At a Glance
Glutaric Acidemia Type 1 (GA-1)

Deficient enzyme: Glutaryl-CoA dehydrogenase

Metabolites*: Elevated concentrations of 3-hydroxyglutaric acid, glutaric acid, glutaconic acid and glutaryl carnitine

Restricted Amino Acid: Lysine

Clinical presentation, in untreated patients: brain atrophy, macrocephaly, striatal necrosis, dystonia, hypotonia

Goal Treatment Range: Plasma lysine- maintain at low end of normal range
Plasma free carnitine- maintain within normal range

*Boy N, et al. Proposed recommendations for diagnosing and managing individuals with glutaric aciduria type 1: second revision. J Inherit Metab Dis (2017) 40: 75-101

Nutrient Needs by Age*

Age	Lysine mg/kg/d	Total Protein g/d
0-6 mo.	65-100	2.75-3.0
6-12 mo.	55-90	2.5-3.0
1-4 yrs.	50-80	1.8-2.6
4-6 yrs.	40-70	1.6-20
>6 yrs.	Consider liberalization of protein intake to age-appropriate DRIs**	

* Bernstein, LE. Nutrition Management of Glutaric Acidemia Type 1. In LE Bernstein, F Rohr, S van Calcar (Eds.) *Nutrition Management of Inherited Metabolic Diseases (2nd Edition)*. Springer: 2021

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Illness in GA-1*

Emergency treatment is often needed at the first sign of illness such as decreased intake, fever, vomiting or diarrhea. Delaying emergency treatment is associated with significant risk for a neurologic crisis. Patients should be counseled to call their metabolic team for guidance at the first sign of illness.

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Starting a GA-1 Diet

1. Determine goals for Lysine (mg), Total Protein (g), Energy (kcal)
   - to estimate mg of lysine from grams of protein use the conversion 35 mg lysine = ~1 g protein
2. Calculate amount of intact protein source (breast milk, infant formula, food) needed to meet lysine goal.
3. Calculate amount of medical food needed in addition to the intact protein source to meet total protein goal.
4. Calculate energy intake from intact protein and medical food sources to ensure total energy goals are met.

Glutaric Acidemia (GA-1)

Medical Food Therapy

	Abbott abbottnutrition.com	Mead Johnson Hcp.meadjohnson.com	Nutricia NutriciaMetabolics.com	Vitaflo www.VitafloUSA.com
Infant (0-1 yr)	Glutarex®-1	GA	GA-1 Anamix® Early Years	
Toddler & Young Children	Glutarex®-1 Glutarex®-2	GA	GA-1 Anamix® Early Years GlutarAde™ Junior GA-1 Drink Mix GlutarAde™ Essential GA-1 Drink Mix GlutarAde™ Amino Acid Blend	GA gel™ GA express® 15
Older Children & Adults	Glutarex®-2	GA	GlutarAde™ Junior GA-1 Drink Mix GlutarAde™ Essential GA-1 Drink Mix GlutarAde™ Amino Acid Blend	GA express® 15

Nutrition Supplementation*

L-Carnitine
-100 mg/kg/d

L-Arginine
-Supplied by medical food, no evidence for benefit of additional supplementation

Riboflavin
-No standard protocol for evaluating responsiveness although certain individuals may show biochemical improvement

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Laboratory Monitoring*

Plasma Amino Acids[1, 2]
Carnitine[1, 2]
Albumin[3]
CBC[3]
Calcium[3]
Phosphorus[3]
Ferritin[3]
B12[3]

[1] Every 3 months until age 1 yr.
[2] Every 6 months until age 6 yrs., then annually thereafter
[3] As indicated

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