

## Jejunioleal Diverticulosis: A Review

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### Abstract

Jejunioleal diverticulosis (JID) is an acquired condition affecting up to 0.06-1.3% of the population of the United States, with a peak incidence of 60-80 years old. While often asymptomatic, JID can present in numerous ways ranging from chronic mal absorption to acute hemorrhage to perforation and sepsis. Treatment of JID is best catered to the presenting symptoms. The following review details the epidemiology, pathophysiology, diagnostic testing, presenting signs and symptoms, and treatment options for this uncommon disease.

### Introduction

The earliest reference to jejunioleal diverticulosis (JID) is found in the works of Soemmering and Baile in 1794. Early reports were simply presented as findings, anatomical variants of no clinical import. Little fanfare accompanied these reports until the late 19th century, when some of the fathers of modern surgery including Osler, Virchow and Laterjet added case reports and published notes on the condition [1]. While JID is most commonly an incidental finding; autopsy series estimate a prevalence ranging from 0.3-1.3% [2]. JID is found twice as often in men [3]. The incidence of JID peaks in the elderly, with most cases found in men and women between the ages of 60 and 80. The goal of this review is to discuss the history, proposed pathophysiology, presentation and therapy of this uncommon illness.

### Pathophysiology

The most commonly held theory for the formation of JID is similar to that of colonic diverticulosis; increased intraluminal pressures affect weak areas of the wall, leading to extrusion of the mucosa and false diverticulum formation [4]. On histological analysis, Edwards found disruption of the muscularis mucosae by the vasculature resulting in a weakening in the wall, or as he termed it "locus minoris resistentia" [1]. This weak point allows for mucosal extrusion from the lumen, resulting in the "out-pouching." Interestingly, the jejunum is known to have a larger diameter of vessels comprising the vasa recta supplying it. The larger diameter of the weakened site may contribute to the observation that diverticula are more common in the proximal small bowel than the distal. Once a point of decreased resistance has been created, elevated luminal pressures create aneurismal dilation of the intestinal wall, resulting in false diverticula.

Krishnamurthy et al. proposed a second theory of the pathophysiology of jejunioleal diverticulosis [5]. In this study, small intestine samples of seven patients were examined under light microscopy to explore the possibility of smooth muscle dysfunction. Four of these patients were found to have fibrosis and decreased quantity of smooth muscle cells. Two more demonstrated fibrosis associated with degenerative smooth

muscle cells. The final patient sample had neuronal and axonal degeneration. These findings were consistent with those of progressive sclerosis, systemic neuropathy and visceral myopathy, respectively. Concomitant existence of autoimmune or motility disorders with JID suggests an association with the formation of the diverticula.

The majority of patients with small bowel diverticulosis are noted to have isolated duodenal disease; in a 1997 retrospective study of two hundred eight patients, 79% of patients had isolated duodenal disease [6]. 18% of patients had jejunioleal disease and 3% had the all three segments of small bowel affected.

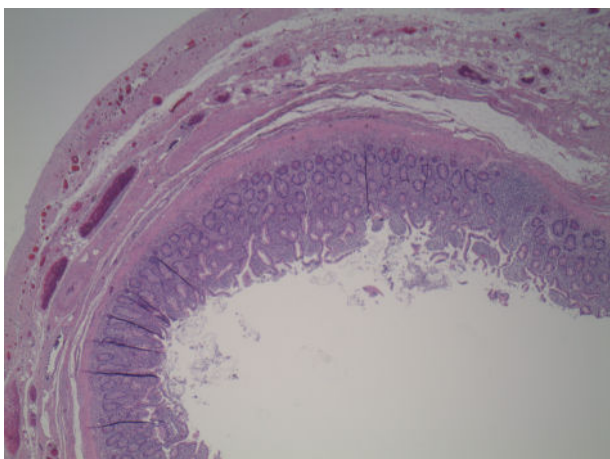
While the majority of this patient population is the elderly, there are more isolated documented cases of intestinal diverticula in children and in family groups [7,8]. These cases have been used to postulate inherited, genetic, or embryological causes for the disorder. Anderson et al described a family of eight siblings, six of whom had small intestinal diverticula [7]. Four subjects had concomitant immunologic diseases, specifically rheumatoid arthritis, ulcerative colitis, thyroiditis and non-viral hepatitis. Barton et al. reported a family of patients with JID in multiple generations [9]. Six of 10 siblings in this cohort had symptomatic JID, and one symptomatic patient had monozygotic twin children with symptomatic JID suggesting an autosomal dominant inheritance pattern. This study included a pathology specimen demonstrating extensive true diverticula of the jejunum (Figures 1-3), suggesting a mechanism more consistent with that of Krishnamurthy et al [5].

### Symptoms

While the majority of cases of JID are asymptomatic, patients can present with a multitude of symptoms [4]. Tsiotos et al. describe a series of 112 patients with JID, 47 of whom were diagnosed incidentally [10]. Of the remaining 65 patients, 45 presented with symptoms of malnutrition or chronic pain while the remaining 20 presented with acute complications of JID.



**Figure 1:** Surgical evaluation demonstrates large diverticula, predominantly along the antimesenteric border of the small bowel.

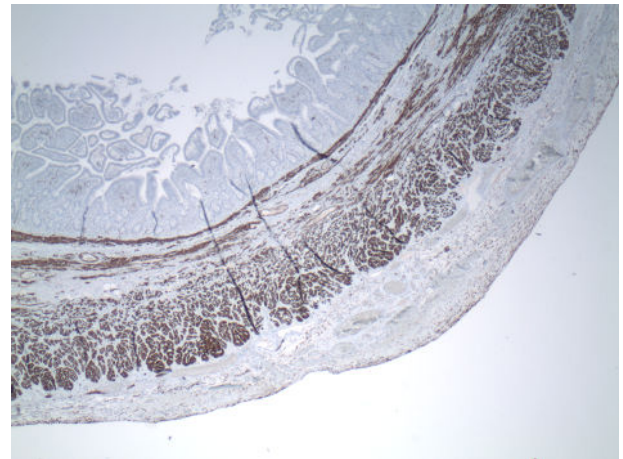


**Figure 2:** H&E stain demonstrates true diverticulum with all layers of bowel wall represented.

An estimated 10% of patients with small bowel diverticula will present with acute clinical symptoms [11]. Jejunal diverticula are 4 times more likely to present acutely compared to other small bowel diverticula. Diverticulitis, sepsis, liver abscess, perforation, fistula formation, volvulus and intussusception are all reported presentations of acute jejunoileal diverticulitis [12]. Patients with JID may also present with lower gastrointestinal bleeding, similar to colonic diverticulosis. In these settings, prompt admission and management of shock is paramount, as life-threatening arterial hemorrhage may occur.

Malnutrition in these patients is thought to be associated with stagnation of luminal and intra-diverticular contents resulting from intestinal hypomotility [13]. This stagnation leads to bacterial overgrowth. The excess bacteria consume B12 and deconjugate bile salts leading to a megaloblastic anemia and impaired uptake of fats causing steatorrhea [10]. Interestingly, the authors were not able to find documented evidence of a significant difference in either B12 or

albumin levels in symptomatic patients when compared to those diagnosed incidentally.



**Figure 3:** Desmin stain demonstrates muscularis mucosa (red arrow) and muscularis propria (\*) within the diverticulum, consistent with true diverticulum.

Chronic JID diverticulitis may present with inflammatory and infectious symptoms, abscess formation or frank perforation with pneumoperitoneum [12]. Barton et al. described a family of patients presenting with chronic pneumoperitoneum and associated malnutrition, without evidence of peritonitis [9]. The malnutrition in this patient population appeared to be secondary to chronic pain and decreased oral intake secondary to the chronic pneumoperitoneum, based on the patient history.

In a majority of cases, the diagnosis of JID is made incidentally [12]. Commonly, diverticula are found on upper GI series, endoscopy and laparotomy. In these cases, exploration is made for symptoms that are unable to be attributed to the diverticula. Among the 47 patients incidentally diagnosed with JID in the Tsiotos series, 39 patients (82%) remained asymptomatic for a mean follow-up time of 4.8 years after diagnosis [10].

## Diagnosis

Computed tomography (CT) has become the mainstay of diagnosis of JID, secondary to its ubiquitous nature [14]. CT should be performed in the abdomen and pelvis with oral contrast to determine the extent of diverticulosis, as well as IV contrast to assess for evidence of diverticulitis such as fat stranding or evidence of extraluminal air. Unfortunately, CT scan underestimate the severity of JID, as diverticula can hide in the mesentery or be mistaken for small bowel loops.

Magnetic resonance imaging (MRI) and enterography (MRE) use is becoming more widespread in the United States [14]. MR has the advantage of improved soft tissue evaluation over CT scan, but carries disadvantages of time and cost. Additionally, patients with claustrophobia tolerate MR poorly due to the smaller tube used during imaging.

Enteroclysis by fluoroscopy, CT or MR carries improved diagnostic accuracy [14]. The infusion of either single or double-contrast with water-soluble fluid and air can demonstrate diverticula throughout the bowel. Despite this, the benefit of enteroclysis over CT scan is limited,

as improved delineation of the diverticula is unlikely to change the plan of management. Additionally, enteroclysis requires the placement of a nasoenteric tube, and is poorly tolerated by patients. Owing to these disadvantages; the authors do not recommend the routine use of enteroclysis in the evaluation of JID.

While endoscopy remains a mainstay of evaluation for colonic diverticulosis, the challenges associated with endoscopic evaluation of the small bowel limits its utility in JID [14]. Double-balloon enteroscopy allows the endoscopist to intubate the small bowel, but this procedure is very invasive and yields little additional information in the treatment of JID. Capsule endoscopy does allow evaluation of the small bowel, but caution should be used when one is suspicious of JID, as Hanna et al have reported a case of capsule retention in a patient with severe, symptomatic JID with chronic pneumoperitoneum, necessitating small bowel resection [15].

Nuclear medicine tagged red blood cell studies and angiography may be of diagnostic value in the setting of active hemorrhage, but have little value in non-bleeding patients [14]. In spite of these many diagnostic modalities, the gold standard for diagnosis of JID remains laparotomy and direct visualization (Figure 1) [16].

## Treatment

As most people with JID are asymptomatic, reassurance remains the treatment for the majority this patient population [17]. Patients with known JID should be cautioned with regards to the symptoms previously outlined in situations where acute complications are suspected, including pain, signs of hemorrhage, and infection.

Patients with hemorrhage should be admitted, with prompt and goal-directed resuscitation. Reversal of any anticoagulation may lead to spontaneous resolution of the bleeding. Tagged red blood cell scans and mesenteric angiography may localize the site of bleeding [18]. Angiography carries the added benefit of allowing either embolization of the site of bleeding, or administration of methylene blue or other compounds to dye the area of hemorrhage and aid surgical resection. In patients with refractory bleeding, operative intervention with either diverticulectomy or bowel resection is mandatory, as endoscopic intervention is unlikely to stop hemorrhage in the small bowel [16].

Acute jejunoileal diverticulitis can be treated non-operatively in the absence of sepsis and peritonitis. Antibiotics, fluid resuscitation and bowel rest lead to improvement in symptoms in 75 percent of patients [10]. Other agents such as anti-spasmodic agents, analgesics, B12 supplementation and antacids are utilized for symptomatic control. In these patients, resection is should be considered as a last resort as resection carries a disproportionately higher morbidity and mortality. Peritonitis or severe sepsis warrants operative management with bowel resection and primary anastomosis [4].

Patients with severe malnutrition, chronic pneumoperitoneum or chronic abdominal pain present a greater therapeutic challenge. Some of these patients will see improvement of their symptoms with intermittent antibiotics to manage their bacterial overgrowth. Tsiotos et al reported that patients with symptomatic malabsorption were treated with a two-week antibiotic regiment resulting in 75% resolution of symptoms [10]. Discomfort associated with chronic pneumoperitoneum can be managed with intermittent paracentesis.

Patients with symptoms refractory to these treatments require surgical therapy [17]. Evaluation should include nutritional assessment and consideration of hyperalimentation prior to intervention. The

choice of open versus minimally invasive surgery is dependent on surgeon experience and comfort. In the majority of patients, simple resection and primary anastomosis is curative. However, the surgeon must be cognizant of the length of involved small bowel, as excessive resection may lead to short gut syndrome post-operatively.

## Conclusion

While jejunoileal diverticulosis is most commonly asymptomatic, awareness of the multiple symptoms can lead to better therapy. JID should remain on the differential diagnosis in the setting of gastrointestinal hemorrhage or acute perforation with sepsis, and resection may be life-saving. A heightened awareness of JID in patients with new onset of failure to thrive, chronic abdominal pain or pneumoperitoneum may lead to improved treatment. While no consensus currently exists for the timing of surgical treatment for patients with chronic, symptomatic JID, resection can lead to significant improvement in symptoms and quality of life.

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