Hereditary fructose intolerance

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Hereditary fructose intolerance. ORPHA:469

Hereditary fructose intolerance (HFI) is an autosomal recessive disorder of fructose metabolism (see this term), resulting from a deficiency of hepatic fructose-1-phosphate aldolase activity and leading to gastrointestinal disorders and postprandial hypoglycemia following fructose ingestion. HFI is a benign condition when treated, but it is life-threatening and potentially fatal if left untreated.