Pathology of the Ileoanal Pouch
Marco Novelli

Overview

• Clinical aspects:
  - What is an ileoanal pouch?
  - Which patients get ileoanal pouches?
  - Function of ileoanal pouches.
  - Complications of pouch surgery.

• Pathological aspects
  - Anatomy of the anus.
  - Histopathology of the ileoanal pouch.
  - Reporting of pouch biopsies.

What is an ileoanal pouch?

Restorative Proctocolectomy
Pouch Configuration

Proctocolectomy without ileostomy for ulcerative colitis
S S PARKS, W J NICHOLLS
Arch Dis Child. 1993; 68: 40-8

Summary and conclusions
An operation has been developed that people need not undergo and that avoids the need for a permanent ileostomy. The ileoanal pouch is the remaining rectal mucosa incorporated into the pouch. A temporary stoma is made. One of eight patients so treated, four were available for follow-up, and four of these were followed up with asymptomatic disease. The remaining three were asymptomatic and had no evidence of pouch disease.

Introduction
Excessive reliance on supportive surgery, and its morbidity, has led to the development of operation. The aim of the operation is to restore function to the rectum and to create a permanent anal sphincter. This is achieved by dividing the rectum and creating a pouch from the anal canal.

Restorative Proctocolectomy
Pouch Configuration

J S W

Parkà and Nichollà pouch
Restorative Proctocolectomy
Pouch Configuration

How much is the volume increased?

Ulcerative Colitis
Restorative Proctocolectomy

Which patients get ileoanal pouches?

- Familial adenomatous polyposis
- Ulcerative colitis
- Indeterminate colitis
- Crohn's disease
- Colon cancer
Which patients get ileoanal pouches?

- Familial adenomatous polyposis
- Ulcerative colitis
- Indeterminate colitis
- Crohn's disease
- Colon cancer

Reconstruction After Colectomy

The Ileoanal Pouch

Parks AG & Nicholls RJ
BMJ 1978; 2:65-8

'Gold Standard'
Ulcerative colitis, FAP

Benefits
- Normal route of defecation
- Good functional outcomes

Restorative Proctocolectomy

Functional Outcome

'Pouches convert a colitic with a colon to a colitic with a pouch'

Restorative Proctocolectomy

Functional outcomes

<table>
<thead>
<tr>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sepsis</td>
</tr>
<tr>
<td>5% - 27%</td>
</tr>
<tr>
<td>Obstruction</td>
</tr>
<tr>
<td>5% - 22%</td>
</tr>
<tr>
<td>Anastomotic</td>
</tr>
<tr>
<td>Stricture</td>
</tr>
<tr>
<td>5% significant</td>
</tr>
<tr>
<td>Pouchitis</td>
</tr>
<tr>
<td>5% - 50%</td>
</tr>
<tr>
<td>Failure</td>
</tr>
<tr>
<td>2% - 10%</td>
</tr>
</tbody>
</table>
Restorative Proctocolectomy Complications

- Sepsis: 5% - 27%
- Obstruction: 9% - 22%
- Anastomotic Stricture: 5% significant
- Pouchitis: 5% - 50%
- Failure: 2% - 10%

Pouchitis predominantly effects pouches of patients with UC.

Anal canal

- Surgical definition: Level of pelvic floor (anorectal ring) to the anal opening ~ 4cm.
- Histological: Upper to lower border of internal anal sphincter ~ 3cm
Anal cuff

- Shortened crypts.
- Mild crypt architectural distortion.

Restorative Proctocolectomy

Stapled versus handsewn anastomosis

<table>
<thead>
<tr>
<th>Schluender SJ</th>
<th>86 vs 98</th>
<th>Meta analysis (4 RCT’s)</th>
<th>Similar anal physiology</th>
<th>Similar functional outcomes</th>
</tr>
</thead>
</table>

Cuffitis (UC involving the anal cuff)

Anal cuff with Paneth cell metaplasia

Pouch Dysfunction

Differential diagnosis

- Primary pouchitis
- Secondary pouchitis
  - pelvic sepsis
  - pouch-anal fistula
  - anastomotic stricture
- Cuffitis
- Crohn’s disease (5%)
- Pouch ischaemia
- Enteric infection
- Functional disorders
  - overflow incontinence
  - delayed emptying
  - irritable pouch syndrome
**Pouchitis**

- Much commoner in patients with UC.
- Almost certainly related to bacterial flora / overgrowth.
- Treated with antibiotics
  - Ciprofloxacin
  - Metronidazole
  - Steroid enemas
  - Probiotics

**Presentation and Diagnosis**

**Pouchitis**

- **Symptoms**
  - Frequency
  - Urgency
  - Incontinence
  - Anorexia / fever
  - Extra-IM

- **Diagnosis**
  - Symptoms
  - Endoscopic Findings
  - Histopathology

**Differential Diagnosis**

- Secondary pouchitis
- Sepsis pelvic
- pouch-rectal fistula
- Anastomotic stricture
- Cuffitis
- Functional disorders
- Irritable pouch syndrome
- Enteric Infection
- Crohn's disease (5%)

**Histopathology of the ileoanal pouch**
Granular pigment in TI useful marker

Adaptive changes
- Villous blunting / mild crypt hyperplasia
- Increased chronic inflammatory cell infiltrate
- Patchy mild acute inflammation
- (Colonic metaplasia)

Adaptive type changes

Pouches in FAP
- Pouchitis is uncommon ~ 5%
- Look out for adenomas

Pouchitis
- Villous flattening / crypt hyperplasia
- Active chronic inflammation
- Ulceration
- Crypt architectural distortion
- Ulcer-associated cell lineage

Pouchitis
- Villous flattening / crypt hyperplasia
- Active chronic inflammation
- Ulceration
- Crypt architectural distortion
- Ulcer-associated cell lineage
Consistent with a clinical diagnosis of pouchitis

Consistent with a clinical diagnosis of pouchitis

**Table: Scoring system for histopathological changes in active pouchitis.**

<table>
<thead>
<tr>
<th>Histological Score</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute inflammatory infiltration</td>
<td>0</td>
</tr>
<tr>
<td>Mild and patchy ulcer in the surface epithelium</td>
<td>0</td>
</tr>
<tr>
<td>Moderate with crypt abscesses</td>
<td>0</td>
</tr>
<tr>
<td>Severe with crypt abscesses</td>
<td>0</td>
</tr>
<tr>
<td>Ulceration</td>
<td>4</td>
</tr>
<tr>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td>Mild superficial</td>
<td>0</td>
</tr>
<tr>
<td>Extensive</td>
<td>2</td>
</tr>
<tr>
<td>Chronic inflammatory cell infiltration</td>
<td>0</td>
</tr>
<tr>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td>Mild and patchy</td>
<td>0</td>
</tr>
<tr>
<td>Moderate</td>
<td>2</td>
</tr>
<tr>
<td>Severe</td>
<td>4</td>
</tr>
<tr>
<td>Villous atrophy</td>
<td>6</td>
</tr>
<tr>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td>Mild and patchy</td>
<td>0</td>
</tr>
<tr>
<td>Extensive</td>
<td>4</td>
</tr>
<tr>
<td>Irregular villous architecture</td>
<td>6</td>
</tr>
<tr>
<td>None</td>
<td>0</td>
</tr>
<tr>
<td>Mild and patchy</td>
<td>0</td>
</tr>
<tr>
<td>Extensive</td>
<td>4</td>
</tr>
</tbody>
</table>

Acute score $\geq 4$ correlates with clinical diagnosis of active pouchitis.

Cases with inflammation not amounting to pouchitis

Shepherd score (e.g., Acute 1+1 = 2, Chronic 2+2 = 4)

The features do not meet the histological criteria for a clinical diagnosis of acute pouchitis. Further clinicopathological correlation would be useful here.

Ulcer-associated cell lineage (UACL) as a marker of previous ulceration.

All looks fairly simple so what is the problem?
Site of biopsies

- Pre-pouch
- Pouch
- Anal cuff

Inflamed mucosa ¿ what is the pathology?

- Pre-pouchitis.
- Active pouchitis.
- Cuffitis.

Biopsy: "Neoterminal ileum"
Clinical: Restorative proctocolectomy and pouch. ¿ Pouchitis.

Is this:
- i) Patchy inflammation?
- ii) Normal pre-pouch with pouchitis?
- iii) Normal pouch with cuffitis?
**Clinical histories provided for same patient**

**11/07/08:** Moderately inflamed. Rectal strip. Pouchitis. Moderate colitis on rectal stump.

**05/08/08:** Pouchitis on EUA. No rectal cuff. Confirm pouchitis.

**Clinical histories provided for same patient**

**11/07/08:** Moderately inflamed. Rectal strip. Pouchitis. Moderate colitis on rectal stump.

**05/08/08:** Pouchitis on EUA. No rectal cuff. Confirm pouchitis.

→ Clinicians not always sure where they are!!
Any features that can help?

- UACL much more common in the small intestine.
- Crypt architectural distortion tends to be more severe in cuffitis.
- Paneth cells tend to be more plentiful in the small intestine.

Patient X 11/07/08

- Pouch biopsies: No evidence of pouchitis
- Rectal stump biopsies: Cuffitis

Consistent with active pouchitis

Patient X 11/07/08

- Pre-pouch biopsies: Normal
- Pouch biopsies: Pouchitis

Differential diagnosis of acute inflammation/ulceration in pouch

- Crohn's disease.
- Superinfection.
- Trauma.
- Intra-abdominal sepsis.
- Mucosal prolapse.
- Ischaemia.

- Pouchitis mimics Crohn's disease ++.
- Fistulae are common following pouch construction.
- NEVER MAKE A DIAGNOSIS OF CROHN'S BASED ON POUCH PATHOLOGY ALONE.
Differential diagnosis of acute inflammation/ulceration in pouch

- Crohn’s disease.
- Superinfection: Clostridium difficile, CMV.
- Trauma.
- Intra-abdominal sepsis.
- Mucosal prolapse.
- Ischaemia.

Neoplasia in the ileoanal pouch

- Carcinoma is rare in ileal pouches in the literature 10s-100s of cases.
- Most carcinoma is actually in residual anal cuff:
  - UC mostly in anal cuff (75%).
  - FAP mostly in anal cuff (75%)
- Adenomas in FAP both anal cuff++ and pouch.

Differential diagnosis of acute inflammation/ulceration in pouch

- Crohn’s disease.
- Superinfection.
- Trauma: Pouch intubation with S pouches.
- Intra-abdominal sepsis.
- Mucosal prolapse.
- Ischaemia.

Role of the pathologist

- Corroborating a clinical diagnosis of active pouchitis.
- Excluding other pathologies: CMV, C.Diff.
- Excluding dysplasia


Reporting of pouch biopsies

- Clinical history.
  - UC or FAP (or indeterminate colitis).
  - Symptoms.
- Endoscopic appearances.
  (Ideally endoscopy report with photos).
- Sites of individual biopsies.
- Discuss at IBD MDT.