



**glycosade**<sup>®</sup>



Innovation in Nutrition

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A practical guide for overnight use of Glycosade<sup>®</sup>  
in hepatic Glycogen Storage Diseases



Vitaflo in Association  
With You

Supporting education in the  
dietary management of rare diseases



## Disclaimer

This is a Practical Guide for the overnight use of Glycosade in individuals diagnosed with hepatic Glycogen Storage Diseases (GSD).

**For use by Healthcare Professionals only.**

**Not for use by parents of children with GSD or for individuals with GSD.**

For general information only and must not be used as a substitute for professional medical advice or treatment.

Glycosade should only be used in GSD Type Ia & Ib, GSD Type 0, III, VI & IX. Glycosade is indicated for use over the age of 5 years in the USA.

All product information was obtained from manufacturer's information available as of the date of publication and is subject to change.

For specific product information, please consult the manufacturer.

## Collaborators

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## Abbreviations

<b>GSD</b>	Glycogen Storage Disease
<b>IBD</b>	Inflammatory Bowel Disease
<b>CHO</b>	Carbohydrate
<b>BGL</b>	Blood Glucose Level
<b>UCCS</b>	Uncooked Cornstarch
<b>IBW</b>	Ideal Body Weight
<b>HCP</b>	Healthcare Professional

## Blood Glucose Unit Conversion

mg/dL	mmol/L
40	2.2
50	2.8
60	3.3
70	3.9
80	4.4
90	5.0

## Foreword

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 abnormal synthesis or degradation of glycogen, and this results in severe and potentially life threatening hypoglycemia.

Cornstarch or uncooked cornstarch (UCCS) was a major breakthrough in the dietary management of GSD I. It has been well studied and considered the standard dietary therapy in North America since the 1980's.<sup>1-6</sup> Methodology and guidelines for establishing the recommended intakes and prescribing UCCS in GSD have been documented.<sup>7-12</sup> Cornstarch is also used for the management of hypoglycemia in GSD 0, III, VI and IX.<sup>4,13-15</sup>

Despite the positive impact of UCCS for patients with GSD type I, it has a limited duration of action. For maintenance of metabolic control, patients must receive UCCS every 4–5 hours, and one study suggested that UCCS only maintains normoglycemia for a median time of 4.25 hours in children.<sup>10</sup> Children must awaken in the middle of the night for UCCS intake and delayed administration can be associated with the development of hypoglycemia, seizures, and neurologic injury. Even as adults, almost all individuals still require UCCS at least every 4–6 hours, and overnight administration is required in approximately 90% of individuals to maintain metabolic control.<sup>1,7,9,10</sup>

Vitaflo has carefully developed and researched a new long acting starch suitable for GSD (Glycosade). Glycosade provides a further breakthrough in the management of GSD in that it offers a significant advantage over UCCS by providing a slower release of glucose, and therefore an extended period of normoglycemia. The main benefit of using Glycosade at night time is the potential

for a longer period of sleep avoiding the need for waking for additional overnight administration of UCCS<sup>16,17</sup> due to the prolonged period of normoglycemia. This can therefore maximize the potential of improved quality of life and lower the risk for missed overnight therapy. For individuals whose glucose is managed using a continuous overnight enteral tube feed, Glycosade may also be beneficial when compared to the negative impacts associated with continuous intragastric drip feeds.<sup>16,17</sup>

Glycosade was originally approved as a Food for Special Medical Purposes in the United Kingdom in 2009, and we now have over 7 years' experience of using this product in children through adulthood in the United States with GSD types 0, Ia, Ib, III, VI and IX. Vitaflo has therefore developed a practical guide on the use and introduction of Glycosade for night time use in the dietary management of children over the age of 5 years and adults. As with any change in dietary management, individuals with GSD will respond differently. Therefore, it is important to monitor individual metabolic response and adjust dose until the most appropriate intake for maintenance of metabolic control is achieved. Following the suggested step wise system for initiating Glycosade can support successful introduction and maintenance of metabolic control.

**David A. Weinstein**, M.D., M.M.Sc.

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## 1.0 Introduction to Glycosade

### 1.1 What is Glycosade?

Glycosade is a form of cornstarch with a high amylopectin content, which has been heat and moisture treated using a specialized patented process to produce a slower release starch compared to UCCS. Glycosade is a medical food currently labeled for use from 5 years of age in the USA.

### 1.2 Presentation of Glycosade

Glycosade is available in 60 g packets providing 53 g of carbohydrate.

Select Nutrients	per 100 g	per 60 g
Calories (kcal)	352	211
Protein (g)	0	0
Carbohydrate (g)	88	53
Sugars (g)	0	0
Fat (g)	0	0

Ingredient: Cornstarch.

### 1.3 Indications for use of Glycosade

Glycosade is indicated for use in individuals with GSD 0, I, III, VI and IX, who experience periods of hypoglycemia or have difficulty in maintaining normoglycemia and require long acting starch dietary management. To date it has been primarily studied for use as an overnight source of glucose in GSD Ia and Ib<sup>18</sup> and GSD III, VI, IX and 0 in the USA.<sup>19</sup> The majority of the individuals studied have been over the age of 5 years and there is limited published evidence for use in children younger than 5 years of age.

## 1.4 Principles of calculating Glycosade requirements

The amount of Glycosade required to maintain normoglycemia will vary according to the type of GSD, age, weight, activity level, individual response and other factors. It is therefore important to monitor individual clinical and metabolic response, and adjust the dose of Glycosade accordingly.

Ideally an individual with GSD is admitted to the hospital for initial dose evaluation of Glycosade. This allows for careful evaluation and monitoring in a controlled environment. It also minimizes the risk of hypoglycemia since metabolic instability may occur if the therapy is not tolerated.

The following sections include a suggested process for an individual transitioning to Glycosade from UCCS for the overnight period and are based on evidence from clinical studies,<sup>16-19</sup> case reports<sup>20,21</sup> and personal experience from the collaborators.

## 2.0 Use of Glycosade in GSD

### 2.1 In GSD Type Ia & Ib established on UCCS

If an individual is already established on UCCS and in good metabolic control, it is suggested that the initial Glycosade dose be based on the total amount of UCCS the individual is taking overnight.

The goal of overnight dietary administration of Glycosade is to prolong the period of normoglycemia supporting an individual to sleep through the night without awakening for an additional cornstarch dose. Therefore, the initial suggested Glycosade dose should be calculated based on the combined 2 overnight UCCS doses.

**55 g UCCS is equivalent to approximately 60 g Glycosade**

Clinical evaluation comparing Glycosade to UCCS in 106 individuals with GSD Ia and Ib showed that Glycosade<sup>18</sup>:

- ▶ Significantly prolongs the duration of normoglycemia to support longer overnight fasting.
- ▶ Supports metabolic control, which is expected to positively impact long-term health outcomes.

The table below outlines the Glycosade dosing ranges by age group used in this cohort of GSD Ia and Ib patients. Ranges can be used to cross check the calculation made from an individual's usual combined overnight UCCS dose.

**Table A**

Suggested intake of Glycosade for night time dose in GSD type I	
Age	Dose (g)
5–6 years	60–75
7–8 years	75–90
Pre-pubertal	90–120
Pubertal	135–150
Adults	120–150

### Considerations for Glycosade starch load test

- ✓ Metabolic control should ideally be optimal.
- ✓ Hospital admission for 2 nights is ideal.
- ✓ On night 1 of admission the individual should undergo a starch loading test of their usual UCCS dose. The duration of normoglycemia and metabolic control needs to be documented. This will then form a baseline for comparison to effectiveness of Glycosade.
- ✓ On night 2 of admission the individual should undergo the Glycosade starch loading test.
- ✓ Ensure individuals with GSD Ib do not have raised inflammatory markers (i.e. not experiencing a flare up of IBD).

## 2.2 In GSD 0, III, VI & IX

In GSD 0, III, VI and IX glucose homeostasis is usually less severely affected than in GSD type I<sup>13</sup> and in general, the required intake for UCCS and therefore Glycosade will be lower than for GSD type I.

Clinical evaluation comparing Glycosade to UCCS in 16 individuals with GSD 0, III, VI and IX showed that Glycosade<sup>19</sup>:

- ▶ Significantly prolongs the duration of normoglycemia to support longer overnight fasting.
- ▶ Supports metabolic control, which is expected to positively impact long-term health outcomes.

It is important to emphasize that not all patients with GSD 0, III, VI, and IX require middle-of-the-night cornstarch doses. Individuals with these types of GSD should monitor morning glucose and ketone levels. **Glycosade use should be limited to those with morning hypoglycemia or ketosis.**

The principles of determining the initial overnight Glycosade dose are the same as for GSD Type I.<sup>11</sup> The initial suggested dose should be equivalent to the combined 2 overnight UCCS doses. Below is the suggested dosage range from Ross et al. for Glycosade<sup>22</sup>:

### **The suggested dose range for GSD Types III, VI, IX, and 0 is 30–75 g Glycosade**

*Note: the individual dose is NOT age- or weight-based*

If the calculated dose falls outside of the suggested range, take into consideration previous overall metabolic control and adherence to dietary management. In general, if the calculated dose is below 30 g, give an initial dose of 30 g Glycosade. If the calculated dose is above 60 g, initiate Glycosade dose at 60 g.

#### **Considerations for Glycosade starch load test at home**

If the following criteria apply:

- ✔ History of adherence to medical recommendations and metabolic control.
- ✔ The calculated dose falls within the recommended range.
- ✔ The medical team and family is confident that the starch loading test could be carried out at home.

A similar careful monitoring program with clear instructions and documentation should be provided to the individual/family for a home-based starch load test. Close communication with the medical team is recommended according to local clinical guidelines.

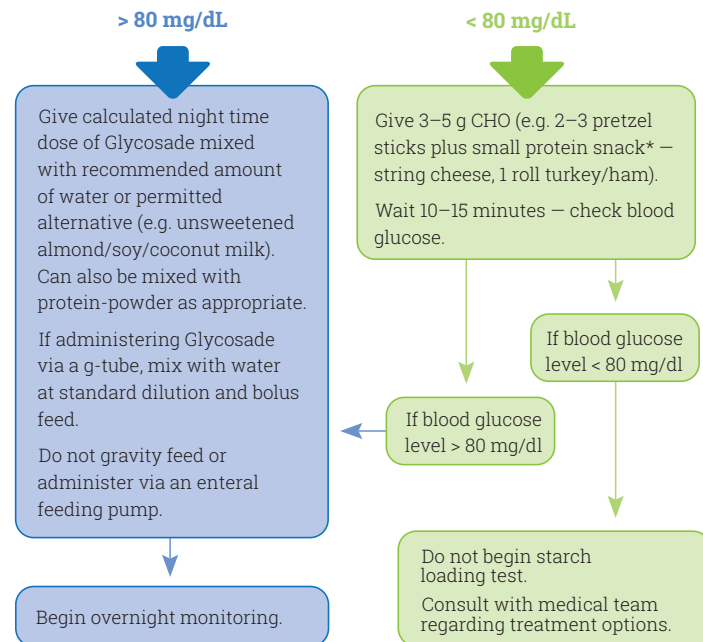
## 2.3 Calculating and initiating the Glycosade dose (all GSD types)

- 1 Determine initial dose: convert total overnight intake of UCCS into grams of Glycosade.

Usual UCCS dose (g)	Equivalent Glycosade dose (g)
55	60
60	65
65	71
70	76
75	82
80	87
85	93
90	98
95	104
100	109
105	115
110	120
115	125
120	131

- 2 Establish how the patient will take Glycosade: oral or bolus via an enteral feeding tube.

- 3 Check blood glucose level. If...



\*protein included for GSD types III, VI, IX

**Note:** If the calculated dose falls outside of the suggested intakes outlined in section 2.1 & 2.2, it is at the health care professionals' discretion to choose the initial dose. Consider the following:

- ▶ If the child is below the average weight and height percentile for age, the Glycosade dose is likely to be nearer the lower end of the suggested range.
- ▶ If the child is above average for weight and height, the Glycosade dose is likely to be nearer the upper end of the suggested range.
- ▶ History of metabolic control and adherence with dietary management.

## 2.4 Suggested overnight monitoring during a planned hospital admission

Ideally an indwelling intravenous (IV) catheter is inserted for the ease of taking blood draws hourly through the night.

Local policy/procedures should be followed regarding insertion and maintenance of IV access.

### All GSD types

#### Monitor blood glucose levels hourly

If blood glucose level...	Action
Is 80 mg/dL or above	No action, continue monitoring hourly
Drops between 75–80 mg/dL	Monitor every 30 min
Drops between 70–75 mg/dL	Monitor every 15 min
Drops below 70 mg/dL	Stop the starch loading test and administer carbohydrate as per standard local practice. Document at what hour this occurred.

**Note:** If blood glucose stays > 80 mg/dl until breakfast time, give usual breakfast and cornstarch dose as per individual feeding protocol.

### GSD Type Ia and Ib

#### Monitor lactate levels hourly.

The goal is to maintain normal lactate concentrations. Lactates above the normal reference range, or above baseline for the patient, even in the setting of blood glucose levels >80 mg/dL should prompt the medical team to stop the starch load test and provide a source of carbohydrate according to standard local practice. Document at what hour this occurred.

### GSD Type 0, III, VI and IX only

#### Monitor ketones at baseline and then every 3–4 hours.

Ketone levels above 0.2 mmol/L: Stop the starch-loading test and provide a source of carbohydrate per standard local practice. Document at what hour this occurred and monitor blood glucose levels hourly.

**Note:** Glycosade starch loading protocols have been described by Ross et al.,<sup>18,19</sup> Correia et al.,<sup>16</sup> Bhattacharya et al.<sup>17</sup> Cornstarch loading protocols have been described by Chen<sup>1</sup> and Wolfsdorf.<sup>2,3,7</sup>

## 2.5 Discharge home

### Starch load test successful

Whether the overnight starch loading test is successful with Glycosade will be determined by both the medical team and individual/caregivers. This may be when normoglycemia lasts for any amount of time longer than the usual UCCS dose, which is perceived beneficial to the quality of life of the individual/family:

- ▶ Send the patient home with a starter kit of Glycosade.
- ▶ Document prescribed dose and instructions for mixing and storage.
- ▶ It is recommended that the patient repeats the trial at home for one or two nights to see if the same response is observed.
  - This should be undertaken with close monitoring of glucose and ketone levels if appropriate (depending on GSD type), and communication with the medical team.



- Glycosade must be taken immediately after preparation and at the time prescribed by the HCP.
- Hands must be washed prior to taking each finger prick test (follow HCP instructions):
  - It is recommended that the medical team provides a detailed at home monitoring protocol following the principles outlined in sections 2.3 and 2.4.
- Individuals/caregivers should communicate with the team if a problem occurs.

### Starch load test unsuccessful

If blood glucose levels have dropped through the night (at the same time or earlier than usual) requiring the starch load test to be stopped, discharge home on dietary management with usual UCCS regimen.

If the medical team suspects that timing and/or dosing was incorrect, repeat the starch load test in the hospital setting, either with an increased or decreased dose of Glycosade on a different date.

### 2.6 GSD type Ia & Ib not established on UCCS or on an overnight enteral tube feed

If an individual is not established on UCCS, has a history of non-adherence to dietary management, and as a result there is no historical reference UCCS dose to form a basis for the calculation, a good starting point is to calculate glucose requirements per hour. First calculate CHO requirement for the overnight period based on recommendations per kg body weight. This may be cross checked against the suggested dose ranges in Table A in section 2.1.

Glucose/hr goal*		
Age (years)	Weight (kg)	Grams of glucose per hour
5-6	20	7.5
7-8	25	8.4
9-10	30	9
10-12	30-40	10
Adult	-	10-11

\*The glucose requirements in this table have been calculated using the equation published by Bier et al.<sup>22</sup>

**Note:** Requirements may go up to 15 mg glucose/hour in puberty.

Convert total glucose required overnight to grams Glycosade needed:

60 g Glycosade is equivalent to approximately 53 g Carbohydrate

Follow initiation, overnight monitoring and discharge instructions as detailed in sections 2.1-2.3.

## 3.0 Evaluation and monitoring

### 3.1 General considerations

It is recommended that patients on Glycosade are monitored according to local clinical practice procedures or as outlined in the most recent national/international guidelines for management.<sup>13, 23, 24</sup>

Individual response to Glycosade will vary according to age, growth, type of GSD and activity level. Periodic dose adjustment may be required to maintain metabolic control.

Additional monitoring will be required if illness is suspected, or during illness. Changes in dietary management, including dose adjustment and other interventions may be considered according to local/standard clinical practice.

If fewer overall feeds are administered as a result of inclusion of Glycosade in the diet regimen, protein intake may be less than estimated requirements. Protein requirements and intake should be reviewed and any deficit should be replaced. Checking prealbumin concentration may be a good indicator of protein status for patients with ketotic forms of GSD.

### 3.2 Practical tips

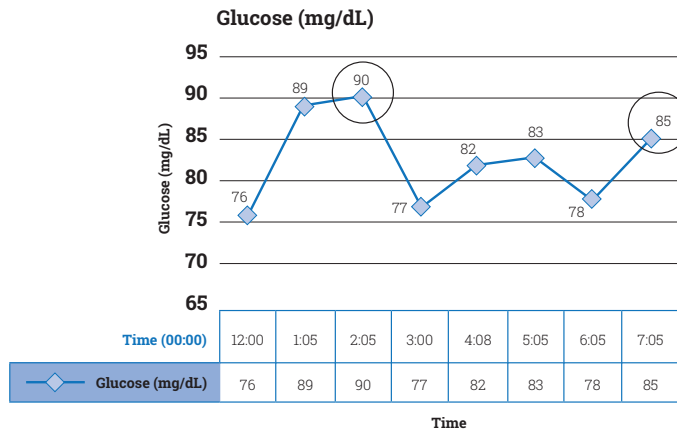
- ✔ It is helpful to try Glycosade with different flavor options to determine preference before evaluation of overnight dose in the hospital setting.
- ✔ Glycosade can be mixed with water, unsweetened almond or soy milk, flavored with a sugar free flavoring or liquid water enhancer drops. It can also be mixed with yogurt or sugar free pudding if permitted. Ask your Vitaflo representative for the Glycosade recipe guide for mixing ideas.

- ✔ The liquid mixed with Glycosade should be cold or at room temperature; **never mix with hot liquid or food as this can break down the starch structure and may alter the metabolic response.**
- ✔ Glycosade should be taken immediately after mixing. If Glycosade has been left to stand, the powder settles, making the full amount difficult to consume. It is important to shake the container to fully mix and drink immediately.
- ✔ Glycosade should be taken immediately before bedtime, as metabolic response may vary if the individual remains active and awake.

### 3.3 Checking the dose

The collaborators' clinical experience suggests that successful dosing of Glycosade is associated with 2 peaks in blood glucose concentration. The following graph illustrates the observed peaks in blood glucose concentration.

Figure 1: Starch load test of a 21 year old female with GSD Ia



- ▶ If a significant drop in blood glucose level is observed after 3–5 hours, this may indicate that the Glycosade dose is too high or too low. Consider the following:
  - If lactate levels have remained low – this may indicate that the dose was too high.
  - If lactate levels have risen – this may indicate that the dose was too low.

**Is a different dose needed?**

- ▶ Try a different dose and evaluate again on a different date as detailed above in section 2.1.
- ▶ Increases/decreases in dose increments of 5 and 10 grams per night is suggested.

**Note:** There are situations where caution must be exercised when trialling a new starch source:

- ▶ In GSD type 1b when experiencing a flare up of IBD
- ▶ If illness is suspected (e.g. fever, diarrhea, vomiting)
- ▶ Pubertal growth spurts
- ▶ Under 5 years of age

**3.4 Dos and don'ts**

Do	Don't
Store in a cool dry place.	Heat Glycosade.
Mix by shaking or stirring into permitted liquid.	Mix with hot food or liquid.
Mix and take immediately.	Mix with very acidic food or liquid; (the acid may alter the starch structure).
Carefully seal the packet if not all Glycosade is used.	Use a mechanical blender to mix.
Use within 24 hours of opening packet.	Use after expiration date.

**4.0 Preparation guidelines**

**Dosage and Administration**

To be determined by the clinician or dietitian following appropriate assessment (a fasting Glycosade load is recommended) and is dependent on the age, body weight and medical condition of the patient.

Each 60 g packet of Glycosade provides 53 g of carbohydrate, and is equivalent to approximately 55 g of UCCS.

**Important Notice**

Must only be used in patients with proven hepatic GSD where the use of a long acting starch is indicated under strict medical supervision.

Suitable from 5 years of age.

Not for use as a sole source of nutrition.

Not for intravenous use.

For enteral use only.

Glycosade may be given through a feeding tube. Not intended for use as a continuous enteral feed. Administer as a bolus to prevent possible tube blockage.

**DO NOT HEAT OR WARM GLYCOSADE, AS THIS DESTROYS THE PROPERTIES OF THE STARCH.**

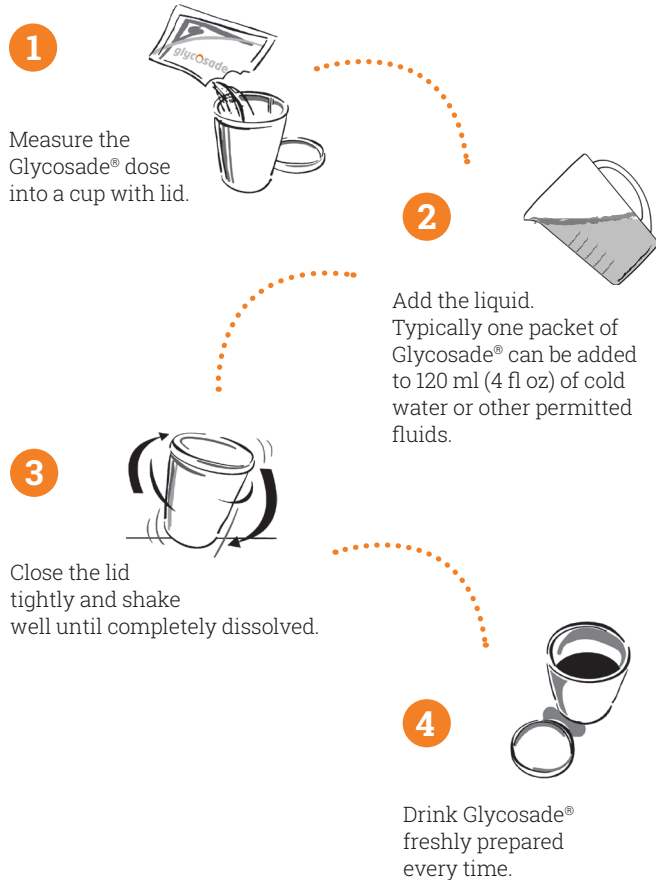
**Storage**

Store in a cool dry place. Ideally packets should be used immediately after opening. Any unused powder in an open packet should be kept in an airtight container or sealed with a clip and used within 24 hours.

**Pack Size**

30 x 60 g (2.1 oz) packets

## Glycosade® taken as a drink:



Tube feeding instructions and additional serving suggestions are available on request.

## 5.0 Case studies

### Case Study, GSD Type Ia female

#### Medical background:

Diagnosed with a liver biopsy on 7<sup>th</sup> day of life in 1994. Prior to referral, many hospitalizations occurred due to poor metabolic control. Patient was on continuous overnight feeds until 15 months of age.

**2011:** 16 years and 7 months of age

#### Anthropometrics:

Weight: 68.1 kg, Height: 163.1 cm, BMI: 25.6 kg/m<sup>2</sup>

#### Dietary recommendations:

Limited carbohydrate diet with emphasis on complex carbohydrates. Avoidance of simple carbohydrate, limitations on sucrose, fructose, and galactose, protein per DRI for age and EER for energy.

Cornstarch regimen	
Time	
7:30 am	65 g cornstarch
11:00 am	50 g cornstarch
2:00 pm	50 g cornstarch
5:00 pm	30 g cornstarch
7:30 pm	30 g cornstarch
10:00 pm	65 g cornstarch
2:30 am	65 g cornstarch

#### 2012 Glycosade trial:

At age 18, the medical team and family agreed to trial Glycosade for the patient to sleep through the night without awaking for middle-of-the-night cornstarch administration to maintain normoglycemia. The patient was admitted to the hospital for a 2-night starch loading test. On the next page are results of the overnight trial with Glycosade.

## Overnight starch loading test

Time	Glucose (mg/dL)	Lactate (mmol/L)	Comment
8:30 pm	96	1.8	
9:30 pm	92	1.3	
10:30 pm	69*	1.6	120 g Glycosade
11:30 pm	72	2.8	
12:30 am	73	2.5	
1:30 am	81	1.8	
2:30 am	72	2.1	
3:30 am	65*	2.1	
4:30 am	73	2.0	
5:30 am	74	1.6	
6:30 am	76	1.7	
7:30 am	58*	2.2	Breakfast and cornstarch given

\*Nursing staff closely monitoring

Glycosade trial was considered successful prolonging the overnight fasting period for a total of 8 ½ hours. This is over 4 hours longer than with the use of cornstarch. The patient was discharged home on the following regimen:

Time	
7:30 am	65 g cornstarch
11:30 am	60 g cornstarch
3:30 pm	60 g cornstarch
7:30 pm	60 g cornstarch
11:00 pm	120 g Glycosade

## Biochemistry

Date	AST	ALT	TG	Cholesterol	Uric Acid	Lactate
Aug 2006	109	86	505	217	9.2	n/a
July 2012	22	20	258	216	6.7	2
July 2018	25	25	191	218	6.8	2.2

## Case Study, GSD Type IIIa female

### Medical background:

Diagnosis in August 2010, s/p liver biopsy at 23 months of age. The child was referred from another medical center in May 2011 and presented with splenomegaly, hepatomegaly, and presumed hypertension.

Biochemistry March 2010 (before diagnosis)	
Lab	Value/unit
AST	2815 U/L
ALT	1052 U/L
Alk Phos	426 U/L
Bilirubin	0.9 mg/dL
Total Protein	7.1 g/dL
Albumin	3.8 mg/dL

### Dietary recommendations:

High protein diet and cornstarch dietary therapy were initiated. Family counseled on strategies to optimize protein intake, emphasis was placed on selecting complex carbohydrates and avoidance of excessive intake of simple carbohydrates.

Recommended protein intake of 3–4 g/kg/day and cornstarch at 1.6 g/kg; EER for age and sex. Multivitamin supplement initiated.

2014: Example cornstarch and protein regimen (6 years of age)	
Time	
7:30 am	25 g cornstarch with 25 g protein
12:30 pm	25 g cornstarch with 25 g protein
5:00 pm	25 g cornstarch with 25 g protein
9:30 pm	30 g cornstarch with 25 g protein
2:30 am (middle-of-the-night therapy)	30 g cornstarch with 25 g protein

### 2015 Glycosade trial:

At age 7, the medical team and family agreed to trial the child on Glycosade for the patient and family to sleep through the night without awaking for middle-of-the-night cornstarch intake to maintain normoglycemia. The child was admitted to the hospital for a 2-night starch loading test. Below are results of the overnight trial with Glycosade.

### Anthropometrics and biochemistry:

Weight: 56.2 kg, height: 152.4 cm, BMI: 24.2 kg/m<sup>2</sup>

Lab	Value/unit
AST	258 U/L
ALT	333 U/L
Alk Phos	416 U/L
Bilirubin	0.6 mg/dL
Total Protein	7.3 g/dL
Albumin	4.1 mg/dL
Prealbumin	13 mg/dL
TGs	162 mg/dL
Total Cholesterol	148 mg/dL

### Overnight starch loading test

Time	Glucose (mg/dL)	Lactate (mmol/L)	Ketones (mmol/L)	Comment
11:00 pm	73*	0.3	0.1	75 g of Glycosade
12:15 am	97	1.6	–	
1:05 am	95	1.4	–	
2:05 am	77	1.0	0.2	
3:05 am	89	0.9	–	
4:05 am	90	0.7	–	
5:05 am	68	0.8	0.2	
6:00 am	68	0.5	–	
7:05 am	60	0.4	0.6	Breakfast and cornstarch

\*Given 3-5 g of carbohydrate: glucose rechecked after 15 minutes and was above >80 mg/dL

Glycosade trial was considered successful prolonging the overnight fasting period for a total of ~8 hours. The child was discharged home on the following regimen:

Time	
5:30 am	25 g cornstarch with 25 g protein
8:30 am	25 g cornstarch with 25 g protein
12:30 pm	25 g cornstarch with 25 g protein
5:00 pm	30 g cornstarch with 25 g protein
9:00 pm	75 g Glycosade with 25 g protein

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