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# Characterization of megapouch in patients with restorative proctocolectomy

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## Abstract

**Background** Megapouch is a rare functional complication of restorative proctocolectomy with ileal pouch-anal anastomosis characterized by pouch ± small bowel dilatation with no evidence of obstruction on endoscopy and imaging. Little is known about clinical characteristics and outcomes of this entity.

**Methods** We included all patients diagnosed with megapouch at our institution, identified from a pouch database. Data on baseline characteristics, management, and outcomes were documented and analyzed from electronic medical records. Appropriate statistical measures were used. $p < 0.05$ was considered significant.

**Results** Twenty-three patients with megapouch were identified. The mean age was 40.7 years; 95.6% had underlying ulcerative colitis; most common indication for colectomy was medically refractory disease (56.5%). Abdominal pain (82.6%) and bloating (52.2%) were most common presenting symptoms. Most common finding on pouchoscopy was pouch dilatation (81.8%), while barium or gastrografin enemas and MRI/CT mostly revealed dilatation of pouch and/or small bowel. Fourteen (66.7%) patients required some forms of surgery—six patients required pouch excision and three required either pouch redo or revision. Rates of pouch failure and IBD-related 1-year hospitalization were higher among patients managed surgically versus those managed medically ($p = 0.007$ and 0.024, respectively), while need for escalation of IBD-therapy was comparable between the groups ($p = 0.133$). No deaths were reported and no patient had recurrence of megapouch. IPAA revision or redo did not lead to more IBD-related morbidity.

**Conclusions** Majority of our patients with megapouch required surgery. In selected patients, redo pouch offered cure. Rates of pouch failure and IBD-related 1-year hospitalization were higher among patients managed surgically.

**Keywords** Ileal-anal pouch anastomosis · Megapouch · Pouch redo · Pouch revision · Pouch failure

Restorative proctocolectomy with ileal pouch-anal anastomosis (IPAA), first described in 1978, is the surgical treatment of choice for patients with medically refractory ulcerative colitis (UC), UC with dysplasia or neoplasia, familial adenomatous polyposis, and some cases of refractory and/or complicated indeterminate or Crohn's colitis [1–9]. IPAA is often associated with benefits like reestablishing intestinal continuity and avoiding a permanent ileostomy, decreasing requirement for disease-modifying therapies and their adverse effects, reducing risk of dysplasia and colon cancer and improving overall quality of life [5, 8, 10–13].

Despite its potential merits, IPAA can often result in a variety of mechanical, inflammatory, functional, dysplastic, and metabolic complications, which can cause significant morbidity and on occasions lead to pouch failure and loss [2, 10, 12, 14–16]. Pouchitis is the most common complication of IPAA, followed by Crohn's disease (CD) of the pouch, cuffitis, and irritable pouch syndrome (IPS) [2, 3, 8–10, 12, 14–20].

Functional disorders of pouch include IPS, anismus, pouchalgia fugax, and pouch pseudo-obstruction [2, 10, 12, 14, 16]. Pouch pseudo-obstruction or megapouch (MP) is rare, possibly under-recognized, complication characterized by pouch ± small bowel dilatation with no obvious obstruction on endoscopy and imaging. Literature review revealed little information on risk factors, clinical features, diagnosing modalities, management, and outcomes of this unusual condition.

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## Patients and methods

### Ethical considerations

All patients with megapouch were identified from our prospectively maintained, Institutional Review Board-approved pouchitis database. A waiver of requirement for informed consent was obtained from our Institutional Review Board since this observational study involved only retrospective chart reviews with minimal risk to the participants.

### Inclusion and exclusion criteria

All patients ≥ 18 years old with history of IPAA and findings suggestive of MP were identified from our prospectively maintained pouch registry. MP was defined by a combination of obstructive symptoms and abnormal dilatation of the ileal pouch ± small bowel on imaging (contrast enema, CT/MRI scan) and/or pouchoscopy, in the absence of mechanical obstruction. Abnormally elevated first sensation, first urge and/or maximum tolerated volumes on anorectal manometry (ARM), though not essential for the diagnosis of MP, were considered to be corroborative findings.

Patients with evidence of mechanical obstruction on imaging and/or pouchoscopy were excluded from the study. Patients without adequate follow-up data were excluded from analysis of outcomes. Patients with non-dilated pouch were chosen as unmatched controls to perform multivariable analysis to identify possible risk factors for the development of megapouch.

### Demographic and clinical data

Information regarding patient demographics, inflammatory bowel disease (IBD) characteristics and therapies, IPAA surgery and complications, clinical presentation, and outcomes of MP was collected from electronic medical records. Data were collected retrospectively regarding medication-use, non-IBD illnesses, IBD phenotype and therapies, IPAA (e.g., indications, type of pouch), and pouch-related complications (surgical, inflammatory, functional, dysplastic or neoplastic, infectious and metabolic complications).

### Diagnosis of megapouch

Unlike megacolon, there are no set up criteria for megapouch in the literature. In this study, the criteria for megapouch were defined as a pouch diameter > 12 cm measured at the mid pouch level, with or without proximal small bowel dilation (> 6 cm). Information regarding clinical features of MP and findings on pouchoscopy, barium/gastrografin enema studies, magnetic resonance imaging (MRI) and/or computed tomography (CT) scans, and ARM were recorded. Details regarding management of MP and outcomes including pouch failure, IBD-related hospitalization(s), need for escalation of IBD therapy, and death(s) were documented.

### Outcome measurement

In this study, creation of a new pouch with or without excision of patient's original pouch was termed *redo*, while reconstruction of a salvaged original pouch was termed *revision* of IPAA. *Pouch failure* was defined as the need for permanent diverting ileostomy, pouch excision or revision. *IBD-related hospitalizations* included all admissions to the hospital for IBD-related complications within one year of MP diagnosis, except those for initial MP surgery. *Escalation of IBD-therapy* was defined as the need for initiation of new disease modifying agent(s), addition of new agent(s) and/or increase in their dose on follow-up after management of MP.

### Statistical analysis

Routine descriptive statistics including measures of central tendency were used. SPSS software package (version 22.0) was used for statistical analyses. Mean and standard deviation were used to describe parametric, while median and inter-quartile range (IQR) were used to describe the distribution of non-parametric variables. Fisher's exact test, Mann–Whitney test and independent samples *t* test were used where appropriate. Multinomial logistic regression was used to identify factor(s) predictive of development of megapouch when compared to unmatched controls from our registry with non-dilated pouch. $p < 0.05$ was considered to be significant.

## Results

From a total of 3122 patients in our pouch registry, we identified 23 patients who met our inclusion and exclusion criteria for MP (prevalence of 0.74%). The mean age at diagnosis of MP was 40.7 ± 7.2 years. Of these, 14 (60.9%) were females and 22 (95.7%) were Caucasians. Five (21.7%) patients smoked more than seven cigarettes per day and two (8.7%) consumed more than one alcoholic drink per day after IPAA. Data regarding baseline characteristics are listed in Table 1.

### Clinical characteristics

An overwhelming majority of these patients had UC ($n = 22$, 95.7%) as the underlying diagnosis; only one patient had

**Table 1** Demographics and baseline characteristics ($n=23$)

| | |
|---|---|
| Demographics | |
| Age (years) | 40.7 ± 7.2 |
| Female gender | 60.9% |
| Caucasian race | 95.7% |
| Current smoking (> 7 cigarettes/day) | 21.7% |
| Alcohol (> 1 drink/day) | 8.7% |
| Medication-use (daily use post-IPAA) | |
| Opioid(s) | 17.4% |
| Antidepressant(s) | 13.0% |
| Benzodiazepine(s) | 13.0% |
| Phenothiazine(s) | 8.7% |
| Calcium channel blocker(s) | 4.3% |
| Co-morbidities | |
| Diabetes mellitus | 13.0% |
| Recent major medical illness | 4.3% |
| Recent major non-bowel surgery | 4.3% |
| IBD characteristics type of IBD | |
| UC | 95.7% |
| CD | None |
| IC | 4.3% |
| Family history of IBD (1st degree relative) | |
| UC | 26.0% |
| CD | 8.6% |
| UC phenotype | |
| Pan-colitis | 100% |
| Extra-intestinal manifestations | |
| Arthritis/arthralgia | 8.6% |
| Skin manifestations | 13.0% |
| Primary sclerosing cholangitis | 4.3% |
| Previous therapies | |
| Anti-TNF | 35.7% |
| Immunomodulator(s) | 34.7% |
| Indications for colectomy + IPAA | |
| Refractory colitis | 56.5% |
| Toxic megacolon | 13.0% |
| Neoplasia | 4.3% |
| Perforation peritonitis | 4.3% |
| Fulminant colitis | 4.3% |
| Steroid dependence | 4.3% |
| Unknown | 13.0% |
| Median interval from IBD diagnosis to IPAA (years) | 6.5 (IQR 1.9–9.0) |
| Type of anastomosis | |
| Hand-sewn | 60.0% |
| Stapled | 40.0% |
| Pouch type | |
| J Pouch | 87.0% |
| S pouch | 4.3% |
| K pouch | 8.7% |
| Stages of IPAA surgery | |
| 1 stage | 13.3% |
| 2 stage | 40.0% |
| 3 stage | 46.7% |
| IBD therapies (> 3 months post-IPAA) | |
| 5-Aminosalicylic acid | 33.3% |
| Topical steroids | 5.3% |
| Systemic steroids | 55.6% |
| Immunomodulator | 11.1% |
| Infliximab | 27.8% |
| Adalimumab | 15.8% |
| Vedolizumab | 21.7% |
| Antibiotics | 75.0% |

*n* adjusted for missing values while calculating percentages

indeterminate colitis (IC) and none had Crohn's colitis. Seven patients had family history of IBD in a first-degree relative—UC in five, CD in one, and both UC and CD in one. Among the nine patients with data regarding baseline IBD phenotype, all had UC with pancolitis. The most common extra-intestinal manifestations in our patients were arthralgia (8.6%), IBD-associated skin lesions (13.0%), and primary sclerosing cholangitis (4.3%). 35.7% patients had received anti-TNF therapies and 38.4% were on one or more immunomodulator(s) prior to IPAA surgery. Medical refractory UC (56.5%) and toxic megacolon (13.0%) were the most common indications for colectomy and IPAA in our patients. The median interval between IBD diagnosis and IPAA was 6.5 years (IQR 1.9–9.0). Most of our patients had J-pouches (87.0%) with only 2 (8.7%) patients with K-pouches and 1 (4.3%) with S-pouches. IBD therapies post-IPAA included 5-aminosalicylic acid (33.3%), topical corticosteroids (5.3%), systemic corticosteroids (55.6%), immunomodulator(s) (11.1%), infliximab (27.8%), adalimumab (15.8%), vedolizumab (21.7%), and antibiotics (75.0%).

## Characteristics of concurrent pouch disorders

The most common mechanical complications in our study subjects were strictures (39.1%), pouch prolapse (28.6%), and anastomotic leaks (23.8%). Pouchitis (78.3%), cuffitis (23.8%), and CD of the pouch (18.2%) were most common among inflammatory pouch complications. Functional abnormalities included anismus (20%) and irritable pouch syndrome (9.5%). Three (13.0%) patients had *Clostridium difficile* infection of the pouch. Metabolic complications were quite common—21 patients had anemia, including 10 (47.6%) patients with anemia of chronic disease and 8 (38.0%) patients with iron deficiency anemia. Four patients had (18.2%) malabsorption, 6 (28.6%) had hypokalemia and

**Table 2** Pouch complications ($n = 23$)

| Pouch complications | (%) |
|---|---|
| Mechanical complications | |
| Anastomotic leak | 23.8 |
| Pelvic abscess | 9.5 |
| Sinus/fistula | 19.0 |
| Stricture | 39.1 |
| Afferent/efferent limb syndrome | 22.7 |
| Pouch prolapse | 28.6 |
| Inflammatory complications | |
| Pouchitis | 78.3 |
| Cuffitis | 23.8 |
| CD of pouch | 18.2 |
| Inflammatory polyp | None |
| Functional compications | |
| Irritable pouch syndrome | 9.5 |
| Anismus | 20.0 |
| Neoplastic complications | |
| Dysplasia of pouch | None |
| Dysplasia of anal transition zone | None |
| Infectious complications | |
| Cytomegalovirus | None |
| *Clostridium difficile* | 13.0 |
| Metabolic complications | |
| Iron deficiency anemia | 38.0 |
| Anemia of chronic disease | 47.6 |
| Osteoporosis | 4.5 |
| Malabsorption | 18.2 |
| Vit. B12 deficiency | None |
| Hyperparathyroidism | None |
| Hypothyroidism | None |
| Hypercalcemia | None |
| Hypokalemia | 28.6 |
| Hypomagnesemia | 27.3 |

*n* adjusted for missing values while calculating percentages

6 (27.3%) had hypomagnesemia. Table 2 lists the pouch-related complications noted in our patients.

## Clinical characteristics and diagnosis of megapouch

The median interval between IPAA and presentation with MP was 12.0 years (IQR 3.8–18.2). The most common presenting symptoms were abdominal pain (82.6%) and bloating (52.2%), followed by difficulty with stool evacuation (39.1%), rectal bleeding (17.4%), and fever (17.4%). Pouchoscopy was helpful in diagnosing this condition, revealing abnormal pouch dilatation in 18 (81.8%) of the 22 patients who underwent the procedure (Fig. 1A, B). Inflammatory mucosal findings on pouchoscopy included ulcerations in 10 (45.4%), edema in 6 (27.3%), granularity in 4 (18.2%), friability in 5 (22.7%), loss of vascular pattern in 2 (9.1%), and mucus exudates in 1 (4.5%) patient(s). Sixteen patients underwent barium enema studies, revealing enlarged pouch in 9 (56.2%) and dilated proximal small bowel in 11 (68.8%) patients (Fig. 2). Cross-sectional imaging with MRI and/or CT showed dilated small bowel in 71.4% and pouch dilatation in 28.6% of patients (Fig. 3). Other associated findings on imaging included stricture (28.6%) and sinus (14.3%) formation.

ARM was performed in 12 patients. All patients had normal average resting and squeeze pressures. Results of volume studies were variable, with the most common abnormality being an increase in first urge volume (50.0%). Two (16.7%) patients had increased first sensation volume and 1 (8.3%) patient had increased maximum tolerated volume. Defecometry revealed paradoxical contractions in 50.0% patients. Table 3 describes the presenting characteristics of our patients with MP.

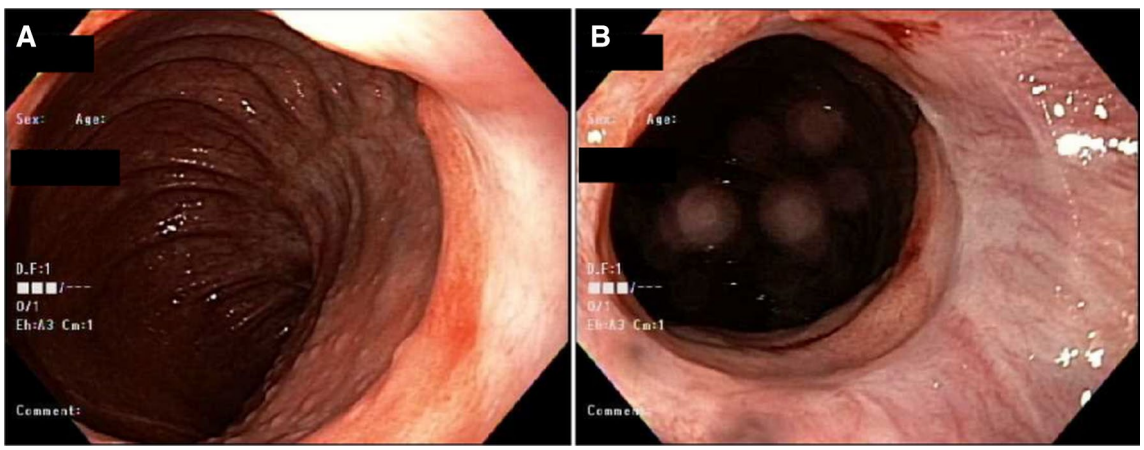


**Fig. 1** **A, B** Pouchoscopy showing dilated pouch with fluid pooling

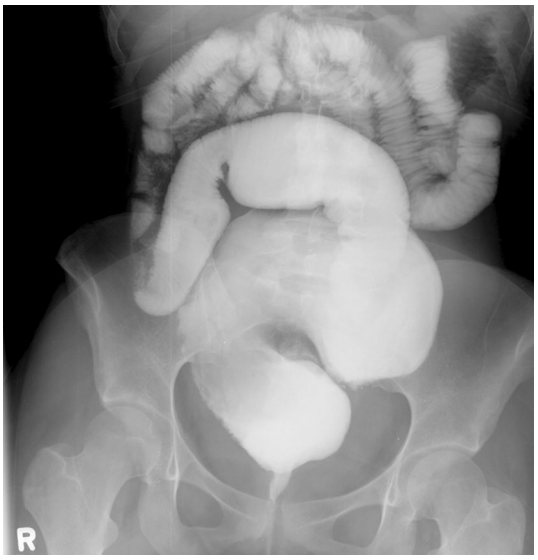


**Fig. 2** Barium enema study showing dilated pouch

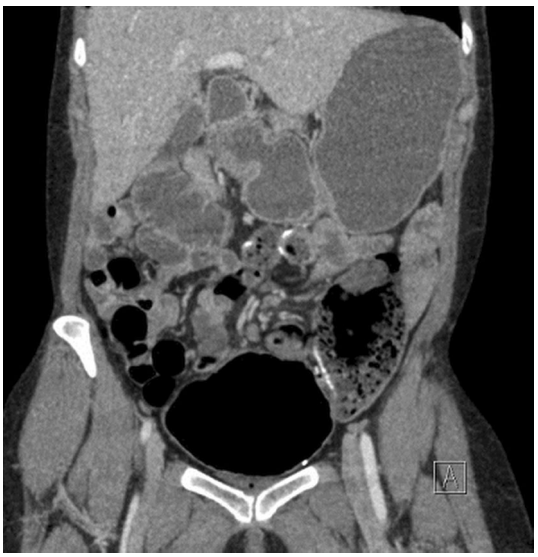

**Fig. 3** CT scan of abdomen and pelvis showing dilated pouch and proximal small bowel

### Potential risk factors for megapouch

Univariable analysis was performed to assess the risk factors for megapouch, with consecutive patients with no megapouch ($N=38$) as controls (Table 4). Subsequently multivariable analysis was performed and only a family history of IBD was found to be significantly predictive of development of megapouch in a logistic regression model ($p=0.026$).

### Management of megapouch

Among the 21 patients on whom we had data regarding management and outcomes, 14 (66.7%) underwent some form of surgical intervention for the treatment of MP, while 7 (33.3%) patients were managed medically with bowel rest and supportive measures. The most common surgeries in this setting were pouch excision with definitive end-ileostomy ($n=4$) and temporary diverting loop ileostomy with subsequent take-down ($n=4$), followed by pouch excision with creation of new pouch ($n=2$), definitive diverting loop ileostomy ($n=2$), and abdominoperineal pouch disconnection and redo of pouch without excision ($n=1$). Among surgically managed patients, 6 (42.8%) required excision of their pouch and 6 (42.8%) patients could salvage function of their initial pouch. One patient underwent pouch revision without excision, while two patients had pouch redo following excision of their initial pouch (Fig. 4).

### Outcomes of megapouch

Average duration of follow-up was 2.9 years. Pouch failure (i.e., need for permanent diversion, pouch excision or revision) occurred in 9 (42.8%) patients, 38.1% required escalation of IBD-therapy, and 52.4% patients required IBD-related hospitalization within 1 year. Pouch failure and IBD-related 1-year hospitalization rates were higher among patients managed surgically ($p=0.007$ and 0.024, respectively), while rates of IBD-therapy escalation were comparable with those managed conservatively ($p=0.133$) (Table 5). No deaths were reported. No patient had recurrence of MP.

Of the surgically managed patients, the rate of pouch failure was higher ($p=0.016$) among those who underwent excision of their initial pouches. However, all other outcomes were comparable, irrespective of whether patients underwent pouch excision or revision/redo (Table 6). All patients who had either pouch redo ($n=2$) or revision ($n=1$) had functioning pouches at latest follow-up, with no recurrence of megapouch.

## Discussion

Megapouch (also known as pseudo-obstruction of pouch) is a rare, and possibly under-recognized, functional complication of IPAA with very few cases reported so far in literature [21–23]. There is no established consensus regarding the diagnosis and management of this entity. In this study, we define MP as a functional disorder presenting with an enlarged, dilated pouch on endoscopy and/or imaging, without any evidence of mechanical obstruction or a well-defined

**Table 3** Clinical and diagnostic features of megapouch

| | |
|---|---|
| Presenting symptoms ($n=23$) | |
| Abdominal pain | 82.6% |
| Bloating | 52.2% |
| Difficult evacuation | 39.1% |
| Rectal bleeding | 17.4% |
| Fever > 100 °F | 17.4% |
| Pouchoscopy ($n=22$) | |
| Edema | 27.3% |
| Granularity | 18.2% |
| Friability | 22.7% |
| Loss of vascular pattern | 9.1% |
| Mucus exudates | 4.5% |
| Ulceration | 45.4% |
| Dilated pouch | 81.8% |
| Barium enema ($n=16$) | |
| Dilated small bowel | 68.8% |
| Dilated pouch | 56.2% |
| Concurrent stricture | None |
| MRI/CT ($n=14$) | |
| Abscess | None |
| Stricture | 28.6% |
| Sinus | 14.3% |
| Dilated small bowel | 71.4% |
| Dilated pouch | 28.6% |
| Anorectal manometry | |
| Pressure studies ($n=12$) | Mean ± 2 SD |
| Avg. resting pressure (mmHg) | 59.15 ± 23.52 |
| Avg. squeeze pressure (mmHg) | 144.52 ± 68.32 |
| Volume studies ($n=12$) | Median (IQR) |
| First sensation volume (mL); (normal—10–60 mL) | 30 (22.5–40) |
| First urge volume (mL); (normal—10–100 mL) | 60 (27.5–155) |
| Max. tolerated volume (mL); (normal—200–300 mL) | 200 (90–200) |
| Volume compliance (mL/mmHg); (normal—5–15 mL/mmHg) | 10 (3–10) |
| Defecometry ($n=10$) | |
| Normal contraction | 50.0% |
| Paradoxical contraction | 50.0% |

**Table 4** Comparison of characteristics between patients with megapouch and their unmatched controls with non-dilated pouch

| | Megapouch ($n=23$) | No megapouch ($n=38$) | $p$-value |
|---|---|---|---|
| Family history of IBD | 7 (30.4%) | 2 (5.3%) | 0.020 |
| J-pouch | 20 (87.0%) | 33 (86.8%) | 0.450 |
| Pouch fistula | 4 (17.4%) | 1 (2.6%) | 0.062 |
| Afferent/efferent limb syndrome | 5 (21.7%) | 0 | 0.006 |
| Pouch prolapse | 6 (26.1%) | 0 | 0.002 |
| Pouchitis | 18 (78.3%) | 23 (60.5%) | 0.173 |
| Cuffitis | 5 (21.7%) | 2 (5.3%) | 0.093 |
| Crohn's disease of pouch | 4 (17.4%) | 6 (15.8%) | 0.999 |
| Irritable pouch | 2 (8.7%) | 0 | 0.138 |
| Difficulty evacuating | 9 (39.1%) | 3 (7.9%) | 0.006 |
| Paradoxical contractions on defecometry | 5 (21.7%) | 4 (10.5%) | 0.262 |

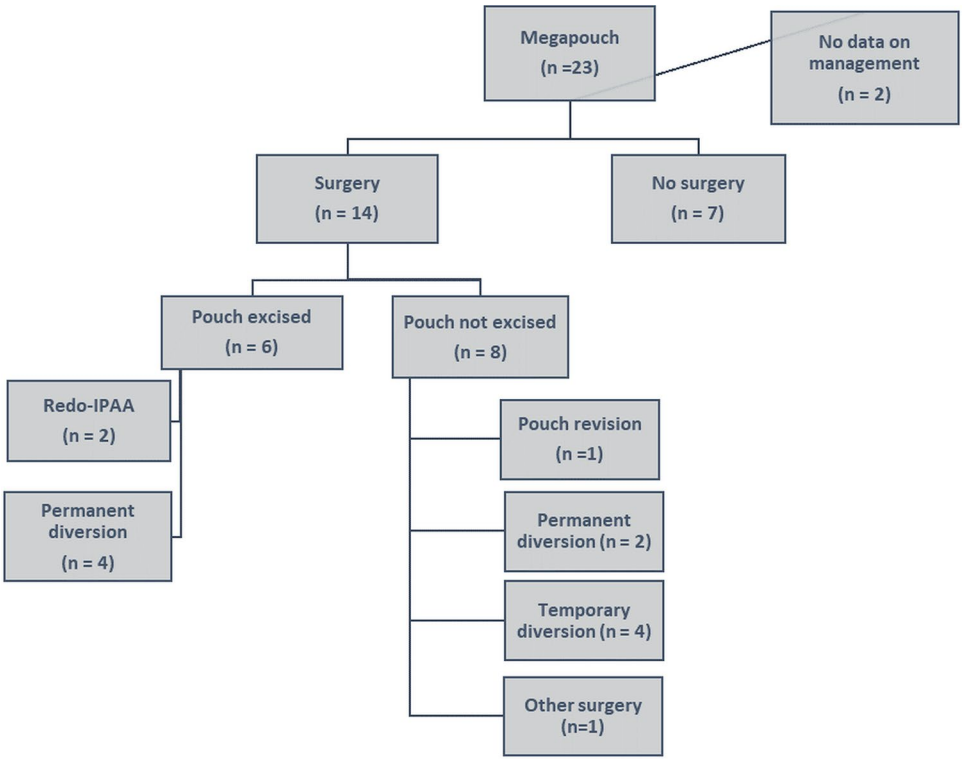


**Fig. 4** Management of our patients with megapouch

transition point above or below the pouch. It may or may not be associated with dilatation of the small bowel proximal to the anastomosis.

In our study, patients with MP presented with variable symptoms and signs which were often clinically indistinguishable from myriad other pouch-related complications. A combination of contrast enema, cross-sectional imaging, pouchoscopy and manometry studies were needed to diagnose MP and rule out various other inflammatory, mechanical and infectious pouch complications. Conservative management, including bowel rest, fluid and electrolyte replacement, and treatment of associated inflammatory and infectious complications, resulted in resolution of MP in a third of our patients, while two-thirds required some form of surgery. Among our surgically managed patients, 43% required excision of their pouch, and 21% underwent redo or revision of IPAA. Rates of pouch failure and IBD-related hospitalization were higher among patients who required surgery, likely reflective of the more severe presentation in this subset. Revision or redo of IPAA did not lead to higher rates of MP recurrence or other IBD-related morbidity, suggesting that such reconstructive procedures offer a reasonable alternative to permanent diversion, while significantly improving quality of life and patient satisfaction.

The etiology and pathogenesis of this disease entity are not clear. We attempted to identify the risk factors for the disorder. In the limited multivariable analysis, only family history of IBD was found to be a risk factor. We speculate roles played by GI dysmotility, abnormal function of interstitial cells of Cajal, and small bowel bacterial overgrowth. Though colonic pseudo-obstruction has been found to occur most commonly among men in their 60 s, majority of our patients with MP were females with average age in the 40 s [24]. Though this study was not designed to identify risk factors for the development of MP, extrapolating from studies on colonic pseudo-obstruction, possible predisposing factors for MP may include severe illness or major surgery, electrolyte and other metabolic imbalances, and medications such as opioid analgesics, benzodiazepines, phenothiazines, calcium channel blockers, antidepressants etc. [25–28]. In our study, only one patient each had preceding major illness or non-bowel surgery. Four (17.4%) patients were on opioids, while 3 (13.0%) each were on benzodiazepines and antidepressants. Though there is no definitive evidence in the literature to support the same, IPAA surgery-related factors including type of pouch, anastomosis, and stages of surgery may influence the risk of MP. Most of our patients had J-pouches. The three previously reported cases of MP were all described in patients with J-pouches [22, 23]. Maddireddy et al. [23] proposed that a long limb length (> 15 cm) of J-pouch predisposed to the development of MP in his patient. Manilich et al. [29] reported that IPAA patients undergoing hand-sewn anastomosis and three-stage surgeries were at higher risk for pouch failure. A total of 60% of our patients had hand-sewn anastomosis and close to 50% underwent three-stage surgery. Paradoxical

**Table 5** Comparison of baseline characteristics and outcomes between MP patients managed conservatively versus surgically

| | No surgery ($n=7$) | Surgery ($n=14$) | $p$-value |
|---|---|---|---|
| Demographics | | | |
| Female gender | 71.4% | 57.1% | 0.656 |
| Average age (years) | 41.8 ± 13.2 | 41.5 ± 10.0 | 0.967 |
| Caucasian race | 85.7% | 100% | 0.333 |
| Type of IBD | | | |
| UC | 85.7% | 100% | 0.333 |
| Indeterminate colitis | 14.3% | | |
| Previous anti-TNF therapy | 33.3% | 33.3% | > 0.999 |
| Type of pouch | | | |
| J-pouch | 57.1% | 100% | 0.030 |
| S-pouch | 14.3% | | |
| K-pouch | 28.6% | | |
| Duration from IPAA to megapouch (in years) | 14.5 ± 8.9 | 10.9 ± 3.9 | 0.477 |
| Clinical features | | | |
| Abdominal pain | 57.1% | 92.9% | 0.088 |
| Bloating | 57.1% | 50.0% | 0.757 |
| Difficulty evacuating bowel | 42.9% | 35.7% | 0.751 |
| Rectal bleeding | 42.9% | 7.1% | 0.088 |
| Fever > 100 F | 28.6% | 14.3% | 0.574 |
| Anemia | 66.7% | 85.7% | 0.549 |
| Pouchoscopy | | | |
| Edema | 14.3% | 28.6% | 0.624 |
| Mucosal friability | 28.6% | 21.4% | 0.717 |
| Ulceration | 28.6% | 42.9% | 0.656 |
| Dilated pouch | 71.4% | 92.9% | 0.247 |
| Barium enema | | | |
| Pouch dilatation | 66.7% | 72.7% | 0.837 |
| Small bowel dilatation | 33.3% | 76.9% | 0.214 |
| MRI/CT scan | | | |
| Dilated pouch | None | 33.3% | 0.490 |
| Dilated small bowel | None | 75.0% | 0.308 |
| Stricture | None | 25.0% | 0.569 |
| Anorectal manometry | | | |
| Volume studies (mean values) | | | |
| First sensation volume (mL); normal—10–60 mL | 20.0 | 57.8 | 0.705 |
| First urge volume (mL); normal—10–100 mL | 20.0 | 94.4 | 0.464 |
| Maximum tolerated volume (mL); normal—200–300 mL | 60.0 | 175.5 | 0.204 |
| Compliance (mL/mmHg); normal—5–15 mL/mmHg | 3.0 | 8.7 | 0.241 |
| Outcomes | | | |
| Escalation of IBD therapy | 14.3% | 50.0% | 0.133 |
| IBD-related hospitalization within 1 year of MP diagnosis | 14.3% | 71.4% | 0.024 |
| Average duration to 1st IBD-related hospitalization | 231 days | 228 days | 0.989 |
| Median no. of hospitalizations in 1st year (IQR) | 0 (0) | 1 (2.5) | 0.024 |
| Pouch failure | None | 64.3% | 0.007 |
| Recurrence of megapouch | None | None | – |
| Death | None | None | – |

**Table 6** Outcomes among patients managed surgically ($n=14$)

| | Pouch excised ($n=6$) | Pouch not excised ($n=8$) | $p$-value |
|---|---|---|---|
| Pouch failure | 100% | 37.5% | **0.016** |
| Pouch function retained at last follow-up | 33.3% | 75.0% | 0.119 |
| Recurrence of MP | None | None | – |
| Mortality | None | None | – |
| Escalation of IBD therapy | 66.7% | 37.5% | 0.592 |
| IBD-related hospitalization within 1 year | 66.7% | 75.0% | 0.733 |
| | Pouch re-done or revised ($n=3$) | Pouch not re-done or revised ($n=11$) | $p$-value |
| Pouch failure | 100% | 54.5% | 0.145 |
| Pouch function retained at last follow-up | 100% | 45.5% | 0.091 |
| Recurrence of MP | None | None | – |
| Mortality | None | None | – |
| Escalation of IBD therapy | 33.3% | 54.5% | 0.515 |
| IBD-related hospitalization within 1 year | 66.7% | 72.7% | 0.837 |

puborectalis contractions on ARM were noted in 50% of our patients and could possibly contribute to pouch and small bowel dilatation in absence of structural abnormality [30, 31]. Pouchitis was a frequent complication among our patients, as were mechanical complications like stricture and pouch prolapse. Shen et al. previously proposed that the effects of these functional, mechanical, and inflammatory complications could be bidirectional on each other [32]. We have noted that patients with functional pouch disorders are more likely to have underlying depression/anxiety and be concurrently treated with antidepressants, anxiolytics, and narcotics, and these psychosocial and medication-related factors could contribute to the development of MP [2, 16, 33]. Metabolic complications, mainly anemia and less frequently hypokalemia and hypomagnesemia, were observed. However, whether they contributed to the development of MP in our patients is not clear.

Clinical presentation of MP can be variable. Most of our patients had abdominal pain, bloating, and difficulty evacuating stool (dyschezia) as their presenting complaints. Similar presentation was seen in the cases of MP reported by Dayton et al. [22] and Maddireddy et al. [23]. Copley et al. reported regarding a chronic non-functional large ileo-anal pouch presenting with chronic abdominal distension, diarrhea, weight loss, and malnutrition [21].

Diagnosis of MP and functional pouch disorders are usually difficult. We used a combination of pouch endoscopy, barium enema, MRI, CT, and ARM to diagnose MP. Dilated pouch and/or dilated small bowel were the most common findings on pouchoscopy, barium enema, and cross-sectional imaging, similar to findings observed in previously reported MP cases. Tang et al. and Shen et al. concluded that complementary modalities such as MRI, CT enterography, gastrografin enema, and pouch endoscopy are needed to best evaluate complex pouch disorders [10, 12, 14, 16, 34, 35]. ARM with defecometry also contributed to our diagnosis. All our patients had normal pressure studies but some had abnormal volume studies. Khanna et al. suggested that manometric evaluation with balloon expulsion be performed in pouch patients with prominent dyschezia symptom [36].

A majority of our patients (66.7%) failed conservative measures and ended up requiring surgical intervention to treat MP. The types of surgery undertaken in our patients are listed above. Redo or revision of IPAA was successfully performed in three patients in our series—one patient had revision with pouch salvage while two patients had excision with creation of new pouch. Previous reports also suggest that MP can be successfully treated with redo or revision of IPAA [21–23]. The functional outcome and quality of life is better in patients undergoing redo or revision of IPAA for malfunctioning pouches and is a suitable alternative to pouch failure with lower subsequent pouch loss rates [22, 37, 38]. Patients also prefer this option over permanent ileostomy. In our study, pouch redo or revision were not associated with increased rates of pouch failure, IBD-related hospitalization, escalation of IBD therapy, or recurrence of MP. Fonkalsrud et al. concluded that reconstruction of dysfunctional ileo-anal pouches due to large size or long efferent limb had resulted in marked improvement in intestinal function in > 93% of his patients and reduced the need for late pouch removal [39]. Patients who fail initial redo surgery can either have a second redo or permanent ileostomy as last option [22, 38, 40]. In our study, pouch failure, defined as the need for permanent diversion, pouch excision, or revision, occurred in 42.8% patients. Rate of pouch failure and IBD-related 1-year hospitalization were higher among our patients who were managed surgically, likely reflective of greater severity of disease presentation in this sub-group.

Besides surgery, strategies that could be tried include treating coexisting inflammatory, mechanical, or metabolic complications with antibiotics, biofeedback, nutrient and electrolyte supplementation, and other supportive measures. Addressing underlying psycho-social factors might also help in select cases. Measures directed at initial pouch surgery like choosing stapled over hand-sewn anastomosis, 1 or 2 stage over 3 stage IPAA and restricting pouch limb length to 12–15 cm, may be protective. In our study, there was no increase in mortality due to MP.

This is the first study of its kind describing the clinical presentation, diagnostic features, management, and outcomes of MP, a rare disorder with only a few cases reported in the literature to date. Our study suggests that while some patients with MP can be successfully managed conservatively, most require surgical intervention. IPAA can be revised or redone, with good outcomes and better quality of life, if patients are properly selected. A high index of suspicion and appropriate utilization of a combination of diagnostic modalities are required to accurately diagnose this uncommon, and likely under-diagnosed, entity. We hope our findings will help contribute to increase the awareness among gastroenterologists and colorectal surgeons about the recognition and treatment of this pouch complication.

Our study had several limitations. Since this was a retrospective observational study conducted at a large referral center, we could have selected patients with more severe disease and thus our findings may not representative of the general population. Due to lack of matched controls in the study, we could not draw inferences regarding risk factors for the development of MP. Our sample size was small due to the rare nature of this disease entity.

In conclusion, MP is a rare complication after IPAA. Precise etiology and precipitating factors are unknown. Clinical presentation can vary and diagnosis usually requires a combination of modalities. Treatment is surgical if conservative measures fail.

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## Compliance with ethical standards

**Disclosures** Preeti Shashi and Bo Shen have no conflicts of interest or financial ties to disclose.

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