Systemic Juvenile Idiopathic Arthritis Diagnosis and Management

Systemic Juvenile Idiopathic Arthritis (sJIA) is a joint disease that usually presents in childhood. Patients present with increased ferritin as well as other markers of inflammation. Management may include glucocorticoids, tocilizumab, or anakinra. Complications include joint deformities, limb-length discrepancies, uveitis, and macrophage activation syndrome. This disease has a highly variable prognosis.



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Laboratory Findings

Increased Ferritin
Up-arrow Ferret-tin
Ferritin is a marker of inflammation, and can be quite elevated in sJIA. Other signs of inflammation include elevated ESR and CRP. Left-shift, leukocytosis, thrombocytosis and anemia are also often present.

Management

Glucocorticoids
Quarter-on-steroids
Glucocorticoids are indicated for severe or drug resistant cases of sJIA. This includes sJIA with perimyocarditis, polyarticular SJIA, and "shock therapy" in seriously ill children at the beginning of long-term drug therapy.

Tocilizumab
Top-hat-lizard-mob
Tocilizumab is a monoclonal antibody against the interleukin-6 (IL-6) receptor. Interleukin-6 is a cytokine that regulates inflammatory reactions in the human body and is increased in inflammatory arthritides.

Anakinra
Anakin-Skywalker
Interleukin-1 (IL-1) is a pro-inflammatory cytokine that is increased in patients with SJIA. Anakinra is a novel drug that acts as an IL-1 receptor antagonist, preventing it from causing further inflammation.

Complications

Joint Deformities
Deformed Joints of Hand
sJIA leads to joint-specific movement restrictions and compensatory maladaptive postures. They must be recognized and treated at an early stage in order to avoid contractures and permanent joint deformities.

Limb-Length Discrepancy
Unequal Limbs
sJIA can cause stunted growth and lead to short stature, especially in severe systemic courses. Growth is locally stimulated or inhibited by chronic arthritis in a joint-specific manner which leads to limb length discrepancies.

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Uveitis

UV-glasses

Anterior uveitis can be associated with an outwardly inconspicuous eye, especially if it is chronic, and can therefore only be reliably diagnosed during slit lamp examination. Due to the risk of blindness, ophthalmological screening and treatment of these complications are of particular importance.

Macrophage Activation Syndrome

Activated Macrophage in Rage

Another complication is macrophage activation syndrome (MAS), characterized by sudden onset of high fever, enlarged liver and spleen, clouding of consciousness, and rashes.

Considerations

Variable Prognosis

Grave and Doctor with Good Prognosis

sJIA has a highly variable prognosis. Some cases remit early whereas some develop into chronic arthritis.

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