Complications of the Pelvic Pouch

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Key Concepts

- Pelvic sepsis is the most common cause of pouch loss.
- Patients with IPAA dysfunction should undergo a structured assessment.
- Pouch dysfunction is often inappropriately ascribed to a diagnosis of Crohn's disease.
- Fecal diversion may palliate disabling symptoms from pouch dysfunction.
- Pouchitis is the most common complication of IPAA.

Introduction

The ileoanal pouch reservoir (IPAA) made stoma-free living a reality in patients, who desire to maintain bowel continuity, requiring the removal of the colorectum [1, 2]. This operation was specifically created with a vision to achieve a higher quality of life after proctocolectomy, providing the patient with an alternative to a permanent lifelong stoma and restoring the natural route of defecation. Contemporary improvements have enhanced the operation since its popularization in the 1980s, but the goals of surgery remain the same: to cure disease while providing the highest possible quality of life (QOL) for the patient.

Failure of the IPAA is an uncommon but devastating situation for patients undergoing restorative proctocolectomy [3]. Patients with IPAA dysfunction, in whom local corrective measures fail, have traditionally been managed with permanent fecal diversion, with or without excision of the failed pelvic pouch [4]. However, advancements in the understanding of pouch failure have opened the door for surgical revision; selected patients who are decidedly motivated to avoid permanent conventional ileostomy may be considered for surgical pouch salvage with a reasonable expectation of good results [5–8]. Critical to the success of pouch revision is the understanding of why pouches fail, which is an evolving topic, as the treatment of pouch complications varies greatly depending on etiology. The management of pouch-related complications, including pouch salvage surgery, is challenging and is best approached in a multidisciplinary, patient-centered fashion with input from both the patient and experienced IPAA clinicians for best results.

Risk Factors for Pouch Dysfunction

The success of an IPAA procedure and its long-term functional outcomes are very much dependent upon adequate healing and maintenance of integrity of the many staple or suture lines required. Anastomotic disruption, with the resultant development of peri-pouch sepsis, is a dreaded complication of staple or suture line failure and typically has marked detrimental effects on long-term functional outcomes [9, 10]. Pelvic sepsis is reported to occur in up to 25% of IPAA patients and is due primarily to the disruption of the pouch-anal anastomosis or, less frequently, the staple line at the tip of the J-pouch [11]. Thirty percent of these patients will experience pouch failure, making it the most common cause of pouch loss [1, 12].

Much emphasis has been placed on avoidance of anastomotic complications by identification of adverse risk factors. Strategies of avoiding or delaying pouch creation in patients taking higher doses of corticosteroid and/or biologic therapy and performing this restorative procedure using a staged approach have been recommended for this reason [13]. Despite patient optimization and technical perfection, anastomotic leak is a known consequence of IPAA surgery, and strategies for management are necessary. The ramifications

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of a leak are great in terms of long-term function; equally concerning and burdensome are the associated financial strains on the patient and healthcare system as these complications often lead to a delay in ileostomy closure and a need for additional radiographic intervention, pouch revision or excision, and prolonged hospital convalescence [14, 15].

Pouch failure due to structural non-septic complications or functional issues is a less studied cause of IPAA dysfunction. One source of these complications may be due to technical missteps performed at the index pouch surgery. Examples include inappropriate rotation or twisting of the small bowel mesentery as it runs into the pelvis (180° or 360° rotation) that causes obstruction or ischemia or an elongated rectal cuff (or S-pouch outlet) causing outlet obstruction and pouch emptying issues. An emphasis on proper technique of IPAA surgery and a vigilance during these challenging cases can prevent these complications from occurring or identify them early so they may be corrected.

Finally, patient selection is crucial to achieving success after pouch surgery. Those patients who have pelvic floor or anal sphincter compromise may not fare well in terms of pouch function and quality of life, and one should be realistic when discussing outcomes when these issues are present. Additionally, a diagnosis of Crohn's disease should generate a thoughtful, individualized assessment of the risk prior to pouch surgery.

Approach to the Patient with a Dysfunctional Pouch

Initial Evaluation

First and foremost, patients referred with a diagnosis of IPAA dysfunction should undergo a comprehensive and standardized evaluation, understanding that the previous diagnosis may be incorrect. Commonly, a patient with non-specific pouch issues is labelled as having chronic pouchitis or Crohn's disease and undergoes a long-term treatment on this basis, without significant improvement in symptoms. Other causes of pouch dysfunction such as chronic pelvic sepsis may be easily missed.

A complete history should be obtained including a full review of the patient's symptoms, treatments that have been attempted prior to the present evaluation, and response to each treatment. Operative reports should be obtained and reviewed, with specifics of surgery and convalescence noted. Any indication of technical difficulty must be thoroughly explored, as a technical complication of the initial pouch surgery may be mistaken for pouchitis. One should pay particular attention to the condition of the patient at the time of pouch creation and the use of covering ileostomy, as large doses of immunosuppression negatively affect pouch healing; anastomotic complications may result in occult sinus tracts or chronic anastomotic leaks with symptoms mimicking pouchitis.

It is not unusual for an empiric diagnosis of "Crohn's disease" or "chronic pouchitis" to be given to patients with symptoms of pouch dysfunction, with limited or no support from endoscopy or other imaging studies. One drawback of this approach is that patients may be given a presumptive diagnosis of Crohn's disease that is never confirmed and medical therapy considered unsuccessful. Therefore, it is important to establish the etiology of compromised pouch function when it begins, even if the symptoms have been longstanding. The correct diagnosis accounting for pouch dysfunction is crucial as treatment options are at times vastly different for each complication [16].

Endoscopic evaluation is performed to look for key identifiers of pouch dysfunction. One must approach pouchoscopy in a standardized fashion, so as to carefully examine the rectal cuff, pouch body, and afferent limb to identify both mucosal changes as well as clues to structural abnormalities such as an elongated rectal cuff, a strictured or twisted afferent limb, or a prolapsing anterior body wall. Pouch endoscopy is very helpful when done in conjunction with an anoperineal exam under anesthesia to identify fistulae, abscesses, or other anal pathology not as easily seen in the endoscopy suite. Contrast enemas and pelvic MRI may reveal or rule out anastomotic complications, fistulae, sinuses, or chronic leaks that may be the source of symptoms.

Next, the surgeon must assess the patient's health status and quality of life during the initial patient encounter, even if the etiology of pouch dysfunction is still unclear. Patients are often referred to the surgeon after years of medical treatments that have left the patient malnourished, decompensated, and mentally exhausted. These individuals may benefit from surgical intervention such as fecal diversion sooner rather than later.

Finally, it is important to have an honest and straightforward discussion with the patient regarding expectations. It must be emphasized that surgical outcomes depend on many factors, especially the etiology of pouch failure. Expectations must be discussed and agreed upon prior to embarking on surgical correction.

Multidisciplinary Approach to Diagnosis

When a patient presents with IPAA dysfunction and the etiology of failure is in question, a multidisciplinary approach is in order. After preoperative evaluation with history, physical, and radiographic testing as outlined above, an evaluation with an anoperineal exam under anesthesia and pouchoscopy

is performed as a team including a colorectal surgeon and gastroenterologist. The anoperineum, pouch-anal anastomosis, pouch body, and afferent limb (complete to the ileostomy closure site) may be examined with members of both specialties in the operating room, enabling both perspectives and respective expertise to be utilized. Any clinical signs of pouchitis or any other IPAA complications are noted (anastomotic sinus or fistula, stricture, pouch prolapse, Crohn's disease, etc.), many of which may cause similar symptoms. Biopsies are obtained for pathologic review. At the completion of the exam, the findings are discussed with the patient and family member, and a patient-centered treatment strategy begins to develop. This multidisciplinary team approach is ideal for the patient as he/she is often presented an immediate plan for treatment, with opportunity for discussion with members of both specialties. The strategy can always be tailored at a later time as pathology results and/or recommendations from a multidisciplinary case conference are made.

The Case for the "Thoughtful" Ileostomy

Fecal diversion is an effective way of alleviating symptoms in patients suffering from a failing IPAA and buying time while investigation continues and decisions are made regarding pouch salvage. Any patient suffering significant health consequences or with poor quality of life is a candidate for ileostomy during any part of the pouch dysfunction evaluation. This approach may provide symptomatic relief as mucosal inflammation is lessened by diversion of the fecal stream and anoperineal excoriation as a result of frequency of bowel motions that may be controlled. The pouch is left in place, allowing for relief of symptoms without committing to a major pelvic operation. This approach can also be useful in patients with little chance of pouch salvage who are not initially accepting of a permanent stoma. A loop ileostomy without "burning the bridge" may convince the patient that fecal diversion will dramatically improve their life as compared to a dysfunctional IPAA and ease the psychological transition to ultimate pouch excision [17].

Loop ileostomy provides many benefits. First, it allows for relief of symptoms related to a dysfunctional pouch in a manner that can be temporary and somewhat easily reversed if the patient is not pleased. In many cases, it may be completed with a laparoscopic approach, even if open IPAA had previously been performed; this will usually shorten convalescence and minimize adhesion formation in case repeat laparotomy for pouch revision or excision is desired. Second, patients are able to experience or have a reminder of what life is like with an ileostomy and may choose to keep the ileostomy on a more permanent basis.

Exploration at the time of ileostomy allows for a thorough exam of the abdomen and small bowel to identify any pathology missed on prior imaging that may be the source of the patient's symptoms. Possible sources are mesenteric twists, afferent limb adhesions, abdominal wall or pelvic mesh adherent to the ileal pouch, or large ovarian cysts thought to be part of the ileal pouch on preoperative imaging. In each of these situations, the pathology can be operatively addressed.

When creating an ileostomy in this setting, it is important to consider the next potential surgical steps in the patient's future. The site of the ileostomy should be made with the most dependent portion of the small bowel and at least 15 cm proximal to the pouch, such that this enterotomy could be used for a new pouch-anal anastomosis in those who may be candidates for pouch revision or recreation in the future.

Etiology and Management of Pouch Complications

Structural Complications of the Pouch

Afferent Limb (AF) Complications

Complications involving the pre-pouch ileum, or the afferent limb (AL) of the pouch, have historically been termed afferent limb syndrome (ALS). ALS is becoming a more recognized and diverse group of findings after pouch surgery in which patients present with symptoms of bowel obstruction, abdominal pain and cramping, and dyschezia but have an otherwise normal pouch body and outlet on endoscopy or distal contrast study. The causes of ALS were initially thought to include only a displacement of the pre-pouch ileum posterior to the pouch causing obstructive symptoms; but a more contemporary understanding of ALS has shown a growing number of potential abnormalities of this portion of the bowel, many of which can be difficult to identify on initial evaluation. For example, a fibrotic stricture causing ALS may be easily seen during pouchoscopy, but angulation of the afferent limb due to adhesions or an inappropriate rotation of the small bowel mesentery is sometimes only identified at laparotomy with findings of partial or complete obstruction [18].

There are several important discussion points regarding ALS that should be highlighted. First, although CD occurring in the pre-pouch ileum can cause fibrostenotic or inflammatory changes of the AL, many of these changes may NOT be due to CD. Chronic nonsteroidal anti-inflammatory medications and Crohn's disease-like conditions (CDLC) can cause similar radiographic and endoscopic findings; clinicians should be careful not to label a patient as having CD of the pouch unless there is reasonable certainty in the diagnosis (Fig. 50.1). Many patients given a label of "CD of the pouch" are branded with this negative stigma and are only offered pouch excision, when in actuality, they may be candidates for salvage procedures.

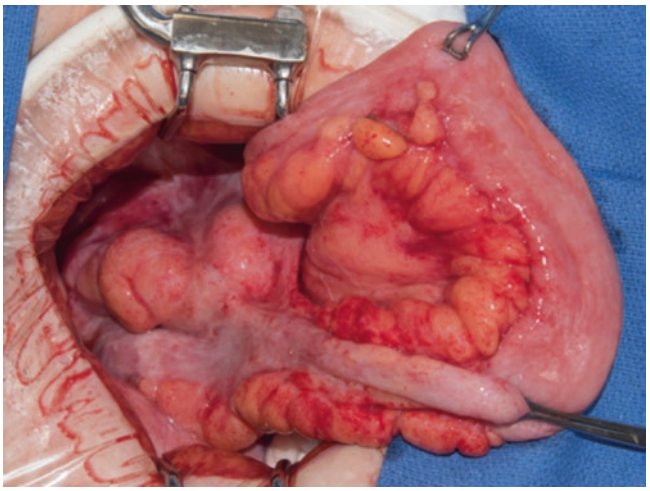

Fig. 50.1 Chronic inflammation of the afferent limb of the ileal pouch forming a strictured segment and causing obstructive symptoms

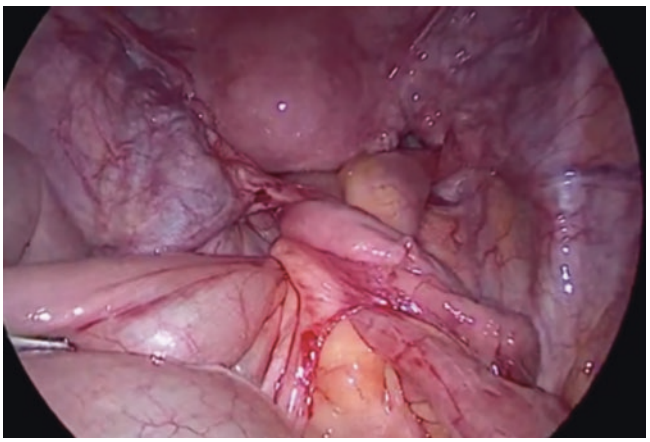

Fig. 50.2 Adhesive bands causing volvulus of the afferent limb of the J-pouch. Note the dilated upstream small bowel (left) due to obstruction of the distal small bowel. A laparoscopic lysis of adhesions with reduction of volvulus was successfully performed

Dysfunction of the AL may also be caused by factors external to the bowel wall (Fig. 50.2). Volvulus or trapping of a redundant AL underneath the small bowel mesentery may result in acute ischemia of the limb or a more chronic intermittent obstructive pattern; patients may be intolerant of large meals, but the pouch is typically normal on radiographic imaging and/or pouchoscopy [19].

Another variant of this is a 180° or 360° rotation of the small bowel mesentery at the time of pouch creation. This incorrect rotation can cause external compression of the mesenteric edge of the AL, often in an intermittent fashion, which makes diagnosis difficult unless the patient is having overt obstructive symptoms. At endoscopy, insufflation of the AL may overcome the external compression and a patent; otherwise normal appearance is suggested. Acute angulation of the AL without mesenteric rotation, "accordioning" of a floppy or redundant pouch, or a rotation of the anterior wall of the pouch can cause obstructive symptoms. Pouch-pexy with creation of a temporary diverting ileostomy to pull the pouch upright "on stretch" helps to secure the pouch in a proper orientation [20–22].

Many patients who suffer from ALS are candidates for endoscopic and/or surgical correction and will not require pouch excision. AL strictures may be assessed for endoscopic balloon dilation, although many strictures persist and require surgical correction. Such strictures are amenable to either surgical resection with primary anastomosis of the pre-pouch ileum, stricture plasty, or a bypass to the pouch inlet. Those with a diagnosis of CD may benefit from ongoing, postoperative medical therapy to prevent inflammatory recurrence. In this way, pre-pouch strictures are dealt with similarly to fibrostenotic CD in other locations [23, 24].

Issues of the Pouch Body

Pouch-Anal Anastomotic (PAA) Defect

One of the most common and devastating complications of IPAA surgery is a leak of the pouch-anal anastomosis. When this occurs in the acute setting (immediate postoperative period), patients may present with fever, leukocytosis, pelvic pain, or other signs/symptoms of sepsis (chills/night sweats), often prompting CT imaging which reveals a pelvic abscess and/or staple line leak. Soluble contrast enema of the pouch, pelvic magnetic resonance imaging (MRI), or examination under anesthesia (EUA) are also helpful to better characterize an anastomotic leak if one is suspected in the early postoperative period [25]. The presentation may be more indolent in some cases, with patients exhibiting indirect symptoms of pelvic sepsis, such as prolonged ileus or urinary retention. Upper pelvic or abdominal abscesses may be percutaneously drained with a CT or ultrasound-guided percutaneous technique; surgical drainage may be required for those not amenable to image-guided measures.

For lower pelvic abscesses or collections obviously associated with an anastomotic disruption, EUA with gentle anoscopy and placement of a flexible mushroom drain through the anastomotic defect is strongly recommended. Care must be taken to avoid drainage approaches that would lead to complex fistula formation; the transanal approach is preferred over percutaneous measures for lower pelvic abscesses for this reason. In patients exhibiting peritonitis or hemodynamic instability, exploration in the operating room with pelvic washout and wide drainage is indicated. This approach, when necessary, is associated with poor pouch outcomes; the rate of pouch excision exceeds 40% with an associated low likelihood of ileostomy reversal [26].

Swift recognition and treatment of anastomotic disruption are paramount to preserve optimal IPAA function and avoid the known long-term sequelae of pelvic sepsis. In cases

where local sepsis is quickly controlled, pouch function is typically preserved, whereas a delay in management risks chronic inflammation with peri-pouch fibrosis and poor compliance of the pouch [11].

When an anastomotic disruption does not heal with the conservative measures described above, patients may develop pouch sinuses, fistulae, strictures, or a number of other pouch-related complications that require further management. Anoperineal fistulae may present as chronic pelvic or anoperineal sepsis which usually require source control with mushroom or draining setons. This presentation is often very similar to CD, but there are subtle and critically important differences. In a patient with AL, sepsis originates from the pouch-anal anastomosis, with the majority of the fibrosis or chronic inflammation at the anastomosis itself; the distal pouch and anal canal are otherwise soft and supple. The anoperineum may be excoriated similar to CD, but without a bluish hue, and there is lack of other CD findings such as waxy skin tags. There is typically (but not always) more fixed fibrosis of the distal pelvis in patients with complications truly attributable to CD.

These subtle clues may help distinguish between CD and sepsis due to AL; but in many cases, these two conditions are indistinguishable, leading to management conundrums. Patients are best evaluated and managed using a multidisciplinary approach involving experienced pouch surgeons and gastroenterologists. Studies have shown that up the three quarters of IPAA patients diagnosed as CD of the pouch were reclassified as having AL after secondary evaluation at a specialty IPAA center and underwent pouch salvage with good results.

Less commonly described, but equally challenging to manage, is the anastomotic sinus-a blind-ending track resulting from an anastomotic dehiscence (Fig. 50.3). It typically presents months to years after IPAA surgery that was complicated by an anastomotic leak, even if the initial leak was not appreciated or documented. It is reported to occur in 2.8–8% of patients undergoing a pelvic pouch procedure, may threaten the integrity of the pouch-anal anastomosis, and is an important predictor of pouch failure [14, 27]. The most common location of a pouch sinus is the posterior portion of the pouch-anal anastomosis and is often associated with presacral inflammation or fibrosis. Pouch sinuses may present as asymptomatic findings on imaging obtained for other indications (e.g., routine evaluation prior to stoma closure) or may exhibit a wide range of symptoms including pelvic or tailbone pain, fecal urgency, night fevers, and other symptoms of pouch dysfunction or failure.

Asymptomatic sinuses require differing management strategies depending on the circumstances of the patient. Sinuses detected in patients without symptoms may be left alone without any intervention, assuming they are not diverted. Those discovered during preoperative evaluation prior to closure of a covering ileostomy will likely heal with a conservative approach; a delay in ileostomy closure of 3–6 months and a repeat pouchogram prior to closure are advised [14, 27, 28].

Sinuses that persist despite these measures can be very difficult to manage for the surgeon and both morbid and frustrating for the patient. Contrast studies of the pouch, pelvic MRI, and EUA are helpful for further delineation of the tract [29]. Treatment begins with periodic incision and drainage of the sinus with healing by secondary intention over a course of usually 6–9 months. Some sinuses may be amenable to endoscopic debridement with sinusotomy (needle-knife therapy) with or without fecal diversion [27, 30]. Revisional or redo pouch surgery via a transabdominal/transanal approach may be considered for refractory cases.

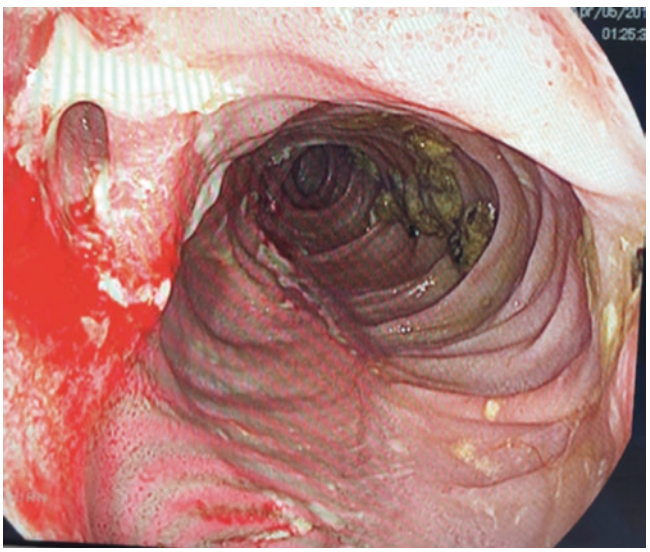

Fig. 50.3 A sinus tract of the pouch-anal anastomosis

"Tip of J" Pouch Leak

The tip of the J-pouch is the anatomic end of the small bowel and may be at risk for ischemic injury or staple line dehiscence, which can lead to sinus formation or persistent staple link leak. If these occur, patients often present with an abscess in the upper pelvis adjacent to the tip of J or with a persistently draining lower midline abdominal wound associated with a low-volume enterocutaneous fistula. These leaks typically do not heal without intervention; although there are reports of endoscopic repair, most will require surgical revision and (much less commonly) full recreation of the pelvic pouch [31, 32].

Incomplete healing of the anterior (or common channel) suture or staple line is a less commonly noted complication of the pelvic pouch. These present as a pelvic abscess, often in the mid-pelvis, that persists despite drainage and without an obvious defect at the pouch-anal anastomosis. Imaging studies may show a connection to the midportion of the

pouch body or even, more rarely, a fistulous connection to other staple lines of the pouch including the tip of J. A full surgical repair of the pouch is usually required, as these also typically do not resolve without intervention. One must be prepared to fully revise or redo the pouch body when embarking on corrective surgery for this complication.

Failure of Pouch "Scaffolding"

The anterior portion of the pouch will usually have more redundancy than the posterior portion, as the shorter, "limiting" axis is typically the posterior aspect of the pouch/small bowel mesentery. Because of this, the anterior portion of the pouch is "floppier" and may become tethered to the presacral fascia, which can limit capacity, or produce internal anterior wall prolapse and cause outlet obstruction. The pouch may also exhibit full thickness external prolapse and protrude from the anus similar to that observed in rectal prolapse [33, 34].

Patients may present with a wide range of symptoms including inability to fill or empty the pouch, pelvic pain, and obstructive symptoms. These patients are often diagnosed with chronic pouchitis. A comprehensive evaluation as described above may identify the abnormal configuration, but sometimes the clues are limited, and this is found only on abdominal exploration. Pouch-pexy techniques (sometimes with an "ileostomy on tension" as described above) is helpful to maintain the appropriate pouch "scaffold." Reports describing mesh or other matrix fixation of the pouch have been published; larger studies with longer follow-up are needed to assess the success and safety of this corrective approach [33, 35].

180°/360° Mesenteric Rotation

Incorrect orientation of the pouch and small bowel mesentery as it descends into the pelvis may cause external compression of the AL or compression/limitation of the volume of the pouch body, resulting in either frequent pouch emptying and/or obstructive symptoms with difficult filling of the pouch. This complication is thought to occur at the time of pouch-anal anastomotic creation, if the pouch inadvertently is allowed to rotate posteriorly (180° defect) or with complete revolution (360°). Patients may be at increased risk for pouch ischemia if the mesenteric blood flow is compromised and may have undue tension on the posterior portion of the anastomosis, a location already prone to anastomotic dehiscence [36, 37].

This complication, as is the case with most others, is better prevented than remediated; appropriate orientation of the mesentery should be assured at the time of anastomotic creation, especially during a laparoscopic or robotic approach, since the abdominal portion of the mesentery is often out of view. To correct this complication, a complete detachment and recreation of the pouch-anal anastomosis is required.

Efferent Limb (EL) Problems

Complications involving the pouch outlet, or efferent limb complications (EL), are likely to present as inability or difficulty emptying the pelvic pouch, with obstructive symptoms, straining with bowel motions, and feelings of incomplete emptying.

Efferent Stricture

Pouch-anal anastomotic strictures usually develop within the first 9 months after surgery. They are more commonly noted after mucosectomy with creation of hand-sewn pouch-anal anastomosis but can be seen in up to 17% of all pouch patients [38, 39]. Development of stricture after IPAA is similar among the commonly used stapler sizes (28/29 mm vs 31/33 mm) used to create the pouch-anal anastomosis [40]. Soft, weblike strictures are often seen after a diverted stapled anastomosis and are amenable to gentle digital dilation. They generally do not recur after restoration of intestinal continuity or are responsive to daily self-dilation [41]. Long, fibrotic strictures commonly result from perioperative pelvic sepsis or pouch ischemia. These generally do not respond to repetitive dilation in the long term, and surgical options are often considered and include (transanal or transabdominal) pouch advancement or pouch revision. Pouch excision with permanent ileostomy may be considered if patient factors are not favorable or if the patient desires this option [42].

Elongated S-Pouch Outlet/Elongated Rectal Cuff (Pouch-Rectal Anastomosis)

At times, efferent limb issues may be caused by technical errors made at the time of pouch creation. These are most commonly in the form of an S-pouch outlet that is made too long or a rectal cuff that is left too long (pouch-rectal anastomosis; Fig. 50.4). Again, meticulous surgical technique dur-

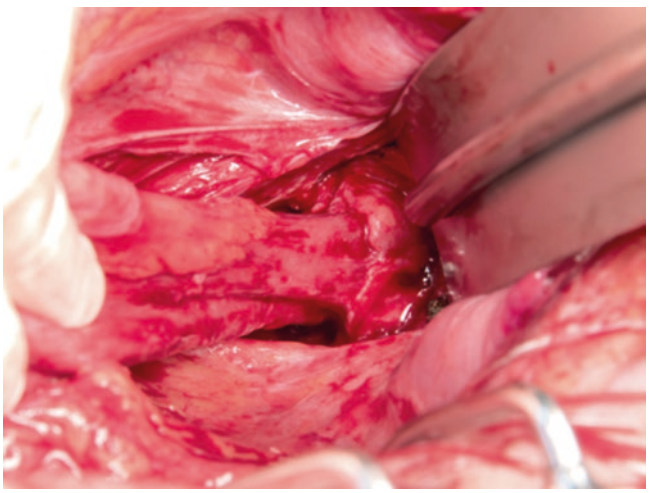

Fig. 50.4 Efferent limb syndrome caused by an elongated rectal cuff (pouch-rectal anastomosis)

ing the de novo pouch operation is critical and can help to avoid these difficult to manage complications. When they do occur, nonoperative maneuvers such as transanal intubation to evacuate the pouch may be offered for amelioration of symptoms but are not likely acceptable as a long-term option to patients and can cause tissue trauma with repetitive use over the years.

Transabdominal/transanal pouch revision is typically employed to shorten the elongated segment. In the setting of an elongated rectal cuff, the surgeon may have the option to restaple a very long cuff with a 30 mm linear stapler and maintain the anal transition zone. If so, a redo double-stapled pouch-anal anastomosis is achievable, and a transanal approach (and mucosectomy) avoided [43].

A situation becoming more commonly seen (and debated) is creation of a pelvic pouch after a proctectomy that preserves the mesorectum and leaves it in place. Although some surgeons argue that this serve as a space filler so that the pouch does not rotate and is "protective" of the retroperitoneal or presacral structures, in actual practice, the residual mesorectum may act as a "collar" around the distal pouch and can contribute to evacuation issues. The correction of this requires transabdominal completion mesorectal excision with a recreation of a new pouch-anal anastomosis.

Inflammatory Complications of the Pouch

Pouchitis

Pouchitis is the most common long-term complication of IPAA surgery. Despite the absence of a surgical "cure" for refractory or chronic pouchitis, the surgeon's role in the multidisciplinary management of pouchitis is crucial to aid with diagnosis and offer options for symptom management.

Although the etiology and pathogenesis of pouchitis are not entirely clear, pouch creation may provide an "inflammation-prone" environment as the distal ileum is converted to storage reservoir. About half of patients who undergo IPAA surgery for UC will develop at least one episode of pouchitis in their lifetime. Approximately 40% of patients experience a single episode (increased frequency of loose bowel movements, tenesmus, rectal bleeding, lower abdominal cramping, and malaise) and respond to a 2–4-week course of oral antibiotics. The remaining 60% will follow a relapsing course; half of these patients will suffer from refractory pouchitis and require treatment with second-line therapies such as chronic antibiotics, steroids, or biologic agents.

A small minority of patients with treatment-resistant pouchitis does not find relief with medical therapy and may desire surgical options for treatment and alleviation of symptoms. These patients should undergo a comprehensive pouch failure evaluation to rule out diseases with similar presentations as outlines earlier in the chapter and be offered the appropriate treatment options depending on the most likely etiology of failure. If refractory pouchitis is suspected, patients may be considered for fecal diversion as a means of alleviating symptoms from mucosal inflammation and perianal excoriation from frequent bowel motions. These patients may also be considered for pouch excision or a loop ileostomy with the pouch left in situ, depending on individualized risk factors for either pouch neoplasia (pouch in situ) and wound healing (pouch excision) [44]. One should be hesitant to offer a redo pouch surgery in this setting without a clear reason to hope for a different outcome with a second pouch.

Pouchitis may be divided into three categories that consider the presumed etiological and pathogenic factors of the condition. Classic pouchitis occurs from dysbiosis of commensal bacteria or infection from bacterial, viral, or fungal pathogens. Patients in this category present with homogeneous, diffuse inflammation of the pouch with or without ulcers. Immune-mediated pouchitis is typically found in the setting of primary sclerosing cholangitis or concomitantly with other autoimmune conditions. Inflammation in this category is found both within the pouch body and afferent limb, with some cases exhibiting concurrent cuffitis [45, 46]. Ischemia-associated pouchitis is thought to occur when undue mesenteric tension on the pouch causes a chronically ischemic environment in the pouch and is characterized by inflammatory changes of only one limb of the pouch in a vascular distribution. Alternatively, one may note ulcerations along the common channel staple line. The typical patient in this category is an obese male with excessive visceral adiposity [47].

A helpful algorithm to treat patients suffering from pouchitis is to initiate a 2-week course of oral ciprofloxacin and Flagyl. Those who are antibiotic-responsive and have resolution of symptoms may undergo additional antibiotic treatment as symptoms recur. Those who respond but are dependent on medications for remission should continue a monitored maintenance antibiotic/probiotic regimen. Patients who continue to have symptoms despite antibiotic therapy should be evaluated for pathogen-induced pouchitis (CMV/C difficile infection) or immune-mediated pouchitis and undergo the appropriate treatment depending on presumed etiology [45]. When ischemic pouchitis is suspected, the patient should be encouraged to pursue loss of visceral adiposity [48].

Cuffitis

Cuffitis is the symptomatic inflammation of the remnant rectal cuff that remains in UC patients after they undergo stapled IPAA. Symptoms can occur in up to 6% of patients with a double-stapled anastomosis and are at times confused with those caused by pouchitis. In these cases, patients may be inappropriately diagnosed and treated for refractory pou-

chitis, when better medical and surgical treatments are available for cuffitis. Medical options of topical steroid enemas, suppositories, or aminosalicylate (5-ASA) drugs are effective. In rare cases when symptoms are not responsive, surgical intervention is warranted. The residual rectal mucosa can be removed with a transanal mucosectomy, followed by ileal pouch advancement with pouch-anal handsewn anastomosis or by transabdominal/transanal pouch redo with anal canal mucosectomy. The success of surgery is increased if the initial stapled anastomosis is no more than 3–4 cm above the dentate line [27] and a tension free anastomosis is fashioned [49].

Crohn's Disease of the Pouch

A diagnosis of CD of the pouch does not necessarily require one to pursue pouch removal with permanent conventional ileostomy as a first step. Disease phenotype heavily influences degree of pouch retention. A study of 65 patients with de novo CD of the pouch reported that 57% were able to maintain their pouch with acceptable function despite this diagnosis. However, the presence of fistulae at the time of diagnosis of CD of the pouch and early diagnosis of Crohn's disease after initial pouch surgery were independent risk factors for pouch failure [50].

Surgical therapies for CD of the pouch may be employed independently or in combination with medical and endoscopic treatments. Regardless of approach, it is most important to consider the desires of the patient and his or her individualized definition of quality of life, as this should be the ultimate measure of treatment success. Many patients diagnosed with CD of the pouch desire pouch preservation and should be offered an evaluation in an IBD center with surgeons experienced in treating this challenging scenario. Those who are not interested in pouch preservation and choose to pursue a permanent conventional ileostomy should be equally supported in this endeavor as well. In either case, the majority of patients with CD of the pouch benefit from both medical and surgical therapy in parallel.

Diagnosis with Exam Under Anesthesia

Making an accurate diagnosis of CD of the pelvic pouch is the cornerstone of success in these challenging patients. An examination under anesthesia (EUA) is often the best first operative option in these patients, allowing the surgeon to establish the correct diagnosis, control sepsis control, and obtain biopsies. Fistulae from a CD pouch are easily confused with pelvic sepsis from a chronic pouch-anal anastomotic leak, and distinguishing between these is critical as the treatment and prognosis are vastly different. It is generally accepted that pelvic sepsis within 3–6 months following ileostomy closure after IPAA is likely a postoperative complication rather than a sequela of CD of the pouch, which is more likely to manifest more than 12 months after IPAA [16, 51].

Control of Sepsis

An initial EUA allows the surgeon to carry out the next critical step of managing CD-related pouch fistulae, which is the control of sepsis. This can be performed using carefully placed mushroom drains in abscess cavities and non-cutting silastic setons to manage fistula tracts (Fig. 50.5). Indiscriminate injury or division of the anal sphincter complex should be avoided, as the risk for fecal incontinence is high. Cautious and gentle completion of these local procedures may control symptoms, improve quality of life, and help to maintain the best chance for pouch preservation for the future [51].

Fecal Diversion

As mentioned previously, a diverting loop ileostomy with pouch in situ is an effective method of controlling symptoms

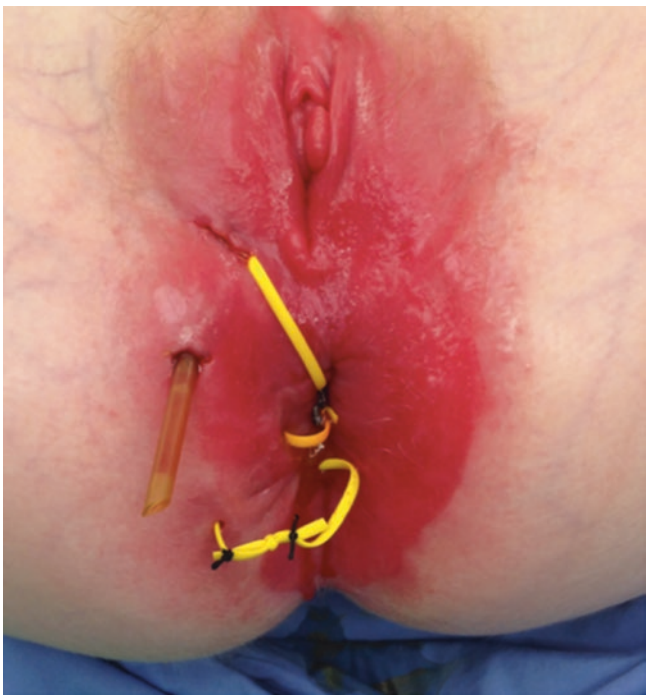

Fig. 50.5 Anoperineal sepsis in a patient with Crohn's disease of the pouch. Control of sepsis is achieved with thoughtfully placed drains and setons

from fistulizing CD of the pouch when the patient has failed medical/endoscopic therapy and/or is not ready to commit to pouch excision or pouch revision (if an option). It is important to emphasize that fecal diversion *improves* but does not necessarily *resolves* anorectal symptoms, as patients may experience ongoing mucous drainage and untoward symptoms from diversion pouchitis [52].

Pouch Excision

Pouch excision is, at times, a necessary surgical option when medical, endoscopic, and local surgical therapies fail but comes with a high morbidity rate. Pathologic confirmation of CD of the pouch is not always confirmed after pouch excision, as shown in a series of 35 such patients, with only 7 cases achieving pathologic diagnosis of CD [53]. A morbid complication of pouch excision is the nonhealing perineal wound and subsequent development of perineal sinus, which can be more difficult to manage than a pouch left in situ. This occurs in up to 40% of patients, and the risk for this troublesome complication should be considered when developing a surgical strategy (Fig. 50.6) [54]. Fecal diversion with staged pouch excision may help reduce the risk for nonhealing.

Pouch Revision in the Setting of CD

Carefully selected patients suffering from fistulizing CD of the pouch may be candidates for pouch revision, either with perineal/perianal repair of the existing pouch or major corrective surgery with recreation of a new pouch-anal anastomosis and/or new pouch.

Any surgical repair of a pouch fistula first requires control of sepsis to normalize tissue quality, followed by medical therapy to reduce inflammation and promote healing. During this time, response to therapy is monitored and assessed, and discussions regarding the next steps must establish reasonable patient expectations and the goals of surgery in these very challenging cases.

Local procedures such as seton placement, mucosal advancement, and fistulotomy have been studied as a means to mitigate symptoms of CD-related pouch fistulas. Although there is evidence to support the use of local procedures for CD-related complications of the pouch, the presence of a fistula at the time of CD diagnosis was an independent risk factor associated with pouch failure [50].

Data regarding pouch revision for CD pathology of the pouch are very limited. Unpublished data regarding patients undergoing redo IPAA for CD revealed pouch retention rates were lower than index pouches (<60% vs 79% at 5 years) but perioperative complications and functional outcomes were comparable. This highlights the critical importance of proper patient selection for this process. Additionally, acceptable outcomes of revisional IPAA surgery for CD of the pouch can be achieved in very carefully selected patients who present for surgery with no active anoperineal disease, limited small bowel disease, and an uncompromised anal sphincter.

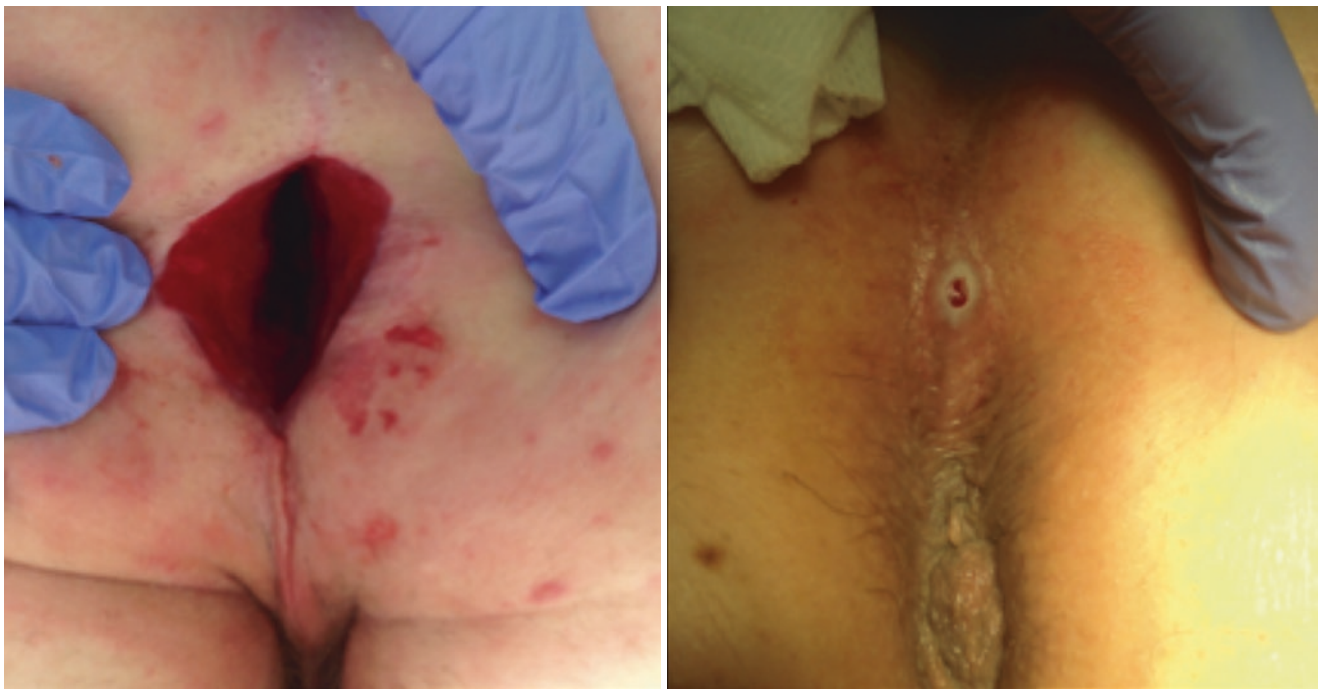

Fig. 50.6 A nonhealing perineal wound (left) and persistent sinus tract (right) after pouch excision for Crohn's disease

Above all, any patient undergoing pouch revision for fistulizing CD must have insight as to the complexity and limitations of redo surgery in this setting and accept the increased risk for postoperative complications, eventual pouch loss, or need for long-term medical therapy.

When considering surgical options for CD of the pouch, it is important to re-emphasize that many patients are misdiagnosed with CD, when failure is actually due to technical complications at the pouch-anal anastomosis. These patients are commonly good candidates for a redo pouch but were not considered owing to an incorrect diagnosis of CD.

Functional Complications of the Pouch

Irritable pouch syndrome is a clinical scenario in which a pouch patient suffers from symptoms of diarrhea, urgency, pelvic pain, and cramping in the absence of endoscopic or histologic findings of mucosal inflammation. It is thought to be related to visceral hypersensitivity and hypermotility of the pouch, but the true etiology is poorly understood. In one study of 61 patients after RP IPAA, 43% exhibited the clinical symptoms described above, with postoperative complications, pouchitis, and CD ruled out. Although an effective treatment strategy is still being elucidated, patients often benefit from a combination of systemic and topical antidiarrheal, antispasmodic, and anticholinergic therapies, in addition to cognitive behavioral therapy [55, 56].

Dyssynergic defecation (DD) or nonrelaxing pelvic floor dysfunction, in which the puborectalis muscle fails to relax during defecation, can cause dyschezia and straining in pouch patients. DD can occur as a primary disorder (idiopathic) or secondary disorder (associated with inflammatory pouch conditions like pouchitis or cuffitis). Anal manometry commonly shows paradoxical contractions of the muscle and failure of the balloon expulsion test. Pelvic floor physical therapy (biofeedback) is helpful in many cases of both primary and secondary DD, along with the treatment of underlying inflammatory conditions that may coexist [57–59].

Neoplasia of the Pouch

Cancers of the pelvic pouch are poorly understood, difficult to detect even with routine endoscopic surveillance, and have a poor prognosis. Studies on the topic are sparse and primarily consist of case reports and small series. One of the largest studies of over 3000 pouch patients reported a cumulative incidence for pouch dysplasia of 0.8% at 5 years and 2.2% at 20 years after pouch construction [60]. Pouch dysplasia is primarily noted at the anal transition zone or rectal cuff and is more likely to occur in IBD patients whose original indication for proctocolectomy was dysplasia or cancer.

Less than 50 cases of pouch cancer have been reported in the literature, with the majority being adenocarcinoma located in the ATZ (64%) or pouch body (19%). Cumulative incidence for pouch cancer has been reported as 0.2%, 0.4%, and 2.4% at 5, 10, and 20 years in one large study of over 3000 UC patients [60], with similar results in other studies [61].

Outcomes of Surgical Management of Pouch Complications

The decision regarding what surgical options to offer a patient with a failed IPAA is extremely complex with life-altering consequences for the patient. On the one hand, pouch excision offers the hope of a better quality of life (QOL) but requires the acceptance of a permanent conventional ileostomy and risk for wound healing issues. Conversely, pouch repair and revision maintain continuity of the intestine but sometimes require a commitment to undergo multiple major operations over an extended period of time. The literature informing the optimal approach to pouch failure with regard to pouch excision vs pouch redo is limited. There are no randomized trials available or studies that directly compare the two approaches. The large majority of available studies are retrospective and descriptive experiences of specialized, high-volume IPAA centers.

The traditional approach to pouch failure has been to offer the patient pouch excision with a permanent conventional ileostomy, often in one operative setting. However, several studies on the topic have reported significant postoperative morbidity after this operation. A retrospective review from the Mayo clinic of 147 patients undergoing pouch excision reported short- and long-term complication rates of 57% and 37%, respectively, with 11% requiring a return to the operating room due to complications within the immediate postoperative period [4]. This is consistent with a prior study from St Mark's Hospital reporting a 25% and 53.7% early and late postoperative complication rate, respectively. Over half of the patients required readmission, with greater than 50% of

these patients requiring reoperation. Persistent perineal wounds were reported in 40% and 10% at 6 and 12 months, respectively [54]. Another retrospective report highlighted the difficult challenge of postoperative perineal wound healing in their study of 47 patients undergoing pouch excision. Of these, nearly 30% suffered from perineal wound complications, including perineal wound infections (100%), perineal sinus tracts (28%), and perineal hernia (7%) [62].

The significant morbidity and need for permanent conventional ileostomy are major drawbacks of pouch excision for pouch failure [63], thus making pouch redo an attractive alternative in highly selected patients. Remzi reported the largest experience describing outcomes of redo pouch surgery performed in over 500 patients spanning three decades. The large majority suffered from sepsis-related pouch dysfunction. Postoperative complications occurred in 53%, with pelvic sepsis the most common. Ileus/bowel obstruction (16%), anastomotic leak (8%), and wound infection (8%) were the most common short-term complications (along with pelvic sepsis). A total of 20% of patients had redo IPAA failure, but 83% of patients had a functional IPAA at most recent follow-up, with 5- and 10-year pouch survival noted to be 90% and 82%, respectively. This report is one of few that examined QOL and functional outcomes and reported that more than 90% of patients recommended surgery to others and would undergo the surgery again if needed [5]. Overall, these results support the important role of pouch revision surgery in carefully selected patients. Many patients with IPAA failure may have a second chance to achieve stoma-free living with acceptable bowel function and quality of life with the redo pouch.

Other series report similar positive results, albeit with smaller number of patients and more limited follow-up. One recent study of 81 patients undergoing pouch revision reported a predicted 5- and 10-year pouch survival of 85% and 65%, respectively, and pouch loss of 23%. The overall (early and late) complication rate was 35.6%, with most the common complications being ileus/bowel obstruction and recurrent fistula [6]. Another study described the outcomes of 51 patients undergoing pouch salvage, 23 of these undergoing transabdominal redo. Of these, 69% were reported to have acceptable functional results, with septic events described as the most notable and morbid postoperative complication [8]. Others have also reported successful redo IPAA with good functional outcomes and patient satisfaction with acceptable rates of complications [64–67].

Patients with IPAA dysfunction should be offered the opportunity to undergo comprehensive evaluation in an IPAA center (experienced in revisionary pouch surgery), with discussion of multidisciplinary management options. A patient's decision to pursue an improved QOL by accepting pouch excision with a permanent lifelong ileostomy should be honored without exception and without persuasion otherwise. For appropriately selected patients desiring an attempt at pouch salvage, pouch revision and redo are good options with a high likelihood of success and require a thoughtful and honest discussion between patient and clinicians to set shared goals and expectations for care.

One additional tool in the toolbox of the reoperative pouch surgeon is the continent ileostomy (CI). CI is an intra-abdominal ileal reservoir made with a continent nipple valve that allows for patient control of stool evacuation (Fig. 50.7a–c). A catheter is inserted into the pouch several times daily by the patient to empty the pouch, at private and convenient times. In this way, patients are able to maintain continence with improved lifestyle and body image as compared to a permanent ileostomy [68–71]. Although less commonly created in contemporary times than the J-pouch, it remains a good option for selected patients desiring a control of bowel habits but who are not candidates for a pelvic pouch.

Despite its many benefits over conventional ileostomy, CI is a complex procedure that carries significant risk of postoperative complications as well as a long-term need for reoperation to repair nipple valve slippage, the commonest complication and indication for reoperation in these patients [70–72]. Patients must undergo extensive preoperative counseling to confer understanding of the associated risks and accept a realistic vision of life with CI. In carefully selected and motivated patients, CI continues to be a durable option, with long-term pouch survival rates approaching 80% [73]. CI patients enjoy greater QOL than others with a conventional ileostomy and that 95% would choose to undergo the procedure again and recommend it to others [74, 75].

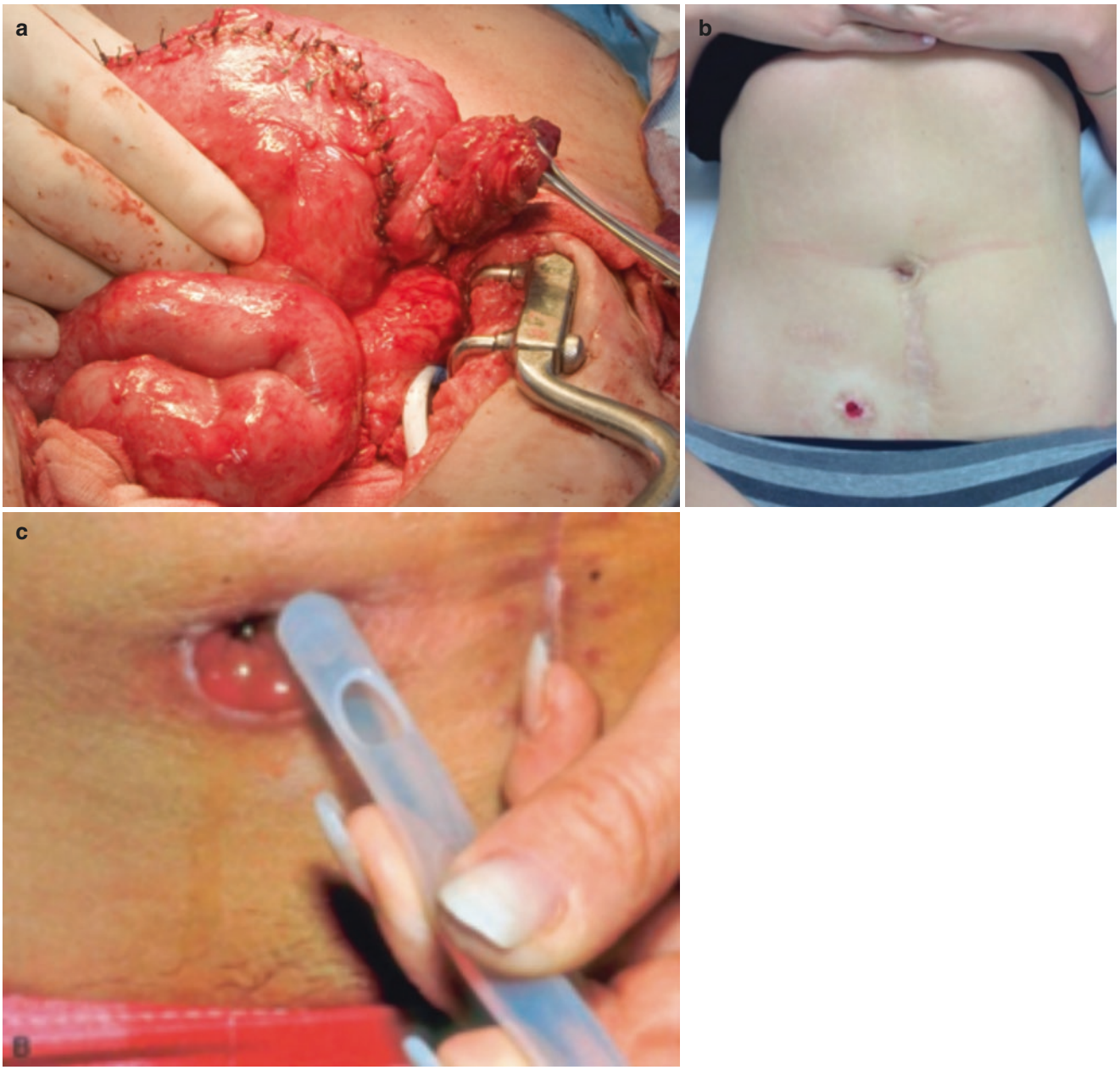

Fig. 50.7 A continent ileostomy created in a patient not suitable for a pelvic pouch (**a**). The inconspicuous stoma (**b**) gives patient control of continence with intermittent intubation and improved quality of life (**c**)

Conclusion

The debate as to how best to approach IPAA failure is multifaceted and ongoing, with limited comparative studies on which to base important decisions. One of the major barriers to mastering this topic is the remarkable uniqueness of every IPAA failure patient. Each patient is different with a distinctive etiology of pouch dysfunction coupled with personal desires and QOL aspirations. Further studies are necessary to continue to learn how to approach the patient with a failed or failing pouch; an individualized plan of care is necessary to achieve the best outcomes.

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