Family Practice Grand Rounds

Ulcerative Colitis

Charles R. Clark, MD
James Graham, MD
Springfield, Illinois

DR. CHARLES CLARK *(family practice resident):* The patient is a 46-year-old white man who presented to the Family Practice Center for a complete physical examination. He and his wife have been married for approximately 25 years and have four children. The patient's education includes two years of high school, and he is employed as meat department manager for a supermarket. He was a heavy smoker, having smoked one to two packs per day for approximately 25 years, but he gave cigarettes up eight years ago. He was a heavy drinker: six to eight beers a day. He was on no medication and had no known allergies.

For approximately the last five years the patient has had frequent stools. This was an intermittent problem, but over the past four to six months it has been more marked and more constant. The bowel movements were described as extremely loose, often diarrhea. He had seen only some small flecks of blood on the stool. He usually had moderate gas and cramping prior to a bowel movement and this had been increasing in frequency. No foods seemed to aggravate the problem. He was bothered more in the morning, often having two or three bowel movements before going to work. He had had no nausea or vomiting and no upper abdominal pain. His father died at age 60, during an operation for cancer of the colon.

His past history and system review were unremarkable. On physicial examination, the patient was 67 inches tall and weighed 214 lbs. He was a friendly, cooperative, moderately obese, white male. Examination revealed no abnormalities of the pulmonary or cardiovascular systems. His abdomen was obese and nontender with active bowel sounds. There was no hepatosplenomegaly and no palpable masses. Rectal exam revealed normal sphincter tone. Prostate was not enlarged. The hemoccult was positive. His hematocrit was 46 percent. We were, at this point, left with a history of "colitis" and a hemoccult positive stool. Proctosigmoidoscopy to 24 centimeters showed an extremely hyperemic, friable mucosa throughout. There were no ulcers or neoplasms. Barium enema revealed the entire descending colon and rectosigmoid to be involved with what appeared to be a granulomatous colitis. (Figure 1) Upper gastrointestinal and small bowel x-rays were normal. The patient was admitted to St. John's Hospital for colonoscopy by Dr. Graham. Dr. Graham, could you tell us what you found on colonoscopy?

DR. JAMES GRAHAM *(Clinical Professor, Department of Surgery)*: I rated this patient as Group 2, Class B. My groupings, one through four, relate to the permanency or irreversibility of such pathologic changes as fibrosis, metaplasia, and neoplasia. My classification, A through D, relates to the severity of reversible inflammatory activity such as edema, hemorrhage, ulceration, and friability. Continuity and total length of pathologic changes are important criteria. Colonoscopy for ulcerative colitis includes serial photography of the colon. In this case, colonoscopy revealed the typical picture of moderately active ulcerative colitis with edema of the mucosa, evidenced by the smooth, glistening appearance and the rounding of the contours of the folds and valves; and multiple, pinhead sized submucosal hemorrhages.

DR. CLARK: The patient was started on prednisone in high doses as well as Azulfidine and he has been gradually tapered over the last two months to a lower dose of prednisone. He is now down to 20 mg of prednisone every other day, and eight gm of Azulfidine daily. Proctoscopic examinations have indicated remarkable improvement. The patient feels better, although I would like to emphasize that he presented himself with minimal symptoms. I would like to leave the rest of the time for Dr. Graham to talk about ulcerative colitis.

DR. GRAHAM: Membranous ulcerative colitis can be a grim and discouraging disease even though the symptoms may seem unimportant at first: nothing more than mucus, a little bleeding, or frequent stools. However, when the disease has set in fully, the colon, all four feet of it including the rectum, becomes painfully bloated and inflamed. Hundreds of ulcers erode and denude the membrane. The ulcers bleed. They exude pus. The congested membrane turns out great quantities of mucus, which in small amount is the colon's natural lubricant. Water absorption through the edematous ileal and colonic membrane fails, a fault which, when combined with excessive mucus, allows liquid feces to pass through the colon unchanged. The result is a daily ordeal of 15 to 20 bloody, liquid stools mixed with mucus and pus, an irritated rectum, and eventually, a completely exhausted and depleted person.[1]

Months after the onset, or possibly after only weeks or days, the spongy, ulcerated colon can bloat, inflate as a thin, gas-filled tube, then burst and spill its contents into the peritoneal cavity. If this occurs, the patient is suddenly feverish, bedridden and deathly sick, a state of affairs that is called toxic megacolon with peritonitis. At this point medication is too

From the Department of Family Practice, Southern Illinois University, School of Medicine, Springfield, Illinois. Requests for reprints should be addressed to Dr. Charles R. Clark, Department of Family Practice, 419 North Seventh Street, Southern Illinois University, School of Medicine, Springfield, Ill 62702.

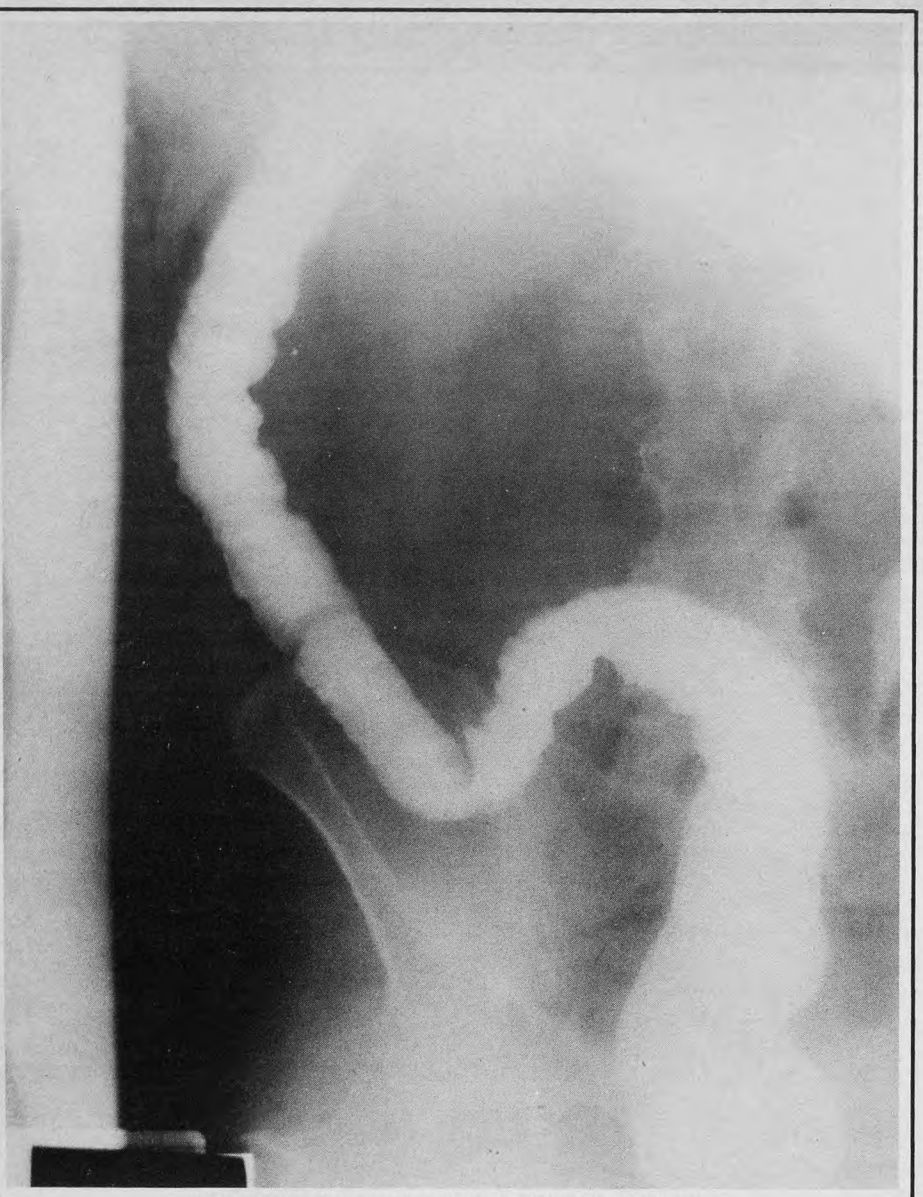

Figure 1. Barium enema of descending sigmoid colon.

late. Only surgery for removal of the diseased colon will help. Surgery becomes imperative as a life-saving measure. The operation, understandably, is hazardous but there is no choice. The patient is in a critical emergency.

Fortunately, ulcerative colitis is not necessarily this severe. Three out of four patients respond to medical management. Only five percent develop toxic megacolon. The disease may even self-correct, or just go away. And it does just this for one out of four patients.

Varieties of Ulcerative Colitis

What about most people who come down with ulcerative colitis? How many are cured? How many will have a lifetime of illness? Is surgery ever recommended early in the game and not just as an emergency? Are all cases of ulcerative colitis pretty much the same, or are there different forms of this disease?

There are many forms of ulcerative colitis, either slow or fast, steady or irregular, mild or violent. The disease affects all kinds of people, both the young and the old, the excitable and the placid. Most often (with half of the patients) the disease comes on slowly. Symptoms are imperceptible in the beginning, and they increase so gradually that the nature of the condition may not be apparent for some time, even for a year or more. In contrast to this, some patients (about 15 percent) are struck suddenly and violently. A person can be transfigured abruptly from health to serious illness in a matter of weeks.

Symptoms vary widely. Diarrhea, for example, is a frequent symptom, but it is not necessarily a consistent or characteristic mark of the disease. Patients can also be constipated with infrequent, firm, and bloody stools. The passage of mucus and pus is almost always a symptom. Weight loss can be severe or it may be insignificant. Ulcerative colitis may attack the slender as well as the obese.

Exacerbations and remissions are the rule. The intervals between attacks are unpredictable. For some patients, long spells of good health are not unusual. For others, flare-ups occur frequently and symptoms seem almost continuous and without interludes.

While ulcerative colitis is mainly a young person's disease, it is by no means confined to the teens and the twenties. Persons in their fifties and sixties also get ulcerative colitis. Four of every 10,000 persons in the United States and Canada have the disease.

Usually (in about 70 percent of patients), the disease begins in the rectum as ulcerative proctitis. Inflammation extends inward from the rectum in a reverse direction up into the colon for several inches or for several feet, all the way to the cecum.

Infrequently (in about five percent of patients), ulceration occurs only in the rectum. This is ulcerative proctitis.

Transmural ulcerative colitis (granulomatous colitis, Crohn's ileocolitis) – about 30 percent of patients – takes an opposite direction. It begins in the cecum or in both the ileum and the cecum. It is more mural than membranous. The colonic wall becomes thickened and stiff. As Crohn's disease spreads along the colon from the right side toward the rectum, it often skips long or short sections. The final result is a colon partly normal and partly diseased, like good and bad links in a chain. Generally, the rectum is not so severely inflamed and ulcerated in persons who have Crohn's colitis. However, abscesses and fistulas around and outside the rectum can be very troublesome.

Distressing complications develop in organs other than the colon when ulcerative colitis settles into a chronic course. Among these are hepatitis (liver impairment), arthritis, painful and lumpy skin inflammation (derma-

titis), bone softening, anemia, body salt alterations, and protein deficiency. Retarded growth and retarded sexual development can be serious problems with adolescent patients. Complications, including toxic megacolon, may develop in either form of colitis, membranous or granulomatous.

Outlook – Four Groups

There are four groupings for ulcerative colitis patients. These groups rank patients according to the seriousness of their disease. Each group represents about one fourth of the total number of patients who come down with ulcerative colitis. It is important to bear in mind that an individual patient with ulcerative colitis does not necessarily remain in one grouping over the years. He may change from high to low grouping or vice versa.

First Group

The first group, that is, the first fourth of patients, will experience only one attack of the disease. After recovering from this initial episode, they will be free from ulcerative colitis for the remainder of their lives. The time required for recovery from this encounter differs widely, from a few weeks to a year or longer.

Second Group

The second group of patients (one fourth of all those with ulcerative colitis) will experience recurring mild symptoms for a lifetime. The symptoms may reappear frequently and quickly or at long intervals, but they will be mild. Many patients in this group remain well and without symptoms for prolonged spells, sometimes years. To such persons, colitis is not an intolerable and disabling condition. It does represent, however, a handicap. It is a lifelong nuisance and an unpredictable disease.

Third Group

Patients in a third group, another one fourth of all ulcerative colitis patients, remain chronically sick. These people suffer from recurrent bouts of active and distressing ulcerative disease. Each flare-up responds more or less to treatment, sometimes quickly, sometimes stubbornly. The symptoms, however, do not leave completely, but hang on to some extent throughout the intervals of freedom from active inflammatory colitis. Patients in this group are forced to give in to their disease. They fashion their lives around their colitis. There are irreversible pathologic changes in the colon: scarring that has produced foreshortening or strictures; disseminated polyposis, and pseudopolyposis.

Fourth Group

The final one fourth of patients suffer almost continuous chronic disease. There is progressive involvement of more and more of the colon until the entire organ is ulcerated and infected. Complications appear – strictures, rupture of the intestine, hemorrhages, fistulas from the intestine and the rectum out through the vagina and the urinary bladder, or toxic megacolon. Many of these patients become severely debilitated. Weight loss can be frightful. The frequent and sudden urges for bowel movements end in loss of sphincter muscle control. Patients in this group are the 25 percent of colitis sufferers who come to surgery for removal of the colon and rectum.

Predicting the Outcome

The outcome is often foreshadowed by the severity of the initial attack. The more severe the beginning, the more likely it is that the final outcome will be bad. On the other hand, it is possible for a mild attack to worsen rapidly into an explosive and rampaging disease. Just as unpredictably, a nearly fatal attack of toxic megacolon may be resolved as mild ulcerative colitis that never threatens seriously again. In general, the outcome for a particular patient is not predictable.

Age is a factor – the younger the patient at the outset of the illness, the worse the outlook. Duration of the disease makes a difference. The longer a patient has had either persistent symptoms or has had frequently recurring flare-ups, the more likely it is that the condition will end up badly. Put another way around, the fewer the symptom-free remissions and the longer the time between remissions, the worse the outlook.

The amount of colon involved in the ulcerative process is related directly to the seriousness of the disease. If most of the colon is involved, the outlook is poor.

There are no tests for predicting whether an initial attack is to be a solo episode and a "one time only" ulcerative colitis, or is the forerunner of recurrent bouts over the years ahead.

Medication

The most frequently used drug is salicylazosulfapyridine, from two to 12 gm daily in divided doses.

Steroids play an important role in suppressing inflammation. They help stabilize body processes and assist in mobilizing strength for weathering an attack or for getting into better condition for surgery. This is of great importance when toxic megacolon sets in. The shock of this complication can be devastating, and here steroids can be used for emergency support.

Steroids are not without danger. They cause water retention, body salt abnormalities, stomach ulcer, bleeding, and retarded growth. The hazards are reduced by using steroids for only short periods of time and discontinuing them as soon as the symptoms of colitis improve.

The effectiveness of steroids wears off. In a relatively short time, their harmful effects can outweigh the good. The length of time steroids can be used safely for bringing an acute attack under control becomes a matter of judgment with each patient. In the presence of toxic megacolon, steroids should be continued only so long as there is unequivocal improvement as shown by eight and 12 hourly abdominal x-ray films. Otherwise perforation can be masked.

Follow-up

The ordinary ulcerative colitis patient, one not experiencing an acute attack, may be checked at intervals of three to 12 months, depending upon the severity of the symptoms.

Follow-up is primarily by sigmoidoscopic examination. This is performed every six to 12 months. X-ray examination is not necessary this often unless symptoms are changing rapidly. Then, simple x-ray pictures are taken rather than the "barium enema."

Sigmoidoscopic examination is supplemented by colonoscopic viewing of the deep interior of the colon. With this flexible, fiberoptic viewing instrument, the physician can see the interior of the entire colon and determine the extent and inner limit of the disease, that is, how far the ulcerations have advanced up the colon.

Cancer occurs more frequently than usual in patients with long standing ulcerative colitis. Cancer is not a threat in the early stages of the disease. It is more likely to be a factor in a patient whose colitis began early in life and in whom colitis has been persistent for ten years. Cancer is also more likely to develop in a colon that is involved totally from the cecum to the rectum and anus.

Surgery

One of every four persons with ulcerative colitis will need surgery. These are the people in the fourth group. The reasons for surgery are: (a) an acute complication develops suddenly, or (b) a progressively debilitating course sets in. Surgery is always a serious matter because it requires removal of the entire colon and rectum.

For many persons who will need an operation – those with acute complications – the need for surgery may arise soon after the illness starts. This is not to say that acute complications are confined to the early years of ulcerative colitis. Any of the serious complications may appear after many years of recurring "attacks" of colitis.

For the remainder of the patients – those with slowly advancing but relentlessly debilitating colitis – the turning point at which surgery becomes inescapable may not be reached for years.

Urgent Complications

Complications can appear with dramatic suddenness. Even so, there may be time for intensive medical treatment in the hope of avoiding removal of the colon. Just as likely, there may be no time for treatment and a decision for surgery might have to be made within hours. Hopefully, surgery will never be needed, but the patient must have knowledge of the nature and consequences of surgery in case the need for a quick decision should arise.

Perforation presents a very urgent acute complication. Here, one of the ulcers, large or small, erodes the colonic wall deeply enough to penetrate the full thickness. Peritonitis develops immediately.

Bleeding can be critical. Bleeding from the spongy, ulcerated lining membrane may be so persistent and steady over a period of weeks, or it may be so heavy for a few days that loss of blood becomes critical.

Fibrosis may seriously narrow the colonic lumen. An obstruction may have to be corrected by removal of the entire colon because of problems with healing. In the case of granulomatous colitis, an eight inch margin of good colon on either side of an obstruction may suffice.

Toxic megacolon is a most serious complication. If a patient does not respond unequivocally to steroids within hours, the situation is critical. Proctocolectomy in one or more stages is risky, but is less risky than a prolonged trial with steroids. The inflated, thinned colon can disintegrate and burst without warning.

The surgeon may choose to remove the colon and rectum in two stages. At the first operation, temporary openings are made in the ileum and the colon. The purpose is release of pressure and inflammation. Later, the colon and rectum are removed with less risk of shock and peritonitis.

Chronic Complications

In the more chronic case, that of the colon cripple, a decision for surgery is not made quickly. It comes after months or years of suffering with colitis. For these people ulcerative colitis proves plainly unmanageable. Despite prolonged and intensive treatment, frequent hospitalizations, severe restrictions of diet, and restrictions of physical and social activities, together with loss of time from work, the patient finds himself crippled by his disease. It becomes evident that life with a colon that is this badly diseased can never be satisfactory. The turning point is not well defined. The decision evolves gradually.

What makes the decision so difficult with membranous colitis is that although proctocolectomy will cure the disease, the patient will need a substitute opening, an ileostomy.

Every person with ulcerative colitis has the right to an understanding at the beginning of his illness that there is some chance that surgery will be necessary eventually. The chance is real indeed in view of the fact that one in every four persons with ulcerative colitis will need to have the colon and rectum removed. The patient and his family must be suitably prepared psychologically and emotionally. All concerned must be ready for a solid decision if and when the time comes. This is essential, since the question of surgery can arise unexpectedly and then a decision must be made under stress. The consequences of surgery should have been thought through ahead of time.

Ileostomy

Ileostomy is an important facet of ulcerative colitis, so much so that any discussion of the problems of ulcerative colitis must include ileostomy. To the person whose colitis has overwhelmed him, ileostomy can be a way out of a miserable situation, considerably less than ideal to be sure, but a welcome option.

How much of a handicap is an ileostomy? Surprisingly, it is minimal. It will not alter a person's work or job capabilities. It will not interfere with family and social life. It need not mark a person as unusual. People in all walks of life have ileostomies – businessmen, housewives, nurses, doctors, secretaries, entertainers, public figures. No job or position is excluded.

The key to life with an ileostomy lies in proper instruction and training. Efficient and meticulous care have to be mastered. Initial training is followed by continuing educational and informational programs. Stoma therapists in major hospitals are ready with technical help and assistance to patients and surgeons.

DR. WILLIAM STEWART *(Chairman, Department of Family Practice):* I'd like to ask Dr. Clark if there was any evidence or indication of problems in this patient's emotional make-up? Did he have any problems at home or at work?

DR. CLARK: I have had no indication of psychological problems. He did have a history of beer drinking when he first started seeing me, and I insisted at the start, before we had even done the proctoscopic examination, that he cut down on his drinking, which he did very easily. He is now drinking maybe one or two beers a week. I think he was drinking out of habit and I don't have any feeling of real psychological problems with this man.

DR. STEWART: Dr. Graham, what do you think about the role of psychological problems in patients with ulcerative colitis?

DR. GRAHAM: Well, my answer is

that I think it's the other way around. Anyone who has to go to the toilet ten or 15 times a day passing bloody mucus, will be nervous. It is a fact that we do see more ulcerative colitis in persons who are intellectually trained and disciplined, college graduates. The incidence is higher in the Jewish population. There is no question that emotional tension and tight situations can aggravate ulcerative colitis. I do not look to emotional factors as the cause of ulcerative colitis.

DR. F. PAUL LAFATA *(Clinical Associate Department of Family Practice):* In your evaluation, you classify Group 3 as irreversible. Would you say that all of irreversibles are surgical candidates, or should be?

DR. GRAHAM: This is the gray group. Many persons in this Group 3 who are colon cripples would have a much nicer life, and their families would have a nicer life, with an ileostomy. These people, the colon cripples, can be brought into contact with persons who have gone down the road of ulcerative colitis and now have an ileostomy.

DR. LAFATA: Is the ulcerative colitis ever ushered in so suddenly that a person could be well today, and tomorrow have a temperature of 104F and bloody diarrhea?

DR. GRAHAM: Absolutely. That's one of the modes of onset of ulcerative colitis.

DR. LAFATA: A third question. Can patients with ulcerative colitis use alcohol?

DR. GRAHAM: I haven't restricted it. The patient presented here drinks, but statistically I think there are just as many people who drink as much as he did and who don't have ulcerative colitis. I have not felt that it's a big factor. It's up to the family doctor to decide whether the patient is using too much alcohol, not from the standpoint of ulcerative colitis, but otherwise. I don't think there is a direct connection. I would like to say a word about alcohol and the ileostomate or the colostomate. I have been to conventions with these people and they eat popcorn and drink martinis, just like anyone, without difficulty.

DR. CLARK: This man's symptoms did clear to a certain extent, as far as the diarrhea was concerned, when he cut down on his alcohol. He improved quite a bit. But the proctoscopic examination did not.

DR. CLINTON TOEWE *(Assistant Chairman, Department of Family Practice)*: Dr. Graham, you stated that the ileorectal anastomosis is usually the preferred surgical procedure for granulomatous colitis. Is this also true for membranous colitis or is there a place in this disease for partial colon resection?

DR. GRAHAM: If there is not too much involvement from a sigmoidoscopic standpoint of the distal four or five inches of rectum, and then at laparotomy the distal rectum is quite pliable, isn't thickened, isn't shortened, and the mesorectum is not very indurated, you can give very serious consideration to ileorectal anastomosis. This is true for both membranous and granulomatous colitis.

DR. RONALD STAUBLY *(family practice resident)*: Dr. Graham, is the choreoembryonic antigen test used with ulcerative colitis? Are the results variable, and is there any prognosis attached to the results?

DR. GRAHAM: We have not used CEA routinely with either carcinoma or ulcerative colitis either to detect or to follow-up. There are too many variables at present.

DR. CLARK: Dr. Graham, after you had done the colonoscopy on this man, you shared your philosophy about treating these people with steroids and Azulfidine with me, and I'd like you to share that with the rest of the group. Your feeling was that you have to do something for him.

DR. GRAHAM: I think it is very important to treat the patient positively. The physician shouldn't hesitate to make his patient better as quickly as he can. As soon as you can make a diagnosis, start treatment and make the patient better. Then he will settle down and stay with you. He will be better off for not wandering around, and he'll have a better understanding of his condition. I put these patients on adequate doses of steroids, get improvement right away, and then take them off the steroids quickly.

DR. STEWART: Do you consider the patient presented today to have granulomatous disease or membranous ulcerative colitis?

DR. GRAHAM: I classed him as membranous, but one can certainly be wrong.

DR. CLARK: What about follow-up studies? I understand that your basic tool is sigmoidoscopy. At what point would you want to get more x-rays or would you want to do colonoscopy again?

DR. GRAHAM: You don't need colonoscopy again until it is indicated clinically. The follow-up is clinical history and sigmoidoscopy. X-ray and colonoscopy are auxiliaries. Their use depends upon how the patient is responding clinically: weight, gastrointestinal symptoms, and how he feels. If you've put your message across about ulcerative colitis and what one can expect for a lifetime, the patient doesn't need to come back every month. Perhaps after you have the disease under control, he can come back every month for two or three times just to get settled down. Then he can go for six months and a year.

DR. LAFATA: I have one more question about Azulfidine. Do you use it on all your patients, and for how long a period of time do you use it?

DR. GRAHAM: I try to get clearance with steroids at first. Whether I follow through with Azulfidine will depend upon the clinical history of number and character of bowel movements, cramps, weight, and general well being. If I can get them off Azulfidine, fine. Some patients get down to about four Azulfidines a day. When I try to taper them, they don't feel as well so I go back up to four. They stay at that level for quite a variable period. I am always trying to get them off medication.

DR. STEWART: What is current thought with respect to aggravation of ulcerative colitis by milk? Is this the lactase deficiency?

DR. GRAHAM: I suppose so. Some people just do much better when they are off milk.

DR. CLARK: What about follow-up as far as carcinoma of the colon?

DR. GRAHAM: The general rule is ten-year history and total involvement. Here, one does more frequent x-ray examinations and colonoscopy. If by either method, questionable polyps are identified, the patient may be in for losing his colon. Stricture is another tip-off to carcinoma.

DR. CLARK: When you say ten years, is this ten years of continuous disease?

DR. GRAHAM: Intermittent.

References

1. Bercovitz ZT, et al: Ulcerative and Granulomatous Colitis. Springfield, Ill, Charles C Thomas, 1973