

# Uncooked cornstarch—efficacy in type I glycogenosis

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## Abstract

**Uncooked cornstarch (UCCS) loads in 14 patients with type 1 glycogenosis revealed that satisfactory glycaemia was achieved for a median of 4.25 hours (range 2.5 to 6). Length of glycaemia was related weakly to UCCS dose, but not to patient age or measures of metabolic control. Careful monitoring is required during UCCS treatment.**

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For the past decade, uncooked cornstarch (UCCS) has been used to treat hypoglycaemia associated with type I glycogenosis (GSD-I).<sup>1 2</sup> It is hydrolysed by pancreatic  $\alpha$ -amylase to release glucose slowly and may maintain normoglycaemia for up to nine hours.<sup>2</sup> However, our impression is that UCCS has not been as effective in maintaining normoglycaemia in GSD-I patients. Consequently, we reviewed the results of UCCS loads performed in 14 GSD-I patients to assess efficacy and investigate factors that may influence response.

## Methods

A retrospective review was made of 14 patients with liver biopsy proven GSD-I who had UCCS loads performed at Great Ormond Street Hospital, London, between 1990 and 1994. At the time of the loads, all were on continuous overnight nasogastric feeds of glucose polymer and regular two hourly daytime feeding regimens. Before the loads, patients were started on small amounts of UCCS once

a day, the quantity was gradually increased to the full dose over four to six weeks to minimise side effects of bloating and diarrhoea.

Patients were admitted to hospital the evening before the load tests, an intravenous cannula was sited, and they were given their usual overnight feed. Within one hour after the end of the overnight feed, the dose of UCCS was given. An additional 10 g of carbohydrate from food or glucose polymer was given to prevent any hypoglycaemia associated with cessation of the night feed or the initial slow release of glucose from UCCS. Baseline plasma total cholesterol and triglyceride and blood glucose concentrations were measured before the UCCS load. Thereafter blood glucose was measured at hourly intervals until the test was terminated when this fell below 3.0 mmol/l.

The period of satisfactory glycaemia was examined in relation to the dose of UCCS per kg body weight and measures of metabolic control: height standard deviation score (SDS), plasma cholesterol, and triglyceride concentrations. Relations between variables were evaluated using Spearman's rank correlation coefficient ( $r$ ).

## Results

The results are shown in table 1. As a group they were short and had hyperlipidaemia with median height SDS of  $-1.41$ , plasma cholesterol 5.1 (range 2.6 to 7.9) mmol/l, and plasma triglycerides 5.0 (2.2 to 16.8) mmol/l. Blood glucose remained at 2.9 mmol/l or above for a median of 4.25 hours. There was a weak correlation between dose of UCCS and duration of normoglycaemia ( $r = +0.32$ ). There was no relation between length of time of satisfactory glycaemia and the patient's age ( $r = +0.02$ ),

Table 1 Details of 14 patients with GSD-I. Age, sex, height SDS, minimum carbohydrate (CHO) treatment before and after the introduction of UCCS, dose of UCCS given in load, and duration of euglycaemia

Patient No	Type	Age (years)	Sex	Ht SDS	CHO intake (g/kg/h)			UCCS (g/kg)	Length of euglycaemia (hours)
					Before: overnight	Before: daytime	After: daytime		
1	1b	4.4	M	-0.05	0.30	0.35	0.48	2.0	4.5
2	1a	5.5	F	-1.50	0.25	0.30	0.30	2.0	4
3	1a	5.6	F	-1.57	0.38	0.60	0.50	2.0	3
4	1b	5.7	F	-0.45	0.22	0.30	0.52	2.0	5.25
5	1a	6.8	M	-1.51	0.33	0.50	0.48	2.0	4.5
6	1a	8.8	M	+2.31	0.19	0.16	0.16	0.7	3
7	1a	9.3	F	-0.22	0.20	0.20	0.22	1.0	2.5
8	1a	10.5	M	-2.47	0.30	0.52	0.52	2.0	4.25
9	1a	10.6	F	+0.40	0.18	0.20	0.43	2.0	6
10	1a	11.6	F	-3.09	0.25	0.37	0.46	1.8	4.75
11	1b	11.7	M	-1.67	0.26	0.21	0.37	1.7	5.5
12	1b	13.0	M	-2.03	0.31	0.36	0.36	2.0	3
13	1a	15.3	M	+0.68	0.12	0.12	0.23	1.0	4
14	1a	16.6	M	-4.75	0.14	0.28	0.25	1.0	4

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nor between response to UCCS and height SDS ( $r = -0.07$ ) or plasma triglyceride concentration ( $r = -0.20$ ). There was a weak negative relation with plasma cholesterol ( $r = -0.42$ ).

### Discussion

Hypoglycaemia is the major biochemical consequence of deficient glucose-6-phosphatase activity in GSD-I. The aim of treatment is to prevent hypoglycaemia and ameliorate secondary lactic acidosis, hyperlipidaemia, and hyperuricaemia, as well as growth failure and hepatomegaly. This may be achieved with continuous enteral glucose polymer infusion at night and intermittent glucose polymer or UCCS in the daytime, or intermittent UCCS throughout the 24 hours at doses equivalent to hepatic glucose production.<sup>1 2</sup>

The glycaemic index<sup>3</sup> is used to compare blood glucose response following ingestion of various foodstuffs. Cornstarch is a long chain infrequently branching polymer of glucose with a high amylose/amylopectin ratio. Raw, it has a low glycaemic index and a long lasting glycaemic response compared to other complex carbohydrates.<sup>2</sup> It is slowly degraded within the intestine primarily by pancreatic  $\alpha$ -amylase: the amylose component to maltose and maltotriose; the amylopectin to  $\alpha$ -dextrins. Brush border enzymes control further hydrolysis to release glucose.<sup>4</sup>

In GSD-I, UCCS is introduced usually from 2 years of age onwards<sup>1</sup> because it is thought that insufficient pancreatic  $\alpha$ -amylase is produced before this age.<sup>5</sup> Satisfactory glycaemia has been reported to last from four to nine hours after ingestion.<sup>1 2</sup> Doses vary between 0.5 and 2.5 g/kg<sup>1</sup> and it may be given in water, milk, or yoghurt. It must not be cooked as this disrupts the starch granules, making them more readily hydrolysed and therefore absorbed earlier.

The results of UCCS loads in our patients were disappointing, with a median efficacy of 4.25 hours. Several factors could have influenced the observed results. Pancreatic  $\alpha$ -amylase activity may be impaired and intestinal absorption may be abnormal in these patients,<sup>6</sup> as glucose-6-phosphatase is normally expressed in the intestine, although its function

there is not clear. The integrity of the brush border enzymes and the glucose transporter may be abnormal. The method of administration of UCCS may also be important. Sometimes it was given mixed with squash, which itself could stimulate insulin secretion, or with milk, which may delay gastric emptying.

Although a weak correlation between dose of UCCS and duration of normoglycaemia was observed, this does not guarantee larger doses will extend normoglycaemia. In addition, larger UCCS doses may be associated with excessive weight gain and suppression of appetite. Currently it is recommended that the UCCS dose should be equivalent to hepatic glucose production, which decreases with age. Consequently smaller doses were given to older patients. Carbohydrate doses from UCCS were similar to previous intakes from glucose polymer (table 1). Some groups recommend that blood glucose concentrations should be maintained at 4 mmol/l.<sup>7</sup> If the tests had been terminated at this level, the median duration of the UCCS loads would have been only 3.5 hours.

In conclusion, UCCS seems less effective at maintaining normoglycaemia than previously described. This is important for GSD-I patients and may be pertinent for other disorders associated with hypoglycaemia treated with UCCS. We recommend close monitoring of glycaemic response when UCCS is prescribed. Further studies of digestion and absorption are necessary to optimise its glycaemic effect.

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