

## Adult Onset Still Diseases

S.K. Mohanasundari

### Abstract

Adult-onset Still's disease is an inflammatory disease that may affect many joints, internal organs, and other parts of the body. Adult Still's develops most often in people before age 45, but can first occur in later years as well. The cause of Still's is unknown and there are no known risk factors. It is thought that a virus or other type of infectious agent may trigger Still's disease, but there is no proof. Although some features are similar, adult-onset Still's disease is different than Still's in children. In children, Still's disease is considered a form of juvenile rheumatoid arthritis and referred to as systemic-onset juvenile rheumatoid arthritis. Less than one in 100,000 people develop adult-onset Still's each year and it is more common in women. It is manifested by fever, joint pain, warmth, and swelling, joint pain, warmth, and swelling, severe muscle ache, and sore throat. It can be diagnosed with blood test, imaging study. The treatment is Symptomatic as this disease as no cure as such.

**Keywords:** Adult; Arthritis; Inflammation; Rash; Infection & Swelling.

### Introduction

Adult Still's disease is a rare type of inflammatory arthritis that is similar to rheumatoid arthritis. It shares characteristics of systemic-onset juvenile idiopathic arthritis, but it begins in adulthood. Inflammation may affect a few joints at first, but may advance to include more joints over time. Some people may have only one bout of the illness followed by lasting remission, while others may develop chronic arthritis.

The cause of adult Still's disease is unknown. Some research suggests that it may be triggered by an infection.

**Author's Affiliations:** Nursing Tutor, College of Nursing, AIIMS, Jodhpur, 342005, Rajasthan.

**Corresponding Author: S.K. Mohanasundari,** Nursing Tutor, College of Nursing, AIIMS Jodhpur, AIIMS residential complex 201/38. Type-II quarters, Basniphase-II. Jodhpur, 342005. Rajasthan

E-mail: [roshinikrishitha@gmail.com](mailto:roshinikrishitha@gmail.com)

**Received on 19.09.2016, Accepted on 12.10.2016**

### Definition

*Adult-onset Still's Disease (AOSD)* is a rare systemic inflammatory disease characterized by the classic triad of persistent high spiking fevers, joint pain and a distinctive salmon colored bumpy rash. This inflammation can destroy affected joints, particularly the wrists.

### Incidence

- Fewer than 1 out of 100,000 people develop adult-onset Still's disease each year.
- Incidence peaking twice: once from 15 to 25 years and again from 36 to 46 years
- Most often in people before age 45, but can first occur in later years as well
- It affects women more often than men.

### Risk Factor

- Age is the main risk factor for adult Still's disease, No other risk factor has been identified.

*Causes*

The cause of adult Still’s disease is unknown

*Symptoms*

Almost all people with adult-onset Still’s disease have fevers, joint pain, sore throat, and a rash. But

the type, pattern, and severity of symptoms vary from person to person and even from month to month for the same person. For example, symptoms may come and go. And, at first patient may have just a few symptoms, then later patient may have more.

If you have a high fever, rash and achy joints,

- 
- |   |  |
|---|--|
| <ul style="list-style-type: none"> <li>• <b>A fever</b> (equal to or greater than 102 degrees) that comes on quickly once per day, usually in the afternoon or evening. For most people, these fevers resolve without treatment.</li> <li>• <b>Joint pain, warmth, and swelling</b> affecting a few joints at first -- often knees and wrists, then several joints. Morning stiffness often lasts for several hours.</li> <li>• <b>A salmon pink-colored skin rash</b> that usually comes and goes with the fever and usually doesn't itch. Flat spots or both flat spots and small, raised bumps may appear on your torso, upper arms or legs, or face.</li> <li>• <b>Severe muscle aches</b>, which also may ebb with the fever.</li> <li>• <b>A sore throat</b> that can be severe, constant, and burning</li> </ul> | <ul style="list-style-type: none"> <li>• Abdominal pain and swelling</li> <li>• Pain when taking a deep breath</li> <li>• Swollen glands (lymph nodes)</li> <li>• Unexplained weight loss</li> </ul> |
|---|--|
- 

see your doctor. Also, if you have adult Still’s disease and develop a cough, difficulty breathing, chest pain or any other unusual symptoms, contact your doctor.

*Investigations and Diagnostic Measures:*

Adult-onset Still’s disease can only be diagnosed after many other diseases (such as infections and cancer) are ruled out. You may need many medical

tests before a final diagnosis is made.

A physical exam may show a fever, rash, and arthritis. The health care provider will use a stethoscope to listen for changes in the sound of your heart or lungs.

The following tests can be helpful in diagnosing adult Still’s disease:

Blood test	Imaging studies
<ul style="list-style-type: none"> <li>• Complete blood count (CBC), may show a high number of white blood cells and reduced number of red blood cells.</li> <li>• C-reactive protein (CRP), a measure of inflammation, will be higher than normal.</li> <li>• ESR (sedimentation rate), a measure of inflammation, will be higher than normal.</li> <li>• Ferritin level will be very high.</li> <li>• Fibrinogen level will be high.</li> <li>• Liver function tests will show high levels of AST and ALT.</li> <li>• Rheumatoid factor and ANA test will be negative.</li> </ul>	<ul style="list-style-type: none"> <li>• Other tests may be needed to check for inflammation of the joints, chest, liver, and spleen:</li> <li>• Abdominal ultrasound</li> <li>• CT scan of the abdomen</li> <li>• X-rays of the joints, chest, or stomach area (abdomen)</li> </ul>

*Treatment and Selfcare*

*Goal of the Treatment:*

To control symptoms and the course of the disease.

To help prevent or lessen any complications.

*Treatment*

Early treatment for adult-onset Still’s disease is aimed at controlling symptoms of arthritis with nonsteroidal anti-inflammatory drugs (NSAIDs). NSAIDs include ibuprofen (Advil, Motrin), naproxen(Naprosyn, Aleve), and high-dose aspirin. Analgesics, or pain drugs, may also be prescribed. if disease is severe or doesn’t respond to NSAIDs, Patient may require corticosteroids, such

as prednisone,

In case of chronic case of Still’s, patient may need medications to suppress immune system and to control arthritis and other symptoms. Patient may take more than one medication at the same time and may need to take these for a long time. Few of these medications can be taken by mouth and others by injection.

These are examples of medications:

- Methotrexate (Rheumatrex)
- Hydroxychloroquine (Plaquenil)
- Sulfasalazine (Azulfidine)
- Azathioprine (Imuran)

- Cyclophosphamide (Cytoxan)
- Cyclosporine (Neoral)
- Anakinra (Kineret)
- Antitumor necrosis factor therapies: adalimumab (Humira), etanercept (Enbrel), infliximab (Remicade)

If adult-onset Still's disease affects your heart or lungs, doctor may also need to prescribe medication to treat these problems.

#### *Self-care*

With adult Still's disease, the medications may need to be taken even after symptoms go away. This is called maintenance therapy. It is important to keep the inflammation under control to prevent more damage to the body. If prednisone is taken for a long time, a doctor may recommend taking calcium and vitamin D to prevent thinning of the bones.

#### *Complications*

- Pericarditis
- Pleural effusion
- Macrophage activation syndrome
- Arthritis in several joints
- Liver disease
- Spleen enlargement

#### *Prognosis and Recurrence*

- It isn't yet possible to prevent Still's disease, and there is no cure.
- Some people have just one episode of adult Still's disease. In other people, the condition persists or recurs.

- 1 in 5 people have symptoms that go away and never come back.
- About 1 in 3 have symptoms that go away but come back several times over several years. These relapses are often less severe and shorter than the first episode.
- 1 in 2 people have symptoms that last a long time, with the disease sometimes affecting vital organs.

#### *Differential Diagnosis*

- Lyme disease,
- Crohn's disease

#### **References**

1. Clarke JT. Other Rheumatologic Diseases. In: Bologna JL, Jorizzo JL, Schaffer JV, et al, eds. *Dermatology*. 3rd ed. Philadelphia, PA: Elsevier Mosby; 2012:chap 45.
2. Gerfaud-Valentin M, Maucort-Boulch D, Hot A, et al. Adult-onset still disease: manifestations, treatment, outcome, and prognostic factors in 57 patients. *Medicine (Baltimore)*. 2014; 93:91.
3. Lee LA, Werth VP. The Skin and Rheumatic Diseases. In: Firestein GS, Budd RC, Gabriel SE, et al, eds. *Kelley's Textbook of Rheumatology*. 9th ed. Philadelphia, PA: Elsevier Saunders; 2012: chap 43.
4. Cecilia Giampietro; Bruno Fautrel. "Review Article: Anti-Interleukin-1 Agents in Adult Onset Still's Disease". *International Journal of Inflammation*. 2012 (317820).
5. Luthi F, Zufferey P, Hofer MF, So AK. "Adolescent-onset Still's disease": characteristics and outcome in comparison with adult-onset Still's disease". *Clin. Exp. Rheumatol*. 2002; 20(3):427-30.