CONDITION

Adult still disease

Learn about adult Still disease, including its fever, rash, and joint symptoms, how doctors diagnose it, available treatments, and signs that need urgent care.

WHAT IT IS
Rare whole-body autoinflammatory disease

BODY SYSTEM
Immune system

MAIN SYMPTOMS
High fever, salmon-pink rash, joint pain, sore throat

WHAT CAUSES IT
Unknown, linked to immune overactivity

HOW IT IS DIAGNOSED
Clinical assessment and exclusion of similar conditions

MAIN TREATMENT
Anti-inflammatory and immune-suppressing medicines

CAN IT BE CURED?
No, disease can relapse after improvement

IS IT AN EMERGENCY?
Sometimes, with severe trouble breathing

LONG-TERM OUTLOOK
Variable, from one episode to relapses or damaging arthritis

Overview

Adult Still disease is a rare inflammatory condition that can affect the whole body. It is also called adult-onset Still disease, or AOSD. It is related to systemic juvenile idiopathic arthritis, a similar condition that begins during childhood.

Adult Still disease is considered an autoinflammatory disease . This means part of the immune system becomes overactive and causes inflammation without an infection or another clear trigger. The inflammation can lead to high fevers, a temporary skin rash, joint pain, and other symptoms throughout the body.

The disease varies greatly from person to person. Some people have one episode that settles with treatment. Others have repeated flares separated by periods of improvement. Some develop ongoing arthritis, which can damage joints if it is not controlled.

Adult Still disease matters because its symptoms can resemble infections, cancers, and other immune-related illnesses. There is no single test that confirms it. Diagnosis therefore requires a careful medical evaluation. Early control of inflammation can relieve symptoms, limit steroid exposure, protect joints, and reduce the risk of serious complications.

Symptoms

Symptoms may appear suddenly or develop over several weeks. They can change from day to day. A typical flare may include:

The rash often affects the trunk, arms, or legs. It may be difficult to see on darker skin and can look different from the classic salmon-pink description. It is usually not very itchy, although itchy or longer-lasting rashes can occur.

Joint symptoms may affect the wrists, knees, ankles, elbows, shoulders, fingers, or other joints. In some people, arthritis becomes the main long-term problem. Morning stiffness may make movement difficult after waking or resting.

Inflammation can also affect internal organs. This may cause abdominal discomfort from an enlarged liver or spleen. Inflammation around the lungs, called pleuritis , can cause pain with deep breathing. Inflammation around the heart, called pericarditis , may cause chest pain that changes with position or breathing.

These symptoms are not unique to adult Still disease. Fever, rash, swollen glands, and joint pain have many possible causes, so they should be assessed rather than used for self-diagnosis.

- A high fever, often reaching its highest point in the late afternoon or evening
- A flat or slightly raised salmon-pink rash that may appear during a fever and fade as the temperature falls
- Joint pain, swelling, warmth, or stiffness
- Muscle pain, especially during fever
- A severe or persistent sore throat
- Fatigue and a general feeling of illness
- Swollen lymph nodes
- Reduced appetite or unintentional weight loss

Causes and risk factors

The exact cause is not known. Adult Still disease is not caused by one identified infection, and it is not contagious.

Researchers believe the condition develops when the innate immune system-the body's rapid, first-line defense system-becomes overactive. Inflammatory messenger proteins, including interleukin-1, interleukin-6, and interleukin-18, appear to play important roles. These proteins help coordinate inflammation but can cause widespread symptoms when their activity is excessive.

A genetic tendency may contribute, but adult Still disease is not usually inherited in a simple or predictable pattern. Having an affected relative does not mean that another family member will develop the condition.

Some people report an infection or other stressful event before symptoms begin. Such events may act as triggers in a person who is already susceptible, but no single trigger has been established. There is no proven lifestyle choice that causes adult Still disease.

The condition can begin at different ages in adulthood. Age, symptoms, or a possible trigger cannot confirm who has it.

Diagnosis

There is no single blood test, scan, or biopsy that proves a person has adult Still disease. A rheumatologist, who specializes in inflammatory and autoimmune conditions, often coordinates the evaluation.

Diagnosis begins with a detailed history and physical examination. The clinician may ask about the timing of fever, whether the rash appears with fever, which joints are affected, recent travel, infections, medicines, and other health conditions. Photographs of a short-lived rash and a written temperature record may be useful.

Blood tests may show:

A very high ferritin level can support the diagnosis, but it is not specific to adult Still disease. Ferritin may also rise with infection, liver disease, cancer, and other inflammatory disorders. Tests for rheumatoid factor and antinuclear antibodies are often negative, but these results do not confirm or exclude the condition.

Depending on the symptoms, evaluation may include blood cultures, viral tests, chest imaging, ultrasound, CT or MRI scans, an echocardiogram, or tests of joint fluid. A skin, lymph node, bone marrow, or liver biopsy is sometimes needed when doctors must investigate another possible illness.

Doctors may refer to classification systems such as the Yamaguchi or Fautrel criteria. These combine several clinical and laboratory features. They can support a structured assessment but do not replace medical judgment.

An important part of diagnosis is excluding conditions that can look similar. These include serious infections, lymphoma and other cancers, medication reactions, lupus, rheumatoid arthritis, vasculitis, and other inflammatory diseases. This process may take time, especially early in the illness.

- High levels of C-reactive protein or an elevated erythrocyte sedimentation rate, both signs of inflammation
- An increased white blood cell count, often with a high proportion of neutrophils
- An increased ferritin level; ferritin stores iron but can rise sharply during inflammation
- Abnormal liver enzyme results
- Anemia or other changes in blood cell counts

Treatment and management

Treatment aims to control inflammation, ease symptoms, preserve joint function, and prevent complications. The plan depends on whether the disease is mainly systemic, mainly affects the joints, or is causing organ complications. Medicine availability and regulatory approval differ by country.

Nonsteroidal anti-inflammatory drugs like ibuprofen or naproxen , or NSAIDs, may help pain and fever in selected cases. They are often not enough to control significant disease by themselves. NSAIDs can affect the stomach, kidneys, blood pressure, and cardiovascular system, so they should be used with medical guidance.

Corticosteroids such as prednisone can suppress inflammation quickly. They may be used when symptoms are substantial or organs are affected. Long-term steroid exposure can cause infection risk, bone loss, high blood pressure, diabetes, cataracts, and other problems. Clinicians generally try to reduce steroids like prednisone or hydrocortisone once the disease is controlled rather than stopping them suddenly.

Disease-modifying antirheumatic drugs, or DMARDs , may be used when arthritis continues or when a steroid-sparing treatment is needed. Methotrexate is one example. These medicines act more slowly and require monitoring for possible effects on blood cells, the liver, and other organs.

Biologic medicines target specific inflammatory signals. Interleukin-1 inhibitors, such as

anakinra or canakinumab, and the interleukin-6 inhibitor tocilizumab are important options for Still disease. Current specialist recommendations support considering targeted treatment early, particularly when disease is active or steroids cannot be reduced. The choice depends on disease features, previous treatment, other health conditions, pregnancy plans, access, and local approval.

Before immune-suppressing treatment, clinicians may check for infections such as tuberculosis or hepatitis. Vaccination needs should also be reviewed. Live vaccines may not be appropriate during some treatments, so vaccination decisions should be discussed with the treating team.

Regular movement can help preserve strength and joint flexibility. During an active flare, activity may need to be adjusted rather than stopped completely. A physical or occupational therapist can suggest joint-friendly exercise, pacing strategies, or tools that make daily tasks easier.

Adequate sleep, balanced nutrition, smoking avoidance, and emotional support can improve overall health, although they do not replace anti-inflammatory treatment. Keeping a record of fevers, rashes, joint symptoms, and medicine effects may help identify patterns.

Complications

Uncontrolled inflammation can lead to persistent arthritis and joint damage. The wrists and other joints may lose movement or function over time. Prompt treatment and rehabilitation can help reduce this risk.

Inflammation may affect the liver, lungs, or heart. Possible complications include hepatitis, fluid around the lungs, pericarditis, inflammation of the heart muscle, or lung inflammation. Medicine side effects can sometimes contribute to liver or blood-test abnormalities, so these results need medical interpretation.

A particularly serious complication is macrophage activation syndrome , or MAS. It is a severe, rapidly progressing state of immune overactivity related to hemophagocytic lymphohistiocytosis. It can cause persistent fever, falling blood cell counts, liver problems, abnormal bleeding, confusion, and failure of multiple organs. Ferritin often rises markedly, but diagnosis relies on the full clinical and laboratory picture.

Seek emergency care for severe trouble breathing, fainting, new confusion, a seizure, uncontrolled bleeding, blue or gray lips, or intense chest pain. Urgent same-day assessment is also appropriate for rapidly worsening illness, unusual bruising, yellow skin or eyes, persistent vomiting, or fever accompanied by marked weakness. These symptoms do not always mean

MAS or another severe complication, but they should not wait for routine follow-up.

Rarely, long-lasting inflammation can lead to amyloidosis, in which an abnormal protein builds up in organs such as the kidneys. Better control of inflammation appears to have made this less common.

Follow up

Routine follow-up is important even when symptoms improve. Adult Still disease can relapse, and treatment may need to be adjusted gradually.

Appointments may include a review of fever patterns, rash, joint swelling, morning stiffness, energy, daily function, and possible medicine side effects. Blood tests may monitor inflammation, blood cell counts, liver function, kidney function, and ferritin. The tests needed and their timing depend on the disease and treatment.

People taking corticosteroids may also need attention to bone health, blood pressure, blood sugar, eye health, and infection prevention. Those receiving biologic medicines or other immune-suppressing treatments should report possible infections promptly and discuss vaccines before receiving them.

Contact the treating clinician between routine visits if fever or rash returns, joint symptoms steadily worsen, new swelling develops, or treatment causes concerning effects. Do not stop corticosteroids suddenly unless a clinician directs you to do so.

Pregnancy planning should be discussed in advance because active inflammation and some medicines can affect pregnancy decisions. A rheumatologist and obstetric clinician can review treatment options before conception and during pregnancy.

Sources

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