Management of pouch dysfunction in a tertiary centre

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<u>Abstract</u>

Aim Restorative proctocolectomy (RPC) with ileal pouch anal anastomosis is the operation of choice for ulcerative colitis (UC) and some cases of familial adenomatous polyposis (FAP). Offering improvement in quality of life and high patient satisfaction, pouch surgery is also associated with significant morbidity. The aim of this study was to describe the management of patients referred to a tertiary centre with pouch dysfunction. **Method** All patients referred with pouch dysfunction from other institutions, between October 2006 and November 2014, were included in this retrospective study. Information regarding initial diagnosis prior to pouch procedure, type of procedure, symptoms leading to referral, relation of symptom appearance with ileostomy closure, investigations, final diagnosis, treatment and follow-up was reviewed.

Results One hundred and twenty one patients were included, with RPC mostly for UC (94%), and with diverting ileostomy (83%). The most frequent reasons for referral to our clinic were high defaecatory frequency in 83 (69%) patients, abdominal pain and incontinence in 45 (37%) each and perianal pain in 44 (36%). The principal investigations performed were pouchoscopy in 97 (80%) patients , examination under anaesthesia (EUA) in 62 (51%), pelvic MRI in 56 (46%) and pouchogram in 45 (35%). The commonest diagnoses were pouchitis (primary and secondary) in 24 (21%) patients and anastomotic leak in 26 (22%). After full investigation a cause for the symptoms could not be found in 24 (20%) patients, resulting in the diagnosis of exclusion of 'irritable pouch syndrome' or functional disorder .

Treatments given were long term antibiotic therapy in 29 patients (25%), ileostomy in 19 (16%), use of Medina catheter in 17 (15%) and 12 (10%) underwent dilatation under anaesthetic. Six patients (5%) underwent revision pouch surgery with defunctioning ileostomy and another 6 (5%) had pouch excision.

Conclusion Patients with ileoanal pouch dysfunction often have multiple symptoms. This study shows that a wide range of investigations and treatment modalities need to be available to manage such patients, with a specialised approach in a multidisciplinary setting.

'What does this paper add to the literature'

Although associated with a low operative mortality, RPC has an associated morbidity as high as 30%.

Most series addressing complications of RPC focus on management and outcome of particular pathologies. The aim of this study was to describe the overall approach to investigating patients with pouch dysfunction referred to a tertiary referral centre.

Introduction

Restorative proctocolectomy (RPC) with ileal pouch anal anastomosis is a way of preserving intestinal continuity as an alternative to a permanent ileostomy and is the operation of choice for ulcerative colitis (UC) and some cases of familial adenomatous polyposis (FAP). Despite offering improvement in quality of life and high patient satisfaction, pouch surgery can be associated with a wide range of complications, both early and late.

Although associated with a low operative mortality, RPC has an associated morbidity as high as 30%^(1,2). The most frequent complications include primary pouchitis, anastomotic leak and chronic peripouch sepsis. Other less frequently described complications associated with RPC include pouch vaginal, pouch perineal, pouch urinary or enterocutaneous fistulas, pouch inlet or outlet stenosis, previously unrecognised Crohn's disease, prepouch ileitis, prolapse, retained rectal mucosa and cuffitis, impaired anal sphincter function, small volume pouch, diversion pouchitis, bile salt malabsortion,

irritable pouch syndrome, and pouch malignancy.

The overall pouch failure rate described in the literature is between 3 and $12\%^{(1,2)}$. Various therapeutic options are available for the management of the various causes of pouch dysfunction. We have previously developed an algorithm⁽¹⁾ to guide investigation and management of these challenging patients (Fig 1).

Most series addressing complications of RPC focus on management and outcome of particular pathologies (eg primary pouchitis, chronic sepsis), rather than the overall approach to a patient with pouch dysfunction. ^(3,4,5)

<u>Aims</u>

The aim of this study was to describe the management of patients referred to a tertiary centre with pouch dysfunction.

Methods

We performed a retrospective review of patients referred with pouch dysfunction from other institutions, first seen between October 2006 and November 2014. The information was taken from the patients' hospital records. Data collected included age, gender, initial diagnosis prior to pouch procedure (with or without covering ileostomy), symptoms leading to referral, relationship of symptom appearance with ileostomy closure, investigations, final diagnosis, treatment and follow-up.

<u>Results</u>

One hundred and twenty one were included, 68 women (56%) and 53 men (44%), aged between 17 and 71 years (median 42 years). All had undergone RPC at another institution. The initial diagnosis prior to pouch procedure was UC in 112 patients (93%), FAP in 7 (6%) and indeterminate colitis (IC) in two (1%). One hundred and four (86%) patients initially had a covering ileostomy.

The most frequent reasons for referral to our clinic were high defaecatory frequency in 83 (69%), patients, abdominal pain or incontinence in 45 (37%) each, perianal pain in 44 (36%), difficult evacuation in 33 (27%), bleeding from the anus in 30 (25%) or urgency in 24 (20%). Other less frequent symptoms were also described (Table 1).

The majority of patients reported multiple (up to 9) symptoms (Fig 2).

In the group of patients who had a covering ileostomy at the time of original RPC (104 patients) symptoms of pouch dysfunction occurred within three months of stoma closure in 52 (50%). In those who had initial acceptable pouch function for 3 months after stoma closure, symptoms commenced later, between 3 and 345 months after ileostomy closure, with a median time to appearance of 39 months. In the group of patients who had not had a covering stoma (13 patients) symptoms of pouch dysfunction started between 1 and 216 months (median 1 month) from the initial pouch operation.

The most frequent investigations performed at our institution were pouchoscopy in 97 (80%) patients, examination under anaesthesia (EUA) in 62 (51%), pelvic MRI in 56 (46%), pouchogram in 45 (37%) and barium follow through in 33 (27%) (Table 2).

We found that most patients had more than one final diagnosis. The most frequently encountered were pouchitis (primary and secondary) in 24 (20%) patients and anastomotic leak in 26 (22%). After full investigation a cause for the symptoms could not be found in 24 (20%) patients, resulting in the diagnosis of exclusion of 'irritable pouch syndrome' or functional disorder.

In the anastomotic leak group, 24 (92%) of the leaks were found in the pouch-anal anastomosis and two (8%) in the blind end of the pouch. We identified 12 (10%) patients with chronic peripouch sepsis, 10 caused by anastomotic leaks already included in the former group and two without an identifiable leak. Fourteen (12%) patients had a pouch-vaginal fistula, four from the anastomosis, again already included in the anastomotic leak group. Pouch outlet stenosis was found in 15 (12%) patients and significant retained rectum with and without cuffitis in 10 (8%) each.

Two patients were found to have previously unrecognised malignancies causing outlet stenosis. Other less frequent diagnoses were also identified (Table 3). Most patients had more than one diagnosis.

Seventy five patients (62%) were referred purely with symptoms but 42 (37%) were referred with a diagnosis that was confirmed in 21 (50%). In 21 patients (50%) the final diagnosis was different.

Concerning treatment, the most frequent was long term antibiotic therapy in 29 patients (24%), ileostomy in 19 (16%), use of Medina catheter in 17 (14%) and 12 (10%) underwent dilatation under anaesthetic. Six patients (5%) underwent revision pouch surgery with defunctioning ileostomy and another 6 had pouch excision (5%). Other treatments included seton placement in 8 (7%), prescription of bile salt sequestrants in 8 (7%) and drainage of peripouch sepsis in five (4%), oral immunosupressants and biofeedback in three each (3%), enemas and suppositories (anti-inflammatories or alicaforsen), barrier creams, botulinum toxin injection and advancement flap for fistula in two each (2%) and diltiazem ointment, enterocutaneous fistula repair and fistulotomy in one patient each (1%). Two patients are awaiting pouch excision following neoadjuvant chemotherapy and defunctioning stoma (Table 4).

With regard to follow-up, 18 (15%) patients had only one appointment in our department. Ten of those chose to continue follow-up in their referring hospital, three did not attend the scheduled follow-up appointment and three declined the proposed surgical treatment and were discharged. Two were discharged after one attendance only. Ninety nine (82%) other patients have been followed-up at our institution.

Discussion

Although associated with an improvement in quality of life, RPC can result in significant morbidity. Symptom appearance can be almost immediate, happening in the first three months after ileostomy closure, but can also be delayed and appear many years later. In our cohort, some patients had a very late onset of symptoms, appearing at a median time of 39 months and up to 345 months after ileostomy closure.

There are many symptoms associated with pouch dysfunction and the majority of patients present with several. Due to the overlap of symptoms and complexity of the problems resulting in pouch dysfunction, a number of investigations may be necessary to reach a diagnosis. Likewise, a wide range of heterogeneous pathologies can be associated with pouch dysfunction.

It is interesting to note that, in our series, 50% of those given an initial diagnosis of the cause of their symptoms had that diagnosis changed after full investigation. This reflects the need for a specialised approach with the use of an algorithm of management⁽¹⁾. In this study, 20% of patients remained with the diagnosis of exclusion of 'functional pouch disorder' despite full investigation. We found two cases of unrecognized adenocarcinoma of the pouch outlet and no histologically confirmed cases of Crohn's disease. A wide variety of treatments were employed, ranging from simple pouch evacuation techniques to pouch revision or excision.

This study has a number of weaknesses, particularly referral bias, as many straightforward problems will have been successfully managed at patients' local hospitals, so tertiary referral will not have been required. No meaningful data can be gleaned on the effectiveness of various management strategies, as the symptoms and pathologies identified were so numerous that the number of each is very low.

This series does, however, show the diversity involved in managing pouch dysfunction patients in a tertiary referral centre. A wide range of investigations and treatment modalities need to be available to manage such patients effectively. This is best done in a multidisciplinary setting by a team experienced in managing pouch dysfunction, including surgeons, gastroenterologists, endoscopists, radiologists and pathologists. Given the relatively low numbers of RPC's performed, and the even lower frequency of subsequent problems, it would seem logical that such cases are managed in specialist units.

Fig 1 Algorithm for the investigation of pouch dysfunction



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<u>Fig 2</u> Number of symptoms at presentation



Table 1 Symptoms leading to referral

Symptom	N (%)
High defaecatory frequency	83 (69)
Abdominal pain	45 (37)
Incontinence	45 (37)
Perianal pain	44 (36)
Difficult evacuation	33 (27)
Bleeding from the anus	30 (25)
Urgency	24 (20)
Watery faeces	22 (18)
Mucous anal discharge	18 (15)
Faecal vaginal discharge	17 (14)
Purulent anal discharge	8 (7)
Vomiting	8 (7)
Enterocutaneous fistula	6 (5)
Purulent perianal discharge	5 (4)
Abdominal distension	5 (4)
Pouch prolapse	3 (3)
Weight loss	3 (3)
Pneumaturia/ fecaluria	2 (2)

Table 2 Investigations performed

Investigations	N (%)
Flexible pouchoscopy	97 (80)
EUA	62 (51)
MRI pelvis	56 (46)
Pouchogram	45 (37)
Barium follow through	33 (27)
CT abdomen and pelvis	20 (17)
Anal physiology	14 (12)
MRI enterography	13 (11)
Endoanal ultrasound	9 (7)
Defecating pouchogram	8 (7)
SeHCAT test	6 (5)
Diagnostic laparoscopy	4 (3)
Fistulogram	3 (3)

Table 3 Final diagnosis

Final diagnosis	n (%)
Anastomotic leak	26 (21)
Functional disorder	24 (20)
Primary idiopathic pouchitis	24 (20)
Pouch outlet stenosis	15 (12)
Pouch-vaginal fistula	14 (12)
Chronic peripouch sepsis	12 (10)
Cuffitis	10 (8)
Bile salt malabsortion	8 (7)
Retained rectum without inflammation	7 (6)
Impaired anal sphincter function	6 (5)
Anal fissure	6 (5)
Prepouch ileitis (without pouchitis)	4 (3)
Fistula in ano	4 (3)
Entrocutaneous fistula	4 (3)
Pouch inlet stenosis	3 (3)
Small bowel obstruction	2 (2)
Pouch prolapse / solitary ulcer	2 (2)
Small pouch	1 (1)
Diversion pouchitis	1 (1)
Pouch-urethral fistula	1 (1)

<u>Table 4</u> Treatment

Treatment	N (%)
Long term antibiotics	29 (24)
Defunctioning ileostomy	19 (16)
Medina catheterisation	17 (14)
Dilation under general anaesthesia	12 (10)
Seton placement	8 (7)
Bile sequestrants	8 (7)
Redo pouch	6 (5)
Pouch excision	6 (5)
Drainage of peripouch sepsis	5 (4)
Immunosupressors (oral)	3 (3)
Biofeedback	3 (3)
Dietary change advised	2 (2)
Anti-inflammatory / alicaforsen enema	2 (2)
Barrier creams	2 (2)
Botox injection of sphincter	2 (2)
Advancement flap	2 (2)
Defunctioning and neoadjuvant therapy for	2 (2)
malignancy (pouch excision pending)	
Diltiazem (topical)	1(1)
Enterocutaneous fistula repair	1 (1)
Fistulotomy	1(1)

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