Advances in the diagnosis and treatment of hemophagocytic lymphohistiocytosis

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Hemophagocytic lymphohistiocysis (HLH) comprises a group of inherited ("primary") and non-inherited ("secondary") disorders of the immune system characterized by the dysregulated activation of T cells and macrophages that copiously secrete cytokines and mediate significant tissue damage. Historically, HLH has been challenging to diagnose due the similarity of its manifestations to those seen in other more common disorders. Similarly, HLH has proven challenging to cure with a large proportion of patients dying due to disease or the complications of its treatment. Thanks to the dedicated efforts of clinical and basic investigators working in the field, the last 25 years have witnessed tremendous advances in our understanding of normal immune cell function and the pathogenesis of HLH. Together, this knowledge has informed many novel and innovative approaches to HLH diagnosis and treatment. Several of these advances will be discussed in this presentation including: 1) development of flow cytometric methods for the rapid detection of primary HLH; 2) development of multi-agent chemo-immunotherapeutic, immune cell-depleting, and cytokine-targeting regimens for the treatment of HLH; and 3) implementation of reduced intensity conditioning approaches for use in hematopoietic stem cell transplantation.