INTRODUCTION

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An IPAA is commonly performed as a two- or three-stage procedure [5]. In the traditional two-stage procedure a proctocolectomy
is performed and the IPAA constructed with a diverting loop ileostomy. The second stage is where the loop ileostomy is closed 6–8 weeks later, restoring intestinal continuity. A modified two-stage procedure is where a subtotal colectomy is performed first. A completion proctectomy and IPAA is performed without a loop ileostomy at a second stage. In a three-stage procedure, a subtotal colectomy is performed first. The second stage is completion proctectomy and formation of the IPAA with a diverting loop ileostomy. The third stage is closure of the loop ileostomy. A minority of patients will have their proctocolectomy and IPAA formed without an ileostomy as a one-stage procedure.

Most patients will have a good outcome after IPAA, but a significant proportion of patients have a poor quality of life [6]. After creation of a pouch it can take time for adaptation to achieve optimum function [7]. Approximately one in 10 pouches will ‘fail’ at 10 years, requiring excision or permanent diversion [8,9]. There is a further group of patients who continue to experience a poor quality of life due to a suboptimally functioning pouch – what we term ‘the pouch behaving badly’. This can impinge on a patient’s psychological, emotional and physical well-being and lead to pouch failure.

Patients present with a variety of symptoms including urgency, high defaecatory frequency, bleeding, abdominal pain, weight loss, watery stool, difficult evacuation, seepage, continence issues and fatigue [10,11]. The possible underlying pathologies have overlapping symptomatology and it can be challenging for both gastroenterologists and surgeons to investigate and manage these patients.

These patients often require long-term medication to control symptoms of a badly behaving pouch, such as analgesia, rotating antibiotics and constipating drugs. Alleviating patients’ symptom burden and improving their quality of life is the therapeutic goal, but this can be a difficult place to navigate to.

This review will cover the risk factors for the pouch behaving badly, an approach to management and then an overview of the possible underlying pathology.

### RISK FACTORS FOR THE POUCH BEHAVING BADLY

The risk factors for the pouch behaving badly are the same as those which lead to eventual failure of the pouch [10]. Pouch failure is defined as the need for construction of a permanent stoma, with or without excision of the pouch or the need for an abdominoperineal pouch revision [12]. The commonest cause of pouch failure is pelvic sepsis [13–15]. Other causes include poor function (of unidentified cause) and chronic pouchitis, uncontrolled by medical therapies [15]. Fistulas, mechanical causes [14] and Crohn’s disease (CD) of the pouch may also be significant contributors to cases of pouch failure [16]. Nondiversion at initial surgery, low hospital volumes and female gender have been associated with pouch failure in a Danish national cohort [9].

Primary sclerosing cholangitis (PSC) is associated with lower quality of life after IPAA, with poorer function and increased rates of pouchitis [17]. PSC also increases mortality and the risk of postoperative sepsis after IPAA [18].

### APPROACH TO MANAGEMENT

#### Clinical assessment

The patient with a badly behaving pouch may present with multiple symptoms [10] and therefore a clear picture of the symptomatology should be constructed. Key questions to ask in the history are outlined in Table 1. We find it useful to construct a timeline of the patient’s past medical history, including gastrointestinal function prior to the development of ulcerative colitis (UC) and surgery. The presence of extra-intestinal manifestations of UC should be noted. Particular attention should be paid to the perioperative period including any complications that may have occurred.

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**FIGURE 1** Configuration of a J-type ileal pouch anal anastomosis (image courtesy of SP)
A history of early pouch-related septic complications should be noted as well as pouch function following closure of the diverting loop ileostomy and restoration of bowel continuity. Preoperative corticosteroid use should be noted, as this is associated with pouch-related septic complications [19]. Other aspects of the history, including medication, psychological morbidity, smoking status and travel, should be covered.

Assessing the patient’s quality of life in an objective and consistent way is an important part of the clinical assessment. It informs the patient and clinician of the overall impact of the disease and also allows treatment efficacy to be determined along the patient’s journey. Tools such as the Pouch Dysfunction Score [20] and the Oresland score [21] are specific to quality of life after IPAA.

Examination should focus on the abdomen and perianal area and include digital examination of the anus and pouch–anal anastomosis. Often the perianal skin is sore in patients with poor pouch function due to seepage, frequency of stools and cleaning practices. Examination under anaesthesia may allow more detailed assessment, particularly if the patient is in pain. A note should be made of the sphincter integrity and tone. The pouch–anal anastomosis should be assessed with regard to patency, integrity and height (and hence the length of the rectal cuff). The presence of sepsis, fistula or induration should be noted.

**Investigations**

Investigations should be guided by the clinical assessment of the patient. An overview of relevant investigations is given in Table 2. We find it useful to perform some initial blood tests to rule out pathology that may have been overlooked in managing a difficult pouch. Patients routinely have a full blood count, haematinsics, biochemistry, liver function tests, coeliac serology, thyroid function tests and examination of inflammatory markers. Faecal calprotectin levels are a clinically useful marker of inflammatory activity [22]. A stool culture including *Clostridium difficile* toxin is recommended.

| Table 1: Key points in the history and examination |
|-----------------------------------------------|
| **History** | **Rationale** |
| Preoperative history | | |
| Bowel symptoms before surgery and IBD diagnosis (functional symptomatology) | Concomitant irritable bowel syndrome |
| Episodes of incontinence before surgery and IBD diagnosis | Incontinence preoperatively may indicate sphincter insufficiency postoperatively |
| Original indication for colectomy | Pouchitis is uncommon in FAP. Pouch function is worse in indeterminate colitis and CD |
| Postoperative history | | |
| Histology of the colon and rectum | Higher risk of pouch complications if CD |
| Did the pouch ever work well? | Dysfunction from the outset suggests an early pouch-related septic complication such as an anastomotic leak |
| Extraintestinal manifestations | Extraintestinal manifestations of active inflammatory bowel disease (primary idiopathic pouchitis or CD) |
| Mouth ulcers | May suggest active CD |
| Difficulty emptying the pouch | Tenesmus and difficulty evacuating may suggest a stricture or functional outflow obstruction |
| Response of symptoms to antibiotics | Resolution of symptoms will occur in both primary idiopathic pouchitis and septic complications such as leak and chronic pelvic sepsis |
| Other aspects of the history | | |
| Travel history | Gastrointestinal infection |
| Drug history | NSAIDs can cause pouchitis and ulceration. Patients with irritable pouch syndrome are more likely to be taking anxiolytics and antidepressants. Ask regarding constipating drugs and barrier creams. Preoperative steroid use is associated with early pouch-related septic complications |
| Psychiatric history | A significant proportion of patients will have concomitant anxiety and depression, which needs to be recognized and addressed |
| Examination | | |
| Perianal skin and anal canal | External openings, induration, skin tags, prolapse, chronic inflammation of perianal skin, soiling |
| Height of anastomosis | Length of the rectal cuff |
| Sphincter tone | Anal sphincter insufficiency |
| Pouch–anal anastomotic stricture/anastomotic defect | A stricture may respond to gentle dilatation with a digit |

Abbreviations: CD, Crohn’s disease; FAP, familial adenomatous polyposis; IBD, inflammatory bowel disease; NSAIDs, nonsteroidal anti-inflammatory drugs.
Pouchoscopy is a useful investigation which can identify a number of underlying pouch pathologies. It will identify pouchitis, prepuce ileitis, cuffitis and strictures, and allow biopsy to assess for cytomegalovirus (CMV) and to identify granulomas pathognomonic of CD.

Pelvic MRI is the investigation of choice to assess the tissues around the pouch. It will identify chronic pelvic sepsis which can masquerade as primary idiopathic pouchitis or as CD of the pouch [23,24]. MR or CT enterography allows assessment of the remaining small bowel for CD if this is suspected and identifies areas of stricturing.

Endoanal ultrasound allows assessment of the anal sphincter muscles and manometry provides objective testing of function. This is helpful in those patients with frequent small-volume incontinence after a pouch. It should be considered where damage to the anal sphincter may have occurred, for example after vaginal delivery or during the IPAA procedure.

A defaecating pouchogram shows the size and anatomy of the pouch, especially small-volume pouches (that cause frequency), as well as documenting pouch emptying. This may reveal functional outflow obstruction such as anismus (which causes difficult evacuation and tenesmus) or prolapse. However, what constitutes a ‘normal’ defaecating pouchogram after a pouch is not known. A nondefaecating pouchogram will also show the size and shape of the pouch as well as leaks.

**Diagnosis**

Many patients have more than one cause of poor pouch function; conversely up to 20% of patients, despite assessment and investigation, will have no cause found [10].

| Investigation                  | Rationale                                                                 |
|-------------------------------|---------------------------------------------------------------------------|
| Thyroid function, coeliac serology, haematins, full blood count, routine biochemistry, liver function tests and inflammatory markers | Diagnose unrecognized concomitant pathology and establish baseline function and inflammation |
| EUA                           | Assess perianal area, anal canal, rectal cuff and pouch                     |
| Flexible pouchoscopy           | Assess pouch in detail and biopsy                                          |
|                               | Allow biopsy of pouchitis, cuffitis, prepooce ileitis, strictures           |
|                               | Also exclude cytomegalovirus                                               |
| MRI pelvis                    | Diagnose peripouch sepsis and missed leaks                                |
| Nondefaecating pouchogram     | Explore pouch anatomy, integrity and diagnosis of urological fistulas      |
| Defaecating pouchogram        | Delineate the pouch anatomy and its function – will demonstrate a small-volume pouch as well as functional outflow obstruction |
| MR/CT enterography            | Identify small bowel Crohn’s disease and other pathology such as a dominant adhesion |
| Stool culture                 | *Clostridium difficile* and other infectious causes                         |
| Breath test                   | Bacterial overgrowth                                                       |
| Faecal elastase               | Chronic pancreatitis                                                       |
| Anorectal physiology          | Anal sphincter insufficiency                                               |
| Endoanal ultrasound           |                                                                           |

**CAUSES OF THE POUCH BEHAVING BADLY**

Here we give an overview of the pathology that can cause a pouch to behave badly. We divide the pathology into surgical, inflammatory, mechanical, functional and dysplastic causes (see Table 3).

**Surgical causes**

Anastomotic leak and pelvic sepsis

Pelvic sepsis occurs when the anastomosis or a suture or staple line separates in the postoperative period. An identifiable site of leakage or contained leakage may not be found in a minority of cases [10]. Pelvic sepsis is responsible for up 50% of cases of pouch failure [15].

**Table 2** Investigations for the badly behaving pouch and their rationale

Abbreviation: EUA, examination under anaesthesia.
The diagnosis of a leak may be made early in the postoperative period and present in the classic manner: abdominopelvic pain, fever, tachycardia and generalized peritonitis. However, patients may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis [25]. Some leaks are may have mild or no symptoms (particularly as many have a diverting loop ileostomy) and this may delay diagnosis.

In the acute setting, treatment options include radiologically guided drainage of associated collections, novel techniques such as Endosponge-assisted closure [28] and a diverting loop ileostomy. Antibiotics are recommended. In the chronic setting, the pouch may require examination under anaesthetic with curettage of the cavity, widening of the opening to the cavity and drainage of any pus. Pouch excision can be offered to those patients in whom the above treatments have not succeeded. Some patients may prefer to continue with long-term antibiotics. It is important that patients diagnosed with ‘pouchitis’ are not started on biological or immunosuppressant therapies before cross-sectional imaging to rule out chronic pelvic sepsis.

**Fistulation**

The development of a pouch fistula is a morbid complication associated with a high rate of pouch failure [29,30]. Fistulas may occur between the pouch and perianal region or the pouch and vagina, or may be complex and connect other spaces (such as the bladder or abdominal wall).

There is a wide range of time to presentation of fistulas in the literature, with the average time from IPAA to fistula presentation reported as less than 1 year [31], 2 years [32] and 6.9 years [33]. The incidence of pouch fistula was reported as 4.7% in the 2017 Ileoanal Pouch Registry Report [34]. It is important to identify the aetiology of the fistulizing process as this will affect the principles of management [35].

There is controversy in the aetiological classification of pouch fistula. We have proposed a rational classification based on aetiology [35] (see Figure 2). Previously, these fistulas were commonly thought to arise from an anastomotic leak or CD. A number of features have been suggested to distinguish fistulas arising from CD and anastomotic leak. These features tend to perpetuate the belief that fistulas arising soon after pouch creation or following reversal of ileostomy are secondary to an anastomotic leak and those that present later represent CD (Table 4). This has led to an overdiagnosis of pouch fistula due to CD, with the implication that many patients are treated with biologicals without substantial evidence. Lightner et al [24] retrospectively reviewed pouch histology following pouch excision in patients thought to have complications originating from CD. Only 20% of patients had histological evidence of CD. Of the group with a diagnosis of CD, 46% of cases were diagnosed based on the presence of a fistula. Retrospective review of clinical notes found that 90% had evidence of postoperative septic complications [24]. Aetiological classification of these fistulas should take into account that fistulas may arise from pelvic sepsis years after pouch creation. Most of the pouch fistulas arising from an anastomotic leak (Type 1) originate from the pouch–anal anastomosis, the tip of the J-pouch or the longitudinal staple line. There will be evidence of sepsis on MRI fistula such as peri-pouch sepsis, pelvic or suprarelevator collection [35].

| Aetiology | Diagnosis |
|-----------|-----------|
| Surgical  | Anastomotic leak |
|           | Chronic pelvic sepsis |
|           | Anastomotic fistulas |
| Inflammatory | Primary idiopathic pouchitis |
|           | Prepouch ileitis |
|           | Cuffitis |
|           | Crohn’s disease of the pouch |
| Mechanical | Large or small pouch |
|           | U-bends |
|           | Twists |
|           | Prolapse |
|           | Stricture |
| Functional | Weak sphincter |
| Dysplasia and cancer | Irritable pouch syndrome |
|           | Anismus and functionally impaired defaecation |
|           | Coexisting psychiatric diagnosis |
| Other less likely causes | Pouch cancer |
|           | Bacterial overgrowth |
|           | Coeliac disease |
|           | Thyroid dysfunction |
|           | Pancreatic insufficiency |
|           | Cytomegalovirus or *Clostridium difficile* infection |
|           | Ischaemia |

Table 3: Aetiology and diagnoses underlying the pouch behaving badly

MRI or CT to evaluate the tissues around the pouch and confirm surrounding infection.
Pouch fistulas may rarely be related to inflammatory bowel disease (IBD; Type 2). CD-related fistulas should present with either histology pathognomonic for CD, such as transmural lymphoid aggregates or nonnecrotizing granulomas, or there should be evidence of discontinuous small bowel disease on CT/MRI enterography or pouchoscopy [24,36]. There may be other sequelae of CD, including mouth ulcers. In the absence of features definitive for CD, a fistula may occur in the presence of IBD related to primary idiopathic pouchitis or cuffitis [37].

Type 3 cryptoglandular fistulas may present at any time after pouch creation and will arise with an internal opening at the dentate line [36,38]. Type 4 fistulas arising from a healthy pouch many years following pouch creation should raise suspicions of a fistulizing cancer of the pouch body or anal transition zone, especially in the presence of a pouch created for FAP or in the presence of a UC pouch with historical evidence of dysplasia. Expedient investigation and diagnosis may prevent the requirement for morbid exenterative surgery in these patients.

Essentially, pouch fistulas mainly arise from an anastomotic leak, cryptoglandular disease, malignancy and rarely IBD.

The management of fistulas should be tailored to the aetiology. Type 1 (anastomotic) fistulas should not be treated with biologics.

**TABLE 4** Factors to aid the diagnosis of pouch fistula

| Factors               | Type 1 Anastomotic                                                                 | Type 2 IBD related               | Type 3 Cryptoglandular        | Type 4 Malignant               |
|-----------------------|-----------------------------------------------------------------------------------|---------------------------------|-------------------------------|-------------------------------|
| **Timing**            | Often diagnosed in the postoperative period but may be mistaken for CD or pouchitis years later | Can occur at any time           | Can occur at any time         | May occur years after normal function with an unusual presentation of abdominopelvic pain, anal or vaginal symptoms |
| **Internal opening**  | Internal opening at anastomosis, tip of J or longitudinal staple line              | Internal opening may be anywhere along the pouch body or at the anastomosis | Internal opening at dentate line | Potentially may occur anywhere |
| **Preoperative diagnosis** | Any diagnosis                                                             | CD or indeterminate colitis     | Any diagnosis                   | Risk factors for neoplasm - IBD-related cancer, family history, a long rectal cuff and PSC |
| **Strictures**        | A pouch–anal (outlet) stricture may be present                                   | Discontinuous disease and structuring of the small bowel are suggestive of CD | None                           | Malignant stricture             |
| **Pelvic sepsis**     | History or radiological evidence of pelvic sepsis. May be missed until much later | No history or radiological evidence of pelvic sepsis | Not present | Not present |
| **Small bowel disease** | Not present                                                                   | Small bowel disease on MR enterography | Not present | Not present |
| **Response to biologics** | May become worse                                                               | Responds                        | No response                    | No response                    |
| **Histology**         | Chronic inflammation                                                            | Pathognomonic granuloma in 10%–12% in CD [65] | Chronic inflammation, granulation tissue, stratified epithelium | Neoplastic change              |

**Abbreviations:** CD, Crohn's disease; IBD, inflammatory bowel disease; PSC, primary sclerosing cholangitis.
immunosuppressants and biologicals as this likely to exacerbate chronic pelvic sepsis. Treatment of Type 1 fistulas consists of permanent pouch diversion with an ileostomy, pouch excision or, in selected cases, a redo pouch. Type 2 fistulas may respond to a trial of immunosuppression and biological therapy. This should be offered in the first instance before resorting to permanent pouch diversion or excision, depending upon patient choice. In Type 3 (cryptoglandular fistulas), surgical management should aim to close the fistula tract without compromising continence [33]. Management of Type 4 malignant fistulas is dealt with in the section on neoplasia.

**Inflammatory causes**

**Primary idiopathic pouchitis**

Primary idiopathic pouchitis is a nonspecific inflammatory condition of the ileal pouch with a poorly understood pathophysiology. It is characterized by symptoms of frequency, urgency, incontinence, seepage, abdominal cramping and pelvic pain [39].

Patients may present with extraintestinal manifestations such as arthralgia, and this can aid diagnosis of primary idiopathic pouchitis [40]. There may be an association between PSC and pouchitis [40]. Other points to note in the history include if the patient is a nonsmoker [41], use of nonsteroidal anti-inflammatory drugs (NSAIDs) [41,42] and previous extensive colonic disease [42]. Pouchitis is seen less often after IPAA for FAP than for UC [43,44]. Pouchitis is a common complication, with a cumulative risk of 15.5%, 22.5%, 36% and 45.5% at 1, 2, 5 and 10 years, respectively, after IPAA [45].

Many patients will have single episode of pouchitis, but between 10% and 15% of patients with acute pouchitis will develop chronic pouchitis. This is a poorly defined diagnosis but the most common definition is where the symptoms of pouchitis persist after 4 weeks of treatment [46]. There are further recognized problems with the diagnosis of pouchitis. Firstly, the patient’s symptoms, flexible pouchoscopy findings and the histology findings do not always correlate. Additionally, how pouchoscopy is performed and reported is not consistent [47].

The mainstay of treatment for acute primary idiopathic pouchitis is antibiotics – most commonly ciprofloxacin or metronidazole [48,49]. Chronic primary idiopathic pouchitis is treated with a combination of antibiotics. Patients may require long-term antibiotics that need rotating. The therapeutic benefit of long-term antibiotics must be weighed against the risks of antibiotic resistance and side effects [50]. Oral budesonide and oral beclomethasone have been shown to be effective alternatives to antibiotics in chronic pouchitis [51,52]. They are unsuitable for long-term use due to side effects [53]. Other therapies such as biologicals should be considered. A systematic review of the management of chronic refractory pouchitis found a pooled remission rate of 53% with biological therapy [46].

Importantly, secondary causes of pouchitis should also be considered: infections (such as *Clostridium difficile*, *Campylobacter*, *Salmonella*, *Candida* and cytomegalovirus [54]), pelvic sepsis, CD (with the above caveats noted), faecal stasis, ischaemia or drugs (particularly NSAIDs) [54].

**Prepouch ileitis**

Prepouch ileitis affects 6% of patients with IPAA [55] and usually occurs with primary idiopathic pouchitis [55–57]. It presents with similar symptoms to pouchitis. Diagnosis is with endoscopy of the prepouch ileum or small bowel imaging, and treatment principles are similar to pouchitis. Prepouch ileitis should be differentiated from CD. There is also evidence that prepouch ileitis does not predict CD of the pouch at long-term follow-up [57].

**Cuffitis**

In forming a pouch, a small amount of residual rectum (the ‘cuff’) is retained to which the ileal pouch is anastomosed. In a hand-sewn technique, the rectal mucosa can be excised entirely, although islands of rectal mucosa are left even with this technique (a mucosectomy). Cuffitis is inflammation of the cuff anastomosed to the ileal pouch.

Patients with cuffitis will present with symptoms similar to that of pouchitis, with frequency, urgency and passage of blood in the faeces. Cuffitis can be difficult to diagnose as flexible endoscopy may not visualize the area well; careful digital examination and pouchoscopy can aid diagnosis.

Shen et al [58] treated patients with cuffitis with mesalazine suppositories, with good tolerance and response. In 2013 Wu showed that 33% of all patients with cuffitis responded to 5-ASA or steroid treatment, 18% were dependent on treatment and 48% were refractory to treatment [59]. Importantly these studies were based on small numbers with heterogeneity in the definition of cuffitis and hence should be interpreted with caution.

**Crohn’s disease of the pouch**

CD of the pouch is probably overdiagnosed [24]. It is possible that a pouch is performed with a preoperative diagnosis of CD but it has been estimated that pouch excision rates for this are 45%–55% in patients who have a preoperative diagnosis of CD [60,61]. In those who originally underwent restorative proctocolectomy for presumed UC, 2%–8% had their original diagnosis changed to CD. Importantly, the criteria utilized to diagnose CD are varied. Some studies have defined CD of the pouch as including: inflammation of the pouch that is resistant to antibiotic treatment, strictureting of the afferent limb, strictureting of the small bowel or fistulating disease [62–65].
CD of the pouch should only be considered when there is conclusive histology (i.e. granulomas supporting CD) and/or the presence of characteristic skip lesions in the small bowel. To help understand the likelihood of CD of the pouch, the timing of the CD-like problems of the pouch can often aid diagnosis. As a general rule, fistulas that occur within 2 years of pouch formation are likely to be related to the surgery itself, whereas CD-like changes beyond this may represent CD, but this requires thorough investigation and is less likely. The semantics are often unhelpful for the patient; fistulas and CD-like complications regardless of cause will need a multidisciplinary approach and careful assessment.

Mechanical causes

Stricture

Stricturing of the pouch tends to develop at two locations – the pouch–anal anastomosis and at the pouch inlet between the pre-pouch ileum and pouch itself. Other less common locations include the pouch body (midpouch) and the afferent limb at the previous diverting ileostomy site [66,67]. Sometimes this latter site can be erroneously thought to be a CD-related stricture.

The incidence of pouch strictures has been reported to be as high as 38% [68]. Large retrospective cohorts found early stricture to occur in 5% and late stricture in 11% [12,69].

A systematic review of pouch strictures highlights how the cause of the stricture can be gleaned from the anatomical site. Subsequent management is then tailored in a stepwise fashion. Pouch inlet strictures are likely to be inflammatory or fibrotic. Inflammatory strictures can be treated initially with medical therapy (such as thiopurines or biologicals) and fibrotic strictures can be managed with endoscopic dilatation or surgical approaches [67].

Pouch–anal anastomotic strictures will respond to digital and instrumental dilatation; this can be highly successful but requires repeat dilations [68]. The likely cause is postoperative sepsis or surgical causes such as tension or ischaemia at the anastomosis. Sometimes there is no clear cause [67]. Medena catheters will also allow patients to evacuate at home but patients must be taught how to use them [70].

Midpouch strictures are poorly understood in terms of their aetiology and there is no clear evidence for definitive management. They may be treated with a mixture of endoscopic and surgical approaches [67].

Anal sphincter insufficiency

The anal sphincter should be assessed preoperatively before offering patients IPAA. Incontinence, seepage, frequency and urgency may all be encountered by the patient when there is anal sphincter insufficiency. Damage may occur to the anal sphincter complex at the index operation or during vaginal delivery. Elderly patients are more likely to have anal sphincter insufficiency. This can be confirmed objectively with endoanal ultrasound and anal manometry. Treatment is difficult, but referral for biofeedback can be considered and constipating drugs such as loperamide (titrated according to effect) should be tried. Other options include anal plugs [71] and surgical repair of the external anal sphincter [72]. Sacral nerve stimulation has been attempted for incontinence, with success in a small number of patients [73].

Structural problems

The pouch itself may be poorly constructed and cause the patient difficulty in evacuation and tenesmus. Issues include a pouch that is either too large or too small. Small pouches often cause frequency, and this can be demonstrated with a pouchogram. Further problems include ‘U-bends’, where the linear stapler-cutter has failed to cut, creating a J configuration that is not actually a pouch internally which may obstruct normal defaecation; these can be diagnosed at pouchoscopy or on cross-sectional imaging if the anatomy is unclear. Other structural problems can involve twisting of the pouch [74,75]. Structural problems may be alleviated with a diverting ileostomy first and patients can be offered subsequent pouch excision.

A further problem can be a long cuff and cuffitis (see Figure 3). However, a noninflamed rectum may also cause problems with adequate emptying of the pouch. This is increasingly common in laparoscopic stapled pouches [76].

Neoplasia

Dysplasia can occur in both UC and FAP despite IPAA. The stapled technique leaves a rectal cuff which may undergo chronic inflammation and dysplastic change. Rectal mucosa may also regrow after mucosectomy and hand-sewn anastomosis. Dysplasia may also occur in the pouch body itself or arise from the anal squamous epithelium.

Pouch-related adenocarcinoma is rare in UC. A systematic review of the evidence found the pooled incidence rate to be 0.35% 20 years after IPAA [77]. Neoplasia at colectomy was the strongest risk factor for development of cancer. The incidence of pouch carcinoma at 10 years’ follow-up is 1% in FAP [78].

However, patients with an unusual presentation of abdominopelvic pain, anal or vaginal symptoms or fistula after many years of normal function should be considered for flexible pouchoscopy and cross-sectional imaging. British Society of Gastroenterology guidelines for IBD recommend annual pouchoscopy in certain high-risk groups such as IBD-related cancer, family history, a long rectal cuff and PSC. Otherwise no or 5-yearly surveillance can be undertaken depending on discussions with the patient [48].

Treatment for carcinoma is very difficult. It is challenging to achieve a complete resection (R0) because of disruption to surgical
planes and difficulty in interpreting imaging. Treatment should be in a complex cancer centre and neoadjuvant treatment is recommended.

**Functional causes**

**Anismus**

Anismus is the inappropriate failure of the relaxation of the anal sphincter complex. This is a functional obstruction where the normal coordinated process fails and the patient strains against their own contracted anal sphincter. However, mechanical causes of difficult evacuation should be investigated with clinical examination and treated first. Defaecating pouchography should then be considered if the patient’s symptoms do not resolve. Patients with anismus may benefit from biofeedback [79].

**Irritable pouch syndrome**

All patients should get advice on diet, fluid and electrolyte replacement, constipating medication and barrier creams to optimize function even in the absence of any pathology. However, up to a fifth of patients will have no demonstrable pathology accounting for their symptoms despite clinical, endoscopic, radiological and histological investigations [10]. These patients are given a diagnosis of exclusion: irritable pouch syndrome. Patients with irritable pouch syndrome are more likely to be taking anxiolytics, antidepressants and opioid medication than those with inflammatory pouch conditions or normal pouches [80]. Treatment is aimed at alleviating symptoms with constipating drugs to reduce frequency and urgency. Referral to a dietician to improve the diet, exclude certain foods and alter timing of meals may also help. Biofeedback may have a role [81].

**CONCLUSION**

The badly behaving pouch is a challenging clinical problem to solve for both gastroenterologists and surgeons. The patient may have been on a difficult journey with poor pouch function for many years and will be desperate for improvement. The importance of a clear and detailed history, careful examination and specialist investigation has been demonstrated. There are a number of potential pathologies with overlapping symptoms that can lead to misdiagnosis and potentially dangerous treatments. The importance of patient-centred care is stressed - the aim here is to improve quality of life and that may be achieved with a diverting loop ileostomy.

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**CONFLICT OF INTERESTS**

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MD, JS, GW, OF, SC and AH conceived and designed the review. All authors wrote the paper. All authors critically appraised the paper and edited the final version. AH is the guarantor of the paper.

**REFERENCES**

1. Parks AG, Nicholls RJ Proctocolectomy without ileostomy for ulcerative colitis. Br Med J. 1978;2(6130):85–8. https://doi.org/10.1136/bmj.2.6130.85
2. Derikx LAAP, Nissen LHC, Smits LJ, Shen B, Hoentjen F Risk of neoplasia after colectomy in patients with inflammatory bowel disease: a systematic review and meta-analysis. Clin Gastroenterol Hepatol. 2016;14(6):798–806.e20. https://doi.org/10.1016/j.cgh.2015.08.042
3. Moertel CG, Hill JR, Adson MA Surgical management of multiple polyps: the problem of cancer in the retained bowel segment. Arch Surg. 1970;100(4):521–6. https://doi.org/10.1001/archsurg.1970.01340220197033
4. Bess MA, Adson MA, Elveback LR, Moertel CG. Rectal cancer following colectomy for polyposis. Arch Surg. 1980;115(4):460–7. https://doi.org/10.1001/archsurg.1980.01380040084015
5. Zittan E, Ma GW, Wong-Chong N, Miligrom R, McLeod R s, Silverberg M, et al. Ileal pouch-anal anastomosis for ulcerative colitis: a Canadian institution's experience. Int J Colorectal Dis. 2017;32(2):281–5. https://doi.org/10.1007/s00384-016-2670-y
6. Muller KR, Prosser R, Bampton P, Mountifield R, Andrews JM. Female gender and surgery impact relationships, body image, and sexuality in inflammatory bowel disease: patient perceptions. Inflamm Bowel Dis. 2010;16(4):657–63. https://doi.org/10.1002/ibd.21090
7. Heuschen UA, Hinz U, Allemeyer EH, Lucas M, Heuschen G, Herfarth C. One-or two-stage procedure for restorative proctocolectomy: rationale for a surgical strategy in ulcerative colitis. Ann Surg. 2001;234(6):788–94. https://doi.org/10.1097/00000658-200112000-00010
8. Ikeuchi H, Uchino M, Matsuoka H, Bando T, Takesue Y, Tomita N. Clinical outcomes after restorative proctocolectomy with ileal pouch anastomosis using ultrasonically activated scalpel for ulcerative colitis. Int Surg. 2012;97(3):210–8. https://doi.org/10.9738/CC60.1
9. Mark-Christensen A, Erichsen R, Brandsborg S, Pachler FR, Nørager CB, Johansen N, et al. Pouch failures following ileal pouch-anal anastomosis for ulcerative colitis. Colorectal Dis. 2018;20(1):44–52. https://doi.org/10.1111/codi.13802
10. Ouro S, Thava B, Shaikh I, Clark SK Management of pouch dysfunction in a tertiary centre. Colorectal Dis. 2016;18(12):1167–71. https://doi.org/10.1111/codi.13352
11. Navaneethan U, Shen B Diagnosis and management of pouchitis and ileoanal pouch dysfunction. Curr Gastroenterol Rep. 2010;12(6):485–94. https://doi.org/10.1007/s11894-010-0143-y
12. Fazio VW, Kiran RP, Remzi FH, Coffey JC, Heneghan HM, Kirat HT, et al. Ileal pouch anastomosis: analysis of outcome and quality of life in 3707 patients. Ann Surg. 2013;257(4):679–85. https://doi.org/10.1097/SLA.0b013e31827d99a2
13. Uchino M, Ikeuchi H, Sugita A, Futami K, Watanabe T, Fukushima K, et al. Pouch functional outcomes after restorative proctocolectomy with ileal-pouch reconstruction in patients with ulcerative colitis: Japanese multicenter nationwide cohort study. J Gastroenterol. 2018;53(5):642–51. https://doi.org/10.1007/s00535-017-1389-z
14. Lightner AL, Shogan BD, Mathis KL, Larson DW, Duchalais E, Fletcher JG, Pemberton JH, Mathis KL, Raffals LE, Smyrk T. Crohn’s disease of the pouch: a true diagnosis or an oversubscribed diagnosis of exclusion? Dis Colon Rectum. 2017;60(11):1201–8. https://doi.org/10.1007/D01000000000918
15. Mark-Christensen A, Erichsen R, Brandsborg S, Pachler FR, Nørager CB, Johansen N, et al. Pouch failures following ileal pouch-anal anastomosis for ulcerative colitis. Colorectal Dis. 2018;20(1):44–52. https://doi.org/10.1111/codi.13802
16. Ouro S, Thava B, Shaikh I, Clark SK Management of pouch dysfunction in a tertiary centre. Colorectal Dis. 2016;18(12):1167–71. https://doi.org/10.1111/codi.13352
17. Pavlides M, Cleland J, Rahman M, Christian A, Doyle J, Gaunt R, et al. Pouch excision: indications and outcomes. Colorectal Dis. 2017;19(10):912–6. https://doi.org/10.1111/codi.13673
18. Worley G, Nordenvall C, Askari A, Pinkney T, Burns E, Akbar A, et al. Restorative surgery after colectomy for ulcerative colitis in England and Sweden: observations from a comparison of nationwide cohorts. Colorectal Dis. 2018;20(9):804–12. https://doi.org/10.1111/codi.14113
19. Gaertner WB, Witt J, Madoff RD, Mellowgan A, Finne CO, Spencer MP. Ileal pouch fistulas after restorative proctocolectomy: management and outcomes. Tech Coloproctol. 2014;18(11):1061–6. https://doi.org/10.1007/s10151-012-0885-7
20. Zittan E, Kelly OB, Kirsch R, Milgrom R, Burns J, Nguyen GC, et al. Low fecal calprotectin correlates with histological remission and mucosal healing in ulcerative colitis and colonic Crohn’s disease. Inflamm Bowel Dis. 2015;22(3):623–30. https://doi.org/10.1097/MIB.0000000000000652
21. Li Y, Wu B, Shen B. Diagnosis and differential diagnosis of crohn’s disease of the ileal pouch. Curr Gastroenterol Rep. 2012;14(5):406–13. https://doi.org/10.1007/s11894-012-0282-4
68. Lewis WG, Kuzu A, Sagar PM, Holdsworth PJ, Johnston D. Stricture at the pouch-anal anastomosis after restorative proctocolectomy. Dis Colon Rectum. 1994;37(2):120–5. https://doi.org/10.1007/BF02047532

69. Prudhomme M, Dozois RR, Godlewski G, Mathison S, Fabbro-Peray P. Anal canal strictures after ileal pouch-anal anastomosis. Dis Colon Rectum. 2003;46(1):20–3. https://doi.org/10.1007/s10350-004-6491-7

70. Perry-Woodford ZL, McLaughlin SD. Ileoanal pouch dysfunction and the use of a Medena catheter following hospital discharge. Br J Community Nurs. 2009;14(11):502–6

71. Segal JP, Leo CA, Hodgkinson JD, Cavazzoni E, Bradshaw E, Lung PFC, et al. Acceptability, effectiveness and safety of a Renew® anal insert in patients who have undergone restorative proctocolectomy with ileal pouch-anal anastomosis. Colorectal Dis. 2019;21(1):73–8. https://doi.org/10.1111/codi.14422

72. Glasgow SC, Lowry AC. Long-term outcomes of anal sphincter repair for fecal incontinence: a systematic review. Dis Colon Rectum. 2012;55(4):482–90. https://doi.org/10.1097/DCR.0b013e3182468c22

73. Mege D, Meurette G, Vitton V, Leroy A-M, Bridoux V, Zerbib P, et al. Sacral nerve stimulation can alleviate symptoms of bowel dysfunction after colorectal resections. Colorectal Dis. 2017;19(8):756–63. https://doi.org/10.1111/codi.13624

74. Shen B. Problems after restorative proctocolectomy: assessment and therapy. Curr Opin Gastroenterol. 2016;32(1): https://doi.org/10.1097/MOG.000000000000235

75. Wu X-R, Kiran RP, Mukewar S, Remzi FH, Shen B. Diagnosis and management of pouch outlet obstruction caused by common anatomical problems after restorative proctocolectomy ☆, ☆☆. J Crohn’s Colitis. 2014;8:270–5. https://doi.org/10.1016/j.jcch.2013.08.012

76. Joyce MR, Fazio VW, Hull TT, Church J, Kiran RP, Mor I, et al. Ileal pouch prolapse: prevalence, management, and outcomes. J Gastrointest Surg. 2010;14(6):993–7. https://doi.org/10.1007/s11605-010-1194-y

77. Selvaggi F, Pellino G, Canonicò S, Sciudone G. Systematic review of cuff and pouch cancer in patients with ileal pelvic pouch for ulcerative colitis. Inflamm Bowel Dis. 2014;20(7):1296–308. https://doi.org/10.1097/MIB.000000000000026

78. Friederich P, de Jong AE, Mathus-Vliegen LM, Dekker E, Krieken HH, Dees J, et al. Risk of developing adenomas and carcinomas in the ileal pouch in patients with familial adenomatous polyposis. Clin Gastroenterol Hepatol. 2008;6(11):1237–42. https://doi.org/10.1016/j.cgh.2008.06.011

79. Stellingwerf ME, Maeda Y, Patel U, Vaizey CJ, Warusavitarne J, Bemelman WA, et al. The role of the defaecating pouchogram in the assessment of evacuation difficulty after restorative proctocolectomy and pouch-anal anastomosis. Colorectal Dis. 2016;18(8):O292–300. https://doi.org/10.1111/codi.13431

80. Makkar R, Graff LA, Bharadwaj S, Lopez R, Shen B. Psychological factors in irritable pouch syndrome and other pouch disorders. Inflamm Bowel Dis. 2015;21(12):2815–24. https://doi.org/10.1097/MIB.0000000000000552

81. Segal JP, Chan H, Collins B, Faiz OD, Clark SK, Hart AL. Biofeedback in patients with ileoanal pouch dysfunction: a specialist centre experience. Scand J Gastroenterol. 2018;53(6):665–9. https://doi.org/10.1080/00365521.2018.1454508

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