

## Composite Score to assess disease activity in patients with Familial Mediterranean Fever

|  |   |   |  |   |                              |                             |
|--|---|---|--|---|------------------------------|-----------------------------|
| <b>Frequency of attacks</b>  | <input type="checkbox"/> ≤2/year  | <input type="checkbox"/> 3-6/year   | <input type="checkbox"/> 7-11/year   | <input type="checkbox"/> ≥12/year   |                              |                             |
| <b>School/work days missed due to FMF</b>  | <input type="checkbox"/> ≤ 5/year   | <input type="checkbox"/> 6-10/year  | <input type="checkbox"/> 11-20/year  | <input type="checkbox"/> >20/year   |                              |                             |
| <b>Increased acute-phase reactants during the attack-free period<sup>1</sup></b> | <input type="checkbox"/> CRP ≤2x UL<br><input type="checkbox"/> SAA ≤14 mg/L                      | <input type="checkbox"/> CRP >2 and ≤4x UL<br><input type="checkbox"/> SAA >14 and ≤50 mg/L | <input type="checkbox"/> CRP >4 and ≤8x UL<br><input type="checkbox"/> SAA >50 and ≤100 mg/L | <input type="checkbox"/> CRP >8 x UL<br><input type="checkbox"/> SAA >100 mg/L    |                              |                             |
| <b>Chronic sequelae<sup>2</sup></b>  |   |   | <input type="checkbox"/> new or worsening sequelae   |   |                              |                             |
| <b>Evaluation</b>  | <input checked="" type="checkbox"/> for all items<br><b>Remission or minimal disease activity</b> | <input checked="" type="checkbox"/> for ≥1 item<br><b>Mild disease activity</b>             | <input checked="" type="checkbox"/> for ≥1 item<br><b>Moderate disease activity</b>          | <input checked="" type="checkbox"/> for ≥1 item<br><b>Severe disease activity</b> |                              |                             |
| <b>Global assessment: target reached?</b>  |   |   |  |   |                              |                             |
| <b>Physician<sup>3</sup></b>   | <input type="checkbox"/> yes  | <input type="checkbox"/> no   |  |   | <input type="checkbox"/> yes | <input type="checkbox"/> no |
| <b>Patient<sup>4</sup></b>   | <input type="checkbox"/> yes  | <input type="checkbox"/> no   |  |   | <input type="checkbox"/> yes | <input type="checkbox"/> no |
| <b>Consequences</b>  | <b>Path 1</b><br><i>Consider paths 5/6 if applicable</i>  | <b>Path 1: observe closely</b><br><i>Paths 2/3 might be considered</i>                      | <b>Paths 2-4</b>   |   |                              |                             |

<sup>1</sup> ≥2 weeks after the last attack, measured at least two times 1 month apart

<sup>2</sup> Amyloidosis, growth retardation (<3<sup>rd</sup> percentile), symptomatic anaemia requiring therapy, thrombopenia due to splenomegaly, persisting arthritis lasting > 6 weeks

<sup>3</sup> Are you satisfied with the current FMF-related health status of your patient?

<sup>4</sup> Is the patient satisfied with their current FMF-related health status?