Hemophagocytic lymphohistiocytosis (HLH) is a rare, life-threatening condition caused by an overactive, abnormal response of the immune system. The immune system is the body's natural defense system against foreign or invading organisms or substances. The immune system is a complex network of cells, tissues, organs, and proteins that work together to keep the body healthy. In hemophagocytic lymphohistiocytosis, the immune system responds to a stimulus or 'trigger', often an infection, but the response is ineffective and abnormal. This ineffective, abnormal response, causes a variety of signs and symptoms, which, if not treated, can potentially become life-threatening. Some affected individuals may have a genetic predisposition to developing hemophagocytic lymphohistiocytosis. This is known as the primary or familial form. In other individuals, the disorder occurs sporadically usually when there is an underlying predisposing condition or disorder. This is known as the secondary form. The secondary forms are more common than the familial forms. Hemophagocytic lymphohistiocytosis most often affects infants from birth to 18 months, but can affect individuals of any age. Early diagnosis and prompt treatment is essential. Hemophagocytic lymphohistiocytosis most often affects infants or young children, but can affect individuals of any age. It affects boys and girls in equal numbers. In adults, it affects men slightly more often than women. The exact incidence and prevalence is unknown. Rare disorders often go misdiagnosed or undiagnosed making it difficult to determine the true frequency in the general population. About 25% of the people with this disorder, have the familial form.