Recent Experience with Choledochal Cyst

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This report details an 11-year experience with 17 patients ranging from newborn to 17 years with choledochal cyst. Two distinct groups were noted: an infantile group (mean age: 3 months) with obstructive jaundice identical to biliary atresia and a late onset group (mean age: 9 years) with various combinations of pain, mass, and jaundice. Two patients had cystoduodenostomy performed and both required revision. One of six patients who had Roux-Y cystojejunostomy required revision. All seven patients who had primary cyst excision and two patients who had secondary cyst excision with Roux-Y hepaticojejunostomy have been followed prospectively and have done well. The follow-up period ranges from 1–11 years with an average of 5.8 years. Cyst excision should be performed as a primary or secondary procedure whenever feasible. The rare patients with intrahepatic ductal dilatation (Caroli's disease) are best approached by hepatic lobectomy when possible, and those with choledochocele should be treated by unroofing the cyst as indicated by the anatomy encountered.

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VATER FIRST DESCRIBED a choledochal cyst in 1723.[1] Then in 1852, Douglas reported a case of dilatation of the common bile duct which he believed was congenital in nature.[2] It turned out to be a relatively rare entity as Alonzo-Lej et al. in 1959 reported only two cases and analyzed 94 others, the only cases reported up to that time.[3] He presented a classification of choledochal cyst into three varieties based on the anatomy of the then-reported cases. The most common form, according to the Alonzo-Lej classification, is congenital cystic dilatation of the common bile duct without associated intrahepatic ductal dilatation. His Type II classification was diverticular malformation of the common bile duct and Type III classification was choledochocele associated with ampullary obstruction. With the advent of modern radionuclide imaging studies, percutaneous transhepatic cholangiography (PTC) and endoscopic retrograde cholangiopancreatography (ERCP), at least two additional types of choledochal cyst have been identified including multiple cysts of the intra- or extrahepatic ducts, or both, and a fifth type involving single or multiple intrahepatic cysts, the latter being referred to most commonly as Caroli's disease.[4–6] (Fig. 1).

Choledochal cyst is still relatively rare in the United States and other cultures where the majority of individuals are white. As is the case with biliary atresia, choledochal cyst would appear to be much more common in Orientals, as evidenced by the abundant Japanese literature on this subject. In recent years, the Japanese have reported a large number of successful cases of choledochal cyst treated by cyst excision rather than bypass, and this has influenced workers in the United States as well.[7] This report describes our recent experience with choledochal cyst using these methods.

Clinical Material

Seventeen patients ranging in age from newborn to 17 years at the time symptoms became manifest have been followed up from 1–11 years with an average of 5.8 years. Of the seven patients who had cyst excision, the average follow-up period is 3.5 years. Thirteen of 17 patients were female, which is consistent with the usually reported female preponderance of this malformation. Two patients were black and the remainder were white. The seven patients who had primary cyst excision performed have been studied prospectively and have been compared with the remaining 10 patients who had other forms of treatment studied retrospectively.

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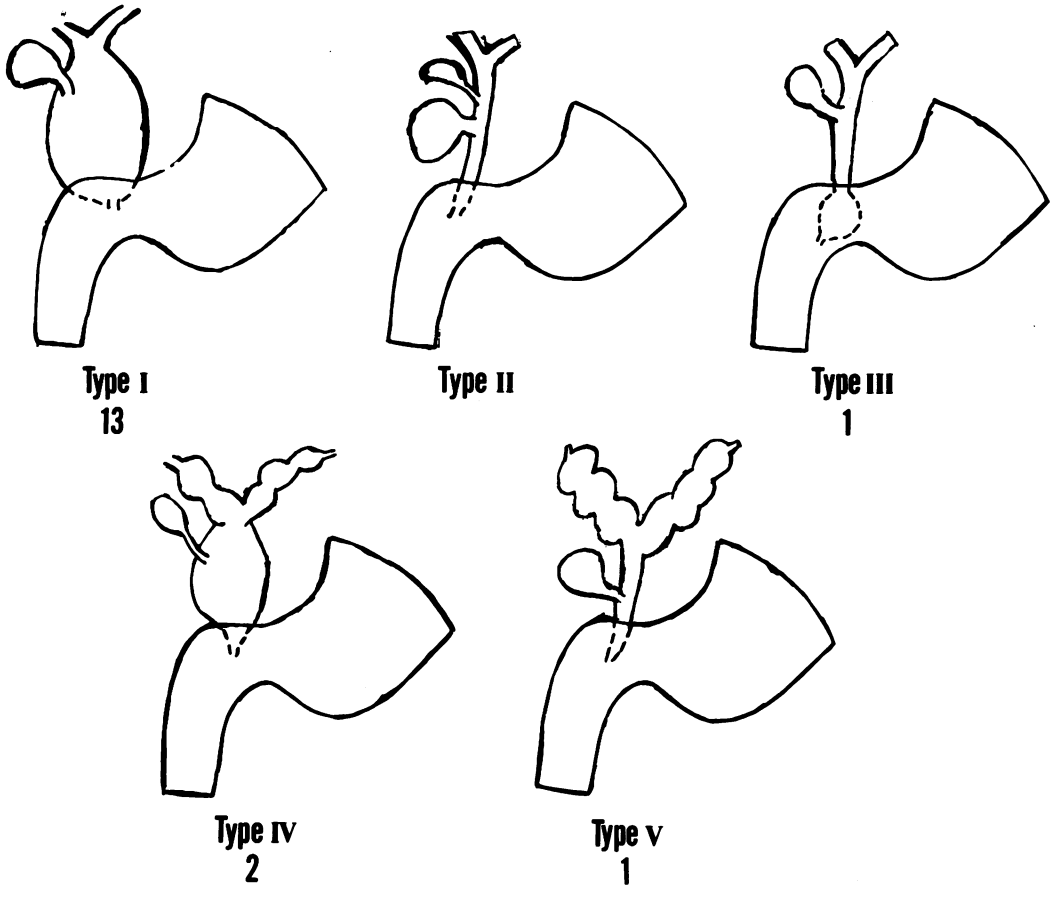

FIG. 1. Variations of these five types of choledochal cysts have been defined by modern imaging studies. Note that Type I, cystic dilatation of the common duct, either with complete or incomplete obstruction, is the most common anatomy encountered.

Signs, Symptoms, and Clinical Findings

Alonzo-Lej and co-workers described what they believed was a classic triad of pain, mass, and jaundice characteristic of patients with choledochal cysts.[3] As experience has been gained through the years, other forms of presentation have been recognized. However, the patients have generally fallen into one of two distinctive groups, late onset and infantile. Only three of nine so-called late onset patients ranging in age from 2–17 years had the triad of pain, mass, and jaundice. Two others had right upper quadrant and midepigastric intermittent abdominal pain associated with mild intermittent jaundice, and three patients had vague digestive complaints and mild upper abdominal discomfort associated with a palpable right upper quadrant mass but no jaundice. The ninth patient who presented after 2 years of age had only very mild intermittent jaundice.

The second younger group of eight patients presented from newborn to 9 months of age. Seven of eight patients had obstructive jaundice, acholic stools, and hepatomegaly and were presumed to have biliary atresia. The eighth patient, one in whom a prenatal ultrasound diagnosis was made, had a mass and mild jaundice. No other signs or symptoms were present in this infantile group.

No patients had any significant associated congenital malformation. Although two patients are known to have splenomegaly on follow-up examination and are presumed to have some degree of biliary cirrhosis and portal hypertension, this is as yet clinically insignificant.

Diagnosis

Laboratory Studies

At presentation, 15 of 17 patients had either constant or intermittent elevations in serum bilirubin levels, primarily conjugated in nature, as well as elevations in alkaline phosphatase levels characteristic of obstructive jaundice. The two remaining patients did not have any elevation of bilirubin or alkaline phosphatase levels. None of the patients had chemical findings indicative of biliary cirrhosis, and none were anemic or had elevations in white blood cell counts. A 17-year-old patient with mild intermittent abdominal pain and a recent episode of jaundice that was clearing when she was seen also had a slight elevation in serum amylase level.

Imaging Studies

A variety of imaging studies were helpful in the management of this group of patients. Although upper gas-

trointestinal x-ray studies with contrast material were believed to be helpful in the past, none of our patients had such studies that we believe are unreliable.

Abdominal ultrasound studies were used for screening purposes in all patients over the last 7 years and were found to be diagnostic in every instance. The diagnosis was made on prenatal ultrasound study in two patients even though the finding was disregarded in one of these two patients and only later appreciated in retrospect when the infant presented with jaundice at 6 weeks of age.[8] Once the diagnosis was suspected on ultrasonography, ^{99}Tc DISIDA scintigraphy was performed although PTC or ERCP were performed first in four older patients. In every patient, operative cholangiography was performed at operation to define the precise anatomic nature of the malformation.

In those occasional instances where imaging studies have been believed to be indicated after operation, ultrasonography and DISIDA scintigraphy have been sufficient to define anatomy and patterns of excretion.

Computerized tomography (CT) was also used in two instances although it probably did not add anything to the information gained on ultrasound. In one instance, CT was combined with intravenous cholangiography since intrapancreatic disease was suspected although not proven.

Pathology

Fifteen of 17 patients had cystic dilatation of the common duct, and in seven patients the obstruction was apparently complete at the level of the distal portion of the common bile duct. Two of 15 patients with dilatation of the extrahepatic biliary tree also had some degree of associated intrahepatic ductal dilatation. One patient had a choledochocele associated with pain and mild pancreatitis, and one patient had cystic dilatation of the left intrahepatic ducts associated with manifestations of bile stasis and infection. In those patients with cystic dilatation of the extrahepatic biliary tree, an apparent "common channel" was apparent in seven instances on cholangiography as described by Ono et al.[9]

Biopsy of the cyst wall or cyst excision was performed in 16 patients and the typical pattern was a thick-walled cyst consisting of dense connective tissue and some smooth muscle. Usually some degree of inflammatory reaction was present within the wall and no mucosal lining was present, although small patchy areas of columnar epithelium were noted in some instances as well as microscopic bile ducts within the wall of the cyst. In the patient with a choledochocele, the cyst was lined with duodenal mucosa and there was associated obstruction of the common bile and pancreatic ducts which entered separately into the wall of the cyst.

Culture of the cyst contents was positive in only one instance when diphtheroids grew out. In addition, cyst fluid was analyzed in two neonates, which was found to contain trypsin but no amylase in both instances.

TABLE 1. *Choledochal Cyst: Operations**

Procedure	No. of Operations	No. Revised
Cystoduodenostomy	2	2
Cystojejunostomy	6	1
Primary cyst excision	7	—
Lobectomy	1	—
Excision of choledochocele	1	—

* 16 patients had cholecystectomy.

Liver biopsies were performed in all 17 patients. Evidence of mild periportal fibrosis was noted in two patients, but otherwise the findings were nonspecific and only indicative of partial or complete biliary obstruction.

The gallbladder was normal in all patients and neither the gallbladder itself nor the cystic duct were dilated in any of these patients.

Operative Treatment (Table 1)

Over 11 years, three surgical approaches have been used for the treatment of 15 patients with cystic dilatation of the extrahepatic biliary tree. Two patients treated early in the series had cyst duodenostomy performed. One of these patients, a 16-year-old girl, had revision of a previous cyst duodenal anastomosis to a Roux-Y cystojejunostomy after having three separate bouts of obstructive jaundice associated with cholangitis. Another patient, age 17 years, had a similar problem after cystoduodenostomy performed some years earlier, and she was able to have revision by cyst excision and Roux-Y hepaticojejunostomy. Six patients had primary Roux-Y cystojejunostomies, and one of these was revised to a cyst excision with hepaticojejunostomy because of jaundice associated with stenosis of the original anastomosis. Seven patients have been treated by primary excision of the choledochal cyst, three by intramural resection as described by Lilly[10] and Todani et al.[6] and four by total cyst resection. The patient with cystic dilatation of the left intrahepatic ducts was treated by left hepatic lobectomy, and another patient with a choledochocele was treated by unroofing of the choledochocele and sphincteroplasty of the common bile and pancreatic ducts, both of which were stenotic.

Cholecystectomy was performed in all patients except for the patient with Caroli's disease. No abdominal drains were used after operation except in the patient who had hepatic lobectomy performed.

Results of Treatment and Follow-up (Table 2)

In this 11-year period, there have been no deaths and no long-term complications. Both patients who had cyst

TABLE 2. *Choledochal Cyst; Operative Follow-up 1–11 Years (Mean: 5.8 Years)**

Procedure	No. of Patients	Result	Mean Follow-up (years)
Cystojejunostomy	6	Well	7.1
Cyst excision	9	Well†	3.5
Lobectomy	1	Well	6
Excision of choledochocele	1	Well	4

* No mortality occurred in this series.
† Biliary cirrhosis, 2 patients.

duodenostomies performed had recurrence of cholangitis and jaundice even though the symptoms were very mild, and both required revision. Follow-up thus far has been favorable in both patients, one of whom was revised to cystojejunostomy, whereas the other had cyst resection. Of the six patients who had primary choledochal Roux-Y cystojejunostomies, only one had stenosis and required revision, and all six are doing well up to 10 years after operation. Hence, we now have a group of six patients who have ended up with cystojejunostomies and all are doing well at this time. The seven patients who have had primary total cyst excision and the two patients who were revised to this form of treatment are all doing well up to 7 years after operation with no long-term complications recognized thus far. This latter group of patients has been followed closely prospectively and even the two patients who are believed to have mild biliary cirrhosis are doing well and have had no progression of disease.

Comments

In general, patients with Type I choledochal cyst involving varying degrees of cystic dilatation of the extrahepatic biliary tree, have presented in two ways. One way has been an infantile group of patients generally 3 months of age or younger who had obstructive jaundice and a clinical picture indistinguishable from that of biliary atresia. Several of these patients were believed to have biliary atresia initially but with imaging studies were accurately determined to have choledochal cysts. It is of interest that one of these infantile patients had a choledochal cyst prenatally and even though the serum bilirubin level was 5.6 mg/dL at age 11 days, the conjugated fraction was only 0.6 mg/dL and alkaline phosphatase level was normal. ^{99}Tc DISIDA scintigraphy demonstrated a choledochal cyst with delayed visualization of the small bowel at approximately 2.5 hours, but at 24 hours complete emptying had occurred into the duodenum. This study was performed at 4 days of age. At age 11 days, an operative cholangiogram demonstrated the choledochal cyst with prompt emptying into the duodenum. It is tempting to speculate that this infant may have had complete obstruction if she had been allowed to go for a longer period. This is suggested by the other patient in whom a prenatal ultrasound study demonstrating a choledochal cyst was misinterpreted, and that infant had a totally obstructed biliary tree at age 6 weeks. In this infant, jaundice was not noted until approximately 3 weeks of age. More infants shown to have choledochal cyst *in utero* will need to be followed up to confirm this impression that the extent of obstruction may progress over time in some patients. It does appear that choledochal cysts do not appear until the last trimester of pregnancy.[8]

The second group of patients, who have had a different mode of clinical presentation, have presented after age 2 years. In these patients, jaundice has been intermittent and less severe than in the infantile patient group, and additional findings, including upper abdominal pain and the delineation of a right upper quadrant mass, have appeared. Abdominal pain has characteristically been mild and lacking specificity so that many of these patients have symptoms for years before being recognized as having a significant clinical problem.

The exact cause of choledochal cyst is unknown. Although the female preponderance of three or four to one over males with this disorder is highly suggestive of a sex-linked genetic association, there are no familial reports or other studies that have implicated genetic factors further. Additionally, the striking lack of associated congenital malformations in this series and most other series reported strongly suggests that whatever the cause, it occurs either late in gestation or after birth.

Initially, choledochal cysts were believed to result from obstructive factors, but this did not account for the large number of cases that are not symptomatic early and in which obstruction does not appear to be present at the level of the ampulla of Vater. It was then believed by Alonzo-Lej and others that the defect was the result of a primary weakness of the bile duct itself, and there was some precedent for this in the form of primary ectasia of portions of the gastrointestinal tract.[3] Around the time that embryologic studies were defining the solid and canalization stages of intestinal development, Yotsuyanagi suggested that an inequality of proliferation of epithelial cells with overproliferation of the proximal end of the bile duct as compared with the distal portion left an abnormally dilated proximal end when the cannalization stage of bile duct development occurred.[11] However, in the last 5 years or so, ERCP has provided information from a large number of cases in which the anatomy of the biliary and pancreatic ductal systems has been visualized. As a result, Babbitt[12] and Todani et al.[13] have suggested that patients with choledochal cysts have anomalous entry of the pancreatic ducts into the bile duct system in such a fashion that there is a narrowed distal portion going into the duodenum, providing a true common channel situ-

ation resulting in easy reflux of pancreatic juice into the bile duct due to absence of the sphincter of Oddi around the union. In recent laboratory studies, Iwai and co-workers demonstrated no sphincter function at the anomalous pancreaticobiliary junction, which tends to confirm the common channel theory.[14] Furthermore, Kato and coworkers have suggested that reflux of pancreatic enzymes in the fetus is responsible for destruction of a part of the wall of the bile duct.[15] If this is the case, it is probably the result of digestion by trypsin since analysis of the cyst contents in newborn patients in this series revealed that trypsin, but no amylase, was present in the newborn. Type II or the diverticular form of cyst may represent some form of prenatal rupture of the common duct. A reproducible experimental model will probably need to be devised before the precise cause can be determined.

The incidence of choledochal cyst is in the vicinity of one in 2,000,000 live births.[8] However, the incidence is much more frequent in the Orient than in Western populations, and the same is true of biliary atresia, which raises the additional question of the possibility of an infectious, perhaps viral, cause.

On the basis of the experience gained from managing this group of 17 patients, the diagnostic approach to patients of this sort is straightforward. In the infantile group of patients with obstructive jaundice verified by blood chemistry determinations, ultrasound evaluation may be all that is required in the way of preoperative imaging studies. In some instances, ultrasound may not demonstrate the malformation, as when there is intestinal distention, and under these circumstances, radionuclide scanning with DISIDA may be helpful. It is rare that PTC or ERCP will be required in the infantile patient group. On the other hand, in the group of patients who present after 2 years of age, where jaundice is intermittent and pain is a prominent feature of presentation, additional approaches may be helpful. Ultrasound is still the best screening study under these circumstances, but DISIDA excretion studies, PTC, or ERCP may be very helpful. At times, intravenous cholangiography imaged by CT may also be diagnostic as used in this series of patients.

Ultrasound would appear to be an accurate prenatal diagnostic study as shown by two patients in this series. There does not appear to be any need to deliver such patients early but rather to allow them to come to term since the liver appears to be protected *in utero*. Once such infants have been delivered, ultrasound is confirmatory and operation may be done electively once the infant is stable. In one of our patients, operation was performed on the eleventh day of life and obstructive jaundice was not a problem as yet in that infant. The other infant who had a diagnostic prenatal ultrasound that was misinterpreted initially did have obstructive jaundice at age 6 weeks. This is the only information currently available regarding how long it may take for obstructive jaundice to develop in this infantile group of patients.

Generally, postoperative imaging studies have not been performed in patients who have been doing well without evidence of jaundice or cholangitis. In those patients who have been studied after choledochal cystojejunostomy, the dilated choledochal cysts have generally diminished in size although not to normal size. We have found that DISIDA excretion studies have been the best way to follow patients who have had either cystojejunostomy or cyst excision. In all instances thus far, the few patients who have been studied have shown prompt excretion.

With regard to treatment, previous studies performed by one of us have shown that choledochal cystoduodenostomy is not an effective method of providing permanent biliary drainage.[16] Furthermore, symptoms and signs suggestive of failure are subtle and difficult to determine, so that often many years pass before it is realized that revision of cystoduodenostomy is needed. This is supported by the fact that both patients who had cystoduodenostomy in this series required revision. In 1978, Spitz reported excellent results on long-term follow-up in over two thirds of his patients who had Roux-Y cystojejunostomy.[17] This also turns out to be the case in the six patients in this series who had cystojejunostomies performed since only one of these patients required revision because of subsequent stenosis at the anastomosis. Continuing long-term follow-up in this particular group of patients will be required, however, before we can be certain that these patients with cystojejunostomies will have a good lasting result. Additionally, the continuing presence of the choledochal cyst in this group of patients is somewhat of concern because of the potential for malignant transformation within the wall of the cyst.[6]

In the last 7 years, we have preferred to perform cyst excision as first reported by McWhorter in 1924,[18] but it was not done very often because of excessive mortality associated with this procedure before the 1970 report of Kasai et al.[7] of 14 patients who had cyst excision. In the group of seven patients followed prospectively who had primary cyst excision and two additional patients who had secondary cyst excision after cystoduodenostomy and cystojejunostomy, respectively, the long-term results thus far have been excellent with no complications encountered, although these patients will also require longer follow-up.

The technique of cyst excision has varied according to the age of the patient and the degree of fibrosis and inflammatory reaction evident. In the very young patient with little or no periductal adherence to surrounding tissues, complete excision of the cyst has been performed with simple closure of the distal end and construction of a Roux-Y hepaticojejunostomy. On the other hand, in older patients with a thicker cyst wall and adherence to

surrounding vasculature, the dissection has been performed in an intramural fashion as described by Lilly[10] and Todani et al.[6] with the plane being entered at the level of the cystic duct entry into the wall leaving the external portion of the cyst wall in place. Then, the distal part of the duct has been closed and a similar hepaticojejunostomy performed. Although it is presumed that the concern about malignant transformation of the cyst wall is eliminated by cyst excision, one must acknowledge that malignancy might still be a possibility at the level of the cyst stump at the hepatic hilus. The patients in this series and reports from around the world seem to suggest that the incidence of cholangitis and anastomotic stricture after excision and hepaticojejunostomy is exceedingly low when compared with the other forms of surgical treatment.[19-21]

Whether Roux-Y hepaticojejunostomy or hepaticoduodenostomy is the best approach is unknown since hepaticoduodenostomy as reported by Todani et al. has not yet stood the test of time.[22] Additionally, there would be concern with the latter method of drainage that duodenobiliary reflux might cause late inflammatory complications.

The less common forms of choledochal cyst are best approached depending on the anatomy involved. In those rare instances of Type II choledochal cyst involving diverticular malformations, excision of the diverticulum and primary repair of the duct is sometimes possible as performed in a previous case by one of us (JAO). Others have reported excision of the ductal system including the diverticulum with Roux-Y hepaticojejunostomy. The treatment of the Caroli's form of the disorder involving intrahepatic ductal dilatation is a difficult problem to solve but selective hepatic lobectomy is probably the most effective approach when it is possible to perform. Unfortunately, many patients with Caroli's disease cannot be treated in this fashion and must be treated by long-term antibiotic therapy or by transhepatic drainage. In those patients who have choledochoceles, the best approach is unroofing of the intraduodenal portion, oversewing the edges. However, it is important in all of these cases to determine whether sphincteroplasty of the pancreatic and bile ducts will be needed in addition because of stenosis associated with this malformation.

Since patients with choledochal cysts have the potential for stasis within the biliary tree if the cyst is not excised, cholecystectomy is best performed in all cases in which cyst excision is not performed. Otherwise, late stone formation and complications will inevitably occur.

Another advantage of cyst excision is that the procedure requires thorough exploration of the ductal system. Operative cholangiography should always be performed since it is well known that in the infantile form of this malformation, intrahepatic biliary atresia may coexist with cystic malformation of the extrahepatic biliary tree. If the patency of the intrahepatic ducts is not determined during operation in such infants, the diagnosis may be missed. These patients should be treated with the conventional approach to biliary atresia by the Kasai operation. Although we did not have any patients with biliary atresia in this series, two patients do have mild biliary cirrhosis that is as yet asymptomatic. Long-term follow-up will still be needed to be certain that these results hold up and that the current recommendations remain valid. Many patients do not appear to have difficulty until as long as 10 or more years after operation. Currently, however, we are encouraged by the results with cyst excision and Roux-Y hepaticojejunostomy and believe that it is the preferred approach to management of patients with choledochal cyst regardless of age. In patients who have had previous cystojejunostomies requiring revision, cyst excision is also the preferred approach rather than revision of the cystojejunostomy.

Portal hypertension has not been a prominent feature in this series, although the two patients with mild biliary cirrhosis are presumed to have mild associated portal hypertension on the basis of splenomegaly. However, this has been asymptomatic thus far. The incidence of portal hypertension in the survey of choledochal cyst by the Surgical Section of the American Academy of Pediatrics reported by Kim in 1981 was 12%.[19]

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Discussion

DR. FRANK G. MOODY (Houston, Texas): Dr. O'Neill and his colleagues have provided for us a very timely presentation of an unusual condition in childhood and also in adulthood.

I have had experience with 12 patients over the age of 15 and up to the age of 34. There were a few differences that I believe should be emphasized, and I will ask Dr. O'Neill to comment.

Clearly, ultrasound is the way to detect a choledochal cyst. The computed tomography (CT) scan adds little to the diagnosis, but in some of the older patients with sepsis, a CT scan is very important in terms of picking up infection within the liver.

The biliary scan is helpful in terms of identifying the dynamics of the emptying of the cyst and in detecting intrahepatic cysts, which are common in the adult. Eighty per cent of adults have a deformity of the intrahepatic biliary tree.

I would advise against only removing the cyst lining. The cyst should be excised by a meticulous technique because there are small bile ducts that come into the cyst high up and usually posterior. If one first takes down the gallbladder, then gets behind the cyst, freeing up the portal vein and the hepatic artery separately, it is simple to transect the cyst and then to complete the dissection proximally. In the adult, one should take out as much of the cyst as possible, and that is why it is very important to have an endoscopic retrograde cholangiopancreatography (ERCP).

You must know the relationship between the pancreatic duct and the cyst because in the adult it is amazing how it seems to creep up into the cyst itself. In this situation then, one must start the dissection distally and then fold the cyst in on itself, examining the mucosa very carefully to be sure that there are no excrescences.

The incidence of cancer in these cysts in the adult is very high in the Japanese series and is up to 20% in some reported series. Therefore, at the time of excising the cyst one should look up in the intrahepatic biliary tree with a flexible choledochoscope. I wonder if you have had experience with that in your older children.

Finally, in your manuscript, Dr. O'Neill, you were a little bit enthusiastic about possibly putting a Roux limb on to the cyst. Does it not concern you that maybe as a function of time these individuals will develop cancer in this residual cyst that is left in place?

DR. LESTER MARTIN (Cincinnati, Ohio): I appreciate the opportunity to discuss Dr. O'Neill's paper. Most surgical residents gain considerable experience in biliary disease, but on the basis of my own experience participating in oral examinations for the American Board of Surgery, the subject of congenital biliary tract anomalies probably deserves greater emphasis in our teaching programs. Choledochal cysts as well as other anomalies occasionally present for the first time in adult life; one of our patients with a choledochal cyst was 32 years old at the time of his first symptoms.

I have been particularly impressed with the minimal degree of gross and microscopic liver disease in the patients with obstructive disease from choledochal cysts, and this simply underscores the good prognosis for the condition. I try to emphasize in our resident teaching, that for these patients, the operation must provide free drainage of bile and not predispose the patient to ascending cholangitis. The operation must be designed to last for 75 years since these patients have a normal life expectancy.

The evolution of the surgical treatment that Dr. O'Neill outlined has evolved over the past 30 years, beginning with simple cystoduodenostomy, then Roux-en-Y drainage, then excision of the cyst, and now intramural or mucosal excision of the cyst with Roux-en-Y choledochojejunostomy. The Japanese deserve a great deal of credit for the development of this operation since the condition is much more common in their population.

For the rare intrahepatic cyst that cannot be excised because of the many huge bile ducts that enter into the cyst, we prefer a simple Roux-en-Y drainage.

One of our patients had portal hypertension and bleeding esophageal varices caused by obstruction to the portal vein by the choledochal cyst. Evacuation of the choledochal cyst provided sufficient decompression of the portal system so that a shunt operation was not required.

My question to Dr. O'Neill relates to the many diagnostic modalities available to us today, including endoscopic retrograde cholangiopancreatography, hydascan, transhepatic cholangiogram, intravenous cholangiogram, computed tomography, and magnetic resonance imaging (which we used to call the NMR). How many of these studies do you advise for a patient with a choledochal cyst?

DR. MARTIN A. ADSON (Rochester, Minnesota): Dr. O'Neill, I have been drawn to the podium because you have said little about cancer and other things that may complicate this disease. Some of my observations may be redundant, but I am compelled to bring up the questions that both Dr. Moody and Dr. Martin have raised.

I believe that this is the same disease in the child that it is in the adult. It is congenitally the same anomaly, but it does present differently at different times in life. Two years ago, Dr. David Nagorney studied our institutional experience with 29 adult patients (16+ years of age) treated at our institution.

(Slide) The incidence of *pancreatitis* in this group of 29 adult patients was 34% (9 patients). Interestingly, pancreatitis did not develop until *after* the patients had biliary-intestinal drainage done without excision of the cyst.

(Slide) The next slide shows the cause of pancreatitis: a long common channel downstream of abnormal conjeries of biliary and pancreatic ducts, three or more cm away from the ampulla. Nine of ten patients whom we studied radiographically had this abnormality.

I am convinced that, despite the cost of ERCP, there is advantage in doing this study before operation. On two occasions, I have been shown cancers in the dependent portion of the cyst 1 cm from its junction with the pancreatic duct. Then it is nice to know where that junction is because the junction can occur the other way around with the pancreatic duct inserting as a "T" on the bile duct, and it is necessary to know how far to go with safe excision of the cyst.

The next slide shows the incidence of malignancy to be 28%. In this group of 29 patients, there were eight such patients, and cancer has developed in another patient since publication of that paper. Unfortunately, cancer cannot always be prevented by taking out the cyst, because cancer developed in half of our patients intrahepatically upstream of the choledochal cyst.

The need for long-term follow-up is obvious; not just long follow-up but extended observation. The mean age of the adult patients that we have studied in whom cancer developed in association with choledochal cyst was 30 years. This means that the patients who have been treated