

# Spontaneous free perforation of the small intestine in Crohn's disease

Hugh J Freeman MD

HJ Freeman. Spontaneous free perforation of the small intestine in Crohn's disease. *Can J Gastroenterol* 2002;16(1):23-27.

Spontaneous free perforation of the small intestine is a rare but often dramatic event in the clinical course of Crohn's disease. Fifteen new cases of spontaneous free perforation of the small intestine – nine female patients and six male patients – were discovered in a series of 1000 consecutively evaluated patients with Crohn's disease seen during a period spanning 20 years, for an estimated frequency of 1.5%. Spontaneous free perforation was the presenting clinical feature of Crohn's disease in nine (60%) of the newly discovered cases. Most perforations were located in the ileum rather than in the jejunum, and there were no duodenal free perforations. One patient with extensive intestinal disease presented with concomitant free perforations of the jejunum and ileum, while a second patient had two free ileal perforations that developed independently, separated by about six years. No perforations were the result of a superimposed malignant process, ie, adenocarcinoma or lymphoma. There have been no mortalities, and the subsequent clinical course of these patients has been limited to a minority requiring corticosteroid or immunosuppressive medications, or further surgical resections.

**Key Words:** *Crohn's disease; Free perforation; Ileal perforation; Intestinal ulceration; Jejunal perforation; Peritonitis*

## Perforation libre spontanée de l'intestin grêle et maladie de Crohn

**RÉSUMÉ :** Les perforations libres spontanées de l'intestin grêle constituent un accident rare mais souvent dramatique de l'évolution clinique de la maladie de Crohn. Quinze nouveaux cas de perforation libre spontanée de l'intestin grêle chez neuf femmes et six hommes ont été relevés dans une série de 1000 patients souffrant de la maladie de Crohn, examinés consécutivement sur une période de 20 ans (fréquence : 1,5 %). La perforation libre spontanée a été la manifestation clinique de la maladie de Crohn qui a motivé la consultation dans neuf (60 %) des nouveaux cas. La plupart des perforations touchaient l'iléon plutôt que le jéjunum, et aucune ne se situait dans le duodénum. Un patient porteur d'une atteinte intestinale étendue présentait des perforations libres concomitantes du jéjunum et de l'iléon et un autre a connu deux perforations iléales libres, indépendantes, qui se sont produites à six ans d'intervalle. Aucune perforation n'a résulté d'un processus malin surajouté, c'est-à-dire d'un adénocarcinome ou d'un lymphome. On n'a pas enregistré de décès, et l'évolution clinique s'est limitée, chez un petit nombre de patients, à un traitement aux corticostéroïdes ou aux immunodépresseurs ou encore à la résection chirurgicale.

Department of Medicine (Gastroenterology), University of British Columbia, Vancouver, British Columbia  
Correspondence and reprints: Dr Hugh Freeman, Gastroenterology, ACU F-137, University of British Columbia Hospital,  
2211 Wesbrook Mall, Vancouver, British Columbia V6T 1W5. Telephone 604-822-7216, fax 604-822-7236  
Received for publication July 23, 2001. Accepted August 21, 2001

Crohn's disease is characterized by a transmural intestinal inflammatory process with deep ulcers and the formation of abscesses or fistulous tracts into adjacent, often adherent intestinal and nonintestinal structures. Free spontaneous perforation into the peritoneal cavity is a dramatic event that is, fortunately, rare but necessitates urgent surgical intervention. It was initially described in 1935 as a fatal case of free perforation of the ileum (1). Since then, about 100 cases have been described. In part, this is due to differing criteria for the clinical definition of free perforation because sealed perforations or ruptured abscesses, as noted by others (2,3), may have been included in earlier reports. Moreover, there is only limited information on the subsequent clinical course of patients who have suffered from this intestinal complication, because of its rarity in patients with Crohn's disease.

In a previous report from this hospital (the University of British Columbia) (4), a clinical database of patients with Crohn's disease was described, including classification of the initial 877 patients. In this tertiary care teaching hospital setting, young adults, predominately females (56.1%), were affected with a high rate of intestinal complications. The present report further explores this specific complication of spontaneous free perforation of the small intestine in a single clinician series of Crohn's disease, with 1000 consecutively evaluated patients. This series not only provides, for the first time, data from a Canadian experience with this complication in patients with Crohn's disease, but also allows the evaluation of the longer term clinical course after initial treatment for this intestinal complication.

#### PATIENTS AND METHODS

A total of 1000 consecutively evaluated patients with Crohn's disease were evaluated. All patients were seen directly by the author, and, as previously noted (4), a diagnosis of Crohn's disease was established on the basis of defined criteria (5). Free perforation of the small intestine was defined by surgical and pathological definitions of a focal small intestinal perforation with peritonitis, and excluded sealed perforations or ruptured abscesses. No patient had undergone prior small intestinal instrumentation, and pathological evaluation of resected tissues in all patients confirmed the pathological diagnosis of Crohn's disease. No patient had another or superimposed cause for small intestinal ulceration or perforation, including carcinoma (6,7) or lymphoma (3,8).

Patients with colonic perforation or appendiceal disease were excluded, and no patients have been seen to date with spontaneous free esophageal or gastroduodenal perforation (9,10).

#### RESULTS

Fifteen patients (nine female [60%] and six male [40%]) with Crohn's disease were seen with spontaneous free perforation of the small intestine in this series of 1000 consecutively evaluated patients (1.5%) – consistent with the estimated frequency of this complication in the literature

**TABLE 1**  
Clinical features of Crohn's disease in 15 patients with spontaneous free perforation of the small intestine

Patient	Age at Dx (years/sex)	Extent of disease	Steroids*	Perf <sup>†</sup>	Other
1	22/M	Jejunem, ileum, colon	Yes	No	Renal calculi
2	22/F	Ileum, colon	No	No	
3	31/F	Ileum, colon	No	Yes	
4	21/F	Jejunem, ileum, colon	No	No	
5	37/M	Ileum	No	Yes	
6	19/F	Ileum	No	Yes	
7	34/M	Ileum, colon	No	No	Renal calculi
8	19/M	Ileum	No	Yes	
9	19/F	Ileum, colon	Yes	No	Two perforations
10	29/F	Ileum, colon	No	Yes	
11	14/F	Ileum	No	Yes	
12	22/F	Jejunem, ileum, colon	No	Yes	
13	32/F	Ileum, colon	Yes	No	
14	25/M	Ileum, colon	No	Yes	
15	17/M	Ileum, colon	No	Yes	

\*Taking steroids at the time of perforation (Perf); <sup>†</sup>Presentation of Crohn's disease with free perforation. Dx Diagnosis; F Female; M Male

from the United States and the United Kingdom (2,3,11). In one patient (patient 12), two perforations were detected at the time of the event, one in the jejunum and one in the ileum; for the other 14 cases, there was a single perforation with only one other jejunal free perforation. One patient (patient 9) had two independent episodes of free ileal perforation, separated by a period of six years.

#### Crohn's disease

Table 1 details the characteristics of the Crohn's disease for each patient. In this series, the mean age of diagnosis of Crohn's disease was 23.2 years (range 14 to 32 years) for female patients and 25.6 years (range 17 to 34 years) for male patients. All patients had ileal involvement, while three also had extensive small intestinal disease in both the jejunum and the ileum; most also had colonic disease. In nine of 14 patients, spontaneous free perforation of the small intestine was the presenting clinical manifestation of

previously undiagnosed Crohn's disease. In three of the remaining six patients who had a pre-existent diagnosis of Crohn's disease, initiation of corticosteroid therapy (such as oral prednisone) was recorded during the month preceding free perforation. Except for renal calculi in two cases, extraintestinal manifestations of Crohn's disease were not recorded. All patients are currently alive, and none has developed a superimposed malignant disorder. Two patients have moved from the province and are no longer followed up by the author.

### Free perforation

Table 2 details the clinical features of the small intestinal perforations. In 14 cases, the perforation was usually a single dramatic clinical event. In addition, one patient in this series (patient 9) had an independent, second ileal free perforation six years after the first free perforation.

Recurrent free perforation of the ileum has been previously recorded, but it was not clear whether Crohn's disease had been present (12). In the present series, all patients had single free perforations of the small intestine with peritonitis, except for one patient (patient 12) who had multiple perforations, as recorded elsewhere (3), in the jejunum and ileum. Moreover, almost all of the perforations occurred in the ileum, except in two of three patients who had extensive jejunal involvement with Crohn's disease and an associated jejunal perforation. Associated colonic free perforation was not seen. In all patients, the cause of the perforation was attributed to the Crohn's disease and not to a superimposed carcinoma (6,7) or lymphoma (3,8). All patients required urgent surgical treatment with resection and anastomosis or diversion; no patient was treated with plication alone. Subsequent laparotomy was required to re-anastomose bowel for those requiring an initial diversion, and two patients (patients 4 and 12) required further surgical treatment for a subsequent enterocutaneous fistula. In this series, most patients required a modest small intestinal resection, with only three having more than 50 cm of small intestine removed (see Table 2).

### Subsequent clinical course

Table 3 details the clinical course of patients after free perforation. The mean length of follow-up for all patients was 11.4 years (range three to 22 years). During their course, all patients were treated with a form of 5-acetylsalicylic acid-containing medication such as sulfasalazine or mesalamine, but only 40% required corticosteroids or immunosuppressive medications. In addition, 40% had a further ileocolic resection for fibrostenosing disease or an enteric fistulous tract. One patient with an ileal perforation required a second resection for another ileal free perforation. Interestingly, in spite of a dramatic presentation with free small intestinal perforation, over 40% of patients with follow-up data for over 10 years have had no significant clinical disease requiring either corticosteroid treatment or further surgical treatment.

**TABLE 2**  
Features of free perforation (perf) in 15 patients with Crohn's disease

Patient	Age at perf (years)	Site of perf	Single or multiple perf	Initial treatment*
1	23	Ileum	Single	Ileocolic resection (45)
2	44	Ileum	Single	Ileocolic resection (30)
3	31	Ileum	Single	Ileocecal resection (25)
4	27	Jejunum	Single	Jejunostomy/dr and ileocolic resection (60)
5	37	Ileum	Single	Ileocecal resection (20)
6	21	Ileum	Single	Ileocolic resection (50)
7	34	Ileum	Single	Ileostomy/dr and ileocolic resection (60)
8	19	Ileum	Single	Ileocecal resection (26)
9	28	Ileum	Single	Ileostomy/dr and ileocecal resection (30)
	34	Ileum	Single	Ileocolic resection (30)
10	30	Ileum	Single	Ileocecal resection (10)
11	18	Ileum	Single	Ileocecal resection (20)
12	22	Jejunum, ileum	Multiple (two)	Jejunal and ileocecal resections (21)
13	32	Ileum	Single	Ileostomy/dr and ileocolic resection (10)
14	25	Ileum	Single	Ileocolic resection (90)
15	17	Ileum	Single	Ileocolic resection (30)

\*Numbers in parentheses are the lengths of small intestinal resection (cm). dr diversion

## DISCUSSION

Spontaneous free perforation of the small intestine in patients with Crohn's disease is an uncommon, even rare clinical event, occurring in less than 2% (3). In a large

**TABLE 3**  
**Follow-up of patients with Crohn's disease after free perforation**

Patient	Current age (years)	Total years seen	Later medications	Later complications or surgery
1	45	13	5-ASA, prednisone	Stric resection 11 years later
2	54	9	5-ASA, prednisone, 6-MP, budesonide, infliximab, flagyl	Rectovaginal fistula
3	45	5	5-ASA	None known*
4	45	17	5-ASA	Enterocutaneous fistula and stric; resection 7 years later
5	56	19	5-ASA	None to date
6	21	3	5-ASA, prednisone	Stric resection 3 years later
7	62	14	5-ASA, prednisone	Stric resection 8 years and 13 years later
8	21	3	5-ASA	None to date
9	46	15	5-ASA, prednisone, 6-MP	2 resections for perforations
10	50	21	5-ASA	None to date
11	33	22	5-ASA	None to date
12	29	6	5-ASA	Enterocutaneous fistula and stric resection 1 year later
13	36	4	5-ASA, prednisone	Ileosigmoid fistula and stric resection 4 years later
14	56	10	5-ASA	None to date
15	41	10	5-ASA	None known*

\*Patients moved. ASA Acetylsalicylic acid; 6-MP 6-Mercaptopurine; Stric Stricture

group of 1010 patients reported from Mount Sinai Hospital in New York, New York, from 1960 to 1980, that is comparable with the present series, free intestinal perforation was seen in 15 patients, but only six in the small intestine, including two in the jejunum and four in the ileum (2). Similarly, five small intestinal perforations occurred in 360

cases of Crohn's disease reported from Birmingham, United Kingdom, from 1943 to 1972 (13). Using similar rigorous criteria for definition of free perforation, 15 patients with 16 independent episodes of spontaneous free small intestinal perforation were seen in 1000 consecutive cases of Crohn's disease observed during a similar time frame of over 20 years. While large series of patients with Crohn's disease with this complication are limited (2,3,13), this Canadian experience confirms the unusual frequency of this intestinal complication reported from American and British hospitals.

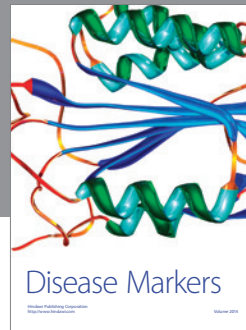
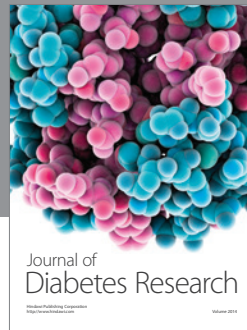
The site distribution and multiplicity of free perforations were reported in this series. Although there were two jejunal perforations (patients 4 and 12), similar to the number in the Mount Sinai series (2), there were 15 free ileal perforations, including one occurring concomitantly with a jejunal perforation (patient 12) and one occurring as a second independent episode of free ileal perforation (patient 9). Others have also recorded a significant preponderance of ileal perforations compared with other sites in the gastrointestinal tract (4,13). In a previous study of 877 patients (4), approximately 13.1% of patients with Crohn's disease had involvement of the upper gastrointestinal tract that frequently included the jejunum. The relative proportion of jejunal and ileal perforations in this series suggests that patients with Crohn's disease do not have a specific site-related propensity to develop spontaneous free small intestinal perforation. Rather, the dramatic appearance of this complication likely reflects the overall distribution of involvement with Crohn's disease in the gastrointestinal tract.

The pattern of clinical behaviour for patients with a free perforation was evaluated. Previous studies of Crohn's disease have suggested that patients with either fibrostenosing (stricturing) or fistulizing (penetrating/perforating) disease tend to redevelop a similar pattern of recurrent fibrostenosing or fistulizing disease. Although one patient developed a second spontaneous free ileal perforation about six years later as a completely independent event, this is rare. In those who required subsequent surgical treatment after the initial treatment for the perforation, it was usually for a subsequent stricture rather than for a perforating or penetrating complication, eg, fistula or abscess. Perhaps most significant in the present series was the frequent occurrence of free perforation as the initial event in the clinical course of Crohn's disease and the relatively benign clinical course after surgical treatment for this initial event, even though extensive ileocolonic disease was usually present. Only six cases have required recurrent surgical treatment to date, after over 10 years of follow-up, and most have not required any form of immunosuppression, including corticosteroids. Earlier reports (2,3,14) emphasized the high morbidity and mortality of this complication in patients with Crohn's disease, possibly reflecting the historical reluctance to treat surgically or the use of suture plication of the defect alone as the definitive treatment – approaches that were not used in the present series. In spite of the initial emergent clinical presentation for each case, the prognosis appeared to be sat-

isfactory. There were no mortalities, and only a limited number of cases have required any further surgical resections.

## REFERENCES

1. Arnheim EE. Regional enteritis with perforation, abscess and peritonitis. *J Mt Sinai Hosp* 1935;2:61-3.
2. Greenstein AJ, Mann D, Sachar DB, Aufses AH Jr. Free perforation in Crohn's disease. A survey of 99 cases. *Am J Gastroenterol* 1985;80:682-9.
3. Katz S, Schulman N, Levin L. Free perforation in Crohn's disease: a report of 33 cases and review of literature. *Am J Gastroenterol* 1986;81:38-43.
4. Freeman HJ. Application of the Vienna Classification for Crohn's disease to a single clinician database of 877 patients. *Can J Gastroenterol* 2001;15:89-93.
5. Lennard-Jones JE. Classification of inflammatory bowel disease. *Scand J Gastroenterol Suppl* 1989;170:2-6.
6. Mauer I, Miller H, Levitan R. Small bowel perforation in regional enteritis resulting from adenocarcinoma. *Arch Intern Med* 1972;130:275-6.
7. Heathcote J, Knauer CM, Oakes D, et al. Perforation of an adenocarcinoma of the small bowel affected by regional enteritis. *Gut* 1980;21:1093-6.
8. Shaw JHF, Mulvaney N. Hodgkin's lymphoma: a complication of small Crohn's disease. *Aust NZ J Surg* 1982;52:34-6.
9. Fisher J, Mantz F, Calkins WG. Colonic perforation in Crohn's disease. *Gastroenterology* 1976;71:835-8.
10. Katz S, Talansky A, Kahn E. Recurrent free perforation in gastroduodenal Crohn's disease. *Am J Gastroenterol* 1983;78:722-5.
11. Alexander-Williams J. The place of surgery in Crohn's disease. *Gut* 1971;12:739-49.
12. Cormack JD, Melsom MA, Winwick JG. Recurrent perforation of the ileum. *Gut* 1970;11:685-7.
13. Steinberg DM, Cook WT, Alexander-Williams J. Free perforation in Crohn's disease. *Gut* 1973;14:187-90.
14. Nasr K, Morowitz DA, Anderson JGD, Kirsner JB. Free perforation in regional enteritis. *Gut* 1969;10:206-8.



**Hindawi**  
Submit your manuscripts at  
<http://www.hindawi.com>

