

INDICATION

Predicts the likelihood of a diagnosis of haemophagocytic lymphohistiocytosis (HLH)

ADDITIONAL INFORMATION

The best cutoff value in the literature was an HScore of 169. This demonstrated a sensitivity of 93% and specificity of 86%.

INTERPRETATION

SCORE	PROBABILITY OF HAEMOPHAGOCYCTIC SYNDROME
≤90	<1%
91-100	Around 1%
101-110	1-3%
111-120	3-5%
121-130	5-9%
131-140	9-16%
141-150	16-25%
151-160	25-40%
161-170	40-54%
171-180	54-70%
181-190	70-80%
191-200	80-88%
201-210	88-93%
211-220	93-96%
221-230	96-98%
231-240	98-99%
≥241	>99%

CALCULATION

Total score = sum of scores for each question

QUESTION		POINTS
Immunosuppression: HIV / Immunosuppressive therapy	Yes	+18
	No	+0
Temperature	<38.4°C / <101.1°F	+0
	38.4-39.4°C / 101.1-102.9°F	+33
	>39.4°C / >102.9°F	+49
Organomegaly	No	+0
	Hepatomegaly or splenomegaly	+23
	Hepatomegaly and splenomegaly	+38
Number of Cytopenias: Haem ≤92g/L or WBC ≤5 × 10 <sup>9</sup> /L or ≤110 × 10 <sup>9</sup> /L	1 lineage	+0
	2 lineages	+24
	3 lineages	+34
Ferritin (µg/L or ng/mL)	<2,000	+0
	2,000–6,000	+35
	>6,000	+50
Triglyceride	<1.5mmol/L / <132.7mg/dL	+0
	1.5-4mmol/L / 132.7-354mg/dL	+44
	>4mmol/L / >354mg/dL	+64
Fibrinogen	>2.5g/L / >250mg/dL	+0
	≤2.5g/L / ≤250mg/dL	+30
AST (U/L)	<30	+0
	≥30	+19
Haemophagocytosis features on bone marrow aspirate	Yes	+35
	No	+0