	The diagnosis of HLH is established by fulfilling one of the following 2 criteria
1	A molecular diagnosis consistent with HLH (e.g. PRF mutations. SAP mutations) OR
2	Having 5 of the following 8 features
A	Fever
B	Splenomegaly
C	Cytopenia affecting ≥2 cell lines (Hb <9 gm/dL, platelet count <1 lac/µl, neutrophils <1000/µl)
D	Hypertriglyceridemia ≥265 mg/dL or hypofibrinogenemia ≤150 mg/dL
E	Hemophagocytosis on bone marrow, spleen, or lymph node without evidence of malignancy
F	Low or absent natural killer cell cytotoxicity
G	Hyperferritinemia (≥500 ng/mL)
H	Elevated soluble CD25 (IL-2Rα chain, ≥2400 U/mL)

ACADEMIC P.E.A.R.L.S

Pediatric Evidence And Research Learning Snippet

HLH (Hemophagocytic Lymphohistiocytosis) in PICU

Hemophagocytic Lymphohistiocytosis in a PICU of a Developing Economy: Clinical Profile, Intensive Care Needs, Outcome, and Predictors of Mortality. Pediatr Crit Care Med. 2020 Oct 8.

Setting:
Retrospective case series in PICU of a tertiary care teaching hospital in North India.

Patients:
Children 2 months to 12 years old with the diagnosis of HLH admitted to PICU from Jan 2012 to April 2019.

Results:
•62 children with HLH (60 secondary and 2 primary).
•Aetiology: Infections (82.3%): scrub typhus (29%), dengue (17.7%), bacterial sepsis (14.5%), enteric fever (6.5%), and other infections (14.5%). Systemic-onset juvenile idiopathic arthritis (9.7%), malignancy (4.8%)
•Treatment: Steroids (77.4%), IV immunoglobulin (25.8%), mechanical ventilation (74.2%); vasoactive drugs (71%), and RRT (24.2%).
•Complications: Shock (71%), AKI (66.1%), ARDS (41.9%), DIC (54.8%), CNS dysfunction (54.8%), MODS (82.3%), and HCAI (Health care associated infection) (14.5%).
•PICU stay 5 days (2.5-9.5 d). Mortality 59.7%.

Conclusion:
HLH in PICU is commonly secondary to tropical infections & is associated with high mortality. Treatment of underlying infection and a less intense immunosuppressive therapy (steroids ± IV immunoglobulin) are suggested options.

EXPERT COMMENT

•In PICU, high index of suspicion for diagnosing HLH is required in children with prolonged fever, cytopenias, organomegaly, and new or progressive organ dysfunction, not responding to conventional treatment.
•Tropical infections are common etiology.
•Treatment of infection & less intense immunosuppression usually suffice.

Dr Suresh Kumar Angurana, MD, DNB, IDPCCM, FIMSA, FCCP, DM
Pediatric Critical Care
Assistant Professor, Division of Pediatric Critical Care,
Department of Pediatrics,
APC, PGIMER, Chandigarh

DR MANINDER S DHALIWAL
Editor – Academic Pearls
pedpearls@gmail.com

Reference

Parajuli B, Angurana SK, Awasthi P, et al Hemophagocytic lymphohistiocytosis in a PICU of a Developing Economy: Clinical Profile, Intensive Care Needs, Outcome, and Predictors of Mortality. Pediatr Crit Care Med. 2020 Oct 8. doi: 10.1097/PCC.0000000000002539.