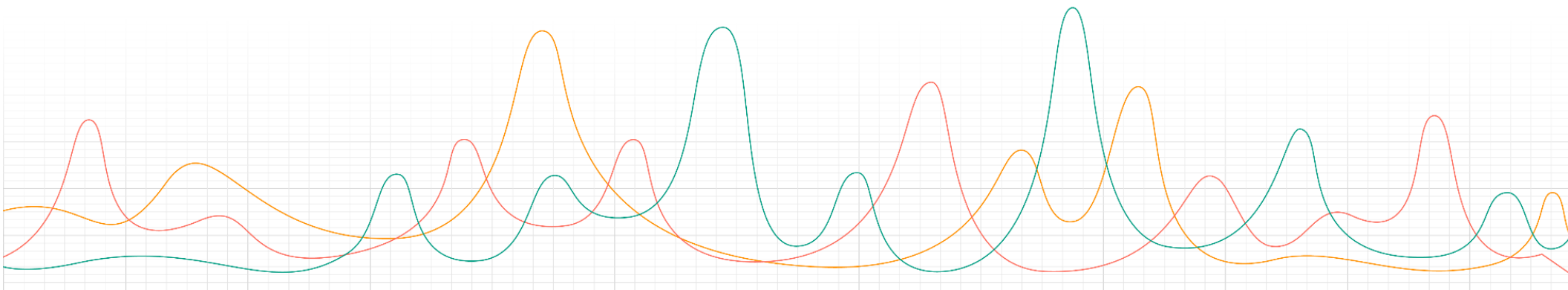




DISEASE ACTIVITY CALCULATOR

Details, Diagnosis Criteria, and References



Calculating Disease Activity in Patients with Systemic Disease:

By using the disease activity calculator you can assess disease severity and need for therapy. Having a disease activity measure or score can help the patient, family or clinician understand:

- A. Severity of disease
- B. Need for aggressive therapies (such as corticosteroids [steroids, prednisone] or biologic therapies [IL-1 or IL-6 inhibitors])
- C. Eligibility for enrollment in clinical trials (drug studies)
- D. When to consider tapering or withdrawing therapy

While several scoring methods have been proposed, there is no universally accepted measure of disease activity in AOSD or SJIA. We provide a disease activity calculator that is based on disease manifestations used to calculate activity.

The Still's Disease Activity Calculator is provided as a service and guide to physicians, clinical trialists and patients for their educational, informational and research use. The disease activity results acquired here should be shared with your rheumatologist or treating physician who is expert in the diagnosis of Still's disease and Autoinflammatory diseases to confirm the diagnosis and to develop a treatment plan tailored to the patient.

Diagnosis Scoring Criteria

| Manifestations [#] | SDII* | mPouchot* | SMS* | SFS* |
|---|-------|-----------|------|------|
| Daily or nightly fever 37.5–38°C (99.5-100.3°F) | - | - | 1 | 1 |
| Daily/nightly fever 38–39°C (100.4-102.1°F) | - | 1 | 2 | |
| Daily/nightly fever 39–40°C (102.2-103.9°F) | 1 | | 3 | |
| Daily/nightly fevers above >40°C (104°F) | | | 4 | |
| Muscle pains (myalgia) | - | 1 | - | - |
| ≥2 swollen joints (inflammatory synovitis) | 1 | 1 | | |
| Still's Rash (nonfacial; seen by physician) | 1 | 1 | 1 | 1 |
| Weight loss (>10% total body weight) | 1 | - | - | - |
| Sore throat (current or recent within 2 wks.) | 1 | 1 | 1 | |
| Pleuritis or pleural effusion | 1 | 1 | 1 | 1 |
| Pericarditis or pericardial effusion | | 1 | | |
| Pneumonitis (by CXR) | | 1 | | |

| | | | | |
|---|---|---|---|---|
| Peritonitis | | - | | |
| Generalized lymphadenopathy | 1 | 1 | 1 | 1 |
| Hepatomegaly or splenomegaly | | 1 | 1 | 1 |
| Elevated hepatic enzymes (AST or ALT > 1.5 ULN) | 1 | | - | - |
| Elevated CRP (≥ 20 mg/L) | 1 | - | - | 1 |
| Elevated ESR (> 50 mm/hr.) | | - | - | 1 |
| Elevated Ferritin (≥ 500 mg/ml) | | 1 | 1 | |
| Elevated Platelet count > 600×10^3 /L | - | - | | 1 |
| Elevated WBC ≥ 12.5 | 1 | 1 | - | 1 |
| Anemia (low hemoglobin <9 g/dl) | - | - | 1 | 1 |

*SDII: Systemic Disease Inflammatory Index; mPouchot: modified Pouchot criteria; SMS: systemic manifestation score; SFS: Systemic Feature Score; CXR: chest x-ray; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; WBC: white blood cell count

#Modifications: for ease of "calculation" several criteria were modified from their original definitions. The modified Pouchot required a ferritin level > 3000 and a WBC > 15. The SFS required a CRP > 10, ESR > 20, Platelet count > 400, WBC > 12 and anemia (hemoglobin) < 11.

Diagnosis Scoring Criteria

| | SDII | mPouchot | SMS | SFS |
|----------------------|---------------|----------|-----|-----|
| Remission score | 0 | 0 | 0 | 0 |
| Active Disease score | ≥ 4 | ≥ 4 | - | - |
| Maximum Activity | 10 | 12 | 10 | 10 |
| Major improvement | ≥50% decrease | | | |
| Improved | ≥2 decrease | | | |
| Flare | ≥2 increase | | | |

Systemic Disease Inflammatory Index (SDII)

Systemic Disease Inflammatory Index (SDII), also known as the Still's disease inflammatory index, this "score" is proposed by Dr. J Cush and is meant to reflect inflammatory (systemic) disease severity either at the outset or during the course of disease. This was devised to assess Still's disease activity, but can also be applied to other autoinflammatory disorders and periodic fever syndromes. An SDII of > 4 points can be used as an indicator of need for aggressive (biologic) therapy or inclusion in a clinical trial of Still's disease.

Scoring: 1 point each for

1. Fever > 102 oF (> 39°C)
-

-
2. Current/recent sore throat (w/in 2 wks.)
 3. Current rash (erythematous, nonfacial, observed by MD)
 4. Weight loss >10% of basal weight (w/in 3-12 mos.)
 5. Serositis: pleuritis, pericarditis, peritonitis, or pneumonitis by (exam or imaging)
 6. Reticuloendothelial system (RES) involvement: Hepatomegaly, splenomegaly, or generalized lymphadenopathy (> 2 chains)
 7. Inflammatory synovitis > 2 joints
 8. Elevated WBC > 12.5 x 10³ cells/mm³
 9. Elevated ESR > 50 mm/hr. OR CRP > 2.0 mg/dl
 10. Elevated AST or ALT > 1.5 ULN

Interpretation: a score of 4 point or higher is active systemic disease that merits aggressive treatment. The most active (maximum score) is 10 points. Remission is indicated by a score of zero (0) and low activity by a score of 1-3. A “flare” or worsening of disease is defined as an increase in SDII > 2 points. For those under treatment, major improvement is defined as a decrease of >50%; “Improved” would be a decrease in SDII of 2 points or more.

Modified Pouchot-Activity Score

Modified Pouchot-Activity Score was originally devised by Pouchot and later the scoring parameters were modified by Rau et al and is a useful clinical tool for distinguishing between acute and chronic courses of AOSD and sepsis. The primary difference between the original Pouchot score and the Rau modification is the addition of ferritin to the scoring parameters. This activity score is based on one point each for 12 typical disease parameters: fever, evanescent rash, sore throat, arthritis, myalgia, pleuritis, pericarditis, pneumonitis, lymphadenopathy, hepatomegaly or pathological liver function tests, leucocyte count > 15,000/ μ l, and serum ferritin > 3000 μ g/l. Three different disease states were defined: active disease, partial remission and remission. This score has been used for adults and children with Still’s disease.

Systemic Manifestation Score (SMS): was proposed by Saccomanno et al and was used to quantify the activity of systemic disease in systemic JIA patients. The SMS ranges from 0 to 10, where 0 = absence of systemic manifestations and 10 = maximum activity of systemic manifestations. The SMS was used to correlate inflammatory activity features with outcomes. Scoring is based on Still's features that excluding arthritis. More points were given for higher degrees of fever "(It was decided to assign a greater weight to fever owing to its greater effect on a child's well-being and major importance in driving treatment decisions)":

- Fever
 - 1 point if 37–38°C
 - 2 points if 38–39°C
 - 3 points if 39–40°C
 - 4 points if > 40°C
- Rash = 1 point;
- Generalized lymphadenopathy = 1 point;
- Hepatomegaly and/or splenomegaly = 1 point;
- Serositis = 1 point;
- Anemia (hemoglobin < 9 g/dl) = 1 point;
- Platelet count > × 109/l or ferritin > 500 ng/ml = 1 point <https://www.jrheum.org/content/jrheum/46/4/416.full.pdf>

Systemic Feature Score (SFS)

Systemic Feature Score (SFS): was proposed by Vojinovic et al. to evaluate disease activity and to be used at 2-4 week intervals to assess disease severity and response to therapy. The SFS consists of 5 clinical and 5 laboratory parameters, each assigned a score of 1 (present) or 0 (absent).

Clinical features

-
- Fever
 - Rash
 - Enlargement of lymph nodes
 - Enlargement of liver or spleen size
 - Serositis.

Laboratory features

- ESR ≥ 20 mm/hour
- CRP ≥ 10 mg/liter
- White blood cell (WBC) count $\geq 12 \times 10^9$ /liter
- Hemoglobin ≤ 11 gm/dl
- Platelet count $\geq 400 \times 10^9$ /liter

“During treatment phases and at follow-up visits, the laboratory parameters were scored as follows: for ESR, score of 0 if < 20 mm/hour or if decreased by $\geq 30\%$ compared to baseline, score of 1 if increased or if decreased by $< 30\%$ compared to baseline; for CRP, score of 0 if < 10 mg/liter or if decreased by $\geq 30\%$ compared to baseline, score of 1 if increased or if decreased by $< 30\%$; for WBC count, score of 0 if $< 12 \times 10^9$ /liter or if decreased by 20% compared to baseline, score of 1 if increased or if decreased by $< 20\%$; for hemoglobin level, score of 0 if < 11 gm/dl or if increased by 20% compared to baseline, but score of 1 if decreased or if increased by $< 20\%$; for platelet count, score of 0 if $< 400 \times 10^9$ /liter or if decreased by 20% compared to baseline, score of 1 if increased or if decreased by $< 20\%$ ”

References

Cush JJ. Still's (Systemic) Disease Inflammatory Index, presented during the "Autoinflammatory Disorders:

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Saccomanno B, et al. Predictors of Effectiveness of Anakinra in Systemic Juvenile Idiopathic Arthritis. *J Rheumatol*. 2019 Apr;46(4):416-421.

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