South African Journal of Clinical Nutrition 2023; 36(2):76–83
https://doi.org/10.1080/16070658.2022.2114404

SAJCN
ISSN 1607-0658 EISSN 2221-1268

ARTICLE

Nutritional interventions and outcomes of children with short bowel syndrome in a tertiary hospital setting in South Africa

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Objectives: To describe the impact of nutritional interventions on the outcomes of children with short bowel syndrome (SBS).
Design: This was a retrospective descriptive observational review where data were obtained from the patient's medical records.
Subjects and setting: Children with SBS between the ages of 0 and 24 months who obtained this diagnosis between January 2005 and December 2015 at a tertiary paediatric hospital in Cape Town were investigated.
Results: There were 46 patients (62% male, 38% female) included in the study. The median duration of parenteral nutrition (PN) support was one month (0.6, 2.2 months), after which 83% of patients were weaned from PN. Enteral nutrition was commenced in 96% of patients, with the majority ($n = 36$; 82%) starting on day six (±6; range 1–29 days) postoperatively and 80% of patients attaining full feeds at median 1.2 months (0.2, 36 months). Patients displayed a mean weight and length gain of 15 g/day (±4; range 19–92 g) and 2 cm/month (±1.4; range 0.25–4 cm) respectively. The main complications were PN-associated cholestasis ($n = 17$), fat malabsorption ($n = 13$) and vitamin D deficiency/insufficiency ($n = 5$).
Conclusion: This study showed that early initiation of PN support was attained, and that most patients were able to achieve enteral autonomy.

Keywords: short bowel syndrome, paediatrics, nutritional outcomes

Introduction

Short bowel syndrome (SBS) is defined in the literature as a malabsorption disorder that occurs because of extensive resection of the small bowel.[1] The diagnosis of SBS is routinely made when more than 70 cm (approximately 23%) of small bowel length is lost and/or the patient remains dependent on parenteral nutrition support for more than six weeks. Small bowel length has been reported to be approximately 150–300 cm in term neonates.[2] Although emphasis is placed on the length of bowel it is important to note that SBS is the loss of small bowel function rather than a small bowel anatomical disorder.[2,3] Other factors that are relevant to the development of SBS include the functionality of the residual bowel, the underlying diagnosis, the type of bowel segments that remain, the presence of the ileo-caecal valve (ICV), presence and location of the stoma and the functionality of the remaining colon. All these aspects will influence bowel adaptation, which further impacts on the functionality of the gastrointestinal system and feeding options.[4,5]

Patients with SBS primarily present with substrate malabsorption as well as electrolyte imbalances, bacterial overgrowth, micronutrient deficiencies and bile malabsorption.[6] This is a direct consequence of the reduction of the absorptive and digestive surface, which results in a reduced availability of digestive enzymes and transport proteins.[7]

SBS management is aimed at promoting adaptation of the remaining short bowel, villous hyperplasia via enteral feeding, promoting appropriate growth and development, optimising the absorptive surface of the bowel with non-transplant surgical techniques, maintaining fluid and electrolyte balance, and promoting good quality of life.[8,9] The literature further highlights the following factors as predisposing to successful adaptation: (1) younger patient age; (2) longer residual bowel length; (3) intact ileo-caecal valve; (4) absence of gastrointestinal mucosal inflammation; (5) absence of cholestasis and (6) normal gastrointestinal motility.[10] It is also anticipated that adaptation will successfully take place in those patients with > 30 cm (10%) residual bowel with an intact ICV and > 70 cm (23%) residual bowel in the absence of an ICV.[11,12]

It is essential for parenteral nutrition (PN) to be started early post small bowel resection as it is the primary means to meet the patient's dietary requirements until bowel adaptation allows for transition to full enteral feeds. The duration of PN support will depend on the measure of intestinal adaptation that has been achieved. The length of time a patient remains dependent on PN will, however, increase the cumulative risk of long-term complication such as sepsis, cholestasis and death. The literature reports that liver function tests may start to increase within 1–4 weeks of starting TPN.[7,13,14]

Enteral feeding, either orally or via nasogastric tube, should be initiated as soon as possible after bowel resection, once the postoperative ileus has resolved, as it is seen to be the primary treatment of SBS. The type and timing of feed will be influenced by the residual bowel length and function, type of stoma, presence of ICV and colon. The presence of nutrients in the intestine is essential in promoting gut adaptation as adaptation is driven by the increased load of fatty acids, carbohydrates and proteins on the enteroglucagon-producing cells found in the ileum.[9] The goal of feeding is to reduce the incidence of parenteral nutrition associated cholestasis (PNAC), decreasing diarrhoea and steatorrhea and dehydration, improving intestinal adaptation and reducing sepsis episodes.[3,7,15]

Infants with SBS may have challenges in maintaining appropriate growth due to malabsorption, bearing in mind the increased dietary requirements for growth and brain development during the first two years of life.[16] Close monitoring of all anthropometric parameters should continue after enteral autonomy has been reached as patients who show appropriate weight and length gains reflect adequate intestinal adaptation and absorption. Failure to thrive may be seen as an indicator to restart specialised nutritional support such as enteral supplementation and/or partial PN if required.[8] Better growth trends are observed during the period of full PN support as the effects of malabsorption are minimised. Poorer growth trends have been reported and are generally expected when enteral nutrition is introduced and the dependence on PN support is decreased, especially when PN support is aggressively weaned. It is thus essential that any decision to wean from PN should take into consideration the adequacy of a patient's growth and the severity of any PN-related complications that may be present[17,18]

The aim of this study was to highlight the nutritional management strategies and assess the nutritional outcomes of patients with short bowel syndrome within a tertiary care setting.

Methods

Study population and sampling

This was a retrospective descriptive observational study, which was conducted at Red Cross War Memorial Children's Hospital. The study population consisted of inpatients (aged 0–24 months) who underwent extensive small bowel resection and were subsequently classified as having short bowel syndrome between January 2010 and December 2015. The same patients were evaluated for one month post-discharge at the outpatient clinic. Short bowel syndrome was defined as the loss of > 70% of their small bowel length or ≤ 100 cm of small bowel length remaining.

Data collection

Demographic information

Data were extracted from the databases of the Department of Paediatric Surgery, the Department of Dietetics and the medical records of the identified patients. The medical records were utilised to obtain information on the patient's age, gestational age and primary diagnoses that resulted in short bowel syndrome. Patient medical records were further reviewed to obtain all surgical operation notes that contained detailed information confirming the remaining bowel length, presence of an ileo-caecal valve, presence and type of stoma, continuity of bowel and presence of colon.

Nutrition-related complications

Liver function, vitamin D levels and faecal histology were recorded to identify possible complications related to nutritional intervention. Liver function tests were evaluated to identify the development of parenteral nutrition associated cholestasis. Parenteral nutrition associated cholestasis was defined as a total bilirubin level > 34 µg/l. Severe parenteral nutrition associated cholestasis was defined as total bilirubin > 68 µg/l, AST > 60 IU/l and ALT > 35 IU/l.[19,20]

The diagnosis of fat malabsorption was based on examination of faecal histology results. Moderate to abundant fat content or a raised stool steatocrit value (> 25%) was used as a positive diagnosis of fat malabsorption.[21]

Where vitamin D levels were available these was classified according to the following criteria: severe deficiency < 5 ng/ml 25(0H) D, mild to moderate deficiency 5–15 ng/ml 25 (OH)D, insufficiency 16–20 ng/ml 25(OH)D, sufficiency 21–100 ng/ml 25(OH)D, excess 101–149 ng/ml 25(OH)D and toxicity > 150 ng/ml 25(OH)D.[22]

Anthropometry

Weights were recorded daily, and length and head circumference data were collected as available during inpatient stay. On discharge, weights, length and head circumference were recorded, as available, for every follow up visit, for a one-month period. All weight measurements, length and head circumference were plotted against the WHO growth charts for age to compare patients against their relevant Z scores. Anthro (https://who-anthro.software.informer.com/3.2/) and INTERGROWTH-21st software (http://intergrowth21.ndog.ox.ac.uk/) was used to assist with these calculations.[23,24]

Ethics approval

The study was submitted for approval to the Health Research Ethics Committee of Stellenbosch University (S16/03/053), the University of Cape Town (744/2016) and the management of Red Cross War Memorial Children's Hospital. A waiver of informed consent was granted as this was a retrospective study, and all data were obtained from patient records only.

Data analysis

MS Excel 2016 (Microsoft Corp, Redmond, WA, USA) was used to capture the data and STATISTICA 13 (TIBCO Software Inc. (2017). Statistica (data analysis software system), version 13. http://statistica.io.), was used to analyse the data.

Summary statistics were used to describe the variables. Descriptive statistics (means, medians and standard deviations, minimum and maximum for continuous variables) were used to describe patient demographics, bowel length characteristics, PN intake amongst age groups, total energy and protein intake, and demographics around complications (PNAC, Vitamin D deficiency and fat malabsorption).

Spearman correlations were done to assess the relationship between short bowel length and total days PN, short bowel length and length of hospital stay, short bowel length and days to full feeds. Non-parametric kernel regression was done to model the relationship between length of stay and short bowel length. Kruskal–Wallis analysis was used to assess the relationship between bowel continuity (presence of jejunostomy or ileostomy or primary anastomosis) and total days PN. One-way ANOVA analysis was performed to assess a comparison of intake between the residual small bowel lengths in terms of days to full feeds and total days PN.

Results

Patient demographics

Between January 2010 and December 2015, 62 patients met the inclusion criteria and 46 (73%) complete medical records were available for evaluation. Of these $n = 46$, $n = 38$ (83%) patients were alive.

The study population of 46 individuals had a male predominance (62%) and the mean gestational age of patients was 34 weeks (±4.4; range 27–40 weeks). The median age on admission

to hospital was 14 days (4, 40 days) and median length of stay was 50 days (27, 107 days) (Table 1).

The main diagnosis resulting in small bowel resections and consequently short bowel syndrome was necrotising enterocolitis (NEC) (48%). After bowel resections the median residual short bowel length was 64 cm (47, 80 cm) with a range of 11–100 cm. In the total patient summary 78% of patients had an intact ileo-caecal valve, 91% had an intact colon, 44% had undergone a primary anastomosis and 59% had a stoma (78% ileostomy, 19% jejunostomy, 4% colostomy).

When comparing the length of stay and residual small bowel length, an inverse correlation was found between length of stay (in days) and the remaining bowel length ($r = -0.29$, $p = 0.07$) (see Figure 1). Further non-parametric regression analysis found that the average effect of residual small bowel length on hospital stay was that an increase in bowel length led to a 2.5-day shorter hospital stay ($p = 0.002$). For every 10 cm shorter than 60 cm residual SBL, length of hospital stay increased by 50 days.

Dietary intervention

Parenteral nutrition support

The majority of patients (46%) started early PN support on the day of resection or one day after resection (39%). The longest duration before starting PN was on day three after resection (7%). The median duration of total PN support was one month (0.6, 2.2 months) with a range of 0.2–49 months. All patients were started on an all-in-one single lipid emulsion (soya bean oil based) complete PN solution.

Thirteen (28%) patients required a second course of PN support for an average of 2.1 weeks (±0.7; range 1–2.8 weeks) after their first course of PN was completed. Of these, $n = 5$ (39%) patients were restarted on PN during their initial admission, $n = 3$ (23%) patients were readmitted from home and $n = 5$ (38%) patients were restarted after being transferred from another hospital. Results therefore show that these patients (28%) did not achieve enteral autonomy after their first course of PN.

The majority (83%) of patients were weaned off PN support and $n = 8$ (17%) patients remained PN dependent with $n = 7$ (88%) of the PN-dependent patients dying during their hospitalisation. Patients who were weaned off PN received a mean duration of PN of 2.8 months (±6; range 0.2–36 months) whereas patients who were not weaned received a mean duration of 8 months (±15; range 0.2–49 months) of PN support. Three (30%) patients required home-based parenteral nutrition with $n = 2$ (67%) of these patients weaned off PN after bowel lengthening surgery.

Factors affecting PN weaning

Spearman's correlation was done to assess the relationship between short bowel length and duration of PN. An inverse correlation was found between short bowel length and total number of days on which patients received PN ($r = -0.32$; $p = 0.03$) (Figure 2).

A residual short bowel length of 60 cm was identified as the clear threshold for duration of PN administration. Patients ($n = 20$; 43%) with a < 60 cm residual short bowel length received PN for a mean duration of 7 months (±12; range 0.2–49 months). Patients with ≥ 60 cm residual short bowel length ($n = 26$; 57%) had a mean duration of PN of 1.1 months (±0.7; range 0.3–3.1 months).

With further comparison of bowel characteristics, a non-significant difference was found for PN days in patients who had undergone a primary anastomosis versus patients with a stoma (mean days 109 vs. 120; $p = 0.92$).

Table 1: Patient demographic information

Clinical characteristic	***n* = 46**	
Gender, *n* (%):		
Male	29 (62)	
Female	18 (38)	
	Mean (min–max)	Median (25th percentile, 75th percentile)
Gestational age, weeks	34 (27–40)	35 (30, 38)
Birthweight, grams	1867 (830–3300)	1780 (1280, 2608)
Age on admission, days	40 (1–420)	14 (4, 40)
Length of hospital stay, days	83 (8–357)	50 (27, 107)
Residual small bowel length, cm	62 (11–100)	64 (47, 80)
Diagnosis, n (%):		
Necrotising enterocolitis	22 (48%)	
Jejunal atresia	11 (24%)	
Mid-gut volvulus	8 (17%)	
Intestinal mid-gut malrotation	3 (6%)	
Shocked gastroenteritis	3 (6%)	
Small bowel obstruction	2 (4%)	
Hirschsprung's disease	1 (2%)	
Gastroschisis	1 (2%)	
Bowel perforation	1 (2%)	
Bowel necrosis	1 (2%)	

Enteral nutrition support

In 44 patients (96%) enteral feeds were initiated, with the majority ($n = 36$; 82%) starting on day six (±6; range 1–29 days) postoperatively. Two patients died (4%) before feeds could be initiated.

The majority of patients ($n = 35$; 80%) achieved full feeds at median 1.2 months (0.5–2.6 months). This includes the $n = 2$ (4%) patients who were started on home-based PN, while awaiting bowel lengthening surgery. They were able to attain full feeds only after 36 and 17 months respectively. Three patients were transferred to neonatal units where full feeds were achieved.

The types of feeds that were initiated are as follows: expressed breast milk (EBM) in 48% of patients, extensively hydrolysed infant formula (EHF) in 32% of patients, polymeric infant formulas in 14% of patients and amino acid-based formula in 6% of patients. Only 17% maintained exclusive breastfeeding on attainment of full feeds. Approximately half of the patients (49%) reached full feeds on amino acid (AA) based infant formula.

Similar to previous results, an inverse correlation was identified between residual small bowel length and days to full feeds ($r = -0.40$, $p = 0.02$) (Figure 3). Patients with 40–100 cm residual SBL attained full feeds earlier at 45 days (±43; range 7–206 days), $p = 0.07$ when compared with the other SBL classifications.

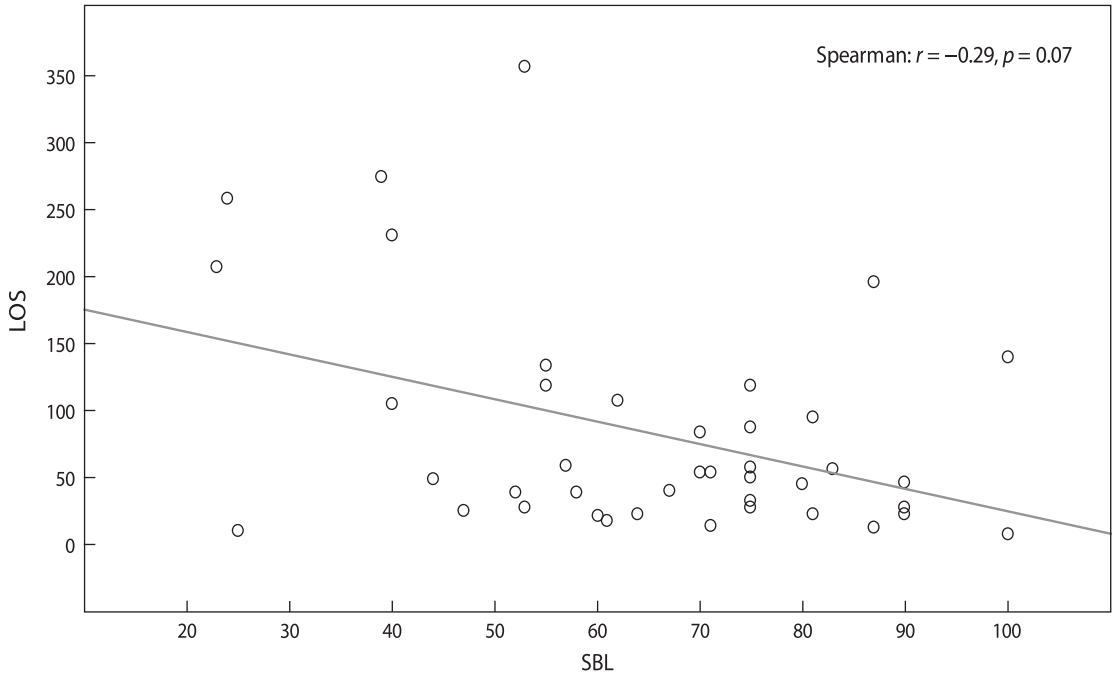

Figure 1: Correlation between SBL and LOS. SBL: short bowel length; LOS: length of stay.

Energy and protein intake

The total energy and protein intakes (PN &EN) were evaluated per age group; premature (<37-week gestational age), term infants (>37-week gestational age) and older children (>1 year) (Table 2).

Premature infants received a mean daily energy intake of 89 kcal/kg/day (±17; range 48–111 kcal/kg/day) which was less than the ESPGHAN recommendations of 110–130 kcal/kg/day. They met only 67% of their protein intake requirements at a mean intake of 2.5 g/kg/day (±0.5; range 1.3–3 g/kg/day).

Term infants received 95% of their recommended intake with a mean total energy intake of 91 kcal/kg/day (±22; range 30–122 kcal/kg/day). Protein intake met the recommended intake with a mean intake of 2.5 g/kg/day (±0.7; range 0.9–3.6 g/kg/day). There was only one patient > 1 year and this patient met 98% of the recommended energy goals at 78 kcal/kg/day and 100% of the protein requirement at 2.1 g protein/kg/day.

Parenteral nutrition associated cholestasis (PNAC)

Liver function tests were evaluated only in those patients who were receiving PN for > 14 days as PNAC is more prevalent in

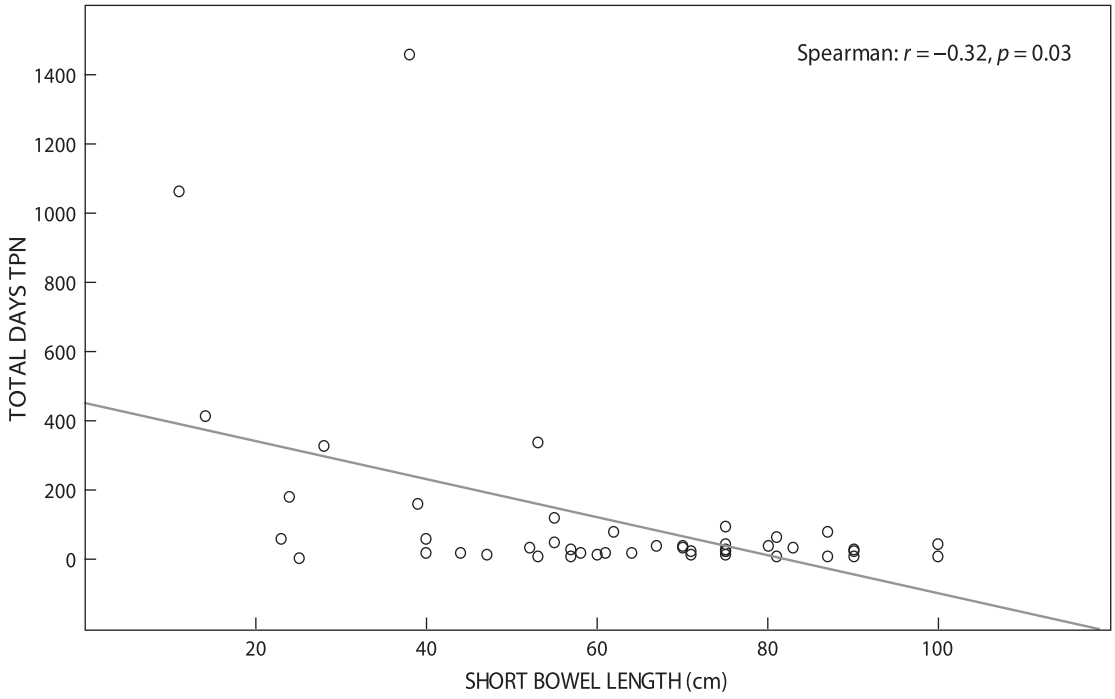

Figure 2: Correlation between short bowel length and total days PN. PN: parenteral nutrition; cm: centimetres.

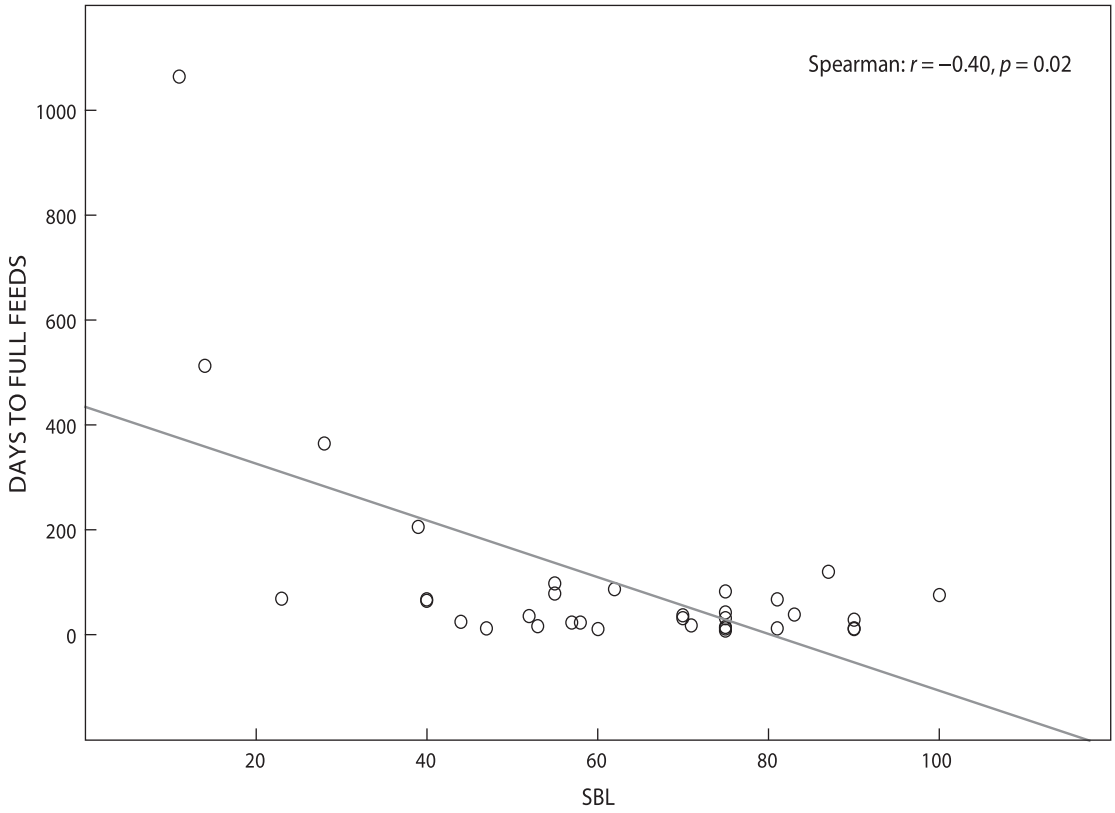

Figure 3: Correlation between short bowel length and days to full feeds.

patients who require long-term PN support.[2] Of the 37 (80%) patients who received PN for > 14 days, $n = 17$ (46%) patients developed PNAC based on raised total bilirubin, conjugated bilirubin, AST and ALT. These patients developed cholestasis 19 days (±23; range 10–58 days) after initiating PN. The mean duration of PN support in cholestatic patients was 7.6 months (±13; range 0.5–49 months).

Patients with NEC ($n = 9$; 53%) and jejunal atresia ($n = 6$; 35%) were more prone to develop cholestasis. Most of these patients had an intact colon ($n = 12$; 71%), an intact ICV ($n = 10$; 59%) and a stoma present ($n = 7$; 41%).

Management strategies aimed at treating and preventing cholestasis included cyclical PN ($n = 7$; 41%) and the use of a multi-lipid PN containing soya bean oil, medium chain triglycerides, olive oil and fish oil ($n = 6$; 35%) (Table 3).

Vitamin D deficiency and insufficiency

Vitamin D [25(OH)D] levels were available in only $n = 5$ (11%) patients. Of these five patients, $n = 3$ (60%) presented with vitamin D deficiency (≤ 15 ng/ml) and $n = 2$ (40%) patients presented with vitamin D insufficiency (16–20 ng/ml).

The mean gestational age of the patients in the vitamin D deficient group was 34 weeks (±4; range 30–38 weeks) and 32.5 weeks (±4.5; range 28–37 weeks) in the insufficient group, respectively. The patients who developed vitamin D deficiency spent an average of 6.3 months (±3.9; range 0.8–12 months) hospitalised. Two (40%) patients had fat malabsorption and $n = 4$ (80%) had cholestasis.

The lack in availability of vitamin D level results could be attributable to these not being regularly available before 2010 at the institution where the study was undertaken.

Table 2: Total energy and protein intake

		Actual intake			
Recommended intake[25,26]		**Mean % of requirements met**	**Mean daily intake ± SD**	**Minimum intake**	**Maximum intake**
Premature infants (< 37-week gestational age), $n = 28$:					
Energy (kcal/kg/day)	110–130	81	89; ±17	48	111
Protein (g/kg/day)	3.5–4	67	2.5; ±0.5	1.3	3
Term infants, $n = 17$:					
Energy (kcal/kg/day)	96–120	95	91; ±22	30	122
Protein (g/kg/day)	2–4	83	2.5; ±0.7	0.9	3.6
Older child ≥ 1 year, $n = 1$:					
Energy (kcal/kg/day)	80	98	78; ±17	60	104
Protein (g/kg/day)	1–3	100	2.1; ±0.5	1.5	2.8

SD: standard deviation.

Table 3: Demographics of patients developing cholestasis

Clinical characteristic	*n* = 17
Mean PN duration (months), mean (min–max)	7.6 (0.5–49)
Mean days to development of cholestasis after PN support started, mean (min–max)	19 (10–58)
Mean small bowel length (cm), mean (min–max)	60 (11–90)
Residual small bowel length characteristics, *n* (%)	
ICV present	10 (59%)
Intact colon	12 (71%)
Primary anastomosis	6 (35%)
Stoma present	7 (41%)
Mean gestational age (weeks), mean (min–max)	33 (28–40)
Diagnosis, n (%):	
NEC	9 (53%)
Jejunal atresia	6 (35%)
Midgut volvulus	1 (6%)
Malrotation	1 (6%)
Patients started on cyclical PN, n (%)	7 (41%)

PN: parenteral nutrition; ICV: ileo-caecal valve; NEC: necrotising enterocolitis.

Fat malabsorption

Thirteen (30%) patients were diagnosed with fat malabsorption. These patients presented predominantly with NEC ($n = 7$; 54%) followed by jejunal atresia ($n = 4$; 31%) and most patients had a residual small bowel length < 50 cm ($n = 8$; 62%). Ten (77%) patients also presented with cholestasis.

On average, fat malabsorption was only diagnosed 74 days (±42: range 30–150 days) after feeds were started. At the time that fat malabsorption was diagnosed, $n = 9$ (69%) patients were receiving an amino acid-based feed (containing 8% medium chain triglycerides), $n = 3$ (23%) patients were receiving an extensively hydrolysed feed (containing 33% medium chain triglycerides) and $n = 1$ (8%) patient were on expressed breast milk. Pancreatic enzymes (*Creon*) were started in $n = 9$ (69%) patients as treatment option for fat malabsorption.

Anthropometry

Anthropometry during hospital stay

Weight data were available for all patients, with length measurements available in $n = 23$ (50%) of these patients. No head circumference data were available to evaluate. On admission $n = 23$ (50%) patients were classified as normal weight for age (−1 to 0SD), $n = 7$ (15%) as severely underweight for age (≤−3SD), $n = 8$ (17%) as moderately underweight for age (−2 to −3 SD) and $n = 8$ (17%) as mildly underweight for age (−1 to −2 SD). We chose to classify weight data only as there were only $n = 5$ (11%) patient's length data available on admission.

Furthermore, the anthropometry was classified of those patients who were weaned off PN ($n = 38$; 83%). Their weight data were classified at attainment of full feeds, on discharge and one-month post-discharge. On attainment of full feeds, the majority of patients, $n = 19$ (51%), were severely underweight and $n = 8$ (22%) had a normal weight for age. On discharge the majority of patients [$n = 22$ (59%)] were severely underweight (Table 4). Due to the lack of length data, weights only were classified at these selected end points.

The overall weight and length gain trajectory fell within recommendations of 15 g/day (±4; range 19–92 g) and 2 cm/month (±1.4; range 0.25–4 cm) respectively.[26] However, a slower trend was seen in those patients with a short bowel length of <60 cm, without a colon present and in patients with a diagnosis of NEC. This group showed an average weight gain of 38 g/week or 5 g/day ($p = 0.012$).

Anthropometry one month after discharge

Post-discharge weight data were available for $n = 25$ (54%) patients, with height data available for $n = 9$ (20%) patients. Sixteen (64%) patients were classified as severely underweight for age, $n = 2$ (8%) as moderately underweight for age and $n = 7$ (28%) patients as normal weight for age.

Where length was available, $n = 7$(78%) were severely stunted (< −3SD) and $n = 7$(78%) had normal weight for height. Patients showed an average weight gain of 23 g/day (±13; range −6.25–44 g/day) which is more than the mean weight gain velocity that was displayed during their stay in hospital (15 g/day ±4; range 19–92 g).

Discussion

This study was undertaken as there has been an increase in patients being diagnosed with short bowel syndrome and patients surviving long term. The management of these patients is known to be resource intensive as it requires long-term hospital stay, multiple surgical procedures and specialised nutritional intervention, which all predominantly occur within the first year of life.[27] The objective of this study was to describe the impact of nutritional interventions on the outcomes of children with short bowel syndrome. We further sought to identify the factors that influence nutritional intervention and to highlight selected dietary-associated complications.

When evaluating nutrition interventions, we were able to display early initiation of both PN and EN support. A high weaning rate of PN was achieved in 83% of our patients, of whom 80% of those were able to achieve full feeds. Only one patient remained PN dependent, requiring home PN support.

Parenteral nutrition is essential in the management of SBS, but unfortunately long-term PN dependence is one of the main contributors to complications and mortality in this patient group. It was therefore important to identify possible predicting factors that could contribute to earlier PN weaning, which would be beneficial in guiding long-term management. Residual bowel length was identified as the main factor that influences nutritional interventions as it inversely affected duration of both EN and PN support. Short bowel length of ≥ 60 cm was identified as an indicator of shorter PN dependence. Patients who had undergone a primary anastomosis were also shown to have fewer mean days' PN support (109 days) compared with stoma patients (120 days), although this was not found to be statistically significant ($p = 0.92$). These data

Table 4: Comparison of weight for age at full feeds and discharge

Weight for age, *n* = 38	Full feeds	Discharge
Severe UWFA, n (%)	19 (51%)	22 (59%)
Moderate UWFA, n (%)	7 (19%)	6 (16%)
Mild UWFA, n (%)	3 (8%)	1 (3%)
Normal, n (%)	8 (22%)	8 (22%)

UWFA: underweight for age.

correlate with the literature, which has identified longer short bowel length (> 10% of predicted small bowel length) and establishment of intestinal continuity to be independent predictors for reaching enteral autonomy.[5,7] Fallon *et al*. also showed that patients with at least 50 cm of short bowel had a 88% probability of being weaned at 12 months of PN in comparison with those patients with < 50 cm of short bowel who had only a 23% probability of being weaned at 12 months of PN support.[26] The beneficial role of intestinal continuity via a primary anastomosis may be attributed to the improvement in the absorption surface area, improvement in colonic adaptation, improved electrolyte and water absorption and an increase in the absorption of short-chain fatty acids.[1]

A low incidence of complications was seen, with 46% of the study population developing PNAC, 30% developing fat malabsorption and 11% patients presenting with vitamin D deficiency and insufficiency. This low incidence of complications is welcome, as the literature reports most of the costs attributed to the care of patients with SBS to be due to the management of resulting complications.[28] The patients who were more prone to develop complications were premature infants, patients with a prolonged hospital stay, diagnosis of NEC, jejunal atresia and SBL $\leq$ 60 cm.

Fat malabsorption may develop as a result of inactivation of pancreatic enzymes due to gastric acid hypersecretion.[26,29] In this study population, 13 patients (30%) presented with fat malabsorption and, of these, 10 (77%) patients also presented with cholestasis. Bile constituents are essential for the efficient intestinal absorption of fat. As cholestasis is characterised by disturbances in bile formation and secretion, it will have a significant impact on lipid metabolism. This may particularly be due to the absence of bile salts in the proximal intestine, which is essential for micelle formation.[26]

The most important causative factors for vitamin D deficiency in malabsorption are inadequate intake of vitamin D, inadequate sunlight exposure for UVB-dependent synthesis of vitamin D and impaired intestinal absorption of vitamin D.[29,30] Vitamin D absorption takes place mainly in the jejunum and terminal ileum. In our vitamin D deficient patients, only one patient had no terminal ileum and three patients had decreased jejunum length of 11 cm, 20 cm and 20 cm respectively. As the majority of both the vitamin D deficient and insufficient group were premature it is of value to highlight prematurity as a potential contributing factor to low vitamin D levels. The premature infant is unable to endogenously produce 25-OHD and is entirely dependent on mother-to-child transfer of vitamin D. The trans-placental transfer of vitamin D occurs mainly in the last trimester of pregnancy, therefore premature infants are inherently born with a vitamin D deficit.[31]

Despite these favourable nutrition interventions our patients had a poor nutritional status during hospital stay. On attainment of full feeds, most of our study population were classified as severely underweight for age when compared with their age peers. We were unable to report accurately on the level of stunting in our group due to the lack of regular length measurements during hospital stay.

These results are in keeping with other studies investigating the long-term outcomes of patients with SBS, who have also demonstrated lower weight-for-age scores and poor linear growth when compared with healthy controls, even when corrected for gestational age. The literature cites the following as possible contributory factors to poor growth in this population: (a) inadequate calorie and protein intake; (b) receiving adequate calories but poor utilisation due to increased gastrointestinal losses; (c) under-recognised micronutrient deficiency and (d) increased metabolic requirements caused by infections. It has also been demonstrated that length gain in particular in patients with SBS has proved to be one of the anthropometric parameters that is the most challenging to improve even when providing sufficient calories.[18,32]

When further analysed, a lower weight gain trend was seen in those patients with a short bowel length of < 60 cm, without a colon present and in patients with a diagnosis of NEC. This group showed an inappropriate mean weight gain of 38 g/week ($p = 0.012$). This is in keeping with data from several studies that report a higher incidence of poor growth in neonates with NEC when compared with controls. Hintz *et al*. report significant growth delay (in weight, length and head circumference) in infants that required surgical intervention for NEC, compared with medically managed NEC patients, as well as patients with a higher staging of NEC, when defined according to the Bell staging for NEC.[27] When Varma *et al*. evaluated the feeding practices of infants who had undergone gastrointestinal surgery they similarly found growth failure in infants with NEC and spontaneous intestinal perforation. They attributed these growth findings to (a) changes in metabolism that are commonly found during critical illness, (b) inadequate delivery of prescribed nutrition due to feeding intolerance, (c) loss of functional gastrointestinal mass post-surgery, which leads to malabsorption and (d) increased energy needs.[33] When analysed according to gestational age, premature infants in our study group received a mean total calorie intake of 89 kcal/kg/day, which met only 73% of their recommended daily energy requirements when evaluated against the ESPGHAN recommendation of 110–135 kcal/kg/day.[23] This low-calorie intake together with the predominance of fat malabsorption in this subgroup (30%) could be seen as one of the main contributors to poor growth in these patients. As this study did not report on the extent of intestinal losses, we are unable to provide this as a potential causative link for poor growth.

On discharge our patients were able to maintain a favourable weight gain trajectory at an average weight gain of 23 g/day (±13; range −6.25–44 g/day). It is important to evaluate post-discharge anthropometry, as maintaining appropriate growth after reaching enteral autonomy is seen as an important indicator of successful intestinal adaptation.[7]

Conclusion

Short bowel syndrome is a complex disorder of malabsorption requiring multidisciplinary input from paediatric surgeons, nurses and dietitians to name but a few. Multidisciplinary management is associated with improved outcomes and earlier attainment of enteral autonomy as it utilises standardised coordinated care through the use of evidence-based management protocols.[33] The dietitian is an essential part of this team, especially in the guidance of appropriate PN and EN interventions. The goal of nutritional management of these patients is to support optimal nutritional status, facilitate intestinal adaptation and limit morbidity and mortality (associated with long-term PN support) by promoting enteral autonomy.

This is the first study that describes nutrition intervention in this population group in the South African setting. Although the retrospective nature of the study placed limitations on the availability of medical records and limited our sample size, we were still able to display favourable results in this area that were comparable to international studies. We demonstrated that it is possible for most patients to be weaned from parenteral nutrition. We further identified residual bowel length, gestational age, diagnosis and length of hospital stay as the main factors influencing the outcome of nutritional interventions.

As medical management strategies changes with time a further prospective evaluation of this patient group is advised to provide more perspective on their outcomes.

Disclosure statement – No potential conflict of interest was reported by the authors.

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Received: 19-04-2022 Accepted: 10-08-2022