NUTRITION MANAGEMENT IN GSDIa

A guide for dietitians

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GSDIa Is a Rare Metabolic Disorder That Impairs Glucose Production

GSDIa is a rare, autosomal recessive metabolic disorder that leads to impaired glucose production.[1] It is caused by a deficiency in the enzyme glucose-6-phosphatase (G6Pase) due to pathogenic variants of the *G6PC* (*G6PC1*) gene.[1] G6Pase is a critical enzyme in the production of glucose, as it catalyzes the final step of both glycogenolysis and gluconeogenesis.[1]

Maintaining glucose levels and metabolic control is critical

Patients with GSDIa must avoid fasting and adhere to an around-the-clock cornstarch regimen.[2] Strict adherence to the dosing schedule is vital, as even one skipped or delayed dose may result in life-threatening consequences of hypoglycemia.[1,2]

A modified diet that limits sucrose, fructose, lactose, and galactose, and focuses on balancing complex carbohydrates with protein and fat is important to maintain glycemic and metabolic control.[2,3]

Feeding considerations in infants

Infants younger than 6-12 months of age lack sufficient levels of amylase needed to digest cornstarch and are initially managed with a soy-based formula, free of sucrose, fructose, and lactose, every 2 to 3 hours on demand to prevent hypoglycemia[2]

Infants who begin to sleep longer than 3 to 4 hours can receive a nasogastric (NG) tube or gastrostomy tube (G-tube) to allow caregivers to administer feeds during sleep, when the child is sick, or when the child refuses or is otherwise unable to eat by mouth[2]

GSDIa, glycogen storage disease type Ia.

Estimating Daily Carbohydrate Needs

Cornstarch and tube feeding requirements vary by age.[1] The following weight-based formulas may be used to determine dosing:

Current guidelines for cornstarch and tube feedings*

Cornstarch[1,2,4]	
Young child	1.0-1.6 g/kg (ideal body weight) every 3-4 hours
Older child/adult	1.7-2.5 g/kg (ideal body weight) every 4-5 hours
Tube feedings[2]	
Infant	Glucose infusion rate of 8-10 mg/kg/min
Older child/adult	Glucose infusion rate of 4-8 mg/kg/min

*This table provides a compilation of nutritional strategies from international publications and is intended for guidance only. Nutritional recommendations should come from the patient's healthcare provider to meet individual needs. Additional recommendations for hypoglycemic recovery, exercise, and illness are not included here.

Glucose needs may also be estimated using the following formulas

Children <8 years of age (the Bier equation)[4-6]

$Y = 0.0014X^3 - 0.214X^2 + 10.411X - 9.084$,

where Y = mg of glucose/min
and X = body weight (kg)

Older children (>8 years of age) and adults[4]

10-11 g of glucose/hour

Estimated glucose needs in g/hour may be applied to:

Cornstarch dosing | Overnight tube feeding rate | Carbohydrates per meal/snack

Note that 1 g of cornstarch has 3.8 kcal and 0.9 g of carbohydrates.[7]

Many Factors May Impact Blood Glucose Levels

Metabolic needs evolve with time and in response to specific situations.[1] Ongoing reassessment of cornstarch dosing and your patient's overall nutrition plan is critical to help prevent both overtreatment and undertreatment.[1]

Factors that may affect glycemic control include[1,2]:

- **Changes in activity level**, such as starting a new sport or exercise routine
- **Adjustments to cornstarch doses and/or frequency**
- **Acute illness**, which may alter the patient's metabolic needs and/or their ability to maintain their dietary intake
- **Hormonal changes**, such as puberty and pregnancy
- **Age**, with cornstarch requirements typically decreasing in adulthood
- **Stress**, which may impact metabolic control

Developing a Personalized Meal Plan

You can help your patients customize healthy, balanced meals and snacks that support their personalized nutrition plan.

International nutrition guidance for GSDIa*

Diet[1,2]	
Carbohydrates (including uncooked cornstarch [UCCS])	60%-70% of total calories
Protein	10%-15% of total calories
Fat	Remainder of calories
Nonutilizable sugars	Sucrose (fructose) and lactose (galactose) are restricted to varying degrees, and sugar alcohols (especially sorbitol) are avoided
	Patients should avoid fasting by consuming small, frequent meals and snacks, properly balanced in complex carbohydrate, protein, and fat content
Supplements[1,2,5]	
Daily supplementation with a sugar-free multivitamin is recommended; additional calcium and vitamin D may be needed, and other supplements may be added based on assessment	

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Consistent cornstarch preparation helps maintain glucose stability

Cornstarch should be consumed uncooked, as heat may disrupt starch granules and reduce effectiveness, potentially impacting blood glucose levels.[2,3]

Cornstarch should be weighed using a gram scale and mixed with water, sugar-free soy milk, sugar-free drinks, or formulas free of sucrose, fructose, and lactose.[2]

Building a GSDIa Nutrition Plan

	General guidelines	May be advised to limit or avoid
UCCS[1,2]	• Complex carbohydrate; typically a sizable portion of total daily carbohydrate intake	• Switching between brands, as there may be variability • Mixing with acidic beverages
Other dietary carbohydrates[1,2,5]	• Accounts for the remaining carbohydrate intake but intake should be carefully controlled, as exceeding carbohydrate requirements may result in over-treatment, increased glycogen storage, and glycemic and metabolic instability • Options may include most nonstarchy vegetables, whole grains and starches, and legumes	• Fruit and vegetables high in sugar (eg, sweet potatoes, corn, peas, carrots) • Food containing fructose and galactose, as well as other sugars that break down into them (eg, sucrose, lactose) • Added sugar, sorbitol, honey, maple syrup, molasses, or high-fructose corn syrup • Dairy
Protein[2]	• Mostly lean meat, poultry, and fish	• Fatty and processed meats
Fats[2]	• Canola and olive oil • Avocados, seeds, and nuts	• *Trans* fats and saturated fats

Working closely with patients to understand their preferences, goals, and changing metabolic needs can help you individualize their plan. Consider discussing:

New foods they'd like to incorporate into their diet

Changes in exercise routines they are considering

Preferences for adjusting cornstarch schedule to better fit lifestyle

GSDIa, glycogen storage disease type Ia; UCCS, uncooked cornstarch.

Ongoing Monitoring Can Help Guide Nutrition Planning

Ongoing monitoring of blood glucose, metabolic parameters, and nutritional status helps identify trends and issues and guide adjustments to the treatment plan.[1–3] While hypoglycemia is the immediate concern for patients with GSDIa, it is important to identify and address other common issues, including[1,2,8]:

OBSERVATION	WHAT IT MAY INDICATE
Excessive weight, increased hepatomegaly, increasing hyperglycemia, and/or rebound hypoglycemia	Overtreatment with UCCS and/or diet[2,5,9]
Poor or faltering growth and development	Undertreatment and/or poor metabolic control; assess for feeding issues; referral to a feeding specialist may be indicated[2]
Blood glucose spikes after meals and/or the ingestion of UCCS	Dosing, spacing, and timing of meals and/or UCCS needs to be adjusted[9,10]
Increasing triglyceride levels	Poor metabolic control related to UCCS or inadequately balanced diet[2,11–13]
Chronic lactic acidosis	Poor metabolic control requiring UCCS and/or diet adjustments[12]

Based on the above assessments, a revised nutrition plan can be created to update:

- Caloric goals[1,2]
- Hypoglycemic rescue treatment[2]
- Dosing and frequency of UCCS[1,2]
- Timing and composition of meals and snacks (carbohydrate goals)[1,2]
- Other nutrient goals (eg, protein, fat, vitamins/minerals)[1,2]

REMEMBER!

The emergency plan to treat hypoglycemia and sick-day plan should be regularly reviewed and updated based on your patient's changing metabolic needs[1,2]

GSDIa, glycogen storage disease type Ia; UCCS, uncooked cornstarch.

Nutrition Is a Vital Component of Managing GSDIa

GSDIa is a rare metabolic disorder that impairs glucose production, resulting in a risk of life-threatening hypoglycemia and long-term complications[1]

Management of GSDIa relies on a strict nutrition plan and cornstarch regimen[1]

Careful monitoring and ongoing assessment can help you adjust your patient's nutrition plan to meet their changing needs[1,3,8]

Working closely with patients and their care team to understand preferences, goals, and changing metabolic needs can help you personalize their nutrition plan

Scan to uncover more about GSDIa, or visit **understandinggsdiahcp.com**

GSDIa, glycogen storage disease type Ia.

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