Sclerosing Cholangitis

Anatomical Distribution of Obstructive Lesions

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The cholangiograms of 36 patients with sclerosing cholangitis were reviewed. The mean age of the patient group was 43 years, and the mean disease duration was 4.5 years. Seventeen of the patients had associated inflammatory bowel disease. The mean serum bilirubin was 6.8 mg/dl, the mean SGOT was 105 IU/L, the mean SGPT was 108 IU/L, and the mean serum alkaline phosphatase was 534 IU/L. The cholangiograms demonstrated involvement of the extrahepatic bile ducts in 33 patients, involvement of the hepatic duct bifurcation in 33 patients, and involvement of the intrahepatic bile ducts in 35 patients. The cholangiograms were graded as to the areas of the most severe obstructive involvement. In 24 patients the area of most severe involvement was the hepatic duct bifurcation. In eight additional patients the hepatic duct bifurcation, along with the extrahepatic ducts and/or the intrahepatic ducts, were felt to be the areas most severely affected. This predilection for severe obstructive disease at the hepatic duct bifurcation in sclerosing cholangitis held for both patients with and without inflammatory bowel disease. Thus, most patients with sclerosing cholangitis have cholangiographic evidence of diffuse extrahepatic and intrahepatic biliary tract disease, with the hepatic duct bifurcation being the area generally most severely affected.

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SCLEROSING CHOLANGITIS is an idiopathic disease of the hepatobiliary system characterized by inflammatory strictures of the biliary tree, and often by liver parenchymal changes varying from portal inflammation (pericholangitis or triaditis) to biliary cirrhosis.[1,2] It occurs most often in young men, frequently is associated with inflammatory bowel disease, and usually presents with jaundice.[3-5] Presently any patient with one or more inflammatory biliary tree strictures in whom iatrogenic injury or malignancy can be excluded, is considered to have sclerosing cholangitis. Liver, bile duct, and gallbladder histology can be helpful in confirming the diagnosis, but all are relatively nonspecific. Currently, the most accurate means of making the diagnosis of sclerosing cholangitis is by cholangiography. Multiple strictures, the appearance of beading, small lumen caliber, and the failure of the biliary tree to dilate proximal to a stricture are all characteristics that allow one to diagnose sclerosing cholangitis on the basis of cholangiography. Recently we have noted a characteristic distribution of obstructing biliary tract lesions in sclerosing cholangitis. This report reviews our patients with sclerosing cholangitis with an emphasis on location of major biliary tract lesions.

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Clinical Material

Between 1973 and 1983, 36 patients with sclerosing cholangitis were seen at The Johns Hopkins Hospital with cholangiograms adequate to evaluate both the intra- and extrahepatic biliary tree (Table 1). Twenty-four of the patients were male and 12 were female. Thirty were white and six were black. Age of onset of biliary tract symptoms ranged from 3 to 69 years, with a mean of 38 years. Disease duration prior to evaluation at Hopkins ranged from 1 month to 21 years, and averaged 4.5 years. Past history was available for 34 of the 36 patients. Seventeen patients had inflammatory bowel disease. Thirteen patients had ulcerative colitis, two patients had Crohn's disease, and in two patients the inflammatory bowel disease was unclassified. Duration of the inflammatory bowel disease at the time of evaluation at Hopkins ranged from 1 to 29 years, with a mean of 13.5 years. None of the 17 patients with inflammatory bowel disease had a family history of inflammatory bowel disease. Five of the 36 patients had a history of chronic pancreatitis, or repeated bouts of acute pancreatitis. Nineteen patients had had a total of 24 prior biliary tract procedures. Cholecystectomy and common duct exploration were the operations most commonly performed. Only one patient underwent T

TABLE 1. *Thirty-six Patients with Sclerosing Cholangitis*

Number	36
Sex	24 male
	12 female
Race	30 white
	6 black
Age at onset	38 years (mean)
Duration of disease	4.5 years (mean)
Associated diseases	
Inflammatory bowel disease	17
Pancreatitis	5
Prior biliary surgery	19

tube insertion for long-term drainage. Five of the 36 patients underwent total proctocolectomy for their inflammatory bowel disease. In four of the five patients, the diagnosis of sclerosing cholangitis predated the colectomy by a mean of 2.8 years. The fifth patient had undergone a colectomy for Crohn's colitis 16 years prior to the onset of her sclerosing cholangitis.

Symptoms at the time of diagnosis were available for 35 patients (Table 2). In 30 patients the first symptom was jaundice, although the serum bilirubin was elevated in only 19 at the time of admission to Hopkins. In the other 11 patients the bilirubin had returned to normal, demonstrating the episodic nature of jaundice in sclerosing cholangitis. Fever was a presenting symptom in 12 patients. Ten of these 12 patients had had prior biliary tract surgery. Only two patients without prior biliary tract surgery had fever as a prominent part of their disease presentation. Abdominal pain, chills, and weight loss were present less frequently. Interestingly, three patients were asymptomatic and had their disease diagnosed only because of a markedly elevated serum alkaline phosphatase level. Liver function tests were available at the time of evaluation at Hopkins for 34 of the 36 patients (Table 3). Serum bilirubin levels ranged from 0.1 to 38.0 mg/dl and averaged 6.8 mg/dl; serum glutamic oxaloacetic transaminase (SGOT) ranged from 22 to 326 IU/L and averaged 105 IU/L; serum glutamic pyruvic transaminase (SGPT) ranged from 20 to 344 IU/L and averaged 108 IU/L; and the serum alkaline phosphatase ranged from 101 to 1092 IU/L and averaged 534 IU/L.

TABLE 2. *Presenting Symptoms (N = 35)*

Symptom	Number of Patients
Jaundice	30
Fever	12
Chills	8
Abdominal pain	7
Weight loss	5
Ascites	1
Esophageal bleeding	1
Asymptomatic	3

TABLE 3. *Liver Function Tests (N = 34)*

Test	Mean (Range)
Total bilirubin (nl < 1.1 mg/dl)	6.8 mg/dl (0.1–38.0 mg/dl)
SGOT (nl < 20 IU/L)	105 IU/L (22–326 IU/L)
SGPT (nl < 20 IU/L)	108 IU/L (20–344 IU/L)
Alkaline phosphatase (nl < 105 IU/L)	534 IU/L (101–1092 IU/L)

Cholangiographic Findings

The distribution of disease in the biliary tree in the 36 patients in this series was evaluated on the basis of 57 cholangiograms. Approximately three-quarters of these studies were performed at Hopkins. The remaining studies were performed elsewhere but were reviewed at Hopkins. Twenty-seven patients underwent successful percutaneous transhepatic cholangiography. Nineteen patients underwent successful endoscopic retrograde cholangiography. In six patients outside T tube cholangiograms were available for review. Five patients had operative cholangiograms. Four patients had cholangiograms for review performed by three of the above four routes; 13 patients had studies performed by two of the routes; and the remaining 19 patients had one cholangiogram available for review.

The extrahepatic biliary tree, bifurcation, and intrahepatic biliary tree were reviewed for extent of involvement and severity of involvement (Table 4). The area of the bifurcation was considered to include 2 cm of the right hepatic duct, 2 cm of the left hepatic duct, and 2 cm of the common hepatic duct adjacent to the bifurcation. In many instances the entire biliary tree was not visualized on a single cholangiogram. However, no patient was included unless his entire biliary tree could be evaluated by reviewing and combining all cholangiograms available. The biliary tree was considered involved if there

TABLE 4. *Cholangiographic Findings in 36 Patients with Sclerosing Cholangitis*

	Number of Patients
Involved areas	
Extrahepatic ducts	33
Bifurcation	33
Intrahepatic ducts	35
Area(s) of most severe involvement	
Extrahepatic ducts	0
Bifurcation	24
Intrahepatic ducts	3
Bifurcation and extrahepatic ducts	3
Bifurcation and intrahepatic ducts	2
Bifurcation and extrahepatic ducts and intrahepatic ducts	3
Extrahepatic ducts and intrahepatic ducts	1

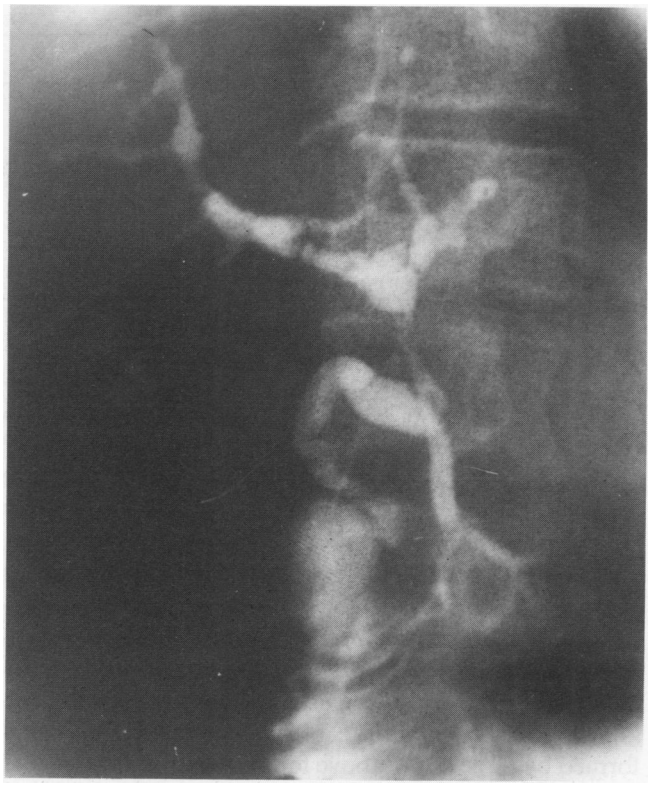

FIG. 1. Percutaneous transhepatic cholangiogram of a 34-year-old man with inflammatory bowel disease of 3 years duration, and jaundice of 3 years duration. Cholangiogram demonstrates diffuse intra- and extrahepatic biliary tree involvement, with almost total biliary obstruction just beyond the confluence of the right and left hepatic ducts.

was evidence of beading, stricture formation, scalloping, or irregularity of the smoothly tapered biliary tree. In 33 of the 36 patients the extrahepatic biliary tree was diseased; in 33 patients the bifurcation was involved; and in 35 patients the intrahepatic biliary tree was involved. Severity of involvement with sclerosing cholangitis was difficult to compare cholangiographically from one patient to another. Comparison of one patient with sclerosing cholangitis to another is probably best done on the basis of clinical course, liver function tests, and liver biopsy. Therefore, severity of involvement was evaluated only within each cholangiogram, comparing the disease involving the extrahepatic biliary tree, the bifurcation, and the intrahepatic biliary tree. An effort was made to judge severity on the likelihood of a lesion or lesions causing significant obstruction of the biliary tree. Wherever the most restrictive lesions were located, was considered to be the most severely involved. By this criterion, the bifurcation was the area most severely involved in 24 of the 36 patients (Figs. 1–6). In five other patients the bifurcation was judged to be the area most severely involved along with the extrahepatic biliary tree in three patients and the intrahepatic biliary tree in two patients. In addition, in three patients the bifurcation, the extrahepatic biliary tree, and the intrahepatic biliary tree were judged to be equally severely involved. Thus, in 32 of the 36 patients with sclerosing cholangitis the bifurcation was at least as involved as the extra- or intrahepatic biliary tree, and in most instances more severely involved. Of the four remaining patients the area of most severe involvement was the intrahepatic biliary tree in three patients, and the intrahepatic and extrahepatic biliary tree in one patient.

Clinical Course

During or following their initial evaluation at Hopkins, 22 of the 36 patients underwent surgery at Hopkins. Nineteen of these 22 patients were amongst those individuals whose most severe disease was at the hepatic duct bifurcation. These 19 patients underwent resection of their hepatic duct bifurcation and extrahepatic biliary tree, and were reconstructed with the aid of silastic transhepatic biliary stents. One patient died 2 months after surgery of liver failure. The remaining 18 patients were discharged from the hospital and have been followed for a mean of 18.7 months. Details of the follow-up of the first 11 patients operated upon have been previously published.[3]

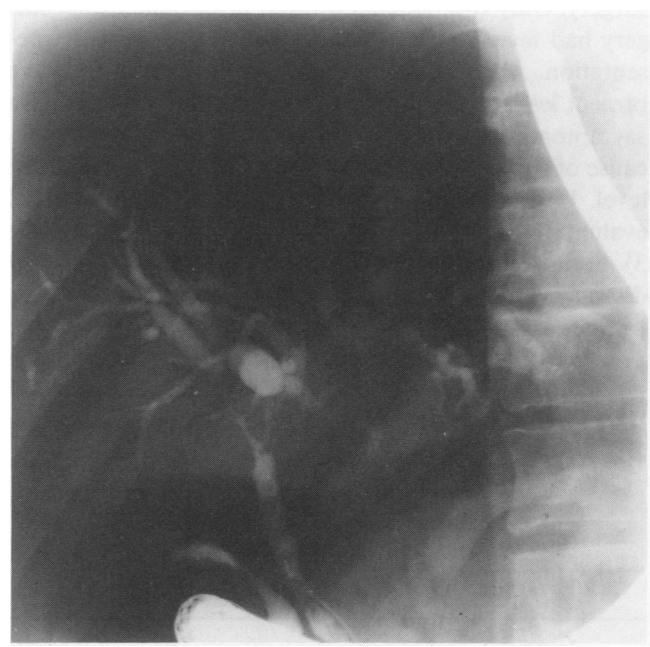

FIG. 2. Endoscopic retrograde cholangiogram of a 42-year-old man. Patient has diffuse disease involving the intra- and extrahepatic biliary tree, but the area of most severe narrowing clearly involves the hepatic duct bifurcation. Initially thought to represent a proximal biliary tumor, an exploratory laparotomy and 5-year follow-up without progression have demonstrated the lesion to be benign.

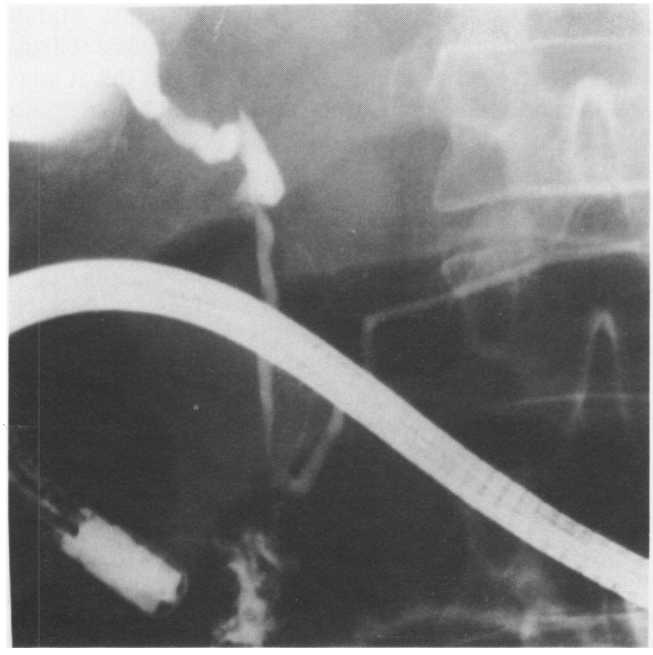

FIG. 3. This 49-year-old man had had Crohn's disease for 20 years, at the time of his presentation with jaundice. This endoscopic retrograde cholangiogram represents a common finding with sclerosing cholangitis. The contrast media fills the extrahepatic biliary tree up to the area of the hepatic duct bifurcation, where it stops because of total obstruction. The gallbladder is becoming distended as the endoscopist tries to force contrast media above the bifurcation.

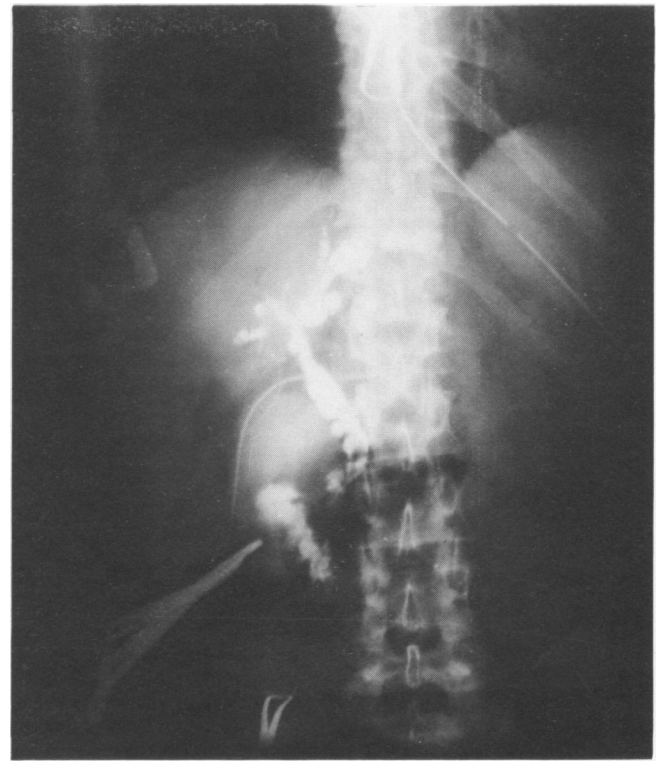

FIG. 4. This 40-year-old man had undergone total proctocolectomy for ulcerative colitis in an unsuccessful effort to stabilize his sclerosing cholangitis. This operative cholangiogram demonstrates diffuse intra- and extrahepatic involvement with almost complete obstruction at his bifurcation.

Three additional patients underwent surgery at Hopkins. One had a bifurcation lesion that at surgery proved to be an unresectable proximal biliary carcinoma. This followed a 6-year course and two prior operative procedures at another hospital. Cholangiograms during the 6-year period were characteristic of sclerosing cholangitis. Whether this represents sclerosing cholangitis with the subsequent development of a bile duct tumor, or merely a prolonged course of a bile duct tumor cannot be determined with certainty; however, we favor the former. This patient had a Ring catheter inserted percutaneously and was treated with radiotherapy. Another patient, who had had a T tube in place for several years, was operated upon and a biliary-duodenal fistula divided and repaired. A sphincteroplasty was performed, and another T tube inserted. The T tube was removed 1 month later and the patient has done well over a 3-year follow-up. The final patient underwent a choledochojejunstomy which strictured 5 months later. The stricture was dilated with a Gruntzig catheter and the patient is doing well 14 months later.

Fourteen of the 36 patients have not undergone surgery since their initial evaluation at Hopkins. One of those 14 patients was markedly jaundiced and died 2 months after admission of liver failure. The remaining 13 patients did

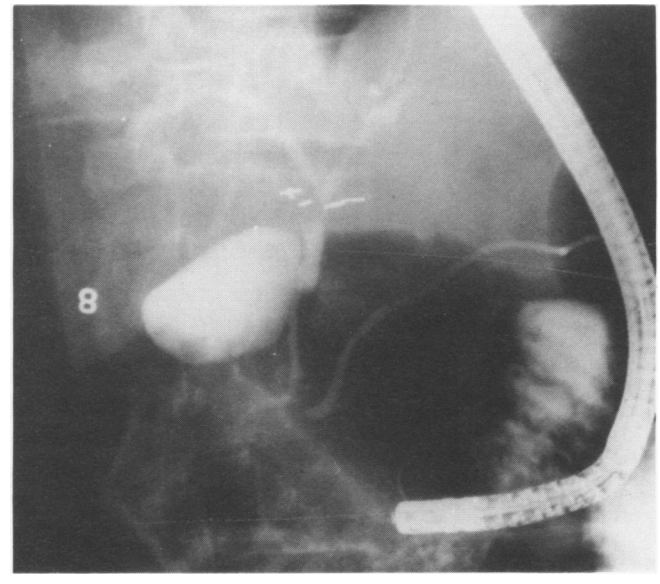

FIG. 5. This 50-year-old man with ulcerative colitis was deeply jaundiced. Endoscopic retrograde cholangiogram demonstrates almost total obstruction of his hepatic duct bifurcation. Serum bilirubin returned to normal following resection of his bifurcation with reconstruction.

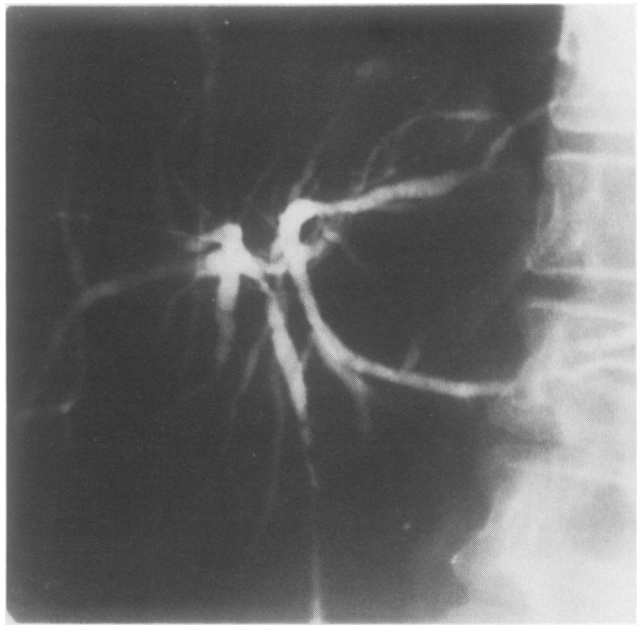

FIG. 6. This 61-year-old woman with ulcerative colitis had a serum bilirubin of 18 mg/dl. A percutaneous transhepatic cholangiogram revealed almost total obstruction of her hepatic duct bifurcation. Because of fibrosis throughout her biliary tree, proximal dilatation did not occur. At surgery, her hepatic duct bifurcation was resected with reconstruction carried out over silastic stents. Her serum bilirubin returned to near normal levels. The pathology demonstrated fibrosis and scarring.

not have persistent jaundice and were therefore not considered to be surgical candidates. Eleven of the 14 patients are alive a mean of 27.3 months since their initial evaluation and are doing well. Even though intermittent episodes of fever and jaundice occur, persistent jaundice has not developed in any. Two patients have been lost to follow-up.

Discussion

The diagnosis of sclerosing cholangitis is inexact, and criteria for inclusion or exclusion vary from one author to another. The clinical presentation is nonspecific, with patients generally coming to medical attention because of jaundice. Despite the name sclerosing cholangitis, fever and chills are unusual unless the patient has undergone prior biliary tract surgery. Only two of the 12 patients in our series with fever as a prominent feature of their presentation had not undergone prior biliary surgery. The insidious onset of jaundice, which often runs an episodic course, is the most common means of presentation. Sclerosing cholangitis cannot be distinguished clinically from other forms of benign or malignant biliary obstruction. The presence of inflammatory bowel disease may suggest the diagnosis of sclerosing cholangitis, but ulcerative colitis is also associated with cholangiocarcinoma.

Operative findings are obviously important aids in confirming the diagnosis of sclerosing cholangitis, and in excluding cancer. A thickened fibrotic extrahepatic biliary tree suggests sclerosing cholangitis. However, this is nonspecific and a malignancy or an iatrogenic bile duct injury can produce the same findings. Histology of the biliary tree is also nonspecific. Extensive fibrosis and scarring are generally seen, and the gallbladder may contain a lymphocytic inflammatory picture. There are, however, no pathognomonic histologic findings. In addition, many of the bile duct cancers produce a great deal of scarring and histologically can be difficult to differentiate from sclerosing cholangitis. The liver biopsy usually shows pericholangitis, consisting of acute and chronic inflammatory cells in the portal triads with varying degrees of fibrosis. In later stages of the disease biliary cirrhosis may be present. However, in some patients intrahepatic cholestasis may be the only histologic abnormality, and in a few individuals the liver biopsy may be normal.

One is thus left with cholangiography as the gold standard for the diagnosis of sclerosing cholangitis. Any patient who has one or more biliary tract strictures, in whom iatrogenic injury or malignancy can be ruled out, currently is considered to have sclerosing cholangitis. The presence of biliary tract stones, or the history of prior biliary surgery, should not exclude the diagnosis. Most patients with sclerosing cholangitis have involvement of both their extrahepatic and intrahepatic biliary tree. In our series only three of the 36 patients did not have involvement of their extrahepatic biliary tree cholangiographically. If one includes the bifurcation with the extrahepatic bile ducts, this number drops to one. Likewise, only one of the 36 patients had no cholangiographic evidence of disease involving their intrahepatic biliary tract. In the past, before sizeable series of patients with sclerosing cholangitis were collected, involvement of only the extrahepatic biliary tree, with sparing of the intrahepatic ducts, was thought to be a common pattern. Our series suggests that is not the case and that virtually all patients have diffuse disease.

Estimating the severity of disease involvement in the three areas (extrahepatic, bifurcation, intrahepatic) chosen in this study is somewhat arbitrary, and the grading is subjective. Nevertheless, criteria were adopted that could be adhered to consistently. A single tight stricture in a peripheral intrahepatic biliary radical was not considered as severe a lesion as a similar tight stricture occurring in the right or left hepatic duct, at the bifurcation, or in the distal common duct. The potential for a lesion to cause significant biliary obstruction was considered in the grading of severity. As a result this grading system would be more likely to consider disease at the bifurcation or in the extraheptic biliary tree as severe. However, if there were diffuse disease throughout the intrahepatic biliary tree without significant stricturing at the bifurcation or in the extrahepatic ducts (as in three patients in our series),

the intrahepatic disease was considered the most severe. Using these criteria, in 24 of the 36 patients the disease was most severe at the bifurcation. In an additional eight patients the disease at the bifurcation was just as severe as that in the rest of the biliary tree. Although our grading system is subjective, this study clearly demonstrates that the bifurcation is involved with significant stricturing in the majority of patients with sclerosing cholangitis.

Distribution of disease was documented by cholangiography in 47 patients with sclerosing cholangitis from the Mayo Clinic.[1] Diffuse disease was present in 81% (38/47), with 19% (9/47) having only extrahepatic involvement. In their series they had no patient with only intrahepatic involvement, but stated they had reviewed outside cholangiograms on two patients with only intrahepatic disease. The Mayo Clinic group did not comment on involvement of the bifurcation. A series of patients with sclerosing cholangitis reported from UCLA documented 33 patients with diffuse disease involving the extra- and intrahepatic ducts.[2] The series also included four patients (11%) with involvement primarily of the distal common duct. Interestingly, the authors in describing the cholangiographic findings in their patients with diffuse disease make the statement, "Often the most prominent stricture was at the confluence of the hepatic ducts."[2]

It is likely that one of the reasons the predilection of the bifurcation for severe disease involvement in sclerosing cholangitis has not been documented better in the past is the failure of significant biliary tract dilatation to occur proximal to a stricture. Because of invariable involvement of the intrahepatic bile ducts with this fibrotic process, the usual dilatation of the bile ducts that occurs proximal to an obstruction is inconstant, and often does not occur (Figs. 1–6). This failure of the intrahepatic biliary tree to dilate may distract one from identifying the bifurcation stricture and understanding its significance in the progressive course of the disease.

The bifurcation involvement in sclerosing cholangitis is often suggestive of the radiographic appearance of a proximal biliary tumor, the so-called Klatskin tumor. In one of our patients the lesion at the bifurcation did indeed prove to be a tumor. This patient had had a 6-year course of sclerosing cholangitis with both extrahepatic and intrahepatic duct involvement, well documented by several cholangiograms and two operative procedures. She then developed bifurcation involvement, became jaundiced, was operated upon, and was found to have a bile duct cancer at the bifurcation. It seems most likely that the cancer developed on the background of sclerosing cholangitis, but it is possible that the tumor had been present for the entire 6 years and the cause of the biliary tract disease.

Nineteen of the 24 patients with severe bifurcation involvement have undergone resection of the bifurcation and extrahepatic biliary tree with reconstruction using silastic transhepatic stents. In all 19 of these patients the bifurcation stricture was available for histologic examination and in each instance the stricture was benign. Interestingly, in both sclerosing cholangitis and biliary tract cancer, the most common site of involvement is the bifurcation of the hepatic duct. Another similarity of these two diseases is their association with ulcerative colitis. The significance of these associations is unknown, but they may be important in understanding the pathogenesis of both disorders.

TABLE 5. *Comparison of Patients with Sclerosing Cholangitis (SC) with and without Inflammatory Bowel Disease (IBD)*

	SC (N = 19)	SC and IBD1 (N = 17)
Sex	12 M, 7 F	12 M, 5 F
Race	16 W, 3 B	14 W, 3 B
Age	42 years (mean)	43 years (mean)
Age at onset	37 years (mean)	39 years (mean)
Duration of disease	5.3 years (mean)	3.9 years (mean)
Presenting symptoms		
Jaundice	16/18	14/17
Liver function tests		
Bilirubin	5.9 mg/dl (mean)	8.6 mg/dl (mean)
SGOT	109 IU/LL (mean)	101 IU/L (mean)
SGPT	113 IU/L (mean)	103 IU/L (mean)
Alkaline phosphatase	485 IU/L (mean)	586 IU/L (mean)
Area of most severe cholangiographic involvement		
Bifurcation	14/19	10/17
Biliary surgery	14/19	8/17

The association of inflammatory bowel disease with sclerosing cholangitis is interesting, and in recent series has been documented to occur in approximately 50%.[1-3,5] Most of these patients have ulcerative colitis, but two of the 17 patients with inflammatory bowel disease in our series had Crohn's disease and in two others it was unclassified. It is tempting to propose that in patients with associated inflammatory bowel disease and sclerosing cholangitis there may be a different pathogenesis and natural history of the disease from sclerosing cholangitis occurring in the absence of inflammatory bowel disease. The heterogenous nature of sclerosing cholangitis suggests that the disease as currently classified may consist of several different disorders. However, a comparison of the patients in our series with sclerosing cholangitis with (17 patients) and without (19 patients) inflammatory bowel disease demonstrates no differences in terms of sex, race, age of onset of disease, or disease duration (Table 5). Likewise, in terms of presenting symptoms, liver function tests, and distribution of disease cholangiographically, there were no distinguishable differences. Finally, in terms of the need for surgery and subsequent course, there were no significant differences between the two groups. Thus,

from these data it would appear that sclerosing cholangitis is the same disease with or without inflammatory bowel disease. Five of our 36 patients had a history of chronic pancreatitis or repeated bouts of acute pancreatitis. The relationship of pancreatic disease to sclerosing cholangitis is not known, but it appears to be the second most commonly associated disorder.

The natural history of sclerosing cholangitis is not well documented. In the UCLA series of 37 patients, eight patients (22%) died over a mean follow-up of approximately 5 years.[2] Six of the eight patients who died survived 3 or more years after their disease was diagnosed. In the Mayo Clinic series adequate follow-up was available for 39 patients.[1] One-third of these patients (13) died a mean of 57 months following their diagnosis. Most of the deaths (11/13) were from liver failure. One patient died of bile duct carcinoma 13 years after the diagnosis of sclerosing cholangitis. In the present series from Hopkins there were only two deaths amongst 36 patients. Both patients died of liver failure, one 3 years and the other 16 years following the onset of their biliary tract symptoms. Mean follow-up for the entire group was 5.5 years. Even though the disease appears to be chronic, with survival possible for many years after the onset of symptoms, the development of progressive liver disease seems inevitable and many patients eventually succumb to liver failure. What role large duct obstruction plays in this progressive hepatobiliary disease is unclear. However, the recognition that significant stricturing involving the bifurcation occurs in the majority of patients with sclerosing cholangitis, along with the progressive changes of secondary biliary cirrhosis seen in most patients who die of this disease, suggest that large duct obstruction may be important. Our successful experience[3] with 11 patients previously reported with bifurcation disease and significant hyperbilirubinemia treated by bifurcation resection and reconstruction, supports this concept. If one looks at sclerosing cholangitis as a disease that involves the extrahepatic biliary tree, the bifurcation of the common hepatic duct, and the intrahepatic biliary tree, the disease can be totally eliminated from two of these three areas by resecting the bifurcation and extrahepatic biliary tree. On the basis of our present understanding of the pathogenesis of this disease, and its chronic course, this would seem to be a more prudent approach than other more extensive surgical procedures. Recurrence or extension of disease, with significant intrahepatic involvement, would of course limit the efficacy of this approach. Percutaneous transhepatic balloon dilatation of strictures in sclerosing cholangitis has been reported by others.[6] In our experience positioning of the dilating catheter in these small ductal systems with multiple strictures is difficult, and may be impossible, and the tough fibrous strictures often are refractory to balloon dilatation. Therefore, currently we are not optimistic about this approach.

Addendum

We have recently operated upon three patients with sclerosing cholangitis who had cholangiographic evidence of involvement *only* at the hepatic duct bifurcation. Before surgery the x-rays suggested a Klatskin tumor, but the resection specimens demonstrated only fibrosis and scarring.

References

1. Wiesner RH, LaRusso NF. Clinicopathologic features of the syndrome of primary sclerosing cholangitis. Gastroenterology 1980; 79:200–206.
2. Thompson HH, Pitt HA, Thompkins RK, Longmire WP. Primary sclerosing cholangitis. A heterogenous disease. Ann Surg 1982; 196:127–136.
3. Cameron JL, Gayler BW, Herlong HF, Maddrey WC. Sclerosing cholangitis: biliary reconstruction using silastic transhepatic stents. Surgery 1983; 94:324–330.
4. Pitt HA, Thompson HH, Thompkins RK, Longmire WP. Primary sclerosing cholangitis: results of an aggressive surgical approach. Ann Surg 1982; 196:259–268.
5. Epstein O, Chapman RWG, Lake-Bakaar G, et al. The pancreas in primary biliary cirrhosis and primary sclerosing cholangitis. Gastroenterology 1982; 83:1177–1182.
6. Martin EC, Fankuchen EI, Schultz RW, Casarella WJ. Percutaneous dilatation in primary sclerosing cholangitis: two experiences. Am J Radiol 1981; 137:603–605.