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# Management of Hypoglycemia in Von Gierke's Disease

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**Abstract :** Introduction:Glycogen Storage Disease Type-1 (GSD-1) is a rare phenomenon primarily affecting the liver and kidney. Excessive accumulation of glycogen and fat in liver, kidney, and intestinal mucosa is noted in patients with deficiency of Glucose-6-phosphatase deficiency. Patients with GSD-1 have a wide spectrum of symptoms, including hepatomegaly, hypoglycemia, lactic acidemia, hyperlipidemia, hyperuricemia, and growth retardation. Age of onset, rate of disease progression and its severity is variable in this disease.Case:An 18-year-old male with GSD-1a, Von Gierke's disease, hyperuricemia, and hypertension presented to the hospital with nausea and vomiting. The patient followed an hourly cornstarch regimen during the day and overnight through infusion via a PEG tube. The complaints started at work, where he was unable to tolerate oral cornstarch. He washemodynamically stable on arrival. ABG showed pH 7.372, PaCO2 30.3, and PaO2 92.2. WBC 16.80, K+ 5.8, HCO3 13, BUN 28, Cr 2.2, Glucose 60, AST 115, ALT 128, Cholesterol 352, Triglycerides >1000, Uric Acid 10.6, Lactic Acid 11.8 which trended down to 8.0. CT abdomen showed hepatomegaly and fatty infiltration with the PEG tube in place.He was admitted to the ICU and started on D5NS for hypoglycemia and lactic acidosis. Per request by the patient's pediatrician, he was transitioned to IV D10/0.45NS at 110mL/Hr to maintain blood glucose above 75 mg/L. Frequent accuchecks were done till he could tolerate his dietary regimen with cornstarch. Lactic acid downtrend to 2.9, and accuchecks ranged between 100-110. Cr improved to 1.3, and his home medications (Allopurinol and Lisinopril) were resumed. He was discharged in stable condition with plans for further genetic therapy work up.Discussion:Mainstay therapy for Von Gierke's Disease is the prevention of metabolic derangements for which dietary and lifestyle changes are recommended. A low fructose and sucrose diet is recommended by limiting the intake of galactose and lactose to one serving per day. Hypoglycemia treatment in such patients is two-fold, utilizing both quick and stable release sources. Cornstarch has been one such therapy since the 1980s; its slow digestion provides a steady release of glucose over a longer period of time as compared with other sources of carbohydrates. Dosing guidelines vary from age to age and person to person, but it is highly recommended to check BG levels frequently to maintain a BG > 70 mg/dL. Associated high levels of triglycerides and cholesterol can be treated with statins, fibrates, etc. Conclusion:The management of hypoglycemia in GSD 1 disease presents various obstacles which could prove to be fatal. Due to the deficiency of G6P, treatment with a specialized hypoglycemic regimen is warranted. A D10 ½ NS infusion can be used to maintain blood sugar levels as well as correct metabolic or lactate imbalances. Infusion should be gradually weaned off after the patient can tolerate oral feeds as this can help prevent the risk of hypoglycemia and other derangements. Further research is needed in regards to these patients for more sustainable regimens.

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