

EnteraLite® Infinity® Helps Combat Metabolic Hypoglycemia and Other Comorbidities Associated with Glycogen Storage Disease

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The objective of this case study is to demonstrate the effectiveness of using the EnteraLite Infinity enteral feeding pump in successfully combating Metabolic Hypoglycemia in patients with Glycogen Storage Disease.

Background

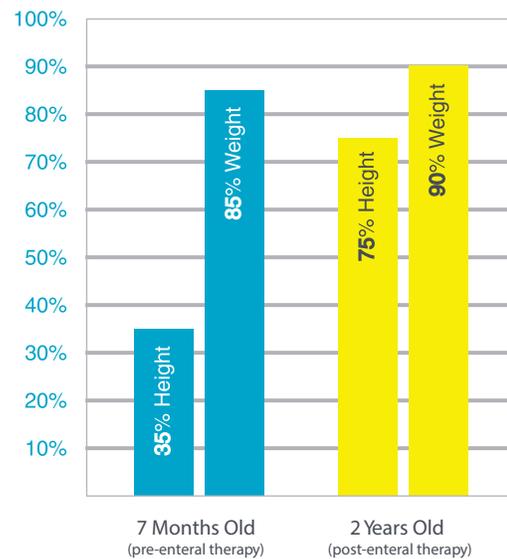
At four months of age, A.N. had a hypoglycemic seizure and was subsequently diagnosed with Glycogen Storage Disease (GSD 1a) type 1a, also known as von Gierke's disease. GSD is an autosomal recessive metabolic disorder that results in a deficiency of glycogen-6-phosphate (G-6-P) enzyme; carbohydrate metabolic pathways are blocked causing glycogen accumulation in the liver and kidneys. Patients afflicted with GSD 1a cannot maintain glucose homeostasis and manifest hypoglycemia, hepatomegaly, kidney enlargement, hyperlipidemia, hyperuricemia and lactic acidemia.

Deficiency of G-6-P blocks the final steps of glycogenolysis and gluconeogenesis resulting in severe hypoglycemia and is one of the few genetic-biochemical causes of hypoglycemia in newborns. Height and growth rate are usually subnormal.

At seven months of age, A.N. was at the 85th percentile for weight and the 35th percentile for length. She had experienced excessive weight gain on a standard infant formula and was transitioned to a soy protein isolate formula with the addition of cornstarch every three hours around the clock. To prevent dumping syndrome, the decision was made to place a gastrostomy tube and admit to home care for enteral feedings.

The patient was provided with the EnteraLite Infinity enteral feeding pump. The Infinity delivered four intermittent feedings during the day and a continuous feeding during the evening. The Infinity allowed overnight institution of uncooked cornstarch necessary for this patient to maintain a constant delivery of glucose calories to prevent hypoglycemia with the assurance that delivery flow would not be interrupted. The family found the Infinity pump was easy to use and allowed A.N. to be active during the day. Blood glucose monitoring was also easier due to the accuracy and manageability of the pump.

A.N. Growth Progress



Conclusion

Enteral therapy is a cornerstone of management for GSD, as carefully planned feedings can reduce liver size, prevent hypoglycemia, reduce symptoms and allow for growth and development. At two years of age, A.N. was at the 90th percentile for weight and the 75th for length. She is tolerating her enteral nutrition regimen well and her family is able to manage her special needs.

The EnteraLite Infinity enteral feeding pump and a precisely defined feeding schedule allowed the essential uncooked corn starch to be manageable for this child and her family.