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## Occult Macrophage Activation Syndrome in Patients with Systemic Juvenile Idiopathic Arthritis

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### *ABSTRACT.*

**Objective.** Macrophage activation syndrome (MAS) is a well described, but purportedly uncommon manifestation of systemic juvenile idiopathic arthritis (SJIA). There is evidence to suggest that macrophage activation is integral to the pathogenesis of SJIA. Accordingly, many patients with SJIA may have evidence of mild MAS that is not appreciated clinically. We investigated the prevalence of occult MAS in children with SJIA by reviewing bone marrow aspirates (BMA).

**Methods.** Patients diagnosed with SJIA who underwent bone marrow aspiration were identified retrospectively. Patients admitted with a diagnosis of fever of unknown origin and discharged with a diagnosis other than SJIA or malignancy, and who had a BMA, were identified as controls. The BMA were reviewed by a single hematopathologist for evidence of MAS, ranging from activated macrophages to frank hemophagocytic cells.

**Results.** Eight of 15 (53%) patients with SJIA had BMA suggestive of MAS. Two of 15 patients (13%) were diagnosed clinically with MAS. Three patients (20%) were noted to have frank hemophagocytosis, only one of whom was diagnosed with MAS clinically. There were no statistically significant differences in the laboratory values for the patients with and without evidence of MAS on BMA. There was no evidence of increased macrophage activity or hemophagocytosis in any of the control BMA.

**Conclusion.** Occult MAS appears to be common in patients with SJIA who undergo BMA. This suggests that macrophage activation may be integral to the pathogenesis of SJIA, with implications for treatment. (First Release Mar 1 2007; J Rheumatol 2007;34:1133-8)

### *Key Indexing Terms:*

MACROPHAGE ACTIVATION SYNDROME  
SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS  
HEMOPHAGOCYTOSIS

CD163 ANTIGEN  
BONE MARROW ASPIRATE  
HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

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