Lysine-Free, Arginine-Enriched Medical Foods for the Management of Glutaric Aciduria Type 1

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Glutaryl CoA-dehydrogenase deficiency (glutaric aciduria type 1, GA-1) is a recessive disorder of lysine and tryptophan degradation that can result in striatal degeneration within the first 2 years of life.

We reviewed dietary prescriptions (n=588), plasma amino acids (n=600), and outcomes for 39 children with GA-1 (49% male, 0.8 to 11.3 years, collective 192 patient-years) managed between 2006 and 2017 on a standard protocol of lysine-free, arginine-enriched medical foods, L-carnitine, and inpatient management of illness with intravenous delivery of 150% maintenance fluids, ~10 mg/kg/min dextrose, and L-carnitine (300-400 mg/kg/day).

Outpatient diets provided (per kg per day) 1.1 ± 0.3 g intact protein, 1.2 ± 0.3 g amino acids from lysine-free, arginine-enriched medical food, and 92 ± 17 mg L-carnitine. Plasma analysis showed reduced lysine (85.6 ± 38.6 µmol/L, reference range 213 ± 48 µmol/L), normal arginine (91.8 ± 33.2 µmol/L, reference range 90 ± 32 µmol/L), and a 50% decrease of the lysine to arginine concentration ratio (from 2.0 ± 1.0 to 1.1 ± 1.2). No child required nasogastric or gastrostomy feeding. One infant suffered acute striatal necrosis at age 7 months and two had delayed ambulation and mild dystonia, but no regressive event. All remaining children (92%) had normal growth and age-appropriate motor development.

For children with GA-1, a systematic approach to longitudinal lysine, arginine, and L-carnitine nutrition, coupled with consistent inpatient emergency care, decisively improves motor outcomes.

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