Short Bowel Syndrome: An Uncommon Clinical Entity and a Therapeutic Challenge—Our Experience and Review of Literature

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Abstract Short bowel syndrome (SBS), one of the commonest types of intestinal failure, usually secondary to extensive bowel resection, traditionally has been associated with a high mortality rate and hence a big challenge for the treating surgeons. It requires comprehensive clinical care to minimise the morbidities and mortality associated with the condition. We report a retrospective review of a series of seven patients with SBS, who presented at our surgical emergency within a period of 1 year and their outcome so as to encourage others in managing such a challenge with more positive mindsets. A retrospective analysis of seven patients with

SBS admitted from January 2014 to January 2015 with a follow-up of 1 year has been done in terms of their demographic characteristics, underlying pathology and clinical outcome. A rising incidence of SBS in the younger age group (71.4%) has been observed in this analysis. Majority of patients (57.1%) had mesenteric ischemia as the underlying cause followed by each case of small bowel volvulus, internal herniation and blunt trauma abdomen. A discharge rate of 71.4% and mortality rate of 28.5% were observed. With this analysis, we believe that SBS is no more an uncommon condition. A structured clinical approach, timely surgical intervention and multidisciplinary postoperative management are essential for managing such frail patients to achieve best possible results. This will encourage others in managing such a critically challenged condition with a more positive approach and thus beneficial for both the patients and the treating surgeon.

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Introduction

Short bowel syndrome (SBS), one of the pathophysiological types of intestinal failure, is defined as length of less than 100–120 cm of small bowel without a preserved ileocecal (IC) junction or as less than 60 cm of remnant small bowel with intact IC junction leading to inadequate digestive and absorptive capacity [1]. It demands comprehensive clinical care to overcome the associated morbidity and mortality. The emerging options of dietary approaches, pharmacotherapies, parenteral nutrition and timely surgical interventions have led to better management of such patients and improved their

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survival outcome; however, the mortality rate still remains high (30 to 40%) [2].

We report a series of seven cases of SBS admitted in our tertiary care hospital in a period of 18 months, illustrating their demographic and etiological aspects as well as short-term outcome. This throws light on the increasing incidence of the condition, the changing demographic trends as well as improving outcome with a holistic management approach.

Method

We did a retrospective analysis of seven cases of SBS from January 2014 to January 2015 admitted at the surgical emergency in one of the surgical units of our hospital with a follow-up for 1 year. They were analysed in terms of their demographic characteristics, underlying aetiology as well as clinical outcome. All of them presented at the surgical emergency with acute abdomen secondary to various underlying primary conditions and required emergency laparotomy and bowel resection leading to SBS.

Results

All of the seven patients were males, of whom five belonged to the age group of 18–24 years while two were in their sixth decades (Table 1). The duration between the onset of symptoms and time of presentation varied among the patients: two presented around 5 to 7 days from the onset of symptoms of acute abdomen and four patients presented within 4 days of onset of their symptoms. One patient was referred to our centre with anastomotic leak and peritonitis following exploratory laparotomy and ileo-ileal anastomosis done at a peripheral centre for a traumatic ileal perforation secondary to blunt abdominal trauma and had presented to us 4 days after the primary surgery. The patients were operated on the day of admission after adequate resuscitation and necessary preoperative diagnostic evaluation. Majority of the cases were secondary to mesenteric ischemia (four patients, 57.1%), while the rest were due to small bowel volvulus, internal herniation and blunt trauma abdomen (Table 1).

The cause of mesenteric ischemia was coagulopathy in two patients, venous thrombosis in one patient and atherosclerosis in one patient. Out of seven, six patients required jejunostomy during the primary surgery (Table 1). The ileocecal junction (IC) could not be preserved in four patients. The length of the remnant small bowel among our patients ranged from 20 to 85 cm (average 50.57 cm).

Of the seven patients, one expired in the immediate postoperative period secondary to septicemia, and another patient expired in the fifth postoperative month secondary to complications of intestinal failure despite necessary management

(Table 1). In the rest five patients, total parenteral nutrition (TPN) was administered for 8–12 weeks until the restoration of intestinal continuity was done. After the second surgery, they required TPN for 10–14 days while they were started on oral nutritional supplementation trials with high-protein diet along with commercially available supplemental elemental formula feeds gradually from day 7 along with intermittent TPN. All of the rest five patients were discharged from the hospital after 3–4 weeks on complete oral diet with required nutritional advice and were followed up to a period of 1 year, with frequent visits with once a biweekly visit for initial 6 months and then monthly visits. However, two of these patients required readmissions in view of dyselectrolytemia which was corrected. During each follow-up visit, they were given parenteral multivitamin and mineral supplements. They were advised acid-reducing agents (proton pump inhibitors), anti-secretory agents (e.g. octreotide) and antidiarrheal medications (loperamide, diphenoxylate) along with special nutritional guidelines such as small frequent meals involving less intake of dietary fibres and carbohydrates with more fatty content, avoidance of high-oxalate food, restricted fluid intake with meals and intake of isotonic fluid such oral rehydration solution, separate solid and liquid intake as much as possible, mineral and vitamin supplement etc. They were discharged and were followed up for 1 year. Thus, we observed discharge rate of 71.4% and a 1-year mortality of 28.5%.

Discussion

Short bowel syndrome (SBS) is the inability to maintain fluid, electrolyte and protein-energy or micronutrient balances while on normal diet due either to inadequate digestion or impaired absorption of nutrients. Such a dysfunction in the adult age group is usually the result of surgical bowel resections required for various pathologies like catastrophic mesenteric vascular events, adhesive obstruction, volvulus, Crohn's disease, malignancy and trauma. About 15% patients who undergo bowel resection develop SBS [3].

SBS has been found to be the major cause of chronic intestinal failure primarily due to reduced intestinal absorptive surface area and found to affect about 75% of patients requiring home-based parenteral nutrition for chronic intestinal failure [4].

Although the published literature on SBS is very exhaustive, there is no consensus about the exact incidence of the disease. The most appropriate estimation for this has been from the number of patients receiving long-term parenteral nutrition and/or intravenous fluid support excluding the weaned off ones and those who did not receive. On this basis, the incidence and prevalence in Europe have been estimated to be 2–3 per million and 4 per million, respectively [5–7].

Table 1 Demographic features and operative details of the cases

Case no.	Age (years)	Sex (M/F)	Preoperative diagnosis	Primary surgery	Restoration of bowel continuity	Resected bowel length	Remnant bowel length	IC preserved or not	Postoperative course
1.	18	M	Postoperative case of exploratory laparotomy done for blunt trauma abdomen with anastomotic leak.	Exploratory laparotomy with adhesiolysis with jejunostomy with distal mucus fistula.	–	300 cm of the small bowel.	20 cm of the jejunum and 10 cm of the ileum.	Preserved	Expired
2.	52	M	Acute small bowel obstruction with mesenteric ischaemia.	Exploratory laparotomy with resection of the mid and distal jejunum and whole of the ileum and ascending colon with end jejunostomy.	Jejunostomy closure via side to side jejuno-colic (transverse colon) anastomosis.	350 cm of the small bowel and ascending colon.	20 cm of the jejunum.	Not preserved	Discharged
3.	23	M	Acute small bowel obstruction with internal herniation.	Exploratory laparotomy with resection of distal two-third of the jejunum and whole of the ileum with end jejunostomy.	Jejunostomy closure with jejuno-colic side to side (ascending colon) anastomosis.	300 cm of the small bowel.	80 cm of the jejunum and 5 cm of the ileum.	Not preserved	Discharged
4.	24	M	Acute small bowel obstruction with mesenteric ischaemia.	Exploratory laparotomy with resection of the jejunum except proximal 4 cm and proximal four-fifth of the ileum with jejunostomy.	Jejunostomy closure with side to side duodeno-ileal anastomosis.	350 cm of the small bowel.	4 cm of the jejunum and 60 cm of the ileum.	Preserved	Discharged
5.	24	M	Acute small bowel obstruction with ileal volvulus.	Exploratory laparotomy with resection of distal two-third jejunum and whole of the ileum with end jejunostomy.	Jejunostomy closure with side to side jejuno-colic (ascending colon) anastomosis.	350 cm of the small bowel.	60 cm of the jejunum and 5 cm of the ileum.	Not preserved	Discharged
6.	22	M	Postoperative case of exploratory laparotomy done for small bowel obstruction with mesenteric ischemia.	Exploratory laparotomy with right-sided hemicolectomy and resection two-third of the small bowel with side to side jejunocolic anastomosis.	–	350 cm of the small bowel caecum, ascending colon and hepatic flexure.	60 cm of the jejunum.	Not preserved	Discharged
7.	55	M	Acute small bowel obstruction with mesenteric ischemia.	Exploratory laparotomy with resection of distal three-fourth jejunum and entire ileum caecum and ascending colon with jejunostomy.	–	400 cm of the small bowel caecum and ascending colon and jejunostomy.	30 cm of the jejunum.	Not preserved	Expired

The spectrum of dysfunction and thus the postoperative prognosis of patients following the bowel resection vary according to the length of the bowel segment removed, residual bowel segment, presence or absence of ileocecal junction, the primary aetiology, presence of any comorbidities, age of the patient and the associated liver disease. However, the primary determinant of the outcome of SBS is the remnant bowel length. SBS commonly develops in those with less than one-third length of the remaining small intestine [8]. The common procedures associated with SBS are end jejunostomy, jejuno-ileal anastomosis and jejunocolic anastomosis. The most commonly done is jejunocolic and the most challenging to manage is end jejunostomies [8].

In cases of doubtful viability or intra-abdominal sepsis after primary surgery, there may be a need for second-look surgery. “Second look laparotomy” was introduced by Shaw, in 1965 to assess adequacy of bowel resection and is considered to be the gold standard for determining further bowel viability [9, 10]. Second-look laparotomy is one of the factors responsible for reduction in mortality rate associated with acute mesenteric ischemia.

Almost all patients of SBS depend upon parenteral/intravenous (PN/IV) nutritional support but in later postoperative period, the process of intestinal adaptation helps in weaning off the PN/IV support. This process of adaptation begins within 12 to 24 h after resection and continues for 1 to 2 years during which the majority of structural and functional changes occur. Permanent dependence on total PN occurs with a small bowel length of <60 cm with ileocecal junction or <120 cm without ileocecal junction.

Surgical options for management of irreversible short bowel syndrome include (A) primarily to increase intestinal absorptive capacity and (B) intestinal transplantation. The former group includes (a) procedures to correct slow transit such as creation of valves [11]; (b) procedures to improve intestinal motility in cases of dilated bowel loops like tapering enteroplasty, longitudinal intestinal lengthening and tailoring (LILT) procedure [12], serial transverse enteroplasty (STEP) and intestinal plication [13]; (c) procedures to slow intestinal transit in the absence of bowel dilatation such as segmental reversal of the small bowel (SRSB) [14] and isoperistaltic colonic transposition; and (d) procedures to increase mucosal surface area like sequential intestinal lengthening and controlled tissue expansion [15]. These procedures have been tried mostly in paediatric age group, and of these, creation of valves and intestinal plication are of historical interest only. Intestinal transplantation is the present standard of care in whom all the bowel rehabilitation measures are exhausted and who are at life-threatening complications of parenteral nutrition [16, 17].

Pironi L et al. conducted a large multicentre survey on 688 adult patients on home parenteral nutrition for benign

intestinal failure, out of which 75% had SBS; the mean age of the study population was 52.9 years with a standard deviation of 15 and was found to be more common in females with the most common underlying condition being mesenteric ischaemia [4]. In another study by Amiot A et al., done over a period of 33 years on 268 patients, the median age reported was 52.5 years, with a majority of female patients, and the most common aetiology being mesenteric ischemia [18]. In our study, however, all the patients were males and majority of them belonged to a younger age group (median age of 24 years).

In a study of 210 patients over a period of 20 years by Thompson J S et al., the underlying primary conditions were postoperative complications (such as postoperative obstruction secondary to adhesion and volvulus, postoperative intestinal ischemia) (25%), irradiation/malignancy (24%), mesenteric vascular disease (22%), Crohn's disease (16%) and other causes (13%) [19]. The most common underlying cause in our study population was mesenteric ischemia secondary to atherosclerosis in two patients belonging to the older-age group and coagulopathy in the two patients of younger-age group.

In the study of Amiot A et al., most patients had undergone jejunocolic anastomosis (67%), while 18% had end jejunostomies and 15% underwent jejunocolic anastomosis [19]. In our series of the seven patients, four underwent jejunocolic anastomosis for restoration of bowel continuity, while one underwent duodeno-ileal anastomosis. There is usually a very low survival rate associated with SBS. Study done by Reinshagen K et al. showed the 1- and 5-year survival in patients on PN 62 to 86% in adults [20]. We observed a lower mortality rate of 28.5% which could be most probably due to majority of patients belonging to younger-age group with no or minimal comorbidities. With the recent advances in the field of nutritional support, good nursing care, surgical interventions and multidisciplinary approaches, the discharge rate from hospital has come up to 70% [20] which was close to that of our study (71.4%).

Conclusion

With the experience of the seven patients admitted to us within a very short span, we believe that SBS is no more an uncommon entity and that there is a changing demographic pattern with increasing presentation in the younger-age group, especially among males. With early detection of the pathology, timely surgical approach and comprehensive postoperative nutritional replacement along with good nursing care the patients with SBS are having better prognosis. Such patients require a structured approach to care with patience, persistence and positive outlook by a dedicated surgical unit in order to achieve the best possible outcome.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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