Congenital Segmental Cystic Dilatation of the Biliary Ductal System

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SEGMENTAL CYSTIC DILATATION of the biliary ductal system leads to bile stasis, predisposes to bacterial growth and by compression of parenchymal cells, impairs liver function. When these lesions are due to a developmental defect in the duct wall they are referred to as "congenital segmental cystic dilatation of the ductal system" (Fig. 1). This term is applicable to both the intra- and extrahepatic bile ducts. Historical reports of patients with this condition, extending back to 1818[14] and long dormant, have received increasing attention since Caroli in 1958[5] described in detail the distressing course of this condition. We agree with others[3,4,12,15] that the defects that are the basis for segmental dilatation are congenital.

One hundred and fifty-four years ago, in 1818, Todd[14] reported from the Dublin Hospital a patient with a diffuse cystic dilatation of the common duct. In 1904 (86 years later) Mayo-Robson[9] called attention to a specimen in the Hunterian Museum in London. It was described by him as "a large tumor of the liver constituted by dilated hepatic ducts which form a series of cysts within the substance of the liver itself." . . . . "There was no obstruction in the main channel." Yotuyanagi[18] in 1936 recorded a patient with a large cystic dilatation of the common duct and an intrahepatic cyst of the right duct. He described the cyst as "the size of an apple" and microscopically as having a single layer of epithelium inside the cyst wall. Another unique and significant case report is that of McWhorter reported in 1924[11] and again in 1939.[10] The patient, a woman of 49, underwent resection of a large dilated common duct. A cholecystectomy and choledocho-duodenostomy were performed. She died at age 69. Autopsy revealed dilatation of major intrahepatic ducts with "cystic degeneration." There was no evidence of stricture of the hepatoduodenostomy anastomotic stoma. Perhaps the most detailed description of congenital segmental dilatation of the intrahepatic ductal system to be published prior to 1958 was the Medical Thesis of Roger Jacques Le Naour[8] in Paris (1941). Tsuchida and Ishida[15] of the University of Tokyo have suggested that more than 500 cases of biliary ductal cysts of all kinds have been recorded since Todd's report in 1818. They reviewed a series of nine patients in Japan with dilatation of the intrahepatic bile ducts associated with congenital cystic dilatation of the common bile duct. They also collected from the literature 21 additional patients with segmental dilatation and cyst formation of the intrahepatic ducts with cystic dilatation of the common duct.

Numerous theories have been advanced as to the origin and nature of these defects commonly held to be congenital. Both intra- and extrahepatic portions of the biliary ductal system are subject to hypoplasia in their embryological development. Incomplete or faulty development may produce a wide range of end results. At one extreme is the complete failure of the cord of cells in the early embryological development to form a lumen.[13] According to Langman[7] there can be a lack of any part of the biliary ducts within the liver or extending from the liver to the duodenum. Gross failure in embryological development is sometimes manifested soon after birth. Segmental occurrence of this, if it be intrahepatic, may well be compensated for if only a portion, perhaps a third, of the liver is without drainage. Turnberg and associates[16] have reported on the increased bile output from a congenital intrahepatic cyst.

In congenital segmental dilatation of the ductal system

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the defect in embryological development appears to be related to the supportive structure of the duct wall (Fig. 2). This wall is considered to be made up of four parts or layers, the first, the innermost, is mucosa; the second, the fibromuscular; the third, a subserous coat composed of dense bands of elastic fibers closely interwoven; and the fourth, the serous coat, or peritoneum. Because of the nature of the fibromuscular and subserous layers they are regarded as the supporting ones. If they are absent or lacking in substance the other components are readily distended. The pressure within the biliary ductal system is known to vary, being lowest in the interdigestive period. It increases when gastric contents pass into the duodenum and the gallbladder empties its contents into the common duct and thence into the duodenum. Intrahepatic cystic dilatations, as well as dilatations of a part or the entire common duct may develop as a result of the pressure changes in those patients who lack integrity in segments of the ductal wall.

The demonstration of the presence of the subserous layer in such dilatations or cysts is difficult because of the attenuation by distention alone. Nevertheless the segmental distribution of cysts when they are multiple and the infrequency of a single duct and its tributaries being involved strongly favors this explanation. It tends to minimize the nerve and vascular elements as etiological factors in their genesis. On the basis that these defects are present at birth[17] one can well justify assuming that some additional factor or factors are usually superimposed that lead to its symptoms or manifestations. The numerous factors that are superimposed have a bearing on the flow of bile into the duodenum. Viscosity of the bile and the mechanical resistance of the distal common duct and its sphincter mechanism during the active phase of digestion may produce a significant

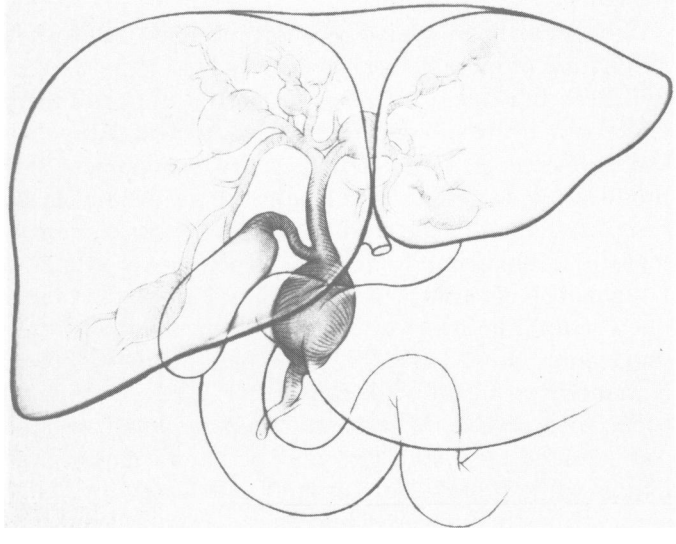

FIG. 1. Schematic drawing of congenital segmental cystic dilatation of the intra- and extrahepatic biliary ductal system.

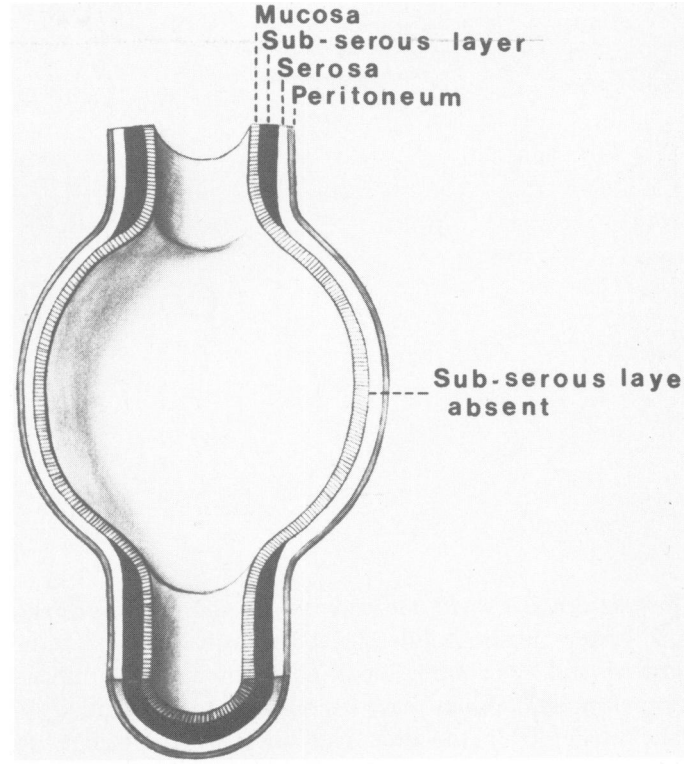

FIG. 2. Schematic drawing indicating structure of bile duct. Most anatomical descriptions of the wall of the ductal system include four components that are identified in the normal common duct. These are: mucosa; subserous layer composed of fibromuscular and elastic fiber elements. Microscopic examination of cyst wall of surgical specimens reveals an absence of subserous or fibromucular layer.

increase in intraductal pressure. Those segments of the ductal system that lack the integrity of their supportive structure either because of deficient fibromuscular and subserous layers or abnormal innervation become dilated and appear cystic when subjected to increased pressure. The duration of the increased intraductal pressure is probably short and occurs during the period bile is passing through the choledochoduodenal junction into the duodenum. Because pressure elevations are of short duration jaundice of the obstructive type does not develop. However, the repetition of the elevation of the ductal pressure several times a day leads to the full exploitation of inadequate segments to become cyst-like in structure.

Clinical Material

From 1941 through 1972, 15 patients with congenital segmental dilatation of the biliary ductal system have been observed at The New York Hospital-Cornell Medical Center (Table 1). There were 12 females and 3 males. The age range was from 4 months to 63 years. Segmental dilatation of a part or the entire extrahepatic bile ducts was present in every patient. Concomitant involvement of the intrahepatic biliary tree occurred in 10 patients. In three patients, insufficient data precluded a decision regarding

TABLE 1. *Congenital Segmental Cystic Dilatation of the Biliary Ductal System*
The New York Hospital-Cornell Medical Center
1941–1972

Patient No.	Age (yrs.)	Duration of Symptoms	Operation	Result	Follow-up
1.	4 mos.	3¾ mos.	Expl. lap & I & D liver abscess	Death	Post-op
2.	4	4 yrs.	Cysto-duodenostomy	Death	9 yrs.
3.	9	3 mos.	Cysto-jejunostomy (Roux-Y)	Well	17 yrs.
4.	20	5 days	None	Death	3 days (in hosp.)
5.	7½ mos.	7½ mos.	Cysto-jejunostomy	Death	6 weeks
6.	38	3 yrs.	Cysto-duodenostomy	Unknown	Lost
7.	63	3 yrs.	Excision & Choledocho-jejunostomy, Roux-Y	Death	23 mos. p.o. Ca
8.	38	19 mos.	Excision	Well	6 yrs.
9.	28	Asymptomatic	Excision	Well	5 yrs.
10.	49	Asymptomatic	Cholecystectomy & Choledocholithotomy	Unknown	Lost
11.	43	2½ yrs.	Cysto-duodenostomy	Well	2 yrs.
12.	20	15 yrs.	Hepatico-jejunostomy (Longmire)	Well	2 yrs.
13.	24	5+ yrs.	Cysto-jejunostomy (Roux-Y)	Well	9 mos.
14.	17	2 mos.	Cysto-jejunostomy (Roux-Y)	Well	9 mos.
15.	61	3 yrs.	Choledocholithotomy	Well	6 mos.

dilatation of the intrahepatic bile ducts. In the remaining two patients, the intrahepatic ducts were apparently normal.

The duration of symptoms prior to evaluation that led to the diagnosis ranged from 5 days in an infant to 15 years in a young woman, age 20. In this small group of patients there was no discernible specific pattern of symptoms. Four of the patients were under 10 years of age. In children, symptoms suggestive of biliary tract disease, particularly when associated with jaundice, should suggest the possibility of congenital segment dilatation of the biliary tree. In adults, the symptoms associated with this condition are usually indistinguishable from those reported by patients with calculous biliary tract disease. In two patients included in this discussion, the cystic lesions in the biliary tree were asymptomatic. A large cyst involving the extrahepatic bile ducts was palpated in the course of a cesarean section in a 28-year old woman. Elective excision of the cyst was accomplished 1 year later. The other patient, age 49, underwent cholecystectomy and chole-

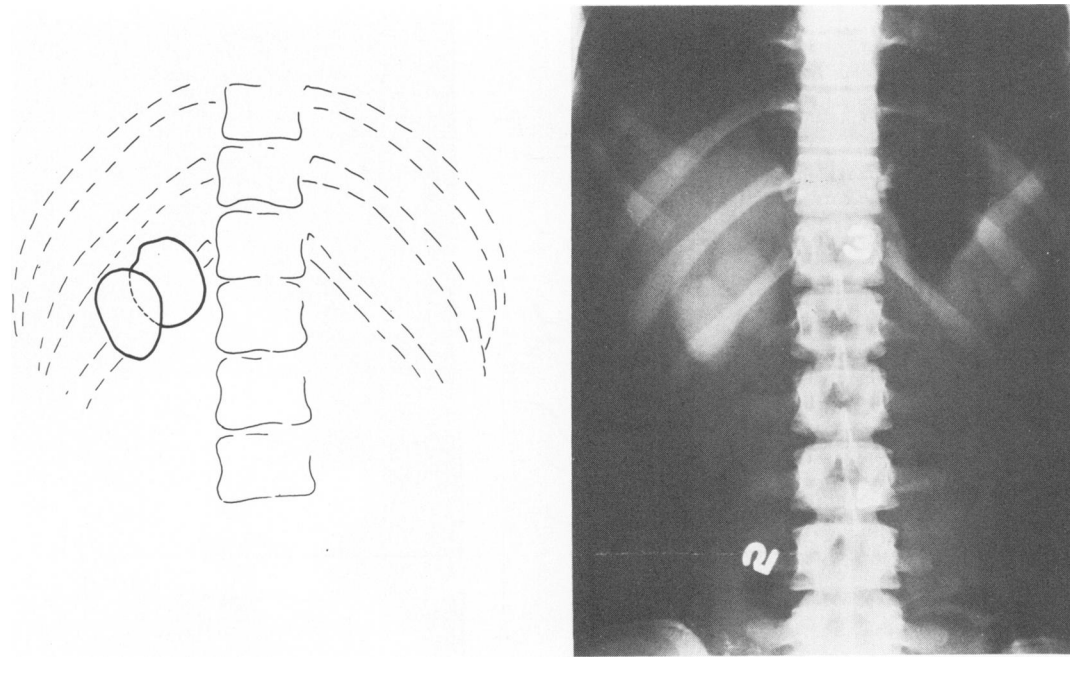

FIG. 3A. J. H. Sex: F Age: 17. Patient had a 7-week history of recurrent episodes of upper abdominal pain. Oral cholecystogram demonstrates a cystic dilatation of the common duct and a normal gallbladder.

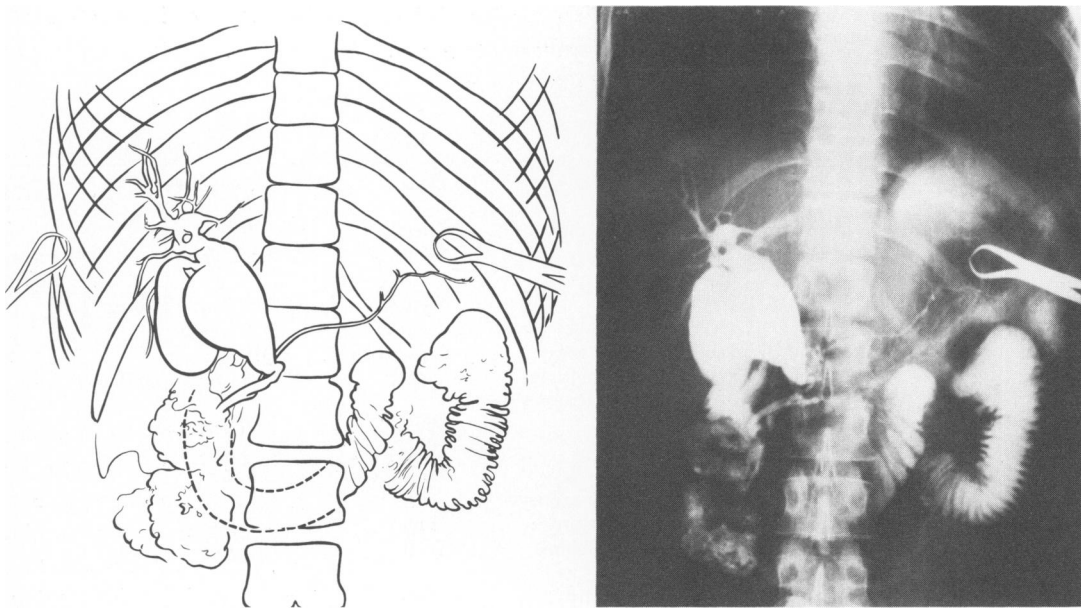

FIG. 3B. J. H. Sex: F Age: 17. Cholangiogram at operation revealed a large diffuse dilatation of the common duct, a normal gallbladder and a small congenital segmental cystic dilatation of the right hepatic duct. Decompression by choledochocystojejunostomy was done.

docholithotomy. The postoperative T-tube cholangiogram disclosed a small cyst at the bifurcation of the right and left hepatic ducts. Further operation for this cyst was not advised and the patient was lost to followup. It should be emphasized that three patients presented with liver abscesses, indicating the serious potential for infection which exists in the presence of bile stasis[1,2] (Fig. 3).

Operative therapy was employed in 14 of 15 patients. The patient not operated upon was a 20-year-old man who had jaundice, abdominal pain and anorexia of 5 days' duration. The diagnosis at the time of admission was infectious hepatitis. The patient died 3 days later because of *E. coli* septicemia. Autopsy revealed congenital cystic dilatation of the intra- and extrahepatic bile ducts with multiple liver abscesses. Another patient, age 4 months, died following incision and drainage of a liver abscess. Autopsy disclosed the congenital cystic dilatation of the biliary tree. Two patients, aged 49 and 61, were operated upon for calculous biliary tract disease. The congenital segmental dilatation of the biliary tree was not appreciated in these patients until their postoperative cholangiograms were reviewed. One patient has been lost to follow-up and the other is well 6 months postoperatively. Definitive treatment of the cystic biliary tract lesions was accomplished by excision in two patients, cysto-duodenostomy in three patients, cysto-jejunostomy

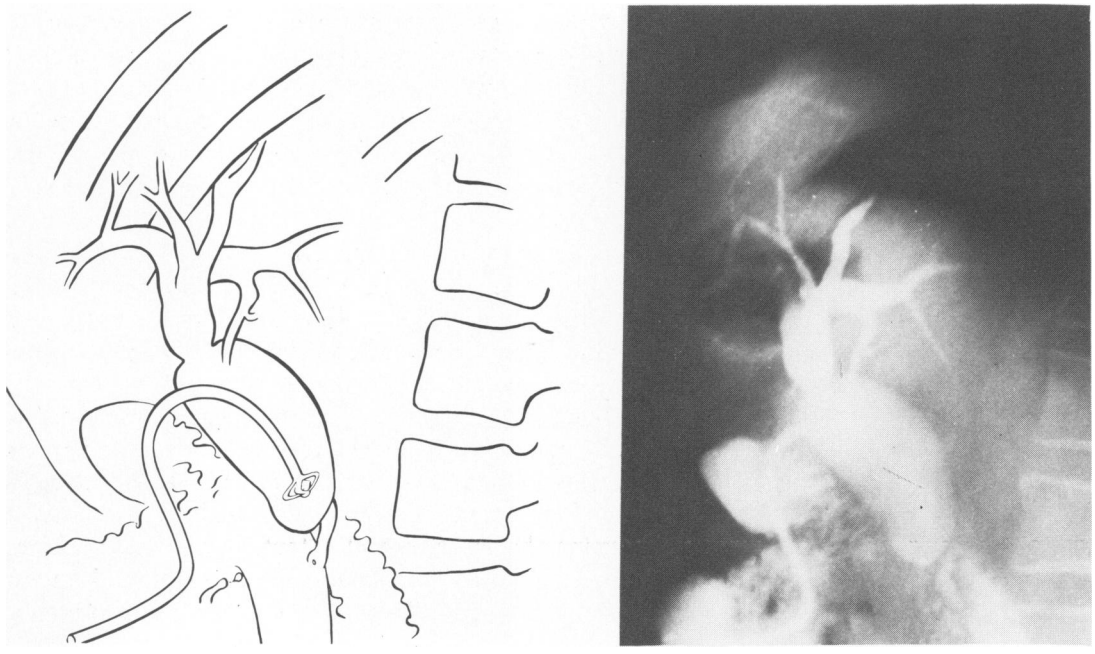

FIG. 4. V. B. Sex: M Age: 43. Operative cholangiogram revealing large choledochal cyst and intrahepatic dilatations. At a previous operation in another hospital this was recognized and decompressed by Malecot catheter. A choledochocystoduodenostomy was done.

(Roux-Y) in four patients, choledocho-jejunostomy, Roux-Y, in one patient and hepatico-jejunostomy (Longmire) in one patient.

In view of the variety of surgical procedures employed in the 11 patients who had had definitive treatment of the segmental cystic dilatations of the biliary tract, firm conclusions regarding the relative merits of each procedure are not feasible (Fig. 4). The two patients who had excision of choledochal cysts are alive and well, 5 and 6 years postoperatively. Of the three patients who underwent cysto-duodenostomy, one has been lost to follow-up, one died 9 years following operation for biliary cirrhosis and portal hypertension, and one is well 2 years postoperatively. Cysto-jejunostomy (Roux-en-Y) was performed in four patients and three are alive and well from 9 months to 17 years postoperatively. The fourth patient, age 7½ months, died 6 weeks following cysto-jejunostomy of intestinal obstruction and hepatic cirrhosis. Another patient, a female age 63, died 23 months after excision and choledocho-jejunostomy, Roux-Y for a carcinoma arising in a choledocho cyst and 27 years after having had the cyst demonstrated at the time that a cholecystectomy was done for cholelithiasis. The one patient subjected to hepatico-jejunostomy (Longmire) is symptom-free 2 years postoperatively.

Discussion

Intrahepatic dilatations of the ductal system have been described as "cystic" involving from one to several centimeters of duct, as well as "cylindrical" *i.e.*, dilatation of the entire ductal system within an anatomical segment of the liver. Patients with most types of congenital segmental dilatations of the biliary tree have been reported to also have biliary calculi. In some instances the calculi have been in the gallbladder, the gallbladder and common duct and in others calculi have been present only in the intrahepatic dilatations.

Calculous biliary tract disease may account for intrahepatic dilatations in those patients in whom there has been injury to the ductal wall due to obstruction caused by calculi or excessive scar formation. Congenital diaphragmatic obstruction with proximal ductal enlargement also may account for intrahepatic calculi. In this treatise we are not specifically concerned with those patients with calculous disease that has been associated with these conditions just mentioned. Rather we are presenting a small series of patients who, we believe, are examples of cystic dilatation of the biliary ductal system not related to calculous disease. Calculi may develop in these cysts or dilatations but are secondary and indeed only incidental so far as their development is concerned.

In the descriptions of a large number of patients with reported dilatations and/or cyst formation of both the intra- and extrahepatic bile ducts it is evident that there are several categories.

These include:

1. Cysts of the common duct.
2. Cystic dilatation of a segment or the entire common duct.
3. Single and/or multiple cysts of the intrahepatic ducts without abnormalities of the extrahepatic ductal system.
4. Intrahepatic cysts with segmental dilatations in the common duct.

Sporatic reports of intrahepatic cysts prior to Caroli's paper in 1958 for the most part did not dwell upon the etiology beeing due to a congenital defect, hypoplasia. Caroli emphasized that in the pure form of congenital intrahepatic cystic dilatation of the ductal system that the common duct frequently was moderately dilated. The term Caroli's disease has come to be applied to this group. It is our contention that the basic defect, hypoplasia involving chiefly the subserous part of the biliary ductal wall, accounts for most dilatations, cyst-like and cylindrical, of any part of the ductal system. We stress that the confusion of this problem is the result of not distinguishing between dilatations due to congenital hypoplasia and those that result from other disease of the biliary tract including those primarily due to calculi. Concomitantly it should be added that calculous disease may develop in patients with congenital hypoplasia and may add to the extent of liver damage as a result of mechanical obstruction and its common accompaniment, infection.

The surgical treatment of congenital segmental cystic dilatation of the biliary tree is dependent upon the location and extent of the disease as well as the presence or absence of acute inflammation (Fig. 5). In those patients with suppurative cholangitis or hepatic abscesses, primary consideration must be directed to drainage of the involved area. Definitive correction of the biliary tract anomaly is deferred pending subsidence of the suppurative process.

Selection of the most appropriate form of surgical therapy in patients with congenital segmental cystic dilatation of the biliary tree should be based upon the detailed anatomic information obtained by operative cholangiography. In the jaundiced patient without evidence of cholangitis, preoperative percutaneous transhepatic cholangiograms may be obtained in place of operative radiographic studies of the biliary tree. Irrespective of the method employed, cholangiograms are essential to determine the location and extent of the cystic lesions, particularly those that are intrahepatic (Fig. 6). In addition, they provide information regarding the presence

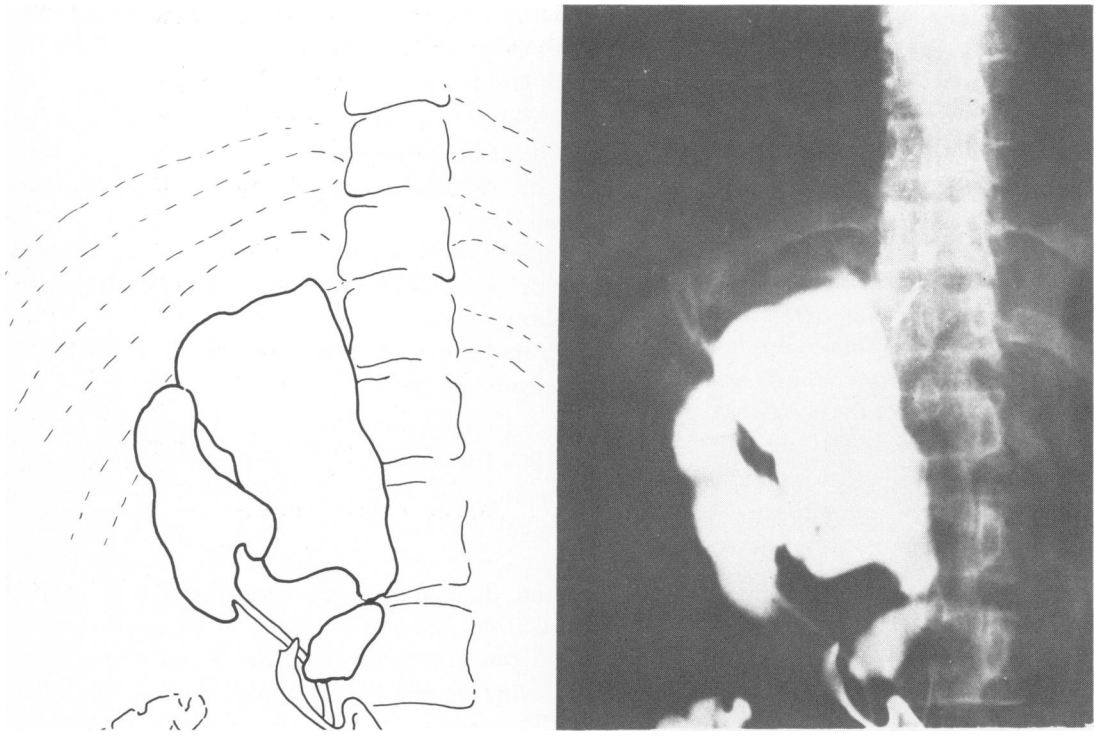

FIG. 5. H. G. Sex: F Age: 24. For 5 years patient had vague abdominal pain associated with nausea and occasional vomiting. Hospitalized for pancreatitis (amylase 236). cholecystostomy was done on 4th day following admission. Cholangiogram through cholecystostomy tube a few days later revealed large choledochal cyst with intrahepatic dilatations. A choledocysto-jejunostomy (Roux-Y) was done.

or absence of other anomalies of the biliary tree and/or calculi.

When technically feasible, excision of the cystic lesion is the procedure of choice. The anatomic prerequisite for successful excision is the presence of a relatively narrow attachment or communication between the cyst and a segment of an extrahepatic bile duct. In patients with cystic disease confined to one lobe or segment of the liver, excisional therapy may take the form of partial hepatectomy. Kasai and associates[6] have called attention to the increased incidence of carcinoma arising in choledochal cysts and advocate excision of these lesions. One patient in our group who had a choledochal cyst remained symptom-free for 27 years. At operation a carcinoma was found at the junction of the cyst and biliary duct.

Cystic lesions confined to the retroduodenal segment of the common duct are perhaps best treated by cystoduodenostomy. In most patients, however, radiographs will reveal that the cystic dilatations are extensive and

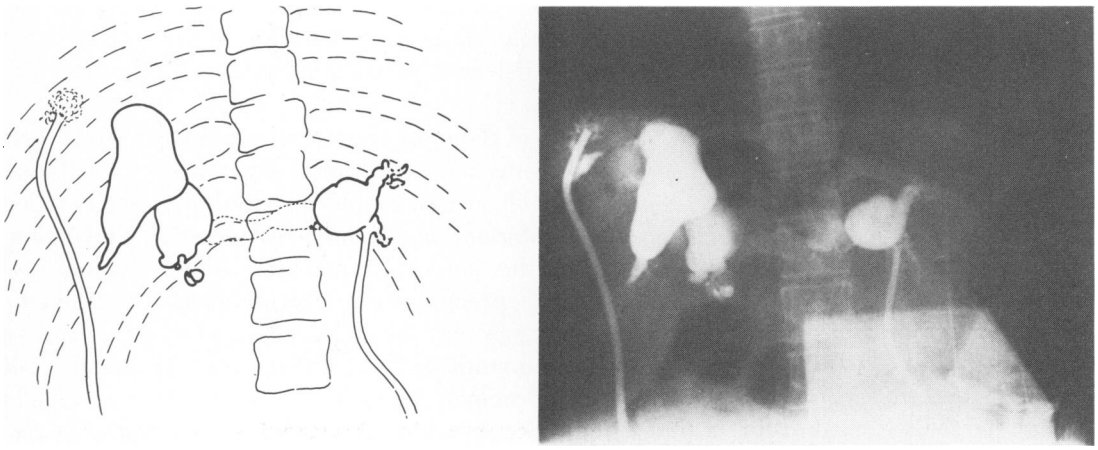

FIG. 6. M. C. Sex: F Age: 20. For 15 years, beginning at the age of 5, patient was subject to bouts of abdominal pain. At age 12 she underwent surgery on the biliary tract in Germany with relief for 2 years. Symptoms then returned and at exploration a subdiaphragmatic abscess was drained. She was again asymptomatic for 5 years. She was then admitted to The New York Hospital because of chills, fever and right shoulder pain. At operation an abscess was found in both the right and left lobes of the liver. These drained into the biliary ductal system. External drainage was established. A hepatico-jejunostomy (Longmire) was done.

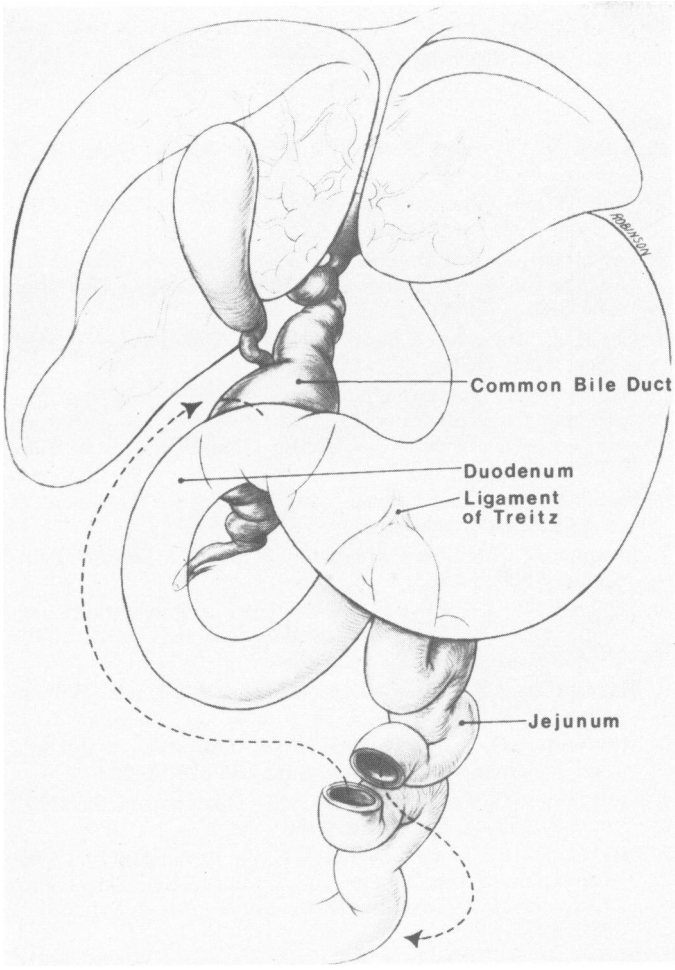

FIG. 7. The jejunum is transected 30 cm. or more distal to the ligament of Treitz. The end of the distal segment as indicated will be anastomosed to the choledochal cyst.

involve both the intra- and extrahepatic bile ducts. In these patients, it is our preference to employ cysto-jejunostomy side-to-end Roux-en-Y. This procedeure establishes decompression of the biliary tract and reduces the further impairment of function of liver tissue that is encroached upon by the cystic dilatation. The hazard of ascending infection is variable and unpredictable. There are those who believe that a stoma that is of sufficient size between the common duct and the intestinal tract, duodenum or jejunum, will not be followed by ascending infection. Our experience over the years does not necessarily support this view. We consider that a Roux-Y limb of 30 cm. and an end-to-side anastomosis does diminish the probability of ascending infection. The essential features of this procedure are depicted in Figure 7 and Figure 8. The cysto-jejunostomy stoma is established in the distal dependent portion of the cystic dilatation and proximal to the choledochoduodenal junction. Any appreciable segment of common duct distal to the anastomosis may be the site of accumulation of sediment material where infection may become established. This may be the cause of pancreatitis.

There is great need for carefully documenting information concerning patients who have been operated upon for this condition. As noted, the majority of these patients are in the younger age group and under ordinary circumstances have a long life expectancy. Of significance, but unknown, is the degree of recovery from impaired liver function following decompression of the biliary tree. Also unanswered is the question of whether or not these cystic dilatations are the site of calculous formation after decompression. It is evident that these patients should be followed at frequent intervals over many years if sufficient data is to be obtained that will be helpful in the management of this condition. In addition to measurement of liver function and estimation of liver size by scan, intravenous cholangiography and barium contrast studies of the cystojejunostomy stoma

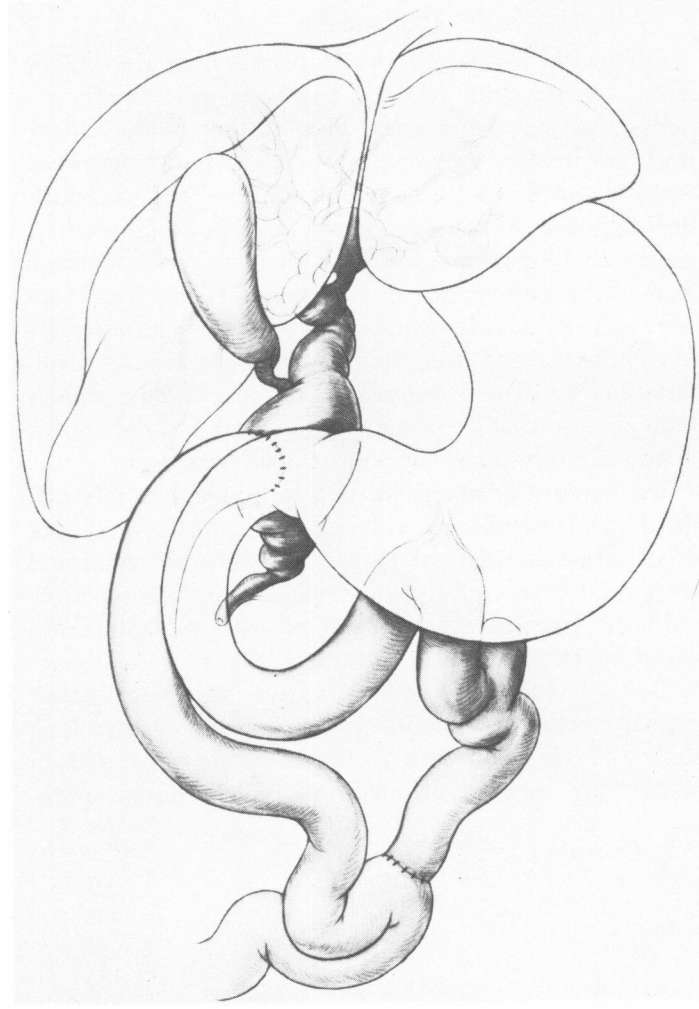

FIG. 8. Decompression by circumventing choledocho-duodenal junction with its sphincter mechanism. A Roux-Y end-to-side choledo-cysto-jejunostomy has been completed with a loop 30 cm. or more from the entero-enterostomy.

are helpful in the follow-up evaluation of these patients.

From presently available data, it is reasonable to assume that some patients with cystic dilatation of the biliary tree will require secondary operations. In those patients who develop cholangitis that persists despite antimicrobial therapy, consideration is to be given to revision of the cystoenterostomy and construction of a longer Roux-Y segment of jejunum. In addition, one must consider the possibility of an intrahepatic abscess in patients who exhibit signs of sepsis following biliary intestinal anastomoses. These may be drained to the exterior without molesting the common duct. The sparse autopsy data presently available indicate that patients who have died from this condition have had (1) suppurative cholangitis and hepatic abscesses; (2) cirrhosis of the liver, only occasionally accompanied by portal hypertension; (3) disease of the pancreas ranging from chronic pancreatitis to atrophy of the gland secondary to obstruction and associated with a deficiency of the external secretion.

Summary

Congenital segmental cystic dilatation of the biliary ductal system until recently has been infrequently reported. As roentgenological visualization of the biliary tract has become common, particularly in the course of surgical operations on the gallbladder and common duct, the diagnosis has become more readily established. A group of 15 patients with this condition are reported from the experience of the surgeons at The New York Hospital-Cornell Medical Center, together with a review of reports gleaned from the literature. The clinical manifestations so closely resemble those of calculous biliary tract disease that at present the diagnosis is dependent almost entirely upon cholangiographic findings.

The concept is advanced that the congenital defect in the ductal system is primarily one of hypoplasia or aplasia of the fibromuscular components of the submucosal and subserosal layers of the duct wall. This is compatible with the frequent involvement of both the intra- and extraphepatic bile ducts in these patients.

Decompression of the biliary tree has been accomplished in most patients by cysto-jejunostomy with a long limb (30 cm.) Roux-en-Y. Fewer patients have been treated by excision of the cystic dilatation or cystoduodenostomy. These patients require prolonged and close observation postoperatively.

References

1. Arthur, G. W. and Stewart, J. O. R.: Biliary Cysts. Br. J. Surg., **51**:671, 1964.
2. Attar, S. and Obeid, S.: Congenital Cyst of the Common Bile Duct. Ann. Surg., **142**:289, 1955.
3. Benintendi, V. and Bressau, G.: Segmental Congenital Cystic Dilatation of the Intrahepatic Bile Ducts. Acta Chir. Ital. **24/5**:601, 1968.
4. Caroli, J.: Diseases of Intrahepatic Bile Ducts. Israel J. Med. Sci., **1**:21, 1968.
5. Caroli, J. and Couinaud, C.: Une Affection Nouvelle, Sans Doute Congenitales des Voies Biliaires: La Dilatation Kystique Unilobaire des Canaux Hepatiques. Sem. Hop. Paris, **34**:136, 1958.
6. Kasai, M., Asakura, Y. and Taira, Y.: Surgical Treatment of Choledochal Cyst. Ann. Surg., **172**:844, 1970.
7. Langman, J.: Medical Embryology. Williams & Wilkins, Baltimore, 1969, p. 263.
8. LeNaour, R. J.: Sur un Cas de Tumeur Polykystique non Parasitaire de Foie. (These de Medecine, Paris, 1941) Paris These, **24**:1942.
9. Mayo-Robson, A. W.: Gall-Bladder and Bile-Ducts. William Wood & Co., New York, 1904, p. 200.
10. McWhorter, G. L.: Congenital Cystic Dilatation of the Bile and Pancreatic Ducts. Arch. Surg., **38**:397, 1939.
11. McWhorter, G. L.: Congenital Cystic Dilatation of the Common Bile Duct. Arch. Surg., **8**:604, 1924.
12. Meeker, W. R., Jr. and Nighbert, E. J.: Association of Cystic Dilatation of Intrahepatic and Common Bile Ducts with Laurence-Moon-Biedl-Bardet Syndrome. Am. J. Surg., **122**: 822, 1971.
13. Popper, H. and Schaffner, F.: Liver: Structure and Function. The Blakiston Div. of McGraw-Hill Book Co., New York, 1957, p. 165.
14. Todd, C. H.: History of a Remarkable Enlargement of the Biliary Duct. Dublin Hospital Reports. Vol. 1, Graisberry & Campbell, London, 1818, p. 325.
15. Tsuchida, Y. and Ishida, M.: Dilatation of the Intrahepatic Bile Ducts in Congenital Cystic Dilatation of the Common Bile Duct. Surgery, **69**:776, 1971.
16. Turnberg; L. A., Jones, E. A. and Sherlock, S.: Biliary Secretion in a Patient with Cystic Dilatation of the Intrahepatic Biliary Tree. Gastroenterology, **54**:1155, 1968.
17. Waller, E.: Idiopathic Choledochus Cyst. Ann. Surg., **66**:446, 1917.
18. Yotuyanagi, S.: Contributions to the Aetiology and Pathogeny of Idiopathic Cystic Dilatation of the Common Bile-Duct with Report of Three Cases: A New Aetiological Theory Based on Supposed Unequal Epithelial Proliferation at the State of the Physiological Epithelial Occlusion of the Primitive Choledochus. Jap. J. Can. Res. **30**:601, 1936.

Discussion

Dr. James E. Pridgen (San Antonio): Dr. Glenn's presentation today is certainly another classic in the archives of biliary surgery.

We would like to agree with Dr. Glenn's contention that these cystic dilatations inside the substance of the liver are similar to the same type of process which occurs in the common bile duct and in the hepatic ducts outside the substance of the liver. Caroli has talked and written about this repeatedly since 1958, describing these cystic dilatations in the liver. However, there had been many others before him who described this process; so that his article, which originated in '58, actually compiled the different forms of information previously reported.

We feel that the dilatations of the bile duct, whether they occur in the liver, outside the liver, or in a combination, are