A quarter of patients surveyed reported mental health issues including: irritability, low self-esteem, and difficulty coping with stress, anxiety, isolation, frustration, and depression.

- “I feel like a ghost of my former self, because I’m not able to do all of the same activities I did before onset. It’s like I’m grieving for that person to come back.” – April, AOSD
- “Having to cancel plans with friends and family because my disease is flaring, or may flare, riddles me with guilt.” – Kelly, AOSD
- “It’s important to remember to stay positive, since stress can increase disease flaring.” – Amanda, SJIA (now adult)
- “As a parent, it’s difficult because I can’t feel what she feels. I can only see what she feels.” – Natalie, parent of Zoe, SJIA

Patient-Reported #MyStills Experiences

- “I get frequent flash fevers throughout the day, often at night. By morning it’s normal.”
- “Joint pain comes and goes, “jumping” to different places. Joint stiffness is worse after sleep and sitting for a long time.”
- “Extreme, chronic, and constant fatigue. I feel exhausted when I wake up.”
- “Sometimes the pain is so bad that it hurts to be touched.”
- “Not all rashes are salmon-colored, flat, and quick to disappear. Sometimes they itch.”
- “Fever, pain, and sore throats often do not respond to over-the-counter medications.”

Want to get involved in our #MyStills campaign? Learn more at aiarthritis.org/stillsdisease. We will continue collecting patient-reported Still’s experiences to use towards the development of future materials.

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International Foundation for Autoimmune & Autoinflammatory Arthritis (AiArthritis)

www.AiArthritis.org
Tax ID: 27-1214308

Give today at aiarthritis.org/donate. Your gift will help us continue improving the lives of those affected by Still’s Disease.

Some studies suggest certain cytokine profiles (interleukins 1, 18 and IL-18BP) and human leukocyte antigen (HLA) genotypes may provide diagnostic clues, but no consistent results are clinically validated.

1 Rheumatoid factor can be positive in healthy people, so having a positive RF should not dictate diagnosis.
WHAT IS STILLS

Still’s Disease is a rare, multi-systemic (whole body) autoimmune condition that can vary from person to person.

Because of a similar pathogenesis, broadly overlapping symptoms, and organ involvement, Still’s Disease represents a disease continuum that includes both Systemic Juvenile Idiopathic Arthritis (SJIA) and Adult-Onset Still’s Disease (AOSD). Those with onset > 16 years of age are categorized with AOSD, while those originally diagnosed with SJIA carry this diagnosis through adulthood. Onset can occur at any age, even presenting after birth.

In some individuals, Still’s Disease develops suddenly, disappears almost as quickly, and may not return for many years, if ever. In others, it is a chronic condition. Symptoms and progression vary per individual. Some patients will have many of these symptoms, some only a few. This makes detecting, diagnosing, and treating Still’s difficult.

Some researchers suspect the condition might be triggered by a viral or bacterial infection or genetic mutations.

We believe the shared experiences from Still’s patients offers a better understanding of the disease and may assist with diagnosis. The content in the symptoms section was developed by analyzing research based on patient-reported disease experiences then reviewed by medical experts from the Center for Autoinflammatory Diseases.

SYMPTOMS

Patients should not be expected to have all of these symptoms. They will vary in degree based on the individual.

Rash - Most commonly a salmon colored rash appearing on the upper anterior torso, arms, and legs that may quickly fade.
- Typically does not itch or mild itch, however patients reported a rash that can itch, be raised, appear as a number of small, flat red spots, and may not quickly fade away. 4
- May not be visible if covered with hair or if fades very quickly.

Fever - High and spiking, rising to 104 degrees F/41 degrees C or higher and can rapidly return to normal levels or below.
- Time of day can vary, but evening and nighttime reported more often.
- May come and go, sometimes twice a daily, but may last a week or longer.
- Could cause for patient-reported excessive sweating/night sweats.5

Fatigue - Reported in > 90%; common in all autoinflammatory diseases, not unique to Still’s.

Myalgia - Reported in > 75% of patients, some described similar to flu-like weakness.

Sore Throat - All age groups, even higher reports in children. 6 Some report difficulty swallowing.

Inflammatory Arthritis - Still’s Disease was once considered “a form of arthritis”, mimicking Rheumatoid Arthritis (and arthritis remains in the SJIA name). However, research shows arthritis may not be present at onset in about 25% of patients - and some may not develop it for years, if ever.
- Joint pain can vary from individual to individual from mild to chronic. Usually polyarticular and symmetrical.
- Swelling, redness, and warmth possible.
- Potential for joint destruction.
- Can cause severe stiffness after rest.

Organ Involvement
- Generalized lymph nodes/glandular swelling.
- Enlargement of the liver and/or spleen.7
- Inflammation of lungs (pleuritis), the saccle covering of your heart (pericarditis), the muscular portion of your heart (myocarditis), eyes, serous tissues.8,9
- Organ involvement may be present at onset.

Gastrointestinal
- Nausea, weight loss, poor appetite.
- 11% of patients surveyed reported canker sores in mouth.

Macrophage activation syndrome (MAS). 10 A massive inflammatory response that overwhelms the whole body, including organs. MAS is more common in SJIA and may be present at onset.

Other: Central nervous system manifestations, seizures, meningismus.

Other: Over 30% of patients reported “brain fog”, which may be associated with inflammation in the brain caused by the immune response.11

1Research based on 2015 Early Symptoms Study (ESS) combined with 2019-2020 focus groups and survey participation, both conducted by the International Foundation for Autoimmune & Autoinflammatory Arthritis.
2There is increasing evidence of atypical rashes: raised, red or purple plaques; itchy: bumps that closely resemble eczema or urticaria. Atypical rashes, in conjunction with other typical Still’s symptoms, should be tested to rule out other causes, such as urticarial vasculitis.
3Some patients reported feeling extremely hot and flushed, without registering a fever.
4Up to 70% of children report sore throat.
5May be cause of abdominal pain, which was reported by approximately 20% of participants.
6May be responsible for patient-reported “shallow breathing” or “chest pain”.
7Serositis refers to inflammation of the serous tissues of the body, the tissues lining the lungs, heart, and the inner lining of the abdomen and organs within.
8Haemophagocytic lymphohistiocytosis (HLH) is termed macrophage activation syndrome (MAS) when associated with rheumatic disease (secondary HLH) and triggers including malignancy and infection.
9Medical reviewers could not confirm this symptom as related directly to Still’s Disease.