

USE OF A NUTRIENT-DENSE FORMULA IN AN INFANT WITH LATE DIAGNOSIS OF CONGENITAL HEART DISEASE AND SEVERE MALNUTRITION

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CLINICAL PRESENTATION

Baby girl R presented to the emergency room at 4 months of age in cardiogenic shock and was diagnosed with a rare congenital heart defect of an anomalous left coronary artery from the pulmonary artery (ALCAPA). In a normal heart, the left coronary artery connects to the aorta and carries oxygen-rich blood to the heart muscle. With ALCAPA, the left coronary artery arises abnormally from the pulmonary artery and supplies the heart muscle with deoxygenated blood from the lungs. This can lead to starvation of the heart muscle, ultimately causing damage to the heart.

Upon diagnosis, she was taken urgently to the operating room for coronary reimplantation. Her intraoperative course was notable for severely diminished left ventricular systolic function for which she required mechanical circulatory support with veno-arterial extracorporeal membrane oxygenation (ECMO) for five days postoperatively. She remained hospitalized for two and a half months for ongoing medical management of her severe ventricular dysfunction, respiratory support, titration of cardiac medications and sedation, and growth optimization.

NUTRITION MANAGEMENT

Baby girl R was born at a healthy weight of 3.5 kg (weight-for-age 70th%ile, z-score 0.55). She had been exclusively bottle-feeding a cow's milk lactose-free infant formula 20 kcal/fl oz, drinking 4 to 6 fluid ounces per feed. Her mom reported she was having trouble gaining weight after 2 months of age, and for 5-7 days before coming to the hospital she was sweating and tiring easily with bottle feeding and vomiting more frequently. Her weight and length on admission at 4 months of age were 4.1 kg (weight-for-age <2nd%ile, z-score -4.04) and 58 cm (length-for-age <2nd%ile, zscore -2.42), and she appeared cachectic. Anthropometrics were consistent with severe malnutrition as evidenced by a weight-for-length z-score -3.07.

Parenteral nutrition was ordered on postoperative day (POD) 1. Trophic feeds were started continuously for 24 hours through a nasoduodenal (ND) tube with a full strength energy-nutrient dense formula (ENDF) after ECMO decannulation (POD 7). ENDF (30 kcal/fl oz, 10.3% protein:energy ratio) was recommended as the caloric content allowed for goal enteral nutrition to be achieved earlier, and the protein-energy ratio was more favorable to promote catch-up growth¹. After 2 days of trophic feeds, enteral feeds were advanced over 5 days to 130 mL/kg/day, which provided 130 kcal/kg/day and 3.4 g protein/kg/day. Parenteral nutrition was weaned off once she reached goal feeds by POD 14. Given her severe malnutrition, utilizing ENDF helped minimize the delay in optimizing her enteral nutrition by avoiding a stepwise increase in caloric density using a standard infant formula. ENDF also gave her optimal nutrition in less volume given her fluid limitations in the intensive care unit and ongoing requirement for diuretics in the setting of heart failure. She continued ND feeds until after she was extubated on POD 14. Her lowest recorded weight after extubation and diuresis was 3.84 kg (zscore -4.96).

Her admission was prolonged while she received milrinone, an inotrope medication for ongoing moderate to severe ventricular function, was slowly weaned, and she was uptitrated on reverse remodeling medications and diuretics were adjusted. Unfortunately,

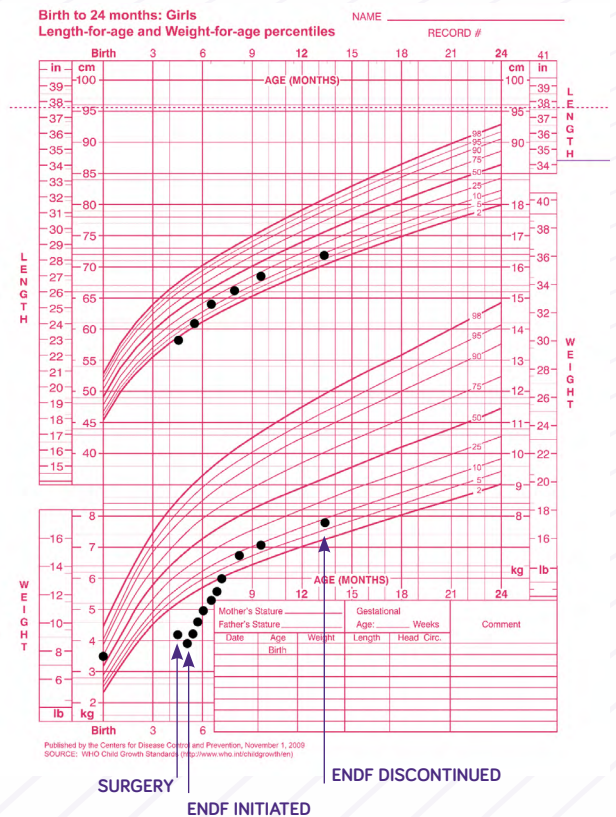


she had a prolonged admission while her doctors continued milrinone, a medication to help with heart contractions and adjusted medical management of her heart failure. She also required a slow sedation wean after a complicated postoperative course on ECMO and prolonged intubation. After her respiratory support was weaned to nasal cannula, her feeding tube was pulled from her duodenum to the stomach. After 24 hours of continuous nasogastric (NG) feeds, she was successfully transitioned to bolus feeds of 90 mL every 3 hours. Bolus feeds were initially run over a pump over 1-hour, but were condensed to run over gravity after a few days.

Baby girl R worked with a speech language pathologist to regain her oral feeding skills during her surgical recovery. She was initially allowed to take a bottle with the remainder of her goal given via NG tube. Feeds were weight-adjusted weekly to 150 kcal/kg/day². Her NG tube was removed one month after surgery once she demonstrated she could maintain adequate oral intake with ENDF. Her caloric and protein intake ad lib ranged from 130-155 kcal/kg/day and 3.4-4 g protein/kg/day, respectively, for the next 6 weeks prior to discharge. After the first 4 weeks on ENDF, she gained an average of 50 grams/day and her weight-for-age z-score improved to -3.22. Following 8 weeks of receiving ENDF, her weight-for-age z-score improved to -1.68 (5th%ile). She also consistently gained approximately 0.5 cm/week in linear growth. Her length-for-age z-score improved to -1.48 after 8 weeks. Her head circumference catch-up growth increased on average 0.5 cm/week. Most notably, her weight-for-length zscore improved from -3.07 to -0.96 after 2 months on ENDF.

Baby girl R had remarkable catch-up growth while exclusively being fed ENDF. The higher percentage of protein from ENDF likely contributed to her impressive catch-up weight and linear growth given protein needs are higher in malnourished patients to support lean body mass acquisition. Her growth contributed to her success in meeting her goals with physical therapy, and she gained the strength to meet her nutrition goals by mouth and have the feeding tube removed weeks before she was discharged. There was a period of a week after milrinone was discontinued that her cardiac output was diminished, leading to a decline in her oral intake and weight gain velocity. However, she still consistently met normal calories (100-120 kcal/kg/day) for her age by receiving ENDF and was able to meet age appropriate growth velocity goals until her cardiac function and nutrition intake improved.

Despite having moderate to severe heart failure after surgery, she had excellent gastrointestinal tolerance to feeds from day one of initiation of ENDF. Her stools were initially loose and intermittently watery but tolerable, and not unexpected while she was weaning sedation. She never required reflux medications. After her milrinone was weaned, she temporarily had some vomiting, but resolved with adjustments to her cardiac medications. Her ECHO at discharge demonstrated low-normal left ventricular function, normal right ventricular function, and mild to moderate mitral regurgitation. At the time of discharge, she began alternating feeds with ENDF and a standard cow's milk infant formula 20 kcal/fl oz due to her robust oral intake, weight gain, and improving malnutrition. She was transitioned to 100% standard infant formula. The total time of ENDF usage was 2.5 months inpatient and 1 month outpatient. ENDF was discontinued 5 weeks after discharge as her growth was normal and malnutrition resolved.



CONCLUSION

ENDF was well tolerated in a severely malnourished and critically ill infant after cardiac surgery. She seamlessly transitioned from ND to NG feeds, and was able to eventually meet her feeding goals by mouth allowing removal of the feeding tube. Use of ENDF ensured she had adequate calories, protein and micronutrients to promote wound healing shortly after surgery, support increased protein requirements during critical illness, and achieve catch-up growth despite higher nutrient requirements associated with heart failure.

REFERENCES

1. WHO. Protein and Amino Acid Requirements in Human Nutrition Report of a Joint WHO/FAO/UNU Expert Consultation. Geneva: World Health Organization, 2007. 2. Kyle UG, et al. Growth failure and nutrition considerations in chronic childhood wasting diseases. *Nutr Clin Pract* 2015;30(2):227-238.