

Juvenile Still's Disease

Still's disease is a form of arthritis occurring most typically in children. In rare cases it can occur in adults, but it is then referred to as adult-onset Still's disease.

This disease is also known as systemic-onset JRA. This means that this is a disease involving the joints, but it typically appears first with body-wide (systemic) symptoms and disease, such as fevers, gland and internal organ involvement.

It is characterized by high spiking fevers, a transient salmon-colored rash, and/or joint involvement.

Symptoms

Systemic (body wide) symptoms usually appear first - extreme fatigue accompanied by high daily fevers (rising to 102 degrees F/39 degrees C or higher, then returning rapidly to normal or below normal temperature.) These fevers often appear at the same time each day.

A faint salmon colored rash typically comes and goes without itching.

Other common symptoms include nausea, loss of appetite and weight loss. Swelling of the lymph glands, enlargement of the spleen/liver and sore throats are also common.

Some, but not all, patients will develop inflammation around the heart and lungs. Occasionally, patients will develop accumulated fluid around the heart and lungs.

All patients with Still's eventually develop joint pain and swelling – usually more than five joints (polyarticular arthritis.) This arthritis often occurs after the rash and fevers have been present some time however, because of the impressive nature of the systemic symptoms, the joint involvement is sometimes overlooked initially. The pain and swelling can occur in any joint, though there are some more likely to be affected than others in Still's disease. Arthritis in the wrists is common, as is the eventual fusion of these joints.

The fevers, rash and other systemic symptoms of the disease usually disappear within several months, however the arthritis often stays on afterwards. The arthritis can become a long term problem, persisting into adulthood.

Diagnosis

A diagnosis of Still's disease is based on the presence of the above mentioned symptoms. The presence of persistent joint involvement (lasting more than six weeks) is needed to conclusively diagnose Still's disease, and other diseases (in particular infections, cancers and other types of arthritis) must be ruled out first.

High white blood cell counts (without the presence of serious infection) and low red blood cell counts are common for Still's disease patients.

Blood tests for inflammation (sedimentation rate and C-reactive protein test) are also often elevated, however rheumatoid factor tests (classic test for rheumatoid arthritis) and antinuclear antibodies tests (indicator for systemic lupus erythematosus) are most usually negative.

Causes

As with many other types of arthritis, the causes of Still's disease are largely unknown. It is believed to be an autoimmune disorder. One school of thought believes it to be brought on by an infection with a microbe.

Frequency

10-20% of all cases of JRA are Still's disease. About 25,000-50,000 children in the US are affected by it.

Frequency of individual symptoms

High intermittent fevers – 100% of patients

Joint inflammation and pain – 100% of patients

Muscle pain with fevers – 100% of patients

Faint salmon-coloured skin rash – 95% of patients

Swelling of lymph glands or enlargement of spleen and liver – 85% of patients

Marked increase in white blood cell counts – 85% of patients

Inflammation of the lungs (pleuritis) or around the heart (pericarditis) – 60% of patients

Severe Anemia (low red blood cell counts) – 40% of patients

Abdominal pain – 20% of patients

Age of onset

Generally this disease appears before age 16. In adult-onset Still's disease the average age of onset is between 20 and 35 years of age.

Treatment

Treatment for Still's disease depends on the specific areas of inflammation. Some symptoms can be controlled with nonsteroidal anti-inflammatory drugs (NSAIDs,) but others may require cortisone (steroid) medications.

In cases where the illness is persistent, immunosuppressant medications (methotrexate, hydroxychloroquine etc.) such as those used for patients with other forms of rheumatoid arthritis may be required. More recently, TNF-inhibitors have been used to successfully decrease signs and symptoms of Still's disease. In some cases, a combination of these treatments may be necessary.

In Summary

Patients always present with high spiking fevers.

Often a non-itchy rash is present

Joint inflammation always develops with Still's disease, though usually does not appear initially.

The causes of Still's disease are not known.

Inflammation of internal organs can occur

Diagnosis is dependent on the presentation of the common clinical features, and the appearance of joint inflammation and pain

Blood tests for other rheumatic diseases are negative

Treatment is aimed at reducing inflammation in affected areas.