Case report

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
48(9) 1–4

DOI: 10.1177/0300060520954726
journals.sagepub.com/home/imr
SAGE

Congenital short bowel syndrome: a rare cause of neonatal intestinal obstruction

Mostafa Zain, Mansour Abdelkader, Ahmed Azab and Mostafa Kotb

Abstract

Congenital short bowel syndrome (CSBS) is an uncommon gastrointestinal disorder in which an unclear aetiology causes considerable intrauterine reduction in small bowel length. As a result of reduced absorptive intestinal length, chronic diarrhoea, vomiting, and consequently, failure to thrive are likely. We report a case of CSBS in a 26-day-old girl who had malrotation and a short bowel with a length of bowel from the pylorus to the ileocecal junction of approximately 40 cm. The patient underwent Ladd's procedure, but she is still dependent on parenteral nutrition.

Keywords

Congenital short bowel syndrome, malrotation, diarrhoea, vomiting, infant, paediatric surgery, parenteral nutrition

Date received: 14 July 2020; accepted: 10 August 2020

Introduction

The precise definition of short bowel syndrome (SBS) has not been fully determined. Nevertheless, SBS is best defined as a multisystemic disorder that is caused by malabsorption of nutrients, resulting from a diminished small intestinal length of less than half of the expected length for gestational age.[1] Fortunately, SBS is rare, but it is a serious condition. Most cases of SBS are secondary to a pathology, such as necrotising enterocolitis, gastroschisis, midgut volvulus, or intestinal atresia.[2] Congenital short bowel syndrome (CSBS)

Pediatric Surgery, Alexandria Faculty of Medicine, Alexandria, Egypt

Corresponding author:
Mostafa Kotb, Pediatric Surgery Unit, Alexandria Faculty of Medicine, 45 Champlion Street, Azarita Medical Campus, Alexandria 21131, Egypt.
Email: mostafa.rashad@alexmed.edu.eg

is an exceedingly rare subtype, where to the best of our knowledge, only 62 cases have been reported in the English literature to date.[3] We report here a case of CSBS in a 26-day-old girl who had malrotation and a short bowel from the pylorus to the caecum of approximately 40 cm.

Case report

A 26-day-old girl presented to our department with persistent vomiting, which started initially as non-bilious and became bilious in the previous 2 days. She was delivered via caesarean section at 33 weeks of gestation to nonconsanguineous parents and her birth weight was 2.25 kg. Her parents reported that they sought medical advice 2 weeks previously because of the persistent non-bilious vomiting and received supportive treatment, but it was not successful. On examination, she showed signs of dehydration and failure to thrive, and weighed only 1.6 kg. The abdomen was distended and non-tender.

An upper gastrointestinal series showed a hugely dilated stomach with secondary reflux up to the level of the cervical oesophagus and malrotation, where the duodenojejunal flexure was positioned in the right midabdomen (Figure 1). After resuscitation, the patient was prepared for surgery.

Exploration confirmed malrotation, and consequently, she underwent Ladd's procedure. The small bowel length was 40 cm from the pylorus to the ileocecal junction (Figure 2). The length of the small bowel was assessed from the pylorus because the duodenojejunal junction was not clearly identifiable after kocherisation of the duodenum. After 3 months of follow-up, the patient had a small increase in weight (approximately 120 g) and still suffers from infrequent attacks of non-bilious vomiting every several days. She is currently receiving enteral feeding via a gastric tube, which started at a low concentration and increased to 0.67 kcal/mL. However, the patient is still dependent on parenteral nutrition.

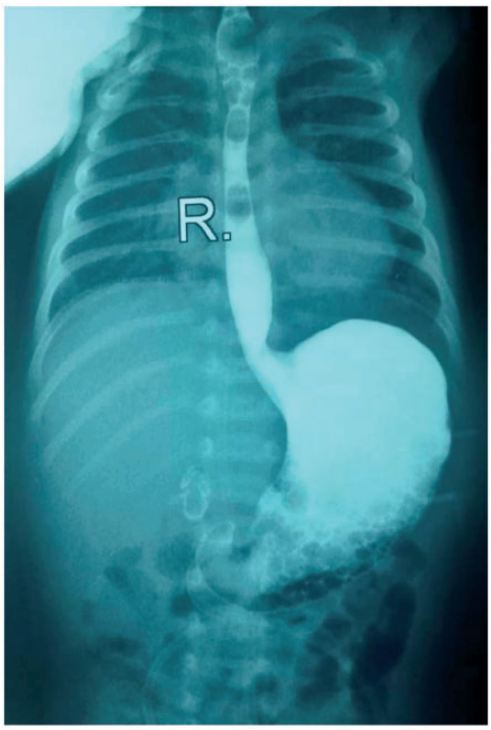

Figure 1. Upper gastrointestinal series showing a dilated stomach and malrotation, where the duodenojejunal flexure is positioned in the right midabdomen.

Discussion

SBS is a complication of many paediatric surgical conditions, such as necrotising enterocolitis, gastroschisis, midgut volvulus, and intestinal atresia.[2] CSBS is a small bowel shortage that is present since birth with preserved bowel continuity and it is an exceedingly rare subtype.[3]

There is no consensus on the definition of SBS. However, a bowel length of less than 100 cm in the first year can establish the diagnosis of SBS and less than 40 cm traditionally indicates therapy in most

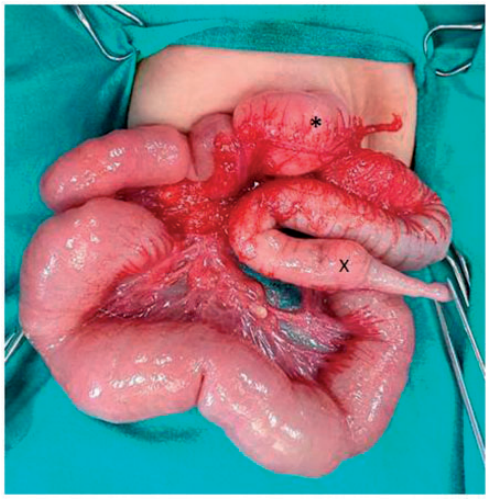

Figure 2. Intraoperative photograph showing a dilated stomach (*) and malrotation with the ileoceacal valve (x) on the left side beside the stomach.

centres.[4] Some centres consider gestational age, rather than absolute numbers, for SBS, and clearly define SBS as a diminished small intestinal length of less than half of the expected length for gestational age.[1]

Most CSBS cases present with diarrhoea, vomiting, failure to thrive, or manifestations consistent with intestinal obstruction.[5] Malrotation is a relatively common feature in CSBS, where it was encountered in all but one case.[3,6] Malrotation could be a possible cause of early embryological insult and it subsequently results in impairment of growth of the midgut with resultant vascular occlusion.[7] Because malrotation is almost always present in CSBS, an upper gastrointestinal contrast study should be performed to confirm its presence.[3] If CSBS is present, Ladd's procedure should be performed to prevent intestinal obstruction and possible volvoulus.[3]

The mainstay treatment of CSBS cases is parenteral nutrition with an attempt of early introduction of enteral feeding.[8] Although parenteral nutrition is crucial for providing essential calories for growth, parenteral delivery has potential complications, with sepsis as the main cause for morbidity and mortality.[9] Early in the postoperative period for CSBS, an adaptive response typically occurs, which increases the length and absorptive capacity of the remaining bowel. This process alone is often sufficient to provide intestinal autonomy. However, when adaptation is insufficient to achieve enteric autonomy, several surgical options, such as bowel lengthening procedures can be performed, and may obviate the need for intestinal transplantation.[10] The prognosis of CSBS highly depends on the length and function of the remaining segment of the small bowel.[11] Regrettably, only one third of patients with CSBS have survived to an average age of 5.8 years and most of the deaths occurred before 1 year of age.[6,12] Fortunately, reports have shown a significantly improved survival rate for patients with CSBS in the past 20 years.[6] This is likely due to improvement in the standards of parenteral nutrition therapy along with development of novel formulas, which not only promote early administration of enteral feeding, but also facilitate intestinal adaptation.[13]

In summary, CSBS is a rare condition that may present with symptoms of small bowel obstruction in the first weeks of life. An upper gastrointestinal study can confirm the presence of malrotation and suggest a short bowel length. Nevertheless, the diagnosis of CSBS is typically established in laparotomy.

Author contributions

All authors contributed equally to the manuscript and read and approved the final version of the manuscript.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

Ethics committee approval was not obtained because this was a case report and no research protocol was applied for. Written and verbal informed consent for publication was obtained from the patient's parents.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Mostafa Kotb https://orcid.org/0000-0002-2558-3551

References

1. Khalil BA, Ba'ath ME, Aziz A, et al. Intestinal rehabilitation and bowel reconstructive surgery: improved outcomes in children with short bowel syndrome. *J Pediatr Gastroenterol Nutr* 2012; 54: 505–509.
2. Wales PW and Christison-Lagay ER. Short bowel syndrome: epidemiology and etiology. *Semin Pediatr Surg* 2010; 19: 3–9.
3. Negri E, Coletta R and Morabito A. Congenital short bowel syndrome: systematic review of a rare condition. *J Pediatr Surg* 2020: S0022-3468(20)30205-0. doi.org/10.1016/j.jpedsurg.2020.03.009.
4. Soden JS. Clinical assessment of the child with intestinal failure. *Semin Pediatr Surg* 2010; 19: 10–19.
5. Palle L and Reddy B. Case report: Congenital short bowel syndrome. *Indian J Radiol Imaging* 2010; 20: 227–229.
6. Hasosah M, Lemberg DA, Skarsgard E, et al. Congenital short bowel syndrome: a case report and review of the literature. *Can J Gastroenterol* 2008; 22: 71–74.
7. Kern IB and Harris MJ. Congenital short bowel. *Aust N Z J Surg* 1973; 42: 283–285.
8. Vanderhoof JA. Short bowel syndrome and intestinal adaptation. In: Walker WA, Durie PR, Hamilton JR, Walker-Smith JA and Watkins JB (eds) *Pediatric Gastrointestinal Disease: Pathophysiology, Diagnosis, Managment*. 3rd ed. Hamilton: BC Decker, 2000, pp.583–602.
9. Calkins KL, Venick RS and Devaskar SU. Complications associated with parenteral nutrition in the neonate. *Clin Perinatol* 2014; 41: 331–345.
10. Sommovilla J and Warner BW. Surgical options to enhance intestinal function in patients with short bowel syndrome. *Curr Opin Pediatr* 2014; 26: 350–355.
11. Gharesouran J, Esfahani BS, Valilou SF, et al. First report of congenital short bowel syndrome in an Iranian patient caused by a mutation in the CLMP gene. *J Pediatr Genet* 2019; 8: 73–80.
12. Howard L, Ament M, Fleming CR, et al. Current use and clinical outcome of home parenteral and enteral nutrition therapies in the United States. *Gastroenterology* 1995; 109: 355–365.
13. Boitano M, Bojak S, McCloskey S, et al. Improving the safety and effectiveness of parenteral nutrition: results of a quality improvement collaboration. *Nutr Clin Pract* 2010; 25: 663–671.