

## INVITED REVIEW

# Role of ileal diversion in pediatric inflammatory bowel disease

Amanda A. Simard<sup>1</sup> | Swetha Kotamraju<sup>1,2,3</sup> | Jennifer R. DeFazio<sup>3,4</sup> |  
Joseph A. Picoraro<sup>1,2,3</sup> 

<sup>1</sup>Department of Pediatrics, Columbia University Irving Medical Center, New York, New York, USA

<sup>2</sup>Division of Pediatric Gastroenterology, Hepatology and Nutrition, Columbia University Irving Medical Center, New York, New York, USA

<sup>3</sup>New York-Presbyterian Morgan Stanley Children's Hospital, New York, New York, USA

<sup>4</sup>Division of Pediatric Surgery, Columbia University Irving Medical Center, New York, New York, USA

## Correspondence

Joseph A. Picoraro, 622 West 168th St, PH17-105G, New York, NY 10032, USA.  
Email: [jp3386@cumc.columbia.edu](mailto:jp3386@cumc.columbia.edu)

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## Abstract

Surgical intervention is often indicated in pediatric inflammatory bowel disease (IBD) for medically refractory disease or complications of severe disease. Specifically, surgical intervention via ileal diversion allows for fecal flow to be redirected away from diseased distal bowel and through an ileostomy. It is utilized in patients who have medically refractory colitis, severe perianal disease, or irreversible bowel damage. In patients with ulcerative colitis, it is primarily performed during a restorative proctocolectomy with ileal pouch anal anastomosis to protect the high-risk anastomoses. In the setting of Crohn's disease, ileal diversion reduces the exposure of diseased distal intestine to pro-inflammatory stool. During perioperative planning, it is crucial for the gastroenterologist to partner early with a multidisciplinary team including surgeons, nutritionists, wound ostomy care nurses, psychologists, and social workers. Patients should be assessed for malnutrition and should be optimized nutritionally with enteral or parenteral nutrition. As they are associated with increased risk of postoperative complications, corticosteroids should be significantly reduced or completely discontinued preoperatively. Though ileal diversion may reduce the complications associated with anastomosis, serious postoperative complications can include diversion colitis and high-output fistulae. This review aims to provide an overview of the role of ileal diversion in the treatment of pediatric IBD to pediatric gastroenterologists to inform their medical decision-making and discussions with patients and families.

## KEYWORDS

Crohn's disease, ileal diversion, pediatric inflammatory bowel disease, pediatric surgery, ulcerative colitis

## 1 | INTRODUCTION

Although medical therapies for inflammatory bowel disease (IBD) have transformed the care of children over the past 25 years, surgical intervention remains a cornerstone of management. In the era of anti-tumor necrosis factor (TNF) therapy, the rate of surgical intervention in children with Crohn's disease (CD) remains up to 26%<sup>1</sup> and in those with ulcerative colitis (UC), 14%–15%.<sup>2</sup> Pediatric IBD is a highly heterogeneous disease in which surgical intervention can be indicated for: (1) medically refractory disease, (2) irreversible bowel damage (e.g., fibrotic

strictures), and (3) complications of severe disease (e.g., complex perianal disease, fistulae). While resection of diseased bowel is typically the central surgical intervention, ileal diversion serves a critical role in the management of IBD. Ileal diversion, in its broadest and simplest terms, is the alteration of the fecal stream away from distal bowel and through a surgically constructed ileostomy. Ileal diversion can be used in combination with bowel resection, either as a long-term stoma or as a temporary measure to reduce the risk of surgical complications. It is also used in CD to provide symptomatic relief and improve disease control by diverting fecal flow away from diseased

segments of the distal intestine. To ensure the optimal use of ileal diversion in children, pediatric gastroenterologists should understand its indications, employ interdisciplinary decision-making, and determine appropriate referral to specialized IBD centers. This review will equip the pediatric gastroenterologist with a comprehensive overview of the role of ileal diversion and its pertinent perioperative considerations in patients with medically refractory IBD and its complications. Management principles in preoperative assessment, perioperative decision-making, and perioperative management are summarized for reference as a checklist (Figure 1).

## 2 | TECHNICAL OVERVIEW OF ILEAL DIVERSION

Ileal diversion is a procedure which surgically redirects the fecal stream through the abdominal wall via creation of an ostomy. This can be accomplished via creation of either an end ileostomy (EI) or a diverting

### What is Known

- Surgical intervention is a cornerstone treatment in pediatric inflammatory bowel disease (IBD) and ileal diversion is a critical option.
- Experience and training in the application of ileal diversion for pediatric IBD among pediatric gastroenterologists is limited.

### What is New

- The pediatric gastroenterologist should know basic principles of preoperative and postoperative management of pediatric patients with IBD that undergo ileal diversion, including indications.
- Multidisciplinary care of pediatric IBD with surgical colleagues optimizes appropriate use of ileal diversion.

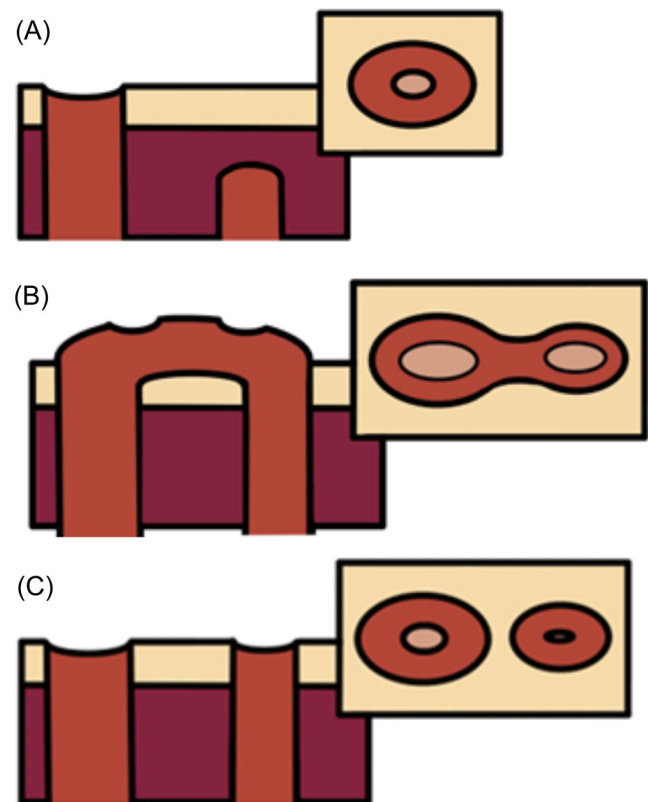
1. Preoperative Planning:
  - Determine indication for ileal diversion
  - Obtain early consultations from interdisciplinary IBD colleagues (surgery, nutrition, wound care, psychology/psychiatry)
  - Assess for malnutrition and incorporate care of a registered dietitian. Consider utilizing standardized nutritional score.
  - Optimize nutrition, preferably enterally
  - Prioritize weaning and discontinuing steroids preoperatively
2. Surgical Planning for the Gastroenterologist:
  - Discuss with surgeon about type of ileal diversion needed
  - In setting of RPC-IPAA, discuss the anticipated stages in creation of IPAA with the surgeon
  - In conjunction with surgery team, discuss surgical options, risks, and benefits with the patient and family
  - With surgery, psychology/psychiatry, and wound ostomy team, discuss expectations of having an "ostomy" with patient and family
3. Post Operative Management:
  - Consider restarting enteral nutrition within 24 hours. If unable, initiate parental nutrition.
  - Ensure initial frequent follow up with gastroenterology, surgery, and wound ostomy team regarding possible complications:
    - Stomal dermatitis
    - Local or systemic infection
    - Anastomotic leak
    - Bowel prolapse
    - Dehydration secondary to high output fistula
    - Bowel hemorrhage/Ischemia
  - Begin early planning for next surgical stage if applicable

**FIGURE 1** Perioperative considerations for ileal diversion in pediatric IBD. IBD, inflammatory bowel disease; IPAA, ileal pouch anal anastomosis; RPC, restorative proctocolectomy.

loop ileostomy (DLI). In creating an EI, the intestine is surgically divided, and the proximal ileum is redirected through a stoma. Meanwhile, the defunctionalized distal portion of the bowel is either converted into a mucous fistula or is partially resected with the possible retention of a long blind segment within the abdominal cavity called a Hartmann's pouch. This pouch consists of the remaining portion of the distal colon through the rectum. If there is significant risk of leak from the stapled-off end of the Hartmann's pouch due to compromised tissue, the creation of a mucous fistula is preferred.<sup>3</sup> EI allows for complete fecal emptying and is often utilized for patients who require a long-term or permanent stoma. When creating a DLI, the goal is to divert some or all of the fecal stream from the distal bowel. This procedure is accomplished by pulling a continuous segment of ileum through the abdominal wall with two lumens, proximal and distal bowel, pulled through the same incision (Figure 2). In this way, the two lumens both drain into one stoma bag. Resection or division of the intestine is not required for the creation of a DLI.<sup>4</sup> The internalization and reanastomosis of a DLI typically entails a smaller incision, and possibly more limited dissection, than reversal of an EI. Therefore, DLI is often utilized to provide temporary bowel rest in the setting of distal inflammation, obstruction, or new anastomosis.

### 3 | INDICATIONS FOR ILEAL DIVERSION

Ileal diversion is primarily utilized in children with (1) severe, medically refractory colitis, (2) complex and medically refractory perianal disease, or (3) in combination with bowel resection for irreversible bowel damage (e.g., fistulae, abscesses, or strictures).<sup>5</sup> For severe UC in which a colectomy is indicated, conventional surgical management is a three-stage or classic two-stage restorative proctocolectomy (RPC) with ileal pouch anal anastomosis (IPAA). During both of these procedures, a diversion ileostomy is created for fecal diversion to minimize the severity of surgical complications, such as anastomotic leak of the IPAA and pelvic sepsis, or to provide the opportunity for medical and nutritional optimization before IPAA surgery.<sup>6</sup> Three-stage RPC with IPAA is typically performed in patients with the most severe disease including risk factors of high-dose immunosuppression, malnutrition, and significant active inflammation. Urgent removal of disease burden is often the priority in these cases. During the three-stage operation, a colectomy is performed with preservation of the rectosigmoid stump and creation of an EI. Once the patient has had the opportunity to recover from the procedure, improve their nutritional status, and taper immunosuppression, the second stage is performed. This stage involves



**FIGURE 2** Comparison of surgical techniques: (A) End ileostomy with Hartmann's pouch, (B) diverting loop ileostomy, and (C) end ileostomy with mucous fistula.

completion proctectomy with preservation of anal sphincter muscles, creation of IPAA, and creation of a DLI. This is followed by subsequent ileostomy closure at the third operation.<sup>7</sup> The goal of performing the procedure over three stages is to mitigate the complications associated with pouch formation and anastomosis in the setting of active inflammation. A two-stage procedure is more commonly performed to avoid repeated pelvic surgery. The classic two-stage procedure consists of performing a total proctocolectomy and creation of an IPAA with a DLI during the first stage. Closure of the ostomy, the second stage of the procedure, occurs once the IPAA has healed 6–8 weeks later.<sup>6</sup>

Due to complications associated with DLIs such as dehydration and obstruction, a modified two-stage and one-stage approach have been employed to avoid diversion at the time of IPAA creation. In the modified two-stage procedure, a colectomy is performed with creation of EI, followed by completion proctectomy with IPAA creation and without a DLI. A one-stage approach includes colectomy, proctectomy, and IPAA creation at the same time without the creation of an ileostomy. While these approaches have been adopted in treatment of adult patients, their outcomes in pediatric patients remains controversial.<sup>8,9</sup>

In CD, RPC-IPAA is rarely performed due to high rates of recurrent disease and pouch failure.<sup>10</sup> Instead, ileal diversion can be utilized to facilitate a beneficial calming effect on the distal bowel via diversion of the pro-inflammatory fecal stream. Fecal diversion is indicated in cases of CD with (1) medically refractory distal bowel inflammation, (2) complex perianal disease, especially in the setting of rectal stenosis, concomitant colitis, or persistent anal sepsis, or (3) complications, such as fistulae, abscesses, and strictures, secondary to complex disease or previous surgical interventions.<sup>11</sup> Complications in CD are common, with a reported 48%–52% incidence rate of complications in patients with CD 5 years after diagnosis.<sup>12</sup> For patients with complex perianal disease, in particular, complications can be difficult to manage medically and a surgical consultation should be obtained early in their care. If both medical and local surgical therapies fail, fecal diversion is indicated. When the fecal stream is diverted via EI or DLI, the distal bowel is exposed to fewer pro-inflammatory factors and bacteria and is thus allowed to “cool down” while medical therapy is maximized.<sup>13</sup> There is significant evidence demonstrating that ileal diversion in these patients leads to significant decrease in disease burden and improvement in quality of life. However, several studies show that there are high rates of relapse after stomal reversal, suggesting that the role of ileal diversion in these patients is largely to provide temporary relief before more definitive surgical intervention.<sup>14,15</sup> Ileal diversion may also be indicated in cases of indeterminate colitis, in which the outcomes of IPAA creation are uncertain and may increase the risk of refractory disease, obstruction, pouchitis, abscess, and fistulae.<sup>7,10</sup> For these circumstances, particularly in children, diversion ileostomy can provide the inflamed colon a period of rest and allow for the natural history of the disease to unfold.<sup>10</sup> It also may have a more prominent role in very-early onset IBD in which immune dysregulation is more common and there is higher variability of disease trajectory.<sup>10,16</sup> This offers symptom relief, facilitates time-sensitive childhood growth and development, and presents an opportunity to better understand the child's underlying disease process.

#### 4 | PREOPERATIVE PLANNING

Overall, surgical intervention in pediatric IBD should be evaluated within the array of treatment options and utilized in a targeted manner to address the specific needs of the child and family. Often, children become surgical candidates when medical approaches have been unsuccessful or due to complications of disease as previously discussed. By this time, issues such as malnutrition, corticosteroid exposure, and infection risk have been compounded. It is imperative for the

pediatric gastroenterologist to partner with their surgical and nutrition colleagues to optimize these modifiable factors and mitigate postoperative complications such as leaks, strictures, and fistulae.

Malnutrition and growth failure are prevalent in all pediatric IBD phenotypes. Malnutrition contributes to poor wound healing, which heightens the risk of anastomotic leaks. Preoperative malnutrition has also been associated with an increased risk of surgical site infection in children with IBD.<sup>17</sup> Despite its clinical relevance, assessing the nutritional status of pediatric patients with IBD can be difficult. The malnutrition observed in these patients arises from a multifaceted pathophysiologic process including poor oral intake, vomiting and diarrhea, malabsorption due to active disease or prior bowel resection, protein loss in the setting of inflammation and steroid therapy, as well as heightened metabolic demand due to underlying disease.<sup>18</sup> There is no current gold standard for assessment of malnutrition in patients with IBD, though many anthropomorphic and biochemical markers have been studied as proxies. Anthropometric parameters like body mass index, mean upper arm circumference, skinfold measurement, and body fat mass have been identified as indicators of malnutrition in patients with active IBD.<sup>18–20</sup> However, other studies show that these measures are not reliable predictors of macro-nutrient or cellular malnutrition, suggesting that anthropomorphic markers of malnutrition should not be interpreted in isolation.<sup>21,22</sup>

Common biochemical indices of malnutrition include complete blood count, serum albumin, pre-albumin, total protein, transferrin, cholesterol, triglycerides, iron, total iron-binding capacity, ferritin, and vitamin levels.<sup>18,19,21–25</sup> Hypoalbuminemia, specifically, has been extensively evaluated for reliability as a marker of malnutrition. Though studies are conflicting, the majority suggest that it is an unreliable indicator of malnutrition in IBD.<sup>17,18,22,24</sup> This unreliability is due to its multifactorial nature; hypoalbuminemia is not only a marker of malnutrition, but also a complication of protein losing enteropathy and a negative acute phase reactant of inflammation. As such, patients with IBD who experience significant inflammation may suffer from hypoalbuminemia, regardless of their nutritional status.<sup>24</sup>

Nevertheless, hypoalbuminemia is a risk factor for surgical morbidity and mortality in patients with IBD. Preoperative hypoalbuminemia in patients with IBD undergoing surgical intervention has been associated with an increased risk for mortality, infectious complications, and nonseptic complications.<sup>23</sup> While the etiology of hypoalbuminemia is multifactorial and should not be interpreted alone as an indicator of malnutrition in patients with active IBD, it is an important component of a preoperative assessment of surgical risk.

Given the complex nature of nutritional assessment in IBD, malnutrition should not be assessed based on single parameter measurements. Instead, providers should consider the use of malnutrition scores, which incorporate both anthropomorphic and biochemical indices, to classify malnutrition more reliably. Nutritional screening scores, such as the nutritional risk screening (NRS) score, have long been utilized to identify patients who may benefit from nutritional support during hospitalization, and the recently developed perioperative nutrition score (PONS) has specifically shown promise as a preoperative nutrition screen in a pediatric IBD population.<sup>17,26,27</sup> We propose utilizing comprehensive screening tools such as these to assess perioperative malnutrition and to guide nutritional optimization to minimize postoperative risk.

Optimizing preoperative nutrition can lead to improved postsurgical outcomes for malnourished patients. A recent American Gastroenterological Association clinical practice update recommends nutrition support via exclusive enteral nutrition (EEN) because it improves mucosal healing, reduces infection risk, and is cost-effective.<sup>4,28</sup> Emerging data in children with CD indicate that preoperative EEN is associated with decreased inflammation in resection pathology and reduced postoperative hospital stays.<sup>29</sup> However, complications and symptoms of IBD like strictures, obstruction, abscesses, and nausea can make adequate enteral caloric intake infeasible. In such cases, parenteral nutrition (PN) may be necessary, although its impact on postoperative outcomes is unclear.<sup>4,30,31</sup> Despite this uncertainty, PN is frequently utilized in practice, as evidenced by a study in which 70%–80% of pediatric patients with IBD received PN within 24 h of urgent surgical intervention.<sup>32</sup> Careful consideration of nutritional strategies is essential for optimizing surgical outcomes for malnourished patients with IBD.

Children with medically refractory IBD are also at risk of high dose and prolonged corticosteroid exposure. Preoperative use of corticosteroids in patients undergoing colorectal surgery dose-dependently increases the risk of postoperative infectious complications.<sup>33–36</sup> Although this may reflect the high disease burden in patients with IBD, corticosteroids may also contribute to delayed wound healing by inducing immune dysregulation and suppressing the hypothalamic-pituitary-adrenal axis.<sup>33,34</sup> Corticosteroid-sparing strategies, including the use of anti-TNFs, are therefore paramount in optimizing children for surgery. The landmark PUCINI study demonstrated that the use of anti-TNF therapy within 12 weeks of surgery did not increase risk for postoperative infection, regardless of serum drug level and disease activity.<sup>37</sup>

Lastly, preoperative abscesses independently elevate the risk of intraoperative and postoperative complications, including intra-abdominal septic complications, such as abscesses, fistulae, and anastomotic

leaks.<sup>38</sup> Percutaneous drainage and antibiotic therapy should be considered to stabilize the patient, decrease inflammation, and optimize surgical readiness.

Due to the elevated risk of anastomotic complications, ileal diversion frequently emerges as an alternative to primary resection and anastomosis in children with malnutrition, high corticosteroid exposure, and increased infectious risk. Preoperative management thus relies upon interdisciplinary decision-making that minimizes disease burden and risk factors and, in light of unmodifiable preoperative and intraoperative factors, considers the role of ileal diversion.

Early, multidisciplinary discussions within a biopsychosocial framework are an essential component of preoperative planning.<sup>39</sup> These conversations should involve surgeons, nutritionists, psychologists, and social workers, each of whom has a defined role. Consideration should be made for referral to a pediatric IBD center that has an integrated multidisciplinary team for any child with IBD who may require surgical intervention. Surgeons play a crucial role in assessing patient candidacy for surgical intervention. They work closely with the gastroenterologists and nutritionists who assess and help manage malnutrition perioperatively.<sup>40</sup> In addition to providing care for the stoma, wound and ostomy care nurses can provide preoperative counseling to patients and their families about having a stoma, which can be very difficult to cope with during the developmental stages of pre-adolescent and adolescent years.<sup>41</sup> Psychologists also offer invaluable support as they help children and families navigate the everyday challenges of chronic illness as well as process the physical and emotional repercussions of surgical intervention.<sup>39,42,43</sup> Finally, social workers help to alleviate stress related to the many follow-up visits, medical supplies, and insurance claims facing patients and their families. Support from each of these team members is particularly valuable given the significant impact of psychosocial stressors on the clinical manifestations of IBD.<sup>43</sup>

## 5 | COMPLICATIONS OF DIVERTED BOWEL

While ileal diversion avoids complications associated with anastomosis, its inherent risks should be carefully considered by the medical team before surgery. Diversion proctocolitis is mucosal inflammation of the bypassed colon that develops after diversion of the fecal stream. In practice, almost all diverted mucosa shows some amount of inflammation, though it can be difficult to distinguish between diversion colitis and IBD.<sup>44</sup> Diversion colitis in patients with IBD is histologically different than that in patients without IBD, with the severity of inflammation appearing to be, at least in part, reflective of IBD activity.<sup>45</sup> Dermatologic

complications such as peristomal dermatitis are also common complications of ileostomy. More significant complications such as nonfunctioning stoma, bowel prolapse, parastomal hernia or stomal retraction occur less frequently but may require surgical attention. High-output fistulae are a severe complication of intestinal diversion which confer significant risk for dehydration, electrolyte imbalance, and protein loss. Finally, bowel ischemia or hemorrhage occur rarely.<sup>46</sup>

Risk also lies in the eventual reversal of the stoma. If stoma reversal is possible given the patient's clinical status, this procedure confers the risks associated with most gastrointestinal surgeries, including anastomotic leak, postoperative ileus, bowel obstruction, enterocutaneous fistula, and infection. Aside from surgical complications, prolonged intestinal diversion can lead to permanent impairment of the distal, temporarily defunctionalized bowel, including poor contractility and villous atrophy.<sup>45,47</sup>

## 6 | CONCLUSION

Ileal diversion addresses severe, medically refractory, or complicated IBD in children. It provides the opportunity to reduce steroid use, improve growth and observe the natural history of the disease in a more controlled manner. It may also enhance quality of life. However, to mitigate the potential surgical complications and anticipated life experience of a stoma, preoperative planning should be done by an interdisciplinary team to identify appropriate indications for diversion and consider surgical risk factors. Crucially, risk factors such as malnutrition, corticosteroid exposure, and infectious risk should be optimized as much as possible before surgery. When appropriately utilized, ileal diversion is a valuable therapeutic option in the management of pediatric IBD.

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## CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

## ORCID

Joseph A. Picoraro  <https://orcid.org/0000-0002-1857-5000>

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