



## Treatment

The goal of treatment is to stop symptoms and normalize inflammation parameters in the blood.

Several treatments may be offered.

Corticosteroids are most commonly used at the very beginning, along with anti-inflammatory drugs such as aspirin.

Currently, international recommendations favor the **anti-interleukin-1 biotherapy** Anakinra as first-line treatment, due to their high efficacy and fewer side effects than corticosteroids.

As a second-line treatment, other biotherapies can be used, such as **anti-interleukin 6** (tocilizumab) or another long-acting anti-interleukin 1 such as canakinumab.

Methotrexate may be added as a supplement to reduce corticosteroid use.

If treatment fails, contact a center specialized in the treatment of Still's disease (CEREMAIA) to discuss alternative treatments.

Powerful treatments are available in case of severe lung, heart, or liver damages and in case of macrophage activation syndrome.



### Our team:

#### Internal medicine

Prof. Sophie GEORGIN-LAVIALLE

Dr. Léa SAVEY

Dr. Marion DELPLANQUE

Dr. Catherine GRANDPEIX-GUYODO

Dr. Yves-Jean Zhu

Dr. Philippe MERTZ

Dr. Robin ECHERBAULT

#### Dermatology & Allergology

Prof. Angèle SORIA

Dr. Jean-Benoît MONFORT

#### Nephrology

Prof. Jean-Jacques BOFFA

#### Anatomopathology

Prof. David BUOB

#### Hepatogastroenterology

Dr. Ingrid POPA

#### **Tenon Hospital**

**Internal Medicine Department**

**4 rue de la Chine**

**75020 Paris, France**

**Tel: +33 (0)1 56 01 74 31**

**Fax: +33 (0)1 56 01 71 46**

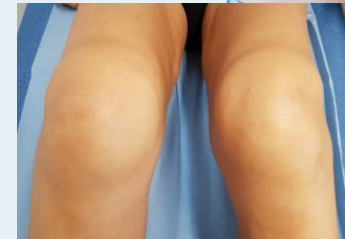
✉ [ceremaia-medecine-int.tenon@aphp.fr](mailto:ceremaia-medecine-int.tenon@aphp.fr)

📷 @CEREMAIA\_Tenon

<https://www.maladiesautoinflammatoires.fr/>



## Still's disease



@CEREMAIA\_Tenon



<https://www.maladiesautoinflammatoires.fr/>



@CEREMAIA\_Tenon



# Still's disease

## Definition

Still's disease is a rare autoinflammatory disease that affects children and adults.

## Epidemiology

Still's disease affects between 1 and 2 per million inhabitants per year in France.

It affects men and women equally.

It can begin at any age.

It is estimated that there are between 500 and 700 patients with the disease in France.

## Pathophysiology

The cause is not yet understood.

No genetic cause has been identified to date.

Still's disease is not infectious and is therefore not contagious.



## Clinical

Still's disease's manifestations combine several of the following signs, which last for 15 to 30 days in the absence of treatment:

- High **fever**  $\geq 39^{\circ}\text{--}40^{\circ}\text{C}$  for more than 7 days, often occurring at the end of the day.
- **Joint** pain or swelling.
- A **rash** on the skin, often pink or salmon pink, occurring during fever spikes.
- Difficulty/pain when swallowing.
- Swollen lymph nodes.

Other rarer symptoms may sometimes be observed:

- Fluid around the heart: pericarditis.
- Fluid around the lungs: pleuritis.
- Liver damage: resulting in increased liver enzymes (cytolysis) and, sometimes, cholestasis.
- Neurological, ophthalmological, or renal damages are very rare.

## Diagnosis

Ruling out infectious, autoimmune, and cancerous causes is needed to establish the diagnosis of Still's disease.

It may be useful to refer to classification criteria such as those developed by Yamaguchi and Fautrel.



## Progression

Still's disease can have several progression profiles:

1. A relatively mild or moderate typical presentation that usually resolves after a single flare-up with prompt treatment using biotherapy.
2. A more severe form with persistent high fever and high blood inflammation measured by CRP, ferritin, and total **interleukin 18**. There may be complicated presentations with macrophage activation syndrome and severe damages of the lungs, liver, and myocardium. In these forms, very powerful treatments are required from the outset and management is recommended in a department specialized in Still's disease. The goal is to reduce the inflammation quickly.

Follow-up must be very regular and include physical examination, blood tests (CRP, ferritin, complete blood count, liver function tests and, if possible, interleukin 18) in a center with a large experience in managing the disease.

In cases of lung, liver and/or myocardial involvement, these organs must be monitored very closely.

The long-term goal is to prevent relapses and avoid organ damage (liver, lungs, heart, joints).

