# Surgical Aspects of Sclerosing Cholangitis

## *Results in 178 Patients*

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**Of 178 patients with sclerosing cholangitis treated since 1950, 88 patients had associated inflammatory bowel disease, 72 had no such history, and 18 had iatrogenic injury or stone disease. A total of 233 biliary operations were performed, with a 75% rate of temporary improvement after initial operation. Subsequent operations resulted in a lower success rate and a higher mortality rate. Radiologic findings included predominant extrahepatic, intrahepatic, and diffuse disease in 29%, 28%, and 43% of patients, respectively; no survival differences were noted. Seventy-five of one hundred three deaths (73%) were related to liver failure, bleeding, or sepsis. Of 14 patients undergoing portosystemic shunt, 13 died of surgical complications or related disease. Orthotopic liver transplantation was performed in 16 patients and resulted in eight deaths, mainly in patients who had previously undergone extensive surgical treatment. No survival differences were seen between the patients with inflammatory bowel disease, those without the condition, or those who had colectomy. Surgical treatment in patients with sclerosing cholangitis should be minimized. Orthotopic liver transplantation should be offered as the treatment of choice for patients with portal hypertension, refractory cholangitis, advanced cirrhosis, or progressive liver failure.**

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SCLEROSING CHOLANGITIS IS an insidious and uncommon inflammatory disease affecting the biliary ductal system that leads to obliteration and fibrosis of the ducts and, ultimately, to biliary cirrhosis. Inflammatory bowel disease (IBD) is associated with most cases of primary sclerosing cholangitis.[1] However other conditions, such as recurrent biliary infections, acquired immunodeficiency syndrome,[2] previous biliary tract surgery, bile duct anomalies, or stone disease, can result in similar lesions and clinical profiles. A significant number of patients have no demonstrable underlying cause. A high index of suspicion clinically combined with biochemical and histologic abnormalities may suggest sclerosing cholangitis;[3] however the presence of specific radiologic abnormalities[4] confirms the diagnosis. Several medical therapies have been suggested for the treatment of patients with sclerosing cholangitis, with little success reported.[5–7] Multiple surgical approaches for the relief of obstruction or cholangitis have been described, but as the number of procedures has increased, less than satisfactory results have been reported.[7–9] Liver transplantation has emerged as the treatment of choice when portal hypertension complicates the patient's course.[10,11] In this report we describe our experience with 178 patients who have undergone treatment for sclerosing cholangitis at the Lahey Clinic Medical Center since 1950.

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### Patients and Methods

A computerized record system was used to identify 178 patients with sclerosing cholangitis seen between 1950 and 1989. The diagnosis was based on clinical, biochemical, and histologic parameters combined with essential radiologic findings of stricture and dilatation of the biliary tree. Vital statistics such as the patient's age when symptoms first occurred, the initial presenting symptoms, and the age at death were recorded.

Symptoms and signs of sclerosing cholangitis (Table 1) included jaundice, pruritus, acholic stools, and cholangitis (fever, chills, and abdominal pain). Portal hypertension

TABLE 1. *Symptoms and Signs of Sclerosing Cholangitis*

| Sign | Primary Sclerosing Cholangitis (% of Patients) | | Sclerosing Cholangitis (% of Patients) |
|---|---|---|---|
| | nonIBD | IBD | Others |
| Jaundice | 76 | 57 | 89 |
| Fever | 77 | 55 | 67 |
| Chills | 79 | 69 | 89 |
| Acholic stools | 86 | 77 | 94 |
| Abdominal pain | 100 | 84 | 89 |
| Cholangitis | 69 | 55 | 89 |
| Ascites | 26 | 32 | 5 |
| Varices | 25 | 30 | 17 |
| Encephalopathy | 28 | 32 | 17 |
| Cirrhosis | 40 | 50 | 11 |

nonIBD, no history of inflammatory bowel disease. IBD, inflammatory bowel disease. Others, iatrogenic injury or stone disease.

was suggested by the presence of ascites, encephalopathy, edema, or documented esophageal or gastric varices. A subgroup of 28 asymptomatic patients was recognized by biochemical abnormalities alone, which prompted cholangiography and the subsequent diagnosis of sclerosing cholangitis.

Abnormal results were found on biochemical tests, which included the standard battery of liver function analyses: bilirubin, serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), alkaline phosphatase, albumin, and coagulation profiles such as prothrombin time and partial thromboplastin time.

Findings of cholangiographic studies were reviewed in all but three patients by one radiologist (FJS) from reports in the records of patients seen before 1978 or films available since that time. These included endoscopic retrograde cholangiopancreatograms (ERCP) in 56 patients and operative cholangiograms in 91 patients, T tube in 22 patients, transhepatic in 5 patients, and intravenous in 1 patient. The predominant sites of involvement were divided into extrahepatic (including bifurcation), intrahepatic, and diffuse intrahepatic/extrahepatic areas.

Pathology reports were available for review in all but 19 patients from liver or bile duct biopsies or from native hepatectomy specimens in those patients undergoing transplantation. Findings consistent with sclerosing cholangitis included pericholangitis, thickened ducts, hyperplasia, and ductal proliferation combined with varying degrees of fibrosis or frank cirrhosis.

Primary sclerosing cholangitis was associated with the presence of coexisting IBD in 88 patients and was confirmed by clinical history, barium studies, and by colonoscopy and biopsy in 33 more recently treated patients. Chronic ulcerative colitis and Crohn's disease were distinguished on the basis of classic radiologic findings or biopsy. When these findings were not typical, the patients were classified as having indeterminate colitis. Types of involvement included pancolitis, left-sided colitis, and small bowel disease. In addition 39 patients who underwent colectomy were assessed for the influence and timing of this procedure on the outcome of treatment for sclerosing cholangitis.

Seventy-two patients with characteristic clinical, biochemical, and cholangiographic findings of primary sclerosing cholangitis were identified who had no history or evidence of inflammatory bowel disease (nonIBD). Another 18 patients (Others) with either well-documented injury to the bile ducts (12 patients) or long-standing recurrent biliary tract stone disease (6 patients) were analyzed as a separate group. The clinical and radiologic characteristics of these patients were indistinguishable from those of sclerosing cholangitis.

The indications for operation, operative reports, and outcomes were evaluated for all patients undergoing surgical intervention, which included 233 biliary procedures, 39 colectomies, 15 portosystemic shunts, 16 orthotopic liver transplantations, and 19 miscellaneous procedures.

The cause of death was documented in all patients not lost to follow-up. The development of bile duct or colon cancer was considered separately and compared between nonIBD and IBD patients. Survival distribution was analyzed using the BMDP1L Kaplan–Meier product-limit method on patients with primary sclerosing cholangitis. Statistical significance of differences between distributions was analyzed according to the BMDP1L log-rank method (Figs. 1 and 2). Univariate survival effects of variables such as the presence of IBD, the effect of colectomy, and the extent of radiologic involvement showed no significant difference on log-rank analysis of stratified Kaplan–Meier distributions. Lack of statistical significance (NS) was defined as two-tailed $p > 0.05$.

Follow-up information was obtained for 151 patients from direct patient contact, referring physicians, or death certificates. Twenty-seven patients were lost to follow-up at a median of 7.5 years (range, 10.2 months to 20 years).

## Results

The nonIBD and IBD groups were predominantly male (98 of 178 patients and 43 of 72 patients, respectively) compared with patients in the Others group (8 of 18 were male). The median age at onset of primary sclerosing cholangitis was 41.6 years (range, 14.9 to 74.5 years) and 46.8 years (range, 28.5 to 77.1 years) in the Others group. Clinical signs and symptoms were similar in all groups, with jaundice, cholangitis, and abdominal pain noted in most patients (Table 1). Complications of portal hypertension and the development of cirrhosis were more frequent in patients with primary sclerosing cholangitis

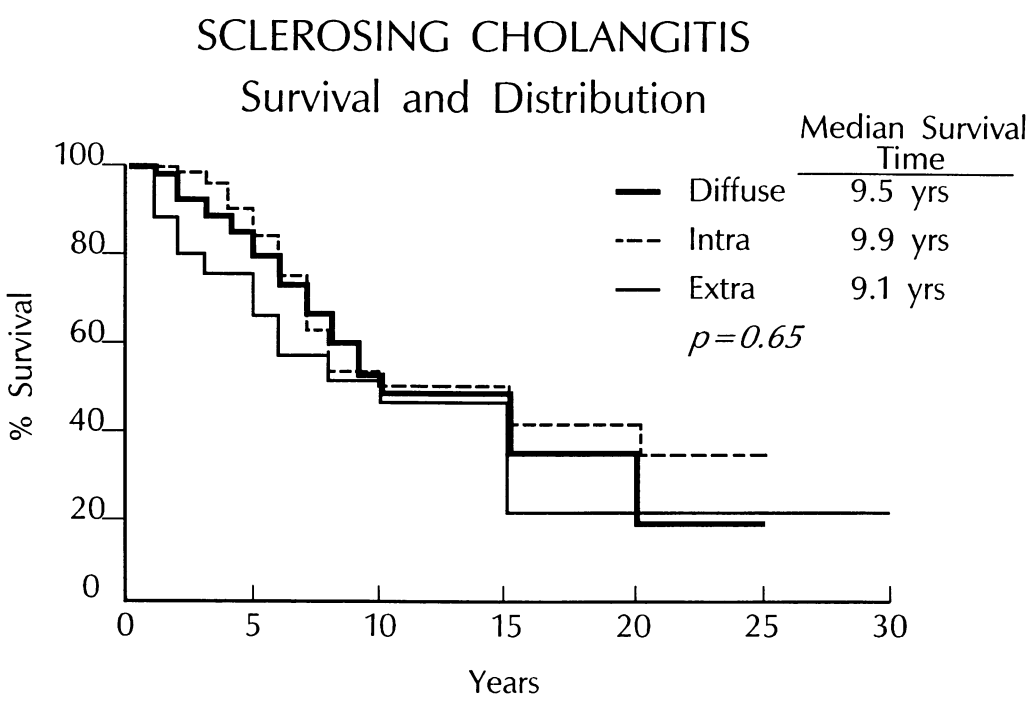


FIG. 1. Patient survival and distribution of sclerosing cholangitis. Significance of differences of distribution was analyzed by the Mantel-Cox log-rank test.

compared with those in the Others group who frequently presented with cholangitis. The bilirubin and alkaline phosphatase levels at diagnosis were elevated in almost all patients.

Surgical treatment included 233 biliary tract operations, 15 portosystemic shunts, 16 orthotopic liver transplantations, 39 colectomies, and 19 unrelated abdominal procedures. The results of the specific biliary tract operations are listed in Table 2. Cholecystectomy and common duct exploration and or dilatation were the initial operative procedures, with jaundice or cholangitis the most common indications. Most patients improved initially and few died. Subsequent surgical procedures included bilioenteric anastomoses, duct dilatation, or insertion of

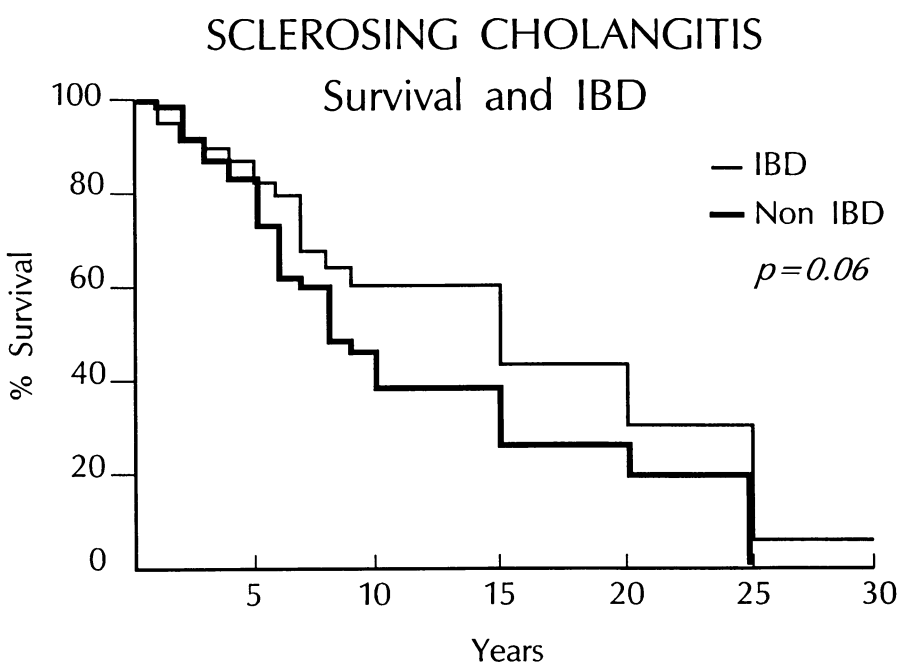


FIG. 2. Survival in patients with sclerosing cholangitis with and without inflammatory bowel disease. Significance of differences of distribution was analyzed by the Mantel-Cox log-rank test.

TABLE 2. *Types and Short-term Results of 233 Biliary Tract Operations*

| | No. of Initial Operations | | No. of Subsequent Operations | |
|---|---|---|---|---|
| | PSC | Others | PSC | Others |
| Cholecystectomy | 32 | 15 | — | — |
| Duct exploration/dilatation | 18 | 3 | 52 | 23 |
| Bilioenteric anastomosis | 5 | — | 36 | 13 |
| Transhepatic stent | 11 | — | 17 | 8 |
| Short-term results | | | | |
| Improved | 75% | 67% | 41% | 39% |
| Unchanged | 24% | 30% | 47% | 46% |
| Died | 1% | 3% | 12% | 15% |

PSC, primary sclerosing cholangitis. Others, iatrogenic injury or stone disease.

transhepatic stents and were associated with less success and more deaths. Fifteen portosystemic shunts were performed in 14 patients, with 13 deaths. Four of these patients died after operation, 3 from recurrent variceal bleeding in 1.5 years, 3 from bleeding during orthotopic liver transplantation up to 4 years later, and 3 from liver failure 1 to 3 years after operation. The cause of death therefore was liver related in all patients.

The overall mortality rate was 58% and appeared to be the highest in the nonIBD group (71%). The median time from diagnosis until death was 10.4 years (range, 1 month to 28 years). Liver failure or sepsis was responsible for more than 50% of the deaths and variceal or postoperative bleeding caused 19% of the deaths. Bile duct cancer was found in 11 patients with primary sclerosing cholangitis (7 nonIBD and 4 IBD) and in no patients in the Others group. Colon cancer was seen in 3 patients with primary sclerosing cholangitis and in no patients in the Others group. Rapid deterioration of otherwise stable disease was noted in 8 patients during a median of 9.4 months (range, 40 days to 15 months).

Radiologic evidence of predominant intrahepatic disease (Table 3) was seen in 49 patients (16 nonIBD, 30 IBD, and 3 Others). These patients underwent a total of 83 operations, with a median survival time of 9.9 years. Diffuse intrahepatic and extrahepatic disease was seen in 76 patients (35 nonIBD, 35 IBD, and 6 Others) who underwent a total of 146 operations, with a median survival time of 9.5 years. Fifty patients with extrahepatic disease only (19 nonIBD, 22 IBD, and 9 Others) underwent 14 anastomoses and 24 dilatations and stentings. Of these patients, 12 died of liver-related causes. The median group survival time was 9.1 years. Survival time therefore was not significantly different among patients with extrahepatic, intrahepatic, or diffuse disease ($p = 0.65$; Fig. 1).

Well-documented IBD was noted in 88 patients. Chronic ulcerative colitis, Crohn's disease, and indeterminate colitis were seen in 70, 15, and 3 patients, respectively. Pancolitis was noted in all 3 patients with indeterminate colitis compared with 60 patients (86%) with chronic ulcerative colitis and 9 (60%) with Crohn's disease. The median duration of IBD was 10.1 years (range, 1 month to 33 years). Differences of 5-year survival rate between patients with IBD and other patients with primary sclerosing cholangitis (Fig. 2) approached significance, with 5-year survival rates of 82.8% and 73.6%, respectively ($p = 0.06$, BMDP1L log-rank). Colectomy was performed in 39 patients for management of severe colitis, except for six asymptomatic patients with rapidly worsening primary sclerosing cholangitis. Of these 6 patients, 4 died of liver-related complications, 1 underwent orthotopic liver transplantation and was doing well at 1.5 years, and 1 had stable disease at 5.6 years follow-up. In 14 patients, colectomy was performed a median of 29.4 months before the diagnosis of primary sclerosing cholangitis (range, 1 month to 15 years) and in 21 patients a median of 7.1 months after the diagnosis (range, 1 month to 14.5 years). Primary sclerosing cholangitis was diagnosed in four patients at the time of colectomy. Symptomatic stomal varices developed in 10 patients, which required local surgical management in 7 patients and included reimplantation of stoma (2 patients) or simple oversewing (5 patients). These procedures failed in all patients. Portosystemic shunt subsequently was performed, which was fatal in all patients. The causes of death included liver failure in five patients and variceal bleeding in one. Another patient died during orthotopic liver transplantation of uncontrollable hemorrhage. The survival of patients who had colectomy was not significantly different when compared with that of other patients with IBD or those with primary sclerosing cholangitis ($p = 0.16$).

The median survival time of patients whose liver biopsies showed fibrosis was 9.4 years (range, 1 to 23 years). For those with the diagnosis of cirrhosis, the median survival time was 4.8 years (range, 4 months to 11 years).

Orthotopic liver transplantation was performed in 16 patients and abandoned in one who was found unexpectedly to have extensive bile duct cancer. The median age at transplantation was 37 years (range, 26 to 62 years) and there was a predominance of women (10 women and 6 men). Inflammatory bowel disease was noted in 10 patients: chronic ulcerative colitis in 8 patients and Crohn's

TABLE 3. *Biliary Tract Disease Distribution and Survival*

| | Extrahepatic | Intrahepatic | Diffuse |
|---|---|---|---|
| No. of patients | 50 | 49 | 76 |
| No. of operations | 54 | 83 | 146 |
| No. of deaths | 28 | 29 | 46 |
| Median survival (years) | 9.1 | 9.9 | 9.5 |

disease in 2. Twelve patients had undergone previous operations, including biliary tract procedures (n = 8), portosystemic shunts (n = 4), or colectomy (n = 3). Six of eight deaths due to orthotopic liver transplantation were in these patients. Three deaths were caused by massive intraoperative hemorrhage.

Abnormal results on liver function tests prompted radiologic evaluation in 28 asymptomatic patients (5 nonIBD, 23 IBD), with a median follow-up of 10.3 years. Surgical intervention was performed in 18 patients and included colectomy in 13 patients (8 of the 13 had colectomy only), portocaval shunt in 1 patient, and 21 biliary tract procedures in 9 patients. Four patients died: one of liver failure, one of variceal bleeding, and two of heart disease.

Of 47 current survivors, 18 patients remain asymptomatic, 16 remain stable despite abnormal findings on laboratory tests and cholangiograms, and 13 have had progression of disease and are being considered for liver transplantation.

## Discussion

This large series allows the evaluation of the management of sclerosing cholangitis over many years. The treatment used has been as varied as the many proposed and yet unknown causes of the disease. The clinical course of sclerosing cholangitis may be indolent for years, with inexorable progression, or can be swift. In either case the ultimate development of complications demands treatment.

The goal of initial treatment is the palliation of cholangitis and obstructive jaundice at an acceptable risk for the patient that permits a reasonably good quality of life. The results in our patients who have had differing distributions of disease indicate that the survival time in patients with diffuse disease is the same as that in patients with extrahepatic disease only, a situation in which relief of obstruction is thought to be mechanically possible.[9] However, in our experience, duct exploration with dilatation and cholecystectomy gives only temporary favorable results in most patients (Table 2). Such treatment can be performed with low risk. With subsequent operative procedures, including operative dilatation, stenting, and anastomosis, the risk increases and the proportion of satisfactory results declines to less than one half.

The role of liver transplantation in the management of sclerosing cholangitis has emerged as a viable choice for these patients. The overall survival rate reported for all adult patients undergoing orthotopic liver transplantation is 75% to 80% at 1 and 2 years.[10] Survival for patients with primary sclerosing cholangitis falls short of this. The Pittsburgh group[11] reported 1- and 2-year actuarial survival rates of 71% and 57%, respectively, after orthotopic liver transplantation and related increased morbidity and mortality rates to previous surgical treatment. It should be noted that 9 of the 18 deaths in this series were due to technical problems associated with the procedures performed before July 1986. Previous biliary tract surgery, portosystemic shunts, and colectomy in a patient with portal hypertension make access to the liver during transplantation difficult and at times impossible. Indeed in our series six of eight deaths in the orthotopic liver transplantation group occurred in patients who had multiple previous operations. It is reasonable to assume that the survival rate for orthotopic liver transplantation in patients with sclerosing cholangitis would approach that for liver transplantation for other indications if the procedure was performed earlier in the course of the disease and in patients who had undergone only one biliary tract procedure without portosystemic shunt or hilar anastomosis.

Percutaneous transhepatic biliary dilatation or occasionally retrograde endoscopic dilatation offer possible solutions to this problem, especially if an anastomosis has been performed earlier in the patient's course and has become subsequently narrowed. Thus, if a patient has undergone initial operative dilatation or anastomosis, has a recurrence of jaundice, and is faced with undergoing a less-than-optimal second operative procedure, percutaneous manipulation would be the preferred treatment before resorting to liver transplantation. Previous reports[12,13] on the usefulness of these techniques have indicated that the results do not equal those for traumatic strictures but that the success rate is sufficient for a trial.

Patients with portal hypertension in whom variceal bleeding, ascites, or encephalopathy develops should undergo orthotopic liver transplantation as an initial procedure. In the past portosystemic shunt was considered in the good-risk patient who presented with variceal bleeding in an attempt to 'buy time' before the need for transplantation. The many complications that occurred after shunt surgery combined with the less-than-optimal results after liver transplantation associated with previous biliary tract surgery have persuaded us to proceed with orthotopic liver transplantation rather than shunt surgery. The main consideration is the avoidance of operation around the liver hilum. If shunts are considered, a distal splenorenal shunt is preferable to a central one. A Sugiura procedure[14] is another alternative.

If a method of prognosticating the course of any given patient were available, one could choose logically between early transplantation and delayed drainage tactics. Wiesner et al.[15] have attempted this through a statistical survival analysis that they characterize as a major first step but not one with great reliability. Cameron et al.[9] have suggested that in patients with rising serum bilirubin values who have had previous intubation, biliary operations, or both, liver biopsy should be performed because the

presence of cirrhosis should prompt the serious consideration of transplantation. Our experience with needle biopsy of the liver leads us to suspect its accuracy and ability to prognosticate. However in this series the prognosis in patients with cirrhosis on open liver biopsy (53 cases with 27 deaths) was 4.8 years survival (range, 4 months to 11 years) compared with 9.4 years (range, 1 to 23 years) with a diagnosis of fibrosis (34 cases with 18 deaths).

As has been found by others,[7,16] colectomy for the treatment of sclerosing cholangitis in patients with IBD has no value. Furthermore it may be followed by the development of stomal varices, a particularly difficult complication. In addition the presence of a stoma can impede exposure for transplantation.

The occurrence of bile duct cancer in this series was 6.2% (11 of 178 patients), which supports the occurrence reported in another series.[7] Sudden deterioration in an otherwise stable patient should be viewed with concern. Routine brushings and cytologic analysis of percutaneous or endoscopic biopsy specimens should be performed. In the patient undergoing orthotopic liver transplantation, distal recipient bile duct sections should be examined by frozen section to avoid unexpected findings at operation.

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### Discussion

DR. HENRY PITT (Baltimore, Maryland): I would like to thank Dr. Martin and her coauthors for the opportunity to review their manuscript and congratulate them on a report of one of the largest series of patients with sclerosing cholangitis.

This analysis is important because it provides data on the natural history of patients seen at one institution over 38 years. In many respects this report is similar to that recently published in *Hepatology* by Weasner and his Mayo Clinic colleagues. They followed 174 patients seen over 15 years and found a median survival rate of 11.9 years, which is similar to the 10.4 years observed in the Lahey Clinic Report.

The Mayo Clinic group emphasized that five factors were important in predicting survival. These factors were age, serum bilirubin, hemoglobin, the presence of inflammatory bowel disease, and the degree of fibrosis or cirrhosis on liver biopsy. These factors were then used to predict whether patients were at low, intermediate, or high risk.

You looked at inflammatory bowel disease and your data differ from those in the Mayo Clinic report. Do you have any information from your data on the effects of age, bilirubin, hemoglobin, or cirrhosis on outcome?

You have also analyzed the cholangiographic distribution of the disease and found no influence on outcome. One problem with this analysis is that your patients were managed in so many different ways over such a long period of time.

In addition you did not analyze separately those patients with primary involvement of their hepatic duct bifurcation. Do you have any data on outcome with respect to that subgroup?

Analysis of your surgical results is also difficult again because so many different procedures were performed over such a prolonged period of time. Nevertheless you have recommended avoidance of biliary tract surgery in favor of transplantation. This recommendation from your analysis is a little hard to understand because 50% of your patients undergoing transplantation did not survive that operation.

In the manuscript you also quote the Pittsburgh transplantation data and suggest that previous biliary tract involvement influences outcome of transplantation. To my knowledge the Pittsburgh data document that previous surgery makes the transplant more difficult but does not affect long-term survivial.

As you know we have recommended resection of the hepatic duct bifurcation and long-term transhepatic stenting for patients with sclerosing cholangitis who have primary involvement of the extrahepatic duct with or without bifurcation involvement and no established cirrhosis.

We continue to perform this operation in carefully selected patients and believe that we actually help them. This slide documents the survival data of 31 of these carefully selected patients operated on at John Hopkins with a median follow-up of 5 years.

According to the Mayo Clinic model these are high-risk patients and should have had 5- and 8-year survival rates of approximately 20%, and 10%, respectively.