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Still's disease

What is Still's disease?

Still's disease is also known as systemic-onset juvenile [rheumatoid arthritis](#). This is just one of the many ways juvenile rheumatoid arthritis can present itself. It is characterised by high spiking fevers and a salmon-coloured skin rash. These symptoms often occur with other systemic symptoms, including swelling of the lymph glands, enlargement of the spleen and liver, sore throat, and inflammation of lungs and heart. Eventually all patients with Still's disease develop joint pain and swelling, the typical signs and symptoms of rheumatoid arthritis.

Who gets Still's disease?

Still's disease accounts for about 10–20% of the total cases of juvenile rheumatoid arthritis. It usually begins in early childhood between 1–4 years. Although rare, Still's disease can develop in adults (adult-onset Still's disease). In 75% of cases this occurs between 16–35 years of age.

What is the cause of Still's disease?

The exact cause of Still's disease is not known. One theory is that the disease is caused by infection with a microbe. Another theory is that Still's disease is a hypersensitive or autoimmune disorder (this is where the body's immune system which normally fights off infections or illness suddenly starts to attack the body's cells and tissues it is suppose to protect).

What are the signs and symptoms of Still's disease?

The main signs and symptoms of Still's disease include:

- Intermittent high fever occurring in all patients. Temperatures may get up to 39 or 40 degrees celcius before rapidly returning to normal levels. These fever spikes often occur at nearly the same time every day and may be accompanied by a feeling of extreme fatigue.
- Skin rash that coincides with the fever develop in about of 95% of patients. Characteristics of the rash are:
 - Small, non-itchy spots or patches up to 5cm in diameter
 - Salmon-pink coloured
 - Usually appear on the limbs and trunk, but may occur on the face or neck.
- Other systemic symptoms include:
 - Swollen lymph glands and/or enlarged spleen/liver in 85% of patients
 - Inflamed lungs (pleuritis) and/or surrounding heart (pericarditis) in 60% of patients
 - Marked increase in white blood cell count
 - Severe anaemia
 - Abdominal pain

In many cases, skin rash and fever may occur intermittently for months or even years before joint pain and swelling develop. All patients with Still's disease eventually develop persistent chronic arthritis even if fever and systemic symptoms have stopped. Arthritis usually affects many joints but in particular the wrists may become severely deformed.

Still's rash



What is the treatment of Still's disease?

There is no cure for Still's disease. The main goals of treatment are to reduce joint inflammation and avoid deformity. Therapy may include both medical and surgical interventions. In addition, the patient and their families need to fully understand the nature of the disease and how to live with it for the rest of their lives.

Drug therapy to control many of the symptoms of Still's disease includes:

- Aspirin (note that young children are susceptible to salicylate toxicity)
- Nonsteroidal anti-inflammatory drugs such as ibuprofen, naproxen and indomethacin
- [Corticosteroids](#) (e.g. prednisone), [methotrexate](#), [gold](#), [azathioprine](#) may be used for more severe symptoms

Non-drug therapy includes physiotherapy, hydrotherapy and simply rest. Patients with Still's disease will experience "good" and "bad" days. The number of "bad" days, that is flare-ups or exacerbations of the disease, can be controlled or reduced by careful planning of activities and rest periods. It has been shown that physical and emotional stress can exacerbate symptoms of Still's disease.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Rheumatoid arthritis](#)

Other websites:

- [International Still's Disease Foundation](#)
- [Still's Disease \(Systemic-Onset Juvenile Rheumatoid Arthritis\)](#) - MedicineNet.com
- [Juvenile Rheumatoid Arthritis](#) - emedicine
- [Juvenile Rheumatoid Arthritis](#) - MedlinePlus

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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