Chapter 18
Management of Ileal Pouch Strictures and Anal Stricturing Disease: A Clinical Challenge

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Abstract Restorative proctocolectomy with an ileal pouch-anal anastomosis (IPAA) has been an ideal surgical option for patients with chronic ulcerative colitis (UC), familial adenomatous polyposis, and selected patients with colorectal cancer and Crohn's disease for nearly four decades. In most cases, patients enjoy excellent quality of life with a durable surgical and functional result, avoiding the need for a permanent conventional ileostomy.

Despite great success, patients with IPAA may suffer from several pouch-related complications that are a challenge for the patient and clinician. IPAA-associated fibrotic stricturing disease is one such challenging complication that requires thoughtful judgment for successful management. Treatment of fibrotic strictures of the IPAA requires a multidisciplinary approach involving medical, endoscopic and surgical input for accurate diagnosis, effective treatment, and improvement of quality of life.

The focus of this review is to provide a structured approach to the challenges that the clinician encounters when faced with a patient with IPAA-associated fibrosis and stricturing disease and to discuss the surgical options that alleviate the morbidity caused by ileal pouch fibrosis when medical treatments fail.

Keywords Ileal pouch · Surgery for IPAA stricture · Ileostomy · Multidisciplinary ileal pouch team · Ileal pouch failure · Pouch disorders · Pouch stricture
Ileal pouch fibrosis

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18.1 Introduction

Restorative proctocolectomy with ileal pouch-anal anastomosis (IPAA) has been an ideal surgical option for patients with ulcerative colitis and familial adenomatous polyposis, and very selected patient with colorectal cancer and Crohn's disease, for over three decades [1, 2]. In most cases, patients report excellent quality of life with a durable surgical and functional result, and are able to avoid a lifelong ileostomy [3]. IPAA has undergone several modifications in its approach since it was popularized in the early 1980s. Over this time, innovative approaches have been applied to IPAA surgery, functional outcomes have improved, and pouch survival has remained high when performed in high-volume centers with surgeons experienced in these types of surgery [4, 5].

When surgery goes according to plan and recovery proceeds without event, patients enjoy excellent quality of life with manageable bowel function and are without major lifestyle limitations [3]. However, circumstances may occur in which patients suffer from immediate or eventual IPAA dysfunction with compromised bowel function and quality of life [1, 6, 7]. One cause of a poorly functioning IPAA that poses great challenges to the patient and clinician alike is development of fibrotic stricturing disease in or adjacent to the IPAA. A proposed etiology, diagnostic approach, and management strategies often employed to address this challenge will be discussed at length in the following text.

18.2 Construction of the Ileoanal Pouch

IPAA surgery consists of removal of the colorectum and creation of an ileal reservoir, which is constructed from the distal ileum (Fig. 18.1). The reservoir is joined, using varying methods, to the anorectal ring to restore intestinal continuity. In patients with severe fulminant colitis or who have poor health, the procedure is performed over an extended time period in multiple stages. This usually involves performing a colectomy with end ileostomy, followed by proctectomy with diverted IPAA when health is restored, usually after a waiting period >6 months. In very carefully selected patients who are otherwise fit and have no risk factors for poor healing, a single-stage IPAA may be a safe option, but this should be a rare occurrence [8].

The first reports of IPAA decades ago described construction of an S-shaped ileal pouch that was secured to the anal canal using a hand-sewn anastomosis [9]. A variety of configurations have been considered over time, including the S, J, W and H configurations (Fig. 18.2) [10]. The J pouch is the most popular configuration presently, as it is the easiest and most expeditious to construct and its construction may be assisted by stapling devices [11]. The S and W pouches necessitate a lengthier segment of distal ileum and typically require a hand-sewn approach to construct the

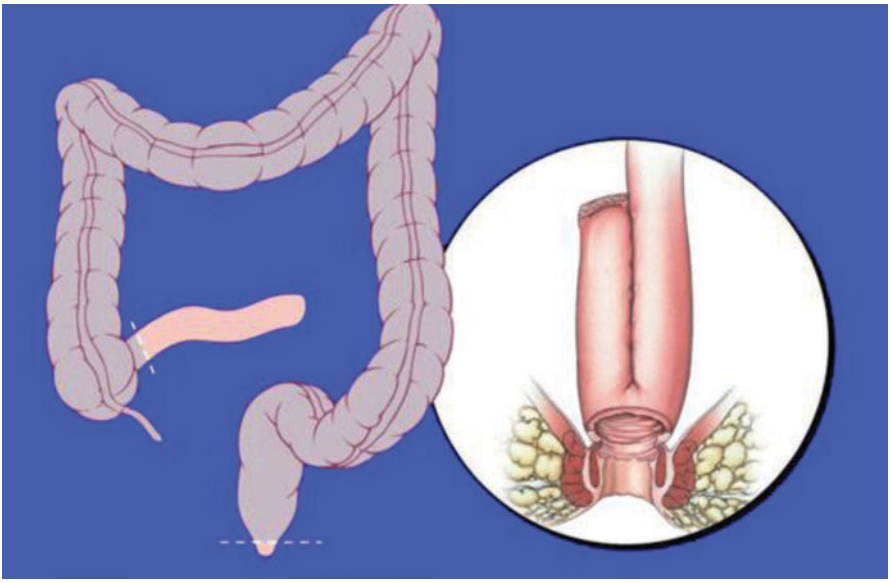

Fig. 18.1 Restorative proctocolectomy with ileal pouch-anal anastomosis (IPAA)

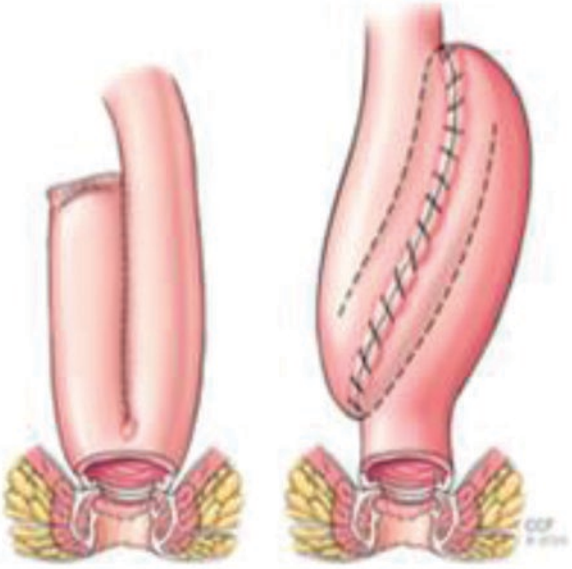

Fig. 18.2 Ileal J pouch (left) and S pouch (right)

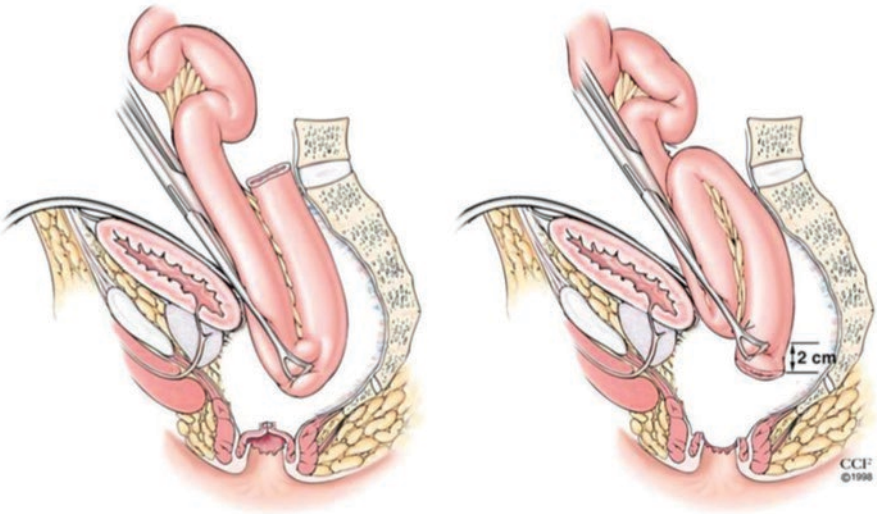

Fig. 18.3 Mesenteric reach with J (left) and S (right) pouch configurations

actual pouch, and thus are more time-consuming and technically challenging to create. The J pouch configuration is most commonly used unless adequate mesenteric length is not available, as creating a tension-free pouch-anal anastomosis is the most critical step to successful pouch surgery. In the case where a J pouch will not reach without tension, an S pouch may be helpful as its configuration allows for a longer reach (2–4 cm longer than J pouch) into the pelvis (Fig. 18.3). A pouch-anal anastomosis created under tension is destined to result in anastomotic leak and pelvic sepsis in the short term, and leads to pelvic fibrosis or chronic pouch ischemia with poor pouch function over time [1].

The ideal method of constructing the pouch-anal anastomosis has long been debated, with the stapled IPAA as the preferred method over hand-sewn IPAA in most instances. The introduction of stapling devices several decades ago made it possible for the stapled IPAA to be less-time consuming and associated with better outcomes than hand-sewn IPAA [12]. In addition, patients with UC undergoing a stapled IPAA rarely develop cancer in the preserved anal transition zone (ATZ) [13]. The stapled IPAA is carried out with either a single or double-stapled approach and the IPAA is joined to the ATZ, thus preserving anal sensory epithelium (Fig. 18.4). Conversely, a hand-sewn IPAA is performed by first removing all anorectal mucosa from the dentate line cranially to the anorectal transection (Fig. 18.5). The IPAA is then delivered into the pelvis and sutured to the internal sphincter at the neo dentate line in a radial fashion. If properly performed, the anal sensory epithelium and all rectal mucosa is removed in this method. However, this method is more likely to exhibit stricture formation at the anastomosis.

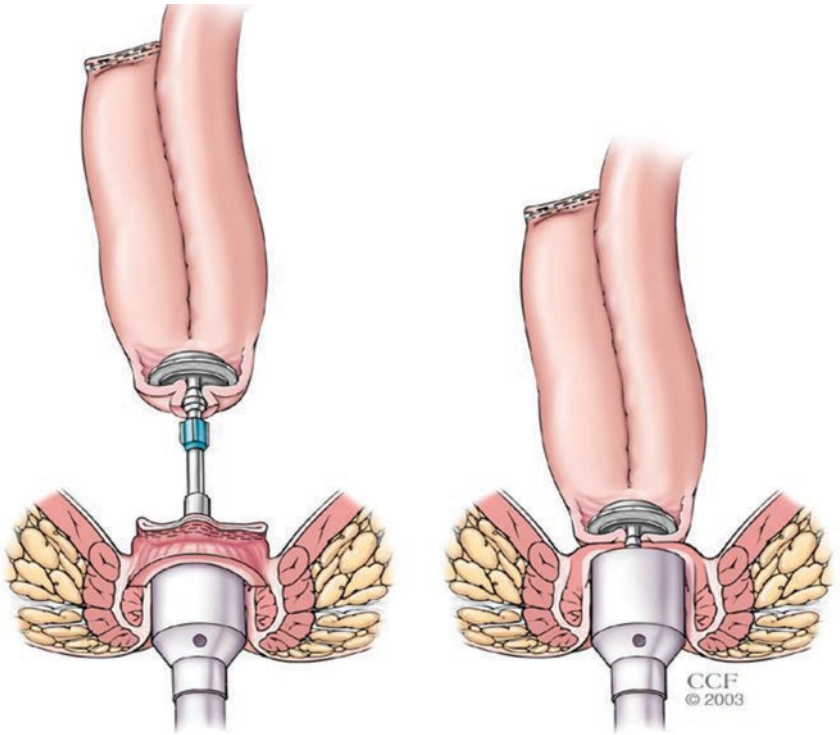

Fig. 18.4 Stapled IPAA

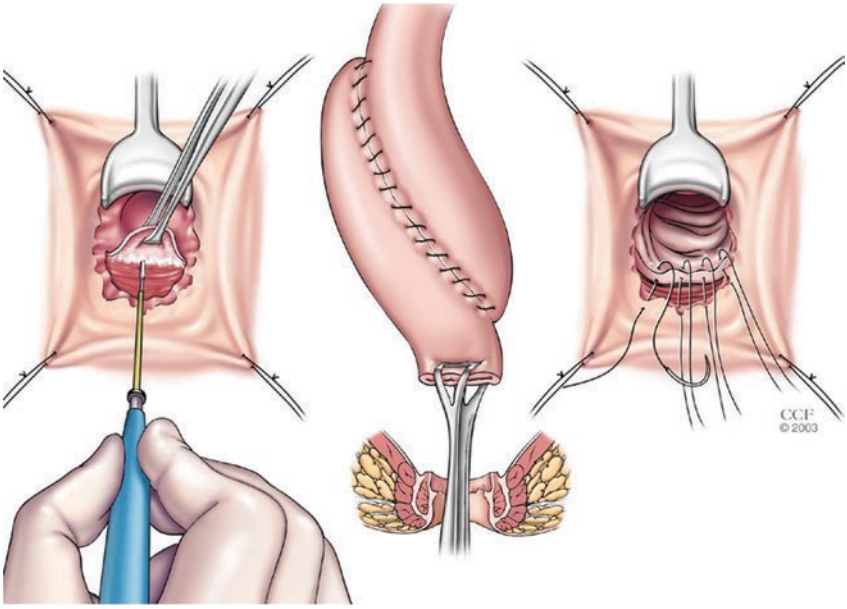

Fig. 18.5 Mucosectomy with hand-sewn IPAA

18.3 Etiology of Fibrotic IPAA Dysfunction

Although many factors may underlie stricture formation, patients who undergo pelvic pouch surgery most commonly develop fibrosis due to the presence of chronic pelvic sepsis. This persistent inflammation in the pelvis and/or anoperineum, if not controlled early, leads to fibrotic changes in the pre-pouch small bowel (afferent limb), pouch body, pouch outlet (efferent limb) or anoperineum [14, 15]. Chronic pelvic sepsis that develops in the months following IPAA surgery is likely the result of technical complications leading to pouch-anal anastomotic leak. Conversely, pelvic sepsis which develops many months to years after IPAA surgery is more likely to be untoward sequelae of Crohn's disease. Regardless of etiology, all pouch-related sepsis necessitates expeditious diagnosis and drainage in order to reduce the risk of stricture development.

Other etiologies have been proposed as causes of IPAA fibrosis and stricture, including weight gain and increased abdominal girth after pouch surgery resulting in excessive mesenteric tension and chronic pouch ischemia [16–18]. In addition, pelvic radiation in the setting of IPAA surgery is associated with pouch fibrosis and subsequent high risk for failure [19].

Regardless of etiology, clinical symptoms from IPAA-related fibrosis depends upon location and severity of inflammation. Fibrotic strictures upstream of the IPAA in the pre-pouch ileum (afferent limb) cause patients to suffer from obstructive symptoms like abdominal pain, cramping, and limited dietary intake of fibrous foods. Bowel motions may be primarily watery or loose, as more bulky components of stool do not pass easily and are detained upstream of the stricture. Fibrosis around or involving the pouch body restricts the ability of the pouch to accommodate and distend, thus reducing its volume and leads to frequent bowel motions. Strictures of the efferent limb (rectal cuff) or anal canal may make pouch emptying difficult, leading to excessive straining, feelings of incomplete emptying, chronic pouch dilation and stretch, and overflow incontinence [20]. Often, a careful and meticulous history can elicit these telltale symptoms from the patient, allowing the clinician to predict the location of stricture even before radiographic or endoscopic evaluation is complete.

An additional site of concern after IPAA surgery is the ileostomy closure site, which may develop stricturing disease due to a subclinical anastomotic leak or ischemia at the time of ileostomy closure, or excessive scar formation after closure (Fig. 18.6). This site must always be interrogated and considered as a part of the

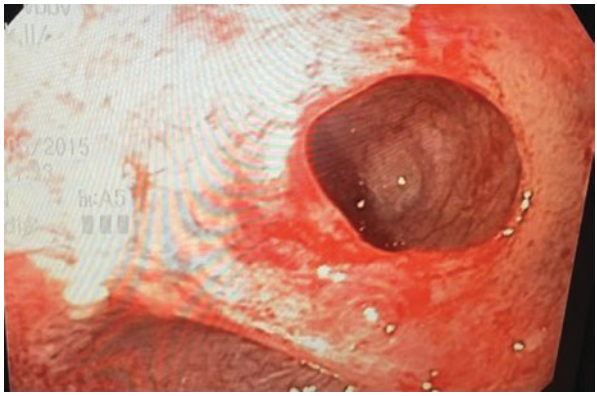

Fig. 18.6 Fibrotic stricture at stapled ileostomy closure site (reuse by permission only JA CCF)

evaluation for IPAA dysfunction as it may mimic obstructive symptoms similar to that of a strictured afferent limb. This would be particularly important in the patient with an endoscopically healthy IPAA but with ongoing obstructive symptoms.

18.4 Evaluation of the Dysfunctional Pelvic Pouch

18.4.1 Initial Evaluation

First and foremost, patients referred with a diagnosis of IPAA-associated fibrosis or stricture should undergo a comprehensive and standardized evaluation for IPAA dysfunction. A complete history should be obtained including a full review of the patient's symptoms, treatments that have been attempted prior to the surgical evaluation, and response to each treatment. Commonly, patients report obstructive symptoms as a result from fibrotic disease of the small bowel and IPAA, with specific complaints dependent on the location of the stricture and fibrosis. They often relay difficulty with abdominal cramping and bloating after meals, intolerance of fibrous foods, and challenges related to storage and emptying of stool from the IPAA.

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 that may lead to fibrosis development may be easily missed. One should pay particular attention to the condition of the patient at the time of pouch creation and the use of temporary fecal diversion. Large doses of immunosuppression negatively affect pouch healing and anastomotic complications may result in occult sinus tracts, anastomotic leaks, chronic inflammation, and subsequent IPAA-associated fibrosis [21–23]. Also important is a review of the patients weight history, with notations regarding weight and body habitus at the time of IPAA surgery and subsequent changes since this time. Weight gain, specifically growth in abdominal adiposity and girth may put undue tension on the small bowel mesentery, resulting in a relative chronic ischemia leading to fibrotic stricture of the pouch-anal anastomosis [16–18].

A thorough physical exam is necessary during evaluation for several reasons. Fibrotic strictures causing obstruction in or around the IPAA may manifest as chronic abdominal distention and tympany on exam. A contracted, fibrotic IPAA with limited reservoir capacity causes increased bowel frequency and severe perineal excoriation from excessive wiping. An anal exam and anoscopy conducted in the clinic setting may reveal a distal stricture, but is often limited by patient discomfort unless sedation is administered.

Selective use of cross-sectional and fluoroscopic imaging studies help to further characterize symptomatic fibrotic disease. CT enterography gives information regarding stricturing disease in the upper gastrointestinal tract and more proximal small bowel that is not reachable by endoscopy. Distal contrast enema with adequate administration of transanal contrast is helpful to identify fibrotic strictures in or around the pouch, reveal the distensibility of the pouch, and identify strictures

upstream at the ileostomy closure site, all of which may cause or contribute to patient symptoms. MRI of the pelvis demonstrates the presence of pouch-anal sinus and fistula tracts that cause inflammation and stricture of the efferent pouch limb or contracture of the pouch body itself, compromising its function.

Pouchoscopy is an effective diagnostic tool and allows for therapeutic intervention in some situations. The exam is best performed under sedation for optimal patient comfort and minimal disturbance if intervention is performed, or in the operating room as discussed later in this section. This study allows one to locate and characterize the severity of the stricture, identify additional contributory pathology, and allows intervention with endoscopic dilation, therapeutic maneuvers such as needle-knife therapy, and tattooing for eventual surgical localization and therapy [24].

A comprehensive exam performed as outlined above allows the clinician to characterize the location (pre-pouch/afferent limb, pouch body, or pouch outlet/efferent limb) and severity of IPAA-associated fibrosis so the appropriate patient-centered treatment strategy may be formulated. In addition, one must assess and consider the patient's health status and quality of life in the decision-making process, even if the etiology of fibrotic pouch dysfunction is still unclear. Patients are often evaluated after years of suffering that have left them malnourished, decompensated, and mentally exhausted. These individuals may benefit from temporary fecal diversion to alleviate symptoms of fibrotic strictures even though a definitive plan for the primary disease has not yet been established.

Finally, it is important for the clinician to have an honest and straightforward discussion with the patient regarding expectations of treatment for IPAA-related fibrotic disease. It must be emphasized that interventions, whether medical, endoscopic or surgical, may mitigate symptoms and improve quality of life, but also risk damaging the pouch, small bowel, or anoperineum. Inadvertent injuries may require repair or temporary fecal diversion, or even lead to pouch excision and permanent conventional ileostomy. Expectations must be discussed and agreed upon prior to embarking on these interventions.

18.4.2 Multidisciplinary Approach to Diagnosis

When a patient presents with symptoms concerning for IPAA-related fibrosis, the authors often use a multidisciplinary approach to evaluate the IPAA. After preoperative evaluation with history, physical, and radiographic testing as outlined above, an evaluation with an anoperineal exam under anesthesia with pouchoscopy is performed as a team by the colorectal surgeon and gastroenterologist. The anoperineum, pouch-anal anastomosis, pouch body, and afferent limb (complete to the ileostomy closure site) are examined with members of both specialties in the operating room, offering both perspectives of expertise. Any clinical signs of IPAA complications are noted (anastomotic sinus or fistula, stricture, pouch prolapse, Crohn's disease, etc.), many of which may cause similar symptoms [25]. Biopsies are obtained for pathologic review. At the completion of the exam, the findings are

discussed with the patient and family member along with a patient-centered treatment strategy. This multidisciplinary team approach is ideal for the patient as he/she is 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 or recommendations from our Multidisciplinary Inflammatory Bowel Disease Conference are available.

18.5 Treatment Strategies for IPAA-Related Fibrosis

Any IPAA-related intervention must be preceded by a frank discussion with the patient regarding the possibility of injury to the pelvic pouch or associated small bowel requiring urgent laparotomy, attempt at repair, and need for fecal diversion. Unfortunately, some situations may result in irreparable injury requiring pouch excision, and the patient must understand and accept this risk prior to embarking on endoscopic or surgical management. Ideally, the patient, endoscopist and surgeon together make coordinated decisions in a multidisciplinary and patient-centered fashion, with a contingency plan present and rehearsed in case surgical exploration is required to address complications.

18.5.1 Pre-IPAA (Afferent Limb/Ileostomy Closure Site)

Choosing the best treatment option for strictures in the pre-IPAA bowel begins with assessment of severity and etiology of disease. Asymptomatic strictures found incidentally on routine endoscopy may be left alone, or gently dilated to prevent progression. Mild to moderate strictures that receive an ileoscope can be treated with balloon dilation for effective yet controlled expansion of the strictured segment. Severe strictures that do not easily receive an ileoscope or allow only a wire to cross may also be treated this way, but must be performed with great caution as risk for perforation or luminal hemorrhage is great and risk should be balanced with benefit of proceeding. Strictures at the ileostomy closure site may be dealt with in a similar way with cautious and gentle balloon dilation to minimize risk for complication [26].

Most recently, endoscopic needle-knife strictureplasty, has been proposed as a method of endoscopically 'coring' the fibrotic ring of the strictured segment instead of breaking the fibrotic ring with outward pressure, as is the case with balloon dilation [27].

This technique requires specialized skill, comfort with the needle-knife technique, and a readily available and willing surgeon experienced in IPAA repair [28]. For these reasons, it is a technique best performed in high-volume referral centers. Initial reports of the success of this technique are few but promising, and long-term efficacy studies are needed [29, 30].

Often, endoscopic therapies, particularly balloon dilation, result in reformation of scar at the strictured site, and subsequent recurrence of symptoms. If a patient enjoys a

relatively long symptom-free period after the initial dilation, one may consider repeat dilation when symptoms do recur. However, progressively shortened symptom-free periods between dilations may prompt the patient and clinician to consider other options for treatment. In these circumstances, surgical intervention should be considered.

Strictures proximal to the IPAA at the ileostomy closure site or afferent limb are best approached by means of strictureplasty or small bowel resection with primary anastomosis (Fig. 18.7) [31]. If strictureplasty is performed, a Heineke-Mickulitz type is most common, in which a longitudinal enterotomy created on the anti-mesenteric segment of the intestine is closed transversely in a handsewn seromuscular fashion, thus expanding the luminal diameter. These are ideal for short-segment strictures. When a small bowel resection is preferred, the surgeon must divide the mesentery just underneath the bowel lumen to prevent compromise of the blood supply to the IPAA. A primary end-to end anastomosis is best employed to recreate intestinal continuity at these locations (Fig. 18.8); however, a stapled anastomosis is

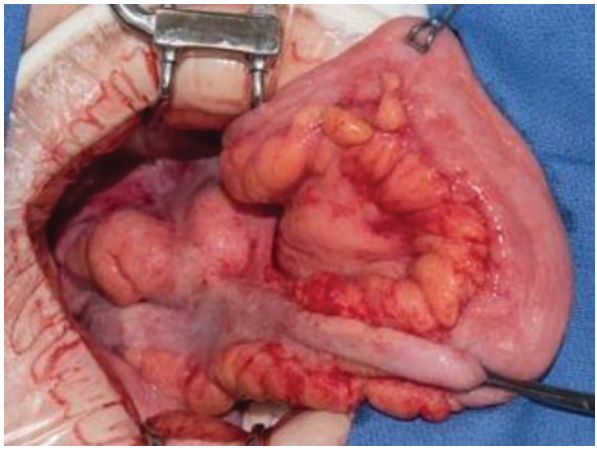

Fig. 18.7 Fibrostenotic Crohn's disease of the afferent limb of IPAA (reuse by permission only JA CCF)

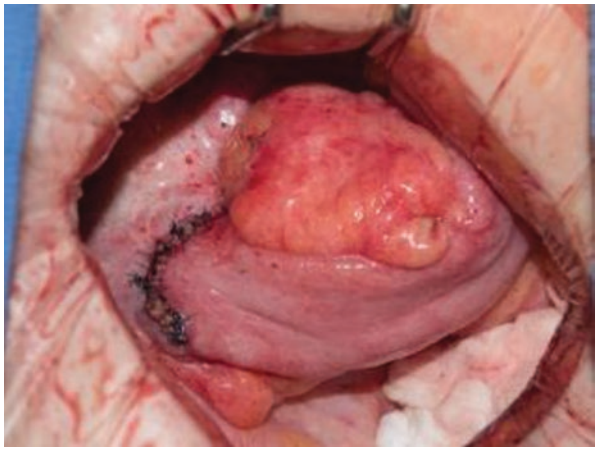

Fig. 18.8 Small bowel hand-sewn anastomosis after resection of afferent limb stricture (reuse by permission only JA CCF)

an effective alternative to this if carefully performed, with transection at the pouch inlet (below the fibrosis) with a linear stapler, careful resection of the affected segment, and joining of the distal ileum to the top of the pouch or tip of J with an EEA circular stapler that is transanally introduced.

Patients who are failing to thrive, immunosuppressed or generally unwell in the perioperative period are best considered for diverting ileostomy at the time of repair to mitigate the potential of anastomotic leak, with ileostomy closure at a later time when the patient has recovered his or her health. Patients exhibiting pre-pouch fibrosis as a result of fibrostenotic Crohn's disease are re-initiated on medical therapy as soon as possible after recovery from surgery.

18.5.2 The Fibrotic IPAA Body

Fibrosis involving the pouch body is a unique and often very morbid situation, and is most commonly a sequelae of chronic ischemia. The IPAA is typically contracted with poor distensibility and is only able to hold low volumes of stool. Patients complain of frequency and urgency of bowel motions, excoriation of the perianum due to frequent soiling and need for frequent cleaning. Those who are severely affected by symptoms should be considered for fecal diversion or pouch excision. In some circumstances, patients may be considered for a redo IPAA if the reason for failure of the first pouch is clearly identified and able to be avoided at the time of a second attempt. One example of this is the situation of a patient who has gained excessive abdominal adiposity since creation of the IPAA. The small bowel mesentery is put on stretch and slowly produces a relative ischemia of the pouch and resultant fibrosis [16, 18]. An acceptable strategy for this patient is for him/her to first pursue adequate weight loss to achieve ideal body weight, followed by redo of the IPAA and strict maintenance of this weight after surgery.

18.5.3 Post-IPAA (Efferent Limb, Anal Canal)

Strictures that are distal to the IPAA are most easily assessed in the outpatient clinic setting, as these are apparent on digital exam. However, although selected patients may be amenable to awake exam or gentle dilation, most will be too uncomfortable for much more than a brief assessment. A thorough exam, typically done in the operating room or sedation suite, is often necessary to determine specifics and etiology of disease, as this is critically important to determining treatment options (Fig. 18.9).

Patients who suffer from fibrostenotic Crohn's disease of the anal canal or pouch-anal anastomosis should first undergo drainage of sepsis with seton or mushroom drains, as soon as possible, to reduce the risk for worsening fibrosis. Those with symptomatic strictures may be offered serial dilations under anesthesia or regular

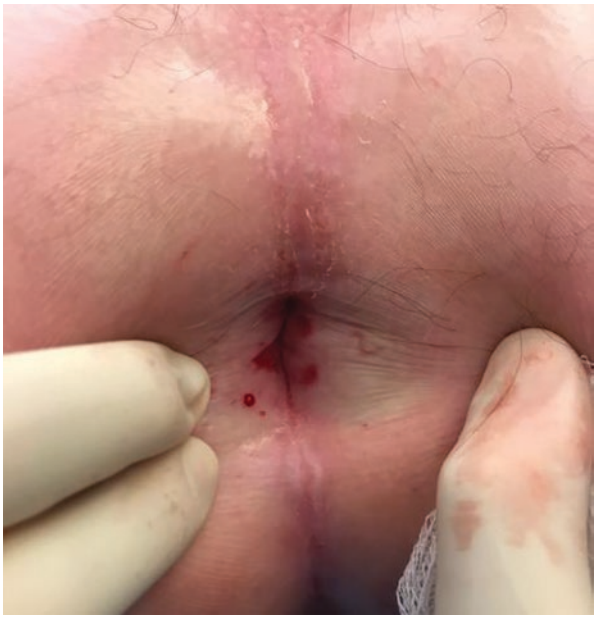

Fig. 18.9 Assessment of a post-IPAA stricture

home dilations performed by patients themselves or willing caregivers [26]. Periodic steroid injection into the fibrotic ring is thought to slow the recurrence of scarring at the time of dilation. Fibrosis will most likely progress, however, and patients deserve a discussion early on regarding the option of fecal diversion to alleviate symptoms if/when they worsen. Severely symptomatic patients are candidates for IPAA excision or proctocolectomy, both with permanent conventional ileostomy.

Outlet strictures that develop for reasons other than Crohn's disease may have treatment options in addition to those described above. Strictures may develop due to chronic ischemia affecting the exit conduit of an S pouch or chronic pelvic sepsis after anastomotic leak in a J pouch. An elongated exit conduit of the S pouch may also develop trauma-related fibrosis over time if transanal intubations are required for emptying. In these cases, surgical correction of the stricture may be performed. Transanal pouch advancement or a combined transabdominal/transanal pouch revision allows for removal of strictured tissues and recreation of a well-vascularized, tension-free anastomosis [3, 7, 32]. Those patients who are appropriate candidates should undergo a full evaluation for redo IPAA if they desire, and should never be dissuaded from pouch excision with permanent conventional ileostomy if they are accepting of this option.

18.6 Conclusion

IPAA-associated strictures present a multi-dimensional challenge that requires a clear understanding of the sequelae of fibrotic disease and is best managed through the combined efforts of physicians and surgeons with appropriate experience and

interest. Medical therapy is not commonly helpful in most cases, and when goals of treatment are not met with endoscopic modalities, early surgical evaluation and intervention is critical to ensure proper treatment and optimal patient outcomes. The ideal surgical approach to IPAA stricture is dependent upon location and severity of disease, as well as the individual goals set forth by this subset of patients with diverse characteristics and desires. The best approach for patients suffering with IPAA-related fibrosis who require intervention involves a multi-disciplinary team approach, early surgeon involvement, and a central focus on goals of curing the disease and avoiding a permanent ileostomy while preserving QOL.

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