# Congenital Short Bowel Syndrome with Malrotation

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Congenital short bowel syndrome (SBS) associated with malrotation and malabsorption is a very rare condition. We report on an infant girl with congenital SBS associated with malrotation and malabsorption. No polyhydraminos was noted during the regular prenatal examination. Protracted postnatal postprandial vomiting with progressive failure to thrive was noted. A laparotomy showed the small bowel was only about 20 cm in length. She eventually survived with short-term parenteral nutrition and use of oral L-glutamine supplementation. To our knowledge, this might be the shortest length of bowel loop ever reported. Currently, she is 15 months of age with a body weight of about 7 kg and good development. *(Chang Gung Med J 2004;27:548-50)*

**Key words: short bowel syndrome, malrotation, parenteral nutrition, glutamine.**

Congenital short bowel syndrome (SBS) associated with malrotation and malabsorption is a very rare condition. Since 1969, only 18 patients with this syndrome have been reported.[1] Among the 18, only 4 patients survived. We present one additional patient who was successfully treated in our institution. The mainstay of management is postoperative parenteral alimentation followed by a very gradual oral feeding regimen.[2]

## CASE REPORT

A baby girl was delivered vaginally at term after a normal pregnancy and labor. Her mother received regular prenatal examinations and no polyhydramnios was noted. The birth weight was 2.6 kg (appropriate to gestational age). She was initially admitted to another hospital at the age of 4 days with a history of frequent postprandial bilious vomiting. Abdominal plain X-ray film showed diffuse dilatation of the small intestine. A lower gastrointestinal series (LGI) showed the caliber of colon was normal with no transitional zone, but obvious obstruction was noted at the level of the ileocecal junction. She was then transferred to our neonatal intensive care unit with a presumptive diagnosis of intestinal obstruction. Repeated abdominal X-ray examinations showed persistently dilated bowel loops with an air-fluid level. A laparotomy was performed on the 6th day after admission (10 days old). During laparotomy, an adhesion band over the ileocecal region was

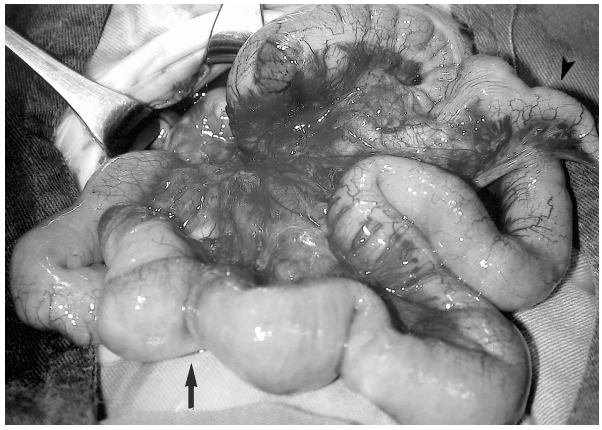

**Fig. 1** Operative photography shows a short dilated small bowel with malrotation (black arrow) and the appendix (arrowhead).

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noted, the small bowel was very foreshortened and malrotation of the small bowel was also noted (Fig. 1). The total length of the small bowel was only about 20 cm.

In the postoperative period, this patient received total parenteral nutrition (TPN) through a central venous (right subclavian vein) catheter. Initiation of oral feeding with glucose water (10% dextrose water) was given about 7 days after surgery and then progressively changed to elementary formula (Progestimil®, Meadjohson Nutritionals Nijmegen, the Netherland). The patient could tolerate a small feeding of 30 cc per meal every 4 hours, but suffered from diarrhea frequently. In addition to the elementary diet, L-glutamine (Sympt-X®, Baxter Healthcare Corporation; Deerfield, IL USA), lactomin (Biofermin®) and beta-galactosidase (Galantase®) were also added to enhance intestinal absorption. The dose of L-glutamine was 0.5 mg/kg/day divided into 3 doses with milk. The TPN was discontinued about 5 months later. The baby grew smoothly and the absorptive function also improved. She was discharged at the age of 7 months with a weight of 5.4 kg (< 3rd percentile). Currently, she is 15 months of age with a weight of 7 kg (< 3rd percentile).

## DISCUSSION

Congenital SBS is a very rare disease. Its etiology, unlike the well-documented acquired SBS, remains unknown. Some authors have proposed a few possible etiologies, including antenatal midgut volvulus and/or vascular injury.[3] Another hypothesis is antenatal intussusception. Familial aggregations have also been reported.[4] Since Hamilton's original report in 1969[1] of 2 sisters with malrotation and congenital short small intestine, there have been 16 other cases reported in the literature. Laparotomy is essential to obtain a definite diagnosis. Patients with congenital SBS described in the literature presented with symptoms related to obstruction, such as bilious vomiting and distended abdomen, mostly caused by dysmotility of the small bowel. The explanation for the functional intestinal obstruction remains unclear. Malrotation with Ladd's band that causes distal bowel obstruction, as shown in our case, has been reported.[2]

Parenteral nutrition should be given initially. Once patients resume intestinal motility, enteral feeding is suggested. Extensively hydrolyzed formula (e.g., Progestimil®) is preferred. If feeding intolerance is present, complete amino acid-based formula may be helpful. Bines et al.[5] reported that this formula could improve diarrhea, reduce vomiting, decrease intestinal permeability to lactulose and increase the disaccharidase level in the intestine. Glutamine also has beneficial effects because it enhances immunity in several critical conditions and enhances intestinal absorption.[6] Glutamine is often referred to as a gut trophic factor and a conditional essential amino acid.

Long-term complications of congenital SBS are catheter-associated sepsis, parenteral nutrition-associated liver disease, local bacterial overgrowth, feeding intolerance, and nutritional imbalance.[2] The survival rate of patients with congenital SBS remains unknown. Our patient, with a small bowel of only 20 cm, survived and achieved normal growth and development following TPN and early enteral feeding with introduction of glutamine treatment. To our knowledge, this might be the shortest length of bowel loop reported in cases with congenital SBS. We conclude that intravenous alimentation along with very gradual oral feeding, perhaps including glutamine, may be the mainstay of therapy for congenital SBS. In cases with malrotation, surgical treatment is mandatory and lifesaving.

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# 先天性短腸症候群合併腸扭轉

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先天性短腸症候群合併腸扭轉及吸收不良是相當罕見的先天性腸道異常，我們報告一個女嬰病例，她的母親接受規則的產前檢查，但並未發現有羊水過多的現象，出生後是以嚴重的嘔吐、生長遲緩以及吸收不良來表現，同時越來越嚴重；在接受腹腔探查手術後發現她的小腸總長度僅有20公分，同時合併有腸扭轉，我們在施行小腸固定手術後給予短期的靜脈營養補充，同時並給予L-glutamine補充在完全水解蛋白配方牛奶中，最後這位女嬰存活下來。目前這位女嬰是15個月大，體重7公斤，發展情況良好。從文獻的探尋中，這個病歷應該是目前所知先天性短腸症的病例中，小腸長度最小的病例。(長庚醫誌 2004;27:548-50)

**關鍵字**：短腸症，腸扭轉，靜脈營養，麩胺。

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