EUROPEAN HEMATOLOGY ASSOCIATION



# *Treatment of hemophagocytic lymphohistiocytosis in adults*

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Hemophagocytic lymphohistiocytosis (HLH) has traditionally been considered a disease of childhood, typically with a genetic cause ('familial HLH'), that occurs extremely rarely in the adult population. However, over the last 15-20 years there has been increasing recognition that HLH does occur in adults and almost always as *secondary HLH*; and as physicians become more aware of the diagnosis there is evidence that it is much less rare than originally assumed.[1] According to a large review on 2197 adult patients with HLH, the most common associated diseases are infections and neoplasms, with autoimmune diseases being the third most common.[1] Diagnosis and treatment of adult HLH has continued to be based almost entirely on the patterns defined in the pediatric population, that are focused on familial HLH. As more studies have been performed, there is growing recognition that the algorithms for diagnosis and treatment in children need to be modified for application to the adult population.[2-6]

Diagnostic criteria for HLH are somewhat more difficult to interpret in adults since many of the findings that are part of the diagnosis (fever, cytopenias, hyperferritinemia) are relatively non-specific, and some features may have different diagnostic implications in adult patients.[7-9] Treatment paradigms also need to be re-evaluated in the adult setting. Since about half of adult HLH occurs in the setting of lymphoma,[1-6] treatment regimens may need to be tailored to therapy of the inciting malignancy, but the adjustments in therapy targeting HLH per se remain controversial.[10] For patients with infection-associated and idiopathic adult HLH, pediatric dosing of HLH therapy may not be suitable for the treatment of older patients.[11] In the end, the role of stem cell transplantation in adult HLH requires evaluation in the light of the unique features differentiating adult from pediatric disease, the relative risk of transplant, and the nature of the disease trigger.[12]

## *References*

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*This review outlines the unique challenges of stem cell transplantation in adults with HLH.*