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Summary of two papers on the long-term safety and efficacy of Glycosade® in Glycogen Storage Disease (GSD)

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Innovation in Nutrition

A Nestlé Health Science Company

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Summary of two papers on the long-term safety and efficacy of Glycosade® in GSD

**Italicised text refers to additional data supplied by authors (K. Ross, 2016).*

INTRODUCTION: Glycogen storage disease (GSD) is a collection of disorders of inborn errors of carbohydrate metabolism. There are two main categories of GSD - hepatic GSDs and muscle GSDs . In hepatic GSDs, there is a life-threatening inability to maintain adequate levels of glucose in the blood. Uncooked cornstarch (UCCS) or cornflour is used by many patients with hepatic GSDs as a slow-release carbohydrate source but is associated with many limitations, most notably an inadequate duration of action (average = 4.25 hours. Lee et al, 1996) which is not long enough to prevent hypoglycaemia during a night's sleep. Exciting new long-term data from the Ross et al shows the clear advantages that Glycosade (an advanced form of cornstarch dietary therapy produced by Vitaflo International Ltd using a patented process) has over UCCS.

Katalin M. Ross, Laurie M. Brown, Michelle M. Corrado, Tayoot Chengsupanimit, Latravia M. Curry, Iris A. Ferrecchia, Laura Y. Porras, Justin T. Mathew, Monika Dambska, David A. Weinstein

Safety and efficacy of chronic extended release cornstarch therapy for GSD type I. *JIMD reports 2015.*	GSD type I	Ketotic forms of GSD (0, III, VI and IX)	Safety and efficacy of long-term use of extended release cornstarch therapy for GSD types 0, III, VI and IX. *J Nutr Ther 2016.*	First published paper using Glycosade outside GSD type I
Most severe form of GSD. Defective release of glucose into the blood, coupled with defective endogenous production of glucose, leading to severe hypoglycaemia. Many long-term complications.		Hypoglycaemia is typically not as severe as type I but many patients still require nocturnal intervention to prevent hypoglycaemia when they are asleep. Many long-term complications.		

Methods (common to both studies)

Patients undertook an open-label overnight challenge using Glycosade

Those deemed to have successfully completed the challenge (>2 additional hours of safe fasting, compared to their response with UCCS, with no compromise in other parameters of metabolic control) were offered the option of entering long-term phase of trial.

**Successful dosing was associated with two peaks in glucose concentration.*

During the long-term phase, Glycosade had to be consumed at least 3 nights per week.

Subjects assessed at baseline and 12 months.

Results (common to both studies)

Significantly longer period of safe fasting when using Glycosade (p<0.001)

**Intake guidelines for Glycosade have not yet been formally established but examples for type I patients are as follows:*

5-6 yrs	*60-75g*
7-8 yrs	*75-90g*
pre-pubertal	*90-120g*
pubertal	*135-150g*
adults	*120-150g*

Loading test data

Type I trial results, n = 106 (93 Ia [43M, 50F]; 13 Ib [7M, 6F]). Age range = 5-60 years.

Mean duration of fasting 7.8hr (Glycosade) vs 4.1hr (UCCS)

Success in 88% of GSD Ia patients (82/93)

Success in 77% of GSD Ib patients (10/13)

Success in 95% females and 78% males

Metabolic markers of control stable

Loading test data

Ketotic forms results, n = 16 ([10M, 6F]: Type 0 = 2, III = 8, VI = 1, IX =5). Age range = 5-60 years.

Mean duration of fasting 9.6hr (Glycosade) vs 4.9hr (UCCS)

Success in 100% of subjects

Metabolic markers of control stable

**Prescribed intake of Glycosade was much lower outside of GSD type I. Individualised doses (usual range 30-60g) which are NOT based on weight.*

Long-term data

Post-trial, long-term data available for 41/82 type Ia patients *(*32 patients aged 5-17 years. 9 patients >18 years.)*

Long-term data is only available for 3/10 type Ib patients with many withdrawals due to GI issues. *(*2 patients aged 5 and 13 years. 1 patient aged 24 years.)*

Long-term data

Long-term data available for all 16 subjects. *(*12 patients aged 5-17 years. 4 patients aged >18 years.)*

Conclusion

Glycosade "appears to improve quality of life of patients with GSD I without sacrificing metabolic control. Avoiding the overnight dose of cornstarch should improve safety in this population."

Glycosade "dramatically prolongs the overnight fast duration, maximises safety from hypoglycaemic events, reduces the possibility of sleep deprivation, and improves the quality of life of patients by eliminating the need to awaken without fail for middle of the night therapy without sacrificing metabolic control."

Additional comments

Open access:
https://www.researchgate.net/profile/David_Weinstein5/publications

Open access:
https://www.researchgate.net/publication/292783898_Safety_and_Efficacy_of_Long-Term_Use_of_Extended_Release_Cornstarch_Therapy_for_Glycogen_Storage_Disease_Types_0_III_VI_and_IX

Cost: Benefit aspects of Glycosade (vs UCCS) discussed - additional cost of Glycosade can be justified if compared to the cost of a single emergency room admission.

Glycosade is a food for special medical purposes. It is for use in the dietary management of GSD where the use of a long-acting starch is indicated.
It must only be used under strict medical supervision.
Intakes in this document are for guidance only; prescribed intakes must always be based on an individual patient assessment.