# Points to consider in hyperinflammation

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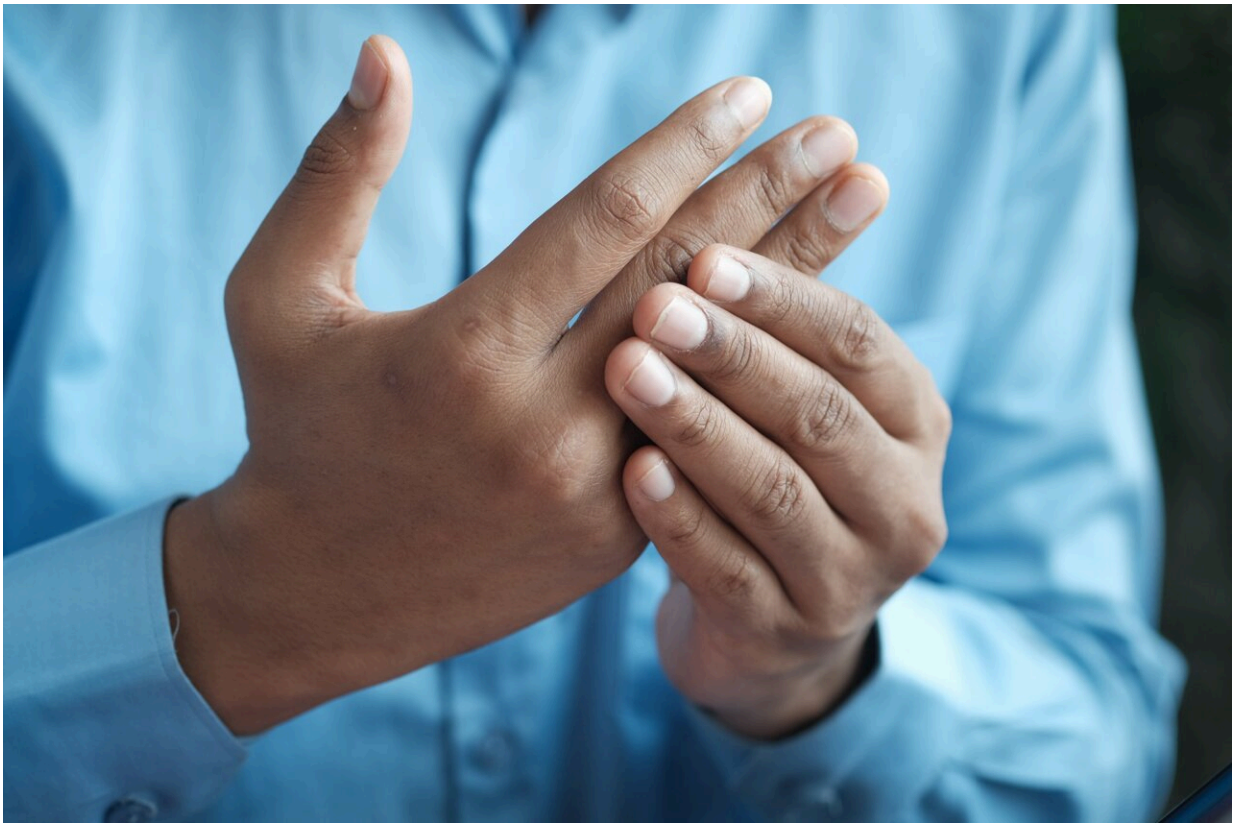

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Hemophagocytic lymphohistiocytosis (HLH) and macrophage activation syndrome (MAS) are life-threatening systemic hyperinflammatory syndromes. Systemic hyperinflammation and HLH/MAS can occur in nearly any inflammatory state, but there are certain predisposing conditions and inflammatory triggers. This includes rheumatic diseases,

malignancies, metabolic diseases, and genetic immune problems.

HLH and MAS are characterized by fever, increased systemic and central nervous system inflammation, low blood cell count, coagulopathy, and hepatitis. This can lead to multiple organ dysfunction, shock, and death. Both HLH and MAS can progress very quickly, so early identification and management are critical. But it is hard to identify at-risk patients.

To support this, EULAR has developed new evidence- and consensus-based points to consider to support clinicians in the diagnosis, treatment, and monitoring of HLH/MAS. The work was completed by an expert task force of adult and pediatric rheumatologists, hematologists, oncologists, immunologists, infectious disease specialists, intensivists, and allied health care professionals—as well as patients and their parents.

The resulting paper, published in the *Annals of Rheumatic Disease* and simultaneously in *Arthritis & Rheumatology* includes six overarching statements and 24 specific points to consider.

Major themes include the need for prompt syndrome recognition and systematic evaluation of underlying contributors, alongside early intervention that targets both the hyperinflammation and its likely contributors. They also stress the need for careful monitoring of progression and complications, as well as the need for expert multidisciplinary assistance. A helpful flowchart is provided to outline the key stages of assessment, investigation, and treatment in people with hyperinflammation and suspected HLH/MAS.

EULAR hopes these new points to consider will help guide the initial evaluation, management and monitoring of people with HLH/MAS in order to halt disease progression and prevent life-threatening

immunopathology.

**More information:** Bita Shakoory et al, The 2022 EULAR/ACR points to consider at the early stages of diagnosis and management of suspected haemophagocytic lymphohistiocytosis/macrophage activation syndrome (HLH/MAS), *Annals of the Rheumatic Diseases* (2023). DOI: 10.1136/ard-2023-224123

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