

A Revised Concept of Acute Regional Enteritis

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AS A RULE, symptoms of regional enteritis begin insidiously but occasionally the disease may start abruptly with symptoms that suggest acute intraperitoneal inflammation. Patients in the latter circumstance were singled out in 1932 by Crohn who termed their form of the disease acute regional enteritis.⁵ Confusion arose in the following years when surgeons operating on suspected appendicitis occasionally found a normal appendix but a terminal ileum involved in an acute inflammatory process. Because of ileal involvement this was accepted as regional enteritis and considered to be the acute form. It was assumed that the findings represented the first manifestations of chronic regional enteritis. It is the purpose of this communication to inquire into the predictability of such a progression. If, in fact, progression is unusual then the question is whether these acute changes found at laparotomy are actually related to regional enteritis.

A report of 34 patients with the chart diagnosis of acute regional enteritis treated at the Presbyterian Hospital from 1932 to 1963 is presented and the literature is reviewed.

Review of Literature

The diagnosis of acute regional enteritis is made infrequently enough so that no author has been able to report and follow more than a limited number of patients.

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Our group of 34 cases is the third largest series of which we are aware. In published reports, the percentage of patients that go on to spontaneous resolution following an attack of acute regional enteritis varies within wide limits and recovery figures from 25%^{4, 6} to 100%^{2, 19} have been recorded. One problem that becomes apparent on reviewing reports of acute regional enteritis, is the lack of rigid diagnostic criteria for this disease. As a rule, the diagnosis of acute regional enteritis must be made in the operating room from the gross appearance of the bowel wall and mesentery without benefit of biopsy. Skip areas, characteristic of the chronic form, are seldom seen in acute cases. It has been stated that the gross appearance of the terminal ileum is not specific and can be the result of tuberculosis, mesenteric lymphadenitis or allergy as well as regional enteritis.²⁵ Rhoads reported a number of patients with marked edema and engorgement of the intestine in association with primary mesenteric lymphadenitis.²¹ The diagnosis of acute regional enteritis rests on even more uncertain grounds in those series that include patients who were diagnosed without laparotomy.

Contrast x-ray studies have been employed to substantiate the diagnosis, but there is no agreement as to their value. In some series of acute cases, no x-ray changes were demonstrable.^{12, 26, 28} Crohn, on the other hand, found evidence of disease in all his 15 patients subjected to roentgen study. It is interesting that when x-ray ab-

TABLE 1. *Previously Reported Cases of Acute Regional Enteritis*

Author	Total No. of Patients	Acute Cases*	Fate of Patients Following Operation		
			Progression to Chronic Disease	No Further Difficulty	Inadequate Follow Up
Koster <i>et al.</i> ¹⁴	17	6	0	2	4
Meyer and Rosj ¹⁷	8	4	1	2	1
Eliason and Johnson ⁹	14	14	1	5	8
Eckel and Ogilvie ⁸	21	11	2	8	1
Sneierson and Ryan ²⁷	22	8	0	8	0
Holloway ¹²	13	5	0	4	1
Smithy ²⁶	5	3	0	2	1
Pugh ²⁰	17	3	0	1	2
Rose ²²	3	3	0	2	1
Homb ¹³	33	33	0	28	5
Crohn, (1949) ⁴	16	16	10	4	2
Armitage and Wilson ¹	34	10	0	10	0
O'Callaghan ¹⁹	8	8	0	3	5
Storrs and Hoekelman ²⁸	8	8	0	7	1
Siegel and White ²⁵	62	42	9	28	5
Bolton-Carter ³	40	12	7	5	0
Austin ²	60	33	0	24	9
Crohn, (1959) ⁶	15	15	5	10	0
Totals	396	234	35	153	46

* Operated on with preoperative diagnosis of acute appendicitis.

normalities are present during an acute episode of ileitis the prognosis is worse than when such changes are not demonstrated.

In some series the acute episode has been mild enough to permit preoperative administration of barium for x-ray studies. This indicates that concepts differ regarding the clinical picture of acute regional enteritis. Many authors^{3, 15, 22} believe that the typical patient should have symptoms and abdominal signs indistinguishable from acute appendicitis so that patients whose conditions permit a substantial period of time for observation and x-ray of the small intestine must present a somewhat different clinical situation. It is not always easy to decide what constitutes an acute case because a history of mild diarrhea, minimal abdominal complaints or a previous rectal operation that preceded the explosive episode can often be elicited on closer questioning. Furthermore, surgeons have sometimes noted changes in the bowel that are

typical of the chronic phase of the disease despite the fact that the onset of symptoms was acute.²⁴ This means that a certain number of previously undetected cases of chronic regional enteritis will first manifest themselves by an acute episode. Thus, the overall prognosis in any acute series will be greatly influenced by the diagnostic criteria employed.

Table 1 summarizes the experience of other authors. Since some studies included both acute and chronic cases the first column indicates the total number of patients in the series. In the second column, an effort was made to limit the acute cases to those that had a clinical picture similar to that of appendicitis and were subsequently explored because of this possibility. Any patient whose follow-up period was less than a year was placed in the last column.

Totals show that progression of the disease is not common. However, simple ad-

dition of patients in the separate series recorded is probably misleading since different concepts of acute regional enteritis are involved.

We were more interested in the fact that 11 of the 17 authors saw no progression to the chronic disease in any of their patients.

Present Series

By reviewing the charts of all patients treated at the Presbyterian Hospital between 1932 and 1963 with a diagnosis of regional enteritis we found 34 patients whose presenting symptoms, signs and history were such that it was considered mandatory to rule out acute appendicitis by immediate operation. The average age in this group was 29; the youngest patient was 8 and the oldest 58.

In 10 of 34 patients, the original laparotomy was performed at another hospital. These 10 when seen here all had the chronic form of the disease. In fact, they were seen at the Presbyterian Hospital because progression of their disease necessitated further treatment. The 24 patients whose acute episodes were treated at this institution present a very different picture in that chronic regional enteritis was noted in only one. Of those 24 patients, 21 were followed for more than 2 years and the average duration of follow up for the group was 9 years. Duration of follow up would therefore seem to have been adequate to detect progression, if such were taking place. Postoperative small intestinal x-ray studies were performed on eight of these patients within 3 months of operation and were considered normal in six. Mucosal abnormalities which regressed on serial examination were described in the other two. It is recognized that some readers will wonder if our series doesn't represent a group with especially mild involvement. However, these cases represent our total experience and no cases were omitted.

In analyzing this series the 10 cases diagnosed as acute regional enteritis at other

hospitals must be considered separately. A careful review of records, if this had been possible, might have shown that the patients had symptoms, prior to the acute episode. If these cases actually represent chronic regional enteritis discovered during an acute exacerbation, the progressive nature of the disease in all 10 patients is not surprising. It is further recognized that the selection involved in these 10 cases renders them unreliable for statistical consideration. They may be a small number from a large population most of whom did not exhibit progression to chronic regional enteritis.

The 24 patients whose acute episodes were treated at Presbyterian Hospital stand in sharp contrast. The failure of progression to the chronic form of the disease coincides with a number of reports by other writers.^{12-14, 28} This observation has led to the suggestion that so-called acute regional enteritis is unrelated to chronic regional enteritis and in fact, represents a separate entity.^{1, 28} The acceptability of this suggestion depends on two points. First, it is necessary to eliminate those acute cases having an acute exacerbation of already chronic, but perhaps previously undiagnosed, disease. This differentiation is sometimes difficult but in any case, the problem is a diagnostic one rather than one of etiology, pathology or disease progression. The other and more critical point concerns true underlying pathologic changes and so long as the etiology of both acute and chronic regional enteritis remains unknown, it will not be possible to state finally whether they are the same disease or are different entities. The evidence of our clinical series leads us to suspect that they are unrelated. The possibility that acute ileitis represents a variant or phase of acute mesenteric lymphadenitis has been proposed by Erskine,¹⁰ but of course there is no way to achieve solid proof.

The question of appendectomy in acute regional enteritis has been discussed fre-

quently in the literature and the danger of fistula formation mentioned. If acute regional enteritis is indeed a separate entity, appendectomy may be performed with impunity. This is borne out in the 24 Presbyterian Hospital patients all but one of whom had their appendix removed at the time of laparotomy. One fecal fistula did develop, but this was in the single patient with chronic regional enteritis mentioned previously. Although this 17-year-old girl presented with acute symptoms, laparotomy revealed that she already had extensive disease including an abscess in the small bowel mesentery. It is worth noting that this fistula was from the diseased terminal ileum rather than the appendiceal stump. Evidently development of a fistula is related to the severity of the disease in the ileum rather than to whether or not an appendectomy has been performed. The origin of these postappendectomy fistulae from the terminal ileum has recently been documented by Marx.¹⁶

The other 22 appendectomies were uncomplicated. The point should be considered that any subsequent pain in the patient whose appendix is not removed may lead to another laparotomy which can be avoided by removing the appendix at the first operation. It is also conceded that the patient who knows his appendix is out is less worried by subsequent mild pain and may not even consult his physician. We believe that our follow-up record in this series is sufficiently good so that failure to report cannot explain the recorded absence of subsequent gastrointestinal symptoms.

In attempting to predict the future course in the postoperative patient, an attempt should be made by further questioning to bring out any minimal symptoms indicating the existence of the disease prior to the sudden episode that led to laparotomy. Roentgen evidence of a diseased terminal ileum suggests that the patient has chronic disease whereas a normal appearance on small intestinal x-rays is indicative

of the acute process. In certain cases the gross appearance of the bowel and mesentery at operation will suggest pre-existing chronic disease. Wedge biopsy of the involved ileum has been performed without complications in acute ileitis.⁷ Wider application of this technic should provide valuable information and further clarify the relationship between acute and chronic regional enteritis.

Even without a biopsy the history, operative findings and postoperative x-rays will usually distinguish those patients whose acute regional enteritis is really the chronic disease discovered during an acute exacerbation from those who recover completely. Acute regional enteritis, as exemplified by the second group, has little relationship to the chronic disease.

Any discussion of acute regional enteritis would be incomplete without a note on patients with free perforation of the involved ileum. While none of our patients presented in this fashion it has been described with increasing frequency since 1960.¹⁸ It must not be forgotten that in many of these patients perforation merely represents an unusual complication in what is clearly chronic regional enteritis.²³ However, there are times when perforation of the ileum is the first sign of the disease.¹¹ Since resection is often forced on the surgeon in this situation, followup studies cannot be used to document spontaneous resolution. In other words, if a patient does well, it is not clear if this represents healing of the lesion or a surgical cure. The relationship between perforation in this acute situation and the subsequent finding of chronic regional enteritis has not yet been determined.

Summary

Thirty-four patients with acute regional enteritis have been reported. Ten were referred from outside hospitals because their disease required further treatment. In the 24 patients whose acute episodes were

treated at Presbyterian Hospital, emergency exploration was carried out for a clinical picture indistinguishable from acute appendicitis. Twenty-three of the 24 have remained free of any sign of progression to chronic regional enteritis.

The literature has been reviewed on this point and 11 of 17 reported series include no cases of progression to chronic regional enteritis. It is suggested that when acute ileitis is encountered in previously well patients being explored for appendicitis a careful search be made for any sign of chronic changes. If these are absent: 1) the appendix should be removed; 2) progression to chronic regional enteritis is most unlikely; 3) a barium study should be done before discharge to complete the search for pre-existing chronic disease.

A certain number of patients with chronic regional enteritis first came to medical attention during an acute episode in the course of their disease. Classification of these patients is difficult but certain features of their illness usually indicate that they belong in the chronic rather than the acute category. The finding of acute inflammatory changes in the terminal ileum of previously well patients being explored for possible appendicitis does not in our experience herald the onset of the chronic disease and indeed may well represent an inflammatory disease unrelated to regional enteritis.

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