

Fellows' Corner

A Rare Differential: Afferent Limb Syndrome in Patients with an Ileal Pouch Anal Anastomosis

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CASE PRESENTATION

A 47-year-old male with a history of ulcerative colitis complicated by acute toxic colitis resulting in an emergent one-stage laparoscopic restorative proctocolectomy with J-pouch formation, presented with a 10-year history of intermittent gastrointestinal obstructive symptoms beginning after his operation. Additionally, he endorsed colicky abdominal pain, nausea, episodic vomiting, bloating, and acid reflux. His symptoms appeared shortly after his J-pouch was created without improvement over the next decade. Multiple cross-sectional imaging studies have been performed but failed to demonstrate any signs of obstruction with transition point, evidence of retained rectum or pouch twist, and no abdominal wall hernia or pelvic sepsis. He was referred to an inflammatory bowel disease surgeon where office exam was negative for paradox puborectalis and repeat endoscopy validated no pouch twist, retained rectum, anastomotic leak and the bowel proximal to his pouch was able to be visualized. A



Image A. Endoscopic view of a J-pouch showing a severely angulated pouch inlet (circle) indicative of afferent limb syndrome.

Photo courtesy of Bo Shen, MD

Gastrografin enema was performed which revealed normal pouch filling, however, after evacuation a large amount of contrast was present in the small bowel proximal to the pouch.

QUESTIONS

1. What is the diagnosis in this patient?
2. What is the underlying pathology of this syndrome?
3. How is this syndrome diagnosed?
4. What are the management options in this patient?

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Image B. Post-evacuation sagittal image of a Gastrografin enema, showing dilated small bowel filled with contrast proximal to the pouch (arrow pointing to retained contrast in small bowel proximal to the pouch).

Question 1.

This patient is suffering from afferent limb syndrome (ALS) after restorative proctocolectomy, which is caused by an acute angulation of the afferent bowel at the pelvic pouch inlet (Image A). ALS differs from efferent limb syndrome (ELS) in patients with a pelvic pouch as ELS is caused by a long exit-conduit from an S-pouch, while ALS is caused by an obstruction proximal to the ileal-anal pouch.

Question 2.

Restorative proctocolectomy and ileal pouch-anal anastomosis (IPAA) is the procedure of choice in patients who have ulcerative colitis, familial adenomatous polyposis, and select patients with Crohn's disease.¹ Despite a low failure rate of 3.4%, there are a variety of possible complications that can result in varying degrees of pouch dysfunction.² Common postoperative complications are pelvic sepsis, hernia, pouch dysfunction due to inadvertent pouch-rectal anastomosis/pouch twists/outlet obstruction/paradoxical contraction, and the most common complication, small bowel obstruction (SBO).³ SBO in patients with an IPAA is frequently diagnosed by symptomatology and cross-sectional imaging and is a consequence of intra-abdominal adhesions, however, some patients



Image C. Gastrografin enema performed status post laparoscopic adhesiolysis, abdominopelvic pouch mobilization, pouchopexy, and intraoperative pouchoscopy with diverting loop ileostomy, and before scheduled ileostomy reversal. Post-evacuation sagittal image of a Gastrografin enema, showing resolution of the dilated contrast-filled bowel proximal to the pouch, with only minimal contrast present in the pouch and the pre-pouch small bowel.

with non-specific abdominal pain and bloating may have ALS. ALS can be due to acute angulation, intussusception of the distal ileum, or adhesions leading to obstruction of the afferent-limb at the bowel of the pouch inlet, typically with bowel trapped between the pouch and the sacrum.⁴ ALS is rare, occurring in just less than 2% of all patients after IPAA creation, and in 12% among patients who are displaying obstructive symptoms.^{4,5} ALS can present acutely after IPAA or years later.

Question 3.

ALS can present as abdominal pain, nausea, and vomiting, similar to symptoms of SBO, but without the typical radiographic findings seen with a bowel obstruction. Patients with ALS have often been worked up for various symptomatology, which typically begins with history, physical exam, and cross-sectional imaging of computed tomography (CT) to assess for adhesive small bowel obstruction. If the initial workup is negative, patients should undergo pouchoscopy to look for signs of Crohn's disease or septic complications.

However, assuming symptoms are secondary to Crohn's disease can lead to years of unneeded medical therapy when a mechanical issue needing corrective surgery is the underlying pathology. For this reason, the patient should be referred to a pouch specialist for formal workup. This includes an exam under anesthesia, flexible pouchoscopy to assess for pouch ulcers, a stricture at the pouch inlet or perianal fistulae to suggest Crohn's disease/pelvic sepsis, a pelvic magnetic resonance imaging to assess for retained rectum or pouch twist, and a Gastrografin enema (GGE) to assess for ALS or stricture. The typical finding for ALS of a GGE exam is contrast retention in a dilated pre-pouch small bowel after the patient has evacuated the Gastrografin (Image B).

Question 4.

The management of ALS involves a multidisciplinary team as endoscopic interventions and/or surgical management may be needed to correct the underlying obstruction at the pouch inlet. Endoscopic balloon dilation can be attempted, typically with a 20mm balloon, though almost half of patients require repeat dilations, and symptoms may continue to ultimately require operative intervention.⁴ Other endoscopic interventions such as needle-knife sinusotomy to divide the chronic scar at the angulated tissue have also had success.¹ Surgically, patients can undergo resection of the angulated bowel with anastomosis, enteroenterostomy to bypass the segment adherent posteriorly to the pouch to avoid pouch mobilization (thus avoiding injury to the pouch or mesentery which can cause pouch ischemia), mobilization of the pouch with a pexy of the pouch and/or proximal small bowel, and pouch excision with the creation of an end-ileostomy.⁴ There is a 40-100% resolution of symptoms after surgical intervention, while around 40% of patients who undergo endoscopic interventions will need repeat endoscopic interventions.^{4,5} Though some series omit endoscopic interventions to correct ALS and refer the patient for surgery, endoscopic intervention by an experienced pouch-endoscopist may obviate the need for surgical intervention and should be attempted initially.^{4,5} This patient was able to undergo a laparoscopic adhesiolysis,

abdominopelvic pouch mobilization, pouchopexy, and intraoperative pouchoscopy with diverting loop ileostomy. He was discharged uneventfully and able to tolerate a low-fiber diet without any abdominal pain or bloating. He underwent a GGE prior to stoma closure (Image C) which showed complete evacuation of the contrast from both his pouch and proximal small bowel. There were no further complaints of any abdominal pain or bloating after stoma closure.

CONCLUSION

Clinicians should be mindful when evaluating non-specific abdominal pain, bloating, or pouch dysfunction in patients who have undergone IPAA, as ALS can be elusive to diagnosis and so its presence must be germane during the workup. Complications such as afferent limb syndrome may be rare, however, may contribute to a delay in diagnosis in this patient population acutely or years after surgery. A multidisciplinary approach is necessary to properly identify the source of pathology and offer medical, endoscopic, or surgical correction. ■

References

1. Holubar SD. Prevention, Diagnosis, and Treatment of Complications of the IPAA for Ulcerative Colitis. *Dis Colon Rectum*. 2018;61(5):532-536. doi:10.1097/DCR.0000000000001094
2. Delaney CP, Remzi FH, Gramlich T, Dadvand B, Fazio VW. Equivalent function, quality of life and pouch survival rates after ileal pouch-anal anastomosis for indeterminate and ulcerative colitis. *Ann Surg*. 2002;236(1):43-48. doi:10.1097/00000658-200207000-00008
3. Ng K-S, Gonsalves SJ, Sagar PM. Ileal-anal pouches: A review of its history, indications, and complications. *World journal of gastroenterology*. *World J Gastroenterol*. Aug 21, 2019; 25(31): 4320-4342 doi: 10.3748/wjg.v25.i31.4320
4. Read TE, Schoetz DJ, Marcello PW, et al. Afferent Limb Obstruction Complicating Ileal Pouch-Anal Anastomosis. *Dis Colon Rectum*: May 1997 - p 566-569 doi: 10.1007/BF02055380
5. Kirat HT, Kiran RP, Remzi FH, Fazio VW, Shen B. Diagnosis and management of afferent limb syndrome in patients with ileal pouch-anal anastomosis. *Inflamm Bowel Dis*. 2011;17(6):1287-1290. doi:10.1002/ibd.21503