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# Clinical Case Reports

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## A case of macrophage activation syndrome twice with starting canakinumab in a child with systemic juvenile idiopathic arthritis

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We present a 4 year old boy who presented initially at 2 years of age with pyrexia; rash and hepatosplenomegaly raised inflammatory markers and later joint swelling. Systemic Juvenile Idiopathic Arthritis (SJIA) was diagnosed after excluding infections and malignancy. He failed methotrexate and tocilizumab treatment, responded to IL blocker (anakinra). Family could not tolerate giving him the daily injections. Treatment was changed to IL  $\beta$  blocker (canakinumab). Presented 26 days later with pyrexia, abdominal distention, raised inflammatory markers, deranged clotting and rising liver enzymes and diagnosed with Macrophage Activation Syndrome (MAS) on clinical grounds and blood investigations. Bone marrow aspirate was negative. He required treatment with methyl prednisolone, etoposide and cyclosporine. After recovery; return to canakinumab treatment was attempted, he developed similar reaction with pyrexia, abdominal distention, raised ferritin, liver enzymes, fibrinogen and triglycerides fulfilling the criteria for MAS with SJIA. He was treated again with methyl prednisolone, cyclosporine and oral steroids. He was then restarted on IL 1 blocker (Anakinra) and remained well. Genetic HLH mutation and NLRP mutations excluded. This is a complex case of SJIA. The pivotal trials of IL  $\beta$  blocker use in SJIA did not highlight increased risk of MAS compared to other biologics. Infection could have been a trigger. Switching from one biologic to another is also an added factor for immune dysregulation. There is a possible unidentified genetic component, which could have made our patient more susceptible to MAS following of IL  $\beta$  blocker. Our case highlights the need for further collaboration.

### Biography

Khulood Khawaja is a Pediatric Rheumatology Consultant at Mafraq Hospital since august 2014. She did her Basic Medical Training and Postgraduate Education in England. She is registered as Sub Specialist in Pediatric Rheumatology in the Specialist Training Authority in the UK. She was a Member of Arthritis Research UK Pediatric Rheumatology clinical study group and was the Principle Investigator for several national research studies in the UK. She is aiming to improve access to care and research in Pediatric Rheumatology in Abu Dhabi and UAE.

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