Journal of Pediatric Surgery 53 (2018) 1345–1350

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Outcomes in patients with short bowel syndrome after autologous intestinal reconstruction: Does etiology matter?☆

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ARTICLE INFO

Article history:
Received 15 January 2017
Received in revised form 26 February 2018
Accepted 1 March 2018

Key words:
Short bowel syndrome
AGIR
Bowel lengthening
Etiology

ABSTRACT

Background: Short bowel syndrome (SBS) is the most common cause of intestinal failure in children. Many factors have been investigated in an attempt to define which parameters influence most survival and ability to wean off parenteral nutrition (PN). The aim of this study was to investigate if aetiology of SBS affects the outcomes in paediatric patients treated with autologous gastrointestinal reconstructive surgery.
Methods: All children with SBS who underwent autologous gastrointestinal reconstructive surgery between 2002 and 2012 were retrospectively reviewed and outcome measures were recorded.
Results: Forty-three patients were divided into 4 groups according to aetiology (gastroschisis, volvulus, necrotizing enterocolitis (NEC), intestinal atresia). No significant differences were found among groups regarding survival and median age at surgery. The volvulus group had a lower pre-operative bowel length in comparison with gastroschisis and intestinal atresia and the lowest percentage of patients off PN (30%). Gastroschisis had the lowest rate of preserved ileocaecal valve (10%), while intestinal atresia had the highest (66%). For children who weaned off PN, intestinal atresia had also the longest time to achieve enteral autonomy (14.5 months), while NEC had the shortest (3.5 months), followed by gastroschisis (8.5 months). None of the patients needed transplant.
Conclusions: In our experience it does not appear that diagnosis is significantly related to outcome and this is consistent with the conclusions of other reviews. However, it should be noted that in our series patients with volvulus had the worse outcome in terms of weaning off PN when compared with intestinal atresia.
Type of Study: Retrospective Study.
Level of Evidence: II

Short bowel syndrome (SBS) is the most common cause of intestinal failure in children. Upon diagnosis, early start of an intestinal rehabilitation program is highly recommended to provide the patients with the best chances to achieve enteral autonomy. Together with the establishment of hepato-sparing parenteral nutrition (PN), early introduction of enteral feeds and central line preservation, some patients might benefit from autologous gastrointestinal reconstructive surgery (AGIR), performed with the aim to slow transit time and, therefore, to increase the contact time of enteral alimentation to the mucosa.

Many predictor factors have been investigated in an attempt to define which parameters have the greatest influence on survival and ability to wean off PN. Among them, the length of remaining small bowel, loss of the ileocecal valve (ICV) and colon, sepsis and development of PN-associated cholestasis are the most debated [1,2]. Aetiology of SBS is often reported in studies for demographic purpose. The main underlying conditions leading to a massive intestinal resection in children include gastroschisis, malrotation and midgut volvulus, necrotizing enterocolitis (NEC) and intestinal atresia. Some authors have pointed out that aetiology of SBS is not predictive of survival nor of the ability to wean off PN [3]. However, some other studies have suggested a worse outcome for patients with gastroschisis [4,5].

The aim of this study was to investigate if the aetiology of SBS affects the outcomes, in terms of survival, achievement of enteral autonomy and need of transplantation, in paediatric patients treated with autologous gastrointestinal reconstructive surgery.

1. Methods

All children with SBS who underwent autologous gastrointestinal reconstructive surgery between 2002 and 2012 at Royal Manchester

☆ There is no financial assistance or any potential conflict of interest, real or perceived.
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https://doi.org/10.1016/j.jpedsurg.2018.03.009
0022-3468/

Children's Hospital were retrospectively reviewed. Most of the patients and some of their data were previously used in other publications from the senior authors [6–10].

Demographic data, aetiology of SBS, intraoperative findings, type and timing of the surgical procedure, surgical complications, post-operative follow-up and outcome measures were recorded.

The patients were divided into groups according to the aetiology causing SBS: gastroschisis (either vanishing or complicated), volvulus associated with malrotation, NEC, intestinal atresia and miscellaneous. In the group of gastroschisis we included the patients with either vanishing gastroschisis or gastroschisis complicated by the co-presence of intestinal atresia, volvulus, or perforation. The miscellaneous group included patients with ganglioneuroma, cloacal exstrophy and aganglionosis.

The patients underwent AGIR procedures, among which the longitudinal intestinal lengthening (LILT) procedure was the most performed, followed by the serial transverse enteroplasty (STEP procedure). Among the other less frequently used procedure there were reversed segment, colonic interposition and tapering of the dilated bowel. All these procedures could occur alone or in combination and were performed by two surgeons with extensive expertise.

Intraoperative starting and final small bowel length measurements were obtained using a sterile silk suture placed along the antimesenteric border of the bowel between the duodenojejunal flexure and the end of small bowel. The percentage increase in length after the procedure was calculated for each patient.

The main outcome measures were:

1. *Survival*: patient survival at the end of follow-up was recorded.
2. *Enteral autonomy and time to wean from PN*: for all patients who weaned off PN and all fluids, the time to achieve enteral autonomy, i.e. 100% of calories taken orally, after the AGIR surgery was calculated.
3. *Complications:* number and type of complications related to AGIR surgery were collected.
4. *Transplant:* patients listed for bowel and/or liver transplantation at the time of publication or at the end of reported follow-up were considered.

In addition, the ability to reach intestinal autonomy was analyzed according to age at surgery, independently of the aetiology. Four intervals of age were defined: 0–6 months, 6–24 months, 24–72 months and 72–144 months. Then, the ability to gain intestinal autonomy was studied according to pre-operative bowel length, independently of the etiology. Three intervals of bowel length were considered: <40 cm; 41–99 cm and > 100 cm.

1.1. Statistical analysis

All continuous data were suitable for non-parametric unpaired-data analyses and were reported as median with range. Binomial data were reported as percentage.

Continuous data were compared with the Kruskal-Wallis test. Percentages were analyzed by using the Fisher's exact test.

$p < 0.05$ was considered statistically significant. Data were analyzed with the software GraphPad Prism 6.0.

2. Results

2.1. Demographic data and aetiology of SBS

During the study period, 46 patients (21 boys and 25 girls) with SBS underwent autologous intestinal reconstructive surgery. This represents our entire cohort of patients with SBS for the period considered, as all patients had at least one form of AGIR surgery. Demographic and clinical data are summarized in Table 1. Twenty-nine patients were inborn, while 17 patients were referred to our department after birth. The causes leading to SBS were gastroschisis in 20 patients, volvulus in 10, NEC in 6, and intestinal atresia in 7. Three other patients, referred from other centers, presented with short bowel syndrome due to sequelae after extensive resection of ganglioneuroma in one case and cloachal exstrophy repair in another; the third one had a total colonic and ileal aganglionosis. These three patients were excluded from analysis due to their heterogeneity and small sample size. The remaining 43 patients were divided into 4 groups according to aetiology.

Median birth weight, available in 29 patients, was 2120 g (range 640–3698) while median gestational age at birth was 36 weeks (range 24–40), being available in 38 patients. Gestational age and birth weight were both significantly lower in NEC when compared to volvulus group ($p = 0.0062$ and $p = 0.03$ respectively). Median length of follow up after definitive surgery was 3.7 years (range 0.16–11) and no difference among groups was identified.

2.1.1. Type and timing of AGIR surgery

Half the gastroschisis underwent controlled bowel expansion, 4/10 patients with volvulus, 2/6 patients with NEC and 2/7 with intestinal atresia. The aim was to achieve circumferential expansion of the bowel to about twice its original size and 5 cm was considered the

Table 1
Demographic data, age at surgery, intraoperative findings, surgical complications and outcomes were grouped according to aetiology. Patients from miscellaneous group were excluded

	Gastroschisis	Volvulus	NEC	Intestinal atresia	Overall
N of patients	20	10	6	7	43
Gender M:F	10:10	3:7	3:3	3:4	19:24
Birth weight (g), median, range	2078 (1650–3698)	2600 (2146–3640)	1030* (640–2900)	2412 (1630–2734)	2120 (640–3698)
Gestational age (wk), median, range	36 (32–40)	38.6 (33–40)	27.4* (24–37.8)	34.8 (32–35.7)	36 (24–40)
Bowel expansion	10/20	4/10	2/6	2/7	18/43
Age at surgery (mo), median, range	6 (2–98)	13 (6–113)	17 (5–56)	21 (3–102)	9 (2–113)
Pre-op small bowel length (cm), median, range	35.0 (10–125)	11.5§ [6–20]	34.5 (6–120)	43.0 (15–120)	24.0 (6–125)
Post-op small bowel length (cm), median, range	55 (32–220)	28§ (10–37)	63.5 (12–210)	80 (30–150)	42 (10–220)
Increased length (%), median, range	80% (38–100)	100% (90–100)	84.5% (75–100)	93% (16–100)	86% (16–100)
Presence of ICV (%)	10%°	50%	33%	66%	30%
Follow up (y), median, range	4.9 (1–10)	2.6 (1–6)	3.0 (1–9)	4.0 (1–10)	3.7 (1–10)
Surgical complications	7	0	2	1	10
Patients off PN (%)	70%	30%#	33%	85%	58.1%
Time to wean off PN (mo), median, range	8.5 (4–96)	9.0 (1–96)	3.5 (1–6)	14.5 (4–36)	9.0 (1–96)
Survival (%)	100%	100%	83%	85%	95%
Transplant	0	0	0	0	0

* $p < 0.05$ NEC vs volvulus for birth weight and gestational age.
§ $p < 0.05$ volvolus vs gastroschisis and intestinal atresia.
° $p < 0.05$ gastroschisis vs volvulus and intestinal atresia.
# $p < 0.05$ volvolus vs intestinal atresia.