

## **Abstract**

Hemophagocytic Lymphohistiocytosis Syndrome is a rare and fatal disease with uncontrolled activation of the immune system resulting in multiorgan dysfunction and high mortality rate. It can be either primary due to autosomal recessive inheritance with perforin gene mutation or secondary due to infections, malignancy, drugs or autoimmune diseases. Systemic Lupus Erythematosus is a common autoimmune disease in females of reproductive age group with multiorgan involvement and a considerable risk to pregnancy and fetal outcome when flares up during pregnancy. The occurrence of HLH in a SLE patient is called Acute Lupus Hemophagocytic Syndrome; thus is a rarer entity with limited case reports and in pregnancy is a challenging diagnosis and management to both the Physician and the Obstetrician.