Small Bowel Dilatation in Pediatric Intestinal Rehabilitation Patients: Surgical Aspects and Proposal of a Treatment Algorithm

Matthias Schunn1, Andreas Busch1, Nalan Darwish1, Simon Scherer1, Ilias Tsiflikas2, Steffen Hartleif3, Jens Gesche1, Ekkehard Sturm1, Johannes Hilberath1, Matthias Kumpf1, Jörg Fuchs1 and Steven W Warmann1*

1Department of Pediatric Surgery and Pediatric Urology, University Children’s Hospital Tuebingen, Germany
2Department of Pediatric Gastroenterology and Hepatology, University Children’s Hospital Tuebingen, Germany
3Department of Diagnostic and Interventional Radiology, University Hospital Tuebingen, Germany
4Department of Pediatric Cardiology, Pulmonology and Pediatric Intensive Care Medicine, University Children’s Hospital Tuebingen, Germany

Abstract

Background: Standardization of surgical indications and procedures concerning bowel dilatation in children with Short Bowel Syndrome (SBS) is difficult due to the variety of individual patient conditions. Aim of this study was to analyze surgery for bowel dilatation in children with SBS treated within our pediatric Intestinal Rehabilitation Program (pIRP).


Results: There were 47 pIRP patients undergoing surgery for bowel dilatation. Most common indications for surgery were bowel/anastomotic stenosis (n=20) and stenotic enterostomies (n=15). Most common procedures were stenosis resection with re-anastomosis (n=26), stoma revision (n=9), bowel tapering (n=9), Serial Transverse Enteroplasty (STEP, n=5), and stricturoplasty (n=4). After a median follow-up of 33 months (0-166), 8 patients achieved complete enteral autonomy, 15 patients were fed predominantly via the enteral route. Seven children needed redo-surgery for intestinal stenosis.

Conclusion: An interdisciplinary approach for the management of children with SBS and bowel dilatation is crucial. Bowel motility represents an essential aspect of the surgical decision making process regarding indications and techniques. We propose an algorithm for the management of pIR patients with bowel dilatation.

Keywords: Short bowel syndrome; Bowel dilatation; Intestinal rehabilitation
Intestinal dilatation is a common phenomenon in children with SBS or intestinal failure. Mechanical and/or functional factors are possible reasons for this pathology. Bowel dilatation in children with SBS has been identified as poor prognostic factor in terms of reaching enteral autonomy. It has also been found to predict prolonged need for Parenteral Nutrition (PN) and mortality and to being associated with mucosal damage, bowel-derived blood-stream infections, and hepatic injury [9-12]. The task of intestinal rehabilitation is therefore on the one hand to identify the underlying cause of bowel dilatation as precisely as possible and on the other hand to address the respective problem by means of individually adapted therapeutic measures.

Surgery is regularly being applied as treatment element in children with SBS and intestinal dilatation; however, surgical procedures are not in every case necessary as initial step during management. Most surgical techniques used in short bowel surgery are routine for many experienced pediatric surgeons. Nevertheless, in addition to the correct technical execution of the corresponding procedures, it is crucial to decide which procedure is appropriate and how this particular surgical procedure should be used in the context of interdisciplinary treatment. The aim of our study was to analyze and to give an overview over the different surgical techniques used in pIRP patients with bowel dilatation and to propose a treatment algorithm.

**Methods**

In a retrospective analysis, we evaluated all children of our pIRP who underwent surgery for dilated bowel between 11/2010 and 12/2019. The study was approved by the institutional ethical Committee of the University of Tuebingen (project No. 493/2020BO). In the study we analyzed patient-specific data, surgical data, surgical outcome, and outcome with respect to underlying diseases.

Initially, all children underwent diagnostic evaluation and all appropriate non-surgical measures of the pIRP were applied (Figure 1). Diagnostic workup was extensive and included combination of measures if necessary (Figure 2 and 3). The assessment of nature and extent of intestinal dysmotility was made on a multifaceted basis, for which the following factors were used: underlying diagnosis, previous clinical course and previous operations, current clinical condition, radiological diagnosis (especially sonography and contrast fluoroscopy), neurohistopathology where appropriate, and endoscopy. Indications for surgical interventions were discussed in an interdisciplinary pIRP conference.

During surgery we always performed a complete adhesiolysis in order to completely explore all intestinal sections. Measurement and documentation of lengths of all bowel sections was done thoroughly. If necessary, an interdisciplinary discussion was carried out during operation, in some cases a simultaneous endoscopy was performed. We made extensive use of intraluminal and abdominal drainage and decompression. Every child was administered to the intensive care unit after surgery. Postoperative care was again administered according to the interdisciplinary standards of our pIRP.

Before surgery every patient was PN-dependent to a varying degree. Since objective determination of surgical treatment success is difficult due to the individuality of patients’ conditions, we registered during follow-up whether there was an improvement of those clinical conditions, which led to the indication of surgery in every single case. We also assessed whether there were further operations necessary for re-occurring dilatation. Finally, we categorized the outcomes after surgery in a descriptive manner and assigned each patient into one of four groups with regard to enteral nutrition: 1) complete enteral autonomy, 2) caloric intake predominantly via the enteral route with some PN substitution, 3) caloric intake predominantly via PN with some additional enteral feeds, 4) complete dependence on PN without tolerance of any enteral nutrition.

**Statistical analysis**

Data analysis was performed using the JMP 11.2 statistical software (SAS Institute, Cary, NC). Quantitative data are summarized with the median, Interquartile Range (IQR), and minimum and maximum values.

**Results**

**Patients’ related data**

Within the study period we treated 184 patients in our institutional pIRP. Of these patients 76 (41.3%) presented with bowel dilatation. Surgery was performed eventually in 47 of the 76 children (61.84%) with dilated bowel.

The underlying diagnoses of the 47 operated children (30 boys, 17 girls) were anatomical SBS in 35 cases (12 Necrotizing Enterocolitis (NEC), 9 gastroschisis, 7 intestinal atresia, 7 volvulus) and chronic intestinal failure in 12 cases (5 Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS), 4 intestinal aganglionosis, 2 Chronic Intestinal Pseudo-Obstruction (CIPO), 1 Microvillus Inclusion Disease). Anatomical reasons for bowel dilatations were intestinal/anastomotic stenosis in 20 cases, stenotic enterostomies in 15 cases, and functional problems together with intestinal adhesions in 12 cases. Five of the 15 stenotic enterostomies had been placed in the former laparotomy wound. In our series we had 5 children who had undergone a STEP procedure elsewhere, in which bowel motility was poor and postoperative dilatation occurred as functional complication. Of the 47 analyzed children, 6 were treated only in our institution, whereas 41 children were submitted to our pIRP from external institutions. The 47 patients had undergone between 1 and 7 abdominal operations before being operated on for bowel dilatation at our hospital.

**Surgery-related data**

Indications for surgery were one or several of the following: persisting mechanical transport impairment with intestinal stasis and/or vomiting, small bowel bacterial overgrowth irrespective to conservative treatment, failure to thrive, failure to increase enteral feeds, pre-stenotic bowel inflammation and/or ulceration as observed on endoscopy irrespective to conservative treatment, and repeated episodes of blood stream infections arising from the dilated bowel.

The 47 patients underwent 56 procedures: Resection of stenosis (intestinal/anastomosis/enterostomy) with direct anastomosis in 26 cases, enterostomy revision in 9 cases, bowel tapering in 9 cases, STEP procedure in 5 cases, stricturoplasty in 4 cases, and adhesiolysis as only surgical procedure in 3 cases. Median operative time was 192 minutes. Median residual small bowel length in the 35 children with SBS was 25 cm.

**Outcome**

Median follow-up was 33 months (Table 1). At last follow up 8 children had reached full enteral autonomy, 15 children were fed predominantly via the enteral route with additional PN. Sixteen children were predominantly on PN with additional oral feeds, and 4 children had only PN without any oral intake. An improvement
of the intestinal conditions, which lead to the indication for surgery, was achieved in 39 patients, whereas there was no improvement in 4 patients despite absence of bowel dilatation after surgery (no data for 4 patients in this regard, see below).

In only one child there was a major surgical complication in form of repetitive postoperative intestinal perforations at different sites (anastomoses as well as bowel suturing for injuries during adhesiolysis). This occurred despite internal and external drainage of the complete intestine. The child underwent 2 subsequent operations, however, in the end the small intestine had to be removed completely and a Hartmann situation had to be established.

**Redo-surgery**

In 7 of the 47 children a redo surgery was necessary due to re-occurring bowel dilatation. The median time between the first and
second operation for dilatation in these 7 children was 23 months (range 3 to 80). Reasons for redo-surgery in the 7 children were anastomotic stenosis (n=4), stenotic stricturoplasty (n=1), stenotic enterostomy (n=1), and functional dilatation (n=1). Accordingly, we performed as redo procedures stenosis resection with direct anastomosis in 3 cases, stenosis resection with direct anastomosis plus STEP procedure in 2 cases, enterostoma revision in 1 case, and bowel tapering in 1 case.

Survival

Forty-three children are alive, 4 of the 47 patients died during follow-up. Two patients died in the direct postoperative course: One died because of postoperative Severe Inflammatory Response Syndrome (SIRS) after STEP, which was uncontrollable by intensive care measures. The second patient who died in the postoperative course had a preexisting liver insufficiency and developed complete liver failure after surgery with uncontrollable ubiquitous nonsurgical bleeding. Both children had a complicated course and were in complex clinical conditions before being admitted for surgery to our institution.

The other two patients died independently from the perioperative stay in our hospital (one because of sepsis from a catheter-associated blood stream infection 46 months after surgery and one because of renal failure 36 months after surgery).

Discussion

Small bowel dilatations in children with chronic intestinal failure can be caused by mechanical and/or functional factors. Dilatations can lead to local (bacterial overgrowth, inflammation, ulceration, intestinal blood loss) or systemic consequences (malabsorption, bacterial translocation with blood stream infections and sepsis, hyperbilirubinemia and inflammation affecting liver function, failure to thrive, acidosis, and anemia). It is known that intestinal dilatation is a possible cause for increased morbidity and contributes to an increased mortality in affected children. Therefore, dilatations have to be considered problematic, and their identification, assessment, and management should be a major goal of Intestinal Rehabilitation (IR), especially if associated complications occur [9-12]. The exact analysis of the causes of intestinal dilatations in children with SBS can be difficult. Often there are multifactorial components and there is only rarely one singular underlying factor. In particular, identification of characteristics and influence of a functional component in contrast to mechanical factors often presents a challenge for diagnostic workup. A mechanical transport disorder is often associated with a functional problem when the intestine is dilated. Conversely, a pronounced functional transport disorder can cause similar symptoms as intestinal stenoses. This must be taken into account on the way to establishing the indication for a surgical intervention or, for example, if the intestinal activity does not immediately resume after surgical correction of a supposedly mechanical cause. Although surgical procedures cannot completely reverse a functional disturbance of intestinal transport, surgery can help to improve the complications of intestinal dilatation caused by functional disorders. Therefore, a consistent and comprehensive diagnostic workup should always be carried out if necessary with a combination of different approaches. At the time of indication for surgery, non-surgical measures should have been exhausted and no further improvement is expected. If a treatment-refractory mechanical component has been identified as the cause of dilatation, then surgery is certainly the decisive treatment step; nevertheless, all IR components should be applied on the way to surgery in order to provide the best possible surgical conditions and to minimize possible risks with regard to effects on the entire organism.

Analysis of the different surgical procedures in our study showed
that conventional techniques are most frequently used by operating surgeons, whereas lengthening procedures are less frequently needed. Intestinal transplants should only be reserved for very few exceptional conditions. Despite the possibility of this procedure at our institution, none of the patients analyzed in this cohort had to be considered for intestinal transplantation. In our opinion, the main purpose of lengthening procedures such as STEP and Longitudinal Intestinal Lengthening and Tailoring (LILT) is not so much to gain every last centimeter of bowel length, but rather to extend the contact time between the intestinal mucosa and nutrients as well as to improve ionization of the intestine. To our understanding, intestinal motility is of exceptional importance when considering the use of an intestinal lengthening procedure. For example it makes little sense to turn a 30 cm long dysmotility intestine into a 45 cm long dysmotility intestine considering the associated operative trauma and the imminent risk of further intestinal dysfunction. A relevant number of patients in our series underwent STEP procedures elsewhere despite having poor bowel motility prior to surgery. Accordingly, these children did not benefit from the lengthening procedure, and the indication for these operations has to be critically reflected. The importance of motility for the application of lengthening bowel surgery is becoming increasingly accepted within several international pIRPs [11-14]. Hukkinen et al. [13] observed that intestinal rehabilitation surgery had significantly poorer results in patients with intestinal dysmotility as compared to patients with SBS and no signs of dysmotility. In a study of patients with neonatal ultra-SBS, the diagnosis gastrochisis was identified as a risk for nutritional failure requiring bowel transplantation [2]. The problem of intestinal re-dilatation after surgery in pediatric SBS patients is a well-known fact [15-17]. Miyasaka et al. [15] identified re-dilatation after lengthening procedures as indicator of poor outcome in children with intestinal failure. Barrett et al. [16] observed that redo lengthening procedures have a poor outcome. Khan et al. [17] observed in a multicenter cohort study, that the magnitude of surgical burden is associated with intestinal failure. In most cases the quantification of intestinal dysmotility represents a serious challenge. For the assessment of intestinal dysmotility there are still no objective standardized examinations. Instead, the assessment is carried out in an interdisciplinary round, taking into account multi-layered factors. In our centre, the procedure (as described in the methods section) is similar. In the future, it will be the task of scientific research on the topic of short bowel syndrome/intestinal failure to establish a corresponding validation. Taken together, we are rather reluctant with performing intestinal lengthening procedures in children with SBS and dysmotility bowel as for example in cases of complicated gastrochisis. A lot of experience of the whole IR-team and an interdisciplinary approach to diagnosis and therapy of affected children is essential. Due to these aspects we confirmed the observation previously described by other groups, that certain surgical procedures in children with SBS are more successful within IRP than outside of corresponding programs and should therefore be performed under the umbrella of a multidisciplinary IRP. Children with SBS and/or chronic intestinal failure benefit significantly from treatment applied by a multidisciplinary IR-team [18-20].

In our cohort some specific aspects were observed regularly as mechanical causes for the formation of intestinal dilatations. In a number of children who had undergone outward surgery, enterostomies were placed in the laparotomy wound and subsequently became stenotic. Since this can develop through the processes of scarring and wound healing without any influence on it, we recommend to place enterostomies using a separate passage through the abdominal wall, even if this means an additional scar in the abdominal area later on.

Our experience has shown that stricturoplasties, if applied in the area of fixed scars or stenotic anastomoses in the intestine, contain a relevant risk to constrict again leading to subsequent bowel dilatation. We therefore perform stricturoplasties only in areas of the bowel where there is no scarring or increased inflammatory activity. In cases of fixed stenoses or inflammatory bowel activity we prefer stenosis resection with re-anastomosis, especially since correspondingly altered sections are usually short and do not contribute significantly to the function of the intestine.

Based on our own experience and considering different observations of other pediatric IR-groups we developed a treatment algorithm for children with SBS and bowel dilatation. Of course there are always situations, in which individual solutions are required, however, good treatment results and favorable outcomes were achieved in our cohort using the presented algorithm as a basic framework:

**Treatment algorithm for children with SBS/chronic intestinal failure and bowel dilatation**

If bowel dilatation occurs, the patient undergoes comprehensive diagnostic workup. If there is a predominant mechanical problem then this is an indication for surgery and the child should be operated on after all non-operative IR-measures have been optimized. Together with removal of the mechanical obstacle tapering surgery should be considered (Figure 4).

If the reason for dilatation is functional and there are no complications, and if the dilatation is stable or regressive then this child can be observed and re-evaluated with therapeutic steps being taken according to the further course.

If in case of bowel dilatation the reason is functional and dilatation is progressive and/or there are associated complications (refractory bacterial overgrowth, failure to thrive, failure to increase enteral feeds and wean off PN, impaired passage, recurring blood stream infections), then the full scale of non-operative IR-procedures needs to be applied. If there is a subsequent improvement, then the child can undergo observation and re-evaluation; if there is no improvement, surgery should be considered.

If tapering surgery is considered and there is an impaired motility of the bowel, conventional tapering of the bowel should be performed irrespective of the bowel length. If bowel motility is good and there is a sufficient bowel length, again conventional tapering of the bowel is sufficient. If bowel motility is good and there is an insufficient bowel length, then a lengthening procedure (STEP, LILT) should be performed.

**References**


