Glycogen Storage Disease I – Von Gierke

Other names for this condition are:
- GSD
- Glycogenosis Type I
- Von Gierke Disease

This condition belongs to a group of disorders known as Glycogen Storage Diseases (GSDs). These disorders are characterised by the inability of the body to use glycogen which is the storage system of energy and glucose for the body. Those affected can develop low blood sugar levels (hypoglycaemia) quickly and have a low tolerance of illness and fasting. Glycogen Storage Diseases are caused by a defect in an enzyme which catalyses glycogen compounds into simple sugars known as monosaccharides. This enzyme deficiency causes an accumulation of glycogen in the tissues. Type 1 can be divided into the two major types, 1a and 1b. In type 1a there is a defect of the enzyme glucose-6-phosphatase in the liver, kidney and intestines which causes an accumulation of glycogen in these organs. Type 1b is caused by a defect in the transport of Glucose-6-Phosphate which causes bacterial infections and ulcers in the intestines due to a decrease in the number of a type of white blood cell known as neutrophils (neutropenia).

Treatment aims to maintain consistent blood sugar levels and prevent a high level of ketones in the body. Cornstarch is often introduced at 4-6 months in small amounts. Significant amounts are not given until they are over 1 year of age. There is no specific age on when cornstarch should be given and this age may vary depending on the individual. For further information regarding treatment please see full summary.

This information is fully sourced and referenced, for more detailed information and references please contact CLIMB by email, letter or telephone.

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