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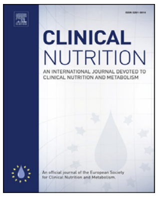

Original article

Long term outcomes of intestinal rehabilitation in children with neonatal very short bowel syndrome: Parenteral nutrition or intestinal transplantation

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SUMMARY

Background & aims: Intestinal rehabilitation is the preferred treatment for children with short bowel syndrome (SBS) whatever the residual bowel length, and depends on the accurate management of long-term parenteral nutrition (PN). If nutritional failure develops, intestinal transplantation (ITx) should be discussed and may be life-saving. This study aimed to evaluate survival, PN dependency and nutritional status in children with neonatal very SBS on PN or after ITx, in order to define indications and timing of both treatments.

Patients and methods: This retrospective cross-sectional study enrolled 36 children with very SBS (<40 cm) who entered our intestinal rehabilitation program from 1987 to 2007.

Results: All the children on long-term PN ($n = 16$) survived with a follow-up of 17 years (9–20). Six of them were eventually weaned off PN. Twenty children underwent ITx: eight children died (40%) 29 months (0–127) after Tx. The others 12 patients were weaned off PN 73 days (13–330) after Tx. Follow-up after transplantation was 14 years (6–28). Seven out of 8 (88%) patients with a history of gastroschisis required ITx. Patients who required ITx had longer stoma duration.

Conclusion: Survival rate of children with very short bowel was excellent if no life-threatening complications requiring transplantation developed. Gastroschisis and delayed ostomy closure are confirmed as risk factor for nutritional failure. Intestinal rehabilitation may allow a total weaning of PN before adulthood. A follow-up by a multidisciplinary team is necessary to avoid PN complications in order to minimize indications for ITx.

1. Introduction

Short bowel syndrome (SBS) is the leading cause of intestinal failure (IF) in children [1]. The normal absorptive capacity of the small intestine is compromised as a result of extensive resection due to either a congenital defect such as intestinal atresia, gastroschisis, or a post-natal ischemia/necrosis secondary to necrotizing enterocolitis, midgut volvulus or vascular thrombosis [2–4]. Small bowel length is estimated at 250 cm ± 40 cm at birth, with a maximal growth during the first year of life [5]. Various definitions of SBS can be found in literature, according either to small bowel length or to parenteral nutrition dependence after surgery [6]. The residual small bowel length has been correlated to PN weaning [7,8].

Although the overall incidence of SBS is only 25/100,000 live births, the mortality rate remains high even with management by experienced teams [9]. Recent data from the pediatric intestinal failure consortium, including 272 infants, reported an incidence of enteral autonomy, death and intestinal transplantation of 47%, 27% and 26% respectively [10].

Abbreviations: IF, intestinal failure; PN, parenteral nutrition; ITx, intestinal transplantation; IR, intestinal rehabilitation; SBS, short bowel syndrome; CVC, central venous catheter; CRBSI, catheter-related bloodstream infection; IFALD, intestinal failure associated liver disease; NPEI, non-protein energy intake; REE, resting energy expenditure.

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Long-term parenteral nutrition (PN) is the first-line treatment for children with IF due to SBS [1]. The aim of PN is to compensate fluids from intestinal losses and provide optimal nutritional supply to support normal growth [11–13].

Patients on long-term PN can develop life-threatening complications such as repeated septic episodes, fluid and electrolyte imbalance, loss of venous access, and intestinal failure-associated liver disease (IFALD) [12]. These patients have recently been categorized as suffering from "nutritional failure" [13]. For these patients, intestinal transplantation is the only life-saving alternative [14].

The Intestinal Transplant Registry reports a 41% graft survival rate at 10 years [15], although individual centers have reported better long-term outcomes [16–18].

This retrospective cross-sectional study aimed to evaluate survival, PN dependency and nutritional status in children with neonatal very SBS (<40 cm) followed in the same center with an integrated and multidisciplinary care, on long term-PN or after intestinal transplantation (ITx).

2. Patients and methods

All children suffering from neonatal SBS born between 1987 and 2007 and referred to our center for intestinal rehabilitation (IR) after extensive small bowel resection in the neonatal period were included. All children started total PN immediately after the surgery. Residual small bowel length was measured along the anti-mesenteric side starting from the ligament of Treitz, either during resection or at the time of ostomies closure. The length for inclusion was defined as less than 40 cm. Beyond this cut-off a predictive value for achievement of enteral autonomy was demonstrated [8,19].

A Broviac-type central venous catheter (CVC) was placed in the superior vena cava of all children enrolled. PN infusion included dextrose, crystalline amino acids, electrolytes, trace elements and vitamins and lipid emulsion. The amino acids were Vaminolact® (Fresenius Kabi, Bad Homburg, Germany) or Primene® (Baxter; Maurepas, France) for children less than 10 years old and Vintene® (Baxter; Maurepas, France) for older children. Lipids were provided by Clinoleic® (Baxter; Maurepas, France) before 2008 (soybean oil and olive oil), and then by SMOF lipid® (Fresenius Kabi, Bad Homburg, Germany) from 2009 (soybean oil, medium-chain triglycerides, olive oil, and fish oil). Lipid intake did not exceed 30% of non-protein energy intake (NPEI). PN was completed with electrolytes, trace-elements (Aguettant®), and vitamins (Cernevit®, Baxter).

Follow-up for patients on PN was performed in multidisciplinary outpatient clinic by a gastroenterologist, pharmacist, dietitian, and a specialized nurse 4 times a year. The adaptation of PN solution were made by clinicians to assure normal growth. Progressive weaning with reduction in weekly infusions and calories was made when possible. The resting energy expenditure (REE) was calculated with Schofield equation [20]. Ratio between NPEI/REE is routinely used in our clinical practice to evaluate level of PN dependency. The PN was suspended when the PN total caloric intake was less than 20% of NPEI/REE, while growth velocity remained normal. Oral feeding in patients on long-term PN was encouraged to promote intestinal adaptation [21]. Enteral tube feeding was used only in children with severe eating disorders. Feeding was started after small bowel resection as soon as intestinal transit had recovered [21], preferentially oral feeding with semi elemental diet containing medium chain triglycerides, such as Pregestimil® (Mead Johnson), Alfaré® (Nestlé) or Peptijunior® (Nutricia). Complementary feeding was progressively increased up to a normal diet. Children were considered for ITx if they developed severe complications following the guidelines of the American Society of Transplantation [22]. The indications were: severe IFALD, recurring life-threatening sepsis, exhaustion of central venous access (thrombosis of two of the four available standard access sites) and impossibility for the care-givers to perform home-PN. All ITx were performed in our institution with the previously described procedures [23].

Data was collected at a single time point, on December 31st, 2016 for the latest regular follow-up or the latest follow-up before transition to the adult department. Data included demographics, growth parameters, intestinal anatomy, liver function tests, liver biopsy if available, complications related to central venous catheter (CVC) – infection, thrombosis, breakage – CVC replacements and date and cause of PN discontinuation.

All measurement of weight was performed using a digital scale, and height using a stadiometer.

Body mass index (BMI) was calculated as weight in kilograms divided by height in meters squared. To evaluate height weight and BMI values across different age and gender groups we used the *Z*-score that were calculated according to the Center of Disease Control and Prevention growth charts of 2000 [24]. Children with BMI values lower than the 5th percentile for age and gender were classified as "underweight", those in the 5th to 85th percentile were classified as "normal weight", those in the 85th and 95th percentile were classified as "overweight" and those greater than the 95th percentile were classified as "obese". Height of both parents was used to calculate the genetic target height, using Tanner's formula [25]. The number of central venous catheters and catheter-related bloodstream infections (CRBSI) for each child was given per 1000 catheter-days.

Liver function tests (transaminases, bilirubin, or gamma-glutamyl-transpeptidase) were monitored every 3 months to screen possible IFALD onset. A liver biopsy was discussed with pediatric hepatologist when abnormal liver function tests persisted for more than six months, especially in the presence of other signs of liver disease (hepatomegaly, portal hypertension) if the screening for other causes was negative. All patients evaluated for ITx underwent a liver biopsy [26]. The METAVIR score was used to grade histological findings of liver biopsies.

2.1. Ethics

This retrospective cross-sectional study was approved by the local ethical committee. All data were made anonymous for the analysis.

2.2. Statistical analysis

Statistical analyses were performed with SPSS version 20.0 (SAS) and Microsoft Office Excel 2010. Results are expressed as median (min, max), mean ± standard deviation or numbers (percentage). Characteristics at referral and outcome data were analyzed by using non-parametric tests: Mann–Whitney, Kruskal–Wallis and Wilcoxon signed rank tests. Chi square distribution was used for comparison of percentages. A Kaplan–Meier curve was used for ITx progression, survival and PN weaning. A p value less than 0.05 was considered as statistically significant.

3. Results

Thirty-six children with ultra-short bowel syndrome were included. Causes were: midgut volvulus ($n = 13$), intestinal atresia ($n = 11$), gastroschisis with or without intestinal atresia ($n = 8$) and necrotizing enterocolitis ($n = 4$).

Median age at referral to our center was 9 months (3–72). Among the patients referred, seventy percent were born premature, but all of them were referred to our center after the completion of term. Small bowel length measurement was made in 85% of cases after term completion.

All patients were managed on long-term PN: 16 patients (group A) remained on PN program (home-PN) and 20 patients (group B) received an intestinal allograft with or without the liver (Fig. 1). Their characteristics are reported in Tables 1–3. As shown volvulus was the most frequent cause of USBS in group A 8 (50%) while gastroschisis the most frequent in group B 7 (35%) (Fig. 2). No significant difference between both groups was found for small bowel length, presence of ileocecal valve and colon integrity, except for a non significant trend towards shorter time with ostomy in children not requiring ITx.

Seven children out of 36 underwent a longitudinal intestinal lengthening procedure: four underwent a longitudinal intestinal lengthening and tailoring and three a serial transverse enteroplasty. Despite the surgical lengthening three patients required an ITx.

The number of CVC replacements and CRBSI per 1000 days of PN were significantly lower in group A than in group B. Two children (11%) in group A and nine children (37%) in group B had CVC-related thrombosis ($p = 0.04$) (Table 3).

Eight patients in group A (50%) underwent liver biopsies (Table 1). METAVIR was F0 in one, F1 in one, F2 in three, F3 in two and F4 in one. Among them 3 (38%) normalized their liver function tests during follow-up, and the one with liver cirrhosis is stable with normal liver function test and weaned from PN. Two patients developed transient abnormal liver function tests at last

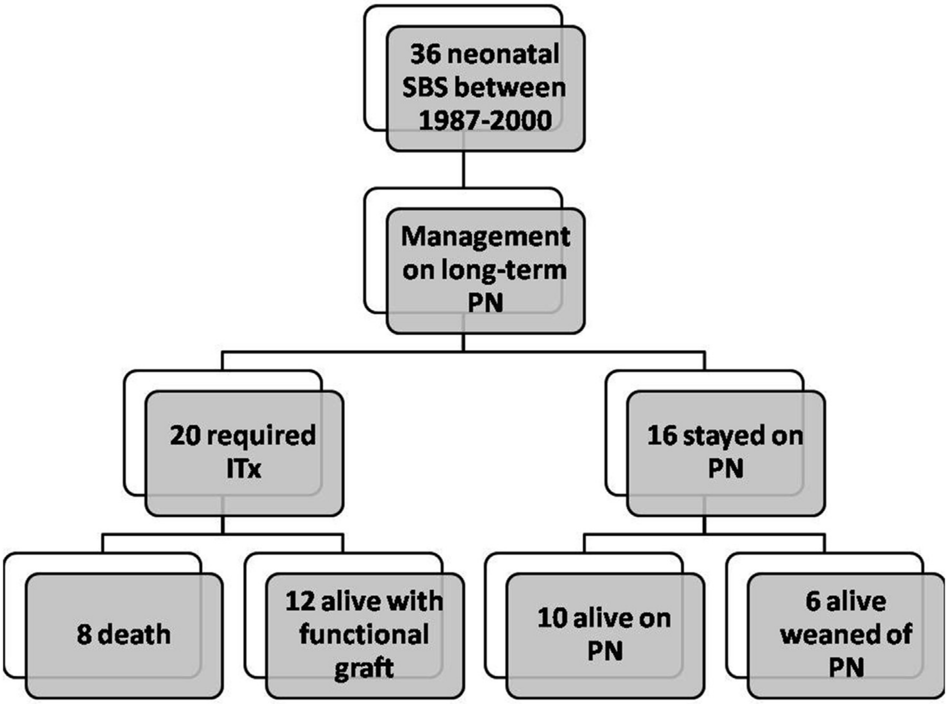

Fig. 1. Patients flow chart. SBS: short bowel syndrome; PN: parenteral nutrition; ITx: intestinal transplantation.

Table 1
Characteristics of patients on long term parenteral nutrition (Group A).

Patient no.	Gender	Age at last FU (y)	Cause of SBS	Bowel length, cm + surgery	ICV	Colon	Time on PN (y)	METAVIR
1	F	18.5	Volvulus	7	Yes	Entire	18	
2	M	17.5	Volvulus	11	Yes	Entire	17.5	
3	M	18	Volvulus	23	Yes	Entire	18	
4	F	20	Atresia	32 + STEP	No	24 cm	20	F3
5	M	20	Atresia	35 + LILT	No	Entire	2	F1
6	F	19.5	Apple peel atresia	30	Yes	Entire	2.5	F4
7	M	17.5	Angiomatosis	25	Yes	Entire	2	
8	F	17	Gastroschisis	24	No	Entire	17	
9	M	19	Volvulus	5	Yes	Entire	19	F3
10	F	9	Volvulus	3	Yes	Entire	9	
11	M	18	Volvulus	37	No	Entire	17.5	
12	M	17	Volvulus	8	No	Entire	17	F2
13	F	11	Volvulus	5	No	Left	11	F0
14	M	11.5	Enterocolitis	34 + LILT	No	Left	11.5	F2
15	M	12	Atresia	15 + STEP	No	Left	12	F2
16	F	10	Apple peel atresia	19	No	Entire	5	

ICV: ileo-caecal valve; PN: parenteral nutrition; SBS: ultrashort bowel syndrome; y: years; M: male; F: female; FU: follow-up.
F1: mild portal fibrosis; F2: bridging fibrosis in less than 50% of portal spaces; F3: bridging fibrosis in more than 50% of portal spaces; F4: cirrhosis.
LILT: longitudinal intestinal lengthening and tailoring; STEP: serial transverse enteroplasty.

Table 2
Characteristics of patients who underwent transplantation (Group B).

Patient no.	Gender	Age at last FU (y)	Cause of SBS	Bowel length, cm + surgery	ICV	Colon	Time on PN (y)	METAVIR	ITx indication	ITx type
1	M	Death 2.5	Gastroschisis	5	No	Left	2.5	F4	IFALD	L-SBTx
2	M	Death 6.5	Atresia	35	No	Entire	6.5	F2	CVC sepsis	SBTx
3	M	Death 16.5	Gastroschisis	25	No	Left	9	F2	IFALD	SBTx
4	M	Death 12.5	Gastroschisis	10 + LILT	Yes	Entire	5.5	F2	CVC sepsis	SBTx
5	M	Death 5	Gastroschisis	4	No	Rectum	4.5	F3	CVC sepsis	SBTx
6	M	Death 14.5	Volvulus	23	No	Entire	8.5	F4	IFALD	MVTx
7	M	Death 4.5	Enterocolitis	15	No	Sigmoide	4.5	F1	CVC sepsis	SBTx
8	M	Death 2.5	Volvulus	15	No	Left	2.5	F4	IFALD	L-SBTx
9	M	19	Atresia	18	No	Entire	3	F4	IFALD	MVTx
10	M	20	Gastroschisis	25 + LILT	No	Left	4.5	F4	IFALD	L-SBTx
11	M	12.5	Enterocolitis	15	Yes	Entire	3.5	F2	IFALD	L-SBTx
12	M	12.5	Enterocolitis	10	No	15 cm	6.5	F4	IFALD	MVTx
13	M	21.5	Atresia	15	No	Left	3	F2	CVC sepsis	SBTx
14	M	18	Apple peel	10	Yes	Entire	6	F2	CVC thrombosis	SBTx
15	M	15.5	Volvulus	10	Yes	Entire	4.5	F2	CVC thrombosis	SBTx
16	M	15.5	Volvulus	5	Yes	Entire	4.5	F2	CVC thrombosis	SBTx
17	M	13	Volvulus	10	Yes	Entire	3	F1	CVC thrombosis	SBTx
18	M	12.5	Gastroschisis	30	No	Entire	5.5	F2	CVC thrombosis	SBTx
19	F	12.5	Gastroschisis	10 + STEP	Yes	Entire	6	F4	CVC thrombosis	SBTx
20	F	18	Volvulus	5	No	Transverse	2.5	F1	CVC sepsis	SBTx

ICV: ileo-caecal valve; PN: parenteral nutrition; USBS: ultra-short bowel syndrome; y: years; M: male; F: female.
F1: mild portal fibrosis; F2: bridging fibrosis in less than 50% of portal spaces; F3: bridging fibrosis in more than 50% of portal spaces; F4: cirrhosis.
IFALD: intestinal failure-associated liver disease.
SBTx: small bowel transplantation; L-SBTx: liver-small bowel transplantation; MVTx: multivisceral transplantation.
LILT: longitudinal intestinal lengthening and tailoring; STEP: serial transverse enteroplasty.

Table 3
Anatomical characteristics and long term PN complications.

	PN, $n = 16$	ITx[a], $n = 20$	p
Female (%)	7 (44%)	2 (10%)	0.020[b]
Age entering intestinal rehabilitation (months)	10 (3–29)	6 (4–72)	0,82
Jejunal length (cm)	10.7 (0–37)	9.5 (0–30)	0.77
Ileal length (cm)	8.5 (0–30)	4.8 (0–35)	0.28
Absence of ICV, n (%)	9 (56)	13 (65)	0.42
Absence of entire colon, n (%)	4 (25)	9 (45)	0.19
Stoma duration (days)	162 (0–1460)	477 (0–4927)	0.07
Follow up of living patients (y)	15.9 (6–20)	15.7 (12–21)	0.87
Number of CVC (n/1000 PN days)	1.8 (0.3–6.7)	4.3 (1.7–15)	<0.001[b]
Rate of CVC related sepsis (n/1000 PN days)	1.6 (0.2–5.4)	4.1 (0.6–12)	0.001[b]
Patients with at least 1 venous thrombosis, n (%)	2 (13)	9 (45)	0.04[b]

PN: parenteral nutrition; ITx: intestinal transplantation; CVC: central venous catheter; ICV: ileo-caecal valve.
Values are expressed in median (range) or number (%).
[a] Before transplantation.
[b] Significant.

follow-up which do not meet the criteria for liver biopsies for the moment.

Twenty children received 21 grafts (group B), 13 of them received an isolated small bowel, four a combined liver and small bowel and three a multi-visceral graft. ITx was performed at a median of 4 years old (4 months to 14 years). All patients who received a liver-containing graft had end-stage IFALD, except for one who had a moderate fibrosis but a liver arteriovenous malformation and severe cholestasis. In the patients who received an isolated small bowel graft, METAVIR was F1 in three, F2 in eight, F3 in one (Table 2). One patient was re-transplanted for acute rejection (multivisceral transplantation) and died of surgical complications.

Because of the long time frame of enrollment (20 years) an analysis of the progression to ITx requirement was performed according to 3 equal time period (Fig. 3). The 3 time period showed a similar rate of progression to ITx of approximately 40% of patients.

3.1. Survival

Overall, 78% of patients survived (Fig. 4). All children in group A were alive at last follow-up at a mean age of 16 years (9–20). Eleven (69%) had achieved their puberty.

The transplanted patients' survival was 60%, not different between isolated small bowel or liver-containing ITx. Death occurred at a mean of 29 months (0–127) after ITx. All but one death (traffic accident) were related to transplantation (acute rejection in three cases, surgical complications in three cases, sepsis in one case). Transplanted patients were 16 years old (12–21) at last follow-up and 14 years (6–28) after ITx.

3.2. Nutritional status

Six children (38%) in group A were weaned off PN after a mean of eight years (Fig. 5). No significant difference of small bowel length were seen between weaned or PN dependant patients. One child needed enteral tube feeding to achieve weaning off PN. Another child needed to re-start PN during puberty because enteral absorption was insufficient to guarantee the caloric needs of pubertal growth acceleration. The other ten children still on PN had a median need of 83% (39–163) of NPEI/REE. The median number of PN infusions was 4 (2–7) per week. Two of them required additional enteral tube feeding. All patients in group B were weaned off PN.

At latest follow-up no significant difference was seen in *Z*-score for height, weight and BMI in alive patients between both groups (Table 4). On a total of 28 alive patients, eight (29%) were defined underweight, those patients were equally divided in group A (31%) and group B (25%). The rest of the patients had a normal weight. Moreover 4 patients (14%) presented a height < 3rd percentile for sex and age. Those patients were equally divided in groups A and B. Weaning of PN was not associated to lower BMI's. Eighteen patients (66%), eleven in group A and seven in group B had completed growth without pubertal delay. Their final height was significantly shorter than their calculated genetic target height ($p < 0.001$) regardless to the group.

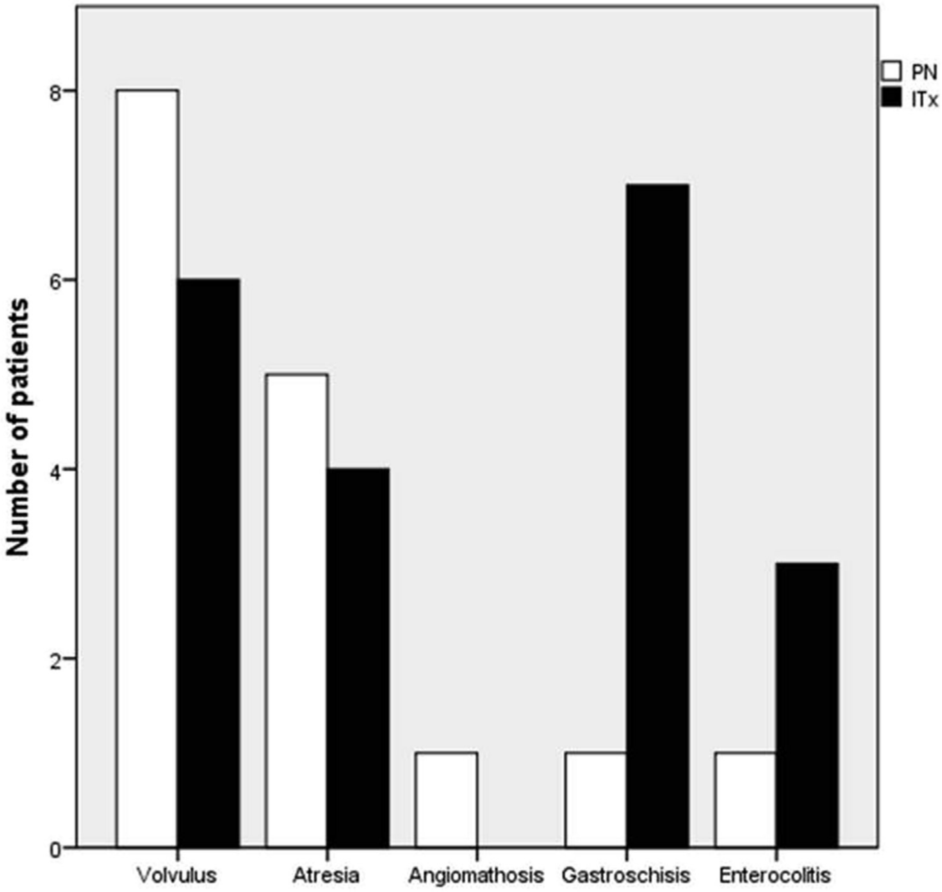

Fig. 2. Short bowel syndrome reason divided according to intestinal transplantation need. PN: parenteral nutrition; ITx: intestinal transplantation.

4. Discussion

This study describes the outcomes of 36 children with neonatal short bowel syndrome after at least 10 years of follow-up in a intestinal rehabilitation center. Seventy-eight percent of the patients survived. All deaths occurred after ITx.

To our knowledge, this is the first cross-sectional study focusing on long-term outcomes in children with neonatal SBS whatever the treatment. The other series focusing on SBS were focusing either on PN [27,28], or on ITx outcomes [29].

The European Society of Parenteral and Enteral Nutrition promoted a large multicenter prospective study on survival rate of patients on long-term PN, published in 2008 [30] and in 2011 [31], with a three and five years follow-up respectively. The study enrolled both adults and children with all causes of IF. The overall survival rate was 92% at 3-year follow-up and decreased to 90% at 5 years. The authors reported a survival rate of 87% that decreased to 73% for ITx candidates with nutritional failure over a 5-year period. In the same study 14% of patients were transplanted and 54% survived. Our patients' survival rate is close to these results. In our cohort, all children suffering from nutritional failure with a very high risk of death underwent intestinal transplantation. No patient died while on the waiting list.

Survival rate of children with SBS on long-term PN (100%) compared to survival rate after ITx (60%) confirms that PN is the primary therapeutic and safest option for these patients. Intestinal transplantation should be considered as a life-saving procedure in cases of life-threatening complications leading to nutritional failure, as already emphasized [13].

Rate of CVC sepsis and thrombosis with consequent CVC replacement are confirmed as prognostic factor of ITx requirement. Results from our cohort seem to suggest that IFALD could be a reversible condition if managed in an expert center for intestinal rehabilitation [32].

A great effort in the prevention of PN-associated complications has been made in the last decade, with the introduction of positive pressure valve needleless connectors [33] and taurolidine locks [34], development of strategies to reduce intestinal bacterial overgrowth and the use of fish-oil based lipid emulsions [35]. Indeed, we observed in our center a reduction of transplant rate for SBS in the last few years. The latest ITx for SBS was performed in 2012. Unfortunately the time frame choice of this study does not allow us to show evidence in this cohort.

Nevertheless patients with nutritional failure should be early referred to a specialized multidisciplinary intestinal rehabilitation center [36]. Intestinal transplantation should be discussed on a case-by-case basis because, even if it remains a difficult procedure, it can offer successful rehabilitation in carefully selected patients [15,23,37,38].

Results from our study, with seven out of the eight gastroschisis included requiring ITX, confirm gastroschisis as a risk factor for the progression to nutritional failure requiring ITx [39]. Stoma duration seems also to play a role in this progression. Due to small numbers we cannot drive conclusions on the impact of small bowel lengthening procedure on the progression to ITx and on the likelihood of achieving enteral autonomy.

Results on PN weaning are also in line with the literature. Pironi et al. reported a 24% weaning rate in a multicenter cohort of adults

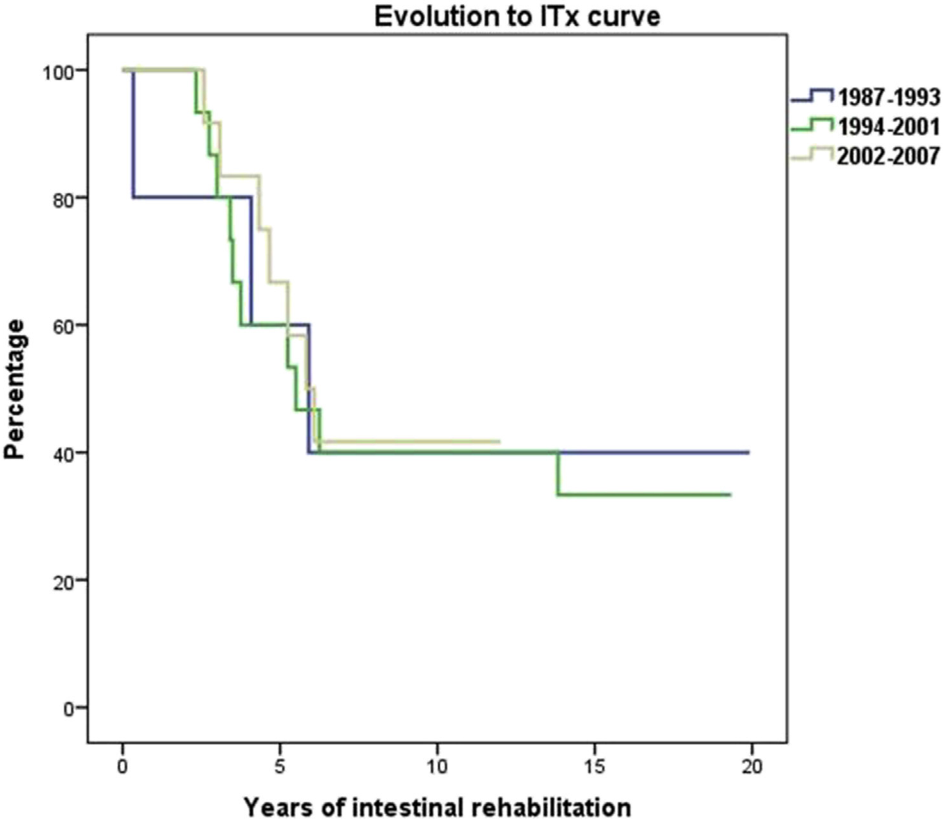

Fig. 3. Progression to intestinal transplantation according to three equal time period. ITx: intestinal transplantation.

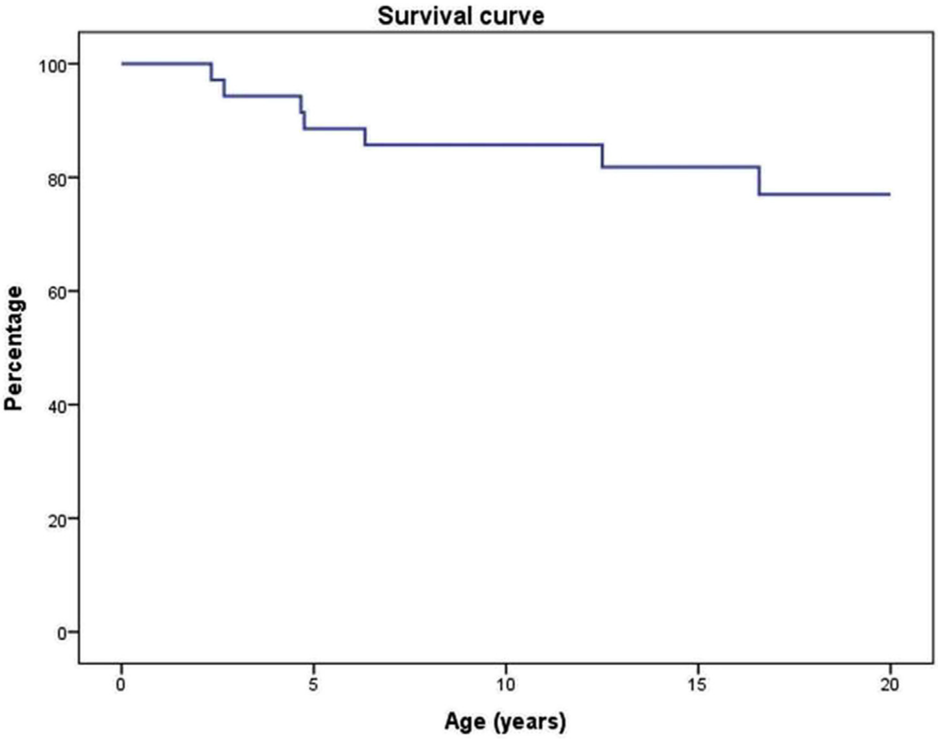

Fig. 4. Survival curve of the cohort.

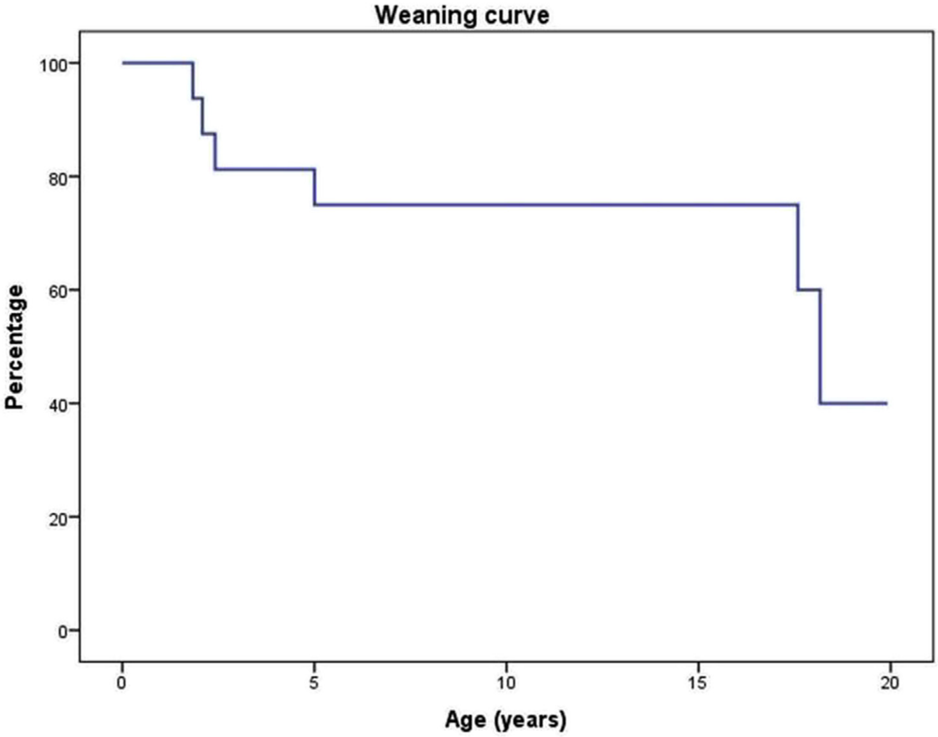

Fig. 5. Weaning curve of patient who stayed on long term parenteral nutrition (n: 16).

Table 4
Nutritional outcome at follow-up.

	PN, $n = 16$	ITx[a], $n = 12$	p
Z-score for height	−0.21 ± 1.4	−0.72 ± 1.1	0.31
Z-score for weight	−0.89 ± 1	−1.26 ± 1	0.31
Z-score for BMI	−0.99 ± 1	−1.12 ± 0.9	0.72

PN: parenteral nutrition; ITx: intestinal transplantation.
Values expressed in mean ± SD.
[a] Alive at follow-up.

and children [31] while a study in children with USBS reported 48% of PN weaning [27]. The two important periods for PN weaning in SBS children are between 2 and 4 years of age and between 16 and 18 years when puberty is achieved (Fig. 5). We observed that even when the remnant small bowel length (<10 cm) could suggest a definitive intestinal failure: a well-managed intestinal rehabilitation may lead to intestinal sufficiency. Thus, trials of PN weaning should be systematic before considering ITx in the absence of short-term life-threatening complications [13].

In the patients on long-term PN, the PN dependency decreased over time to 4–5 weekly infusions improving acceptability and quality of life. Also, these patients might become candidates for drugs increasing intestinal absorption – such as teduglutide – currently tested in children [40].

Long-term growth in these children was comparable to previous reports [19]. However, we observed that approximately a third of patients were underweight and a significant difference between final and target height. No benefic effects of ITx or enteral autonomy on growth were shown. This should be carefully followed through infancy and childhood in order to re-start nutritional support if weight gain is inadequate or growth velocity decreases – especially in the pre- and per-pubertal period. Other factors (prematurity, hormonal changes ecc.) may play a role in growth for patients with neonatal SBS with adequate nutritional intake (enteral and/or parenteral).

Our study has several limitations. Only a small number of patients could be included which limited the statistical analysis. Short bowel syndrome is a rare condition which explains the limited number of patients and the retrospective design of the study. Also, the study covers a very long time frame which might induce a calendar bias. Outcomes of ITx have improved [15,23,41] because of better patient management, new immunosuppressive drugs, improvement in timing for transplantation, surgical techniques as well as diagnosis and treatment of humoral rejection [42]. In the first years of ITx, the indications were also larger because of the lack of specific guidelines [22], which could explain the number of transplanted patients in our cohort. However, the long time frame of the study allowed us to report accurate data about long-term survival, growth and adult height. Finally, because of the retrospective design of the study we cannot exclude that the children who underwent ITx might have died in the short-term if left on parenteral nutrition – although such predictions are hazardous.

In conclusion, we confirm that long-term PN is the first-line treatment for children with neonatal SBS – with good long-term prognosis compared to intestinal transplantation. An expert intestinal rehabilitation team composed by gastroenterologists, dieticians, pharmacists, PN-specialized nurses and both transplant and non-transplant surgeons may be beneficial in the multidisciplinary management of children with SBS. The aim should be the early prevention of intestinal failure-related complications and the achievement of PN weaning with good nutritional status, growth and development, in order to limit the indications for intestinal transplantation. Treatments to increase enterocyte absorption currently under study should further improve the long-term management and outcomes of such patients. Despite its difficulties and insufficient results, intestinal transplantation should be considered in case of nutritional failure with careful timing and management to maximize survival chances.

Source of founding

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Conflict of interest

None.

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