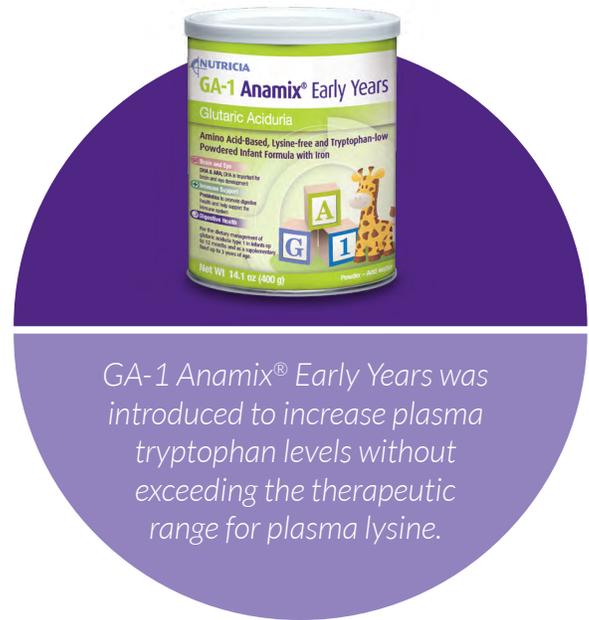


Nutrition management of an infant with GA-1 and transition to a tryptophan-reduced medical food

History

7-day old male referred for management of Glutaric Aciduria type 1 (GA-1) had a family history and confirmatory testing consistent with GA-1 (glutaryl carnitine (C5DC) 5.34 $\mu\text{mol/L}$ and presence of glutaric acid and 3-hydroxy-glutaric acid in the urine). The pregnancy was uncomplicated, and he was born at 37 weeks gestation. He received Similac[®] Advance after birth, eating every 2-3 hours. A non-Nutricia medical food (lysine- and tryptophan-free) was started at seven days of age after receiving the abnormal newborn screen results. Parents noted that before the GA-1 medical food was introduced, the infant was very sleepy and became more alert after starting the medical food. This was the couple's second child with GA-1.



Nutritional Management

The infant was started on a lysine- and tryptophan-restricted diet at seven days of life with carnitine supplementation and an emergency protocol for use during illness as standard of care. Similac Advance and the GA-1 medical food were used initially to provide approximately 88 mg lysine/kg, 3.3 g protein/kg and 120 kcal/kg. He was allowed additional bottles made with only medical food as needed. He later switched to Similac[®] Sensitive due to increased fussiness and gas. At one month of life, plasma tryptophan levels were low (9 $\mu\text{mol/L}$; reference range: 15-90 $\mu\text{mol/L}$) with normal lysine levels (124 $\mu\text{mol/L}$; therapeutic range: 60-120 $\mu\text{mol/L}$) corresponding with dietary intake of 14 mg tryptophan/kg and 83 mg lysine/kg. Repeat laboratory analysis showed both low tryptophan (9 $\mu\text{mol/L}$) and lysine (22 $\mu\text{mol/L}$) levels. The diet was modified to provide 17 mg tryptophan/kg and 98 mg lysine/kg, but plasma tryptophan remained low at 9 $\mu\text{mol/L}$.

At 3.5 months of age, GA-1 Anamix® Early Years was introduced in combination with the non-Nutricia medical food and Similac Sensitive to increase plasma tryptophan levels without exceeding the therapeutic range for plasma lysine. The new product was well tolerated, and parents did not report any constipation, diarrhea or gassiness with the change. Plasma tryptophan levels normalized with the addition of GA-1 Anamix Early Years (levels ranged from 22-47 µmol/L). At 11 months of age, the infant discontinued the non-Nutricia medical food and continued on GA-1 Anamix Early Years, Similac Sensitive and solid foods.

Results

The infant transitioned to GA-1 Anamix Early Years slowly, partly due to insurance concerns. Plasma amino acid profiles showed adequate intake of protein, and tryptophan levels have remained in the reference range since introducing GA-1 Anamix Early Years. He has maintained a normal growth velocity, with normal weight (49th percentile), height (48th percentile), and percentage target weight (99%) with relative macrocephaly, common in GA-1 (OFC 81st percentile) at 14 months of age. The infant had two hospital admissions for fever and vomiting in the first year of life, during which he received IV fluids with D10 and Intralipid®. He has not had a metabolic decompensation and no abnormal movements or tone.

“*He has maintained a normal growth velocity, with normal weight (49th percentile), height (48th percentile), and percentage target weight (99%).*”

This case report* is provided by Krista Viau, PhD, RDN; Boston Children’s Hospital; Boston, MA.

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