Clinical Classification of Perianal Crohn's Disease

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Assessment of the efficacy of therapeutic approaches to anal lesions of Crohn's disease is frustrated by the lack of precise definition of its various manifestations. A classification that is clinical and based on anatomic and pathologic aspects is presented; it has been derived from a 20-year prospective study of anal Crohn's disease in Cardiff. Conceptually, the classification is analogous to the TNM system for cancer. The main classification (U.F.S.) defines the presence of Ulceration, Fistula/abscess, and Stricture, qualified by numeric values reflecting severity (0 = not present, 1 = limited clinical impact, and 2 = severe). A subsidiary classification (A.P.D.) defines Associated conditions, Proximal intestinal involvement, and Disease activity. In addition, the classification may be used in a detailed form for research or comparative purposes or in a simple form defining only the dominant lesions for routine clinical use. General use of the classification would make it possible to compare in detail incidence, management, and results of treatment in different centers. [Key words: Crohn's disease; Anus; Classification]

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Anal lesions were recognized early as a complication of Crohn's disease; Penner and Crohn[1] in 1938 described perianal fistula as the most common fistula complicating ileal Crohn's disease.

Early papers described only the general appearance of the lesions, and definition was no more precise than the presence of an anal abnormality in a patient with intestinal Crohn's disease. The inadequacy of this approach became obvious when it was found that anal lesions could predate the clinical onset of Crohn's disease by many years.[2] An early attempt at objective assessment was that by Fielding[3] (and Fielding JF. An enquiry into certain aspects of regional enteritis. Doctoral Thesis, National University of Ireland, Dublin, 1970, unpublished data), who undertook a specific anal examination in 153 patients and divided lesions into three groups: 1) abscess, sinus, or fistula, 2) fissure, and 3) skin tags. Other workers undertook similar surveys of clinically more obtrusive lesions.[4, 5]

The first classification based on surgical pathology was published from Cardiff in 1978.[6] This paper divided anal lesions into primary—considered to be part of Crohn's disease—and secondary—arising from mechanical and/or infective complications of the primary lesions. It also introduced the concept of disease activity, since clinical assessment of activity based on the presence of edema and active ulceration showed some correlation with the outcome from surgical procedures, particularly in relation to wound healing.

Further experience has broadened our conception[7] to include incidental lesions, coexisting conditions such as hemorrhoids that are unrelated to the Crohn's disease. Rarer entities, such as perianal fistulas originating from ileal rather than anal disease and the complication of malignancy in long-standing cases, are also covered.

THE CARDIFF CLASSIFICATION

The classification defines individual lesions encountered in 150 patients with clinically significant perianal Crohn's disease (PACD) assessed personally over the past 20 years, recorded in detail prospectively. The lesions are defined in a manner that allows characterization of the clinical findings in individual patients.

It is possible to characterize many patients on simple clinical examination, but more complex lesions—particularly where pain is a feature—require examination under anesthesia. While this classification is based on examination of patients with anal disease requiring active management, it applies equally well to the much larger group of patients under our care with PACD causing no serious clinical problems.

THE ANATOMIC BASIS OF PACD

The primary lesions of Crohn's disease are confined largely to the endoanal skin, the cuboidal

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transitional epithelium of the anal canal, and the contiguous 1 to 2 cm of rectal mucosa (Fig. 1). Simple fissures tend to involve the squamous lining of the anal canal, while cavitating ulceration occurs in the upper anal canal or adjacent rectal mucosa.[6] Extension of ulceration outside the anal canal to the perianal skin is rare and occurs in the acute "aggressive" form of the disease, although perianal skin is frequently involved secondarily by means of perianal fistula.

The lowest 1 cm of rectal mucosa may be affected by localized cavitating ulceration and stricture, both of which are very characteristic of PACD. The rectum itself may be involved with contiguous Crohn's disease, but this usually resembles colonic Crohn's disease—in contrast to the specific features of PACD seen in the lowest 1 cm of mucosa.

THE PATHOLOGIC BASIS OF PACD

Basic primary lesions in the anus are similar to those in the small bowel, where ulceration may be predominantly superficial or deep.[8] The first is seen in the ileum as longitudinal or circumferential ulceration of mucosa and submucosa and corresponds to the typical, painless midline fissure of anal Crohn's disease (Fig. 2, U1a). Less commonly it is seen laterally in the anal canal (Fig. 2, U1b). Associated fistulas are low or intersphincteric (Fig. 3).

The second is a penetrating ulcer that characteristically leads in the small bowel to mesenteric abscess and fistulation into adjacent viscera. The corresponding lesion in the anal canal or lower rectum is seen as a cavitating ulcer that leads to fistulous abscess. These cavitating ulcers may be high or low depending on the level of cavitating ulcer in relation to the anorectal ring (Fig. 2, U2a,b). They give rise to high, complicated fistulas (Fig. 4). Anterior ulcers in the female lead to anovulval or vaginal fistula if low and rectovaginal fistula if high (Fig. 5). The former is characteristically long and tortuous and gives little trouble apart from associated infection and abscess formation. The latter tends to be wide, direct, and associated with incontinence of feces or flatus.

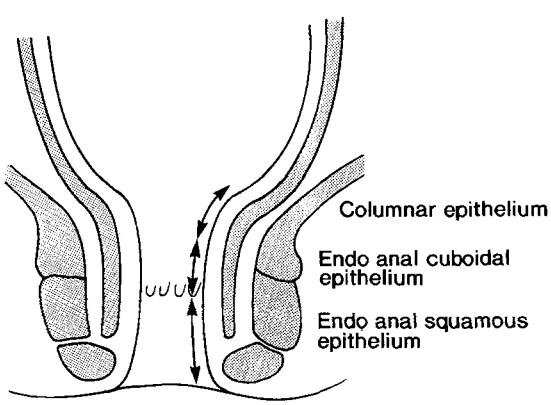

Figure 1. Anatomic sites of primary PACD lesions.

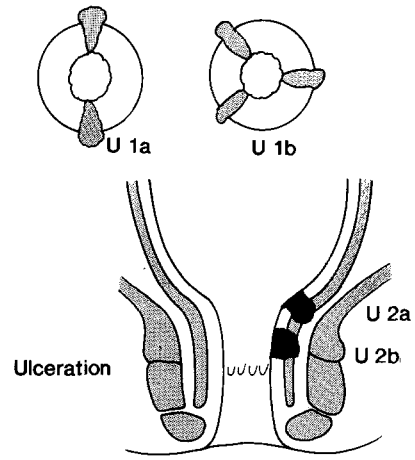

Figure 2. Ulcerating lesions: U1a = midline anal fissure; U1b = lateral anal fissure; U2a = cavitating ulcer in anal canal; U2b = cavitating ulcer in lower rectum.

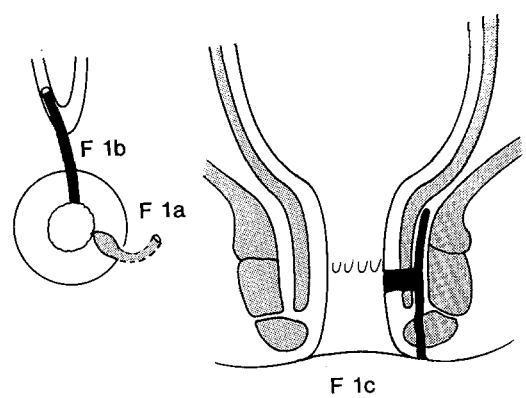

Figure 3. Low fistulas: F1a,b = perianal and anovulval fistula; F1c = intersphincteric fistula.

Aggressive ulceration is an uncommon form that extends rapidly beyond the perianal skin to involve the perineum and often the vulva. It is always associated with cavitating ulcers.

A characteristic of the inflammatory process associated with fistula formation is the extensive burrowing through normal tissue with formation of exuberant granulation tissue but little frank suppuration. A particularly characteristic example of PACD is a collar stud extension of a fistulous tract through the levator complex to form a large blind

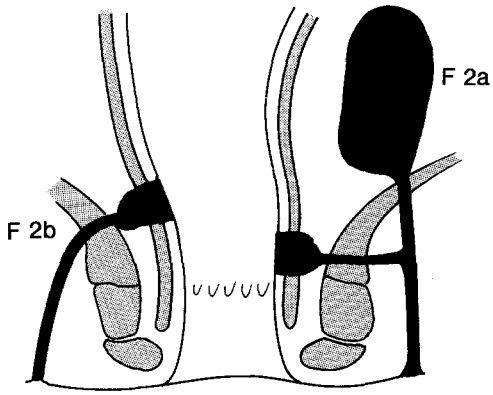

Figure 4. High fistulas: F2a = translevator extension of fistula; F2b = direct anorectal fistula.

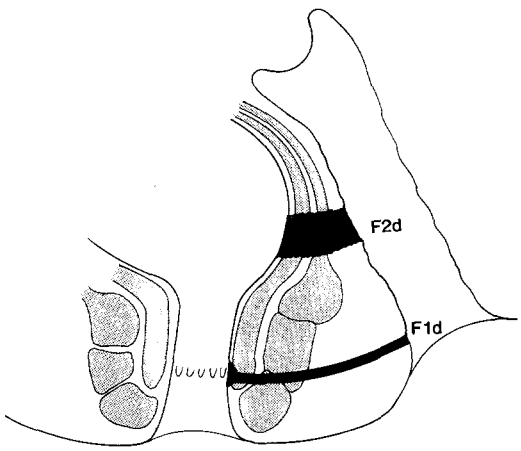

Figure 5. F1d = anovulval fistula; F2d = rectovaginal fistula.

pararectal cavity (Fig. 4, F2a). A long-term consequence of a horseshoe extension of this supralevator sepsis is a dense extrarectal stricture (Fig. 6, S2b).

The pathology of strictures differs between the anus and lower rectum. In the mid anal canal they are usually due to spasm and relax under anesthesia; less commonly an organic stricture is associated with extensive sepsis. Rectal strictures of PACD are organic and of two types, either at the level of the anorectal junction or in the first 1 to 2 cm of rectum. The first is a membranous stricture inside the lumen, presumably resulting from circumferential, superficial ulceration (Fig. 6, S1b); it sometimes extends like a shelf around only half the rectal circumference. The second is a broader extrarectal band of dense, fibrous tissue, presumably secondary to perirectal extension of a deep anal abscess (Fig. 6, S2b). Isolated involvement of the lower 2 cm of rectum should be regarded as

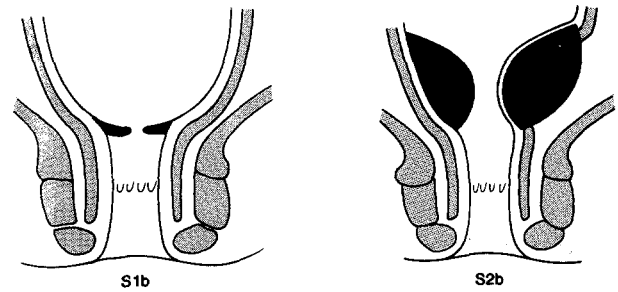

Figure 6. Rectal strictures: S1b = membranous; S2b = extramucosal.

part of PACD rather than rectal Crohn's disease, a term that should be reserved for more extensive contiguous involvement of the rectum.

CLASSIFICATION

The main classification (U.F.S.) is based on the presence of various specific types of Ulceration, Fistula/abscess, and/or Stricture. The associated numbers convey the degree of severity from a clinical point of view: 0 = not present, 1 = lesions of little clinical impact and generally good prognosis, and 2 = lesions associated with greater morbidity and poorer prognosis (Table 1). All the lesions encountered during this 20-year study are included in the comprehensive classification given in Table 2 and illustrated in Figures 2 to 6. A detailed description of them has been given previously.[6, 7] Edematous skin tags are included with ulceration because they arise from the distal margin of superficial fissures. Minor degrees are implied by the classification S1a or S1b; more obtrusive tags are recorded by the addition S1c.

Fistulas are described on an anatomic basis.

Anal spasm with severe pain but without identified cavitating ulcer or sepsis is a definitive clinical syndrome in PACD[9] classified under S1 because of the associated anal spasm and lack of demonstrable abscess.

The subsidiary classification (A.P.D.) given in Table 3 covers Associated conditions, Proximal disease, and Disease activity, which are important in patient comparisons and management and are noteworthy although outside the primary classification of PACD.

Disease activity is a concept not easily grasped, but observation allows ready recognition of the watery edema, pink or violaceous color, and florid ulceration typical of active disease. These appearances contrast with the opaque, dehydrated skin

tags and dry, thinly epithelialized fissures of inactive disease.

Overall, use of the classification has two main advantages. It compels the observer to assess the disease fully and specifically and readily allows the recording of a large amount of information, as exemplified by an illustrative case classified U1a,c, U2b, F2a, S0 (A0, P1, D1). This covers the presence of a posterior anal fissure with gross edematous skin tags, a rectal cavitating ulcer, and a high perianal fistula with a blind supralevator element but no anal or rectal stricture. There are no associated anal lesions, the patient has proximal small bowel disease, and the Crohn's lesions show disease activity.

This degree of detail is necessary for comparison or research but may be simplified for routine use by recording only the clinically dominant lesion of each broad group (Table 4). In that case, the above patient would be recorded for clinical purposes as U2F2S0 (A0P1D1). The concise version loses nothing in clinical significance, since the impact of the condition depends on the most serious lesion rather than the number or site of individual lesions.

Table 1.
Basis of Cardiff Classification of PACD

U. Ulceration	F. Fistula/abscess	S. Stricture
0. Not present		
1. Clinical impact slight/moderate		
2. Clinical impact severe		

DISCUSSION

Advances in management of difficult clinical conditions often come from detailed analysis of the anatomic and pathologic variants. With perianal Crohn's lesions, the tendency has been to lump them together as a single group while recognizing the variety of lesions that might occur. This lack of specific assessment is probably responsible for the widely ranging accounts of management and outcome. For example, metronidazole is regarded with enthusiasm by some but with disappointment by others.[10] The rationale for any surgical approach is questioned by some, and those who report the outcome of specific procedures give widely differing results. Our experience of the past 20 years has led to recognition that specific lesions tend to be associated with predictable outcomes. For example, fissures usually remain symptomless and be-

Table 2.
Cardiff Classification of Anal Crohn's Disease (U.F.S.)

U. Ulceration	F. Fistula/abscess	S. Stricture
0. Not present	0. Not present	0. Not present
1. Superficial fissures (a) posterior and/or anterior (b) lateral (c) with gross skin tags	1. Low/superficial (a) perianal (b) anovulval anoscrotal (c) intersphincteric (d) anovaginal	1. Reversible stricture (a) anal canal—spasm (b) low rectum—membranous (c) spasm with severe pain— no sepsis identified
2. Cavitating ulcers (a) anal canal (b) lower rectum (c) with extension to perineal skin (aggressive ulceration)	2. High (a) blind supralevator (b) high direct (anorectal) (c) high complex (d) rectovaginal (e) ileoperineal	2. Irreversible stricture (a) anal stenosis (b) extrarectal stricture

Table 3.
Subsidiary Classification (A.P.D.)

A. Associated Anal Conditions	P. Proximal Intestinal Disease	D. Disease Activity (in Anal Lesions)
0. None	0. No proximal disease	1. Active
1. Hemorrhoids	1. Contiguous rectal disease	2. Inactive
2. Malignancy	2. Colon (rectum spared)	3. Inconclusive
3. Other (specify)	3. Small intestine	
	4. Investigation incomplete	

Table 4.
Simplified Clinical Classification of PACD

U. Ulceration	F. Fistula/Abscess	S. Stricture
0. Not present	0. Not present	0. Not present
1. Superficial fissure	1. Low/superficial	1. Spasm/membranous
2. Cavitating ulcer	2. High/complex	2. Severe fibrotic

nign. Cavitating ulcers are likely to lead to major symptoms and incapacitating fistulous tracts. Anovaginal fistulas are usually indirect and tortuous, causing problems by abscess formation, while rectovaginal fistulas are wide and direct and produce fecal incontinence. The latter may be readily closed by a simple musculomucosal flap advancement following regression of local signs of disease activity in patients with proximal disease confined to the small bowel, but rectal excision may be necessary where disease activity cannot be controlled because of continguous active rectal disease. The pathology and concepts underlying these two different consequences of rectovaginal fistulas can be appreciated and expressed through the Cardiff classification.

This detailed classification is presented as a simple means of recording clinical data, as a basis for individualizing management and comparison of therapeutic results, and in the planning of clinical trials.

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