Congenital Cystic Disease of the Liver and Biliary System

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CONGENITAL cystic abnormalities may occur throughout the biliary ductal system from the finest intralobular hepatic ducts to the intraduodenal portion of the common bile duct. Regardless of anatomical location, signs and symptoms related to these anomalies, when present, are of two general categories: (1) those related to the size of the cyst or cysts, i.e., vague pressure symptoms and compression of adjacent viscera, and (2) those related to varying degrees of biliary obstruction with or without superimposed infection and cholangitis. On rare occasions, acute symptoms may result from hemorrhage into a cyst or rupture of the cyst into the peritoneal cavity. Lesions arising from the finer intrahepatic ducts and confined to the liver parenchyma may be asymptomatic for many years, even throughout the life of the individual. Although the incidence of associated congenital anomalies is increased in all of these conditions, polycystic disease of the kidney which occurs in 50% of patients with polycystic disease of the liver[13] is by far the most frequently related anomaly. Such disease of the kidneys has a much more serious effect on renal function than polycystic liver disease has on hepatic function, and the general state of the patient's health usually is related to kidney involvement rather than to the status of the liver. Kerr and associates[9] pointed out that congenital hepatic fibrosis, although rare, may accompany any of these cystic ductal anomalies but probably most commonly occurs with cystic dilatation of the intrahepatic ducts.[5] It is often responsible for the development of portal hypertension early in life, thereby adding another dimension to the symptomatology of these congenital diseases.

As many of the cystic lesions of the biliary system are associated with recurrent cholangitis and some degree of biliary obstruction, it may at times be difficult to differentiate the congenital malformations from acquired changes. Visualization of the ducts by x-ray examination following injection of a radiopaque solution may demonstrate similar changes in both congenital and acquired conditions. For example, multiple congenital cystic dilatations of the intrahepatic ducts may be similar to the cystic dilatation, with or without abscess formation, that may be associated with a chronic high-grade biliary obstruction and superimposed intrahepatic cholangitis. Therefore, because of the similarities of the clinical picture with congenital lesions, certain acquired conditions of the ductal system have been included in this discussion.

Historical

The description by Douglas[4] in 1852 of a choledochal cyst generally is regarded as

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the first recognized account of a cystic lesion of the biliary system, although Douglas refers to a similar case previously recorded by Todd in the Dublin Hospital Reports. Todd's case he considered "defective" since the patient died almost immediately after being seen, there was complete obstruction of the duct, and the dilatation appeared to have been uniformly diffused to the common and hepatic ducts. On the other hand, the cyst in the 17-year-old female patient whose clinical course and postmortem findings were reported by Douglas filled the whole right side of the abdomen and contained a half gallon of yellow thin syrupy fluid with an offensive odor. The hepatic ducts were dilated; the cystic duct and gallbladder were not. "The remedies which appeared most to palliate her distress were hot fomentations and purgatives. Blisters were of some service." Repeated trocar aspirations of the cyst were proposed for treatment of future cases.

Four years later in 1856 Bristowe[2] reported a case of nonparasitic cystic disease of the liver and emphasized the association with polycystic renal disease. He was unwilling to speculate on the origin of the cysts but was inclined to view the coexistence of the same disease in the liver and kidneys as "merely a coincidence." Two weeks later at a meeting of the Pathological Society of London, Wilks [18] presented similar pathological specimens of cystic disease of the liver and kidneys from a case on the shelves of the Museum of Guy's Hospital and expressed the opinion that he did not believe these cysts to be the result of blockage of the biliary system.

Moschowitz [16] collected 85 case reports of cystic disease of the liver reported before 1906 and reviewed the theories regarding the etiology of cysts of the liver and kidney. He believed that the hepatic cysts arose from aberrant intrahepatic ducts.

Meyenburg [15] described in 1918 the occurrence in polycystic livers of groups or clusters of small bile ducts in the liver lobules separate from the portal areas which have subsequently become known as Meyenburg Complexes.

Melnick [13] in 1955 gave a comprehensive report of 70 cases of polycystic liver disease seen at the Los Angeles County General Hospital over a 30-year period and on whom postmortem examination had been performed, representing an incidence of 1 case per 687 autopsies. In a series of articles on benign tumors of the liver Henson *et al.*[7, 8] presented the cases of solitary and multiple cysts of the liver of surgical significance seen at the Mayo Clinic. Of 38 patients found to have solitary cysts, 12 had sufficient symptoms to warrant operation. Surgical pathological material was available for study from 29 patients who had polycystic disease of the liver.

Caroli *et al.*[3] are credited with having first described the condition of cystic dilatation of the intrahepatic biliary tree. They proposed the following classification of intrahepatic cystic dilatation and related diseases:

1) True polycystic disease of the liver.

Cysts occur in the parenchyma in association with typical Meyenburg complexes; they do not communicate with the biliary system.

2) Mixed form of polycystic disease.

Same configuration as (1) but associated with painful attacks of cholangitis. Certain cysts contain bile and communicate with the biliary system.

3) Cystic dilatation of the intrahepatic biliary ducts.

They contain bile and are associated with symptoms of pain and cholangitis. Meyenburg complexes are absent. There is dilatation of the main bile ducts.

4) Congenital fibrocystic disease.

Microcysts in broad bands of fibrous tissue (Hamartome biliaire fibreux); absence of lesions in the biliary tract. Signs of portal hypertension.

5) Multiple biliary cysts associated with fibrocystic changes.

Portal hypertension is present; there are cysts in the parenchyma and symptoms of cholangitis.

Kerr *et al.*,[9] as previously mentioned, emphasized in 1961 that congenital hepatic fibrosis was a condition related to congenital cystic liver and often associated with polycystic kidneys.

Foulk,[5] stated in an editorial concerning congenital malformations of the intrahepatic biliary tree in the adult, that when diffuse Meyenburg complexes are seen throughout the liver in association with prominent hypoplasia of portal vein branches, the lesion has been referred to as "congenital hepatic fibrosis." When dilatation and tortuosity of interlobular ducts appear predominant and symptoms seem to relate primarily to the anomaly of these ducts, the term "congenital dilatation of intrahepatic bile ducts" has been applied. Most frequently, dilatation of intrahepatic ducts is associated with congenital hepatic fibrosis, and the dilatation may be one of the causes of intrahepatic stones. Foulk believes that although Meyenburg complexes and intrahepatic fibrosis frequently are present in polycystic disease, the condition is not identical to that of congenital hepatic fibrosis.

Alonso-Lej *et al.*[1] analyzed 94 cases of choledochal cyst and classified the lesion into three major types. Their classification forms the basis of this report on extrahepatic cystic disease.

Case Material

Material for this report has been collected from the records of 58 patients with various types of cystic lesions related to the biliary system seen at the UCLA and Wadsworth Veterans Hospitals between 1956–1971. The classification of cases and the number in each group are presented in Table 1.

TABLE 1

Classification Cystic Condition	No. Lesions
I. Intrahepatic	
A. Primarily parenchymal	
1. Solitary with or without communication with biliary system	9
2. Polycystic disease	23
B. Primarily ductal	
1. Localized dilatation of a major intrahepatic duct	
a. Congenital	3
b. Acquired	2
2. Multiple cystic dilatations of intrahepatic ducts	
a. Congenital	2
b. Acquired	4
II. Extrahepatic	
A. Alonso-Lej Type A. Choledochal cyst	
1. Typical	9
2. Localized	1
3. Fusiform	1
B. Alonso-Lej Type B. Congenital diverticulum	
1. Arising from common bile duct, hepatic ducts, or gallbladder	2
C. Alonso-Lej Type C. Choledochocele	0
D. Multiple cystic dilatations	2
E. Unexplained diffuse ductal dilatation	
1. Congenital	3
2. Acquired	1

Intrahepatic

Primarily Parenchymal

Solitary Cysts. Solitary cysts from nine patients were identified, eight of which were lined with biliary-type epithelium (Fig. 1). One 4 × 5 cm. multilocular cyst which contained clear fluid was diagnosed as a serous cyst lined by thin mesothelial cells. Five of the cysts were from male patients and four from females. Absence of the expected 4 to 1 predominance of females may be accounted for by the fact that three of these patients were from the Veterans Hospital population. Of the remaining six from the University Hospital, four were female patients and two were male. Of the total group, three females and and one male had sufficient symptoms to require treatment. In four of the males and

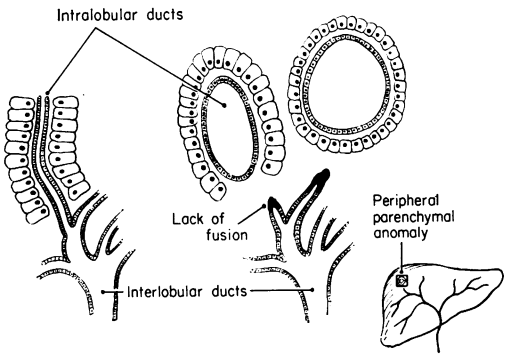

FIG. 1. Diagrammatic representation of hepatic cyst formation from intralobular ducts.

one female, the cyst was an incidental finding at operation or autopsy.

Symptomatic patients complained of fullness after eating or fullness and pain or tenderness in the epigastrium or right upper quadrant. Jaundice or cholangitis was not encountered.

A three-to-four fingerbreadth enlargement of the liver was described in three cases, and there was displacement of the second portion of the duodenum laterally and posteriorly on upper gastrointestinal x-ray examination. The hepatic flexure of the colon was also displaced downward. Liver function tests were normal in all patients. The cysts were totally excised in two patients. In a third patient where there was a two millimeter-sized communication with the biliary tract, the free edge of the cyst was partially excised and the remaining cavity anastomosed to a Roux-en-Y jejunal limb. Another patient was found to be totally asymptomatic one month following the second aspiration of 600 cc. of clear fluid from a cyst in the right lobe. This patient is under observation, and it is assumed that symptoms of fullness and discomfort will return when the cyst refills.

Although total excision is the treatment of choice, this method is hazardous unless the cyst is superficial. The larger cysts which extend deep into the parenchyma of the liver have numerous interlacing trabeculae in their walls which contain sizable biliary-vascular bundles. We encountered one such case in which the division of a large trabecula resulted in an obstinate postoperative biliary fistula. We have believed it safe, when the contents of the cyst are clear without any evidence of bile or infection, merely to operate and deroof the cyst, leaving it in free communication with the peritoneal cavity. When bile is present, the cyst is drained internally into a jejunal limb (Fig. 2C). When infection and purulent material are present, the cyst should be drained or marsupialized until the infection is controlled. Should bile or serous drainage persist, the cyst should then be drained internally into the intestine. Cysts less than 8 to 10 cm. rarely cause symptoms and probably require no treatment other than aspiration if discovered incidentally during operation.

Henson *et al.*[7] reported that the various types of epithelium lining solitary cysts suggest that more than one factor may be involved in the pathogenetic process. Although the lining epithelium in most of their cases was composed of flat or cuboidal cells similar to bile duct epithelium, tall columnar mucus-producing, ciliated, and even squamous epithelium was encountered. In general the symptoms and methods of treatment were similar to those in the present series. Complications included perforation, spontaneous hemorrhage, infection, or torsion if the cyst was on a pedicle.

An illustrative case is that of a 46-year-old woman who complained of fullness after meals for one year. There was no history of jaundice or cholangitis. An 8 cm. firm, rounded, slightly tender mass could be palpated below the right costal margin. There was an external pressure deformity of the first portion of the duodenum. Cholecystogram was normal. A 25 cm. cyst which lay in the interlobar fissure near the edge of the liver was excised and a cholecystectomy performed (Fig. 2, A, B). The

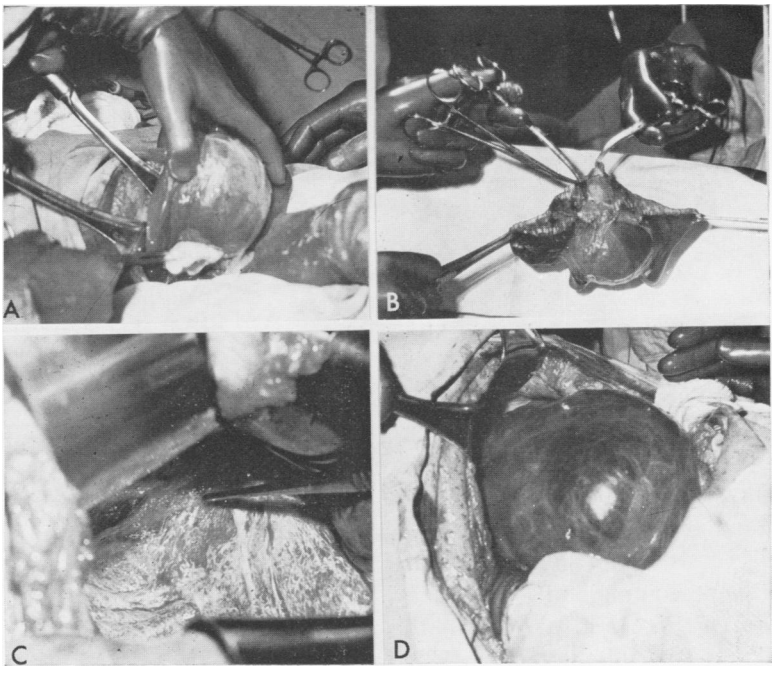

FIG. 2. A. Solitary superficial cyst in interlobar fissure. B. Excised cyst with margin of liver tissue. C. Interior of large solitary cyst. Instrument points to 2 mm. opening of bile duct into cyst. Cyst wall partially excised and remaining cyst anastomosed to jejunal limb. D. Large cyst containing 1,600 cc. clear fluid in liver with polycystic disease. Cyst wall partially excised and interior placed in free communication with peritoneal cavity.

cyst wall was lined with low cuboidal bile duct-type epithelium. The patient has been well for 8 years.

Polycystic Disease. Of 23 patients, there were nine females and 14 males. More than half were between 40 to 60 years of age when under our care. The youngest was 5 months and the oldest was 87 years. In all cases but one, normal hepatic function was preserved over periods of many years despite extensive involvement of the liver. The major disease process was related to polycystic disease of the kidneys in 11 patients, the cystic liver disease being an incidental and unimportant condition in the course of each patient's illness (Fig. 3, A, B).

Cysts of the liver discovered at autopsy or at operation for an unrelated condition were present in seven patients. One patient died after rupture of a congenital cerebral aneurysm and three others of malignant disease with tumors of the colon, pancreas, and brain.

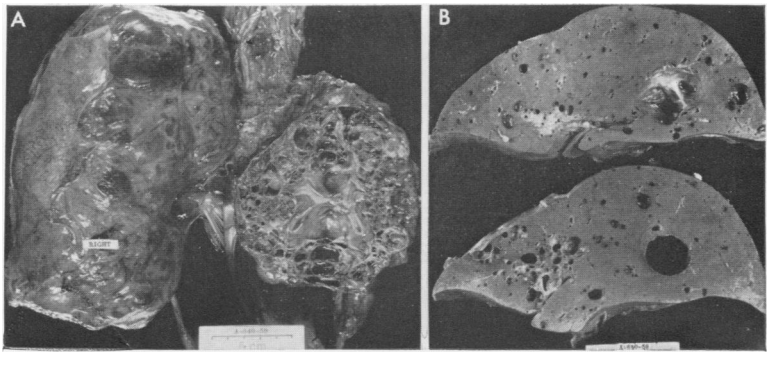

FIG. 3. Polycystic disease of kidneys (A) and liver (B) from patient who died of renal failure. Liver function normal.

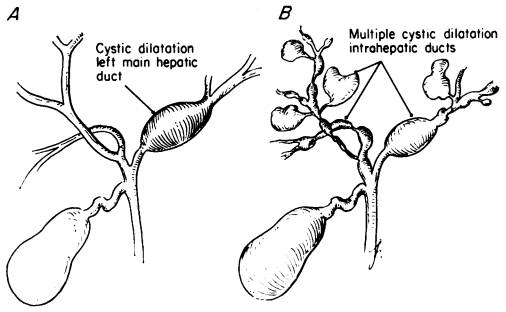

FIG. 4. A. Congenital cystic dilatation left main hepatic duct. B. Multiple cystic dilatations intrahepatic ducts.

Of five patients who underwent celiotomy, the procedure was performed in three for the purpose of establishing the diagnosis of an enlarged liver mass. One patient complained of fullness, discomfort after eating, and a mass in the abdomen. The mother of the final patient was known to have had cysts of the liver and kidneys, and the patient had been aware of an enlarged liver and had had attacks of fever and pain in increasing frequency for 8 years.

In four cases the larger cysts were punctured or the cyst wall partially excised. One such cyst which contained 1,600 cc. of clear fluid was completely unroofed and placed into free communication with the peritoneal cavity (Fig. 2 D). There have been no further symptoms or signs of recurrence in the 8 years since operation.

Internal drainage of a large infected bile-containing cyst into a Roux-en-Y jejunal limb was partially successful in one patient who eventually died of deteriorating hepatic function and portal hypertension from an associated congenital hepatic fibrosis.

Treatment of the cysts found incidentally at operation consisted of biopsy of the wall of a cyst in two cases, puncture of cyst in one case, and external drainage in one case. Histological findings of the epithelial lining were compatible with bile duct epithelium in all cases examined.

The management of this condition has been well expressed by Henson and associates [8]: "The disease generally becomes manifest in adult persons, has a long and benign course, and requires only symptomatic treatment, except in an occasional case in which the contents of large cysts may be aspirated or drained to reduce the weight of retained fluid." We would only add that partial excision of the exposed portion of the cyst, if the contents are clear and uninfected, and internal enteric drainage of bile-containing or infected cysts are methods of treatment we have found helpful.

A 49-year-old woman whose mother had renal and hepatic cysts had an enlarged liver for 8 years with attacks of pain and fever of increasing frequency and severity. Ascites had developed shortly before hospital admission. The liver, which extended to the iliac crest, was hard, nodular, and tender. The tip of the spleen was palpable. Liver function laboratory tests were normal. X-ray examinations of the stomach and intestine showed displacement but were otherwise normal, as were intravenous pyelograms. At operation the liver was noted to be stony hard with extensive cystic involvement. Cysts varied in diameter from a few millimeters to 20 cm. The largest cyst, which filled the dome of the liver, contained cloudy, bile-stained, foul smelling fluid from which micrococci were cultured. A Roux-en-Y jejunal limb was anastomosed to a dependent portion of the cyst. Other clear cysts were unroofed, and the liver was biopsied. The spleen was enlarged, and there was a moderate amount of ascitic fluid. The portal pressure was 240 mm. saline. The lining of the cysts was compatible with bile duct epithelium, and the liver biopsy showed evidence of congenital hepatic fibrosis. Postoperatively, signs of infection were improved but not entirely relieved. X-ray studies indicated the cyst enterostomy was not sufficiently

dependent to completely drain the cyst. Gradually, over a period of months, hepatic function began to deteriorate, multiple varicocele hemorrhages occurred, and the patient died eight months after operation.

Primarily Ductal

Localized Dilatation of a Major Intrahepatic Duct

1) **Congenital.** The three patients in the present series have cystic dilatations in the left main duct (Fig. 4 A); one of these also has other extensive cystic changes of the extra- and intrahepatic system. Two of the patients had long histories involving repeated bouts of jaundice with chills and fever. The third patient did not experience such acute attacks but had persistent mild jaundice with periodic exacerbations and gradual deterioration of hepatic function; she died without definitive surgical treatment. The cystic area was drained internally by anastomosis to the jejunum in one patient, and the left lateral hepatic segment was excised for removal of the cystic area and stones in another.

Repeated attacks of jaundice or cholangitis, at times starting in childhood, with normal common duct exploration should suggest intrahepatic dilatation as a possible factor. Complete operative cholangiography with visualization of the intrahepatic ductal system is essential to establish the diagnosis.

Glenn and Moody [6] reported four cases with dilatation of the main intrahepatic ducts, at times forming a cistern-like pocket which they believe favors stagnation and incomplete emptying of bile with the formation of calculi. Particles of calcareous material constantly are being passed into the common duct. Here these fragments act as a nidus for the formation of large stones which in turn may cause biliary obstruction. The left hepatic duct was involved in three of their patients and the right in one. Two patients were treated by multiple common duct exploration and stone removal. A transhepatic exploration and drainage of the left hepatic duct was performed in the other two patients.

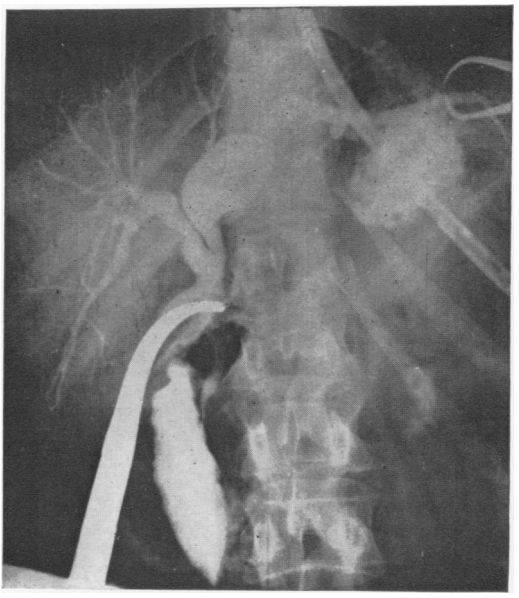

FIG. 5. Nineteen-year-old woman with congenital cystic dilatation left main intrahepatic duct. History of repeated attacks of cholangitis.

A 19-year-old girl * was first operated upon at 2 years of age following repeated bouts of fever, chills, itching, and jaundice. The liver was enlarged, and the gallbladder did not function on cholecystogram. At operation the liver was enlarged, and the gallbladder and extrahepatic ducts were normal. The symptoms cleared for 8 months without specific treatment. When jaundice returned, a needle biopsy was performed, after which jaundice increased. Exploration revealed hemobilia. For 13 years she was well until, once again, jaundice, itching, and fever returned. The gallbladder was removed, and the common duct was explored. A T-tube cholangiogram demonstrated a saccular area in the left main hepatic duct (Fig. 5). Following recurrent

* This case history is included with permission of Dr. John W. Freese, Fort Worth, Texas.

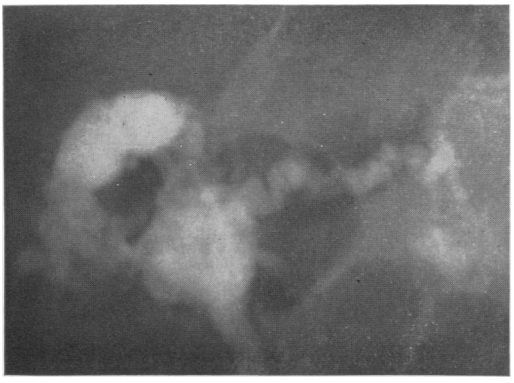

FIG. 6. Acquired dilatation of right intrahepatic duct and sigmoidization of left duct following high traumatic stricture of common hepatic duct.

attacks of jaundice, the cyst was anastomosed to the jejunum in January, 1969. There have been no further attacks.

2) **Acquired.** Similar saccular dilatations of the intrahepatic ducts can arise with prolonged, recurrent obstruction, biliary stasis, and infection. We have encountered two such cases. One patient has developed a moderate saccular dilatation of the right main duct (Fig. 6) following development of a high stricture in the common hepatic duct caused by excision of the remaining extrahepatic biliary system during cholecystectomy. This has been treated with a biliary enteric anastomosis with disappearance of the dilated area.

The second case, previously reported,[11] was a 43-year-old man whose cholecystectomy and common duct stone removal was the first of four operations for choledocholithiasis. At the third operation a side-to-end choledochojejunal Roux-en-Y limb anastomosis was performed. The ductal system both intra- and extrahepatic was filled with stones and debris. Marked liver tenderness, chills, fever, and jaundice led to a fourth operation which disclosed a cystic dilatation of the first portion of the left hepatic duct filled with soft stones and sludge which obstructed the left duct and gave rise to distention of the duct, infection, and numerous intrahepatic abscesses. Removal of stones, prolonged transhepatic cyst drainage, and irrigation have resulted in complete relief of symptoms for 3 years.

Multiple Cystic Dilatations of Intrahepatic Ducts

1) **Congenital.** Two patients with this diagnosis are included in our series (Fig. 4B). One was a 65-year-old woman who complained of right upper quadrant abdominal pain, malaise, anorexia, and 15-pound weight loss. Laboratory tests of liver function were normal, and the right lobe was enlarged on scan. At operation the liver was diffusely enlarged, hard, and scarred. No nodules or masses were palpated. The common duct was moderately dilated. The gallbladder was thickened, but there were no stones in the gallbladder or the duct. Biopsy of the liver disclosed a striking, well-circumscribed peribiliary duct fibrosis of the Meyenburg complex type which did not involve the parenchyma. The finer biliary ducts were dilated. There was no evidence of bile stasis or tumor. A diagnosis of multiple cystic dilatation of intrahepatic ducts was made. Her symptoms have continued, but weight has been stabilized during the past 2½ years.

The second patient, a 47-year-old woman, stated that she had "never been in good health." "Liver medicine" had been prescribed for her 29 years before following her first pregnancy. She had always had an intolerance for fatty foods. Two years before admission she developed daily episodes of green emesis, jaundice, and dull pain in the right upper quadrant radiating into her back. She lost 15 pounds, and the symptoms fluctuated until following a particularly severe attack of pain. A cholecystectomy and liver biopsy were performed. The gallbladder was full of stones, the liver "abnormal," and the spleen was enlarged. Jaundice, pain, fever, and weight loss continued. Liver and spleen were

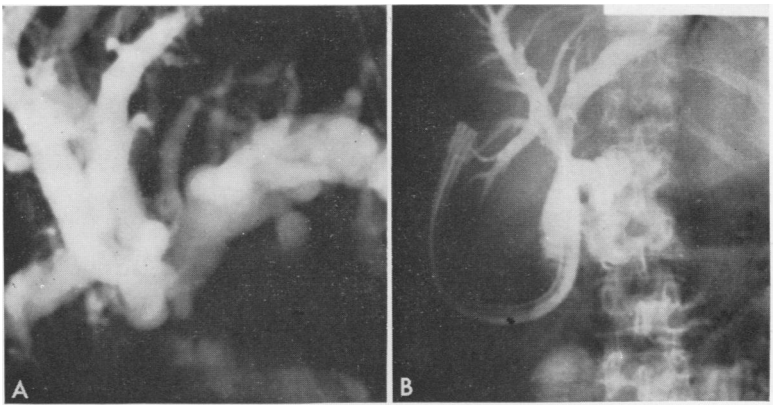

FIG. 7. A. Marked obstructive intrahepatic duct obstruction with saccular areas. B. Marked decrease in caliber of duct following duct-jejunal anastomosis.

markedly enlarged, and there was deep jaundice.

Operative cholangiogram revealed dilated intra- and extrahepatic biliary system with numerous alternating cystic and stenotic areas within the parenchyma and a long area of stenosis in the distal portion of the common duct. Since choledochojejunostomy above this area, there has been a significant improvement in the patient's condition with serum bilirubin levels only slightly above normal. Enlargement of liver and spleen and signs of portal hypertension persist, but her condition is otherwise satisfactory 2 years following operation.

Both of these patients showed evidence of prolonged biliary tract disease with symptoms of cholangitis in one. The liver was enlarged and fibrotic with dilatation of the intrahepatic ducts and portal hypertension. The second case was complicated by a stenosis of the distal portion of the common duct. Duct-enteric anastomosis above this site has relieved the symptoms of cholangitis and biliary obstruction.

Mercadier and associates,[14] in 1968, presented a case of cystic dilatation of the intrahepatic duct and reviewed the management of six additional cases previously reported. Left hepatectomy with anastomosis of the jejunum to the right ductal system or the common duct gave the best results. Following various types of treatment, two of the seven patients died, two were improved, and two were considered cured. Their patient had an excellent short-term result following left hepatectomy and right hepaticojejunostomy.

2) **Acquired.** Similar cystic dilatations have been identified in four patients with prolonged histories of biliary obstruction and infection. Such obstruction produces dilatation and ectasia of the duct which on cholangiogram may appear as cysts. This has been referred to as "sigmoidization" of the ducts. Multiple small abscesses also may develop under these conditions, giving an appearance quite similar to that seen in multiple congenital intrahepatic cysts.

These conditions have been identified as acquired in these cases on the basis of normal or near normal cholangiograms of the intrahepatic ductal system either before the onset of severe obstruction and infection or after symptoms have been relieved (Fig. 7, A, B).

Extrahepatic

Choledochal Cyst—Alonso-Lej Type A

Typical. Of the nine patients with the Type A variety of choledochal cyst (Fig. 8A), seven were females and two were males, indicating the usual preponderance

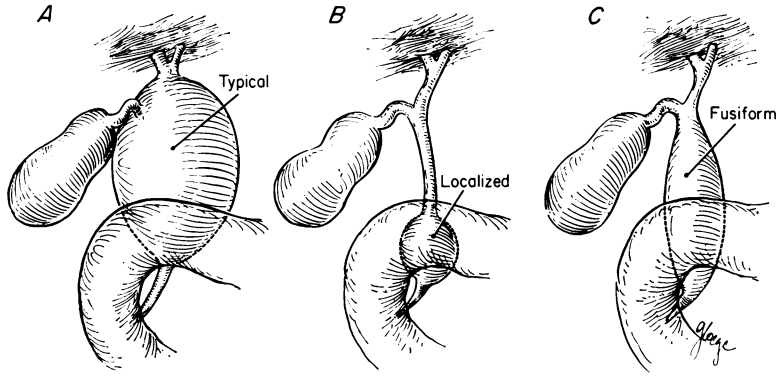

FIG. 8. Varieties of choledochal cysts: A. Typical, B. Localized, and C. Fusiform.

of females. Two patients were under 5 years of age, five ranged between 20 and 40, and two were over 40.

One 30-year-old patient had been well for 5 years following exploration for diagnosis but without any specific surgical treatment for the cyst. X-ray examination of the abdomen showed the cyst to be filled with air, presumably by passage through an incompetent sphincter of Oddi. In another case, that of a 4-year-old girl, excision of the cyst and choledochojejunal anastomosis were performed with good results.

Drainage of the cyst by means of a cystjejunostomy was employed in four patients. One developed acute cholecystitis requiring cholecystectomy 4 months after the drainage procedure. The course of the other three patients has been satisfactory. In our recent cases, cholecystectomy has been performed routinely at the time of cyst drainage.

Three patients in whom the cyst was anastomosed to the duodenum have required reoperation with conversion to a cystjejunostomy, two because of stricture of the anastomosis and recurring cholangitis and one because of severe pain associated with the regurgitation of duodenal contents into the cyst. All three have been asymptomatic following reoperation, although one patient has developed cirrhosis with depressed hepatic function.

Trout and one of the present authors (WPL) [17] recently presented evidence that the long-term results of jejunal decompression have been better than those of du-

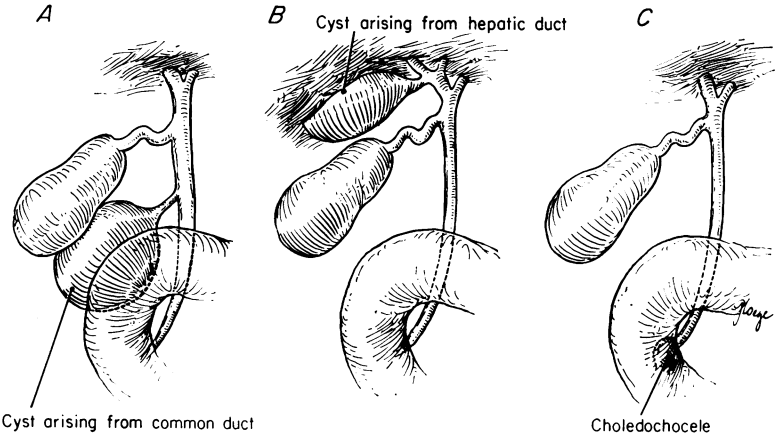

FIG. 9. A. and B. Alonso-Lej Type B. Choledochal Cyst—congenital diverticula arising from common duct (A) and hepatic duct (B). C. Alonso-Lej Type C. Choledochal Cyst—choledochocele. Cyst in duodenal intramural area of common duct.

odenal drainage and that the development of acute cholecystitis has been sufficiently frequent that cholecystectomy should be performed concomitantly with the initial cyst-enteric anastomosis.

Localized and Fusiform. It is important to recognize that the choledochal cyst may not present as the typical sac-like structure that occupies most of the common hepatic and common bile duct from near the junction of the left and right hepatic ducts down to the head of the pancreas, but instead may involve only a segment of the extrahepatic system.

In a 10-year-old girl with repeated attacks of jaundice, pain, and fever in whom a 3 cm. dilatation of the distal end of the common duct was found, anastomosis to a jejunal limb was accomplished with relief of symptoms for 7½ years (Fig. 8, B).

A common duct exploration has been performed six times in a 54-year-old woman with recurrent attacks of pain, jaundice, and fever over a 33-year period. The common duct had been noted to be markedly dilated, but at each exploration, this was considered a result of repeated stones and obstruction. An operative cholangiogram, however, revealed a marked fusiform dilatation of the common duct with normal hepatic ducts, and it was determined that the dilatation of the duct was the primary condition and not secondary to obstruction. Following anastomosis of the dilated duct to a jejunal limb, no further attacks have occurred for 14 years (Fig. 8, C).

Type B—Congenital Diverticulum

Alonso-Lej *et al.*[1] reported four cases of this rare type of cystic diverticulum in which the cyst is connected by a stalk to common duct, the hepatic ducts, or the gallbladder (Fig. 9, A, B).

A 64-year-old man in our series continued to have right upper quadrant pain and jaundice following cholecystectomy and

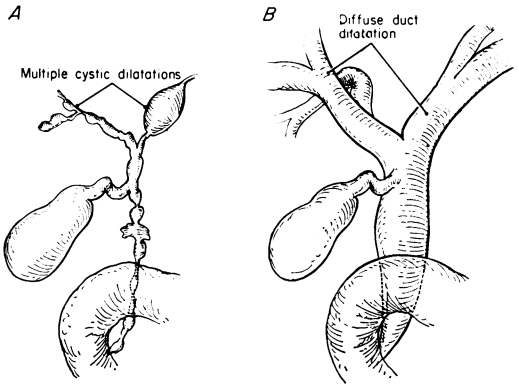

FIG. 10. A. Multiple cystic dilatations of ductal system. B. Congenital dilatation of entire ductal system. The term hypotonia has been applied to this condition.

negative common duct exploration. A T-tube cholangiogram disclosed a sac-like structure which seemed to fill with dye from the right hepatic duct. Upon exploration, the cyst was found embedded in the right lobe of the liver near the gallbladder fossa. At the time of excision, the cyst, which was filled with stones and debris, was thought to be a second gallbladder; however, microscopic examination revealed the cyst wall to be lined with bile duct epithelium. There have been no further biliary symptoms.

Type C—Choledochocele

This rare type of lesion involves only the intraduodenal portion of the common bile duct (Fig. 9, C.). The bile and pancreatic ducts open into the cyst which then open into the duodenal lumen. There is little, if any, dilatation of the biliary tree above the cyst. Since the cyst is said to be lined by duodenal mucosa, there is question as to whether the lesion should be considered a type of choledochal cyst or not. The small opening into the duodenum may cause obstruction. We have not encountered this type of choledochal cyst.

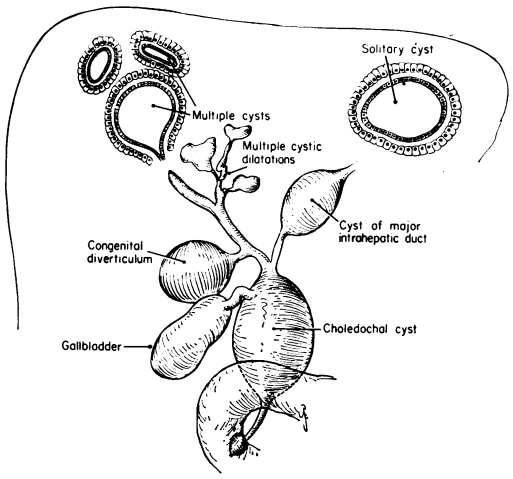

FIG. 11. Composite schematic drawing indicating the location in the biliary system of the various types of congenital cystic lesions.

Multiple Cystic Dilatations

Three cases of this unusual condition have been collected including one personal case.[11] In our case, multiple saccules or diverticula arose from the common and hepatic ducts, and a saccular dilatation was present in the first portion of the left hepatic duct (Fig. 10, A). Radiopaque dye injected into the duct flowed freely into the intrahepatic radicles and into the duodenum without evidence of obstruction. However, the patient continued to have increasingly frequent bouts of obstruction, developed secondary biliary cirrhosis, and recently died of hepatic failure.*

A second patient with similar intrahepatic ductal features recently has been admitted to the UCLA Hospital; however, the entire extrahepatic biliary system is missing following cholecystectomy. Possibly the abnormal appearance of the common duct may have made difficult its identification and thereby contributed to the surgical misadventure.

Unexplained Diffuse Ductal Dilatation

Congenital. Three patients in this category are included in this series; all had long histories of biliary tract disease dating back to childhood or early adult life with repeated bouts of jaundice and cholangitis. Repeated duct explorations disclosed the recurrent formation of soft pigment-type stones and sludge. The entire intra- and extrahepatic duct system became dilated, but no organic obstruction could be demonstrated (Fig. 10, B). For lack of a better name, this condition has been referred to as hypotonia of the common duct. It resembles a choledochal cyst, particularly the fusiform type, in its symptomatology and behavior; however, contrary to the findings in choledochal cysts, the dilatation extends well into the intrahepatic system, and stones and sludge are formed repeatedly and in considerable quantity.

It has generally been recommended that such patients be treated by choledochoduodenostomy, although we have found this to be unsatisfactory. Even an end-to-end choledochojejunostomy has not relieved entirely the apparent effects of bile stasis. It has been suggested that the entire malfunctioning common and hepatic duct be excised and that a high hepaticojejunostomy be used. In our present state of knowledge, we would hesitate to perform this procedure until an anastomosis at a lower level in the duct was found to be inadequate.

Acquired. Dilatation of the common duct from long-standing partial obstruction resulting from a stone or stones may produce an inflammatory thickening of the dilated ductal wall that remains even after removal of the source of the obstruction.[12] This lesion which has been referred to as inflammatory choledochitis may then serve as a reservoir for bile stasis and the formation of stasis stones. When such changes occur in the duct, drainage should be enhanced by the creation of a duct-enteric anastomosis. The differentiation of this condition from congenital hypotonia may be difficult. The onset of biliary disease early in life suggests the congenital type.

*This patient had been followed by Dr. Bernard D. Sherer, Bellflower, California.

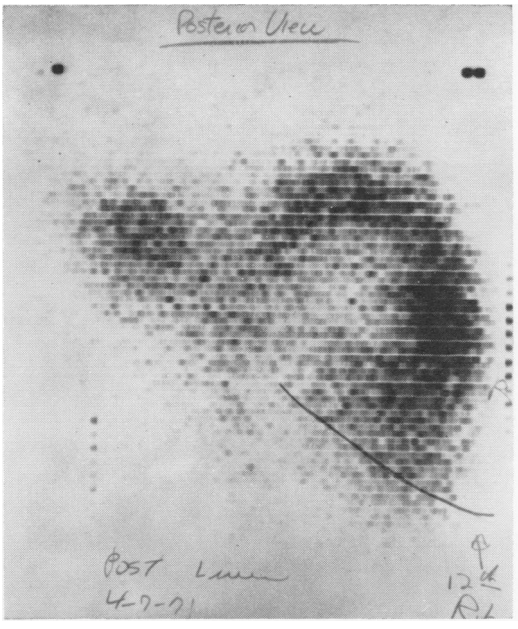

FIG. 12A.

One patient, a 74-year-old woman, has been selected from our files as an example of this process. Thirty-five years before admission, a cholecystectomy had been performed for acute cholecystitis with stones. She had mild attacks of epigastric pain but did fairly well for 15 years when she became jaundiced and was thought to have hepatitis. Two years later stones were removed from a greatly dilated and thick-walled common duct. Postoperative T-tube cholangiograms were negative for stones. Fourteen years later following recurring attacks of pain and jaundice, a number of soft pigmented stones were removed from a widely dilated duct which measured 8 cm. in diameter. A choledochoduodenostomy was performed. The patient had no further biliary tract disease until the time of her death 6 years later of an unrelated disease.

Summary

Cystic biliary malformations may be classified in relation to the levels of the biliary tree. Embryologically, according to Lewis,[10] the distal portion of the liver anlage forms the hepatic parenchyma, and the proximal portion forms the ductal system, which then grows into the rapidly enlarging distal hepatic portion. The terminal branches of the ductal system reach to the liver lobules and form the *inter*lobular bile ducts in the porta septa. Meanwhile, small bile ducts begin to appear in the liver parenchyma, the so-called *intra*lobular bile ducts which connect the bile canalicula to the interlobular bile ducts in the porta septa.

Meyenburg[15] proposed that at one stage in embryological development, many more intralobular bile ducts are formed than are necessary. Failure of these ducts to involute and in the absence of proper distal duct connections, they may undergo gradual dilatation to form a polycystic liver. Solitary cysts of bile duct origin may arise from a similar mechanism.

Mutiple cysts of the intrahepatic ducts may represent a malformation in the most distal portion of the ductal system formed from the duct anlage. Other cystic congenital duct malformations have been listed progressing from that point down the biliary tree, these all being related to malformations in the proximal or ductal anlage (Fig. 11).

Surgical treatment is designed generally

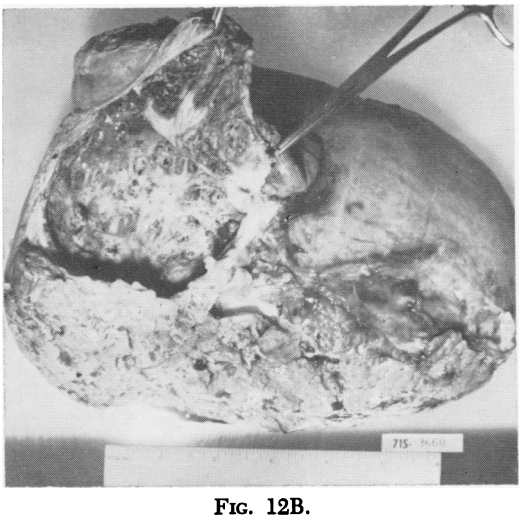

FIG. 12B.

to limiting the size of the liver mass so as to avoid interference with adjacent organs and structures and to maintaining a normal flow of nonseptic bile. Symptomatic improvement can be achieved in the majority of these anomalies despite the fact that they cannot be anatomically corrected.

This report relates these anomalies to a general scheme, presents enough information about each condition to facilitate its identification, and discusses aspects of management.

Addendum

Since submission of this manuscript, an additional 51-year-old man has been treated for Caroli's disease. Following a 10-year history of intermittent attacks of cholangitis, a large filling defect was detected on liver scan in the right lobe (Fig. 12A). Operative cholangiogram demonstrated a large right intrahepatic bile duct cyst with stones and debris in the enlarged hepatic and common ducts. As the condition did not improve after choledochojejunal anastomosis, the cyst and right hepatic lobe were removed at a second operation (Fig. 12B). A subhepatic abscess and continued sepsis resulted in death of the patient after a prolonged complicated course.

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Discussion

Dr. William Rhoads Waddell (Denver): Dr. Longmire, in his characteristic way, has thoroughly described and documented this impressive series of cases, and it will stand as one of the larger and better studied series, but I would point out from the slides that were shown and one of the tables that will be included in the published report that there is only one group that has 20-odd cases, two other groups that have nine cases, and then the remainder are scattered with one, two, or three cases. So there really isn't an opportunity to know too much about these conditions.