

EULAR
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NEW: POINTS TO CONSIDER FOR DIAGNOSIS, TREATMENT AND MONITORING OF SYSTEMIC AUTOINFLAMMATORY DISEASES

EULAR 'points to consider' published in the *Annals of the Rheumatic Diseases*

Diagnosis and management of Systemic Autoinflammatory Diseases (SAIDs) can be challenging, and when left untreated people with severe SAIDs have poor prognosis. But there are life-changing treatments that can significantly improve outcomes. EULAR – The European Alliance of Rheumatology Associations – has developed a set of new points to consider in order to help improve care for people with these rare diseases.

SAIDs are a group of multi-system disorders caused by dysfunction in the innate immune system. A number of these SAIDs are mediated by a cytokine called interleukin-1 (IL-1), including the cryopyrin-associated periodic syndromes (CAPS), tumour necrosis factor receptor-associated periodic syndrome (TRAPS), mevalonate kinase deficiency (MKD), and deficiency of the IL-1 receptor antagonist (DIRA). These rare diseases usually start in early childhood with variable multiorgan involvement.

The new EULAR points-to-consider were developed by a multidisciplinary task force from 7 European countries. The people taking part had expertise in paediatric and adult rheumatology. The group also included allied health professionals and patient representatives. The work was completed in line with EULAR standardized operating procedures. The aim was to establish evidence-based recommendations for diagnosis, treatment and monitoring of people with IL-1 mediated autoinflammatory diseases.

The paper developed by EULAR and published in the month 2022 issue of the *Annals of the Rheumatic Diseases* includes five overarching principles and three sets of points to consider. The principles say that a multidisciplinary team is required to evaluate and manage people with IL-1 mediated SAIDs, and that patients presenting with flares of unexplained systemic inflammation should receive a prompt diagnostic workup including genetic testing and screening for complications. The goal of treatment is to control clinical signs and symptoms and normalize inflammation using a treat-to-target approach, and requires long-term monitoring.

The three sets of points to consider include 14 statements related to diagnosis, 10 on therapy, and 9 focused on long-term monitoring. For diagnosis, the points cover genetic and clinical workup for specific diseases. The statements on therapy give suggestions for the use of IL-1 blocking therapy, each supported by a grade of recommendation and level of supporting evidence. The 9 points to consider on monitoring touch on disease activity and burden, as well as growth and development of children with SAIDs, and the risk of complications and infections. These also cover recommendations for immunizations.

EULAR believe these points to consider can guide diagnostic evaluation, treatment and monitoring of patients, and help to standardize and improve care, quality of life, and disease outcomes.

Source

[Romano M, et al. The 2021 EULAR/American College of Rheumatology points to consider for diagnosis, management and monitoring of the interleukin-1 mediated autoinflammatory diseases: cryopyrin-associated periodic syndromes, tumour necrosis factor receptor-associated periodic syndrome, mevalonate kinase deficiency, and deficiency of the interleukin-1 receptor antagonist. *Ann Rheum Dis* 2022;81:907–921.](#)

About EULAR

EULAR is the European umbrella organisation representing scientific societies, health professional associations and organisations for people with rheumatic and musculoskeletal diseases (RMDs). EULAR aims to reduce the burden of RMDs on individuals and society and to improve the treatment, prevention and rehabilitation of RMDs. To this end, EULAR fosters excellence in education and research in the field of rheumatology. It promotes the translation of research advances into daily care and fights for the recognition of the needs of people with RMDs by the EU institutions through advocacy action.

Contact

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