Diagnosis and management of pouch outlet obstruction caused by common anatomical problems after restorative proctocolectomy☆☆☆

Xian-rui Wu a, Ravi P. Kiran a, Saurabh Mukewar b, Feza H. Remzi a, Bo Shen b,∗

a Department of Colorectal Surgery, The Cleveland Clinic Foundation, Cleveland, OH, USA
b Department of Gastroenterology/Hepatology, The Cleveland Clinic Foundation, Cleveland, OH, USA

Received 26 March 2013; received in revised form 24 August 2013; accepted 25 August 2013

Abstract

Background and aims: Efferent limb syndrome (ELS) after S pouch and pouch-rectal anastomosis (PRA) after J pouch are common anatomical problems after restorative proctocolectomy that lead to pouch outlet obstruction. This study was aimed to evaluate the frequency, diagnosis and management of ELS and PRA.

Methods: Consecutive patients diagnosed with ELS or PRA at our Pouch Center from 2002 to 2011 were included. Demographic, clinical, endoscopic, and radiographic features together with its management and outcomes were studied.

Results: A total of 26 patients met the inclusion criteria, 17 (65.4%) were male. Eleven patients (42.3%) had ELS and 15 (57.7%) had PRA. The median length of the efferent limb/rectal stump for all patients was 6.0 (interquartile range [IQR]: 5.0–8.8) cm, 7.0 (IQR: 5.0–9.0) cm and 6.0 (IQR: 5.0–10.5) cm for S and J pouch patients, respectively (P = 0.025). Dyschezia (n = 15, 57.7%) was the most common presenting symptom, followed by bloating (n = 9, 34.6%), abdominal pain (n = 9, 34.6%), the sense of incomplete evacuation (n = 7, 26.9%) and perianal discomfort (n = 3, 11.5%).

Abbreviations: ATZ, anal transitional zone; CD, Crohn’s disease; CARP, chronic antibiotic-refractory pouchitis; EIM, extra-intestinal manifestations; ELS, efferent limb syndrome; FAP, familial adenomatous polyposis; IBD, inflammatory bowel disease; IPAA, ileal pouch-anal anastomosis; IQR, interquartile ranges; IRB, institutional review board; NSAID, non-steroidal anti-inflammatory drugs; PDAI, the Pouchitis Disease Activity Index; PRA, pouch-rectal anastomosis; QOL, quality of life; SD, standard deviations; TNF, tumor necrosis factor; UC, ulcerative colitis.

Author contributions: XRW contributed to study concept and design, acquisition, analysis and interpretation of data, and drafting of the manuscript. RPK, SM, FHR and BS contributed to study concept and design, analysis and interpretation of data and critical revision of the manuscript for important intellectual content. All authors read and approved the final manuscript.

Grant support: This project was partially supported by the Ed and Joey Story Chair (to. B.S.).

Meeting: Presented to a meeting of the American Society of Colon and Rectal Surgeons, Phoenix, Arizona, April 27 to May 1, 2013.

∗ Corresponding author at: Department of Gastroenterology/Hepatology-A31, The Cleveland Clinic Foundation, 9500 Euclid Ave, Cleveland, Ohio 44195, USA. Tel.: +1 216 444 9252; fax: +1 216 444 6305.

E-mail address: shenb@ccf.org (B. Shen).

1873-9946/$ - see front matter © 2013 European Crohn’s and Colitis Organisation. Published by Elsevier B.V. All rights reserved.
http://dx.doi.org/10.1016/j.crohns.2013.08.012
1. Introduction

Restorative proctocolectomy with ileal pouch-anal anastomosis (IPAA) is the standard surgical treatment for most UC patients who do not respond to medical therapy or develop neoplasia, and for a majority of patients with familial adenomatous polyposis (FAP). This bowel-anatomy-altering surgery substantially reduces the risk for colitis-associated neoplasia and significantly improves patients’ health-related quality of life (QOL) by preserving the natural route of defecation.1–4 However, the construction of the ileal pouch reservoir is often associated with various complications.

J and S pouches are two most commonly used configurations. The anatomy of a three-limb S and a two-limb J pouch is different. The efferent limb of an S pouch is directly linked to the anal transitional zone (ATZ), while the efferent limb of a J pouch is sealed with the “tip” of the J. Efferent limb syndrome (ELS), a rare condition after the creation of ileal pouch, is characterized by the presence of excessively long pouch outlet, leading to complete or partial obstruction of the pouch outlet. It typically occurs in S pouch patients with a dysfunctional or excessively long efferent limb. However, similar symptoms might also exist in J pouch patients with a pouch-rectal anastomosis (PRA) caused by a long rectal stump.5 ELS (S pouch) and PRA (J pouch) are two common anatomical problems after restorative proctocolectomy causing pouch outlet obstruction, due to an elongated pouch outlet. Data relating to the disease course, diagnosis and management of ELS and PRA is scant beyond case reports.6–8 The aim of this study was to evaluate the frequency, diagnosis and management of ELS (S pouch) and PRA (J pouch).

2. Patients and methods

2.1. Patients

This study was approved by the Cleveland Clinic Institutional Review Board (IRB). All eligible ileal pouch patients with ELS or PRA being followed up at our Pouch Center from 2002 to 2011, were identified from the IRB-approved Registry. The subspecialty Pouch Center is a national and international referral center for various ileal pouch disorders, with about 67% of patients coming from outside of the State of Ohio. Demographics, clinical, endoscopic, and radiographic features together with the management and outcomes were all prospectively maintained in the Registry. Both paper charts and electronic medical records would be carefully reviewed should data be missing.

2.2. Inclusion and exclusion criteria

In order to qualify for the study, patients needed to meet all the following inclusion criteria for having: (1) an ileal pouch; (2) a diagnosis of ELS or PRA; (3) regular follow-up at the Pouch Center; and (4) underlying inflammatory bowel disease (IBD). Ileal pouch patients with underlying FAP or other bowel diseases were excluded from this study. Patients with the continent ileostomy or Kock pouch were also excluded.

2.3. Diagnostic criteria

ELS was defined as the presence of a dysfunctional or excessively long efferent limb in S pouch patients, while PRA was defined as the presence of a long rectal stump in patients with J pouch-rectal anastomosis, with main symptoms related to the partial obstruction of the pouch outlet (Fig. 1).5 Both ELS and PRA were diagnosed based on a combined assessment of symptoms, endoscopic, and/or radiographic features.

2.4. Patient groups

All the patients were divided into two groups based on the configuration of the pelvic ileal pouch: the ELS (S pouch) and PRA (J pouch) groups.

2.5. Definitions of variables

Chronic antibiotic-refractory pouchitis (CARP)—defined by a modified Pouchitis Disease Activity Index ≥ 5 points and symptoms lasting 4 weeks or more and failed to respond to a 4-week course of single antibiotic therapy (ciprofloxacin, metronidazole, or tinidazole);9 cuffitis—endoscopic and histologic inflammation of the rectal cuff; Crohn’s disease (CD) of the pouch—diagnosed based on our previously published criteria,10 i.e., the presence of non-surgery-related perianal fistula or inflammation or ulcerations at the pre-pouch neo-terminal ileum or small bowel in the absence of non-steroidal anti-inflammatory drug (NSAID) use, or granulomas on histology; afferent limb syndrome—distal small bowel obstruction caused by an acute angulation, prolapse, or intussusception of the afferent limb at the junction to the pouch, in the absence of intraluminal mucosa associated strictures.5
2.6. Outcome measurement

The primary outcome of this study was to assess the disease course, diagnosis and management of ELS (S pouch) and PRA (J pouch), two common anatomical problems causing pouch outlet obstruction after restorative proctocolectomy.

2.7. Statistical analysis

Descriptive statistics were computed for all variables. These included means and standard deviations (SD) or medians and interquartile ranges (IQR) for continuous factors, and frequencies for categorical factors. Comparisons between two groups were made by using the 2-tail t test (or Wilcoxon rank sum test as appropriate) for continuous variables and chi-square test (or the Fisher exact test as appropriate) for categorical variables. All statistical analyses were performed with the SPSS software version 16 (SPSS, Chicago, IL). P value less than 0.05 was considered statistically significant.

3. Results

3.1. Patient demographics

There were a total of 26 patients who met the inclusion criteria (Fig. 1), including 11 patients (42.3%) who had ELS and 15 (57.7%) who had PRA. During the study period, the Registry from the Pouch Center enrolled 44 S pouch patients and 1195 J pouch patients. Therefore, ELS (25.0%, 11/44) is more frequently encountered in S pouch patients than PRA (1.3%, 15/1195) in J pouch patients (P < 0.001).

All the 26 patients underwent IPAA surgery for underlying UC. Male patients accounted for 65.4% (n = 17) of the whole cohort. The mean age at the diagnosis of UC and at colectomy was 26.2 ± 12.8 years and 31.7 ± 13.1 years, respectively. The indications for colectomy were colitis-associated neoplasia in 1 patient (3.8%) and medically refractory disease in 25 (96.2%). Twenty-three patients (88.5%) had extensive colitis and 3 (11.5%) had left-sided colitis. Eight patients (30.8%) had at least one EIM and only one (3.8%) had PSC. One patient (3.8%) had 1-stage IPAA surgery, 22 (84.6%) had 2-stage surgery and the remaining 3 (11.5%) had 3-stage surgery. Nineteen patients (73.1%) had other concurrent pouch complications, including cuffitis (n = 6, 23.1%), CARP (n = 5, 19.2%), pouch sinus/fistula (n = 5, 19.2%), pouch strictures (n = 5, 19.2%), CD of the pouch (n = 3, 11.5%) and afferent limb syndrome (n = 2, 7.7%). Although a statistical significance was not reached, there was a trend that more patients with PRA suffered from cuffitis than ELS patients (33.3% vs. 9.1%, P = 0.2). The distribution of other concurrent pouch complications was comparable between the two groups (P ≠ 0.05). The majority of the ELS/PRA patients (84.6%, n = 22) had their primary IPAA constructed at outside institutions. There were no differences in other demographic features between the ELS and PRA groups (P ≠ 0.05) (Table 1).

3.2. Clinical presentation and diagnosis

The median length of the efferent limb/rectal stump for all patients was 6.0 (interquartile range [IQR]: 5.0–8.8) cm, 7.0 (IQR: 5.0–9.0) cm and 6.0 (IQR: 5.0–10.5) cm for S and J pouch patients, respectively (P = 0.025). All patients (100.0%) suffered from symptoms presumably due to obstruction of the pouch outlet. Dyschezia (n = 15, 57.7%) was the most common symptom, followed by bloating (n = 9, 34.6%), abdominal pain (n = 9, 34.6%), incomplete evacuation (n = 7, 26.9%) and perianal discomfort (n = 3, 11.5%). A greater proportion of ELS patients had dyschezia compared to PRA patients (33.3% vs. 9.1%, P = 0.2). The distribution of other concurrent pouch complications was comparable between the two groups (P > 0.05). The majority of the ELS/PRA patients (84.6%, n = 22) had their primary IPAA constructed at outside institutions. There were no differences in other demographic features between the ELS and PRA groups (P > 0.05) (Table 1).
patients (7.7%), radiographic examination did not show ELS/PRA despite the findings at pouchoscopy. In 7 patients (26.9%), the efferent limb/rectal stump obstruction was diagnosed with both radiographic examination and pouchoscopy.

### 3.3. Management and outcomes

A total of 15 ELS/PRA patients (57.7%) needed surgical management. The requirement for surgical management was 90.9% (n = 10) in ELS patients versus 33.3% (n = 5) in PRA (P = 0.005) (Table 3). Of the 15 patients, 6 (23.1%) underwent a redo J pouch construction with efferent limb/rectal stump excision, 2 (7.7%) had a redo S pouch construction with efferent limb/rectal stump excision, 2 (7.7%) had a diverting ileostomy as the first stage of surgical management, 3 (11.5%) had pouch excision with end ileostomy and another 2 patients (7.7%) required surgical management but were lost to follow-up. The reasons for pouch excision in the 3 patients were unsatisfactory and dysfunctional IPAA (n = 1, 3.8%), inability for reach of the ileal pouch to the anal canal (n = 1, 3.8%) and concurrent pouch sinus with suspected pelvic sepsis (n = 1, 3.8%).

After a mean follow-up of 3.4 ± 1.4 years, 1 (12.5%) of the 8 patients, who underwent pouch reconstruction with efferent limb excision, developed ischemic pouchitis and subsequently required pouch excision. The remaining 7 patients (87.5%) maintained functional ileal pouches at the last follow-up, of whom 2 (28.6%) patients developed strictures at the new anastomotic site but were successfully treated with endoscopic balloon dilation. Improvement in symptoms related with obstruction of pouch outlet was reported by all the 7 patients.

### 4. Discussion

When restorative proctocolectomy with IPAA was initially introduced to treat UC patients by Parks in 1978,¹¹ the ileal pouch reservoir was fashioned with an S configuration. However, the J pouch has quickly evolved as the most commonly performed reservoir considering that it is relatively technically easier to construct and associated with improved clinical outcomes.¹²–¹⁴ In the initial design of an S pouch construction, the reservoir was deliberately connected to the anus by a long conduit of ileum, termed the efferent ileal limb, to minimize the risk of incontinence.¹¹ With time, this was found to be unnecessary and it became apparent that the efferent limb was itself often responsible for dysfunction, especially difficulty in evacuation.⁶ In the clinical practice at our Pouch Center, we have also noticed that a small proportion of J pouch patients with a long rectal stump often suffered from similar symptoms to those seen in S pouch patients with a long efferent limb.
Since the identification of the association between pouch dysfunction and a long efferent limb or a long rectal stump, the ileal pouch reservoir was redesigned to leave the exit conduit of the pouch not greater than 2.2.5 cm in length.5,15,16 Patients with ELS (S pouch) or PRA (J pouch) were rarely encountered in clinical practice. In the literature, there are limited data on the disease course, diagnosis and management of ELS (S pouch) and PRA (J pouch) patients. In 1990, Nicholls et al.6 evaluated 6 ELS patients who required surgical correction. They concluded that excessive length of the efferent limb of an S ileal reservoir could cause unsatisfactory defecation and this might be improved by resection of the efferent limb and re-anastomosis. In 1996, the same group reported 11 ELS patients out of 16 who had ileal outlet mechanical obstruction, and had a similar finding with their previous study.7 However, none of the above studies compared the diagnosis and management between S (i.e. ELS) and J (i.e. PRA) pouch patients.

In our cohort of 26 patients, 11 had ELS and 15 had PRA. S pouch patients had a greater likelihood of suffering from ELS than J pouch patients suffer from PRA, which was consistent with the notion that ELS typically occurs in S pouch patients but PRA is a rarely recognized disease entity.6 However, the numbers are likely higher than those in the general population as our Pouch Center is a referral center for patients with pouch problems. The median length of efferent limb/rectal stump for all patients was 6.0 (5.0–8.8) cm. This was much longer than that expected in patients with a normal pouch, where the exit conduit of the pouch is expected to be around 2 cm in length.5,15,16

Table 2: Symptoms and diagnosis.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All cases</th>
<th>ELS</th>
<th>PRA</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median length of efferent limb/rectal stump (interquartile range), cm</td>
<td>6.0 (5.0–8.8)</td>
<td>7.0 (5.0–9.0)</td>
<td>6.0 (5.0–10.5)</td>
<td>0.025</td>
</tr>
<tr>
<td>Dyschezia</td>
<td>15</td>
<td>10 (90.9%)</td>
<td>5 (33.3%)</td>
<td>0.005</td>
</tr>
<tr>
<td>Bloating</td>
<td>9</td>
<td>5 (45.5%)</td>
<td>4 (26.7%)</td>
<td>0.42</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>9</td>
<td>2 (18.2%)</td>
<td>7 (46.7%)</td>
<td>0.22</td>
</tr>
<tr>
<td>Incomplete evacuation</td>
<td>7</td>
<td>5 (45.5%)</td>
<td>2 (13.3%)</td>
<td>0.10</td>
</tr>
<tr>
<td>Perianal discomfort</td>
<td>3</td>
<td>0 (0%)</td>
<td>3 (20.0%)</td>
<td>0.24</td>
</tr>
</tbody>
</table>

Table 3: Surgical management.

<table>
<thead>
<tr>
<th>Requirement of surgical management (intention-to-treat)</th>
<th>All cases</th>
<th>ELS</th>
<th>PRA</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Redo J pouch</td>
<td>6</td>
<td>5</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Redo S pouch</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Pouch excision</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Diverting ileostomy as the first stage of surgical management</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Need surgical management, but lost to follow-up</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>
There are a few limitations to our study, particularly relating to the small number of patients. Since all the patients in the current study were seen in the setting of a subspecialty Pouch Center, referral bias is inevitable. Our Pouch Center as a national and international referral center virtually covers patients with a whole spectrum of pouch disorders as well as normal pouch patients. Data on QOL was also not obtained. However, QOL scores after surgical correction should not be expected to be lower than those before surgery as suggested by the previous reports.

In conclusion, based on the results from a specialized Pouch Center that deals with patients with pouch disorders, outlet dysfunction may occur after restorative proctocolectomy after both S and J pouch creation. When comparing ELS after S pouch creation and PRA after J pouch, the former is more likely to be associated with severe symptoms and hence the need for revisional surgery.

Conflict of interest
Disclosure: The authors declared no financial conflict of interest.

Acknowledgment
Dr. Xian-rui Wu is a research fellow from the Department of Colorectal Surgery, the Sixth Affiliated Hospital, Sun Yat-sen University, Guangzhou, China. The study is supported by the Ed and Joey Story Endowed Chair (to B.S.).

References