UNCOOKED SWEET MANIOC STARCH: A CANDIDATE FOR THE TREATMENT OF GLYCOGEN STORAGE DISEASE Ia?

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**Introduction**

Glycogen storage disease type 1a (GSD Ia) is characterized by severe fasting hypoglycemia. The clinical management includes the administration of uncooked cornstarch (UCCS). Although such a diet approach is effective in achieving euglycemia, its impact on the quality of life of patients should be considered. In vitro analyses suggest a longer release of glucose when using sweet manioc starch (SMS).

**Methods**

We compared the efficacy and safety of the administration of SMS and UCCS during a short-fasting challenge in patients with GSD Ia in a randomized, triple-blind, phase I/II, cross-over study. GSD Ia patients aged ≥16 years and treated with UCCS were enrolled. Participants were hospitalized for two consecutive nights, receiving UCCS or SMS in each night. After the administration of the starches, glucose, lactate and insulin levels were measured in 1-h interval throughout the hospitalization period. The procedures were interrupted after 10 h of fasting or in a hypoglycemic episode (<3.88 mmol/L).

**Results**

Eleven individuals (mean age: 21.6±4.3 years; all presenting body mass index >25 kg/m\(^2\)) participated in the study. The average fasting period was 8.2±2.0 h for SMS and 7.7±2.3 h for UCCS (p=0.04). SMS maintained euglycemia for a greater period over UCCS (Figure 1A).

Increased lactate concentrations were detected even in absence of hypoglycemia, not being influenced by the different starches investigated (p=0.17) (Figure 1B).

**Conclusions**

SMS appears to be non-inferior to UCCS in the maintenance of euglycemia, thus emerging as a promising alternative to the treatment of GSD Ia.

**References**
