

Transitioning: TYR infant formula to TYR Anamix® Next: *Seamless transition to a fiber-containing medical food*

Introduction

Tyrosinemia Type I (HT1) is an inherited metabolic disorder that requires a diet restricted in tyrosine and phenylalanine to prevent the buildup of tyrosine and succinylacetone in the body. For individuals with inborn errors of metabolism, transitioning to a new formula can be a challenge. A few reasons to introduce a new formula to a patient include poor formula tolerance, formula availability, age-appropriateness of nutrient composition, introduction of improved formulas on the market, and patient formula refusal. Metabolic healthcare professionals and patients must overcome these challenges daily. It is therefore important that formulas meet macronutrient and micronutrient needs, while at the same time having good patient acceptance. The following case reports on the medical and nutritional management of a patient with HT1 who transitions formula products.



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History

GM is a 2-year-old African-American/Caucasian female who was identified through newborn screening and diagnosed at two weeks of age with HT1. She was born at 39 weeks gestation at an appropriate weight with Apgar scores of 9 at 1 minute and 10 at 5 minutes. Complications included transient hypoglycemia secondary to gestational diabetes in the mother. The patient was started on Enfamil® Infant formula per the mother's preference.

Upon her confirmed HT1 diagnosis, she was placed on standard medical and nutritional management of the disease. Nutritional management ultimately involves a diet limited in phenylalanine (phe) and tyrosine (tyr). GM was started on XPhe, XTyr Analog®, Enfamil® Infant Formula, and Duocal® at 19 days of age. Phenylalanine (phe) and tyrosine (tyr) were both restricted to provide a combined total of 95 mg/kg of body weight per day. Daily calories were provided at 125 kcal/kg of body weight, and daily total protein was provided at 2.6 g/kg of body weight per day. Nitisone (NTBC) treatment was also initiated at standard dose for kg of body weight.

GM maintained good metabolic control on medical and dietary therapy with normal growth and development with no signs of liver dysfunction during her first year of life. Plasma tyrosine levels were maintained between 150-500 µmol/L and no traces of succinylacetone were detected in the urine.

Transition to Solid Foods and Next Stage Formula

Solid foods were introduced at 8 months of age, and at 16 months of age she was transitioned to TYR Anamix® Next due to discontinuation of XPhe, XTyr Analog. Furthermore, TYR Anamix Next was age-appropriate with added docosahexaenoic acid (DHA) as well as a multifiber mix, including prebiotics.

Her formula transition was accomplished in a step-wise process to decrease taste aversions and formula refusal. TYR Anamix Next was added in 2-3 tablespoon (T) increments, while XPhe, XTyr Analog was decreased to provide appropriate protein and calories for age. Her final formula prescription on solely TYR Anamix Next provided an adequate supply of micronutrients, 22 g of protein per day, and 300 kcal per day. In addition, she received 5 fl oz of whole milk and 150 mg of phe from foods for a combined total of phe and tyr at 641 mg per day.

	Step 1	Step 2	Step 3	Step 4
XPhe, XTyr Analog	10 T	6 T	4 T	0 T
TYR Anamix Next	2 T	4 T	6 T	9 T
Whole Milk	6 fl oz	6 fl oz	6 fl oz	5 fl oz
Natural foods	100 mg phe	100 mg phe	100 mg phe	150 mg phe

The formula transition for GM took about 4 months. In general, transition timing depends on the child and how often the clinic can be in contact with the family. GM did not experience any adverse effects nor taste aversions during the transition. She did not experience any negative gastrointestinal symptoms from the fiber provided from the formula. She has continued to thrive and develop in line with her peers on TYR Anamix Next.

Conclusion

Patients with HT1 require strict dietary limitations of phe and tyr intake. The challenge of providing adequate macronutrients and micronutrients in such a restricted diet can only be alleviated by the proper formulation of medical foods. In this patient's case, tyr- and phe-free formula selection was made based on meeting nutrient requirements, formula availability, and patient acceptance. GM had an easy transition to TYR Anamix Next and has continued to thrive with HT1.

This case report is provided by Grace Stuhrman, MS, RD, LD; Tulane University School of Medicine – Hayward Genetics Center.

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