

Newly Diagnosed IVA Baby

Using standard infant formula and the new IVA Anamix[®] Early Years for dietary management

History

9-day old female referred for management of Isovaleric Acidemia (IVA) after both a positive newborn screen and confirmatory testing (isovalerylcarnitine 1.27 $\mu\text{mol/L}$ and isovaleryglycine 73 mmol/mol creatinine). The pregnancy was uncomplicated and she was born 39 weeks gestation. She breastfed exclusively for 2 days after birth, and then discontinued as she began to spit up with feeds. The infant switched to Similac[®] Sensitive, consuming approximately 3 fl oz Similac Sensitive every 3-4 hours when she presented to clinic. Mom continued expressing and storing breast milk. This was the couple's first child diagnosed with IVA.

Nutritional Management

The infant was started on glycine and L-carnitine supplements and a leucine-restricted diet as standard of care at 9 days of life. Expressed breast milk and XLeu Analog[™] were used initially; alternative medical foods were not available. The mixture provided 125 calories/kg, 2.3 g total protein/kg, and 116 mg leucine/kg. The infant was allowed additional bottles of XLeu Analog alone, as needed. Since diagnosis, a new IVA infant formula was brought to market and a decision was made to switch her to this new formula. The infant successfully switched to IVA Anamix[®] Early Years within a few days of initiating the leucine-restricted diet. Product was well tolerated and parents denied any constipation, diarrhea, or gassiness with the change. Due to inadequate breast milk supply, she transitioned to Similac Sensitive before 30 days of age. At 34 days, total protein intake increased to 3.5 g/kg (2.4 g/kg from IVA Anamix Early Years) and leucine to 115 mg/kg concomitant with an increase in appetite. At 2 months of age, she was consuming 126 calories/kg, 3.2 g total protein/kg (2 g/kg from IVA Anamix Early Years), and 120 mg leucine/kg.

Results

The infant successfully transitioned to IVA Anamix Early Years from XLeu Analog without changes in frequency or consistency of bowel movements or increased gas. This infant transitioned quickly without difficulty, which may be related to the short amount of time she was on XLeu Analog. She has maintained a normal growth velocity, with a normal length (58th percentile), weight (76th percentile) and head circumference (48th percentile) at 2 months of age. Plasma amino acid profiles showed adequate intake of total protein, and leucine intake was increased to normalize a low leucine level.

This case report* is provided by Krista Viau, PhD, RDN

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