
Spontaneous Free Perforation and Perforated Abscess in 30 Patients with Crohn's Disease

ADRIAN J. GREENSTEIN, M.D.
DAVID B. SACHAR, M.D.*

DANNY MANN, B.A.
PREM LACHMAN, M.D.

TOMAS HEIMANN, M.D.
ARTHUR H. AUFSES, JR., M.D.

Spontaneous free perforation is an uncommon event in the natural history of Crohn's disease. It occurred in 21 of 1415 patients (1.5%) admitted with Crohn's disease to The Mount Sinai Hospital between 1960 and 1983. The mean duration from onset of Crohn's disease to occurrence of perforation was 3.3 years. Ten patients had small bowel perforation, ten patients had large bowel perforation, and one patient had simultaneous perforation of both ileum and cecum. The incidence of perforation in disease segments of small bowel was 1.0% (jejunum 6.0%, ileum 0.7%), and in the colon, 1.3%. Besides the 21 patients with spontaneous free perforation, an additional nine patients had spontaneous free rupture of an abscess into the peritoneal cavity. The mean duration from onset of Crohn's disease to rupture of abscess was 8.5 years. All 30 patients had surgery within 24 hours of perforation or rupture. All 21 patients with spontaneous free perforations survived, as did all but one of the nine patients with perforated abscess. The cornerstone of the treatment of ileocolonic lesions perforating into the general peritoneal cavity is proximal diversion with delayed reconstruction of intestinal continuity whenever possible. With perforation of the small bowel, primary reanastomosis is possible in selected patients.

FREE PERFORATION OF THE BOWEL in inflammatory bowel disease has been an uncommon but often lethal complication.¹⁻³ Free perforation occurs more frequently in ulcerative colitis^{1,4-6} than in Crohn's disease,⁷⁻⁹ but in ulcerative colitis it invariably occurs in the colon, usually as a consequence of toxic dilation, whereas in Crohn's disease it may also occur in the small bowel, generally without a prelude of intestinal dilatation.

Morgagni in 1769 may have provided the first description not only of Crohn's disease but also of free perforation in Crohn's disease.¹⁰ In 1935, Arnheim reported a patient at autopsy with a perforated abscess due to regional en-

From the Departments of Surgery and Medicine (Gastroenterology) of the Mount Sinai School of Medicine of the City University of New York; and The Mount Sinai Hospital, New York, New York*

teritis.¹¹ Two years later, Halligan described a 40-year-old woman with ileal perforation who also died.¹² Since that time, at least 84 well-documented cases of free perforation have been reported.¹³ Most cases have involved the ileum (62, or 74%),^{3,7,8,12-15} with a smaller number occurring in the jejunum (10, or 12%)^{8,16,17} or colon (12, or 14%).^{13,18-22} The mortality rate of this complication appears to have been decreasing.¹³

We review the data of 30 patients with Crohn's disease with spontaneous free perforation (21 patients) or free perforation of an abscess (9 patients) seen at The Mount Sinai Hospital. We also list the sites of free perforation in another 13 patients who had Crohn's disease before admission to The Mount Sinai Hospital.

Materials and Methods

The records of 1415 patients with Crohn's disease who were admitted to The Mount Sinai Hospital between 1960 and 1983 were obtained from the computer files of the hospital and were reviewed retrospectively. The diagnosis of granulomatous disease was based on the criteria reported by Lockhart-Mummery and Morson,²² and Cook and Dixon,²³ and discussed in a previous publication.²⁴ The clinical features required for the diagnosis of Crohn's disease include radiologic, pathologic, or endoscopic evidence of transverse fissure formation or fistulization, asymmetrical mucosal involvement, aggregated inflammatory pattern, confluent linear ulceration, segmental involvement, right-sided disease, and noncaseating epithelioid granulomas.

Free perforation due to Crohn's disease was defined as spontaneous rupture of the small or large bowel with free

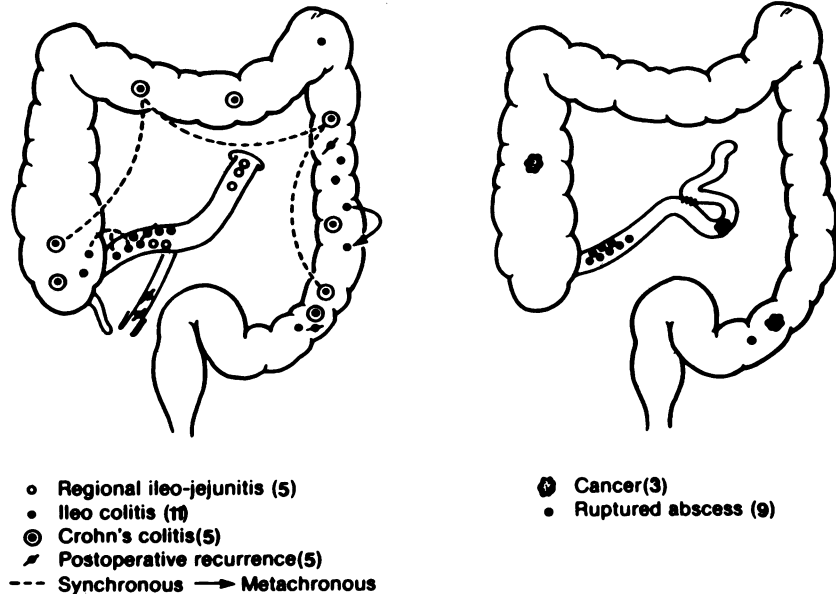
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Reprint requests and correspondence: Adrian J. Greenstein, M.D., Mount Sinai Medical Center, One Gustave L. Levy Place, New York, NY 10029.

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FREE PERFORATION IN PRIMARY (21) OR
RECURRENT (5) CROHN'S DISEASEFREE PERFORATION OF CANCER (3) OR
RUPTURED ABSCESS (9) IN CROHN'S DISEASE

FIG. 1. Sites of free perforation (primary or malignant) or ruptured abscess in Crohn's disease.



flow of intestinal contents into the general peritoneal cavity. Ruptured abscess was defined as spontaneous rupture of an intraperitoneal abscess into the greater peritoneal cavity.

Among the 1415 patients with Crohn's disease admitted to The Mount Sinai Hospital, 53 patients had free perforation of small or large bowel, or rupture of an abscess into the free peritoneal cavity. We excluded from this report 13 patients with free perforation who were not seen at this hospital at the time of perforation (7 small bowel perforations, 4 large bowel perforations, and 2 perforations of abscesses originating in the ileum). Also excluded were five patients in whom the perforation occurred after previous surgery, three patients with iatrogenic perforation after endoscopy, and three patients with perforation due to adenocarcinoma. The three perforated cancers were located in the ascending colon, the sigmoid colon, and in a bypassed loop of jejunum, and will be reported on separately. Among the 23 patients excluded from this study, death occurred in two of the five postoperative patients and in the three patients with perforated cancers.

Results

There were 21 patients with free perforation, 12 men and 9 women. Perforation occurred in five of 554 patients with regional enteritis (0.9%), 11 of 602 patients with ileocolitis (1.8%), and five of 259 patients with Crohn's colitis (1.9%).

The sites of free perforation are diagrammed in Figure 1. Eleven small bowel perforations occurred (one having a cecal perforation in addition), for an overall incidence of perforation in diseased segments of small bowel, in-

cluding regional enteritis and ileocolitis, of 1.0% (11 of 1156 patients). Three jejunal perforations occurred among approximately 50 patients with jejunitis, jejunoileitis, or jejunoileocolitis (6.0%), and eight ileal perforations occurred among 1156 patients with jejunoileitis, ileitis, or ileocolitis (0.7%). The colon was the site of perforation in 11 of 861 patients (1.3%) with Crohn's colitis or ileocolitis, including the patient with simultaneous ileal perforation. In one patient there were four synchronous colonic perforations, and in another patient there were two metachronous colonic perforations (Fig. 1).

Among the 21 patients with free perforation, the mean age at onset of disease was 27.4 years (range: 13–63 years), and at the time of perforation 30.7 years (range: 14–65 years). Perforation was thus a relatively early complication in Crohn's disease, occurring at a mean time of 3.3 years (range: 1 month–10 years) from the onset of disease. Free perforation occurred within 1 year of onset in nine patients (43%), and within 5 years of onset in 17 patients (81%).

Rupture of an established intra-abdominal abscess into the free peritoneal cavity occurred in nine patients (0.6%): eight patients with ileitis and one patient with colitis, 5 males and 4 females. The mean age at onset of disease was 23.3 years (range: 11–36 years) and the mean time of rupture of the abscess was 32.8 years (range: 15–56 years). Thus, the duration from age at onset to rupture was more than twice as long, 8.5 years, compared with 3.3 years for free perforation.

Although the diagnosis of free perforation is usually fairly obvious, the administration of large doses of steroids may sometimes obscure the clinical picture. Fourteen of 21 patients with free perforation had been on steroids

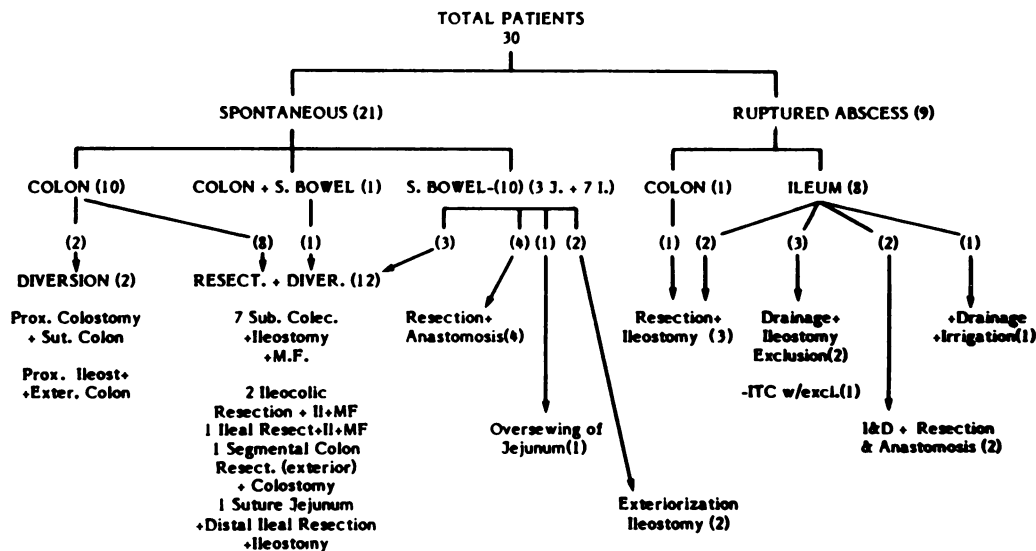


FIG. 2. Surgery for free perforation in Crohn's disease. MF = mucous fistula; II = ileostomy.

before the perforation. In this study, sudden severe abdominal pain occurred in 15 of 21 patients, disappearance of bowel sounds occurred in nine patients, and board-like abdominal rigidity occurred in eight patients. An upright film revealed free air under the diaphragm (associated with loss of liver dullness) in six patients; four patients with colonic, one patient with ileal, and one patient with jejunal perforations. However, all patients had at least one or more of the above findings.

Surgery was performed within 24 hours of perforation in all 30 patients. The operative procedures are shown in Figure 2.

All 21 patients with spontaneous perforations and all but one of the nine patients with perforated abscess survived. There was, however, considerable postoperative morbidity. Among the 21 patients with free perforation, major wound infections occurred in six patients, intra-abdominal abscess occurred in four patients, and wound dehiscence, pulmonary embolism, transient renal failure, transient jaundice, fecal fistula, and seizures occurred in one patient each. Among the nine patients with ruptured abscess, one patient had a wound infection and one patient had postoperative hepatic failure. The one death in the abscess group occurred 60 days after surgery and was due to serum hepatitis with hepatic necrosis.

Discussion

Since the first report from The Mount Sinai Hospital that described the necropsy findings of a ruptured ileal abscess in 1935,¹¹ 11 additional cases of ruptured abscess^{20,25-28} or free perforation^{18,20,26,28} of the bowel in patients with Crohn's disease have been reported from this institution.^{18,20,25-28} Although Crohn had not seen free perforation at this hospital up to 1957,²⁹ he had reported seven cases by 1965, all of which occurred in acute ileitis.^{9,30} However, only a single case was fully described,^{9,30}

and it is not clear whether any of them had been previously reported. An extraordinary feature of Crohn's study was postoperative survival of all seven patients.³⁰ One of the nine abscess cases in the current study has been previously described,¹⁸ and three were previously mentioned²⁷ by us. We have omitted from this current study ten of the cases previously reported from this institution, plus any cases that occurred earlier than 1960.

Our 21 cases of free perforation bring the number of fully described cases to at least 105.^{3,13} We recently reported on 99 cases of free perforation that included 15 cases in the current study.¹³ Although Menguy believed that more than 100 cases had been reported,³ and Kyle collected information on 91 cases and added an additional four,³¹ not all have been described in detail in the medical literature.¹³

The incidence of 21 patients with free perforation in this study of 1415 patients (1.5%) is very similar to previously reported incidences. In 963 patients with Crohn's disease reported or described by Steinberg et al.⁸ in 1973 and others,³²⁻³⁵ there were 13 patients with free perforation (1.4%). However, unlike most previous reports, our patients had as many colonic as small bowel perforations (11 each, one patient having both). However, in patients who had perforation before admission to The Mount Sinai Hospital there were a greater number of small bowel perforations (7 vs. 4). Only 19 cases of colonic perforation have been previously reported,^{19,36} including three cases of ruptured abscess and three postoperative cases.¹³ The high number of colonic perforations in our study probably reflects the high proportion of Crohn's colitis and ileocolitis at our hospital; the colon was involved in 861 of 1415 patients (61%). Moreover, the actual incidence of colonic perforation among patients with colitis and ileocolitis in this study (1.3%) was somewhat higher than the incidence of small bowel perforation in regional enteritis and ileocolitis (0.95%). Perhaps the incidence of

free colonic perforation in Crohn's disease is higher than most previous reports suggest. For example, Fisher et al. reported three cases that occurred within 8 months at the University of Kansas Medical Center.³⁷ They suggested that other cases of free perforation in Crohn's disease may have been mistakenly classified as idiopathic ulcerative colitis. Pneumoperitoneum is a relatively uncommon finding in perforations of the small bowel in Crohn's disease.³⁸ Suk et al.,³⁶ who reported a patient with two colonic perforations and air under the right hemidiaphragm, were unable to establish the true incidence of pneumoperitoneum with colonic perforation. Four of our ten patients demonstrated this sign. This high incidence may be due to an increased amount of air in the colon as compared with the small bowel.

In our review of the literature,¹³ free perforation was the initial symptom of Crohn's disease in 25 of 84 patients (30%).^{9,12,39-41} Free perforation was not the presenting symptom in any of our 21 patients,* having occurred within one year of onset of disease in nine patients and within 5 years of onset in 17 patients. The mean disease duration of 3.3 years before free perforation was similar to the mean interval before development of toxic dilation (5 years or less), but shorter than the duration to development of ruptured abscess (8.6 years) or other perforating complications, such as intra-abdominal abscess⁴² and ileovesical fistula (10 years each).⁴³ The relatively short duration from onset to perforation would tend to support the original theory of Crohn that free perforation (and also perhaps colonic dilatation),^{2,18,44} occurs before the protective granulomatous fibrotic and cicatrizing reactions take place.^{9,45} By contrast, perforation secondary to carcinoma is inevitably a very late complication of Crohn's disease (Table 1) since carcinoma takes so long to develop in these patients.⁴⁶

The underlying mechanism for free perforation may be bowel distention with increased intraluminal pressure proximal to an obstruction. This phenomenon occurred in at least one patient in the current series, and has been previously reported in other studies.^{3,7,8,28,44,46,47} A more speculative hypothesis is that of Harjola et al., who postulated ischemia as the principal pathogenetic factor.¹⁶ Although this circulatory complication occurs in some cases of toxic dilation in which intramural infarction results in perforation, in other cases, perforation may occur in the absence of colonic dilatation.⁴⁸ Kyle has described enteritis of small blood vessels that is consistent with the ischemic hypothesis.^{31,46}

Although the administration of steroids in Crohn's disease has been reported to be associated with an increased morbidity and mortality rate⁴⁹ and a higher recurrence

TABLE 1. Duration From Onset of Disease to Perforation in 24 Patients with Crohn's Disease

	No. of Patients	Age at Onset (years)	Age at Perforation (years)	Duration of Symptoms (years)
Free perforation (primary)	21	27.4	30.7	3.3
Ruptured abscess	9	23.3	31.8	8.5
Free perforation (carcinoma)	3	13.3	44.3	31.0

rate,⁵⁰ such therapy is clearly not the major factor in the development of free perforation. In this study, steroids had been given to 14 of 21 patients, a proportion not very different from that in the overall Crohn's disease population. In a previous study by Steinberg et al.,⁸ only two of seven patients had received steroids. Nasr et al.⁷ reported steroid administration in four of five patients but only two of 36 patients.

The mortality rate of free perforation has always been assumed to be formidable. However, a review of the literature suggests that survival is contingent on correct and appropriate surgical management. The mortality rate associated with simple suture, particularly in earlier reported series, was high (41%).¹³ However, with resection the mortality rate was only 4%.¹³ It is possibly even lower with diversionary procedures with or without resection, as was found in this study, and by Menguy.³ This relatively low mortality rate is in sharp contrast to perforation in ulcerative colitis, which is associated in some reports with mortality rates of over 40%.¹ It is difficult to explain this difference in the mortality rates of perforation between these two forms of inflammatory bowel disease. One may speculate that the large number of jejunoileal perforations in Crohn's disease, with lower anaerobic bacterial counts, is a factor. However, all 11 patients with colonic perforations in our current study survived, and nine of 11 previously reported patients with spontaneous, free colonic perforation without abscess also survived.¹³ Perhaps local adhesions or the focal nature of the lesions in Crohn's disease may contribute to the better prognosis.

The excellent survival rate in this study may be explained by the consistent policy of early surgery with resection or diversion or both. Of the 21 patients with free perforation, four patients had resection and anastomosis and 16 patients had some form of diversion proximal to the perforation. One of the latter patients with suture and distal ileal resection, without proximal diversion, had re-perforation and required reoperation and diversionary jejunostomy. Although a second patient was successfully treated by suture of a jejunal perforation, we cannot recommend this form of treatment in view of the high mortality rates reported. In the patients with abscesses, diversion was the cornerstone of treatment with or without concomitant resection. Delayed resection and reconstruc-

* Since this study was completed one patient has been seen in whom perforation was the presenting manifestation.

tion of continuity is possible after recovery in all patients in whom a diversionary procedure was performed, provided the distal colon, rectum, and perianal areas are not severely diseased.

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