Letter to the Editor

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Poor outcomes in congenital short bowel syndrome with intestinal malrotation: Experience from a developing country

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DEAR SIR

Congenital short bowel syndrome is a rare gastrointestinal disorder characterized by the need for prolonged parenteral nutrition for more than 60 days or the presence of a small bowel measuring less than 25% of the expected length. Ultrashort bowel syndrome is defined as less than 10 cm or less than 10% of the expected length of small bowel for age. [1] The prognosis has improved as medical care and parenteral nutrition have advanced. Nevertheless, the mortality rate remains high. [2]

From January 2000 to December 2022, three infant boys and one girl were diagnosed with congenital short bowel syndrome with malrotation. They were 15, 27, and 30 days old (2 infants). They all had normal pregnancies, ultrasonography, and perinatal courses. Bilious vomiting was the constant alarming symptom. Upon examination, the abdomen was flat in one patient and distended in three with facial dysmorphic features in two patients.

Abdominal X-ray revealed small intestine dilation with multiple air-fluid levels. An abdominal ultrasound was performed in two patients revealing significant gastric dilation; Doppler USG showed normal mesenteric vessels relation. Given the absence of a distended abdomen and isolated gastric dilation, one patient required an upper gastrointestinal series. It revealed a distended stomach with apparent short bowels.

After initial resuscitation, an exploratory laparotomy was performed and the diagnosis of midgut malrotation was confirmed. Although the reduced intestine was viable, an ultra-short bowel syndrome was discovered. The length was 20 cm in one, 30 cm in two, and 35 cm in the fourth (Fig.1).

Due to oral nutrition intolerance, they all received exclusive parenteral nutrition via a central right jugular vein catheter. However, all infants died before discharge after an average of 12.75 days due to sepsis and malabsorption (at 7 days in two infants, 12 days in one, and 25 days in one). The histological examination revealed no abnormalities of the appendix with normal ganglion cells and histological lesions of cystic fibrosis (meconium ileus). The short bowel syndrome in our patients may result from the in-utero volvulus.

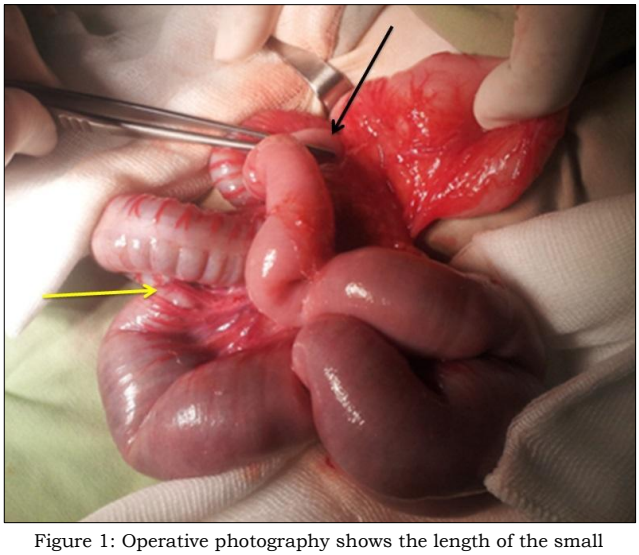

Figure 1: Operative photography shows the length of the small intestine measured from the pylorus less than 30 cm: black arrow shows the pylorus; the yellow arrow shows the ileocecal junction.

The poor prognosis in infants with ultra-short bowel syndrome is secondary to the low absorptive capacity and lethal intestinal failure due to the slow development of intestinal adaptation mechanisms. [2] They require chronic parenteral nutrition as well as gradual oral feeding with hypo osmolar preparations to enhance absorptive capacity and achieve autonomy. [3] Some studies using hormonal modulation have also yielded promising results. [1, 3] Our country still lacks specialized centers that explain the poor prognosis in our cases.

Many surgical options have been proposed to lengthen the intestine and slow its motility. It

consists of a Longitudinal Intestinal Lengthening and Tailoring procedure, a Serial Transverse Enteroplasty procedure, and the Segmental Reversal of the Small Bowel. Otherwise, intestinal transplantation may represent the last resort though it is associated with severe complications which limit its application. [4]

To summarize, survival in infants with short bowel syndrome primarily depends on advances in medical management and the establishment of specialized centers. Further genetic research may aid in genetic counseling, as the genetic basis of familial aggregations has been described. [2,5] Moreover, a prenatal diagnosis could potentially advance fetal procedures and increase long-term survival.

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REFERENCES

1. Dogra S, Peters NJ, Samujh R. Short bowel syndrome in neonates and early infancy. J Neonatal Surg. 2023; 12:8.
2. Negri E, Coletta R, Morabito A. Congenital short bowel syndrome: Systematic review of a rare condition. J Pediatr Surg. 2020; 55:1809-14.
3. Wang Y, Chen S, Yan W, Lu L, Tao Y, Xiao Y, et al. Congenital short-bowel syndrome: Clinical and genetic presentation in China. JPEN J Parenter Enteral Nutr. 2021; 45:1009-15.
4. Goulet O, Abi Nader E, Pigneur B, Lambe C. Short bowel syndrome as the leading cause of intestinal failure in early life: Some insights into the management. Pediatr Gastroenterol Hepatol Nutr. 2019; 22:303-29.
5. Van der Werf CS, Halim D, Verheij JBGM, Alves MM, Hofstra RMW. Congenital short bowel syndrome: from clinical and genetic diagnosis to the molecular mechanisms involved in intestinal elongation. Biochim Biophys Acta. 2015; 1852:2352-61.