Reconstruction Surgery and Traumatology

I. Knee Joint, Pathology, Reconstruction
II. Critical Remarks on Indications in Hip Surgery
III. Miscellaneous

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Editorial

We take great pride in announcing that the Swedish member of our Editorial-board, Docent Dr. SVEN-ERIK BERGENTZ, has been appointed full Professor of Surgery by the King of Sweden at the beginning of this year.

Dr. BERGENTZ, born 1927, has been trained with Docent STIG LINDGREN in Falun and Professor RAGNAR ROMANUS at the Department of Surgery II, University of Gothenburg, Sahlgrenska Sjukhuset. During the last 8 years he has been associated with Prof. LARS ERIC GELIN at the first Surgical Department in Gothenburg. One year was spent with Professor FRANCIS D. MOORE at the Peter Bent Brigham Hospital, Harvard Medical School, Boston, Massachusetts. Dr. BERGENTZ is internationally known for this important contributions in the field of rheology and blood coagulation as related to surgery. His thesis on fat embolism (1961) has become a standard reference. During the past 7 years he has been very active in clinical kidney transplantation, now overlooking one of the largest and finest series in the world. He will continue his important work as transplantation and vascular surgeon as head of one of the departments of Surgery and Director of Experimental Surgery at the Malmö General Hospital, which is one of the teaching Hospitals of the University of Lund.

We heartily congratulate our colleague in Sweden for his great success and wish him all the very best for his future activities.

July 1971
M. ALLGÖWER, R. CALNE and U. F. GRUBER

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Crohn's Disease, with Special Reference to Surgical Management

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Contents

I. Introduction

Perhaps it is necessary to defend the choice of the term 'Crohn's disease', which is the designation persistently used in Britain for this

Table I. Numbers of patients with ileo-caecal tuberculosis and Crohn's disease respectively, treated at the Leeds General Infirmary from 1925–1969 inclusive

	1925–1929	1930–1934	1935–1939	1940–1944	1945–1949	1950–1954	1955–1959	1960–1964	1965–1969
Ileo-caecal tuberculosis	20	18	19	12	3	2	2	1	4
Crohn's disease	–	–	3	13	25	38	72	98	110

condition instead of 'regional or granulomatous enteritis', as preferred in the United States of America, despite the fact that it was in New York that BURRELL CROHN practiced as a physician. This might be regarded as a clear instance of the truth of the old Biblical quotation that a prophet is not without honour save in his own country!

It is true that 19 years before CROHN's classical paper with GINZBURG and OPPENHEIMER, DALZIEL [1913] of Glasgow described 8 cases of this condition, and even before him many other surgeons, such as MOYNIHAN [1907] and MAYO-ROBSON [1908] of Leeds, had also encountered similar lesions in the small or large bowel. Undoubtedly, however, it was the clear and complete account by CROHN *et al.*, in 1932 and subsequently that drew world-wide attention to this entity – a condition that had unquestionably existed previously, though it was not known as Dalziel's disease, but usually as *hyperplastic ileo-caecal tuberculosis*. This is well shown by the records of the General Infirmary at Leeds giving the number of cases of ileo-caecal tuberculosis and of Crohn's disease respectively treated in that hospital during the years 1925–1969 (table I). It will be noted that intestinal tuberculosis has become progressively less common, almost up to disappearing point in the early 1960's, with a slight revival of incidence in the later 1960's, and the incidence of Crohn's disease has steadily increased from 1935 onwards. It is interesting to recall, too, that the diagnosis of tuberculosis was sometimes made in the past in cases in which the pathologist considered the histological appearances to be not quite typical of a tuberculous lesion. Whilst some of these cases may rightly have been diagnosed as tuberculosis, it seems highly probably that many of them so labelled were in fact suffering from Crohn's disease.

Indeed some pathologists, such as WILLIAM BOYD [1955] and SHIELDS WARREN and SOMMERS [1948] have expressed extreme scepticism as to whether there ever was a condition that could rightly be re-

garded as hyperplastic tuberculosis of the intestine. However, the total rejection of tubercle in this connection is certainly unjustified, for ileo-caecal tuberculosis is well recognized to be an extremely common condition in India and Pakistan, the report of ANAND [1956] of Amristar in India being particularly impressive. Actually in the last few years cases of tuberculosis of the ileo-caecal region and other parts of the large intestine have been encountered in Pakistani and Indian immigrants in Britain, the diagnosis often being confirmed by guinea-pig innoculation of material taken from resected specimens. The condition also occurs in the white population of this country, as shown by recent reports by ANSCOMBE et al. [1967] and BYROM and MANN [1970]. The former authors emphasize that the lesions in their patients were invariably confined to the caecum, and that the term ileo-caecal tubeculosis would have been a misnomer for them.

II. Incidence and Site of Lesions

In their original account CROHN et al. [1932] described the disease as occurring primarily in the lower ileum, but it was soon recognized that extension into the caecum and right colon was not uncommon and that the condition could occur in many other parts of the alimentary tract as well – the duodenum, stomach, oesophagus, pharynx or mouth – in fact, as it has been somewhat facetiously expressed, 'anywhere from the back teeth to the anus'. Crohn's type lesions have also been described in situations remote from the gastro-intestinal tract, for example, the skin of the sub-mammary region [MOUNTAIN, 1970]. This wide distribution of Crohn's disease in areas of the alimentary tract other than the intestine itself and in non-alimentary sites renders the use of the eponymous term 'Crohn's disease', rather than 'regional enteritis', particularly convenient.

But perhaps the most striking thing about the distribution of Crohn's disease in recent years has been its increasing incidence in the large intestine. This is well shown by an examination of the site of the lesions in 332 cases of Crohn's disease seen at the General Infirmary at Leeds from 1939–1968 in table II. Overall the small bowel was affected roughly twice as frequently as the large. However, if the relative incidence of disease in the large and small bowel is examined at various stages throughout the 30-year period under review (fig. 1), it will be not-

Table II. Site of lesions in intestinal tract in 332 cases of established Crohn's disease treated at the Leeds General Infirmary from 1939–1968 inclusive [from GOLIGHER *et al.*, 1971] in %

49 {	Small bowel only	34 }	62
	Small bowel mainly	28	
	Large bowel mainly	21 }	38
	Large bowel only	17	

Rectum involved in *31* %

ed firstly, that there has been a great increase in the overall occurrence of Crohn's disease in the Leeds experience in the last 10–15 years, and secondly, that the frequency of this condition in the large bowel, alone or in association with the small bowel, has undergone a quite disproportionate increase during the same period, so that at the present time Crohn's disease is being seen about as often in the large bowel as in the small.

A question that immediately arises is whether the increase in the number of cases of Crohn's disease referred to the Infirmary represents a genuine increase in incidence, or merely reflects either a higher diagnosis rate or an increasing capacity to attract these patients. The weakness of these figures is that they relate to the activities of one institution – and indeed largely to the work of an individual surgeon – rather than to the endemic frequency of the disease in a whole geographical region of the country, and are inflated by referral of many patients from other areas in Britain and even countries overseas. However, statistics on a regional basis have recently been produced by KRAUSE [1971] and BRAHME and WENCKERT [1971] in Sweden and by MYREN [1971] in Norway, showing that the proportion of hospital admissions due to Crohn's disease has undergone a quite striking rise in the last decade or so.

The possibility of more frequent diagnosis in recent times pertains mainly to Crohn's disease of the large bowel. The pathological and clinical features of the disease as it occurs in the colon and rectum, and its differentiation from ordinary ulcerative colitis, were first clearly defined by LOCKHART-MUMMERY and MORSON in 1960 and described more fully by them in 1964. It has been suggested that prior to their work many instances of this condition in the large intestine were possibly misinterpreted as examples of ulcerative colitis, whilst in the last few years they have more usually been correctly recognised as cases of Crohn's disease.
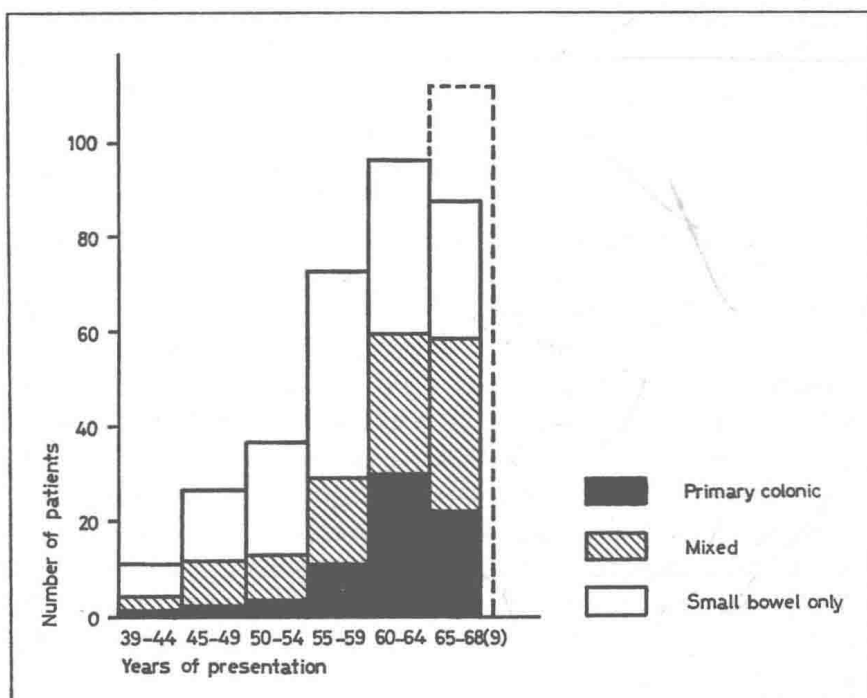
Fig. 1. The increasing incidence of Crohn's disease, especially in the large bowel, during the last 10–15 years [from GOLIGHER *et al.*, 1971].

If this had been the entire explanation for the recent apparent increase in frequency of large bowel Crohn's, one would have expected as a corollary that the frequency of true ulcerative colitis in the same period should have declined. MYREN [1971] has examined this point in his recent epidemiological survey in Norway. He finds that there has been no decrease in the incidence of ulcerative colitis in that country *pari-passu* with the increased incidence of Crohn's disease, so that clearly the greater numbers of cases of Crohn's colitis cannot be explained to any significant extent on that ground, and must represent a real increase in the incidence of this disease in the large bowel.

III. Pathology

This perhaps leads naturally to a consideration of the special morbid anatomical characteristics of Crohn's disease of the large intestine.

Sometimes the lesions of this condition encountered in the colon and rectum closely resemble those in the small intestine, with the same classical gross thickening of the bowel wall and 'cobblestoning' of the mucosa. In addition there may be 'skip' lesions in different parts of the large intestine with or without associated involvement of the small gut. But in other instances the condition does not at first look very different from ordinary ulcerative colitis, for the wall of the colon is thin and there is much mucosal ulceration; the changes, however, are often patchy and asymmetrical. The microscopical distinction between Crohn's colitis and ordinary ulcerative colitis may also be very difficult and depends on the recognition in the former of the sarcoid reaction with non-caseating giant cells, fissures and transmural extension of the disease in the bowel wall. The present authors, not being histopathologists, are by no means competent to discuss in detail this often perplexing diagnostic problem of intestinal pathology. But it is noteworthy that on occasions different pathologists of considerable experience in this field reach different decisions on the same specimen as between Crohn's versus ulcerative colitis, and, further, that most intestinal pathologists admit that they find themselves unable at the present time to classify satisfactorily about 10% of the specimens of colitis presented to them [MORSON, 1971].

The Risks of Malignant Change in Crohn's Disease

An important question related to the pathology of Crohn's disease of the large bowel is whether the now well-documented predisposition of ulcerative colitis to form intestinal cancers applies also to Crohn's disease, particularly in the colon. No firm answer can as yet be given, for, though about 7 or 8 well-authenticated cases of carcinoma arising in areas of Crohn's enteritis have now been reported [SHIEL et al., 1968; PERRETT et al., 1968; JONES, 1969, and TRUELOVE, 1970], compared with the large number of cases of Crohn's disease on record, this does not at present constitute a significant incidence. It must be remembered, however, that Crohn's disease of the large bowel, which is presumably the part of the intestinal tract that would be most likely to show any tendency towards carcinoma formation, has only been seen in substantial numbers of patients in recent years. Perhaps when Crohn's colitis has been widely recognised for a longer time, especially if physicians can produce some more effective medical treatment that will induce patients

to keep their diseased colons instead of having them removed at a fairly early stage of the disease, the crop of carcinomas may increase dramatically and force clinicians to accept malignant change as a genuine danger in Crohn's disease and to take account of it in framing therapy for this condition. But to date this has certainly not been so.

IV. Clinical Features and Diagnosis

The clinical manifestations of Crohn's disease may be extremely variable, as is shown by a recent analysis of the initial symptoms in 332 consecutive cases of this condition treated at the Leeds General Infirmary (table III). It will be seen, however, that the commonest complaints were abdominal pain and diarrhoea. Sometimes the onset of symptoms is very insidious and, moreover, when the patient eventually comes to hospital, the radiological and other special investigations may be negative or equivocal in their results – for not every case of Crohn's disease of the small bowel shows a typical Cantor string sign on barium study. Consequently quite a few patients with Crohn's disease remain incorrectly diagnosed – often labelled as psychoneurotic – for 6–12 months or longer from the commencement of their complaint, as DYER and DAWSON [1970] have recently emphasized.

Alternatively, the onset of symptoms in Crohn's disease may occasionally be dramatically sudden, in the form of severe abdominal pain, localized to the right lower quadrant of the abdomen and mimicking that due to acute appendicitis, so that emergency operation is undertaken, at which the appendix is found to be normal but the lower ileum appears

Table III. Symptoms at onset of disease (per cent of 332 patients)

Abdominal pain	51.8
Diarrhoea	42.7
Weight loss	13.2
Vomiting	12.7
Rectal bleeding	6.9
Abdominal mass	3.6
Appendicitis	4.5
Obstruction	2.4
Perianal sepsis	7.2

thickened and congested. These are the cases of so-called 'acute' Crohn's disease [SJÖSTRÖM, 1971; GOLIGHER *et al.*, 1971]. Another form of acute Crohn's disease is that affecting the colon, and possibly the rectum with the suddenness and severity of a fulminating attack of ordinary ulcerative colitis, from which it may be clinically indistinguishable [JAVETT and BROOKE, 1970; GOLIGHER *et al.*, 1970]. Sometimes acute colonic dilatation or even perforation may occur, and urgent ileostomy and colectomy has been performed on several of these patients.

Another important manifestation of Crohn's disease is the occurrence of an anal abscess or fistula, which was the presenting symptom in 7.5% of the cases at the Leeds General Infirmary, and occurred at a later stage in the course of another 7 or 8%. Clearly the only safe rule for a surgeon dealing with an apparently idiopathic anorectal abscess or fistula is to regard the condition as possibly due to an underlying Crohn's lesion of the bowel till proved to the contrary by histological examination of the granulation tissue of the fistulous track and by sigmoidoscopic and radiological examination of the intestine.

Of particular importance and interest at the present time is the clinical presentation of large bowel Crohn's disease, because this condition has obtruded itself so much in recent years and requires to be differentiated from ordinary ulcerative colitis, which it often resembles, though in most instances it is possible to distinguish the two conditions on clinical grounds. In a case of colitis the points in favour of a Crohn's colitis rather than an ordinary ulcerative colitis are as follows:

(1) The diarrhoea is usually less severe, say 4 or 5 motions instead of perhaps 12 or 15 in the 24 hours, and is thus generally diurnal instead of diurnal and nocturnal.

(2) Bleeding *per rectum* is often absent in Crohn's disease but usually present in ulcerative colitis.

(3) Abdominal pain is frequently a symptom of Crohn's colitis.

(4) In 30 or 40% of the cases of large bowel Crohn's disease the rectum is normal on sigmoidoscopy — which it virtually never is in ordinary ulcerative colitis — or, if involved, the proctitis tends not to be diffuse, as in ulcerative colitis, but patchy with areas of apparently normal mucosa between the discrete ulcers.

(5) Biopsy of any rectal lesions revealed may disclose typical histological appearances of Crohn's disease; but it must be emphasized that the reliability of biopsy depends on the quality of the pathologist examining the specimen, and, unless he is interested in this particular work,

there are likely to be quite a few negative reports on cases of undoubted Crohn's disease.

(6) Anal fissures, abscesses or fistulas are much commoner with Crohn's disease of the large bowel than of the small, being found in about 30% of the former. However, anal complications of this kind are also a fairly frequent complication of ulcerative colitis, which lessens their diagnostic value in differentiating between Crohn's colitis and ordinary colitis. A biopsy of the anal lesion may be helpful.

(7) Much more striking and valuable in diagnosis are the other anal or perineal abnormalities, sometimes observed in Crohn's disease of the large bowel, but never in ordinary ulcerative colitis – namely the oedema and bluish discolouration of the perianal skin and the sometimes very extensive ulceration in this area [CRAY et al., 1964]. Again biopsy of the perianal lesions, if examined by an experienced intestinal pathologist, should confirm the diagnosis.

(8) Barium enema examination may show an absolutely typical appearance of Crohn's disease with 'skipping' lesions, or a spared distal colon and rectum, but on other occasions the appearances are less striking, with fine or gross spiculation, asymmetrical involvement of bowel, or stricture formation, or they may be very difficult to distinguish from ordinary ulcerative colitis. Another radiological point much in favour of Crohn's colitis is extensive involvement of the ileum or disease elsewhere in the small intestine on barium meal and follow-through examination.

(9) Radiological studies of the small bowel are in general less reliable than X-ray examination of the stomach and duodenum or colon, and may sometimes miss lesions of Crohn's disease. In such cases tests for malabsorption, particularly for fat and vitamin B_{12}, may be useful in diagnosis. If gross malabsorption is revealed, even if the small bowel X-ray study was negative in a case of colitis of indeterminate origin, the condition may be assumed to be due to Crohn's disease with considerable implication of the small gut, for ordinary ulcerative colitis, even with some associated 'back-wash' ileitis, never produces significant malabsorption.

As a rough estimate, probably in about 90% of the patients referred with colitis, it is possible to make a firm clinical decision as between Crohn's disease and ulcerative colitis. Incidentally the indeterminate 10% do not correspond precisely with the 10% in whom the pathologist has difficulty in diagnosing accurately as colitis or Crohn's disease on