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Congenital short bowel syndrome

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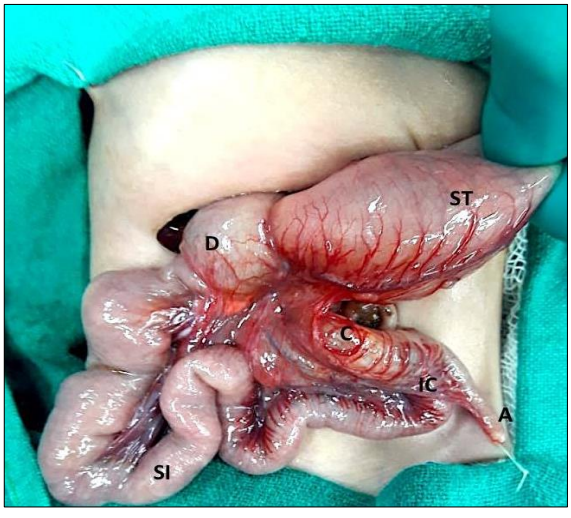

Figure 1: Congenital short bowel with a length of 25cm. ST: stomach, D: duodenum, SI: small intestine, A: appendix, IC: ileocecal valve, C: colon.

A 33-week male preterm neonate with a low birth weight of 1.75kg who was born to a 25yr-old mother, through non-consanguineous marriage, was admitted to our neonatal intensive care unit (NICU). He had passed meconium immediately after birth and feeds were started through an orogastric tube within 2 hours of life. On day 2 of life, the baby had bilious vomiting with abdominal distension and visible bowel loops, followed by persistent bilious aspirates. On evaluation, the septic screen was negative, plain radiograph abdomen showed dilated bowel loops and ultrasound abdomen confirmed the same finding, but the relationship between superior mesenteric artery and vein could not be defined due to the gaseous distension. The baby was taken up for exploratory laparotomy in view of persistent bilious aspirates and feed intolerance. Intraoperative findings revealed a foreshortened small intestine, which was only about 25 cm in length and had malrotation with ileocecal junction in the left upper quadrant (Fig. 1). There was no atresia and the bowel was dilated till the rectum.

Congenital short bowel syndrome (SBS) is a rare intestinal disorder with only 62 cases reported in the literature to date. Normally, the bowel length doubles from 28 to 40 weeks of gestation; the normal small bowel length of a term neonate is usually 250cm, and SBS manifests when the small bowel length is less than 75cms. [1] The shortest reported bowel length was 20cm. [2] The etiopathogenesis is poorly understood and several theories have been proposed including delayed or interrupted intestinal elongation, ischemic injury, or antenatal volvulus, and defective neuro-enteric development with intestinal dysmotility. [1] Familial occurrences were noted in 60% of reported cases with two genes identified in these patients (CLMP gene (61%) with autosomal recessive inheritance and FLNA gene (39%) with X-linked inheritance). [3]

Antenatally, there was no polyhydramnios in these patients, making antenatal diagnosis difficult. [2] Two-third of them had early, acute presentation with intestinal obstruction, who mostly die before 1 year of age. One-third of them had a late, subacute presentation with chronic diarrhea, vomiting, abdominal distension, and failure to thrive. [4] Diagnosis may be suggested by upper GI contrast studies but is confirmed by exploratory laparotomy. [5] Malrotation is seen in all patients except one, but volvulus with gangrene is an infrequent finding. [1] Other associations of congenital SBS are appendicular agenesis, IHPS, bowel adhesions or bands, hemivertebra, dextrocardia, and PDA. [6] Malabsorption and dysmotility are the principal hallmarks of CSBS, making them dependent on prolonged TPN. Surgical options available to increase the bowel length and absorption are serial transverse enteroplasty (STEP) or longitudinal intestinal lengthening and tailoring (LILT), and bowel transplantation.

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