JIA: A Medical Description

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Juvenile Idiopathic Arthritis (JIA) is a chronic disorder of the body joints affecting about one in every 1000 children and adolescents under 16 years of age.

Causes and symptoms

There are several causes of joint inflammation, including an injury or infection of the joint itself (septic arthritis) or, more frequently, infection (e.g., streptococcal throat infection or gastroenteritis) somewhere else in the body (reactive arthritis). These trigger the release of cytokines (mediators of inflammation) such as TNF within the joint; these cytokines induce and perpetuate the inflammation. There is an increased blood flow though the lining of the joint (synovium), which becomes swollen and produces excess joint fluid. Reactive arthritis usually resolves within a month from onset.

In JIA, no trigger is identified, but the inflammation persists beyond six weeks. The child complains of pain in the affected joint(s), which become stiff on waking up in the morning and after periods of inactivity. The affected joints are swollen and warm, and fluid may be elicited in the larger joints (e.g., the knee).

The arthritis may affect a few joints (oligoarthritis) or many joints (polyarthritis). It may spread to other joints (extended oligoarthritis).

JIA is different from adult rheumatoid arthritis. Only about 5% of JIA cases test positive for rheumatoid factor (or anti CCP, a newer marker of RA in adults) in the blood and have a disease course similar to that in adult rheumatoid arthritis.

Arthritis may be the presenting manifestation in psoriasis and in enthesitis-related arthritis (ERA), which is the paediatric equivalent of ankylosing spondylitis in adults.

Systemic-onset JIA can be difficult to diagnose initially as it presents with fever and rash resembling a viral infection. The arthritis, usually involving small and large joints, may present weeks or months later.

Arthritis may also seen in children with Down's syndrome, inflammatory bowel disease, and a variety of other conditions.

Diagnosis and treatments

The diagnosis of JIA is made after a detailed history and examination. There is no single diagnostic test. Some investigations, such as blood picture and x-rays, are done mainly to exclude other causes of arthritis. Other investigations, such as the inflammatory markers ESR and CRP, may be useful in monitoring the disease course.

The autoantibodies ANA (anti-nuclear antibody) and RF (rheumatoid factor; often now replaced by a test called anti-CCP)) may be helpful in determining which subtype of JIA a

child has and in giving a prognosis on the severity and duration of JIA. Patients with ERA are likely to have an HLA B27 positive tissue type. Ultrasound and MRI scans can sometimes give useful information about inflammation in the joints.

Inflammation of the internal lining of the eye (uveitis) is a potentially serious complication of JIA. It is more common in female patients, especially those who are ANA positive, and in patients with oligoarthritis. Regular eye screening is essential to detect its presence at an early stage, as patients remain symptom-free until the eye suffers significant and sometimes irreversible damage.

The treatment of JIA aims at the suppression of inflammation in the joints. Treatments include intra-articular steroid injections and the suppression of the underlying autoimmune process by the administration of medicines such as methotrexate.

There are a number of newer 'biologic' drugs, which have been designed to block a specific part of the immune system implicated in causing inflammation or arthritis.

Drugs	How they work	Uses
Etanercept, Infliximab, Adalimumab, Golimumab	Block TNF, the main cytokine inducing synovial inflammation	Most types of JIA (not usually helpful in cases of systemic JIA)
Tocilizumab	Blocks IL-6	Systemic and polyarticular JIA
Anakinra	Blocks IL-1	Systemic JIA and MAS
Rituximab	Depletes B-cells, which make antibodies	Most likely to be useful if blood tests for RF or CCP are positive
Abatacept	Affects T-cells in the immune system	Polyarticular JIA if anti-TNF drugs are not suitable or effective
Secukinumab	Blocks IL-17	Adult patients with psoriatic arthritis or ankylosing spondylitis; currently in tests to assess its efficacy in treating psoriatic JIA and ERA

Physiotherapy and hydrotherapy can help some children with JIA by building up the muscles and keeping the affected joint(s) supple. Occupational therapists give useful advice on how to keep functioning as normally as possible at home, at school, and in other activities. If grip strength has been affected by JIA, a hand-strengthening programme can be very useful.

Although a cure is not available, modern advances in therapy allow good disease control and prevent long-term joint and eye damage in most patients.