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USE OF CONTINUOUS GLUCOSE MONITORING IN GLYCOGEN STORAGE DISEASES

Authors: Paulina Bartušytė[1], Vijonė Adomaitytė[1,]
Scientific research supervisor: Rimantė Čerkauskienė[2]

1- Vilnius University, Faculty Of Medicine, Vilnius, Lithuania, 2- Center for Coordination Of Rare Diseases, Vilnius University Hospital Santaros Klinikos, Vilnius, Lithuania

Introduction. Fanconi-Bickel syndrome (FBS) is a rare autosomal recessive genetic disorder linked to a faulty glucose transporter 2 (GLUT2) due to SLC2A2 gene mutations. FBS manifests with metabolic challenges like acidosis, fasting hypoglycemia, postprandial hyperglycemia and growth retardation. Yet, there's a lack of data on effective nutritional treatments, underscoring the need for improved clinical assessments.

Objectives. We aimed to evaluate the efficacy of a diet with extended release waxy-maize cornstarch (ERWMC) and continuous glucose monitoring (CGM) on metabolic response, biochemical changes and growth in FBS patients over time.

Materials and methods. We report three FBS cases and their tests findings in a 2 year time period to compare them before monitoring and during it. Patients were treated in Vilnius University Hospital Santaros Klinikos with uncooked corn starch (UCCS) or ERWMC (Glycosade®), carbohydrate-restricted diet (CRD) and applied CGM system using a smartphone.

Results. **Patient 1** 19-year-old female with FBS diagnosed at 10 months. Managed with CRD from 2 years age and UCCS from 4 years age that helped maintain slow but steady growth. Glycemic data insufficient for evaluation. Cholesterol increased with fluctuations (6.41->7.4- >9.04->7.59), and triglycerides initially rose then decreased to 3.26. **Patient 2** 6-years-old female. With FBS diagnosed at 3 months age. CRD and UCCS was prescribed at 1 year 9 months of age, but parents did not comply with prescribed regime and diet. The glucose management with CGM was introduced at 4 years age, but discontinued after a few weeks. Noticeable growth retardation (height growth from 2 cm below 3rd percentile of height by age at 3 months to18 cm below 3rd percentile of height by age at 4 years 9 months). Fasting glucose before monitoring was below the lowest normal limit but later increased over the highest limit(1.9->8.4->6.6->5.6). Cholesterol, triglycerides have prominently increased over a 2 year period. (3.19->6.97->9.32 and 1.61->11.99->8.8). **Patient 3** 4-year-old female, diagnosed with FBS at 1.5 months. Monitored with CRD and UCCS from 8 months age, switched to ERWMC at 20 months age, and started CGM at 2 years 2 months. This led to adequate growth (height growth from 1 cm below 3rd percentile of height by age at 1.5 months to a 3rd percentile of height by age at 4,5 years). Glucose levels fluctuated widely (4.1->8.8->5.3->3.9), cholesterol increased slightly above normal (3.14->3.07->5.31->5.82), and triglycerides was 2.8 times lower than the lowest limit before the monitoring and it has increased during.

Conclusions. In our observation, using a CGM system can benefit FBS patients by helping determine ERWMC doses, making informed dietary choices, and maintaining stable blood sugar levels. This fosters positive metabolic outcomes and supports healthy growth.

Keywords. Continuous glucose monitoring, Fanconi-Bickel syndrome, Glycogen storage diseases, Growth retardation.