# key points

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**Juvenile idiopathic arthritis (JIA) affects approximately** 1-4 per 1,000 children under the age of 16. Girls are more commonly affected than boys. It is a heterogeneous condition, even within JIA categories, ranging from insidious arthritis affecting one to two joints to florid and life-threatening systemic arthritis.

**Oligoarticular disease predominantly affects preschool** children who are generally otherwise well and remain active despite their arthritis. It typically presents with gradual onset arthritis of the large joints. Polyarticular disease may present with systemic symptoms, such as lethargy, malaise and even low-grade fever, and tends to affect multiple large and small joints symmetrically. Systemic onset JIA on the other hand can cause prolonged fever accompanied by a macular rash. Arthritis may be a relatively late clinical finding. Evidence of immune activation may be seen. A small percentage of patients with systemic onset JIA will experience macrophage activation syndrome. This severe, uncontrolled immune response is a medical emergency with significant mortality rates.

**Uveitis affects up to 20% of patients with JIA. It is** usually asymptomatic in the early stages and may precede the onset of arthritis. It can be sight threatening if not detected and treated. Some clinical signs such as cataracts, keratic precipitates, and irregular pupils secondary to synechiae can be observed by direct ophthalmoscopy. Patients should have an ophthalmic assessment within six weeks of initial diagnosis or urgently if any eye signs or symptoms are present.

**All patients with suspected JIA should be referred** urgently to a specialist paediatric rheumatology team. Coordinated, multidisciplinary care within a specialist team is required to achieve optimal outcomes. Most patients with JIA will need rheumatology follow-up into adulthood and transition to adult services usually occurs between 16 and 18 years of age. Planned and coordinated transition is vital for ensuring continuity of care.

**Early, aggressive therapy increases the likelihood of a** response to first-line treatments and of achieving drug-free remission later in life. NSAIDs are used first line for symptomatic treatment. Systemic steroids are used to induce remission and maintain disease control in some patients. Systemic disease modifying medications (e.g. methotrexate, leflunomide, sulfasalazine and cyclosporine) will be required in some cases. Biologic medications are generally reserved as second-line therapy should conventional DMARDs fail. Patients on systemic immunosuppressive therapy are at risk of severe infection. Symptoms may be masked by treatments. Any sign of infection should prompt urgent medical review and early initiation of treatment if needed.