At a Glance
Glycogen Storage Disease (GSD) Type Ia and Ib

Deficient Enzyme: GSD Ia: glucose-6-phosphatase; GSD Ib: glucose-6-phosphatase translocase

Clinical presentation in undiagnosed patients or patients with poor metabolic control:[1]

Hypoglycemia, hepatomegaly, failure to thrive, short stature, lactic acidosis, hypertriglyceridemia, hyperuricemia; GSD Ib-neutropenia

Treatment Goals:

Prevent hypoglycemia (blood glucose goal 75-100 mg/dL)[2]
Correct laboratory abnormalities
Ensure adequate nutrient intake for age

Recommended Macronutrient Composition of Diet[2]

- Carbohydrate (CHO) (60-70% of energy intake)
  - Include calories provided by UCCS
  - Limit to 15 g CHO per meal and 5g CHO per snack †
  - Limit fructose to 2.5 g per meal
  - Limit galactose/lactose-containing foods to 1 serving/day
- Protein (10-15% of energy intake)
  - Offer lean sources of protein
- Fat (<30% of energy intake)
  - Limit saturated fat

Recommended Fasting Times[1]

Infants: 2-3.5 hours
Children and adults: 3-5 hours

Uncooked Cornstarch (UCCS) [2]

- Initiating therapy
  - Begin UCCS at 9-12 months of age
  - Use 1 g UCCS per dose, increase by 1 g increments as tolerated
- For children <8 years old, calculate Bier Equation to determine dose of UCCS
  - Bier Equation
  - $Y=0.0014X^3 – 0.214^2 + 10.411X – 9.084$
  - Y = mg glucose per minute
  - X = weight in kg
  - i.e., X=10 kg; Y=75 mg/min (4.5 g/hr)

For individuals > 5 years old, consider Glycosade, a slow-release form of cornstarch that extends fasting times in some individuals[3]

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Enteral Formulas for GSD I*

	Abbott abbottnutrition.com	Mead Johnson hcp.meadjohnson.com/s/	Nestle nestlehealthscience.us	Nutricia medicalfood.com
Infant (0-1 yr)	Elecare Infant	Prosobee Nutramigen Pregestimil Puramino	Alfamino Infant	Neocate Infant
Toddler and young children	Elecare Jr PediaSure Peptide (unflavored) PediaSure Peptide 1.5	Nutramigen Toddler Puramino Jr (unflavored)	Alfamino Jr Peptamen Jr (unflavored) Vivonex Pediatric Tolerex	Neocate Jr
Older children and adults	Osmolite Ensure Max Protein Nutrition Shake (oral-several flavors)		Glytrol Impact (unflavored) Isosource (unflavored) Nutren (unflavored) Peptamen (unflavored) Vivonex Plus Vivonex RTF Vivonex TEN	

***Ingredients may change, products should be reviewed before making a recommendation**

Supplementation

Sugar-free multivitamins, calcium, vitamin D3 to meet Dietary Reference Intake for age

Consider probiotics

Monitoring

Nutrition:

Anthropometrics, dietary intake, physical findings

Laboratory

Glucose, lactic acid, uric acid, triglycerides, cholesterol, liver function tests,

Markers for anemia (hemoglobin, hematocrit, MCV, ferritin, iron, folate, vitamin B_{12})

Glucose monitors and continuous glucose monitoring (CGM)

Point-of-care blood glucose testing (ie. Freestyle Light) test prior to UCCS dosing

CGM (ie. Dexcom) to assess trends over 24-hour period

References

1. Kishnani PS et al. Diagnosis and management of glycogen storage disease type I: a practice guideline of the American College of Medical Genetics and Genomics. Genet Med. 2014;16(11):e1.

2. Ross KM et al. Dietary Management of the Glycogen Storage Diseases: Evolution of Treatment and Ongoing Controversies. Adv Nutr. 2020;11(2):439-46.

3. Vitaflo. A practical guide for overnight use of Glycosade in hepatic Glycogen Storage Disease. Bridgewater, NJ: Vitaflo USA. LLC.: 2020.