X-linked lymphoproliferative (XLP) syndrome is an extremely rare inherited (primary) immunodeficiency disorder characterized by a defective immune system that is powerfully responsive to infection with the Epstein-Barr virus (EBV). This virus is common among the general population and is relatively well-known because it is the cause of infectious mononucleosis (IM), usually with no long-lasting effects. However, in individuals with XLP, exposure to EBV may result in severe, life-threatening fulminant hepatitis; abnormally low levels of antibodies in the blood and body secretions (hypogammaglobulinemia), resulting in increased susceptibility to various infections; malignancies of certain types of lymphoid tissue (B-cell lymphomas); and/or other abnormalities. The range of symptoms and findings associated with XLP may vary considerably from case to case. In addition, the range of effects may change in an affected individual over time. In most cases, individuals with XLP experience an onset of symptoms anytime from ages about 6 months to 10 years of age. X-linked lymphoproliferative syndrome is an extremely rare disorder that is usually fully expressed in males only. About 400± cases affecting males in more than 80 multigenerational families (kindreds) from several different countries have been reported in the medical literature. Researchers estimate that approximately one to two in every one million males are affected by XLP.