



Reduction in insulin response in a patient with GSD1 using a new modified starch: case report



Charle M Maritz, Alison Cousins, Elaine Murphy

Charles Dent Metabolic Unit, National Hospital for Neurology & Neurosurgery, London, UK

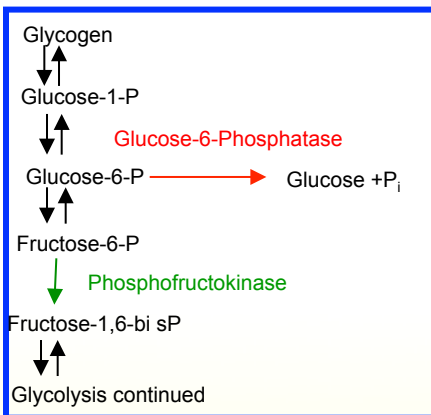
Introduction

Glycogen Storage Disease type 1a is caused by a deficiency of glucose-6-phosphatase resulting in the inadequate production of glucose. Hypoglycaemia will occur after relatively short fasting periods. The primary goal is to maintain normal blood glucose concentrations ($\geq 3\text{mmol/L}$) during the day and night. Management for adults consists of a combination of continuous overnight feeding and uncooked cornstarch (UCCS) during the daytime or UCCS throughout the 24-hour period.

Case Report

Our patient, a 22 year old female, had been diagnosed at 3 ½ months of age. Her metabolic control had never been good and she presented to hospital with recurrent hypoglycaemia and persistently elevated lactate levels. On admission, her blood glucose levels were only controlled by continuous pump feeding (20 hours) with two 2 hour breaks for meals. Prior to her hospital admission she had been taking 85g UCCS 4-5 times daily. The patient had a very limited fasting tolerance of <1.5 hours on a controlled fast. A starch load with 50g UCCS resulted in maximum insulin, lactate and glucose levels of 31.4mmol/L, 6.3mmol/L and 5.8mmol/L respectively. A starch load of 60g Glycosade (a new prescribable modified starch) resulted in maximum insulin, lactate and glucose levels of 7.6mmol/L, 8.1mmol/L and 4.7mmol/L respectively. On 60g of Glycosade she maintained adequate glycaemic control for 5 hours. She has been on 70g Glycosade three times per day since early in her admission and has not required daytime pump feeding. Despite considerable improvement in her daytime glycaemic control she still has considerable problems with eating, resulting in ongoing hypoglycaemia and is currently being considered for a liver transplant.

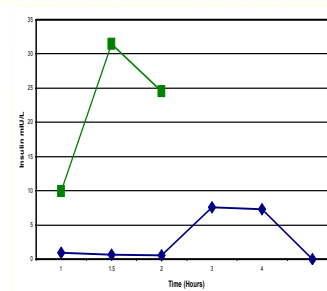
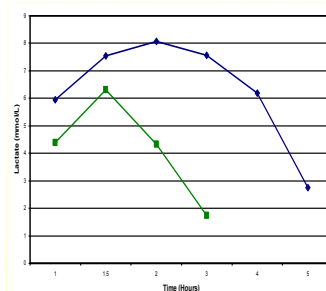
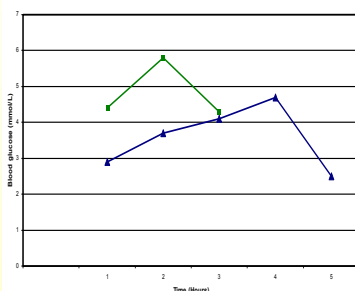
Results



Composition of Cornstarch compared to Glycosade

	UCCS	Glycosade
Moisture content (%)	10.9	11.9
Amylopectin (%)	72.8	99.5
Total carbohydrate, wet base (%)	87.5	82.5
Resistant starch (%)	60.5	67.7
Glycemic index (GI units)	70	30

Glycosade



--- Glycosade
--- UCCS

Conclusion

This case demonstrates longer duration of normoglycaemia with reduced insulin response in this female patient with Glycosade compared to UCCS. However ongoing eating problems necessitate the consideration for a liver transplant.

References