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Outcomes after ileal pouch anal anastomosis in patients with primary sclerosing cholangitis☆

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Abstract

Background and aims: Outcomes after ileal pouch anal anastomosis (IPAA) are not well established in patients with primary sclerosing cholangitis (PSC). We conducted a comprehensive outcomes assessment in these patients.
Methods: A retrospective case note review of complications in all PSC-IPAA (n = 21) and matched ulcerative colitis patients with IPAA (UC-IPAA; n = 79) after surgery in Oxford (1983–2012) was conducted, and functional outcomes (Öresland score) were evaluated (2012). Quality of life [Cleveland Global Quality of Life Questionnaire, Short Form-36 (SF-36)], and sexual function were also assessed (2012) including patients with PSC-associated UC without IPAA (PSC-UC; n = 19). Sub-group analysis of patients with large duct (*ld*) PSC-IPAA (n = 17) was also performed.

Abbreviations: IPAA, ileal pouch anal anastomosis; PSC-IPAA, patients with PSC who have undergone IPAA; *ld*PSC-IPAA, patients with large duct PSC who have undergone IPAA; UC-IPAA, patients with UC and without PSC who have undergone IPAA; PSC-UC, patients with PSC and UC who have not undergone IPAA; CGQOL, Cleveland Global Quality of Life score; SF-36, Short Form 36; FSFI, Female Sexual Satisfaction Index; IIEF, International Index of Erectile Function; SF-36, summary scores; PF, physical functioning; RP, role limitation due to physical problems; BP, bodily pain; GH, general health; VT, vitality and energy; SF, social functioning; RE, role limitation due to emotional problems; MH, mental health; PCS, physical health summary score; MCS, mental health summary score.

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Results: The 1-, 5-, 10- and 20-year risk of acute pouchitis for PSC-IPAA was 10%, 19%, 31% and 65% respectively, compared to 3%, 10%, 14% and 28% in UC-IPAA (p = 0.03). More PSC-IPAA (36%) had poor nocturnal pouch function (vs 2% in UC-IPAA; p = 0.0016). There were no differences in surgical complications, quality of life or sexual function between the 3 main groups. *Ld*PSC-IPAA had poorer pouch function (Öresland score: 7.7 vs 5.4 in UC-IPAA; p = 0.02), and worse quality of life [SF-36 Physical: 42 vs 50.5 in UC-IPAA; 47.7 in PSC-UC; p = 0.03 and Mental Health summary scores: 41.6 vs 51.2 in UC-IPAA; 42.3 in PSC-UC; p = 0.04].
Conclusions: PSC-IPAA suffer more acute pouchitis and have worse functional outcomes than UC-IPAA. *Ld*PSC-IPAA also have poorer quality of life.

1. Introduction

Primary sclerosing cholangitis (PSC) is an immune mediated cholestatic disease characterised by inflammation and fibrosis of the bile ducts.[1] It is closely associated with inflammatory bowel disease, particularly ulcerative colitis (UC). The prevalence of UC in patients with PSC is as high as 75% in Northern European cohorts.[2] PSC incurs an increased risk of colon dysplasia and cancer[3] and the prevalence of PSC is higher in patients with UC undergoing colectomy when compared to those who do not need surgery.[2] Patients with PSC may need to undergo colectomy because of medically-refractory UC or to treat colonic dysplasia and neoplasia. The operation of choice for the management of UC in these patients is a restorative proctocolectomy with ileal pouch anal anastomosis (IPAA). This operation is safe and cures the patients of the intestinal manifestations of their disease.

Post-operative complications including pouchitis can occur, but on the whole, patients with UC undergoing IPAA have good outcomes and quality of life.[4] However, there are conflicting reports in the literature on the impact of PSC on post-operative functional outcomes.[2,5–8] Furthermore, there are limited data on the effect of PSC on the quality of life and sexual function in patients who undergo IPAA for UC.

The aim of this study was to investigate complications, functional outcomes, quality of life and sexual function in patients with PSC associated UC who have undergone IPAA.

2. Patients and methods

2.1. Definitions

2.1.1. Ulcerative colitis (UC)

The diagnosis of UC was based on recognised endoscopic, histological and clinical features.[9] The extent of the disease was described according to the Montreal classification (proctitis: disease limited to the rectum, left sided: disease distal to the splenic flexure and extensive: disease extends proximal to the splenic flexure).[10]

2.1.2. Primary sclerosing cholangitis (PSC)

The diagnosis of PSC was based on characteristic findings on cholangiography (ERCP or MRCP) and histological features on liver biopsy.[11] The presence of cholangiographic findings alone was enough to diagnose large duct PSC (*ld*PSC). Patients were considered to have small duct PSC if they had a normal cholangiogram but the clinical presentation, biochemistry and histology findings were compatible with PSC.

2.1.3. PSC-IPAA

These were patients who were diagnosed with PSC associated with UC and had undergone colectomy and IPAA. The diagnosis of PSC could have been made at any point during the follow-up period.

2.1.4. UC-IPAA

These were patients who were diagnosed with UC (but not PSC) and had undergone colectomy and IPAA.

2.1.5. PSC-UC

These were patients who were diagnosed with PSC and UC but had not undergone colectomy and IPAA.

2.1.6. Pouch dysfunction

This was defined as any episode of symptoms (including diarrhoea, blood in the stool, urgency, abdominal pain, fever) that raised clinical concerns and led to further investigations or treatments. Pouch dysfunction may be the result of many underlying pathologies including surgical complications (e.g. anastomotic stenosis, pouch related pelvic sepsis), Crohn's disease of the pouch, cuffitis (inflammation of the mucosa in the remaining rectal stump due to the underlying UC), irritable pouch syndrome and pouchitis.[12]

2.1.7. Acute pouchitis

This was defined as any episode of pouch dysfunction with endoscopic and histological evidence of acute pouchitis. The presence of any of mucosal oedema, granularity, friability, loss of vascular pattern, exudates and ulceration[13] was considered to be endoscopic evidence of acute pouchitis. Pouch biopsies were assessed according to the system reported by Shepherd et al. in 1987.[14] Briefly, this score evaluates the degree of acute (0–6) and chronic (0–6) inflammatory changes in the pouch mucosa. For the assessment of acute inflammation the extent of mucosal infiltration by neutrophils (0: none to 3: severe with crypt abscesses) and the degree of ulceration (0: none to 3: extensive) are evaluated and a total acute score of ≥ 4 is considered to be histological evidence of acute pouchitis.

2.2. Patient cohorts

PSC-IPAA who had surgery between 1983 and 2012 were identified. This group was matched to UC-IPAA who had