A description of the management and outcomes of infants with short bowel syndrome in a South African context

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Background. Intestinal failure because of congenital or acquired massive bowel loss is an extremely difficult problem to manage and has traditionally been associated with a very poor outcome. Mortality of up to 90% in children failing to achieve enteral autonomy has been reported. Advances in understanding of intestinal adaptation and management of IF has resulted in improved outcomes in North America and Europe, and mortality rates as low as 5 to 10% have been reported. Management in multidisciplinary intestinal rehabilitation programmes (IRPs) has, in particular, resulted in improved survival. Research on survival and achievement of enteral autonomy has only recently been published from a South African (SA) centre and has not yet been investigated in our institution.

The aim of the present study was to describe the current management of short bowel syndrome (SBS) and the factors associated with survival and achievement of enteral autonomy in a South African context. To this end, a retrospective analysis of patients treated for IF at our institution was undertaken.

Factors associated with survival and achievement of enteral autonomy in patients treated for IF were the key elements of this investigation. Previous literature has focused on the role of aetiology, bowel length, presence of an ileo-caelal valve (ICV) and occurrence of IF-associated liver disease (IFALD) as possible risk factors for failure to achieve enteral autonomy and/or non-survival. In the present study, in addition to these factors, catheter-associated bloodstream infections (CABSI), loss of venous access, and surgical complications were examined as possible risk factors.

By identifying risk factors the author hoped to identify key areas that require improvement to improve survival of patients with IF.

Methods

Study design, sample selection and data extraction

Patients included in the study were below 1 year of age at diagnosis, treated at Inkosi Albert Luthuli Central Hospital (IALCH), Durban, SA, and met the case definition of IF: lack of sufficient intestinal function to maintain protein energy, fluid, electrolyte, and micronutrient balance requiring parenteral nutrition for at least 2 months. The cause of IF was determined to be SBS if patients met the following definition: IF resulting from surgical resection; congenital defect; or disease-associated loss of bowel length >50% of that expected for gestational age. Patients were included in the study if they died before 2 months but otherwise met the definition of SBS.

A retrospective electronic chart review of patients who fulfilled the inclusion criteria between November 2015 and February 2023 was undertaken.

Data were extracted from the electronic records by the author and included the following parameters: aetiology; length of stay; anatomical description; management details; complications; and outcomes. Anatomical description included: bowel length; segment of bowel loss; presence of ileo-caecal valve; and colon remaining. Management details included: enteral feeding choice; central venous access used; medical therapies; and bowel-lengthening procedures. Complications recorded were: early re-operation, surgical complications, CABSI, other central venous catheter complications, loss of venous access and occurrence of IFALD. Early re-operation was defined as repeat abdominal surgery within 28 days of the index operation. Finally, outcomes were recorded, i.e. enteral autonomy and survival. Patients with incomplete records were excluded from the analyses. Data were
inserted into an Excel spreadsheet (Microsoft Corp., USA) for analysis.

**Definition of terms**
The percentage of remaining small bowel length was based on normal reference values.\(^{1,12}\) Bowel length was measured at the time of initial surgery using a sterile measuring tape along the anti-mesenteric border.

CABSI was diagnosed if an acute septic episode was associated with the same positive microbiological culture from central venous catheter (CVC) and peripheral venous blood culture or through exclusion of other sources of sepsis in patients with indwelling CVCs. IFALD was defined as a sustained increase in total serum bilirubin >35 µmol/L, with exclusion of an alternative cause of liver disease. Enteral autonomy was defined as sustained growth and adequate fluid balance without parenteral support.

**Data analysis**
Data were analysed from two perspectives. Firstly, to describe the study population in terms of aetiology, length of stay, anatomical factors, management details, complications and outcomes. Secondly, to analyse possible risk factors for failure to achieve enteral autonomy and non-survival. The following risk factors were analysed: occurrence of a surgical complication(s); bowel length; ICV loss; CABSI; occurrence of IFALD; and loss of all venous access. Risk factors were quantified with odds ratios and a p-value <0.05 was considered statistically significant. Stata software (version 17; StataCorp., USA) was used for statistical analysis.

**Ethics**
Ethics approval was obtained from the Biomedical Research Ethics Committee (BREC) at the University of KwaZulu-Natal (UKZN) (ref. no. BREC/00004900/2022).

**Results**

**Patient characteristics**
Twenty-two patients were managed with IF during the study period (Table 1). In all patients, the cause of IF was SBS. All except 1 patient were in the neonatal age group and all patients were under 1 year of age. Diagnoses included the following: type 4 jejunal atresia with 3b component (n=5; 23%); type 3b jejunal atresia (n=5; 23%); type 4 jejunal atresia without 3b component (n=3; 14%); and malrotation with volvulus (n=3; 14%). One patient each had necrotising enterocolitis (NEC), gastrochisis with atresia, intussusception, type 1 jejunal atresia with volvulus, type 3a jejunal atresia and volvulus around an ileostomy (Fig. 1).

**Anatomical description of patient profile**
The median (IQR) bowel length was 36.5 (25 - 41) cm, with a minimum of 8 cm and a maximum of 60 cm. Bowel length distribution is shown in Table 2. Bowel segment loss was jejunum in 2 (9%), ileum in 3 (14%) and jejunum and ileum in 17 (77%) at index surgery. The ICV was present in 17 (77%) patients and absent in 5 (23%). A total of 19 patients (86%) had colon remaining, 1 patient had no colon remaining (5%) and 2 had partial colon remaining (9%).

**Patient management**
All patients received parenteral nutrition (PN) with early initiation of enteral feeds where feasible. Exclusive breastmilk was used in 50% (n=11) of cases and was the preferred feed, 18% (n=4) received polymeric formula, 9% (n=2) received elemental formula and 23% (n=5) patients were never fed. PN was given to all patients via a peripherally inserted central catheter (PICC) or CVC. The majority of catheters were non-tunneled (86%, n=19). Children treated required a median (IQR) of 3 (2 - 4) catheters (minimum 1, maximum 24). Intestinal-lengthening surgery was performed in 7 patients (32%), all were serial transverse enteroplasty (STEP) procedures. Three STEP procedures were done primarily at the time of initial laparotomy; in all cases the diagnosis was jejunal atresia with dilated proximal bowel. Medical adjuncts were used in 20 patients (91%). Two patients (9%) received a proton-pump inhibitor (PPI), loperamide and growth hormone (somatotropin), while another 2 patients received a PPI and loperamide, 16 patients (73%) received a PPI only and 2 patients received no medical adjuncts.

**Complications**
Life-threatening CABSI occurred in 73% of patients (n=16) and loss of central access occurred in 18% (n=4). Further CVC-related complications included threatened limb in 1 patient and an infected thrombus in 1 patient.

Fifteen patients (68%) developed IFALD. No patients progressed to end-stage liver disease; to minimise this complication, enteral feeds were initiated early where feasible. Patients who developed IFALD were managed by advancement of enteral feeds if possible and cyclical administration of PN. During the study period, the majority of patients received only 100% soybean-based PN (Intralipid; Fresenius Kabi, Germany). Soybean, medium-chain triglyceride, olive oil and fish oil-based PN (SMOFlipid; Fresenius Kabi, Germany) was only available towards the end of the study. SMOFlipid was only received by 1 patient – he did not develop IFALD.

Surgical complications occurred in 10 patients (46%) and 11 patients (50%) required early re-operation. One patient required early re-operation but did not have a surgical complication – the patient required an ileostomy after initial damage-control surgery for NEC (Fig. 2).

The median (IQR) length of stay was 57 (35 - 114) days. Enteral autonomy was achieved in 10 patients (45%). Enteral autonomy resulted in survival in all but one patient who died following surgery for adhesive bowel obstruction. Thus survival was achieved in 9 patients (41%). The cause of death was overwhelming sepsis in 9 patients (69%), malnutrition in 1 patient – in whom PN was stopped following loss of all venous access – and 3 patients (23%) died following withdrawal of care owing to critical patient prognostic characteristics (8 cm remaining total bowel (n=1) and only remaining bowel <20 cm of type 3b bowel with inability

### Table 1. Patient characteristics (N=22)

<table>
<thead>
<tr>
<th>Variable</th>
<th>n (%)</th>
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<tbody>
<tr>
<td>Female sex</td>
<td>13 (59)</td>
</tr>
<tr>
<td>Neonatal presentation</td>
<td>21 (95)</td>
</tr>
<tr>
<td>Age at diagnosis (days), median (IQR)</td>
<td>14 (6 - 14)</td>
</tr>
<tr>
<td>Gestational age (weeks), median (IQR)</td>
<td>37 (34 - 38)</td>
</tr>
</tbody>
</table>

*IQR = interquartile range.

**Table 2. Bowel length and survival**

<table>
<thead>
<tr>
<th>Bowel length (cm)</th>
<th>Death, n (%)</th>
<th>Survival, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;20</td>
<td>2 (100)</td>
<td>0</td>
</tr>
<tr>
<td>20 - 39</td>
<td>8 (67)</td>
<td>4 (33)</td>
</tr>
<tr>
<td>≥40</td>
<td>3 (43)</td>
<td>4 (57)</td>
</tr>
</tbody>
</table>

*Unless otherwise specified.
to progress enteral feeds ($n=2$)). With the exclusion of palliated patients, survival improves to 47%.

**Risk factors for mortality**

Risk factors for mortality are shown in Table 3. The most significant risk factors were surgical complication(s) and bowel length.

Ten patients developed surgical complications and 9 of them died. Twelve patients had no surgical complications and 4 of them died. Thus, children with a surgical complication had an 18-fold higher risk of dying, which was statistically significant ($p<0.05$).

Bowel length <20 cm predicted death perfectly. Bowel length >20 cm was not a good predictor of death or survival.

**Discussion**

The purpose of the present study was to describe the current management of paediatric IF secondary to SBS and to provide a current estimate of survival in an SA context. Furthermore, the study aimed to identify risk factors for failing to achieve enteral autonomy and non-survival.

Type 3b jejunal atresia, also known as apple-peel atresia (APA),\(^{13}\) accounted for the largest proportion of patients in this study (45%, $n=10$). Five patients had 3b atresia alone and 5 patients had 3b atresia as part of a type 4 atresia. Type 3b jejunal atresia is a rare anomaly, the reported incidence being 1 in 50 000 live births.\(^{14}\) A high mortality has previously been reported for 3b atresias in a variety of clinical settings.\(^{15,16}\) Intestinal atresias typically account for the second or third largest group of patients with neonatal SBS behind NEC and gastroschisis in many series.\(^{2,5,17-20}\) The reason for the relatively few patients with NEC and gastroschisis in the present study perhaps reflects the high early mortality for children with any of these conditions in our setting.\(^{21-23}\)

The median bowel length of patients in this study was 36.5 cm. Based on a comprehensive evaluation of normal values by Struijs et al.,\(^{12}\) the value of 36.5 cm is 31% and 15% of the expected length for a 30-week preterm and term infant, respectively. This represents severe short bowel length and might explain the high mortality experienced in this study since in much of the literature median bowel lengths of greater than 60 cm have been reported.\(^{2,3,6,8,19,24}\) However, many other studies\(^{4,5,18,20,25}\) reported similar or shorter bowel lengths and experienced much lower mortality. Therefore, other factors need to be examined to account for the high mortality experienced in the present study.

The mortality rate in the present study was 59%, which was high compared with many other reviews,\(^{3-6,8,18,26-28}\) where mortality rates <20% have been reported. The consideration of patient characteristics has been alluded to. Patient management details should be considered too, particularly central venous catheter use. Patients required an average (range) of 4.3 (1 - 24) central lines. The complication rate of CVCs is well known to increase with multiple removals and re-insertions.\(^{29-32}\) Furthermore, while the reasons for catheter removal are difficult to identify on a retrospective review, the author’s experience reflects that catheter removal and re-insertions directly reflect two aspects of CVC care, i.e. (i) the high rate of inadvertent removal and (ii) the high rate of CABSIs. Both issues demand further attention.

Besides catheter-related complications, surgical complications were also frequently encountered ($n=10$; 46%). This study clearly demonstrates the relationship between surgical complications from the initial surgery and mortality in children with IF secondary to SBS. While the cause of...
surgical morbidity is likely multifactorial, it is crucial for surgeons to pay attention to meticulous technique and postoperative care needs to be equally thorough. IFALD was a management-related complication associated with increased mortality in the present study and although not statistically significant is likely to be clinically significant.

The management of intestinal rehabilitation is evidently an overwhelming task incorporating initial surgical care, CVC insertion and maintenance, medical care, nutritional rehabilitation and avoidance of all the complications detailed above. A multidisciplinary team is best suited to managing all these complex issues and initiation of intestinal rehabilitation programmes (IRPs) has been shown to have a significant positive impact on patient outcomes. Indeed, perhaps the lack of an IRP at IALCH was the most significant contributor to the poor outcomes of many patients in the present study.

**Study limitations**

Limitations of the present study include the small sample size and retrospective sampling technique. In particular, it is possible that some patients who met the definition of IF were missed because of rapid rehabilitation. Furthermore, some patients with particularly severe SBS, which was rapidly fatal, may have been missed at the beginning of the study period.

**Conclusions and recommendations**

Evidence from the present study has informed the following recommendations: (i) secure, long-term CVCs with low sepsis rates is crucial for survival; (ii) CVC-related complications are severe and the use of infection control measures, including ethanol locks, single-lumen catheters and antimicrobial-impregnated catheters should be considered; (iii) judicious use of tunneled catheters should be employed; (iv) adherence to insertion and maintenance bundles is crucial, as is the ongoing education on CVC care of nursing and medical staff. Surgical complications have particularly important consequences and surgery should be undertaken with great care by experienced members of the team with vigilant postoperative care. IFALD can be avoided and reversed with advancement of enteral autonomy and survival was achieved in many patients with bowel lengths that would have been considered incompatible with life in previous eras. It is likely that vigilant surgical and CVC care, avoidance of IFALD and initiation of IRPs may improve outcomes further.

**Declaration.** None.

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**Author contributions.** Sole author.

**Funding.** None.

**Conflicts of interest.** None.

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Table 3. Risk factors for death

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>OR</th>
<th>p-value</th>
<th>95% CI</th>
</tr>
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<tbody>
<tr>
<td>Surgical complication</td>
<td>18</td>
<td>0.02</td>
<td>0.01 - 0.61</td>
</tr>
<tr>
<td>Bowel length &lt;20 cm*</td>
<td>3.56</td>
<td>0.3</td>
<td>0.33 - 38.78</td>
</tr>
<tr>
<td>Absent ICV</td>
<td>6.87</td>
<td>0.06</td>
<td>0.93 - 50.78</td>
</tr>
<tr>
<td>IFALD</td>
<td>2.2</td>
<td>0.42</td>
<td>0.32 - 14.97</td>
</tr>
<tr>
<td>Loss of all IV access</td>
<td>2.4</td>
<td>0.48</td>
<td>0.2 - 27.72</td>
</tr>
</tbody>
</table>

OR = odds ratio; CI = confidence interval; ICV = ileocecal valve; IFALD = intestinal failure-associated; CABSI = catheter-associated bloodstream infection(s); IV = intravenous.

*Bowel length <20 cm predicts death perfectly.


