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Perianal Crohn's Disease

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Perianal disease is a frequent manifestation of Crohn's disease and occasionally is the presenting symptom. However, most lesions are or become asymptomatic and remain so for many years. A series of 109 patients with perianal fissures and fistulas has been followed for 10 years. Fourteen have died, 7 from unrelated disorders. Ten have had an excision of the rectum but only 5 for perianal disease. Of the remaining 85 patients, 24 asymptomatic patients were not reexamined, and 61 were investigated extensively by sigmoidoscopy biopsy or manometry. Many fissures and most fistulas had healed, but some had progressed to some degree of anal stenosis, usually asymptomatic. A very conservative policy is proposed.

Perianal lesions are common in Crohn's disease, although it is perhaps surprising that the association has only been recognized recently. Probably the first description of perianal Crohn's disease was by Gabriel [1] who, in 1921, recognized a distinction between perianal tuberculosis and a condition that was probably Crohn's disease. Others had noticed a relationship between perianal disease and inflammatory lesions of the bowel even before the classic paper by Crohn and his colleagues in 1932 [2]. Nevertheless, it was more than a quarter of a century before the clear relationship between Crohn's disease of the small and large bowel and perianal lesions was fully recognized and well documented [3]. It has since been found that perianal lesions could sometimes be the first manifestation of Crohn's disease and for many years the only one [4]. Although many surgeons have advocated surgical intervention for perianal Crohn's disease, the latest trend is towards extreme conservatism in the management of this complication [5].

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The Clinical Features

The typical features of anal disease in association with Crohn's disease are described below.

1. Skin maceration and superficial erosions are common in any patient with chronic diarrhea and are not necessarily specific for Crohn's disease. Secondly, bacterial or fungal infection may cause an acute inflammatory condition that may be painful.

2. Skin tags are common around the anus in patients with Crohn's disease. These are usually simply edematous corrugations of the perianal skin, often associated with some macerations. The skin tags tend to compound the problem, as they make the anal region difficult to clean. Skin tags in Crohn's disease, particularly in the active phase, are often much larger, thicker, and harder than the skin tags without Crohn's disease.

3. Fissures are often found in patients with Crohn's disease if the anal canal is inspected carefully in a good light or with a proctoscope. They are often deep and frequently have undermined edges, a cyanotic hue to the surrounding skin, and they often occur at any angle, not just anteriorly or posteriorly. Sometimes adjacent deep fissures with undermined edges meet, leaving a "flying buttress" type of skin bridge between them. One of the remarkable features of fissures in perianal Crohn's disease is the relative lack of pain despite their alarming appearance.

4. A minor degree of stenosis of the anal canal is frequently encountered when the rectum is examined digitally in patients with Crohn's disease. The anal canal will frequently not admit the examining index finger and, unlike the normal anus, will rarely

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Table 1. Incidence of perianal lesions related to the site of intestinal Crohn's disease (n = number of cases; % incidence = incidence of perianal disease).

Series and year reported	Site of principal lesion						Overall incidence of perianal disease—%
	Small bowel		Large bowel		Small and large bowel		
	n	% incidence	n	% incidence	n	% incidence	
Loygue and Huguier [6] 1971	49	8	25	8	46	22	29
Fielding [7] 1972	118	76	35	94	14	92	80
Lockhart-Mummery [8] 1975	62	32	101	56	141	70	61

admit 2 fingers. Severe long and tortuous strictures of the anal canal are found in Crohn's disease but, in our experience, usually are seen in those patients previously treated surgically. Patients rarely complain of symptoms when there is only a moderate degree of stenosis because they usually pass liquid or semi-solid feces to which the stenosis is no barrier. In addition to the narrowness of the passage to the anal canal, there is often surrounding induration.

5. Fistulas-in-ano, although less common than fissures, are often encountered in patients who have Crohn's disease with no anal symptoms or without a previous history of a perianal abscess. Because they are frequently asymptomatic, lesions require a specific search for their detection. The fistulas usually resemble the ordinary fistulas seen in patients without Crohn's disease, in that there is an indurated opening in the skin that exudes a little pus, and there is a palpable track running in towards the anal canal; the track may be either direct or indirect. In Crohn's disease, there are very frequently multiple openings to the fistulas, often far out in the buttock or even the scrotum, labia, or thigh. As with all the other types of perianal Crohn's disease, the distinctive features are the lack of pain, chronicity, induration, multiplicity, and cyanotic coloration. Occasionally, there is a blockage of the outlet of fistulas and an abscess results. Whenever there is pain associated with perianal Crohn's disease, it is usually because an abscess is draining inadequately.

Although fistulas are often multiple, they can still be usefully categorized as superficial or deep. In superficial fistulas, openings, although they may be multiple, can be traced to an internal opening at the pectinate line and appear to have arisen from infection of an anal gland. Deep fistulas usually track above the pectinate line and either arise from Crohn's ulcers in the rectum or upper anal canal, or follow inappropriate surgical treatment.

Rectovaginal fistulas in Crohn's disease have a similar classification, but are relatively rarely from anal canal gland infection; more commonly they result from direct penetration of the rectal wall by an ulcer.

6. In contrast to some earlier reports, we do not find that patients with Crohn's disease have internal hemorrhoids, although they may frequently be misdiagnosed as such. The fleshy skin tags are often incorrectly classified as hemorrhoids, and some of the serious complications of perianal Crohn's disease that we have seen have been in patients who had what we would regard as an unnecessary hemorrhoidectomy.

Differential Diagnosis

Any anal lesion in a patient who is known to be suffering from Crohn's disease is likely to be perianal Crohn's disease and almost is so by definition. Of course, it is possible for patients with Crohn's disease to develop coincidental conditions, such as venereal proctitis or anal carcinoma, but in our experience of many hundreds of patients with Crohn's disease, this has never occurred. The problem that arises in differential diagnoses is when the perianal Crohn's disease is the first manifestation of the inflammatory bowel disease and so there is no prior diagnosis. In someone who has no evidence of intestinal disease, the most common differential diagnosis is between simple nonspecific types of pruritus ani, fissure, or fistulas-in-ano. Of course, it is extremely difficult to differentiate, but the relative lack of pain, multiplicity of lesions, or eccentricity of fissures should alert the observer to the possibility that the lesion is due to Crohn's disease. The most useful point in differential diagnosis in such a patient is a careful sigmoidoscopy looking for mucosal lesions within the rectum. These naturally should not occur in the absence of inflammatory bowel disease and are usually diagnostic of Crohn's disease. Any mucosal lesion should be subjected to biopsy, and this will sometimes provide the diagnosis. Histologic examination of the fistula track, or the fissure may also give the diagnosis. However, more often it does not give a classic histologic picture and,

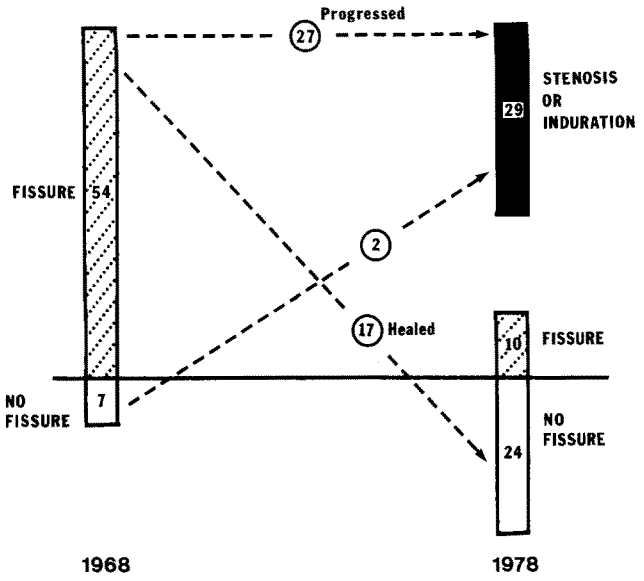

Fig. 1. Graphic representation of the number of patients with and without fissures in 1968, indicating what happened to them 10 years later. Many of the fissures healed and many others progressed to give an indurated anal canal, without the presence of an active fissure.

furthermore, the occasional presence of foreign body giant cell systems in the tracks of nonspecific fistulas may make the diagnosis difficult. Often, the only point in attempting to differentiate is that we would advocate a very much more conservative approach if Crohn's disease is suspected.

The presence of venereal disease in the anal canal should be considered in the differential diagnosis, particularly when there is an unusual type of anal lesion and no known inflammatory bowel disease. It is then necessary to take the appropriate tests, such as a Frei's skin reaction test, serology, or bacteriological tests. In seriously ill patients with unexplained florid perianal disease, the possibility of leukemia or myeloma should be considered. In patients from tropical areas, one should consider Bilharzia, and in some areas of the world tuberculosis is common. In any patient with known tuberculous disease, the presence of perianal tuberculosis should be excluded histologically and bacteriologically before making a diagnosis of perianal Crohn's disease.

Frequency

The incidence of perianal lesions in some reported series is listed in Table 1 [6–8]. The considerable variation in the incidence is related more to problems of the establishment of the criteria of diagnosis and to the fact that many were retrospective surveys, rather than to any true variation in incidence from series to series. In some series, it is only the presence of fistulas or fissures that are listed, skin tags being considered to be a minor variation of normality [7]. Furthermore, in some series, it is only symptomatic perianal disease that is considered; clearly such a definition will give a very low incidence [6]. In other series, when there is some doubt about the differentiation between Crohn's colitis and ulcerative colitis, the inclusion of a large number of patients with ulcerative colitis will cause the incidence to appear to be low. We believe that perianal disease is relatively uncommon in ulcerative colitis, although the nonspecific type of fistula or fissure-in-ano may occur in any chronic diarrheal state. We have found that many patients diagnosed originally as having ulcerative colitis who later had perianal disease were eventually reclassified as having Crohn's colitis, particularly when inflammatory disease of the small bowel declared itself later. The very high incidence of perianal disease in some series is related to case selection, particularly when only seriously ill patients were investigated [9].

Natural History of the Disease

When Fielding made a survey of perianal disease in 156 patients attending our follow-up clinic, he found what he considered to be perianal Crohn's disease in 109 [7]. Patients with skin tags were excluded; he only included those with definite fistula or fissures. We have kept these patients under review over the years and reassessed them after 5 years and recently 10 years. During this time, 10 of the original patients have come to proctectomy, although in only 5 was this done directly because of perianal Crohn's disease. Fourteen have died from coincidental causes or from complications of Crohn's disease, and a few have emigrated. Twenty-four were completely asymptomatic but refused to be reassessed proctologically after 10 years. After allowing for these exclusions, we had 61 patients whose perianal Crohn's disease was assessed thoroughly in 1968, and then reassessed equally thoroughly in 1978. Figure 1 shows a graphic representation of the fate of those patients who had fissures in 1968 as they were when reassessed 10 years later. It will be seen that a large number of those with fissures underwent spontaneous healing and that some of those who healed appeared to have a normal anal canal. However, many who had active fissures in 1968 progressed to some degree of anal stenosis with induration 10 years later, despite the lack of any present breach in the mucosa of the anal canal. A small number of those who had no fissure in 1968 also progressed to a minor degree of anal narrowing.

In Figure 2 we see the fate of patients who had a fistula diagnosed in 1968. Despite having no surgical

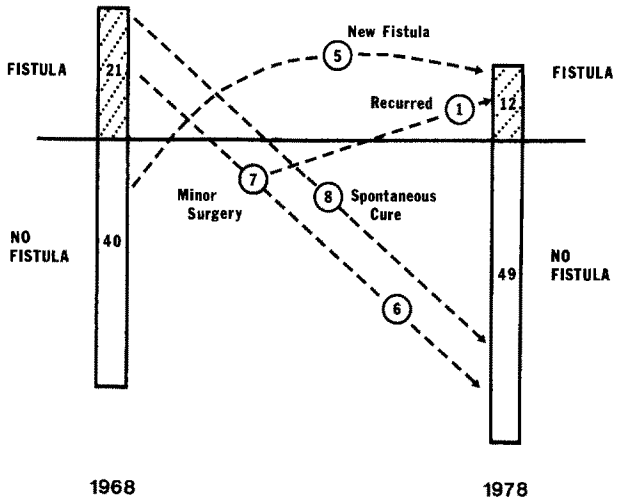

Fig. 2. Graphic representation of the number of patients with and without a fistula in 1968, showing what happened to them 10 years later. Eight patients appeared to have healed their fistula spontaneously, 7 others had minor surgical procedures, and in only 1 of these had the fistula recurred. All the patients with demonstrable fistulas were symptom free.

treatment, 8 of these patients had no detectable fistula-in-ano 10 years later. This is of interest because it appears to be very uncommon for the usual simple type of fistula-in-ano, in patients without Crohn's disease, to heal spontaneously. Seven of the patients with fistula-in-ano were treated surgically, usually by unroofing a low fistula; one of those so treated recurred. As our policy has tended to become more conservative in the last 10 years, we now question whether it was necessary to perform any sort of surgical treatment on these patients; they might well have healed spontaneously or, like most of the other patients, remained asymptomatic even though the fistula was still present.

Complications

Abscess

Perianal abscesses occur from infection in anal glands from deep Crohn's fissures ulcerating through the wall of the bowel, or from deep anal canal fissures with undermined edges. Our experience leads us to believe that whenever perianal Crohn's disease is painful, it is because there is pus or acute tissue inflammation.

Stenosis

As was recorded in the previous section, some degree of anal stenosis is very common, particularly in those patients who have had fissures in the anal canal that have healed with fibrosis. However, stenosis from this complication is usually mild and often gives a little trouble. In our series, severe stenosis, enough to cause subacute intestinal obstruction or severe symptoms, has been confined to patients who have had previous surgical treatment, usually hemorrhoidectomy, wide manual dilatation, or unroofing of the fistula track.

Incontinence

This is probably the most serious complication of perianal Crohn's disease. We have reported previously 16 patients with this complication[5]. Six were completely incontinent of feces and 10 incontinent only when they had diarrhea. Four of this less severe group had had no previous surgical treatment, but the other 6 all had some form of extensive anal surgery. Since publishing our experience in 1975, we have seen only 2 further patients with incontinence, both of them following extensive surgery elsewhere. Therefore, we still hold our previously expressed view that incontinence in Crohn's disease is due to aggressive surgeons and not to progressive disease.

Carcinoma

We have reported recently our experience with carcinoma developing within the track of a chronic rectovaginal fistula in Crohn's disease [10]. Although this was a single case and obviously a rare occurrence, it was important to consider that a very chronic irritation for 20 or more years in any epithelial surface predisposes to development of malignancy [11].

Treatment

Medical

Rest is probably the most important facet of the medical management of Crohn's disease. As with Crohn's disease elsewhere, perianal disease tends to improve if the patient rests in hospital. We have recently admitted a woman who was ill from an acute exacerbation of gross perianal inflammation from Crohn's disease in a defunctioned rectum. The pain was due to a labial abscess associated with a fistula; this was unroofed, the patient was treated by bedrest and simple hygiene, and there was a rapid improvememt.

Immunosuppression by steroids or by azathioprine appears to have a similar beneficial effect to that of rest in patients with perianal Crohn's disease [12]. No controlled trials have been reported, and it is not certain whether these dangerous drugs are bet-

ter than rest alone. We have seen one elderly woman with severe perianal Crohn's disease who did not respond to rest or steroids, but who subsequently responded satisfactorily to azathioprine for 2 years. Eventually, she developed a moderately severe stenosis that needed dilatation.

Antibiotics are helpful during acute exacerbation of perianal Crohn's disease, particularly if there is surrounding induration and tissue tension as a result of bacterial invasion of tissues. Any form of appropriate antibiotic is effective; good results have been reported from our unit with the use of metronidazole [13].

Surgical

We have reported previously on the management of 418 patients with Crohn's disease treated between 1945 and 1973 [5]. There were 68 patients who had had a total of 96 operations for perianal complications. These included 25 drainage of abscesses, 52 laying open of fistulas, 3 wide excisions of fissure, 5 manual dilatations, 7 wide excisions of deep fistulas, and 4 hemorrhoidectomies. In this earlier report, we suggested that many of these operations had been unjustifiably radical and, in view of the complications that had followed, we advocated a more conservative policy. In the past 5 years, the incidence of operations has fallen dramatically. We have performed only 20 simple drainage of abscesses, 2 laying open of superficial fistulas, and 2 anal sphincter reconstructions in patients who have been rendered incontinent by surgical treatment elsewhere.

Surgical intervention is indicated in patients with perianal Crohn's disease for: (a) simple unroofing and decompression of an abscess; if a fistula results, as it commonly does, this is not an indication for further treatment; and (b) dilatation of an anal stenosis. Initially this should be performed under general anesthesia and should be extremely gentle. It should never be with more than 1 finger. Thereafter, the patient can be supplied with a small plastic anal dilator (with a diameter not exceeding 1 cm) for self-dilation. There is no place for forceable multiple finger dilatation of the anus in Crohn's disease.

Surgical repair of iatrogenic incontinence may be worth considering, particularly if there is no active perianal or rectal disease. We have repaired successfully 2 patients whose sphincters had been divided. Surgical repair of large rectovaginal fistulas may also be indicated. We have treated 1 young girl with a large rectovaginal fistula by temporary diversion with an ileostomy, a secondary repair of the fistula, and later successful closure of the ileostomy.

In the management of perianal Crohn's disease, we advocate an extremely conservative policy.

Résumé

Les lésions péri-anales sont fréquentes dans la maladie de Crohn. Elles peuvent être le premier signe de la maladie. Mais la plupart d'entre elles sont ou deviennent asymptomatiques et peuvent le rester pendant plusieurs années. Nous avons suivi pendant 10 ans 109 malades atteints de fissures ou fistules anales. Quatorze patients sont décédés, 7 d'entre eux d'affections sans relation avec le Crohn. Dans 10 cas, une amputation du rectum a été réalisée, mais les lésions anales ne constituaient l'indication de cette exérèse que dans 5 cas. Sur les 85 cas restants, 24, sans aucun symptome, n'ont pas été réexaminés, 61 ont été étudiés par rectoscopie, biopsie ou manométrie. De nombreuses fissures et la majorité des fistules étaient cicatrisées; certaines avaient entraîné une sténose anale, en général modérée et asymptomatique. Nous estimons donc que le traitement doit être conservateur.

References

1. Gabriel, W.B.: Results of an experimental and histological investigation into seventy-five cases of rectal fistulae. Proc. R. Soc. Med. *14*:156, 1921
2. Crohn, B.B., Ginsburg, L., Oppenheimer, G.D: Regional ileitis: a pathologic and clinical entity. J.A.M.A. *99*:1323, 1932
3. Yarnis, H., Marshak, R.H., Crohn, B.B.: Ileocolitis. J.A.M.A. *164*:7, 1957
4. Morson, B.K., Lockhart-Mummery, H.E.: Anal lesions in Crohn's disease. Lancet *2*:1122, 1959
5. Alexander-Williams, J.: Perianal Crohn's disease. Amsterdam-Oxford, Excerpta Medica, 1975, p. 43
6. Loygue J., Huguier M.: Le traitment chirurgical des localisations ano-rectales de la maladie de Crohn. Arch. Fr. Mal. App. Dig. *60*[Suppl.]:29, 1971
7. Fielding, J.F.: Perianal lesions in Crohn's disease. J. R. Coll. Surg. Edinb. *1717*:32, 1972
8. Lockhart-Mummery, H.E.: Crohn's disease: anal lesions. Dis. Colon Rectum *18*:200, 1975
9. Weakley, F.L., Turnbull, R.B.: Recognition of regional ileitis in operating room. Dis. Colon Rectum *14*:17, 1971
10. Buchmann P., Allan, R.N., Thompson, H., Alexander-Williams, J.: Carcinoma developing in a rectovaginal fistula in Crohn's disease. (*In press*)
11. Greenstein, A.J., Sacher, D.B., Pucillo, A., Kreel, I., Geller, S., Janowitz, H.D., Aufses, A.: Cancer in Crohn's disease after diversionary surgery. A report of seven carcinomas occurring in excluded bowel. Am. J. Surg. *135*:86, 1977
12. Brooke, B.N., Cave, D.R., King, D.W.: Place of azathioprine for Crohn's disease. Lancet *1*:1041, 1976
13. Allan, R.N., Cooke, W.T.: Evaluation of metronidazole in the management of Crohn's disease. Gut *18*:422, 1977