Outcome analysis of pediatric hemophagocytic lymphohistiocytosis.

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Objectives: To identify clinical parameters related to mortality and establish prognostic factors correlated with unfavorable outcome in high risk patients whose treatment may fail.
Design: Retrospective review of electronic medical records.
Setting: Single tertiary medical centre in Taiwan.
Patients: Children below 18 years old with the confirmed diagnosis of HLH (as per HLH 2004 criteria) admitted from January 2004 to December 2018

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

Pediatric Evidence And Research Learning Snippet

Hemophagocytic Lymphohistiocytosis (HLH) in Paediatrics

Results:
•51 children identified by HLH-2004 criteria.
•Median age at diagnosis: 7 years.
•**Etiology:** Infections (27), MAS (12), Malignancy (8), Idiopathic (4)
•Infections – EBV (14), Influenza (3), Adenovirus (3), one each of CMV, Parvo, Parainfluenza, Scrub typhus, Mycoplasma, Pseudomonas, & E. coli.
•MAS - SLE (7), JIA (2), Uncertain autoimmune disease (3).
•Malignancy – ALL, Lymphoma, JMML, Aplastic anemia & Renal sarcoma.
•**Treatment:** Standard chemotherapy as per HLH-2004 protocol + IVIG in those with infections, Methylprednisolone pulse therapy in MAS group.
•**PICU stay** 6 ± 20.8 days
•**Mortality** 21.5% (40 survived, 11 died).
•**Predictors of outcome:** Longer aPTT, lower sodium concentration and higher serum creatinine and AST were significantly related to higher mortality. **On multivariate Cox regression analysis, longer aPTT (> 44.35 secs) was an independent risk factor for mortality.** Other poor outcome predictors were need for CRRT and mechanical ventilation.

Conclusion:
•HLH is not an uncommon complication in paediatric patients.
•The most common etiology is viral infections followed by immunological disorders and malignancies.
•Treatment is mostly by IVIG, steroids, chemotherapeutic agents (as per HLH-2004 protocol) and definitive treatment of the underlying cause.
•Outcomes are dependent on the initial presence of a prolonged aPTT, hyponatraemia, raised serum creatinine and AST, need for CRRT and mechanical ventilation.

EXPERT COMMENT

"Paediatricians must have a high index of suspicion for HLH esp. if patients have an unusual presentation with multisystem involvement. Most common aetiology is infections followed by autoimmune diseases and malignancies. A high aPTT(>44.35 secs) at admission is an independent predictor of mortality. Clinicians must remember the importance of testing for liver and renal dysfunction."

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With warm regards,

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Reference
Chen TY, Hsu MH, Kuo HC, Sheen JM, Cheng MC, Lin YJ. Outcome analysis of pediatric hemophagocytic lymphohistiocytosis. J Formos Med Assoc. 2021 Jan;120(1 Pt 1):172-179. doi: 10.1016/j.jfma.2020.03.025.