

Hemophagocytic Lymphohistiocytosis (HLH)

Description

Hemophagocytic lymphohistiocytosis (HLH) is a severe, life-threatening systemic disease characterized by hyperinflammation, fever, organ damage, hypercoagulability, hepatosplenomegaly, and cytopenia. Macrophage activation syndrome and cytokine release syndrome are related disorders. Both conditions are caused by an excessive release of pro-inflammatory cytokines. It usually occurs in association with other diseases (lymphomas, infections, autoimmune diseases, etc.), but sometimes manifests spontaneously. Congenital forms have also been described.

Further detailed information can be found here:

<https://www.onkopedia.com/de/onkopedia/guidelines/haemophagozytische-lymphohistiozytose-hlh/@@guideline/html/index.html>

Diagnostics

- Fever
- Hepatosplenomegaly
- Cytopenia affecting 2 or more cell lines
- Hypertriglyceridemia
- Hypofibrinogenemia
- Severely elevated ferritin levels
- Hemophagocytosis in bone marrow, cerebrospinal fluid (CSF), or lymph nodes

H-Score

The probability of HLH can be calculated using the H-score:

<https://www.mdcalc.com/calc/10089/hscore-reactive-hemophagocytic-syndrome>

Therapy

Therapy is based on high-dose corticosteroids and intravenous immunoglobulin concentrates. Cytokine antagonists (anakinra, tocilizumab) or additional immunosuppression (etoposide, methotrexate, cyclosporine, etc.) are administered depending on the underlying cause. Ruxolitinib, rituximab, or Campath are utilized as second-line therapeutic agents. Intensive care treatment is frequently necessary and should be initiated early.

References

Thomas L, Labor und Diagnose, 2023, Release 5: <https://www.labor-und-diagnose.de/index.html>

Parameterkatalog des Klinischen Instituts für Labormedizin, Med.Univ.Wien und AKH Wien:

<https://www.akhwien.at/default.aspx?pid=3982>

Leistungsverzeichnis der Klinischen Chemie, Univ.Klinikum Ulm:

<https://www.uniklinik-ulm.de/zentrale-einrichtung-klinische-chemie/leistungsverzeichnis.html>

Weitere ausführliche Informationen finden sich hier:

<https://www.onkopedia.com/de/onkopedia/guidelines/haemophagozytische-lymphohistiozytose-hlh/@@guideline/html/index.html>