Perianal fistula and the ileoanal pouch – different aetiologies require distinct evaluation

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Abstract

Aim Restorative proctocolectomy has been widely adopted as the procedure of choice for restoring gastrointestinal continuity following proctocolectomy. It is often associated with improved quality of life and high patient satisfaction; however, the development of a pouch anal fistula can cause significant morbidity. Pouch fistulas are notoriously difficult to treat and there is great heterogeneity in the management reported of these fistulas. A lack of classification, and the assumption that fistulas originating from completely different aetiologies will behave and respond similarly to a particular treatment strategy, precludes meaningful comparison of management outcomes. We aim to introduce consistency in the reporting of pouch fistulas using a novel classification system.

Methods A consensus process involving clinicians experienced in the management of pouch fistulas from two high volume tertiary centres was performed.

Results We propose that pouch anal fistulas should be classified into four distinct groups according to their aetiology: group 1, anastomotic related; group 2, inflammatory bowel disease related, with sub-classifications Crohn’s (type A) and non-Crohn’s (type B) in origin; group 3, cryptoglandular related; and group 4, malignancy related.

Conclusion Classification of pouch fistulas according to their aetiology will provide consistency in the literature and improve the quality of prospective evidence for the management of pouch fistulas.

Keywords Ileal pouch anal anastomosis, pouch fistula, pouch anal fistula, pouch vaginal fistula, Crohn’s disease, classification

Introduction

The ileoanal pouch was first described 40 years ago [1]. With a greater number of pouches being created, pouch dysfunction and pouch failure are increasingly being reported; the most common causes are pelvic sepsis and pouchitis [2].

‘Pouch fistula’ comprising either perianal or vaginal fistula (with some publications also including enterocutaneous fistula to the abdominal wall) are commonly seen in inflammatory bowel disease (IBD) referral centres specialized in managing dysfunctional pouches, but discussion in the literature remains limited. There has been an assumption that such fistulas are generally due to either an anastomotic leak, when diagnosed shortly after pouch formation or restoration of intestinal continuity, or Crohn’s disease, either newly diagnosed or a previously known diagnosis [3]. Low heterogeneity amongst the patient population in studies reporting pouch fistulas hinders interpretation of findings and meta-analysis.

Method

The classification proposed is based on the collective experience of treating high volumes of complex pouch patients in our two centres.

Comparison with other methods of classification

The (non-pouch) anal fistula literature is littered with mixed series containing various morphologies, duration and complexities of fistula of cryptoglandular and Crohn’s origin. These are often analysed together and
treated as though they should respond to therapies in a similar way. Even within the Crohn’s fistula literature, a lack of adequate classification has led to such heterogeneity that comparison of therapeutic techniques and outcomes is almost impossible. Similarly, series of rectovaginal fistula repairs often group multiple aetiological causes together including those of obstetric, Crohn’s related, radiation induced and septic (cryptoglandular, Bartholin’s) origin. Mixed case series may produce a success rate which is not true for any of the fistula or cohorts included, and so represents an unhelpful average.

We herein propose a classification that allows separation of pouch fistulas into groups aetiologically distinct from one another and which would therefore, intuitively, benefit from differing treatment strategies.

We have observed four groups of fistulas in the ileoanal pouch.

**Group 1 – Anastomotic dehiscence and pelvic sepsis related**

These fistulas have two crucial identifying features: (i) anastomotic – they arise from the anastomosis; and (ii) timing – they develop shortly after pouch construction or restoration of intestinal continuity if the pouch construction was diverted (Fig. 1a). Anastomotic dehiscence can occur at the pouch anal anastomosis, at the efferent or afferent seams of the pouch or at the apex of the J-pouch. The internal opening of the fistula is identified at the point of the anastomotic separation and may fistulate into surrounding structures including bladder, prostate, urethra and vagina. Peri-pouch sepsis or a presacral collection may also be present [4]. We have also occasionally seen ‘late’ anastomotic fistulas with small or previously unrecognized radiological evidence of anastomotic abnormality on early MRI. Efforts to understand the aetiological cause centre around the risk factors associated with anastomotic leakage. Treatment strategies include early diagnosis of anastomotic leak with immediate intervention, such as an Endo-sponge® [5], or later surgical repair with pouch advancement or reconstruction, given appropriate features such as an adequate distance between the anastomotic opening of the fistula and the dentate line [4].

**Group 2 – Inflammatory bowel disease related**

These fistulas can appear at any time but will generally appear at least 6 months following pouch construction (Fig. 1b). They may arise from the dentate line, the anastomosis, the cuff (perhaps in the presence of

**Figure 1** (a) Fistula arising from an anastomotic dehiscence. (b) Fistula arising from Crohn’s disease in the presence of proximal ileal disease. (c) Fistula related to cryptoglandular disease with an internal opening at the dentate line below the ileal pouch anal anastomosis (IPAA).
cuffitis) or from the pouch itself and are associated with definite features of active inflammation.

We define two subgroups in this category. Type A fistulas are associated with characteristic features on imaging and/or the histology of Crohn’s disease. Crohn’s can be confirmed by findings of discontinuous small bowel disease on enterography or typical granulomas on histological evaluation [3]. The presence of a (non-anastomotic) fistula and a history of ulcerative colitis (UC) is not, as has been suggested by some, sufficient to diagnose Crohn’s disease in our view [6]. Type B fistulas are a distinct subgroup that are not associated with features consistent with Crohn’s disease but are associated with histological characteristics of chronic inflammation in the pouch or cuff.

Aetiological analysis and treatment will mirror those in non-pouch Crohn’s perianal fistulas, perhaps including appropriate reparative/reconstructive procedures, medical treatment or palliative loose seton insertion. Defunctioning and pouch excision may be necessary more often in the presence of active inflammatory disease.

Group 3 – Cryptoglandular related

These fistulas arise from the dentate line rather than the anastomosis and in the complete absence of luminal inflammation (e.g. cuffitis or pouchitis) [7] (Fig. 1c). They appear cryptoglandular in origin but seem to arise at a greater frequency than that seen in the healthy population. The cryptoglandular theory may explain origination but persistence, particularly at a higher rate than in the healthy population, may be related to factors seen in IBD more generally. This is consistent with a recent report that demonstrated that perianal fistulas were more common following anorectal abscess in people with UC than in the healthy population, although less common than in those with Crohn’s disease [8].

The latter and most fascinating group, like those patients with an anal fistula in the presence of UC, may hold the key to understanding the factors leading to persistence of anal fistulas in IBD, since they may share these with anal fistulas arising in Crohn’s disease (and IBD-related fistulas more generally, including those in group 2). This phenotypic overlap probably correlates with a genotypic or perhaps immunological similarity, highlighted previously [9], which predisposes both groups to fistula occurrence and/or persistence.

Group 4 – Malignancy related

Primary malignancy in the ileoanal pouch or cuff may present with a pouch fistula. This should be considered in the absence of an obvious alternative aetiology especially in those with a background of familial adenomatous polyposis or previous dysplasia or cancer in UC. Primary malignancy presenting as a pouch fistula has a poor prognosis if diagnosis is delayed; therefore, imaging, pouchoscopy, examination under anaesthetic and biopsy should be used early and evaluated with a high clinical suspicion in the presence of a new fistula.

Conclusion

Pouch fistulas are an increasingly important topic of investigation. To adequately define optimal treatment practices and investigate aetiological risk factors, a classification system which allows separation of fistulas into cohorts, suitable for further study, is necessary. We herein describe four primary cohorts of fistulas related to pelvic sepsis, IBD, cryptoglandular disease and malignancy in origin as a starting point for consistent investigation in aetiology, risk factors and treatment paradigms. This will strengthen the results of any future investigation.

Classification of pouch anal and pouch vaginal fistula

Conflicts of interest

The authors have no conflicts of interest to disclose.

Author contributions

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References

1 Parks AG, Nicholls RJ. Proctocolectomy without ileostomy for ulcerative colitis. *Br Med J* 1978; 2: 85–8.

2 Worley GHT, Fearnhead NS, Brown SR, Acheson AG, Lee MJ, Faiz OD. Review of current practice and outcomes following ileoanal pouch surgery: lessons learned from the Ileoanal Pouch Registry and the 2017 Ileoanal Pouch Report. *Color Dis* 2018; 1: 913–22.

3 Lightner AL, Pemberton JH, Loftus EJ. Crohn’s disease of the ileoanal pouch. *Inflamm Bowel Dis* 2016; 22: 1502–8.

4 Mallick IH, Hull TL, Remzi FH, Kiran RP. Management and outcome of pouch-vaginal fistulas after IPAA surgery. *Dis Colon Rectum* 2014; 57: 490–6.

5 Rottoli M, Di Simone MP, Vallicelli C et al. Endoluminal vacuum-assisted therapy as treatment for anastomotic leak after ileal pouch–anal anastomosis: a pilot study. *Tech Coloproctol* 2018; 1: 223–9.

6 Barnes EL, Kochar B, Jessup HR, Herfarth HH. The incidence and definition of Crohn’s disease of the pouch: a systematic review and meta-analysis. *Inflamm Bowel Dis* 2019; 20: 1474–80.

7 Gaertner WB, Witt J, Madoff RD, Mellgren A, Finne CO, Spencer MP. Ileal pouch fistulas after restorative proctocolectomy: management and outcomes. *Tech Coloproctol* 2014; 26: 1061–6.

8 Sahnan K, Askari A, Adegbola SO et al. Persistent fistula after anorectal abscess drainage: local experience of 11 years. *Dis Colon Rectum* 2019; 62: 327–32.

9 Tozer PJ, Lung P, Lobo AJ et al. Review article: pathogenesis of Crohn’s perianal fistula – understanding factors impacting on success and failure of treatment strategies. *Aliment Pharmacol Ther.* 2018; 48: 260–9.