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Congenital short bowel syndrome: A case successfully treated with home parenteral nutrition

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Purpose: Congenital short bowel associated with malrotation and intestinal dysmotility is a quite rare condition with poor prognosis. The aim of this report is to describe a case complicated with pyloric stenosis as a rare associated anomaly and successfully treated with home parenteral nutrition.

Method: A 5-day-old newborn girl with a small intestinal length of 45 cm, malrotation with volvulus, and umbilical hernia, who underwent Ladd's procedure. Total parenteral nutrition was initiated postoperatively with a small amount of enteral feeding with elemental diet. After her weight became more than 5 kg, home parenteral nutrition was started. Parenteral nutrition was weaned depending on the amount of oral intake and her nutritional status.

Results: Although non-bilious vomiting was frequently observed, diagnosis of gastroesophageal reflux due to pyloric stenosis was made at 3 years old. Operative finding indicated no membranous obstruction and muscular hypertrophy, and she underwent pyloroplasty. After the operation, the amount of oral intake gradually increased, and home parenteral nutrition was discontinued at age 5 years. And then, for the past 4 years, her growth and nutritional conditions has been maintained within normal range.

Conclusion: Although early onset within first week of life has been reported to have a fatal outcome, meticulous management of intravenous alimentation and early introduction of enteral feeding may be important for survival.