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Commentary

# Clinical Characteristics of Hemophagocytic Lymphohistiocytosis in Pregnancy

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## DESCRIPTION

Hemophagocytic Lymphohistiocytosis (HLH), often called hemophagocytic syndrome, is a form of hyperinflammatory reaction brought on by primary or secondary immunological diseases. Persistent fever, hepatosplenomegaly, and a decrease in the amount of blood cells are the main symptoms of HLH. There are two types of HLH: hereditary (primary) and acquired (secondary). Primary HLH is more common in children. It is the result of a genetic mutation. In most adult HLH instances, secondary HLH occurs as a result of infection, autoimmune disorders, or malignant tumours. Because HLH is rapidly lethal, with mortality rates ranging from 26.5% to 74.8%, early detection and diagnosis of suspected HLH cases are critical [1].

Secondary HLH was most likely due to the pregnant patients' age. Hemolysis, increased liver enzymes, low platelet count and acute fatty liver are signs of HLH, which are comparable to those of obstetric problems. Furthermore, detecting HLH in pregnancy is complicated due to various causes and related factors that cooperate to create HLH [2]. Clinical therapy of HLH appears to be uneven across published instances, and the impact of drugs on the fetus during pregnancy must be considered. As a result, there is currently no agreement on how to treat HLH during pregnancy.

The most prevalent period of beginning for HLH in pregnancy in our study was in the second trimester, followed by the third trimester, which is consistent with earlier research. Our theory is that pregnancy is a regulatory immunological state in which immunologic changes with increasing pregnancy hinder pathogen clearance, resulting in an increase in the incidence of disease caused by some pathogens [3].

HLH during pregnancy is not currently covered by any guidelines. The majority of treatment options are based on clinical experience and clinical symptoms. In general, immunosuppressive medications and cytotoxic treatments are used to control life-threatening inflammatory reactions, while the underlying cause is treated, or both. HLH-1994 and HLH-2004 are the most extensively utilized standard treatment systems nowadays. The use of medications must be carefully considered. The US Food and Drug Administration classify corticosteroids as category C medications because they reduce immune system activity and suppress the inflammatory response. They are used in the HLH-1994 and HLH-2004 regimens. Women, who take corticosteroids during pregnancy, especially after the first trimester, have a minimal risk of birth abnormalities. Corticosteroids are the primary line of treatment for most individuals with HLH during pregnancy, regardless of the precipitating cause. However, the causes and related factors must be recognized and treated appropriately [4]. Patients with unsuccessful corticosteroid treatment may benefit from etoposide and pregnancy termination.

Etoposide may be administered as soon as possible after delivery to enhance the prognosis of patients, especially those who are severely ill. In prior studies, corticosteroids were used as the first line of treatment for 38 (95%, 38/40) women, with 12 (31.6%, 12/38) showing a curative benefit.

Etoposide, a Topo II inhibitor, is an important component of the HLH-94 and HLH-04 regimens. Adult hemophagocytic syndrome prognostic variables showed that using etoposide as the first-line treatment was related with a better result. Etoposide is an FDA-designated category D anticancer medication that targets the cell cycle [5]. Etoposide is a drug that is often used to treat ovarian cancer, and it is safe for the fetus if given during the second or third trimester.

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