

Review criteria

We search PubMed for original papers focused on systemic juvenile idiopathic arthritis and pathogenesis. Search terms used, alone or in combination were “systemic juvenile arthritis,” “macrophage activation syndrome,” “hemophagocytic lymphohistiocytosis,” “autoinflammatory diseases,” “alternatively activated macrophages” and “S100A proteins.” The search was performed for articles published in English-language journals up to January 2011.