Hemophagocytic syndrome

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Hemophagocytic syndrome. ORPHA:158032

Hemophagocytic syndrome (HPS) is a rare immune disease (see this term) and a potentially life-threatening disorder characterized by cytokine storm and overwhelming inflammation causing fever, hepatosplenomegaly, cytopenia, hypertriglyceridemia, hyperferritinemia, and hemophagocytosis in bone marrow, liver, spleen or lymph nodes. It can be either primary due to a genetic defect (primary hemophagocytic lymphohistiocytosis ; see this term), or secondary to malignancies, to infections, most commonly with viruses such as Epstein-Barr virus or cytomegalovirus, human immunodeficiency virus, or to autoimmune disorders such as systemic lupus erythematosus or adult-onset Still disease (secondary hemophagocytic lymphohistiocytosis) (see these termes).