P-010 - A CLINICAL TRIAL EVALUATING THE USE OF UNCOOKED BRAZILIAN SWEET POLVILHO VERSUS UNCOOKED CORNSTARCH IN GSD IA: PRELIMINARY RESULTS

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INTRODUCTION: Hepatic Glycogen Storage Diseases (GSD) are a group of genetic diseases characterized by fasting intolerance associated hypoglycemia. The most commonly used treatment strategy is the frequent administration of uncooked cornstarch (UCCS). Although this treatment is successful, the adherence rate is not optimal and the use of large amounts of UCCS can lead to overweight and to a decrease in patients and caregivers quality of life, due to the need of taking the starch during the night. In-vitro studies by our group suggested a longer glucose release from uncooked sweet polvilho (SP), a product extracted from cassava, than from UCCS. OBJECTIVE: To evaluate the short-term efficacy and safety of the use of SP in adult GSD Ia patients. METHODS: A randomized, double-blinded, phase I/II crossover study is being conducted, comparing the use of UCCS and SP (100 grams of starch in 200 ml of water). The inclusion criteria are: age ≥16 years, being treated with UCCS and having a genetic diagnosis of GSD Ia. Ten patients were assigned to participate in two sequential overnight sessions. Each individual stays two nights hospitalized: one night using UCCS and one night using SP. After ingestion of each randomized starch, subjects are followed during the night and procedures are stopped after 10 hours of fasting or anytime if patient presents hypoglycemia (plasma concentrations <70 mg/dL). From an intravenous catheter, blood samples are taken every hour for glucose, lactate and insulin concentrations. RESULTS: To date, four individuals [three male (21 to 26 years old) and one female (21 years old)] had finished two starch load procedures. Mean body mass index of participants was 29.53 (25.2 to 37.88). The SP kept the normoglycemia for a period at least equal to the UCCS. No distinction was made between the two randomized products regarding palatability. One subject presented severe hyperlactatemia (≥5.0mmol/l) during SP load procedure. No patient presented serious adverse events. DISCUSSION AND CONCLUSION: Preliminary clinical data showed herein suggest this product may be a good alternative for treatment of GSD. Other six individuals will be evaluated until the completion of this protocol and accomplishment of statistical analysis.