# Acceptance type: Intestinal Failure Poster

# \*Poster of Distinction

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## Inflammatory Bowel Disease-Like Findings in Pediatric Intestinal Failure Patients

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#### Abstract

Introduction: Intestinal failure (IF) in children is an uncommon but highly morbid condition. As IF management improves and long-term survival rate increases, its physiological complications have become more apparent. The development of Inflammatory Bowel Disease (IBD) in this population has been reported but the literature describing it in detail is sparse. The present study was designed to characterize children with intestinal failure who developed an inflammatory bowel disease-like disorder and identify the potential predisposing clinical factors.

Methods: This is a retrospective study based on electronic medical records of pediatric patients seen at the Cincinnati Children's Hospital Medical Center between January 2000 and July 2022.  Demographic and medical history data were collected and compared between a group of short bowel syndrome patients with an IBD-like illness and a control group of sex and age-matched short bowel patients without an IBD-like disorder. Univariate analysis was performed comparing the following variables: gender, race, gestational age, diagnosis, and bowel length.

Results: During the follow up period, 23 children were diagnosed with jejunitis or colitis, and compared with 23 control patients with surgical short bowel syndrome but without jejunitis or colitis. Of the 23 with IBD-like findings, 8 had jejunitis and 7 had colitis. Of the patients with jejunitis or colitis, 12 (52%) were males, with a median age of 4.5 (3, 7) years at diagnosis of their IBD-like illness. Nearly one third of those patients had gastroschisis (31%), followed by necrotizing enterocolitis (26%) and malrotation and volvulus (21.7%). None of the variables had statistically significant differences between the 2 groups. The frequency of metronidazole use to control suspected small intestinal bacterial overgrowth was no difference between the two groups (17 out of 23 versus 15 out of 24, p=0.5). However, more children in the IBD-like group lacked an ileocecal valve and adjoining distal ileum compared to the control group (15 patients ,65% vs. 8 patients, 33%). Moreover, more children in the IBD-like group had undergone a prior lengthening procedure than the non-IBD-like group (5 patients, 21.7% vs.0, respectively).

Discussion: Short bowel syndrome patients are at risk of relatively early onset IBD. The absence of an ileocecal valve (and adjoin ileum) and prior lengthening procedures emerge as factors associated with the risk of jejunitis or colitis in these patients. Therapy with metronidazole appears ineffective in aborting or treating these inflammatory conditions.

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## The Management and Outcome of Catheter Related Bloodstream Infection with Staphylococcus aureus in Pediatric Patients with Chronic Intestinal Failure receiving Home Parenteral Nutrition

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#### Abstract

**Introduction**

Catheter related blood stream infection (CRBSI) with Staphylococcus aureus is known to be a serious infection with a high mortality rate in adults and a high risk of infectious complications. Therefore, removal of the catheter is advised when S. aureus is detected. However, paucity of data in pediatric patients with chronic intestinal failure (IF) receiving home parenteral nutrition (HPN) exists and current management is based on data retrieved from adult patients. Especially in this patient group, removing their lifeline is not preferable due to risk of loss of vascular access. More information about the clinical course in this specific patient group is needed in order to evaluate the effectiveness and safety of the current management in the Emma Children’s Hospital / Amsterdam University Medical Centers which is an expertise center in HPN and chronic IF. Currently, standard care consists of preventive taurolidine catheter lock solution for all patients every day. When they are having a positive blood culture with S. aureus and when they are hemodynamically stable, two weeks intravenous flucloxacilline will be given. In case of recurrent fever, new cultures will be taken. In case of a positive blood culture with S. aureus, the central line will be removed.

**Methods**

This is a retrospective chart review to collect data on prevalence, treatment methods and outcomes including complications (e.g. arthritis, osteomyelitis, endocarditis, abscess, pneumonia) of CRBSI with S. aureus in patients aged < 18 years with chronic IF receiving HPN,  treated in the expertise center Emma Children’s Hospital between October 2013 and October 2022.

**Results**

A total of 74 pediatric patients on home parenteral nutrition were treated between 2013 and 2022 in Emma Children’s Hospital, see table 1. A percentage of 38% (n=28) of all patients were diagnosed with S. aureus bacteremia. These 28 patients had a total of 52 infections with a mean number of 1.9 (±1.2) infections per positive patient. The catheter was removed in 56% of the infections (n=29) and not removed in 44% (n=23). Mean number of days the catheter was removed before a new one was inserted was 2 (±2.4). Out of 28 patients with an infection, 4 presented with complications at admission including arthritis (n=2), febrile convulsions (n=1) and both arthritis and febrile convulsion (n=1). These complications were not related to whether the catheter was removed or not as the complications were present at admission.

**Conclusions**

Our results show the importance of tailor-made management for this specific patient group. Almost half of the cohort diagnosed with S. aureus bacteremia did not undergo catheter removal without extra complications during admission. More research about the clinical course of  S. aureus bacteremia in pediatric patients with chronic IF on HPN is needed in order to create an evidence-based protocol for this patient group to prevent unnecessary loss of access in these children.

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## Are we using all the available techniques for the surgical treatment of SBS patients?

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#### Abstract

Background

The surgical management of short bowel syndrome (SBS) focuses on autologous gastrointestinal reconstructive surgery (AGIR). AGIR follows the principles of bowel conservation, restoration of intestinal continuity, lengthening, tailoring and procedures to slow transit time, but the best surgical option is still unknown. More than one surgical procedure could be combined to achieve a better outcome with tailored treatment. The present study investigates the safety and utility of combined procedures performed simultaneously or serially on SBS patients.

Methods

Data were obtained retrospectively from SBS patients’ databases of two European tertiary centres. All patients examined underwent combined procedures, defined as more than one AGIR used on the same patient. Patient variables included demographics, primary diagnosis, operative information, complications, and outcome. Data were presented as median and IQR. Wilcoxon signed rank was used for all paired analyses. Results Combined AGIR techniques were performed on twenty-one children (12 females), with a median age at surgery of 16 months (IQR: 8-63 months). AGIR procedures were performed simultaneously in 15 patients and sequentially in six. Preoperative small bowel length was 20 cm (IQR: 15–35 cm), and postoperative was 35.5 cm (IQR: 30.75–50.50 cm) (P &lt; 0.001). Intraoperative loss of the second LILT loop due to vascular damage was reported in two patients. Three bowel obstructions were recorded due to anastomotic strictures at 9.2 years (IQR: 7.55–9.78 years) of follow-up. After combined AGIR procedures, two patients were weaned entirely from parenteral nutrition (PN), and the remaining patients were on a weaning regime of 4 nights (IQR: 3–4 nights).

Conclusions

Combined AGIR procedures could be considered effective surgical management to create suitable intestinal length and improve intestinal functionality. These techniques should be individually tailored to each patient and can be performed simultaneously or sequentially. AGIR procedures are essential in the surgical treatment of SBS, but their use should be electively indicated and performed by specialized surgeons within dedicated intestinal rehabilitation centre.

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## Where do SBS/IF patients and caregivers go to receive information, and what do they want more information about: findings from a cross-sectional community-driven survey

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#### Abstract

**Introduction:** It remains unclear where members of the SBS/IF community access disease-specific information and what their information needs are. This study investigated information access and priorities among SBS/IF patients and caregivers.

**Methods:** A cross-sectional survey, developed by community members to capture the needs, priorities, and information-seeking behaviors of SBS/IF patients and caregivers, was disseminated through patient support organizations and relevant social media groups. Bivariate cross-tabulations were calculated between socio-medical-demographic variables and information access and needs; multivariate logistic regression was used to predict information access.

**Results:** 320 eligible respondents completed the survey; a majority were non-Hispanic white (90.6%), female (89%), and located in North America (83.3%). Half of respondents (49.5%) were receiving IRP care; 52.2% were adults with SBS/IF, with the remainder caregivers of children (36.2%) or adults with SBS/IF (11.6%). Regardless of patient- and IRP- status, specialist providers/teams (63.7%) and social media support groups (49.8%) were the most frequently accessed sources of SBS/IF-specific information. In multivariate analyses, IRP management was positively associated with the odds of frequently obtaining information from a specialist provider/team (OR=4.19, p <.001); adult patients and their caregivers had higher odds of frequently accessing information from a GP (OR=3.57, p<.01). Years since diagnosis was negatively associated with the odds of frequently accessing information from specialist providers/teams (OR=0.97, p <.05), peers (OR=0.96, p<.05), social media support groups (OR=0.96, p<.01), and the internet (OR=0.95, p <.01). Regardless of patient- or IRP- status and time since diagnosis, respondents most often reported desiring more information about long-term outcomes (63.8%) and quality of life (60.6%). In bivariate analyses, adult patients and their caregivers less frequently desired information about a range of items; the one exception was where to find experienced providers (51.5% of adult patients/caregivers vs. 31.9% of pediatric caregivers). Information needs were lower among those further out from diagnosis relative to those who were more recently diagnosed. Still, a substantial share of patients/caregivers 12+ years post diagnosis desired receiving information about medications (46%), diet (44.6%), finding experienced providers (43.2%), daily living (41.9%), relevant research (40.5%) and mental health (40.5%).

**Conclusions:** These data suggest that, in addition to specialist providers, social media support groups are a prominent avenue through which SBS/IF patients and caregivers access disease-specific information. Information needs are most pronounced in the first years after diagnosis, but even those many years post diagnosis continue to desire information to better understand and manage their condition and achieve the quality of life they desire.

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## PRELIMINARY RESULTS OF THE REGISTRY OF EUROPEAN CHILDREN TREATED WITH TEDUGLUTIDE

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#### Abstract

Introduction: Teduglutide is now an available treatment in Europe to improve intestinal absorption for children with short bowel syndrome (SBS). The extent of response of children to teduglutide and, and the profile of candidates expected to respond have not been clarified yet. For this reason, we established a European registry on the real-life use of teduglutide with the aim of measuring treatment response and predictors of response, safety and adverse events and cost effectiveness of the drug.

Methods: A multi-center, observational, longitudinal, retrospective and prospective cohort study was designed. A registry was created in order to collect all data from previously treated patients and enrolling prospectively children starting teduglutide from June 2021. Response to treatment was analized based on different parameters.

Results: Children enrolled in the registry are currently 73 from six countries in Europe. We were able to perform analysis on 69/72 who reached a minimum of 6 months treatments. Median age at Teduglutide start was 5.4 years old (IQR: 3, 9). More than a half (64%) were preterm. The most preminent coause of SBS was necrotizing enterocolitis (NEC) (27.5%). The median length of residual small bowel was 26 cm (IQR: 14, 55), 16% had a class I SBS, 59% class II, and 24% class III. Ending stoma was present in a minority of patients 19%. Median parenteral nutrition dependency index (PNDI) was 60% before starting treatment. Reduction of at least 20%, 50% or 75% of PNDI was achieved in 73%, 46% and 33% respectively. The analysis on growth pattern was performed on 46 (67%) patients which achieved the 12-month follow-up. Patients achieving a sustained response of over 75% of PN reduction were 19 (41%) and showed a mean increase of 7.8% of body weight after 3 months of treatment, 14% after 6 months, 23.7% after 9 month and 29.7% after 12 months of treatment, which were significantly higher (p: 0.035) than their non responder pairs (figure 1). When considering a lower response rate of > 20% or >50% PN no significant weight increase difference was observed.

Conclusions: Teduglutide is an effective treatment for children with SBS with nearly one third of patients achieving a stable reduction of more than 75% of PNDI. Children with SBS on home PN display an important weight for age retardation before starting Teduglutide treatment. Children achieving an almost complete PN weaning after Teduglutide show a higher weight gain despite a lowering PN support in the first year after treatment.  Monitoring weight gain during the first year on Teduglutide treatment may help identifying children which will most benefit from Teduglutide treatment.

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## Overview of medication use in patients with chronic intestinal failure

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#### Abstract

**Introduction**Oral drug therapy may be compromised in chronic intestinal failure (CIF) because of alterations in absorption and transit. However, only scarce literature is available on which medication CIF patients take in daily life. The aim was to describe the medication used by patients with CIF on home parenteral support (PS).

**Methods**A medication history was obtained from adult CIF patients treated in our tertiary care CIF center, either face-to-face or by telephone interview. This included all orally and parenterally administered (prescribed and ‘over the counter’) medication and micronutrients on top of their daily supplementation with PS. Patient characteristics, degree of polypharmacy, total monthly medication cost (from a societal perspective), route of administration, formulation, drug classes and Biopharmaceutics Classification System (BCS) classes were analyzed. Patients were divided into SBS and non-SBS patient cohorts.

**Results**From October 2019 until December 2020, 72 patients (35 short bowel syndrome (SBS) and 37 non-SBS patients) were included. Median age and body mass index were 59.5 years [IQR 51.8-69.2] and 20.5 kg/m2 [18.9-22.7], respectively. The majority of patients (76.4%) were female. The median volume of PS per week and amount of days per week on PS were 6.0 L [4.5-11.9] and 5 days [3-7], respectively. Polypharmacy (at least five drug preparations), was seen in 85.7% of SBS and 75.7% of non-SBS patients. The median total cost per month for medication was 281.78 USD [113.66-870.97] and 175.08 USD [115.09-626.09] for SBS and non-SBS patients respectively. Both in SBS (78.0%) and non-SBS (74.9%) patients, most medication was taken orally, requiring gastrointestinal absorption of the active substance to be pharmacologically active. Most of these medications (77.0% in SBS and 80.8% in non-SBS patients) were formulated as a capsule or tablet, requiring disintegration and dissolution in the gastrointestinal tract before absorption of the active substance can take place. The top three drug classes taken by CIF patients were 1/ proton-pump inhibitors (85.7% of SBS and 67.6% of non-SBS patients), 2/ vitamin D (57.1% of SBS patients) or acetaminophen (46.0% of non-SBS patients) and 3/ anti-motility medication (54.3% of SBS patients) or laxatives/benzodiazepines (each in 37.8% of non-SBS patients). About 25% of the medication taken by SBS and non-SBS patients was medication of BCS class I, characterized by a good permeability and solubility.

**Conclusion** Polypharmacy is highly prevalent in SBSB and non-SBS CIF patients. Most medication is taken orally in formulations requiring disintegration, dissolution and gastrointestinal absorption, which could be compromised in CIF. More research is needed to investigate the clinical effects of different oral formulations in patients with CIF, as oral administration will always be preferred by healthcare professionals and patients.

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## Use of teduglutide during breast-feeding after pregnancy in a patient with severe intestinal failure

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#### Abstract

Background: Successful pregnancy in intestinal failure (IF) is possible but requires careful surveillance to optimize nutrition and to minimize complications. Teduglutide during pregnancy might improve life quality and decrease risk of complications, but experiences are lacking.  Potential risks and benefits for the mother and fetus are thus unknown. Teduglutide induces intestinal hyperplasia, increases intestinal blood flow and delays gastric emptying thereby improving absorption in IF. By decreasing the rate of gastric emptying and the intestinal motility pregnancy itself might thus exert some of the positive effect that teduglutide may achieve. During breast-feeding after delivery the water, electrolyte and nutrition needs are increased but other aspects less complicated. Here we describe a patient who successfully used teduglutide during breastfeeding.

Method: The patient was at the age of 33 operated due to intestinal ischemia caused by strangulation related to congenital malrotation of the small intestine during pregnancy in week 16. The child was lost.  During a period with a high jejunostomy (40 cm jejunum left), ascendostomy and gastrostomy stomal flow and intravenous needs were large. Liver tests became pathological after 5 months. After 10 months anastomosis of jejunum to colon ascendens, stricturoplasty and closure of gastrostomy. Liver tests normalized and PN could be decreased. The patient wanted to become pregnant and was early informed that this was possible.

Results: Pregnant 11 months after the reanastomosis. Normal course of pregnancy during careful surveillance and PN adaptation. Delivered by Cesarian section. One month after termination of breast-feeding, teduglutide treatment was started and had good effect. After one year teduglutide was withdrawn due to desire to become pregnant, which succeeds 1.5 years later. Normal course of the pregnancy. Delivery by Cesarian section. Started teduglutide treatment one month after delivery. Continued breast-feeding for another four months during the treatment. PN gradually decreased to intermittent use, but long-term weight stability without PN has not been achieved. Effects of GLP-2 are highly specific for the intestine. Pig studies show abscence of GLP-2 in fetal blood until the very late phase of pregnancy, and exogenous GLP-2 had no effect on the fetus. In line with earlier findings  our patient exhibited no liver pathology during the pregnancy.

Conclusions: Teduglutide could be used with good effect and without side effects during breast-feeding. Experiences from use during pregnancy are lacking but knowledge about biological effects of GLP-2 biology give no reasons to expect harmful effects on the fetus.

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## Accessibility to teduglutide treatment at a single center in Argentina

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#### Abstract

Introduction: Teduglutide (TED) was approved by the Argentinean government´s regulatory agency (ANMAT) in October 2020 for the treatment of intestinal failure (IF) due to short bowel syndrome. Before its approval, TED was prescribed under compassionate special permit and its coverage was not always granted. Our center started prescribing TED in 2014 for adults and in 2017 for children. In Argentina the health care (HC) is provided by a multi-payer system (public, social security and pre-paid health coverage). Visceral disability (VD) is defined in Argentina as the deficit of an internal organ and the limitations, disadvantages and restrictions that it generates. Chronic IF was included as a VD in 2011 allowing full access to HC coverage including the whole spectrum of chronic IF therapies, from home PN to TED and intestinal transplant.

Aim:  to describe patients’ demographics and accessibility to TED treatment in adult and pediatric patients at a single center.

Methods: retrospective observational study with data extracted form a prospectively filled database comprising all patients treated with TED from 6/2014 to 12/2022. Statistical analysis was done using Mann Whitney test, SPSS 20.Continuous variables are expressed as median and Interquartile range (M/IQR).

Results: 26 patients (10 children) were treated with TED in the studied period, all were on home PN and all but one had VD certificate. Four patients (3 children, 1 adult) temporarily discontinued TED due to lack of provision; 3 adults discontinued TED permanently (1 due to lack of coverage, 1 due to lack of adherence, 1 due to lack of clinical response); 1 child and 6 adults electively discontinued TED more than 6 months after achieving intestinal autonomy, and 4 (2 children, 2 adults) are electively under alternating days regime. Table 1 shows type of HC coverage, time form TED prescription to initiation, if an appeal for protection was used for coverage. When analyzing patients that obtained TED through an appeal for protection, time elapsed from prescription to initiation was longer (p 0.13) (appeal for protection (5 patients) median/IQR 17(6.5-23.5) vs.no an appeal for protection median/IQR 4 (3-10) months).Time form from TED prescription to initiation was median/IQR  10(6.2-22.2) months when covered by social security, and median/IQR   4 (3.0-8.8) months when covered by pre-paid health coverage (p 0.042).Time from TED prescription to initiation was median/IQR 4.5 (4.0-8.7) months in children and median/IQR 6 (3-10) months in adults (p 0.39). Patients that started TED after the ANMAT´s approval, obtained it in a significantly shorter period of time (pre-approval median/IQR 9.0 (4.0-20.7) vs. post approval median/IQR 3.5 (3.0-4.0) months, p 0.019).

Conclusion: TED treatment in Argentina is covered by the health care system although it is costly. Time to approval was variable according to the type of health coverage. ANMAT´s approval allowed shortening the approval time by HC coverage.

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## Teduglutide therapy in children, six years of experience in a specialized Intestinal Rehabilitation Unit

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#### Abstract

Introduction: Management of intestinal failure (IF) secondary to short bowel syndrome aims to achieve enteral autonomy and to prevent parenteral nutrition (PN) related complications. Intestinal rehabilitation is accomplished using a combination of different strategies (medical, surgical, and nutritional). A semisynthetic GLP-2 analogue, teduglutide (TED), is an enterohormone that has been recently added to the IF treatment options for children; experience with its use is still limited.

Aim: to describe the results of using TED treatment in children with Type III-IF (IF) due to short gut syndrome.

Methods: Retrospective descriptive study in patients < 18 years treated with TED 0.05 mg/kg/day from 1/2017 to 1/2022 performed at a single center in Argentina. SPSS v24.0 was used for statistical analysis, continuous variables were expressed as median and interquartile range (M/IQR), and Wilcoxon test was used to compare variables between initial and end points.

Results: 10 patients are described, 5 with neonatal onset of IF (congenital GI anomalies) and 5 with childhood onset of IF (3 volvulus, 1 post-surgical complications and 1 trauma). M /IQR time of IF before TED initiation was 6 (2.5/11.7) years; M/IQR residual intestinal length was 18(8.5/21.5) cm, 3 patients had ICV and intact colon. M/IQR time of TED treatment was 1.7 (0.8/4.1) years.

At last follow up 4 patients had electively discontinued parenteral nutrition (PN) at weeks 24, 29, 48 and 186 of TED therapy, and one discontinued at week 168 due to loss of conventional vascular access; one of them electively discontinued TED at week 260, and other 2 are on TED alternating days. The additional 5 patients continue on PN and daily TED, with a reduction on PN support of 26.5% (expressed as ratio between non protein energy intake/ resting energy expenditure) from baseline: M 0.85 (IQR:0.7/1.23) to 0.71 (IQR: 0.52/0.91) (p=0.059); and number of days of PN infusion/week from 6 (IQR: 5/6.5) to 4.8(4/5.5) (p 0.068). Overall Nutritional data is presented in table 1. Adverse events occurred were: abdominal pain in 8 patients, pain/bruises at injection site in 5, transient diarrhea in 5 mild hyperamylasemia in 4, low serum bicarbonate in 4, mild anemia in 3, hypomagnesemia in 2, low Vitamin D levels in 8, recurrent lower GI bleeding in 1 (due to anastomotic ulcers, diagnosed prior to TED initiation), positive fecal occult blood with negative endoscopy findings in 1, cholesterolosis in 1 and cholecystitis in 1.

Conclusion: the use TED therapy allowed achieving intestinal rehabilitation or reduction on PN support with no impact on the nutritional status, in a group of patients unable to obtain further progress with standard medical rehabilitation. The adverse events were similar to those reported in the adult population.

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## Teduglutide in 19 pediatric SBS patients – single center real world data on clinical efficacy and treatment associated challenges

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#### Abstract

Introduction:  GLP2 analog teduglutide is efficacious in short bowel syndrome (SBS) patients providing reduction in parenteral support (PS) and improving quality of life. In contrast to selective and controlled clinical trials, real world studies include patients with various concomitant diseases and adjusted use of the medication.

Methods: After identifying pediatric patients treated with teduglutide from our institutional SBS database, we reviewed individual patient records for PS details, clinical outcomes, and adverse effects. Parenteral energy provision was expressed as percentage of calculated daily requirement.

Results: There were 19 pediatric (age range: 1.4-17 years) patients who had received teduglutide during 2016-2022, with median 15 (range 2-52) follow-up months on medication, Table 1. SBS etiologies were typical for pediatric onset disease: midgut volvulus (26%), NEC (21%), bowel atresia (21%), gastroschisis (21%) being most frequent. All patients were PS dependent with median 16% small bowel remaining and most (78%) had ileocecal valve removed.

Fourteen (74%) patients reached >20% energy reduction of PS. In four (20%) patients feeding disorder associated with suboptimal PS reduction. Four patients (21%), all who had plateaued on less than 30% PS energy prior to treatment, were successfully weaned off PS. Two of them remained free of PS despite stopping teduglutide. Median reduction in daily PS provision of energy and volume was -36% and -35%, respectively. Median number of weekly infusion-free nights increased from 0.5 to 3. One patient discontinued teduglutide due to insufficient treatment response.

Four patients (21%) experienced clinically significant intestinal bleeding while on teduglutide, one of them having to discontinue treatment due to recurring bleeding episodes and non-specific extensive jejunitis. In three patients we observed signs of tubular nephropathy while on teduglutide. Two patients had tubular loss of electrolytes, which abated during follow-up and did not recur after re-introducing teduglutide. One suffered from extensive nephrocalcinosis after cessation of PS. Stomal prolapses occurred in all patients with end-jejunostomies. One patient died of an unrelated cause. At the end of follow-up, 14 (74%) patients continued to use teduglutide long-term.

Conclusions: In majority of children with SBS, teduglutide resulted in clinically significant PS reduction, increasing infusion-free nights. In minority, the effect on PS requirement remained marginal, possibly due to remaining bowel anatomy and coexisting feeding disorders preventing adequate increase of enteral energy provision. Associated adverse effects included intestinal bleeding from anastomotic ulcers and nonspecific mucosal inflammatory changes, uniform occurrence of jejunostomy prolapses and signs of tubular kidney dysfunction.

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## Are Prophylactic Antibiotics During Broviac Repair in Pediatric Patients with Intestinal Failure Protective Against Blood Stream Infection?

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#### Abstract

**Introduction:**Broviac catheter breakage is a frequent complication in pediatric patients with intestinal failure (IF). Standard of care in pediatric IF patients is Broviac repair to avoid line replacement and preserve vascular access. Evidence supports prophylactic antibiotic administration in the pediatric oncology population, but there is a paucity of data within the pediatric IF population. The primary aim was to determine if antibiotic use, administered at the time of Broviac repair, decreased incidence of bloodstream infection (BSI; defined as positive blood culture within 7 days of intervention). The control population was IF patients undergoing Broviac replacement.

**Methods:** Single-center IRB-approved retrospective review of pediatric IF patients with Broviac catheter breaks that underwent repair or replacement from January 2005-December 2021. Demographic, clinical, and operative data including etiology of IF, catheter size, date of original placement and catheter break(s), blood culture and antibiotic administration were collected. A Pearson Chi-Squared test was run to evaluate for association between infection and antibiotic treatment.

**Results:** Seventy-three patients had 419 interventions (repair n=358, replacement n=61). BSI rate was 2.9% and occurred in 3% of repairs (11/358) and 1.6% of replacements (1/61). There were five infections despite antibiotic administration and seven infections without antibiotics. The association between infection and antibiotic treatment was found to be protective, and use of antibiotics decreased the risk of infection (RR = 0.76, 95% CI: 0.25-2.38), although this was not statistically significant (P=0.64).

**Conclusion:** Our data suggest that prophylactic antibiotic administration is protective against BSI. Further investigation is needed prospectively in the pediatric IF population to standardize practice and elucidate risk factors that may contribute to post-repair BSIs and potentially invoke preventative measures, including use of prophylactic antibiotics.

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## Long term reduction in parenteral support requirements with teduglutide therapy in short bowel syndrome-intestinal failure

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#### Abstract

Introduction: Teduglutide has demonstrated efficacy in clinical trial settings for reducing parenteral support (PS) requirements in adult patients with short bowel syndrome-intestinal failure (SBS-IF). We aim to describe the long-term clinical and nutritional outcomes of patients treated with teduglutide at an Australian quaternary centre.

Methods: Consecutive SBS-IF patients who received at least 12 months teduglutide from November 2018 to January 2023 were analysed. Clinical (urine output, stool and stomal output frequency) and nutritional parameters (weight, dietary intake, indirect calorimetry, handgrip strength) were compared with PS frequency and volume at the commencement of teduglutide and throughout treatment.

Results: Ten patients (70% female, median age 49.6 (IQR: 35.2, 63.5) years) completed between 12 and 48 months of therapy. Median duration of reliance on PS (parenteral nutrition (PN) plus intravenous fluid (IVF)) prior to commencing teduglutide was 8.1 (1.5, 11.4) years. Three patients (30%) achieved complete enteral autonomy within 9 months of teduglutide treatment, with a further 3 patients weaned off PN after 18 months of teduglutide, though required ongoing IVF support. PN and/or PS weaning in these 6 patients was maintained at median follow up 3.25 years. A significant reduction in PS volume (>20%) was observed in 90% of patients, and eight patients (80%) achieved at least 1 night off PS per week. Median PS volume significantly reduced from 10.8L per week at baseline to 5.5L (p<0.01). PN calorie provision reduced by 65%, from 6831±3916 kcal to 2241±3602 kcal per week (p<0.01); whereas daily oral intake increased from 1685 kcal to 2000 kcal/d (p=0.017). Median caloric intake met 128% of patients measured (n=7) or estimated (n=3) energy requirements at last follow up. There was no significant change in urine output volume, whilst stool frequency or stoma bag emptying reduced from 10.2 to 6.3 per day (p=0.03). Patients maintained body weight during teduglutide treatment, and there was a non-significant trend towards increased handgrip strength (median increase 13%, p=0.09).

Conclusion: In our small cohort of patients with SBS-IF destined for a lifetime of PS, treatment with teduglutide improved enteral fluid and nutrient absorption resulting in rapid and significant reductions in PS volume, frequency of PS infusion, and PN calories delivered; with therapeutic efficacy sustained over long term follow up. Importantly, hyperphagia was evident amongst patients and allowed the decrease in PS to be achieved whilst maintaining nutritional status and functional muscle measures.

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## Therapeutic challenges of intestinal ulceration in pediatric intestinal failure: a unique case

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#### Abstract

**Introduction:** Intestinal ulcers contribute to chronic anemia in children with short bowel syndrome (SBS). The potential etiologies, including small intestinal bacterial growth (SIBO), anastomotic ulcers and non-specific intestinal inflammation present therapeutic challenges. Herein we present a case of with multifactorial persistent perianastomotic ulceration in the context of chronic SBS-related SIBO preceding the onset of Crohn’s disease.

**Case:** A boy with SBS type II secondary to necrotizing enterocolitis who had achieved intestinal autonomy at 7 months of age, presented at 3 years of age with chronic bloody diarrhea, iron deficiency, hypoalbuminemia, elevated CRP and poor growth. Endoscopy demonstrated a deep anastomotic ulcer as well as ulcers in the jejunum and colon distal to the anastomosis. Histology was consistent with SIBO. After initial clinical, biochemical, radiologic, and endoscopic improvement with cycling antibiotics for SIBO, persistent and progressive ulceration prompted a change in management. In the ensuing 4 years, treatment trials included prokinetics, prebiotics, exclusive enteral nutrition, bowel rest and oral 5-ASA; repeated prolonged courses of oral budesonide had a predictable biochemical response.  Serial endoscopies revealed a persistent gross appearance and histologic changes compatible with SIBO. Poor growth necessitated re-initiation of parenteral nutrition at 6 years of age.  At 7 years of age, he was diagnosed with Crohn’s Disease (CD) of the jejunum and rectum; with the most severe disease being perianastomotic. Treatment with vedolizumab resulted in clinical and biochemical remission, including a normalization of fecal calprotectin (117.5 mg/Kg). PN dependence decreased from 82 kcal/Kg/day (135% dependence) to 61 kcal/kg/day (122% dependence) in the first year on vedolizumab and further decreased to 42 kcal/kg/day (104% dependence) at 3 years. Over that time, height for age z-score improved from -2.1 to -0.92. While annual endoscopy for 3 years revealed resolution of jejunal and rectal ulcerations, deep, serpiginous circumferential anastomotic ulcers persisted despite dose optimization and the addition of budesonide.

**Conclusion:** This case of Crohn’s disease in the context of SBS-related altered anatomy and chronic SIBO demonstrated a significant clinical, nutritional, and biochemical response, and relative endoscopic response to vedolizumab. However, there was treatment failure at the anastomotic site where vascular changes may make healing challenging. This case highlights the varied underlying etiologies and therapeutic challenges of intestinal ulcers in children with short bowel syndrome. Multicenter collaboration is needed to improve our understanding and develop an evaluation and management algorithm for intestinal ulcers in pediatric intestinal failure.

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## REPORT OF 17 YEARS OF EXPERIENCE IN MANAGEMENT OF ADULT AND PEDIATRIC INTESTINAL FAILURE PATIENTS IN A SINGLE REFERRAL CENTER IN ARGENTINA

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#### Abstract

INTRODUCTION: Parenteral nutrition (PN) is the first line treatment for intestinal failure (IF). In the last decades the development of intestinal rehabilitation strategies included both medical-nutritional treatment (drugs, feeding strategies, use of enterohormones [EH]) and surgical procedures (autologous gastrointestinal reconstruction surgery [AGIRS], lengthening procedures, transplantation [ITx]) with the aim of re-establishing intestinal autonomy. The aim of this study is to report the results of 17 years of experience in a specialized single center in Argentina.

METHODS: From February 2006 to February 2023 a retrospective analysis of a prospective database of patients (pts) with IF was performed. Pts were divided into 2 groups (A adults and P pediatrics) reporting the main causes of IF. From those pts with short bowel syndrome (SBS) number of surgeries performed, EH treatment, PN independence and overall survival were assessed. ITx indications and outcomes were also reported.

RESULTS: 530

pts with IF were included (Group A=400, P=130 ). In group A, IF was type I in 48 (12%), type II in 64 (16%) and type III in 288 (72%). SBS was the leading cause in 199 pts (49.8%) followed by intestinal fistula in 66 (16.5%), intestinal dysmotility in 65 (16.2%), mechanical obstruction in 46 (11.5%) and extensive small bowel mucosal disease in 24 (6%).  From the SBS group, 128 pts (56.6%) received AGIRS. Mean post-surgical intestinal length was 144.3 ± 101.1cm;  anatomy type was 1 in 13 (10.2%), 2 in 41 (32.0%) and 3 in 74 (57.8%.) From those pts, 74 (57.8%) were weaned off PN. EH were used in 16 out of 128 pts (12.5%) and 12/16 (75%) achieved intestinal rehabilitation without severe adverse events, with a mean intestinal length of 47.5 ± 43.7cm. The overall survival at 10 years was 77%. Nineteen  out of 288 pts (5.9%) received 20 ITx, and 89% achieved PN independence. ITx survival at 10-year was 41%. In group P IF was type III in all the cases and the leading causes were SBS in 112 (86.1%), intestinal dysmotility in 11 (8.5%) and extensive small bowel mucosal disease in 7 (5.4%). In the SBS group 17 pts received AGIRS (15.2%) and 9 lengthening procedures (8.0%). Mean intestinal length post initial resection was 28.7 ± 27cm. Twenty three out of 112 (20.5%) SBS pts were able to wean off PN and ten (8.9%) required EH treatment, achieving intestinal independence in 5 (50%) The overall survival was 69% at 10 years. Twenty-nine pts received 33 ITx, 69% achieved PN independence at 1 year, and 10 years survival was 44 %.

CONCLUSION: After 17 years of experience 530 pts with IF were treated in our program. The best results were obtained by a multidisciplinary approach, combining surgical, medical, and nutritional strategies. The introduction of EH improved the outcome of SBS pts, allowing to achieve intestinal sufficiency even in those with unfavourable intestinal length and anatomy types. ITx was required in only 10% of the cases.

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## Long lasting effect of Intramuscular (IM) iodine injection in the treatment of goiter in an intestinal failure patient with complete enterectomy and colectomy.

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#### Abstract

**Introduction:** Individuals with intestinal failure are at risk for micronutrient deficiency, especially iodine which is an essential trace element critical for the production of thyroid hormone. Iodine is not available in commercial trace element preparations in the United States. In those with complete parenteral nutrition (PN) dependence, options to replenish and maintain iodine stores are very limited as oral forms are poorly absorbed and IV alternatives are not available. Ethiodized oil (Lipiodol TM) is an iodinated contrast agent injected into a target organ with an unusually long half-life.

We report the successful use of intramuscular (IM) injection of ethiodized oil in the treatment of goiter in a patient with total entero-colectomy.

**Methods:** We reviewed the medical record of a single case identified during routine clinical care.

**Results:** A 20-year-old female with short-bowel syndrome secondary to traumatic intestinal evisceration underwent isolated small bowel transplant at 14 years of age. She went on to have a complicated post-transplant course which included post-transplant lymphoproliferative disorder and rejection resulting in complete enterectomy and colectomy. She was unable to be placed in continuity leaving her with 100% TPN dependence and essentially no functional proximal small bowel.

She developed hypothyroidism and goiter in 2015 with labs remarkable for elevated TSH > 150 (0.7-5.7 IU/ml), low free T4 of < 0.2 (0.89-1.76 ng/dL) and nonexistent total T3 <1 (86-192 ng/ml). Her initial ultrasound in 2015 showed thyromegaly, heterogenous echotexture with lobulated contour, and hyperemia with small cysts. She was placed on IV levothyroxine with normalization in TSH and free T4 after a month but persistent low total T3 <2 and iodine deficiency, with serum iodine levels ranging between 35-50 mcg/L (52-109 mcg/L). Her remaining anatomy impaired appropriate oral iodized salt or oil absorption as iodine is mainly absorbed in the small bowel. After obtaining innovative consent, she received IM Lipiodol TM in 2015 and levothyroxine was discontinued. Her iodine levels were monitored  and normalized 6 months post-injection followed by goiter resolution. She had repeat labs and ultrasound in 2020 which showed normal thyroid levels (TSH: 1.52, free T4: 1.39, total T3: 189) and improved thyroid inflammation, decreased gland size and absent cysts/nodules.

**Conclusion:** Ethiodized oil is a medication designed as a contrast agent that contains iodine combined with ethyl esters. Its long half-life when injected intramuscularly may be beneficial in patients with complete intestinal failure suffering from iodine deficiency where oral and IV forms are not feasible alternatives. Iodized IM injection enabled our patient to be weaned off intravenous thyroid hormone, thereby simplifying her care, treating her goiter and maintaining long lasting thyroid function for 5 years or more.

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## Improved outcome in the past fifteen years of a pediatric intestinal failure Latin American cohort

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#### Abstract

Introduction: Multidisciplinary (MD) Intestinal Rehabilitation Programs are crucial to obtain the best results in children with intestinal failure (IF). International multicenter reports describe better results on mortality, rehabilitation, and less need for transplantation in the last decade. Our aim: to analyze the outcomes of children with IF managed by a MD team at a reference center in Latin America.

Methods: Retrospective review of medical records from January 2008 to December 2022 of pediatric patients with IF managed by an intestinal rehabilitation MD team. Demographic data were collected, together with rehabilitation, mortality and transplantation rates. They were analyzed in the global population and also in two equal time periods, according to initiation of follow-up either before or after June 2015, the middle of the studied period. Early referral (before 6 months of diagnosis) and bowel lengthening procedures were recorded. The descriptive data was analyzed by chi2 test or Fisher's test for categorical variables and by t-test or Mann Whitney for continuous variables, depending on the distribution.

Results: 156 patients with IF were treated during the study period. Median age at first consultation was 0.43 yrs. (IQR 0.1-1.75), 95 males.The IF etiology was short bowel syndrome (SBS) in 127/156 (81.4%), motility disorder in 24 (15.4%) and congenital enteropathy in 5 patients (3.2%). Thirteen patients had type 1 SBS, 91 type 2, and 23 type 3. 86/156 were able to stop parenteral nutrition, with a rehabilitation rate of 55%, 2/91 received teduglutide. Enteral autonomy was achieved in a mean time of 1.03 years (SD ±1.25). Fourteen intestinal transplants were performed in 13 patients, of which 8 are still alive, 4 with functioning grafts. Cumulative mortality during these 15 years was 16% (25/156).

The analysis between the two periods showed better trends in rehabilitation, with a statistically significant difference observed with lower mortality rates and less transplantations performed in patients from the most recent period.

There was also a tendency towards better rehabilitation rate, shorter time to reach it, and higher proportion of patients with early referral,althought the difference was no statitically significant  (Table 1).

Conclusion: Although the possibility of reaching intestinal rehabilitation continues to increase, the most important impact observed in this pediatric cohort with IF, was reduced mortality and less transplantation in the recent times supporting international reports.

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## Healthcare utilization during the initial hospital stay for children with short bowel syndrome with intestinal failure

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#### Abstract

Introduction: Recent efforts of the Intestinal Rehabilitation and Transplant Association led to the formation of the International Intestinal Failure Registry (IIFR). Pilot analysis of IIFR patients provided contemporary data on outcomes within the first year but lacked details regarding the initial hospital stay. We analyzed health care utilization costs during the initial hospital stay for children <1y with short bowel syndrome and intestinal failure.

Methods: We identified a cohort in the Pediatric Health Information System database with a diagnosis code of post-surgical malabsorption, neonatal intensive care unit stay, and parenteral nutrition use for 60 days discharged between 2004 and 2020. Demographics, diagnosis and procedures codes, utilization, and child opportunity index (COI) were collected; COI combines 29 indicators to represent neighborhood opportunity categorized into 5 levels from “Very Low” to “Very High.” Descriptive statistics characterized all variables. Our primary outcome was total standardized costs adjusted to 2021 US dollars. Logistic regression to identified characteristics associated with in-hospital mortality and generalized linear regression determined factors associated with increased costs.

Results: We included 3082 children with short bowel syndrome with intestinal failure (42% female, 30% non-Hispanic White, 52% NEC). Among those with a reported gestational age (70%), the median gestational age at birth was 31 weeks [IQR 26 – 35 weeks]. Median birthweight was 1595 g [IQR 860 – 2430]. Venous thromboembolism (VTE) was found in 13% and central line-associated bloodstream infection (CLABSI) was found in 13%. In-hospital mortality was 7.0%. In multivariate analysis, presence of VTE, earlier discharge year, and non-White race but not CLABSI were associated with increased odds of death during the admission. Median length of stay was 149 days [IQR 112 – 199 days] and median per patient total standardized cost was $26.1 million [IQR 16.5 – 39.2]. In multivariate analysis adjusted for length of stay, increased cost was associated with non-Hispanic Black race vs non-Hispanic White race, necrotizing enterocolitis diagnosis, VTE, and “Very High” COI but not CLABSI.

Conclusions: Children with short bowel syndrome with intestinal failure have long initial hospitalizations, elevated mortality, and very high utilization costs. Many children experience CLABSI and VTE during the initial hospital stay, and VTE is associated with increased mortality and increased total costs. Future work will examine the longer-term outcomes of this cohort.

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## Prevalence of Eosinophilic Gastrointestinal Diseases in Children with Short Bowel Syndrome (SBS) at Children’s National Hospital (CNH)

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#### Abstract

Introduction: Patients with SBS have multiple risk factors for eosinophilic gastrointestinal diseases (EGIDs) including increased risk for dysbiosis and food allergy compared to their counterparts with normal anatomy. However, there is limited data on the prevalence of EGIDs in children with SBS. The aim of this study was to define the prevalence of EGIDs in a SBS cohort and its association with different risk factors.

Methods: A retrospective chart review of patients with SBS at CNH from 2009-2020 was performed. Patients were excluded from the cohort if they did not have an endoscopy during this timeframe. 83 SBS patients met inclusion criteria for analysis. Pathology reports with incomplete information on the number of eosinophils per high power field (HPF) were excluded. Some patients had multiple procedures during the study timeframe. For statistical analysis, only the most recent biopsy specimen for each patient was retained.

Results: The prevalence of eosinophilic esophagitis (EOE), ≥15 eosinophils per HPF, in our SBS cohort was 12.7%. Of the patients that had any eosinophilia on esophageal biopsy, 50% had EOE. Only one patient with biopsy-defined EOE had small intestinal bacterial overgrowth (SIBO) at time of the procedure. SBS patients with history of allergy were more likely to have esophageal eosinophilia on biopsy than patients without allergy (p=0.01). The prevalence of eosinophilic gastritis (EG), ≥30 eosinophils per HPF, in our SBS cohort was 5.6%. Of these patients, one with biopsy-defined EG also had biopsy-defined EOE. There were significant differences in the number of gastric eosinophils on biopsy between SBS patients on full enteral nutrition, mixed enteral and parenteral nutrition (PN), and exclusive PN (p=.01). The prevalence of eosinophilic enteritis (EE), >50 eosinophils per HPF, was 5.89%. One patient with biopsy-defined EE also had biopsy-defined EOE, and two patients also were noted to have SIBO at time of biopsy. SBS patients with history of allergy were more likely to have intestinal eosinophilia on biopsy than patients without allergy (p=0.03). Four patients had eosinophils on colonic biopsies and only one met the diagnostic threshold for eosinophilic colitis (EC), >80 eosinophils per HPF. Seven patients with any biopsy-defined EGID had SIBO at time of biopsy; however, SIBO was not associated with a significant difference in esophageal (p=0.78), gastric (p=0.95), intestinal (p=0.14), or colonic eosinophilia (p=0.41). In addition, length of bowel was not correlated with degree of eosinophilia. Patients on proton pump inhibitor and budesonide therapy did not have significantly less eosinophils on biopsy than those not on therapy.

Conclusion: The prevalence of EGIDs in a  pediatric SBS cohort is significantly higher than in the general population and may be linked to allergy sensitization. Further research is necessary to define the pathophysiology behind the evolution of EGIDs in this special population.

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## An innovative educational program for adolescents on home parenteral nutrition: “the connected ados”

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#### Abstract

Background and aims

Facing with an increasing demand for transition to adult care management and guidelines, our Home Parenteral Nutrition (HPN) team designed a therapeutic group educational program specifically intended for adolescents on long-term HPN. The aim of this study was to report on the first sessions of this program.

Methods

The Adolescent Therapeutic Educational Program (ATEP) was designed in 3 sessions of 5 consecutive days, during school holidays over the year. The ATEP included group sessions on catheter handling, disconnecting and connecting the PN and catheter dressing, dealing with unforeseen events such as fever or catheter injury, but also sessions with psychologist, social worker, sports teacher, fashion specialist, meeting with adults who received HPN since childhood. Specific course for the accompanying parents were also provided. Six months after the last session, a 3-day trip to the Futuroscope, Poitiers, France, was organized without any parental presence.

Results

Along the three training courses, a total of 16 adolescents were enrolled in the ATEP. They were aged between 13 and 17 years (median 14 years IQR: 14-16.25). All but five were on long term HPN starting during the neonatal period, the five others started PN at a median age of 10 year old (IQR: 1-10). At the time of the ATEP, their median PNDI was 105% (IQR: 95.5-120.8) while receiving a median number of 6 infusions per week (IQR: 5-7). Thirteen out of 16 received  Taurolidine lock procedure. At the end of the three sessions, 11 adolescents could be considered as fully autonomous, 4 as partially autonomous and one failed to gain any autonomy. Course evaluation by adolescents or parents was good to excellent.

Conclusion

Through the holistic and multi-professional approach of this training and the group cohesion, the adolescents were not only able to handle catheter care and PN connections but were able to understand and accept better their illness and project themselves into their own future.

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## RESTORE “amendment”: from Argentina to Latin America, new steps of the First prospective Intestinal Failure Registry due to short bowel syndrome.

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#### Abstract

Background: Type III Intestinal Failure (IF) due to short bowel syndrome remains a rare condition.  Few registries worldwide are able to collect prospective information on diagnoses, management, complications and outcomes in the subject.

Aim: to report the progression, updated results and future steps of the RESTORE amendment registry.

Methods: A prospective multicenter observational registry was created in 2017 for adult patients with type III-IF due to SBS in Argentina (RESTORE). In May 2020, it was amended in order to include pediatric patients and to expand to Latin America in 2 steps (first including centers from Chile, Colombia, México, Peru and Uruguay; the second step will include the rest of the countries in the region). Demographics, clinical data, nutritional assessment, home parenteral nutrition (HPN) provision and management, medical and surgical rehabilitation techniques, related complications, overall survival, and freedom from PN survival were analyzed, from 6/2017 to 12/2022, accomplishing the first step. Data were collected in RedCap; statistics were run on SPSS v24.0.

Results:  Of the initially 37 identified centers, 25 obtained approval and enrolled patients; monitored data on 108 patients are analyzed. Table 1 shows data at enrollment, patients are grouped according to age.

Etiologies of the IF were:  surgical complications 35%, intestinal ischemia 24%, trauma 9%, mechanical obstruction 8%,  volvulus 5%, Crohn’s 4% and others 15%, for adults;  atresia 34%, gastroschisis 21%, NEC 21%, volvulus 12%, and others 12%,   for children.  At enrolment all patients were on PN and 83 (77%) were receiving HPN; at the end of the analysis 96 (89%) were on HPN. IF/PN related complications are shown in table 2

The mean time of follow up was 33.7±20.5 months for adults and 8.1±5.6months for children. During the whole period, 32 surgeries were performed on 28 patients (adults: 27 AGIRS, children: 2 STEP and 3 AGIRS). Sixteen patients received enterohormones (1 GLP-1, 15 GLP-2 [4 children]), no severe adverse events were observed. By the end of the period, 19 of 108 patients (17.5%) (all adults) achieved enteral autonomy; 4 of them were on GLP2.  In the remaining 89 patients, weekly PN volume was reduced by 24% and PN calories by 39 % in adults, and 17% and 28 % respectively in children. One adult received a multivisceral transplant. Five patients were lost to follow up. Twelve patients died (10 adults), causes were: sepsis 6, diabetic ketoacidosis 1, post-surgical complications 1, while was not registered in 4. . Overall actuarial survival was 82% (83% for adults and 91% for children).

Conclusion: The RESTORE “amendment” highlights the value of having a registry; it provides unprecedented and novel data on epidemiology, main causes, current medical and surgical approach, as well as outcome and complications related to IF/PN at dedicated centers in the region, providing a system to standardize care.

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## Venting  Percutaneous Endoscopic Jejunostomy as Adjuvant Treatment for Rehabilitation of an Infant with Short Bowel Syndrome secondary to Gastroschisis.

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#### Abstract

**Introduction:** Complex gastroschisis is one of the most frequent causes of short bowel syndrome (SBS) in pediatrics. Motility disorders, intestinal dilatation and hypoplasia are factors that difficult progression and intestinal rehabilitation. We present the case of an infant whom a venting  jejunostomy aided in treatment.

**Methods:** 5-month-old male infant, with antenatal diagnosis of gastroschisis, born at 34 weeks, with finding of associated  intestinal atresia, ischemia and stenosis in the ring and extra abdominal intestine at birth, also absence of colon from cecum to descending portion, with remnant hypoplastic descending and sigmoid colon. Initial surgery was an  intestinal resection leaving 20 cm of remaining dilated small intestine and a side-to-end  jejuno-colonic anastomosis + jejunostomy. At 45 days of life, patient required remodeling   of stoma due to jejunostomy prolapse.   Prolapse recurred few days after the procedure. Since birth patient was managed by the pediatric intestinal failure team with parenteral nutrition and controlled and slow progress of enteral nutrition, due to high output and mucous prolapse  of the stoma. At 80 days of life, the jejunostomy was surgical closed, leaving the intestine in continuity. Postoperative evolution was stationary , with poor oral tolerance, abdominal distension, biliary emesis  and minimal progress despite having daily bowel movements. Contrast images showed dilation of the small intestine with passage of the contrast through the colon and patent anastomosis, without signs of mechanical obstruction. The group decided to place a percutaneous endoscopic jejunostomy tube with a 14 Fr endoscopic gastrostomy kit through an anterograde approach for ventilation, proximal to the jejuno-colonic anastomosis. Endoscopy showed the entire remnant intestine dilated with normal mucosa and a patent wide  jejuno-colon anastomosis. There were no complications (Image 1).

**Results:**After the procedure, the patient was managed with intermittent opening of the tube for decompression, with  a good outcome, tolerating  oral route with normal stools and without abdominal distension or  vomiting (Image 2).

**Conclusions:**Patients with intestinal failure and SBS due to gastroschisis  born with intestinal dilatation and motility disorders represent a therapeutic challenge since lengthening/remodeling surgeries are questionable. The placement of a percutaneous venting jejunostomy is a less invasive procedure and  may be helpful  to promote enteral progression and rehabilitation of these patients, without completely excluding  the distal intestine and without complications such as prolapse that surgical jejunostomies present.

**Image 1.**

**Image 2.**

### 85

## Thirteen cases of enteric dysmotility

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#### Abstract

Objective: Case series study was conducted to investigate the clinical characters of 13 cases of enteric dysmotility (ED). Methods: The medical records of all ED patients who were diagnosed with small intestinal manometry from Aug. 2019 to Jul. 2022 were retrospectively reviewed. The clinical parameters, small intestinal manometry presentations, treatment, and outcome were analyzed. Results: 13 patients were identified. In each case, small intestinal manometry presented 1-3 abnormalities regularly showed in ED. All 13 cases (100%) presented a history of constipation. Among them, five (38.5%) began with idiopathic constipation and had a subtotal colectomy history. Six (46.2%) patients began with upper gastrointestinal disorders but with a long history of constipation. Three (23.1%) patients showed severe chronic abdominal pain with 1 of them opiate dependence. Eight patients were surgically treated and all of them obtained a significant nutritional increase and symptom relief. Conclusion: ED is a concisely defined disease with clear diagnostic criteria. This study indicated that surgery can increase the outcome of some patients.

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## ENDOSCOPIC AND SURGICAL APPROACH IN PATIENT WITH INTESTINAL FAILURE WITH LIFE-THREATENING GASTROINTESTINAL BLEEDING

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#### Abstract

METHODS

Case report

RESULTS

A 16-month-old female patient is reported, with a history of gastroschisis, presence of short bowel syndrome without ileocecal valve, resolved liver failure. Initially with ileostomy with 35 cm of small intestine; Seeking to improve progression of the enteral route and intestinal adaptation, ileostomy closure and subsequent colostomy were performed since there was suspicion of stenosis in the distal colon. It begins with events of sudden anemia, drop of 2 g of hemoglobin per day, requirement of transfusion support, associated with evidence of hematochezia by colostomy, the first endoscopy is performed by colostomy documenting small ulcers with evidence of eosinophilic colitis, she began management with prednisolone, elemental diet, without improvement. A 2nd time endoscopy was performed, perianatomotic ulcers were documented, with no evidence of active or recent bleeding. Complete evaluation of the intestine endoscopically was not possible. Medical management was started with mesalazine, sucralfate, and steroids were continued, with persistence of symptoms. Ischemic colitis is suspected, so the group decided to carry out a new surgical and endoscopic procedure simultaneously, where a segment of the small intestine with erythematous mucosa and an ischemic appearance is documented in the endoscopic assessment that is adjacent to the area of the small intestine anastomosis, which was resected, associated with an erythematous area and ulcer in the endoscopic assessment adjacent to one of the mechanical sutures of the previous intestinal lengthening, which was also resected. Surgery performs end-to-end anastomosis, resection of the proximal end of the narrow sigmoid colon, end-to-end colo-colonic anastomosis. Pathology reports the absence of ischemic changes in resected segments, with the presence of severe eosinophilia (unquantifiable). Patient since the last procedure without new events of gastrointestinal bleeding.

CONCLUSION

Multidisciplinary work in patients with intestinal failure in the process of intestinal adaptation is essential. The performance of a hybrid endoscopic and surgical procedure, by pediatric gastroenterology and pediatric surgery, allowed the adequate identification of the affected segments, so that they were resected more precisely, achieving control of the symptoms in the patient, as well as the progression of the enteral nutrition.

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## The role of the colon in paediatric short bowel syndrome

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#### Abstract

Introduction

Colon has a fundamental role in Short Bowel Syndrome (SBS) because it promotes intestinal adaptation. The importance of the colon is connected to its ability to absorb water and nutrients and as a host to an integral part of the intestinal microbiota. After intestinal resection, the colon becomes an energy-absorptive organ and improves the chances of weaning patients from parenteral nutrition (PN), even in cases of extensive small bowel resections. Finally, the colon, especially the right colon, produces glucagon-like peptide 2 (GLP-2), which inhibits gastric secretion and affects small bowel permeability.

Methods

Data were collected retrospectively from our Intestinal Failure Unit. Collected data were patients’ diagnosis, small bowel length, autologous gastrointestinal surgery, percentage of the remaining colon and volume of parenteral nutrition per week at follow-up. Descriptive and inferential statistical analysis was performed: the Mann-Whitney test was used to assess statistical significance.

Results

We found complete data on a total of 31 paediatric SBS patients; 14 had either the entire colon or the right colon (Group A), while 17 only had the left colon or no colon at all (Group B). The leading causes of SBS in group A were volvulus and intestinal atresia, and in group B, volvulus and NEC. Median small bowel length in group A was 36 cm, and in group B, it was 48 cm. In group A, 6/14 patients had less than 25 cm of small bowel, and in group B, only 3/17 patients had less than 25 cm of small bowel. In group A, half of the patients (7/14) are still on PN at follow-up, while in group B, 59% of patients (10/17) are still on PN. Median PN volumes per week for group A is 5082 ml, and for the group B is 4597. Differences in PN volume among the two groups and subgroups were not statistically significant.

Conclusion

The presence of the colon, in particular the right colon, can be a great asset to improve SBS patients’ condition and their chances to wean from PN thanks to the absorptive functions of the colon and the production of GLP-2. Limitations to this study are the small dimension of the group and the heterogeneity between the two groups. Small bowel length should also be considered a confounding factor since most patients in Group B have more than 25 cm of small bowel. Further studies are needed to better analyse the colon's role in weaning from PN in pediatric SBS patients.

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## Clinical utility of Gastric Alimetry® in the management of intestinal failure patients with possible underlying gut dysmotility

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#### Abstract

**Introduction**

Gut dysmotility is an increasingly common contributor to intestinal failure. However, differentiating true cases of severe dysmotility requiring intensive nutritional support from other causes of symptoms (e.g. gut-brain or visceral hypersensitivity disorders) is clinically challenging. Reliable motility tests are not widely accessible and may be non-contributory, while transit studies can be labile and insensitive for neuromuscular pathologies. Gastric Alimetry® (Alimetry, New Zealand) is a new test of gastric function that was recently shown to define patient subgroups with true neuromuscular dysfunction in chronic nausea and vomiting syndromes.1 This study reports the first application and outcomes of Gastric Alimetry in the management of patients with intestinal failure.

**Methods**

A cohort of 10 adult intestinal failure patients with a presumed diagnosis of gastroparesis were re-evaluated with Gastric Alimetry. Gastric emptying tests were abnormal in all cases (delayed in 8/10; indeterminate in 2/10 due to vomiting). 9/10 were on parenteral nutrition, (1/10 PN recently with-held due to line infection). A standard test protocol of 30 mins fasting, test meal and 4 hr post-prandial recording was performed, with simultaneous symptom logging on a validated App. Gastric Alimetry Rhythm Index (GA-RI), BMI-adjusted amplitude and Principal Gastric Frequency were compared to established reference intervals.2 Patients were divided into those with neuromuscular disorders vs normal gastric function based on Gastric Alimetry results,1 to inform targeted therapy and integrated clinical care.

**Results**

Gastric Alimetry tests shown neuromuscular dysfunction in 3/10 patients (low GA-RI and/or amplitude); normal in 7/10. These results led to changed clinical diagnoses in 6/10 cases, primarily to refocus on disorders of gut-brain interaction in subjects with normal tests. Changed management based on these results facilitated successful weaning of PN in 6/9 patients at median 5 months of follow-up, due to intensive targeted pharmacological therapy and integrated care (including neuromodulators / health psychology); with estimated cost savings of >$100,000 per patient per year.

**Conclusions**

Gastric Alimetry provided clinical utility in the work-up of intestinal failure patients with suspected motility disorders, changing diagnosis and guiding less-invasive management in >50% of patients. Test results facilitated gut rehabilitation, reduced PN dependence, and reduce healthcare costs.

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## BARIATRIC SURGERY COMPLICATIONS: MANAGEMENT IN TWO INTESTINAL FAILURE REFFERRAL CENTERS IN ARGENTINA

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#### Abstract

**Introduction:** Obesity is considered a modern pandemic. Several medical strategies have been developed in the last decades. However, the most successful treatment to lose weight is bariatric surgery (BS). This procedure is not free of medical and surgical complications requiring long term medical treatment and/or many re-interventions to be solved, with increased morbidity and mortality. Some patients could develop intestinal failure (IF) consequently. The aim of this study is to report the surgical and medical complications of BS patients referred to two specialized centers in the management of IF in Argentina.

**Methods:**  A retrospective analysis of a prospective database of 520 adult IF patients was performed from February 2006 to January 2023. Sex, age, type of BS, medical and surgical complications and PN dependency were assessed.

**Results:** Forty two out of 520 patients (8.1%) were referred with complications of BS. Twenty-nine were females (69.0%) and mean age was 47.12 ± 11.57 years. Surgical procedures were gastric bypass (GYBP) in 34 (81.0%) , gastric sleeve in 7 (16.6%) and gastric banding in 1 (2.4%) Thirty-six (85.7%) out of 42 developed IF and 6 (14.3%) intestinal insufficiency. Twenty four out of 36 patients (66.6%) developed chronic IF (CIF) and 12/36 (33.3%) acute IF (AIF) (Table 1) Five out of 24 CIF patients developed medical complications as chronic diarrhea and 19 developed surgical complications (9 hernia, 4 fistula, 2 anastomosis stenosis, 1 ischemia, 1 anastomosis leakage, 1 anastomosis necrosis and 1 abdominal obstruction) Complications appeared in 1 patient in the immediate postoperative period (POP) (within 30 days), in 7 patients in the mediate POP (between 30 days and 6 months) and in 15 patients were on the long term (beyond 6 months). Eight patients underwent autologous gastrointestinal reconstruction surgery and the average number of surgeries required to manage complications was 2.17 (0-3). Fifteen patients achieved PN independence, 8 continued on PN and 1 was lost in follow up.

**Conclusions:** IF can be the result of medical and surgical complications of BS. In this cohort of patients surgical complications were the most frequent cause of CIF. Successful outcomes require a comprehensive and a multidisciplinary treatment in specialized centers in the management of such patients.

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## PREVALENCE OF SARCOPENIA IN A COHORT OF PATIENTS WITH CHRONIC INTESTINAL FAILURE DUE TO SHORT BOWEL SYNDROME.

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#### Abstract

**Introduction:** Sarcopenia is defined as a progressive and generalized loss of muscle mass and function and its presence leads to worse outcomes. Patients with chronic intestinal failure due to short bowel syndrome (CIF-SBS) could develop sarcopenia at diagnosis due to malabsorption, inflammatory states and/or physical inactivity. The aims of our study were to determine the prevalence of sarcopenia in a cohort of adult patients with CIF-SBS and its association with demographic and nutritional parameters.

**Methods:** Adult patients with CIF-SBS were included in the analyses. SARC-F questionnaire was performed. Age, sex, anatomy type, time on parenteral nutrition (PN), current weight, height, Body Mass Index (BMI) and hand grip strength were assessed. In addition, a CT scan was done. Bivariate analysis was carried out to study variable associations. Chi-squared was used for categorical variables. Statistical analyses were performed in SPSS v24.0; significance was considered when p<0.05.

**Results:** From June 2019 to December 2022, 42 adult patients with diagnosis of CIF-SBS were included in the study. Mean age was 49.3 ± 16.1  years and 54.8% were female. Anatomy type was 1 in 92.8% of the patients, followed by anatomy type 3 in 7.2%. Mean BMI was 22.8 ± 4.3 kg/m2 and 52.4% had less than 6 months on PN. All patients presented risk of sarcopenia assessed by SARC-F questionnaire. Hand grip strength resulted in deficit in 57.1%, with no association between time on PN (p: 0.129), anatomy type (0.387) or low BMI (p: 0.158). However, low handgrip strength associated with male sex (p: 0.049). Sarcopenia was diagnosed in 69.0% of the patients using CT scan. There was no association between presence of sarcopenia and sex (p: 0.453), less than 6 months of time on PN (p: 0.588), anatomy type (0.926) and low handgrip strength (p: 0.335). There was an association between low BMI and presence of sarcopenia (p: 0.023) however, 60.6% of the patients that had sarcopenia had normal BMI.

**Conclusion:** Sarcopenia was diagnosed in most of the patients with CIF-SBS. In this cohort of patients low BMI could conduct to its suspicion, however, it should be assessed in all patients as 60.6% of the patients with normal BMI had sarcopenia diagnosis.

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## A Comparison of First-line Intravenous Lipid Emulsions SMOFlipid Versus Intralipid on Incidence and Time to Resolution of Cholestasis in Hospitalized Pediatric Patients

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#### Abstract

Introduction: Lipid emulsions are an important component of pediatric parenteral nutrition (PN). PN-associated cholestasis (PNAC), defined as conjugated bilirubin >2 mg/dL or conjugated bilirubin >20% of the total bilirubin if the total bilirubin is >5 mg/dL, is a major complication of long-term PN use. Intralipid® contains 100% soybean oil and is associated with PNAC due to multiple factors including a high content of omega-6 polyunsaturated fatty acids and phytosterols and relatively low alpha-tocopherol levels; SMOFlipid® contains fish oil which has reduced cholestatic risk factors. The study aim was to determine whether first-line use of SMOFlipid for all hospitalized pediatric patients would reduce the rate of PNAC or have a later onset or faster resolution of PNAC when compared with Intralipid.

Methods: This double-center, retrospective cohort study included all hospitalized pediatric patients who received ≥14 days of lipid emulsion therapy (Intralipid or SMOFlipid). Primary outcome measures were incidence of PNAC, and time to develop PNAC and time to resolve it. Secondary outcomes included lipid therapy effect on weight and laboratory parameters.

Results: Included in this study were 194 hospitalized pediatric patients; 93 received Intralipid and 101 received SMOFlipid. The incidence rate of developing PNAC in SMOFlipid patients was 10% (10/101), which was comparable to 13% (12/93) in Intralipid patients (P=0.651). The time to develop PNAC after lipid therapy initiation was 19.0±13.0 days in the Intralipid group, and 39.7±24.7 in the SMOFlipid group (P=0.02); 83% (10/12) of Intralipid patients developed PNAC at ≤21 days compared with 20% (2/10) of SMOFlipid patients (P=0.008). The time of PNAC resolution was 21.8±21.3 days in SMOFlipid patients, which was not significantly different compared with 33.0±17.5 days in Intralipid patients (P=0.268).

Conclusion: Our study shows that SMOFlipid is associated with a delay in onset of cholestasis in hospitalized pediatric patients receiving SMOFlipid as a first-line intravenous lipid emulsion therapy compared with Intralipid therapy. However, SMOFlipid does not show a statistically significant reduction in the incidence of cholestasis. Based on our study findings, we recommend that neonates expected to be on PN for at least 21 days should be initiated on SMOFlipid therapy to delay PNAC. More studies, ideally randomized control trials, are needed to better evaluate the benefits of using SMOFlipid as a first-line intravenous lipid emulsion therapy for all hospitalized pediatric patients.

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## Iodine and selenium status and hypothyroidism in children with intestinal failure

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#### Abstract

**Background:**  Iodine is essential for thyroid hormone biosynthesis. We investigated the prevalence of iodine deficiency (ID) and hypothyroidism in children with intestinal failure (IF) followed up longitudinally, considering selenium status, which is also essential for thyroid gland function.

**Methods:** Children admitted to an intestinal rehabilitation program and receiving home PN were regularly followed-up for urine iodine concentration (UIC), selenium, and thyroid function tests from April 2019 to June 2022. The outcome variable ID was defined as a UIC value < 100 μg/L. Generalized estimating equations were used to assess the effects of potential variables associated with UIC.  The study was approved by the hospital's ethics committee.

**Results:**Twenty-four patients aged 62.7 (39.1; 79.7) months receiving PN for 46.5 (21.5) months were included. The average energy supply was 81.2 kcal/kg/day, 77.6% of which was provided by PN. An average of 5.2 UIC measurements per patient were performed. The prevalence of ID decreased from the first assessment (83.3 %) to the last (45.8%). Three patients developed hypothyroidism secondary to iodine and selenium combined severe deficiency. In the adjusted analysis, iodine intake from oral or enteral nutritional formulas was associated with UIC (β= 0.71 [0.35, 1.07]; p < 0.001). Normal UIC values were observed in patients who reached ≥ 80% of the recommended iodine intake from nutritional formula.

**Figure 1**- Adjusted linear prediction of iodine intake adequacy from oral or enteral nutritional formulas on urinary iodine concentration (µg/L). The dotted line represents the lower limit of  UIC (urinary iodine concentration).

**Conclusion:**ID is highly prevalent in children with IF who receive long-term PN, and its frequency decreases with iodine intake by nutritional formula. Patients who developed hypothyroidism had severe combined iodine and selenium deficiency. Iodine and selenium status, thyroid function, and iodine intake should be monitored in children with IF.

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## Pumping Up Our Patients With Intestinal Failure

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#### Abstract

Introduction  
Children with short bowel syndrome (SBS) and intestinal failure (IF) require long-term parenteral support and are at high risk of electrolyte abnormalities and dehydration. We present a 6-year-old ex-34 week female with SBS-IF secondary to jejunoileal astresia, s/p small bowel resection, s/p STEP procedure, with poor weight gain, requiring intravenous fluids (IVF) and parenteral nutrition (PN) since birth. Patient’s anatomy is: 35cm small bowel, no ileocecal valve, full colon in continuity. She transferred to our hospital after multiple admissions to other hospitals with significant for metabolic acidosis (CO2 10 mmol/L), hypocalcemic clinical tetany (Calcium 7.4 mg/dL) , and acute kidney injury in the setting of PN being discontinued for three months. Upon transfer to our hospital and after ongoing correction of her electrolytes and acidosis, she was discharged home on daily IVF with weekly monitoring of her electrolytes and IVF adjusted accordingly. She had ongoing electrolyte abnormalities at home including low potassium (range 3-3.6 mmol/L), CO2 (range 12-22 mmol/L), calcium (range 7.9-9.3 mg/dL) and magnesium (range 1.3-2.0) despite receiving large amounts of electrolytes and being increased weekly in her IVF and orally. Parent reported that she was eating well by mouth (regular diet + nutritional supplements) and complained about high urine output overnight. Parent denied any vomiting or diarrhea as well as any missed days of IVF. Stool tests for malabsorption were significant for fat malabsorption and severe pancreatic insufficiency and she was started on pancreatic enzymes. Urine electrolytes revealed high urine potassium suggesting renal loss and she was referred for renal evaluation. Previous renal evaluation had revealed no renal losses. The ongoing electrolyte abnormalities that did not respond to increases of electrolytes in her IVF led us to obtain pump reports to ensure the IVF was being run as ordered.  
  
Methods  
Two pump reports at separate time intervals were obtained from the pump manufacturer through patient’s home care companies (two separate companies). The reports show dates and times the pump was accessed, turned on, turned off, and volume of fluid run through the pump daily.

Results

The first pump report revealed that patient’s IVF were given for 14/22 days (63.6% compliance). Importance of compliance with daily IVF as ordered was reinforced with the patient’s parent. Despite this, the second pump report a few months later revealed that patient’s IVF were given for 2/81 days (2.5% compliance).

Conclusion  
 When patients are demonstrating ongoing electrolyte abnormalities and/or unexplained poor weight gain that is not consistent with the electrolytes/calories being ordered in IVF/PN, pump reports can be useful to check compliance with IVF/PN administration. The utility of obtaining routine pump reports in patients on home IVF/PN to assess compliance needs to be further assessed in future studies.

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## Acrodermatitis enterophatica due to acquired zinc deficiency in a patient recieving long term parenteral nutrition

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#### Abstract

Background: The supply of essential micronutrients plays a critical role in children with intestinal failure (IF), because they are at risk of deficiency owing to reduced intestinal absorption, insufficient intake, losses from diarrhea, drains, fistulas and use of medications. Long term parenteral nutrition in children is associated with a risk of trace element deficiency, specially zinc, which is essential for immune function, antioxidant defenses and protein synthesis.

Methods: For the preparation of this case report, the appropriate information was obtained through consultation of data from the patient's electronic medical records. The study was approved by the ethics committee of the hospital and the consent form was applied to the patient's parents. Due to the clinical picture of the patient, it was not possible to apply an assent term

Results: This report presents a case of a 5- year- old boy, who has been receiving parenteral nutrition, since  birth, due to necrotizing enterocolitis (NEC). He was a premature baby of 25 weeks, weighed 1115 g at birth, with tracheal atresia, needing a traqueostomy. During the first weeks of life, he developed NEC with a massive intestinal resection and needed a jejunostomy. At 7 months of age, an intestinal reconstruction with duodenoileoanastomosis was performed. At 8 months of age, he was discharged home for the first time. He is completely dependent on parenteral nutrition, with poor tolerance for enteral nutrition.  After 5 years in parenteral nutrition, he developed a skin rash, first in perioral area, and quickly evolved into perineal region and around the gastrostomy and tracheostomy. He also presented with alopecia, brittle nails, and irritability. He was receiving the adequate amount of energy and protein requirement in his tailored made parenteral nutrition,with polivitamin and trace elements. His  zinc sulfate prescription was 40mcg/kg/day and the blood level of zinc was in the normal range of 0,5mcg/ml (reference range 0,5 – 1,1 micrograms/ml). We performed blood tests, to look for infection, specially fungus, micronutrient tests, including copper, iron, aluminium, cromium, manganese, vitamins and excluded all of them, including fatty acid deficiency and essential aminoacid deficiency. We also performed a tricology test that revealed zinc deficiency. After nine days of zinc sulfate suplementation with 200mcg/kg/day, he  solved completely the  skin eruption.

Conclusion: In general, it´s assumed that if we provide the ESPGHAN recomendation of trace element products, children will mantain their blood level of micronutrients adequate. However, children with intestinal failure depending on long term parenteral nutrition, might need a tailored provision of trace elements, accordingly to routinely measurements of blood levels and also taking into account their clinical condition with emphasis on those at risk of deficiency.

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## All is not lost: a new route for central venous access

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#### Abstract

Introduction:

We present the case of a 6 months old boy admitted to our intestinal rehabilitation center with short bowel syndrome, severe malnutrition and loss of central venous access.

Methods:

The child was born in Romania. He underwent short bowel resection following midgut volvulus at 2 days of life. The remaining intestine was estimated at 55 cm, with no ileo-caecal valve and the entire colon. He received intermittent parenteral nutrition (PN) and underwent numerous central catheter placements with either thrombosis or vein ligature after insertions which left the local team in the impossibility to insert further central catheters and to administer PN. His parents asked for the transfer to our unit. Because Romania is part of the European Union, the child could benefit from social security in our country.

He was admitted at 6 months of life in a cachectic pre-mortem state. A central catheter was inserted with difficulties in the superior vena cava through the right internal jugular vein. He was then slowly re-fed with PN to avoid refeeding syndrome. He received anti-coagulant prophylaxis and was discharged on home PN two months later.

He progressively developed superior vena cava syndrome with increased head circumference and mild cerebral oedema.

The CT-scan showed that the catheter was placed through a narrow varicose network and was obstructing the blood flow to the superior vena cava. All other central venous access were thrombosed including the inferior vena cava. At that stage, the child was still highly dependent on PN.

Results:

After several multidisciplinary discussions (pediatric gastro-enterologist, cardiac surgeons, anesthetists, cardiolgists and neuro-surgeons), the decision was made to perform a superior vena cava plasty and to insert a cuffed central venous catheter in the right atrium directly through the myocardium with an abdominal exit site (figure 1 and 2). The procedure was performed at the age of 13 months. The surgery was successful but was complicated with a PN pericardial effusion which was firstly mistaken for a chylo-pericardium. A second procedure was undertaken 5 days later to reposition the tip of the catheter which had migrated from the atrium cavity to the pericardium. From then, the PN was administered safely through this catheter for 15 months until the child was totally weaned off PN. Superior vena cava syndrome completely lifted. Anti-coagulant treatment was stopped 6 months after the procedure. Four months after weaning PN, he underwent another cardiac surgery to remove the central line.

Conclusion:

Cardiac surgery with insertion of a central line directly through the myocardium into the right atrium can be a life-saving procedure in infants and children with total loss of central venous access.

Figure 1

Figure 2

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## AETIOLOGY ROLE ON SBS PATIENTS' CLINICAL OUTCOME: A SINGLE EUROPEAN TERTIARY CENTRE EXPERIENCE.

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#### Abstract

Aim: to research aetiology's role on clinical outcomes in a population of patients with Short Bowel Syndrome (SBS) treated in the tertiary centre of Meyer Children's Hospital in Florence, Italy.

Study: we collected and reviewed retrospective clinical records of patients treated in our centre from 2018 to 2021. Patients were divided into five groups depending on the aetiology of SBS, and data were analysed to determine differences in clinical outcomes. Chosen variables were residual bowel length, the need for parenteral nutrition (PN), the percentage of colon left, the presence of ileocecal valve (ICV), and the number of central venous catheter (CVC) infective episodes (IE). Data were analysed by the Kruskal-Wallis test and Fisher exact test when appropriate.

Results: we collected 39 clinical records. Twelve patients had necrotising enterocolitis (NEC; 31%), nine had a volvulus (VO; both because of rotation defect or for other causes; 23%), eight had intestinal atresia (IA 20%), six had other aetiologies, such as mesenteric ischemia and long-segment Hirschsprung disease (OA; 15%), while the remaining four had gastroschisis (GS; 10%). The highest median residual bowel length was found in OA group (62 cm) and the lowest in VO group (33 cm). 50% of patients in NEC and IA groups have the whole colon; all patients in the GS group underwent partial colic resection. 43% of our patients still have an ileocecal valve, with the higher rate in IA group, while all GS patients had it resected. Only three patients in our population underwent total colectomy.

Approximately 50% of all groups except IA (41%) are not weaned off parenteral nutrition, and 27% of patients on PN do not receive it daily. There was no significant difference between SBS types and need of PN in IA group. The higher rate of CVC infections is in the GS group (75% with three or more IE), while the lowest is in the NEC group (16% with three or more IE).

Conclusion: No significant difference was found between groups regarding clinical outcomes. However, we noticed that all patients in the GS group had their ICV resected, had at least two CVC-related infective episodes and were submitted to autologous reconstructive surgery compared to the other groups. Indeed, aetiology contributes to clinical features, such as the presence of ICV or residual colon percentage, recognised as outcome predictors. Statistically, this association still must be demonstrated. Implementing SBS studies with larger populations could be a turning point in understanding this complex pathology.

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## Growth in male teenagers with a history of intestinal failure requiring reinitiation of PN during adolescence

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#### Abstract

Introduction: During adolescence there is an increased requirement for nutrition to go through puberty and achieve adult height. Nutrition also plays a role in the progression of puberty. Expected linear growth increases from 5cm per year to approximately 9.5cm per year for males during puberty [1].  Malnutrition in chronic disease impacts pubertal growth spurt [1]. Short bowel syndrome (SBS) and enteropathies can result in malabsorption of nutrients and in turn affect final adult height.

Methods: We reviewed our home parenteral nutrition (PN) cohort 1987- 2022 in a tertiary children's hospital. Patients were identified who had successfully weaned off PN earlier but required reinitiation of PN due to static growth. Other pathological causes of growth failure were ruled out. Height centile and z-score were documented prior to re-initiation of PN, a year later and at the time when PN was stopped again. Δ z height was calculated. Bone age delay (Tanner Whitehouse/TW2 method) was documented at baseline as a marker of delay in pubertal onset.

Results: Six patients were identified who had successfully weaned off PN, but whose growth had become static during adolescence. PN was restarted at this point following a multi-disciplinary discussion.  All six patients identified were male. Diagnoses were: trichohepatoenteric syndrome (THES) n = 4, SBS n = 2 (median small bowel length 42.5cm). All six patients received PN from the early neonatal period which was stopped at a median age of 5.1 years (range 1.33 years to 11.75 years). Enteral nutrition was optimised before restarting PN. The median age of restarting PN was 14.58 years (range 13.25 years to 15.67 years). Five patients had documented bone age at baseline; four out of five had delayed bone age. The median growth rate in the year prior to restarting PN was 3.5cm per year, in the first year on PN median growth rate was 7.95cm. Five patients stopped PN once adult height was reached. Patient B continues on PN.  Patients with the best growth response to PN at 1 year (C) and at completion of PN (E, F) were younger and all had more pronounced delay in bone age at baseline. The patient (D) with no measurable effect of PN on growth (D) was the oldest when restarting PN and did not have a delayed bone age at reinitiation of PN. Growth in patient A was less pronounced despite significant bone age delay at baseline.

Conclusions: Long term follow up, including bone age and growth monitoring, of patients with a history of intestinal failure in early life is essential in guiding reinitiation of PN in adolescence. Increased nutritional requirements in puberty are greater for males and patient concerns of adult height are factors that can drive this decision. The age at which PN is restarted is important to achieve maximal height velocity.

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## Use of ultrasound liver elastography to monitor IFALD in SBS patients: experience from an Intestinal Rehabilitation Unit

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#### Abstract

Introduction: Intestinal failure-associated liver disease (IFALD) is one of the most common and serious complications of long-term parenteral nutrition (PN) occurring in short bowel syndrome (SBS) patients. IFALD may evolve into end-stage liver disease, a primary cause of morbidity and mortality. Ultrasound liver elastography is a non-invasive technique widely used in adult population to evaluate liver damage. In our study, we compared FibroScan® data with gGT and histopathological findings in SBS patients who underwent autologous gastrointestinal reconstruction (AGIR) procedures to evaluate and monitor liver parenchyma damage. The study aims to establish a correlation between FibroScan®, results and gGT as a measure of long-term liver damage in SBS patients. The goal is to assess the role of FibroScan® in monitoring liver deterioration in pediatric SBS patients.

Methods: Patients admitted to the Pediatric autologous bowel reconstruction and rehabilitation unit between 2019 and 2022 were considered. Inclusions criteria were: SBS patient eligible for an AGIR procedure performed at our institution, patients with more than 90 days of TPN and blood test not older than 6 months. FibroScan® was performed by our dedicated hepatologist, who collected and analysed the results. USS data were compared to liver biopsy taken during surgery and to gGT values.

Results: 8 patients were included in the study, 3 were lost at follow-up. All the results were reported according to the FibroScan® standardized tables. Four patients underwent the SILT procedure, 2 underwent STEP procedure, and 1 underwent colonic interposition. PN support was started for a medium of 4.75 years (range 1-8 years) before FibroScan® performance. All patients presented S0 Steatosis grade, according to CAP score, ranging from 147 dB/m to 226 dB/m. The degree of liver stiffness ranged from 2,7 to 5,8 kPa. Interestingly, biopsy results were negative for steatosis, while minimal periportal fibrosis was found only in one patient. gGT were all within normal values.

Conclusions: Our study supports FibroScan® to be a safe, non-invasive technique to monitor liver parenchyma damage in SBS patients undergoing PN. FibroScan® results were comparable to histopathological analysis. Our patient presented no liver  at blood tests, as confirmed by normal gGT values. Absence of liver damage was confirmed at histopathological analysis and at clinical examination.  Further studies are required to confirm our finding, but this study may be a starting point to shed a light over new minimally invasive technique to prevent IFALD.

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## Conversion from SMOFlipid® to Omegaven® as a Salvage Therapy for Severe Intestinal Failure Associated Liver Disease

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#### Abstract

Introduction

Parenteral nutrition (PN) is a life-saving therapy. One of the most feared complications of intestinal failure and PN is the development of intestinal failure associated liver disease (IFALD). Modern lipid strategies with the use of a mixed lipid emulsion decreased the incidence of IFALD, but some patients still develop liver dysfunction. Omegaven® is an effective treatment of IFALD in patients receiving intralipid®, but little is known about the role of Omegaven® for the treatment of IFALD in patients treated with SMOFlipid®. The aim of this study is to review the role of Omegaven® as a salvage therapy in patients receiving SMOFlipid® that developed progressive IFALD.

Methods

Retrospective study of patients managed by the intestinal failure program at The Hospital for the Sick Children from 2015 to 2022. The study cohort included children that developed advanced IFALD  with the use of SMOFlipid® and converted to Omegaven for at least 7 weeks. Advanced IFALD was defined as conjugated bilirubin persistently over 50 µmol/L (2.9mg/d) and no near future possibility of PN weaning. The primary outcome was conjugated bilirubin <34 mmol/l at 6 months after initiation of Omegaven®. Descriptive statistics were used.

Results

From 2015 to 2022 eight patients developed progressive IFALD while receiving SMOFlipid® and were treated with Omegaven®. Mean conjugated bilirubin at time of Omegaven initiation was 114 mmol/l (range 42-183). We excluded 2 patients from the analysis; one due to short period of treatment (4 weeks until a liver-intestine transplantation) and the other for multi-system disease that may impact the liver (chronic granulomatous disease). Following conversion to Omegaven®, 3 patients (50%) achieved improvement in bilirubin levels (Figure 1). The length of time to achieve conjugated bilirubin below 34 µmol/L (2 mg/dL) was a mean of 18.6 weeks (range 11-25; SD 7.09). In the non-responders (N=3), one patient died and 2 underwent multi-visceral transplantation. Unlike the responders, non-responders had signs of more severe IFALD prior to treatment initiation. These include: previous use of Omegaven® for advanced IFALD while on Intralipid® (n= 2); gastrointestinal bleeding secondary to portal hypertension (n= 2); need for continuous glucose infusion to prevent hypoglycemia (n= 2); and higher range of INR (1.4-2.7 in non responders vs. 1.1 - 1.2 in responders).

Conclusions

Omegaven® reversed progressive IFALD in 3 out of 6 patients (50%) who received SMOFlipid® as their PN lipid source. Treatment failure was common in children with more severe IFALD prior to Omegaven® initiation, and history of previous use of Omegaven® for IFALD reversal. Larger cohorts are required to confirm the findings of this case series.

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## Intestinal Rehabilitation at Home: A Retrospective Review

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#### Abstract

INTRODUCTION: In patients with short bowel syndrome (SBS), chronic intestinal failure (CIF) may be reversible with Intestinal Rehabilitation (IR). IR is a process of maximizing the digestive and absorptive capacity of the remnant short bowel to improve uptake of fluid, electrolytes, and nutrients. The goal of IR is to reduce the need for the routine use of parenteral nutrition (PN) therapy and intravenous (IV) fluids. The traditional home infusion model does not include IR. A national home infusion provider implemented an at home IR program. The purpose of this abstract is to evaluate the impact of this program on PN volume and infusion days.

METHODS: A retrospective review of all patients who completed the IR program from May 2021 through August 2022 was conducted. Patient enrollment criteria included SBS diagnosis, 18 years of age or older, clinically stable, able to tolerate an oral diet, amenable to weekly contact for education and follow-up, interested in weaning PN and under the care of a medical team open to program recommendations. Patients who did not complete the 3-month program were excluded in the review. After 3 months, patients who remained on service with PN and continued to be followed by a certified nutrition support dietitian were also reviewed and analyzed at 6 and 12 months. Upon enrollment, a comprehensive SBS evaluation was completed. An individualized patient plan was created and weekly patient follow-up included intensive diet education and manipulations individualized to each patient’s remaining bowel anatomy and response to prior week’s suggested interventions.

RESULTS: Fourteen patients completed the program; 5 (36%) males, 9 (64%) females, average age of 63. Primary diagnoses resulting in SBS were 4 (29%) inflammatory bowel disease, 4 (29%) ischemic bowel, 3 (21%) cancer, 2 (14%) small bowel obstruction, and 1 (7%) familial multiple polyposis syndrome. The average length of small bowel remaining was 135 cm, 4 (29%) with an unknown length. Seven (50%) had a colon and 3 (21%) had an ileocecal valve.

Table 1 summarizes the IR patient outcomes at 3, 6, and 12 months, including number of patients enrolled at each interval. There was a steady decrease in average mL per day and days per week.

CONCLUSIONS: An at home IR program can be successful in assisting patients reduce PN volume and infusion days, ultimately improving their quality of life. It is a valuable resource for patients who are unable to travel to an IR Center of Excellence or require more intensive follow-up. The ability for patients to have 1 on 1 intensive diet education, medication management, and guidance for managing SBS symptoms by their home infusion provider allows for convenience, continuity of care, improved patient outcomes and cost savings.

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## Tapering enteroplasty or bowel lengthening? – A surgical algorithm for pediatric intestinal rehabilitation patients with bowel dilatation

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#### Abstract

Introduction:

Bowel dilatation regularly occurs in children with short bowel syndrome (SBS). Despite the common nature of this finding, there is no standardized approach for dilated bowel in affected patients with regard to surgical indications and techniques. This is mainly due to the highly individualized conditions of the patients. Several surgical techniques have been proposed for the management of bowel dilatation in pediatric SBS patients including bowel lengthening procedures. However, certain limitations for the different approaches have meanwhile been identified. Aim of this study was to systematically analyze indications and decision making for surgery of children with SBS and bowel dilatation, and to establish an algorithm for the decision making process.

Methods:

We retrospectively evaluated all patients of our intestinal rehabilitation program that underwent surgery for bowel dilatation between 11/2010 and 12/2020. We registered indications and techniques of the surgical procedures and analyzed their outcome with special regard to surgical and functional outcome. Based on these retrospective data we formulated an algorithm for the surgical decision making process, which we then prospectively analyzed for the subsequent operations until 09/2022. Success of surgery was evaluated by matching the individual indication/aim of surgery with the actual postoperative patient condition.

Results:

Of the 203 children treated in our intestinal rehabilitation program,  55 underwent surgery for dilated bowel and were evaluated for this study. Indications for surgery were established in our multidisciplinary treatment board. Most common surgical procedures were resection of stenosis with primary anastomosis (n=28), stoma revision (n=11), bowel tapering (n=10), and Serial Transverse Enteroplasty (STEP, n=9). With focus on the surgical decision making process, the main factors for establishing our algorithm were presence of mechanical obstruction, bowel length, and motility. After retrospective evaluation of patients’ data, this formulated algorithm was successfully used prospectively, resulting in a surgical success rate of 89%.

Conclusion:

We successfully established a surgical algorithm for decision making in children with SBS and bowel dilatation. This algorithm is now being evaluated prospectively in our intestinal rehabilitation program. Further analyses are necessary to evaluate the application of our algorithm in a broader fashion

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## Complications of vascular Ehlers-Danlos Syndrome: Management by an Intestinal Failure Multidisciplinary Team

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#### Abstract

INTRODUCTION

Vascular Ehlers-Danlos syndrome (EDSv) is a connective tissue disease associated with mutations in COL3A1 gene. Clinical features include a characteristic facial appearance, translucent skin and spontaneous arterial, uterine or intestinal rupture. Spontaneous intestinal perforation is commonly described, with frequent surgical complications. Secondary intestinal failure (IF) has been rarely reported.

METHODS

Report of 2 pediatric cases with IF due to complications of EDSv and a literature review with focus on surgical management.

RESULTS

Case Nº1: 13 ys, male. History of laparotomy for jejunal perforation, complicated with anastomotic leakage, ileal and duodenal spontaneous perforation and multiple enterocutaneous fistulae. Palliative treatment with vacuum- assisted closure was indicated. Clinical diagnosis of EDSv was assumed. 9 years after diagnosis, the patient is currently dependent on parenteral nutrition (PN) with a type 1 SBS (100 cm of residual IBL).

Case Nº2: 4 ys, male. Operated for an initial diagnosis of appendicitis, several surgical procedures were needed because of sigmoid colon and multiple jejunal perforations, needing a jejunostomy at 100 cm. EDSv was suspected and COL3A1 gene mutation confirmed. After 18 months of PN, enteral autonomy was achieved following ostomy closure. Gentle tissue handling, protection of anastomotic line with fibrin glue and cautious postoperative care were applied.

Literature review: IF associated with EDSv has been reported in isolated cases. No published reports of patients as young as 4 years old with surgical complications and long term PN requirement were found. Risk of perforation, anastomotic leakage, operative bleeding and fistulae is higher in this group, increasing mortality. A significant delay in the diagnosis of EDSv is the rule. Visceral re-perforation rate after colo-colic anastomosis has been reported to be higher, compared with colectomy and ileo-rectum anastomosis. Adequate exposure, double-line anastomosis, use of fibrin sealants and avoidance of fast- track postoperative management have been recommended.

DISCUSSION

EDSv may have a severe impact on intestinal autonomy, impairing quality of life by multiple operations and complications. Most severe cases may require long term PN. A high index of suspicion of EDSv when faced with recurrent anastomotic leaks or fistulae is crucial. Surgical reconstruction is feasible, although long term follow-up is mandatory to detect recurrence. Early management of severe cases by an intestinal rehabilitation MDT may be recommended to reduce complications and improve outcomes.

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## Surgical Techniques for non-Short Bowel Syndrome Children with Intestinal Failure: Reducing Length and Improving Outcome.

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#### Abstract

Introduction

Patients with pediatric intestinal pseudo-obstruction (PIPO) and congenital diarrheas and enteropathies (CODE), are unique intestinal failure patient populations. Bowel resection is not a routine and integral part of their treatment options. Herein, we describe extensive intestinal resection as a treatment modality in two distinct pathologies of IF - PIPO and microvillus inclusion disease (MVID).

Case reports

Patient 1: A 2-year-old female with MVID (MYO5B mutation) with difficult to manage fluid and electrolyte requirements due to high volume diarrhea refractory to medical management including antisecretory medications. In an attempt to better control the patient’s fluid and electrolyte needs we hypothesized that a reduction in hypersecretory bowel mass with an extensive enterectomy would decrease the amount of electrolyte transport diarrhea. A resection of 178 cm of jejunoileum, ileocecal valve, and a subtotal colectomy was performed. The remaining 40 cm of jejunum was anastomosed to the sigmoid colon. Following this intervention, a significant reduction in parenteral nutrition electrolytes and volume needs were noted. Specifically, sodium, potassium, phosphate, and total fluid intake (TFI) requirements dropped from 37.7 mmol/kg, 6.9 mmol/kg, 2.4 mmol/kg, and 202.5 ml/kg/day respectively, to 10.5 mmol/kg (Na), 2.5 mmol/kg (K), 0.7 mmol/kg (P), and 123.3 ml/kg/day (TFI) 18 months after the surgery (Figure 1). In addition, improved quality of life and easier daily management at home were reported by the parents.

Patient 2: A 15-year-old female diagnosed with PIPO, TPN dependent, and with a terminal ileostomy performed at 3 years of age. Her surgical course was complicated by recurrent episodes of stoma prolapse. Numerous operations were performed including revision, resiting and pexing. In addition, the patient also presented with frequent episodes of mechanical bowel obstruction caused by the dilated, atonic, fluid-filled small bowel kinking at the level of the fascia of the ileostomy site. An enterectomy was performed leaving the patient with 135cm of small bowel to a jejunostomy. A tapering enteroplasty was also performed decreasing the caliber from 8cm to 3cm. Although the patient was primarily PN dependent she enjoyed eating for comfort; therefore, 135cm was chosen to permit some absorptive capacity. Two years after the procedure, the patient has had no episodes of stoma complications or bowel obstruction and has not required further surgical intervention.

Conclusions

These two cases, illustrate less common forms of autologous bowel surgery to improve functional status. The surgical approach was focused on reducing complications related to the intestinal failure's etiology. Consideration of extensive enterectomy in patients with no prospect of TPN weaning, can improve quality of life, reduce morbidity, and optimize PN and IF management.

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## Intestinal Rehabilitation from Severe Enteroperitoneal Tuberculosis a 14-year Prospective Protocol

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#### Abstract

Introduction:

Tuberculosis(TB) is a systemic disease and is on the rampage. Each year about 1.7 million people die of TB and 9 to 13 million new cases occur worldwide. We have found that for every 25 to 30 cases of lung TB there is one case of enteroperitoneal TB(ETB). Humans are the natural reservoir of M. tuberculosis and about 500,000 people worldwide are infected with a multidrug-resistant strain of M. tuberculosis.  
 ETB is a tragic life threatening condition associated with a completely "frozen" abdomen : an abdominal catastrophe. Mortality is up to 26%. Clinical presentation makes even the situation worse for it presents in some cases without concomitant lung disease so the first clinical impression is pelvic malignant disease. The concept that antibiotic/chemotherapies are the only way of curing very serious cases of ETB is full of prejudice, as the corner stone is the association of antibiotic/chemotherapies with NS.

Methods

From 2008 to 2022, 15 patients were included in a strict prospective protocol. We were specially interested in severe cases of ETB. Once they had a diagnosis of ETB, patients were included . Diagnosis: by laparotomy in 10 /15 cases, laparoscopy in 2/15, endoscopy 1/15, 1/15 by polymerase chain reaction in ascitic fluid and 1/15 by CTScan.  
Protocol :  
  
1) Total Parenteral Nutrition(TPN) or Total Enteral Nutrition (TEN) were started at the same time that multidrug treatment was started, with TB specific intravenous antibiotic and chemotherapy treatments.  
  
2) TPN and TEN were given sequentially. Once patients were discharged they were given daily oral supplements with enteral nutrients supplementing oral food.  
  
 3) In case of surgical complications, the operation was performed by the surgeon (3/15 patients) of the NS team, also responsible of supervising NS treatment and follow up until the patient was free of disease.

Results:

Age of diagnosis: 36.2 ±18.91 and 16 to 91 years.

Patients had NS from 13 to 260 days. Mean of 143.92 ± 95.29 days. 11/14 patients started NS with TPN as enteral tolerance was impossible. 2/14 patients could start NS with TEN as TPN was not necessary .

Periods of TEN(Intestinal Insufficiency) were from 41 to 260 with a mean of 177.66 ±104.6 days and periods of TPN(Intestinal failure Types I and II) were from 13 to 240 with a mean of 90.25 ±76.32 days.

3) 13/15 patients collaborated with careful follow up, after discharge were considered cured of the disease at the end of multidrug and NS treatments.

4) 6/15 patients required surgery either for diagnosis or treatment of complications . 3/15 patients had fistulas during the protocol, 2/15 required surgical treatment and 1/15 closed with TPN.

             5) 9/15 patients had no lung TB making diagnosis very difficult

Conclusions:

The corner stone, we firmly believe, to cure severe cases of enteroperitoneal TB is NS as multidrug treatment is the conventional approach that will only be properly delivered once NS is conceived as such, whatsoever.

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## Preoperatory and Postoperatory Nutritional Repletion aiming at Intestinal Rehabilitation : A 5-year Prospective Protocol for Large Retroperitoneal Malignant Tumors

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#### Abstract

Introduction:

Severe cases of solid retroperitoneal tumors may occupy the abdominal and/or pelvic cavities partially and/or completely and may render planned surgical treatment impossible without nutritional support(NS) and/or lead to severe postsurgical complications that may be impossible to manage without nutritional support because of serious intestinal compromise.

Methods

Between 2017 and 2022, 20 adult patients were diagnosed of retroperitoneal tumors and were operated on by a surgeon of a NS Team. They were prospectively studied to ascertain if they needed nutritional support pre and/or postoperatively, as every effort was made to perform radical curative resections. Surgical specimens follow up, with concomitant Surgical and Pathology Consultants evaluation of tumors, at pathology lab, to study their pleomorphic characteristics and if possible, their blood supply, was performed.

Results:

6/20 (30%) patients required NS, enteral and/or parenteral for periods of 4 to 50 days, mean±SD: 16.83±17.9 days. We received even one patient that was deemed inoperable in other referral institution, and did well within the protocol. 19/20 patients were discharged in good condition, 1/20 patients died in spite of nutritional support. Largest tumors of 11.5 and 17.5 kg required NS due to very poor postoperatory conditions and did well. Patients with worst intraoperatory bleeding (3 to 6 liters) did not require NS.

Tumors were 12 sarcomas, 4 lymphomas, 2 stromal, 1 fibrous and 1 fibrotechoma. Photos may show how aggressive and large these tumors were and their large blood supply as well as macroscopic features and organs resected like stomach, kidney, small and large bowel.

Conclusions:

Large retroperitoneal tumors can be dealt with considering that nutritional support is an important weapon before and/or after surgical treatment even in apparently hopeless patients in order to obtain nutritional repletion and therefore intestinal rehabilitation. We advise that protocolized NS should be considered in radical and aggressive surgical techniques for retroperitoneal malignant tumors.