RESULTS:  
- Dosing for overnight Glycosade is comparable to total uncooked cornstarch for the same period. Median dose in adults was 135 grams (range 120-150 grams).
- 58 of 69 patients (84.1%) GSD type Ia experienced improved short term metabolic control and increased duration of fasting on a Glycosade challenge.
- The success rate at extending fasting was 95% for females but 71% for males. No factors could be identified to predict who would benefit from the extended release preparation.
- All markers of long term metabolic control remained stable on the new therapy.
- No episodes of severe hypoglycemia or hospitalizations have been required for subjects treated with the new therapy.
- 1 patient developed rapidly growing adenomas on Glycosade therapy, but it is unclear if there is an association with the therapy.

INTRODUCTION:  
Glycogen storage disease (GSD) type I is caused by deficiency of glucose-6-phosphatase resulting in severe hypoglycemia, hepatomegaly, hyperlactatemia, and hyperlipidemia. Cornstarch has been the main treatment for GSD since 1982. In 2012, a new extended-release cornstarch was released, but there have been no studies on the long-term efficacy and safety of this product.

HYPOTHESIS:  
A new modified starch, Glycosade® (Vitaflo® (Int’l) Ltd.), will allow patients with GSD to have longer uninterrupted sleep and improve quality of life without sacrificing metabolic control.

METHODS:  
A prospective cohort study was performed using all patients that tried overnight Glycosade at University of Florida. A total of 69 subjects with GSD Ia (31 M, 38 F; average age 18.0 years, age range 5-59 years) attempted fasting with Glycosade between 2012 and 2013. For inclusion in follow-up studies, one nighttime dosage of Glycosade extended-release cornstarch needed to be consumed at least 3 nights per week for at least 3 months. Long-term laboratory data are available for 27 of the patients who have transitioned to Glycosade.

CONCLUSION:  
Glycosade has allowed patients with glycogen storage disease type Ia to maintain normoglycemia for an extended period of time and improved quality of life without sacrificing metabolic control.