Hers disease is a genetic metabolic disorder caused by a deficiency of the enzyme, liver phosphorylase. This enzyme is necessary to break down (metabolize) glycogen, a carbohydrate that is stored in the liver and muscle and used for energy. Deficiency of this enzyme results in the abnormal accumulation of glycogen in the body. Hers disease is one of a group of disorders known as the glycogen storage disorders. It is characterized by enlargement of the liver (hepatomegaly), moderately low blood sugar (hypoglycemia), elevated levels of acetone and other ketone bodies in the blood (ketosis), and moderate growth retardation. Symptoms are not always evident during childhood, and children are usually able to lead normal lives. The incidence of glycogen storage diseases is estimated to be between 1 in 20,000 and 1 in 25,000 persons in the United States. The incidence of Hers disease is unknown, although the disorder has a higher prevalence in the Mennonite population. Symptoms of Hers disease are usually not noticed until adulthood although the disorder may present in childhood. A diagnosis of Hers disease is based on a test for activity of the liver phosphorylase enzyme. A small fragment of liver tissue is surgically removed (biopsy) and assayed for the activity of the enzyme. In persons with Hers disease, this enzyme activity will be reduced.