

EAACI statement on the pathogenesis, immunology, and immune-targeted management of the Multisystem Inflammatory Syndrome in Children (MIS-C) or Pediatric Inflammatory Multisystem Syndrome (PIMS).

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Abstract

Multisystem inflammatory syndrome in children (MIS-C) is a rare, but severe complication of coronavirus disease 2019 (COVID-19). It develops approximately four weeks after severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection and involves hyperinflammation with multisystem injury, commonly progressing to shock. The exact pathomechanism of MIS-C

is not known, but immunological dysregulation leading to cytokine storm plays a central role. In response to the emergence of MIS-C, the European Academy of Allergy and Clinical Immunology (EAACI) established a task force (TF) within the Immunology Section in May 2021. With the use of an online Delphi process, TF formulated clinical statements regarding immunological background of MIS-C, diagnosis, treatment, follow-up, and the role of COVID-19 vaccinations. MIS-C case definition is broad, and diagnosis is made based on clinical presentation. The immunological mechanism leading to MIS-C is unclear and depends on activating multiple pathways leading to hyperinflammation. Current management of MIS-C relies on supportive care in combination with immunosuppressive and/or immunomodulatory agents. The most frequently used agents are systemic steroids and intravenous immunoglobulin. Despite good overall short-term outcome, MIS-C patients should be followed-up at regular intervals after discharge, focusing on cardiac disease, organ damage, and inflammatory activity. COVID-19 vaccination is a safe and effective measure to prevent MIS-C. In anticipation of further research, we propose a convenient and clinically practical algorithm for managing MIS-C developed by the Immunology Section of the EAACI.

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