

Surgical pathology and management of anorectal Crohn's disease¹

Professor L E Hughes FRCS

*University Department of Surgery,
Welsh National School of Medicine, Cardiff CF4 4XN*

It is surprising that lesions as distinctive and florid as those seen in severe cases of anorectal involvement with Crohn's disease should have evoked so little comment until the classical papers of the St. Mark's group (Morson & Lockhart-Mummery 1959, Lockhart-Mummery & Morson 1964). Subsequent interest has centred around two questions, incidence and management, but so far has not led to a detailed clinicopathological description. Fielding (1972) has shown that abnormalities in the anal region are extremely common in Crohn's disease. However, some of the lesions described are seen in patients with other gastrointestinal disease or even in normal people, and it is not certain how many of the more minor conditions, e.g. skin tags and minor fissures, may be chance associations. On superficial examination, views regarding treatment vary widely. Alexander-Williams *et al.* (1974) and Ritchie & Lennard-Jones (1976) lay stress on a conservative approach; the former authors believe that many of the serious complications are iatrogenic, and that anal lesions give little trouble if treated conservatively. Lockhart-Mummery (1975) gives a different emphasis, pointing out that treatment as radical as abdominoperineal resection is necessary for severe cases.

A detailed description and classification of these lesions is needed to clarify both questions. To try to apply general concepts of a conservative or a radical approach to a heterogeneous group of conditions may be misleading, since individual elements may require quite different management regimes.

This study has attempted to provide such a classification by analysing the anorectal lesions of Crohn's disease into clearly-defined entities for which the clinical course and behaviour can be ascertained, leading to management policies appropriate to each individual clinicopathological entity.

Patients

The study has been based on observations over a six-year period in the combined inflammatory bowel disease clinic in Cardiff, where approximately 400 patients with Crohn's disease are under the care of Dr John Rhodes. During this period 14 operations have been carried out for anorectal Crohn's disease, excluding simple drainage of abscesses and those patients having rectal excision as part of the management of extensive large bowel disease. The cases treated surgically represent only a small percentage of those with anorectal manifestations.

The operations carried out are shown in Table 1. All operation wounds healed uneventfully, although slowly in some cases. No anorectal problem has recurred in follow up over a period of up to 6 years, but 2 patients have developed reactivity of ileal disease.

The anorectal lesions found in Crohn's disease have been classified into three groups: primary anal and rectal lesions; secondary mechanical problems; and secondary infective

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Table 1. Operations for anorectal Crohn's disease (1971-77)

	No. of operations
Local surgery:	
Superficial 'fistula'	2
Haemorrhoidectomy	2
Abdominoperineal resection:	
Rectal stricture	5
Anorectal fistula	4
Rectovesical fistula	1

problems. The first group may be regarded as primary manifestations of the disease and the remaining two as secondary manifestations.

Primary lesions of anus and rectum (Table 2)

These lesions show two properties not seen with the secondary problems. They reflect activity of the disease, tending to be progressive when proximal disease is active and to heal as it regresses, and they are specific lesions, seen mainly in association with Crohn's disease. They may therefore be of help in making this diagnosis in doubtful cases. Neither association is absolute, but is sufficient to be of marked clinical usefulness.

Table 2. Primary lesions associated with Crohn's disease

Anus:	Crohn's fissures
	Ulcerated piles
	Cavitating ulcer
Rectum:	Cobblestoning
	Transmural ulcer

The Crohn's fissure (Figure 1) has a classical, indolent appearance which immediately suggests the diagnosis. Although it may occur in any quadrant, it is usually posterior or posterior and anterior. The bluish, oedematous and deeply-undermined margin, the pale floor, still containing the fibres of the sphincter muscle, and the oedematous sentinel pile or skin tag at the lower end (often with a small pinhole perforation) are all typical features. The most surprising feature is the painless nature of the fissure, which may lead to its being overlooked – unless specifically sought in the initial part of a rectal examination by carefully feeling the full circumference of the anal wall between the two components of the sphincter. In early lesions the undermined edges are more readily felt than seen. When healed, the edges remain visible, though less precipitous, but the base becomes covered with a fragile, pale epithelium. Lesser degrees of fissuring than this classical description are seen, but their significance is less certain. It is easy to produce a small mucosal split with the strong separation of the buttocks necessary to inspect the anal canal adequately.

The ulcerated pile complex is also characteristic; it consists of three or more large oedematous external skin tags with a linear ulcer on each, extending longitudinally into the anal canal. These ulcers are lined by pale, oedematous granulation tissue but without the deeply undermined edge seen in the anus. Whether the term 'pile' should be used is arguable since they are not related to internal haemorrhoids. But in the absence of a known pathogenesis, the term pile is used in its widest context, since they resemble large oedematous skin tags of external piles, without the internal component.



Figure 1. The Crohn's disease fissure

The third entity is a cavitating lesion where a deep ulcer, eroding whatever lies in its path, penetrates the wall of the anal canal between the deep and superficial sphincters, or in the rectum immediately proximal to the sphincter. These craggy ulcers are readily felt on digital examination, although they are difficult to see and impossible to photograph; they are shown diagrammatically in Figure 2. Unlike the primary fissures, these ulcers are usually painful on defaecation and intensely so on examination, reflecting the sphincter invasion. The cavity itself may feel like an ulcerated anus, so the examining finger may be uncertain as to the direction of the true lumen of the anal canal. This is made more difficult because of pain, and sometimes the situation can be clarified only under general anaesthetic.

The primary lesions in the rectum – cobblestoning and transmural ulceration – do not differ greatly in appearance or behaviour from those elsewhere in the bowel.

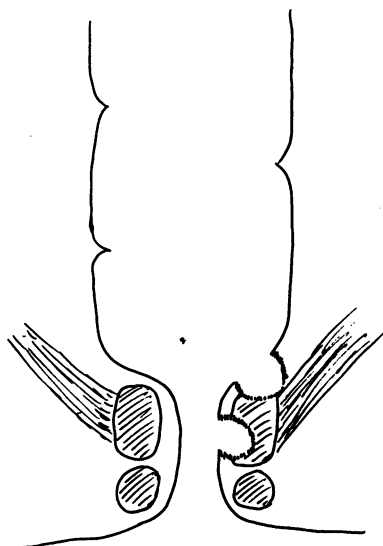


Figure 2. Cavitating anal ulcers

Clinical significance and management

Because these lesions are closely related to activity of the disease, they frequently heal when proximal disease is brought under control, either spontaneously by medical measures, or following surgical resection. The fissure will leave a shallow epithelized defect, and the ulcerated piles will leave large pendulous skin tags (Figure 3) although the ulceration heals and the oedema lessens. The cavitating ulcers may also show healing in response to decreased disease activity, but this may be less apparent, because they are frequently complicated by extensive fistulae and because the destruction of tissue in the anal canal means that healing may occur at the expense of stricture formation.



Figure 3. Resolving oedematous skin tags

Management is not a major problem in uncomplicated cases. As a general principle, local surgery should be avoided when signs of disease activity are present, since healing is likely to be poor. In fact, it may not occur at all, or surgery may precipitate local extension of the disease process. Disease activity is assessed by the usual clinical criteria in relation to the proximal bowel disease, and the anal lesions in terms of oedema, ulceration and indolence on the one hand, or decreasing oedema and signs of healing on the other. For symptomatic cases, local measures may be used, particularly steroid creams or suppositories, but we have not been greatly impressed by the results of such measures. Since these lesions improve spontaneously as the disease responds to general medical measures, it seems that the best approach is to treat the underlying disease in the usual way with systemic steroids and Salazopyrin. Once the active proximal disease has remitted, there is no contraindication to local surgery should this be required because of the development of 'mechanical' complications. Occasionally, such mechanical complications are so severe that one is forced in the presence of local disease activity to operate. If this is the case, surgery should aim to be curative in the sense of excising all active bowel disease. This is dealt with further below.

Secondary mechanical problems (Table 3)

These mechanical complications are responsible for much of the morbidity of anorectal Crohn's disease. They may be regarded as secondary conditions since each arises from one of the primary lesions. In general, these secondary complications result from healing and so arise when the active phase of the disease has passed – albeit temporarily in some cases. They usually require surgical management and there is no contraindication to surgery if the specific Crohn's disease process – in the proximal bowel or the anus – is quiescent.

Table 3. Secondary 'mechanical' problems

	Secondary lesion	Primary lesion
Anus:	Skin tags	Ulcerated pile
	Stricture	Cavitating ulcer
	Epithelized fistula	Crohn's fissure
Rectum:	Stricture	Transmural ulceration
	Epithelized fistula	Cavitating ulcer

Anal skin tags arise when the large oedematous ulcerated pile lesions resolve, the ulcers healing but leaving large skin tags which are still a source of much discomfort (Figure 3). They are readily treated by simple excision but it is wise to be conservative in the removal of the redundant skin.

Severe anal stricture in this series has always been associated with severe perianal infective complications, and this is probably due to the fact that one process, cavitating ulceration, leads to stenosis when it heals and also predisposes to extensive inflammatory disease by opening up tissue spaces beyond the sphincter. In theory they can be treated with a dilator, but they tend to fall into two groups: mild, which need no treatment, and severe, associated with fistulae and recurrent abscesses or deep penetrating ulcers. These are too painful to dilate, and frequently recurrent abscesses make the patient's life miserable. The alternatives are to let the patient soldier on, or to take the radical approach of abdominoperineal resection. In many cases the patient will make the decision and refuse radical surgery, at least in the early stages, but most of these patients come to abdominoperineal resection within a few years of developing severe stenosis. When the patient is willing to accept surgery there is no advantage in prolonging morbidity by excessive delay.

The epithelized anal fistula is a condition which has a specific pathogenesis, causes much pain and discomfort, and may be cured by a very simple surgical manoeuvre. If one looks carefully at the anal fissure of Crohn's disease (Figure 1) it can be seen that deep undermining is present at the distal margin as well as at the lateral edges of the ulcer. This produces a pocket (Figure 4) in which faeces tend to collect and the pressure of defaecation will force faecal material into the subcutaneous tissues. As the disease activity resolves, oedema will lessen but the undermining will tend to become deeper, leading to a deep subcutaneous sinus. The mechanical effect of defaecation keeps the sinus open and causes it to extend, even though the disease has become inactive with epithelization of the fistula tract. The clinical result (Figure 5) is a subcutaneous fistula with multiple openings from which faeces are extruded. It is painful, not from disease activity or inflammation, but due to the constant filling with faecal material, giving distension and irritation. It is important to recognize that this fistula is the result of mechanical forces, and is unrelated (or inversely related) to the activity of the Crohn's disease and therefore suitable for simple surgical deroofting (Figure 6).

Stricture of the rectum is not uncommon, and it is interesting that in this series it has not been associated with colonic disease. The disease has either been confined to the anorectal region, or associated with disease of the terminal ileum. In the early stages it is due to oedema

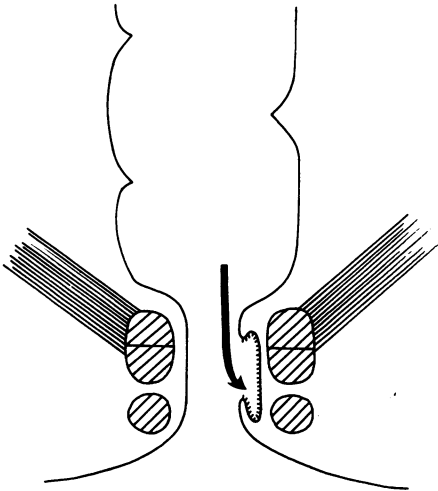


Figure 4. The genesis of the subcutaneous fistula



Figure 5. A subcutaneous epithelized fistula.



Figure 6. Deroofing a subcutaneous fistula

of the inflammatory process, together with muscle hypertrophy. Such a stenosis may give rise to remarkably few symptoms. But sooner or later this tends to be replaced by true fibrosis when the degree of disorganization of the rectal wall and perirectal tissues is extreme. The symptomatology is then largely of partial obstruction, with diarrhoea and poor control because of the lowered rectal capacity and the liquid nature of the motions proximal to the stricture. One of the problems encountered in dealing with these patients is the gradual progression of symptoms, with the patient becoming used to his discomfort. If this is combined with reluctance on the part of the surgeon to offer abdominoperineal resection, by the time the

patient comes to accept surgery, he is at a low ebb both emotionally and physically. Such patients who have had abdominoperineal resection have been uniformly pleased to accept colostomy in exchange for their diarrhoea and incontinence. The possibility of carcinoma must be considered; one of our patients had an unsuspected carcinoma within a stricture of six years' duration.

Fistula from the rectum is usually due to distal obstruction causing a cavitating ulcer to extend into the extrarectal tissues. Obstruction may be due to rectal or anal stenosis (Figure 7). The relative obstruction of an anal sphincter in constant spasm due to ulceration may act as a stricture, thus accounting for the classical site of the cavitating ulcer immediately proximal to the main sphincter mass. In other cases true anal stenosis is present. Where the obstruction is due to rectal stenosis, the fistula will be within the pelvis and may be rectovaginal or rectovesical. Operation in this situation requires removal of the entire area of diseased bowel if possible, and usually this can be achieved by abdominoperineal resection. With all active disease removed, healing is usually uneventful.

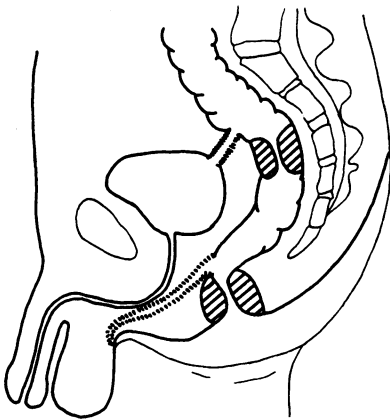


Figure 7. Fistulae associated with anal or rectal obstruction

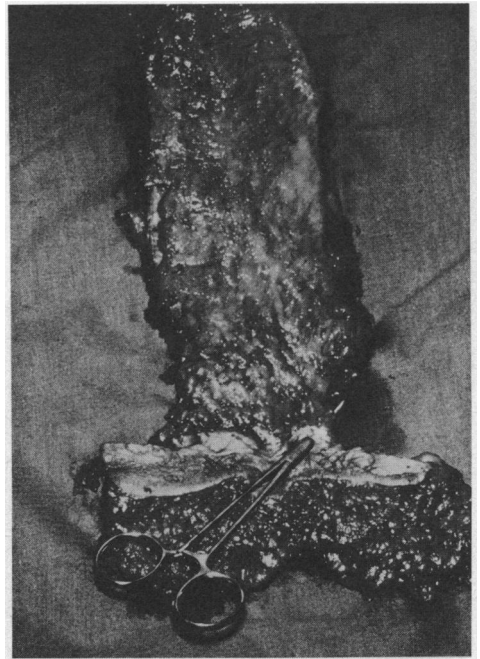


Figure 8. A persisting anorectal fistula

When the internal opening of the fistula is immediately proximal to the anal sphincter, a high anorectal fistula will result, frequently presenting externally in sites such as the scrotum. It may be treated by conservative drainage, but the natural history is for the abscesses to recur until some radical measure is undertaken. This will be so even though the active disease settles completely. This is illustrated in Figure 8, which shows the operative specimen from a patient who had a fistula intermittently discharging pus, faeces and flatus from the base of the scrotum. Recurrent abscesses led to abdominoperineal resection and the specimen showed slight rectal disease but complete healing of the anal disease. The fistula, opening immediately proximal to the dentate line, had epithelized and was kept open by faeces forced into the internal opening on each defaecation. This case illustrates why there is so much confusion with anorectal lesions of Crohn's disease. The active anal disease may resolve completely as the proximal disease resolves, yet the mechanical results may be such as to make the condition worse (in this case the

epithelization of a fistula, preventing healing); hence the clinical experience that a combination of cavitating ulcers (or strictures) and extensive perianal fistulae will usually require radical surgery. Even if the fistula is cured by local measures, during a quiescent phase, further problems can be expected when the disease becomes active again. On the other hand, should an effective medical treatment for Crohn's disease be developed, the approach to these conditions might change. Under these circumstances the lesion shown in Figure 7 should be curable by excision of the fistula by a York Mason approach under cover of a colostomy, so avoiding rectal excision.

Secondary infective problems

The perianal and perirectal abscesses and fistulae of this group obviously overlap with the other categories already discussed. They fall into three groups; the first includes those where there is no active specific lesion present in the anus. These tend to follow the normal pattern of perianal suppurative disease and may be treated in exactly the same way, i.e. as though there were no Crohn's disease present. Many of these are treated some years before a diagnosis of Crohn's disease becomes obvious, and healing is generally uneventful – at least in the first instance. The second group includes those associated with a 'minor' lesion – such as the Crohn's anal fissure; while the third group includes those cases where perianal abscesses occur in association with severe anal disease – either primary, a cavitating ulcer, or secondary complications, especially anal stricture or epithelization of the fistula opening. In the presence of disease activity, healing is unlikely to be uneventful after surgery; extension of the disease may be seen and incontinence may result from inadvisedly extensive surgery. The second group is best treated by as simple a drainage procedure as is reasonable, together with medical treatment for the general Crohn's disease state. Most patients can be managed satisfactorily over long periods in this way. The third group is much more of a problem, as shown in the earlier discussion of patients with strictures and fistulae. They may be managed initially in the same way as the second group, but the overall approach is coloured by the fact that they are likely to need abdominoperineal resection in the long term – at least until an effective medical treatment for Crohn's disease becomes available. Hence one should try to pick the optimal time for recommending a radical approach, in order to avoid both an unnecessarily early colostomy and unduly prolonged suffering on the part of the patient.

Summary

The anorectal manifestations of Crohn's disease comprise a complex and heterogeneous group of conditions which fall into two main categories: primary lesions of the active disease process and secondary lesions resulting from healing of, or infective complications of, the primary lesions. The lesions are classified into distinct clinicopathological entities which may be used in assessing prognosis and determining appropriate treatment. A basic principle of management is that surgery should be avoided as far as possible in the presence of disease activity, but will frequently be necessary, and satisfactory, for secondary complications.

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