In format provided by Grom *et al.* (MAY 2016)

Supplementary Table 1: **Adjudication criteria for MAS cases**

Adjudication	Evidence
Canakinumab clinical program [1]	
Probable	Clinically consistent with MAS with histologic confirmation, laboratory features or meeting current formal HLH guidelines criteria [2], OR Clinical and laboratory features consistent with MAS but without histologic confirmation or meeting current formal HLH criteria
Possible	Laboratory features consistent with MAS but without clinical features, histologic confirmation, or meeting current formal HLH criteria
Unlikely	Some clinical and/or laboratory features of MAS, but with possible alternative explanation
Insufficient information	Insufficient information for adjudication
Tocilizumab clinical trials and postmarketing surveillance [3]	
Definite	Clear case of MAS meeting the preliminary MAS diagnostic criteria [4] and/or current formal HLH criteria [2]
Potential	Event that could be MAS but other potential diagnoses recorded
Not MAS	Event that did not meet the preliminary MAS criteria and other potential diagnoses recorded
Insufficient information	Insufficient information for adjudication

Definitions and associated probability of MAS in patients with sJIA as used in the canakinumab clinical program and tocilizumab phase III clinical trials and postmarketing surveillance in Japan.

Abbreviations: MAS, macrophage activation syndrome; HLH, haemophagocytic lymphohistiocytosis. Modified from Grom, A. A. et al. Arthritis Rheumatol. 68, 218–228 (2016), © 2016 American College of Rheumatology, with permission.

1. Grom, A. A. *et al.* Rate and clinical presentation of macrophage activation syndrome in patients with systemic juvenile idiopathic arthritis treated with canakinumab. *Arthritis Rheumatol.* **68**, 218–228 (2016).
2. Henter, J. I. *et al.* HLH-2004: Diagnostic and therapeutic guidelines for hemophagocytic lymphohistiocytosis. *Pediatr. Blood Cancer* **48**, 124–131 (2007)
3. Ravelli, A. *et al.* Macrophage activation syndrome in patients with systemic juvenile idiopathic arthritis treated with tocilizumab [abstract A56]. *Arthritis Rheumatol.*. **66** (Suppl. S3), S83–S84 (2014).
4. Ravelli, A. *et al.* Preliminary diagnostic guidelines for macrophage activation syndrome complicating systemic juvenile idiopathic arthritis. *J. Pediatr.* **146**, 598–604 (2005).