositis)
- Sarcoidosis
- Metabolic (e.g. osteomalacia (rickets), cystic fibrosis, mucopolysaccharidoses)
- Haematological (haemophilia, haemoglobinopathy)
- Tumour (benign/malignant)
- Developmental/congenital (e.g. spondylo-epiphyseal dysplasia)

Table 2

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 (e.g. walking, handwriting); this can be observed by parents or others, such as teachers or nursery workers, and can suggest 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'. Parents may also notice that joints appear swollen, but this can be subtle and easily overlooked. Mechanical joint pain, in contrast, typically worsens with physical activity, and swelling is uncommon and often transient. The presence of '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[2]. Free educational resources to demonstrate pGALS are available (see www.pmmonline.org/doctor/approach-to-clinical-assessment/examination), as well as a pGALS app, with language translations. pGALS is acceptable in acute paediatric settings, is an effective way to assess all joints and 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 the paediatric Regional Examination of the Musculoskeletal System (pREMS), which is based on the 'look, feel, move, function, measure' approach. Joint swelling caused by effusion or synovitis is the most reliable physical sign of JIA, but can be subtle in very young children, and difficult to assess in joints such as the hip, shoulder and ankle, especially if changes are symmetrical.

Investigation

Laboratory tests are not diagnostic in JIA, but help to exclude other diagnoses and are used by specialist teams to monitor disease activity and the adverse effects of immunosuppressive drugs. Blood tests and radiographs are initially often normal in JIA and can provide false reassurance at the time of presentation. If there is clinical concern of suspected rheumatic disease,