



Comparison of a novel starch Glycosade vs Uncooked Cornstarch on duration of euglycaemia in two children with Glycogen Storage Disease Type 1

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Introduction

Glycogen storage disease Type 1 (GSD 1) is caused by either deficiency of glucose-6-phosphatase or of the glucose-6-phosphate transport system. Glucose production is hence inadequate and hypoglycaemia will occur after short periods of fasting. Clinical consequences include growth retardation, hepatomegaly, as well as long term complications like renal disease, hepatic tumors, osteopenia and polycystic ovaries in females. The aim of dietary treatment is thus to avoid hypoglycaemia and suppress secondary metabolic decompensation. Traditionally, this is achieved through the use of either uncooked cornstarch (UCCS) or continuous overnight tube feeding. UCCS is currently the preferred option for parents as it is easily obtained and economical.

GSD 1 is relatively rare, with an estimated world-wide incidence of 1 in 100,000. In KKH, from year 2000 to date, there are only 4 patients with this condition.

Aims

The objective of this study was to compare a novel starch Glycosade versus UCCS in maintaining euglycaemia in children with GSD 1. Although the incidence of GSD 1 is low and UCCS is easily obtained and economical, the side effects of UCCS include loose stools, flatulence and bowel distension, which could hamper compliance to therapy. Moreover, the reported duration of euglycaemia varies, thus requiring four to six hourly feeds, both day and night. This not only inconveniences both the caregivers and the child, at worst, it could mean hypoglycaemia in the child who is unable to be roused during the night for a dose of UCCS.

Glycosade, a novel starch, has been reported to increase the duration of euglycaemia. This is potentially useful as both caregivers and the child can enjoy a restful night. In addition, it also reportedly causes fewer gastrointestinal side effects, thus potentially increasing compliance to therapy. However, it is costly, costing \$416 per box of 30 sachets (approximately \$14 per sachet vs \$0.40 for an equivalent amount of UCCS). Hence, it was necessary to evaluate the product to determine if the convenience and reduced side effects justified the additional costs.

Method

Of the 4 children with GSD 1, only 2 children were involved in this study as the other 2 children were not compliant to UCCS therapy. Both children were admitted to KKH specifically for the study, and informed consent was taken from parents.

Both children were given a fasting starch load test consisting of 2g per kg body weight of starch over two consecutive nights (UCCS the 1st night and Glycosade the 2nd night). Their meals and snacks during the day were provided by the hospital's catering department, and kept consistent over the two days of testing. Two hours after dinner or supper, depending on the child's usual meal pattern, the baseline blood glucose level (BGL) was measured. The UCCS or Glycosade was then given, and BGLs measured every hour until it reached 4 mmol/L, and then every half hour until it reached 3 mmol/L. The test was stopped 10 hours after consumption of starch, or when BGL reached 3 mmol/L, whichever occurred sooner.

Results

The patients' characteristics and results of the starch load test are as shown in Tables 1 & 2 below. Glycosade did not increase the duration of euglycemia in either child.

Table 1 : Characteristics of patients 1 & 2

Patient characteristics	Patient 1	Patient 2
Demographics	5 yr old male, diagnosed at 8 mths of age	7 yr old male, diagnosed at 15 mths of age
Growth status	Wt : 3 %tile, Ht : <3 %tile	Wt : 75-90 %tile, Ht : 25-50 %tile
Usual UCCS dose	2g per kg every 4 - 7 hrly	1-1.6g per kg every 4 hrly
Symptoms of hypoglycemia	Yes, tired & sweaty	No

Table 2 : Results of starch load test

Starch Load Test (2g per kg)	Patient 1		Patient 2	
Type of Starch tested	UCCS	Glycosade	UCCS	Glycosade
Time from starch dose till BGL reached 4 mmol/L	6 hrs	6 hrs	7 hrs	6 hrs
Time from starch dose till BGL reached 3 mmol/L	7 hrs	6.5 hrs	8.5 hrs	6.5 hrs
GI intolerance	Nil reported	Nil reported	Nil reported	Nil reported

Discussion

Glycosade did not improve fasting tolerance in the study subjects with GSD 1. However, we cannot draw a strong conclusion from this study due to the small sample size.

However, performing the starch load test enabled us to optimize the UCCS dosage for both patients as follows : (i) Patient 1's parents were convinced to decrease the maximum time interval between UCCS doses to 6 hrs, thus relieving his symptoms of hypoglycemia (ii) Patient 2's parents were able to increase the amount of UCCS per dose, as well as the time interval between UCCS doses from 4 hrly to 6-7 hrly, which improved both parents and patient's quality of life as he did not require an early morning dose of UCCS, as well as a dose of UCCS his during school hours.

PATIENTS. AT THE HEART OF ALL WE DO.