

Gastrointest Endoscopy Clin N Am 16 (2006) 175-187 GASTROINTESTINAL ENDOSCOPY CLINICS OF NORTH AMERICA

Sigmoid Volvulus an Update

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Sigmoid volvulus is described as far back as the Papyrus Ebers of ancient Egypt. The Egyptians recognized the natural history by which the twisted colon either spontaneously reduced or "rotted." Hippocrates recognized sigmoid volvulus and advocated using a 12-in long suppository and insufflated air to relieve the obstruction [1,2]. Soranus, an ancient Greek physician, defined the condition as a "severe and dangerous twisting of the intestines." Because of severe pain, volvulus was often referred to as "iliac passion" until the early nineteenth century. In the past, sigmoid volvulus was a rare condition. In the earliest case series reported by Crise in 1872, only 12 of 730 intestinal obstructions were caused by volvulus. Currently, it is the third most common cause of colonic obstruction in the United States after cancer and diverticulitis accounting for approximately 2% to 5% of cases of large bowel obstruction [1,3].

The frequency is higher in other parts of the world, particularly in the so-called "volvulus belt," which encompasses Africa, India, Iran, and Russia. Volvulus ranks as one of the leading causes of lower intestinal obstruction in Western Brazil, Bolivia, and Northern Argentina [4,5]. Worldwide, sigmoid volvulus is reported mostly in young healthy men with a male to female ratio of 2:1 [6–8]. In Africa, India, Iran, Pakistan, Brazil, and Eastern Europe, sigmoid volvulus is usually seen in men with a mean age of 40 to 51 years. Patients from English-speaking countries, including the United States, United Kingdom, and Israel, have a different age distribution. The mean ages in these regions are 62, 67, and 75, respectively. In these regions, sigmoid volvulus usually occurs in older

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individuals of either gender, usually with coexisting medical illnesses. Index of suspicion for volvulus should vary based on where the patient is from. In 10 American series [3,9-17] of 221 patients, 146 (two thirds) were African American and 74 (one third) white. Only one Hispanic patient was reported. In nine of these American series, 54.9% of patients were admitted from private homes, 32.4% from mental institutions, and 12.7% from nursing homes.

Etiology and pathophysiology

The cause of volvulus is multifactorial and colonic length is an important predisposing factor. The loop must be of considerable length and the meso-colon must be long and very narrow at its parietal attachment, so that the two ends of the loop may be brought as close together as possible. More recent studies undertaken by Bhatnagar and coworkers [18] document the anatomic measurements of the sigmoid colon in 70 Indian subjects (51 live and 19 cadavers). They show that the sigmoid mesocolon is brachymesocholic (wider than long). The authors hypothesize that a narrower mesocolic root with a greater vertical length of the mesocolon in the male sigmoid colon makes males more susceptible to sigmoid volvulus and may account for the lower incidence of sigmoid volvulus in females [18].

In addition to gender differences, colonic length may also be influenced by dietary factors, in particular high-fiber diets that may contribute to long redundant colons. In parts of India where volvulus of the sigmoid is common, groups at higher risk tend to have a diet higher in fiber content [19]. Further evidence comes from the changes in the incidence of volvulus noted in Norway during World War II. A marked increase in the number of patients treated for volvulus of the sigmoid colon coincided with a change to a high-fiber diet [19–21]. Andersen [22], Scott [23], and Sinha [24] in India, Saidi [25] in Iran, Bruusgaard [26] in Norway, and Andersen [27], Elmasri and Khalil [28], and Riedl [29] in Africa implicate vegetable fiber in the diet as an etiologic factor.

In addition to anatomic variation, other etiologic factors play an important role in sigmoid volvulus. These factors include chronic constipation [30–32], systemic and local neurologic disease [30,33–36], adhesions [30,37,38], pregnancy [30,31,39,40], toxins [30,41], megacolon [30,35,36,42], and metabolic disease [30,34,35,41]. Sigmoid volvulus is particularly frequent in the high altitude of the Andes [43–47]. In the series presented by Asbun and coworkers [43], 230 consecutive cases of sigmoid volvulus were treated at the Juan XXIII Hospital in La Paz, Bolivia. All patients were natives of the Andes and of the 230 patients, 111 (48%) were 40 to 60 years of age, the male to female ratio was 4.5:1, and 54% were from rural areas with low socioeconomic status. Frisancho [44] proposed the term "high altitude megacolon." It is hypothesized that the unusually high incidence may be secondary to the increased intraluminal pressures seen at altitudes of 13,000 ft. The gas generated in the lumen of the bowel (carbon dioxide, methane, and hydrogen) could contribute to chronic distention of a redundant sigmoid in susceptible individuals, although further studies evaluating intraluminal pressure are needed [43].

Sigmoid volvulus is endemic to areas where American trypanosomiasis or Chagas' disease is prevalent. One such region is Sao Paulo, Brazil. In a series of 365 patients with Chagas' disease in Sao Paulo, volvulus developed in 26.9% of patients [48,49]. The underlying mechanism for volvulus in affected individuals is thought to be loss of sympathetic and parasympathetic neurons in the submucosal and myenteric plexuses of the colon [50]. Subsequent neuron loss leads to disturbances in motility and development of megacolon. As the disease progresses, the colon becomes enlarged in both diameter and length with colonic wall thickening and constipation. These anatomic and physiologic changes, which predominate in the sigmoid colon, are risk factors for the development of volvulus.

Subsequent bowel injury is the same, regardless of etiology. The twist in the mesocolon may be clockwise or counterclockwise. Various degrees of rotation about the mesenteric axis from 180 to 360 degrees can produce a closed loop obstruction [1]. Hyperperistalsis then forces luminal contents into this closed loop and the intra-abdominal pressure and tension on the mesentery increases with progressive abdominal distention. As luminal pressure increases beyond the diastolic venous pressure and mesenteric torsion decreases blood supply, gangrene ensues. The three different points at which gangrene may occur are (1) at the neck of the volvulus, (2) the twisted sigmoid loop, and (3) the lower descending colon and upper rectum from retrograde mesenteric thrombosis [1]. Perforation of the sigmoid is uncommon because a thickened wall is able to withstand higher pressures. If the ileocecal valve is competent, a double closed-loop obstruction occurs with possible perforation of the thinner cecal wall.

Clinical features

Sigmoid volvulus may be acute or chronic with exacerbations and periods of quiescence [51,52]. The variation in presentation is likely secondary to the degree of torsion. There has been no correlation between the nature of the disease (ie, acute versus chronic) and risk of colonic gangrene [51,53]. The duration of signs and symptoms before presentation is also variable and may range from hours to days [12,34,51,54].

The triad of abdominal pain, distention, and constipation is the predominant constellation of signs and symptoms [12,51,54] encountered, and 40% to 60% of patients have a history of prior attacks [37,51,55–60]. Vomiting is a less common finding. Physical findings usually include a tympanitic and distended abdomen, typically more marked in the upper abdomen [19,51,53,61]. The distended colon may be palpable or visible during examination [51,62]. Bowel sounds are nondiagnostic and may be increased or decreased. Rectal examination reveals an empty rectum, and the presence of stool in the rectum makes the diagnosis of

volvulus questionable. The presence of blood during the rectal examination warrants consideration of the possibility of ischemia or bowel necrosis. The abdomen is usually nontender to palpation. Fever, a toxic-appearing patient, tachycardia, hypotension, and abdominal tenderness on palpation are suspicious for gangrene. Most patients come to medical attention before the development of gangrene, and in one series of 54 patients, gangrene was seen in only four patients [63,64]. It is important to remember that pain may not be elicited in the elderly and those with neurologic conditions [63,65].

Diagnosis of sigmoid volvulus

The upright plain abdominal roentgenogram is diagnostic. Radiologic findings in sigmoid volvulus include the "bent inner tube" or "coffee bean sign" deformity arising from the left lower quadrant; usually there is lack of air in the rectum [66]. The "bent inner tube" refers to a large collection of gas in an ahaustral colon that extends from the pelvis to the right upper quadrant as high as the diaphragm (Fig. 1).

Contrast studies are not necessary to make the diagnosis when it is apparent on a plain film and can lead to colonic perforation at the site of volvulus if barium is forced beyond the twist [66,67]. When contrast studies are performed a "bird's beak" deformity or an "ace of spades sign" is usually described. Although a plain film is the test of choice in diagnosing sigmoid volvulus, limitations may preclude its usefulness. Diagnostic difficulties may occur if the degree of proximal dilation is marked and the sigmoid loop is unrecognizable [68]. It may also be difficult to define the sigmoid loop if there is a large amount of fluid and small



Fig. 1. Upright posteroanterior abdominal film revealing dilated sigmoid loop. Note the paucity of air distal to the obstruction.



Fig. 2. The endoscopic "whirl" sign. The mucosa is normal in appearance.

amount of air leading to poor definition of the sigmoid colon [68]. Furthermore, volvulus of the right colon, closed loop small intestinal obstruction, and sigmoid volvulus complicated by peritonitis may simulate sigmoid volvulus. In cases where the diagnosis is in doubt, CT may be a useful adjunctive modality. CT may be useful in identifying the cause and site of obstruction resulting from other pathologies and also in demonstrating ischemia resulting from strangulation [68].

Endoscopy is diagnostic and therapeutic. Endoscopic findings include obstruction, usually 20 to 30 cm from the anal verge. The endoscopic features include spiral convergence of the colonic folds ending in a pinpoint occlusion (Fig. 2), and have been referred to as the "whirl" sign. The mucosa may appear



Fig. 3. Endoscopic view of the obstruction site with marked congestion and lack of vascular pattern.

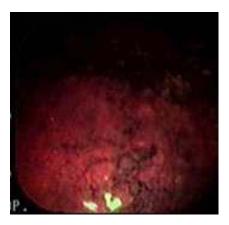


Fig. 4. Endoscopic view of obstruction site with dusky mucosa and dark black spots indicative of necrosis.

healthy (see Fig. 2) or show signs of ischemia (Fig. 3) with congestion and violaceous color. The presence of dark spots and blood suggests necrosis (Fig. 4).

Management of sigmoid volvulus

Prompt intervention is required if volvulus is suspected because death may occur as quickly as 24 hours from the onset of signs and symptoms [26,51]. A mortality rate as high as 14% to 21% has been reported in association with sigmoid volvulus [1,51,69–73] and a 52.8% mortality rate when gangrene of the intestine develops [51,69]. Patients may fare better in developing countries where patients are younger and have fewer coexisting medical conditions [51,69]. Another possible explanation for better outcomes in developing countries is the fact that the disease is more common and the correct diagnosis is made more rapidly [51,74]. In areas where it is not common, the diagnosis is frequently not entertained and patients are often incorrectly treated by blind proximal diversion procedures on a presumptive diagnosis of obstructing neoplasm [51,75–79].

The management of sigmoid volvulus involves relief of obstruction and the prevention of recurrent attacks [20]. Management may be surgical, medical, or endoscopic depending on the viability of the colon. Both options are discussed and newer less invasive techniques, such as endoscopic sigmoid pexy, are described.

Sigmoidoscopy with endoscopic decompression is considered the treatment of choice when the colon is viable. It was first described by Brusgard in 1947 [20] and more recent large series have verified its efficacy [20,26,58,64,65, 69,73,80]. The concept of using a tube for decompression dates back as far as 1859 when Gay found that while performing an autopsy on a patient with sigmoid volvulus, a rectal tube relieved the bowel of its contents and the colon "righted" itself [1,81]. With the patient in the left lateral position, the rigid sigmoidoscope is advanced to the point of volvulus (see Fig. 2). A 40-cm welllubricated rectal tube is inserted beyond the point of obstruction. The distal part of the tube should be connected to a rectal bag to avoid soiling of the operator. A successful deflation is accompanied by a large amount of released gas and liquid stool and immediate relief of abdominal distention. After the procedure, it is necessary to obtain repeat upright abdominal films to confirm reduction and absence of air under the diaphragm. After the procedure, a rectal tube should be left in place for 24 to 72 hours [26,51,54,82,83].

Most obstructions occur within the instrument's reach and have been successfully relieved in 70% to 90% of cases [20,65,69,84]. A flexible endoscope may also be used. Friedman and coworkers [30] emphasizes that flexible sigmoidoscopy may be more beneficial than rigid endoscopy because a greater number of torsion points can be reached. Endoscopic reduction should have a complication rate below 1% and is an acceptable treatment option [3,30,85]. Other methods including enemas and siphon lavage have fallen out of favor because of unacceptable rates of complications.

Any deterioration in the patient's condition should raise suspicion for perforation and immediate surgical exploration should be considered. Risks may be minimized by the use of gentle technique, a soft tube, and visualization of the point of torsion. If bloody stool is returned after the procedure, laparotomy is mandatory.

The recurrence rate after endoscopic decompression is high. In a series of 149 patients who did not have surgery after successful endoscopic decompression, the recurrence rate was 40% to 70% [69,73,85,86] depending on the series. The mortality rate in recurrent volvulus is variable and probably dependents on geography and patient characteristics. Because of the high rates of recurrence, most authors advocate sigmoid resection following decompression. Newer noninvasive techniques can be considered in individuals in whom surgery is not a reasonable option [85,87].

The technique of endoscopic sigmoidopexy has been used in place of surgical resection in certain individuals deemed to be high surgical risks. The procedure involves a technique similar to that used for percutaneous endoscopic gastrostomy placement and is performed by two operators. During the procedure, the distal limb of the redundant sigmoid loop is straightened and distended at colonoscopy and then fixed with a line of four T-fasteners, each separated by a distance of approximately 5 cm [88].

Indications for urgent laparotomy include signs of perforation, evidence of bowel necrosis, and unsuccessful operative technique [20,84,89]. If the decompressed colon is viable, then either prophylactic surgery or future endoscopic decompression may be considered. Deciding factors seem to be patient age and comorbid conditions. Arnold and coworkers [12] cite an operative mortality of 15% from resection after the first episode and a mortality rate of 9% from recurrent episodes of volvulus requiring surgery [90]. They further stratified the studied population by age and found that approximately 67% of deaths occurred in patients over the age of 70. They concluded that in patients over the age of 70, resection should be deferred until the second episode of volvulus. Elective resection during the same hospital stay is advocated for older patients by Bak and

Boley [73], however, who report a 6% mortality compared with a mortality of 30% from recurrent volvulus in those medically managed [90]. Armstrong and Ballantyne [90] make the argument that surgical consideration should be based not only on age but also the overall general health of the individual. They argue that patients above the age of 70 in reasonable health do not benefit from chancing the risks associated with recurrent volvulus and should undergo early surgical intervention. In developing countries where the patient is younger without comorbidities, it has been shown that a conservative approach is appropriate with a low mortality rate. In a series of 151 patients, 43 patients undergoing multiple decompressions on an as-needed basis had no mortality. In the authors' experience, 28% required surgery, of which only two patients required urgent surgery for necrosis.

The surgical procedure that is chosen, whether open or laparoscopic, may further complicate management of these high-risk patients [20,91-95]. If sigmoid resection is performed, some advocate primary anastomosis [20,38,71,96], whereas others advise the traditional teachings of a colostomy with a mucous fistula or a Hartmann's procedure [20,69,74,97]. Mortality rates range from 0% to 33% for primary anastomosis versus 0% to 50% for colostomy [20]. Again, the general condition of the patient and presence of comorbid conditions determine the outcome of the patient regardless of the choice of surgical procedure [20,37,51,71,98-101]. No randomized controlled studies compare different surgical options, so an evidence-based recommendation is not possible. Surgical intervention does not preclude recurrence of volvulus and rates as high as 36% in the postoperative course of 29 patients have been described [102].

Sigmoid volvulus and pregnancy

The incidence of intestinal obstruction during pregnancy varies widely between 1 in 1500 and 1 in 66,431 deliveries [103–106]. Forty-four percent of obstructions in pregnancy are related to the sigmoid colon [37,103]. Sigmoid volvulus during pregnancy was first reported by Braun in 1885 [103]. This patient developed a volvulus during pregnancy and she subsequently died. The diagnosis was ultimately made during autopsy. Since this reported incident, there have been 83 cases of sigmoid volvulus during pregnancy reported in the world literature. Only four cases have been reported since 1985, with the most recent case reported by Alshawi in 2005 [107].

In 1944, Kohn and coworkers [40] performed a statistical analysis of the 78 cases of volvulus during pregnancy. Age ranged from 18 to 41 with most cases under 30 years of age. Three of the cases were primigravidas, 12 were multigravidas, and three authors did not report the parity. This study suggests that multiparity may predispose to volvulus and hypothesizes that this may be caused by relaxation of the abdominal wall musculature [40]. Most cases of volvulus occur at term when the abdomen is distended by the pregnant uterus [40]. Overall, pregnancy is a precipitating factor because the enlarging uterus causes a redundant or an abnormally long sigmoid colon to rise out of the pelvis and twist around its point of fixation on the pelvic side wall [37]. Torsion in pregnancy may vary from 135 to 800 degrees with a greater incidence in the counterclockwise direction [40].

The diagnosis of sigmoid volvulus in pregnancy is often delayed and the median length of time from admission to laparotomy is approximately 48 hours [106]. This may be secondary to pregnancy clouding the clinical picture because abdominal pain, nausea, and leukocytosis may occur in normal pregnancies [106]. Another factor is the hesitancy to perform radiologic examinations in pregnant women.

The diagnosis is suspected when a pregnant woman presents with a clinical triad of abdominal pain, distention, and constipation. The patient may give a history of varying degrees of constipation for several days [40]. This is followed by abdominal pain, which may be colicky, and increases in severity. Vomiting may be a variable symptom depending on the location of the volvulus. Volvulus of the small bowel usually produces early emesis. Colonic volvulus can lead to vomiting in later stages [40]. In the series by Perdue and coworkers [106] of 66 pregnant patients with intestinal obstruction, including cases of volvulus, abdominal pain was present in 98% of patients, vomiting in 82%, and tenderness to palpation in 71%. The plain abdominal roentgenograms may be diagnostic in 80% of cases demonstrating a bent inner tire sign, its apex pointing toward the left lower quadrant [98]. Examination of the abdomen usually reveals distention above the volvulus and abdominal tenderness is generalized if present [40].

As Perdue and coworkers [106] emphasize, prompt management of obstruction is essential and begins with fluid resuscitation, electrolyte repletion, and prophylactic antibiotics. Surgical colleagues should also be involved early because bowel strangulation requiring surgery may be as high as 23%.

When there is no evidence of peritonitis in the first trimester, the treatment is nonoperative and consists of colonic decompression and detorsion and rectal tube decompression [107]. This may be repeated in recurrent cases until the second trimester when sigmoid colectomy is recommended. Surgery is usually postponed if possible until the second trimester because operating in the second trimester is associated with a lower rate of miscarriage [107]. This benefit is offset by the rate of high recurrence by nonoperative management. In the presence of nonviable colon or unsuccessful reduction of volvulus, sigmoid colectomy and Hartmann's procedure is advisable regardless of the gestational age of fetus.

Although volvulus is uncommon during pregnancy, it is a condition that poses significant risk to both mother and fetus. This complication was first reported by Houston in 1830 and again in a review on intestinal obstruction during pregnancy in 1937 [106,107]. The maternal and fetal mortality rates were 21% and 50%, respectively [106,107]. In a more recent retrospective review of 66 cases of intestinal obstruction complicating pregnancy, Perdue and coworkers [106] report that 38% of patients completed term pregnancies after operative resolution of the obstruction. The total maternal mortality rate in this series was 6% and total fetal mortality 26%.

Prompt recognition and intervention are keys to decreasing the mortality rate. Although surgery seems to be advocated in most cases, there are a few case reports where medical management has been successful. Alshawi [107] reports a case of a 22-year-old pregnant Saudi woman, gravida 2, para 1, who presented with sigmoid volvulus at 28 weeks. Colonic detorsion and rectal tube decompression was successful; the patient presented with recurrence at 35 weeks and a repeat procedure was successfully performed [107]. At 38 weeks' gestation and after induction of labor, a healthy baby was delivered. Two weeks later the patient underwent an uncomplicated sigmoid colectomy and was discharged 5 days later [107].

Summary

Although sigmoid volvulus is a rare condition dating back to Egyptian times, it is an evolving disease with an improved prognosis during the past few decades [20,108]. The operative mortality of 75% reported at the beginning of the twentieth century has decreased to about 2% [108,109]. This reduction in mortality is likely caused by early recognition of the disease and advances in endoscopic decompression and surgical techniques. It is difficult to ascribe a single cause or pathophysiologic basis for this disease entity because sigmoid volvulus has a variable presentation based on geography, gender, and age of the patient. It is important that the astute physician keep these factors in mind when trying to diagnose sigmoid volvulus in a particular patient.

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