

Progression

To date, no severe complications have been reported, in particular no inflammatory amyloidosis or excess mortality.

However, patients experience severe fatigue after attacks, and recurrence can have a negative impact on quality of life and work attendance.

Treatment

It has not yet been codified because the entity has been recently described.

Colchicine appears to shorten the duration of symptoms and lengthen the healthy interval between episodes in some patients.

For the most severe cases that are resistant to colchicine, consultation with an expert center at Tenon Hospital is recommended.



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Our team:

Internal medicine

Prof. Sophie GEORGIN-LAVIALLE
Dr. Léa SAVEY
Dr. Marion DELPLANQUE
Dr. Catherine GRANDPEIX-GUYODO

Dermatology & Allergology

Prof. Angèle SORIA
Dr. Jean-Benoît MONFORT


Rheumatology

Dr Philippe Mertz

Anatomopathology

Prof. David BUOB

Tenon Hospital
Internal Medicine Department
4 rue de la Chine
75020 Paris, France
Tel: +33 (0)1 56 01 74 31
Fax: 01 56 01 71 46

 ceremaia-medecine-int.tenon@aphp.fr

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SITRAME syndrome



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SITRAME syndrome



Definition

This is an acquired autoinflammatory syndrome in adults, meaning it is not inherited from parents. Patients experience episodes that are:

- Systemic (with fever);
- Inflammatory (elevated CRP in the blood);
- Recurrent;
- Acute;
- erythematous-macular (red) rash predominantly on the trunk (chest, back) and the root of the limbs.

The acronym "SITRAME" has been proposed in English and stands for:

S Systemic
I Inflammatory
T Truncular
R Recurrent
A Acute
M Macular
E Eruption

Soria A, Amsler E, Boursier G, Georgin-Lavialle S. Provisional diagnostic criteria for systemic inflammatory trunk recurrent acute macular eruption diagnosis. *J Eur Acad Dermatol Venereol*. 2025;39(1):e87-e90

Soria A, Amsler E, Garel B, Moguelet P, Tieulié N, Cordoliani F, Guichard I, Mahé A, Grateau G, Boursier G, Georgin-Lavialle S. Systemic inflammatory trunk recurrent acute macular eruption (SITRAME): A new auto-inflammatory syndrome in adult? *J Eur Acad Dermatol Venereol*. 2023;37(4):e538-e542.



Epidemiology

The disease has so far been described in adult subjects of Caucasian origin. The average age of onset of attacks is between 25 and 50 years old.

The cases described are sporadic: the patients are the only ones affected in their families. To date, there are approximately 50 identified cases in France.



Clinical features

Patients present with recurrent acute episodes of fever followed by a stereotypical erythematous (red) macular (flat, without relief) rash that does not itch, always located in the same areas of the trunk and sometimes the root of the limbs, with the vast majority of cases sparing the periumbilical area.

Inflammation is found in the blood (elevated C-reactive protein) during flare-ups, and patients often experience severe fatigue after an episode.

The rash lasts an average of three days and is accompanied by fever in more than half of cases.

Other symptoms: headaches, abdominal pain, muscle pain, sore throat, and flu-like symptoms may be associated with the rash.



Genetics

To date, no mutation in a gene already implicated in an autoinflammatory disease has been identified by next-generation sequencing (panel) in the blood of patients.



Diagnosis

The diagnosis of SITRAME is **clinical**, based on the combination of **major and minor criteria**:

Major criteria (mandatory):

- 1.Systemic inflammation:** at least one documented episode of CRP > 5 mg/ml during a skin flare-up
- 2.Non-pruritic macular rash of the trunk**
- 3.Recurrence:** at least 3 different episodes
- 4.Acute:** duration of attacks less than 8 days

At least one minor criterion among:

- 1.Fever during flare
- 2.Flare-ups triggered by infections, vaccinations or intense physical exercise
- 3.Asthenia during and/or after flare
- 4.Associated papular rash

It remains a **diagnosis of exclusion**: other causes such as infection, drug allergy, or known dermatologic or autoinflammatory conditions must be ruled out.

Episodes may be triggered by **medications, infections (viral or bacterial), or occasionally vaccination**.

Referral to an **expert center (Tenon Hospital)** is recommended.

