|  |  |
| --- | --- |
| **A.** Molecular diagnosis consistent with HLH | |
| **B.** Diagnostic criteria for HLH fulfilled (five of the eight criteria below) | * Fever * Splenomegaly * Peripheral blood Cytopenia affecting two or three lineages in the peripheral blood: * Hemoglobin < 90 g/L (in infants < 4 week: hemoglobin < 100 g/L) * Platelets < 100 × 109/L * Neutrophils < 1.0 × 109/L * Hypertriglyceridemia (fasting triglycerides ≥ 265 mg/dL) and/or hypofibrinogenemia (fibrinogen ≤1.5 g/L) * Hemophagocytosis in bone marrow, spleen, or lymph nodes * Low or no NK cell activity * Hyperferritinemia (ferritin ≥500 µg/L) * sCD25 (soluble IL-2 receptor) ≥2400 U/mL |

**Table 2:** HLH-2004 diagnostic guidelines [1]. The diagnosis of HLH can be established if criteria A or B is fulfilled.