Abnormalities of the Bile Ducts*

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INJURY OF THE BILE ducts under ordinary conditions is usually due to poor execution of surgical procedures on these structures. However, the occurrence of abnormal arrangements of these ducts, particularly in the presence of severe inflammation, enhances the danger of injury, and in some cases almost precludes satisfactory repair when injury has been inflicted. It is important, therefore, that such abnormalities should be thought of while operations on the gallbladder and the ducts are in progress. Unusual courses of the blood vessels may also lead to bleeding and then to injury, but this will not be considered. The congenital deformities in infants present difficult and often insurmountable problems, but they, too, are outside the scope of this paper.

Eisendrath[8] in 1920 presented an excellent review of injuries to the common duct. He collected 51 cases, some of these had been operated on by eminent surgeons, and he attributed the accident in many of these cases to abnormalities in the duct arrangement.

It is difficult to estimate the incidence of these abnormalities because at operation many of them are not observed and cause no trouble, and in postmortem examinations they would be overlooked unless the pathologist were particularly careful to search for them. Eisendrath[8, 9] has reported that anomalous bile ducts were found in 8 per cent of a series of necropsies, while Michels[29] found accessory ducts in 18 per cent of 200 specimens. Abnormal course of the cystic duct is a frequent occurrence. It may run parallel to the hepatic duct, and this has been reported in autopsy series by Flint,[10] 14 per cent of 200, and by Thompson,[27] 23 per cent of 421 bodies. It may run a spiral course and be so intimately related to the hepatic duct, especially in the presence of inflammation, that identification is quite difficult (Eisendrath[8] and Werelius[28]). It may also be completely absent, as will be reported here in two cases.

The vast majority of these abnormalities are of congenital origin; therefore, the embryological development of the liver and its ducts will be briefly reviewed.

EMBRYOLOGY

To understand various anomalies and abnormalities pertinent to surgery of the biliary system, one must study the complicated development of these structures. Essentially, the biliary system originates from entoderm lining the primitive intestine during the fourth week. Jordan and Kindred[14] state that the hepatic groove develops into the hepatic diverticulum, which is an entodermal outpouching from the site of the future duodenum at its middle part. This becomes a short tubule which in turn gives rise to numerous solid branches of entoderm, and these invade the mesoderm of the septum transversum and the ventral mesentery of the primitive gut.

According to Patten[22] these cell cords grow out between the layers of the splanchnic mesoderm and become tubules, the distal portions of which form the secretory tubules of the liver. Proximally these tubules

* Presented before the Southern Surgical Association, Hollywood Beach, Florida, December 7, 1954.

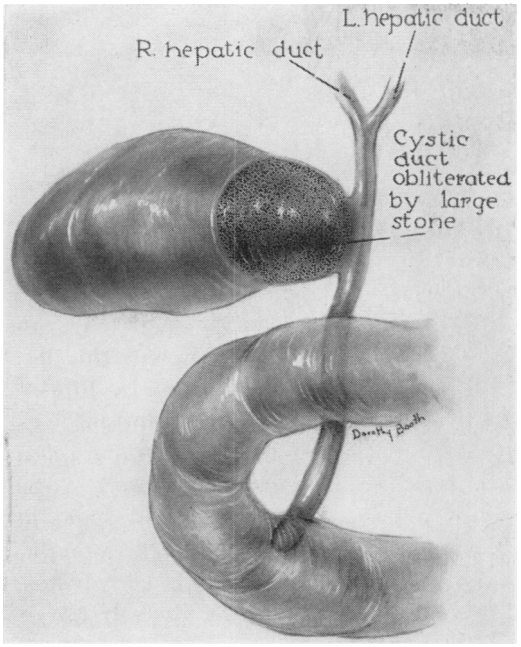

FIG. 1. Illustrating Cases 1 and 2, large stone in gallbladder has obliterated the cystic duct.

become confluent, and in tree-like fashion they taper down to form the hepatic ducts, the common hepatic duct, and the common bile duct. Many of these tubules disappear or coalesce, and it is not difficult to imagine multiple variations in the limbs of the tree due to the manner of their mergence or failure of disappearance. The latter would give rise to the various accessory ducts. Boyden[4] discussed developmental anomalies of the biliary tract in an excellent article on its embryogenesis in 1944.

The gallbladder arises as a local dilatation from the original diverticulum near the site of confluence of the two hepatic ducts, and the proximal part of this dilatation becomes narrowed to form the cystic duct. The liver is made up of the distal fine branches of entodermal tubules which have invaded the mesoderm, and are closely meshed with the sinusoidal capillaries of the vitelline and umbilical veins.

A study of the comparative anatomy of the biliary system in man and the lower animals is of interest in connection with reviewing the embryology. Such a study was made by Mentzer[19] in 1929, when he concluded that about 10 per cent of human beings have anomalous biliary tracts. He stated that most of the anomalous structures found in man represented normal arrangements found in animals, and stressed the importance of a knowledge of the comparative anatomy by the surgeon. It is certain that the surgeon who is faced with abnormalities of the biliary tree will be better equipped to handle the situation if he has knowledge of comparative anatomy and embryology.

REPORTED ABNORMALITIES

In reviewing the literature one finds numerous reported cases of abnormalities of the biliary tract. Almost four centuries ago Blasius[3] described a double gallbladder with separate cystic ducts. More than 300 years ago Huber in Switzerland[13] recorded the first "Y"-shaped type of gallbladder reported in man. Since then there have been so many abnormalities recorded that recently Michels[20] stated, "Many years ago Sir Arthur Keith stressed the fact that in the biliary region, 'variation is rampant'."

Although disease of the gallbladder is not uncommon, anomalies of this organ are relatively rare. Boyden[4] in 1926 collected 19,000 reports of cadavers and patients, finding 20 cases of accessory gallbladder. Gross[11] reviewed 148 cases of congenital anomalies of the gallbladder and discussed variations in its form (double, bilobed, absent) and position (intrahepatic, left-sided, floating). Mayo and Kendrick[17] have recently discussed anomalies of the gallbladder and reported a case of left-sided floating gallbladder with a cystic duct 6 cm. in length.

Most of the reported anomalies of the common bile duct have been the choledochal cyst. Much has been written about this rarity. In 1951 Macpherson[16] reviewed the literature and found 200 reported cases. He added an autopsied case which was more unusual in that two accessory hepatic ducts

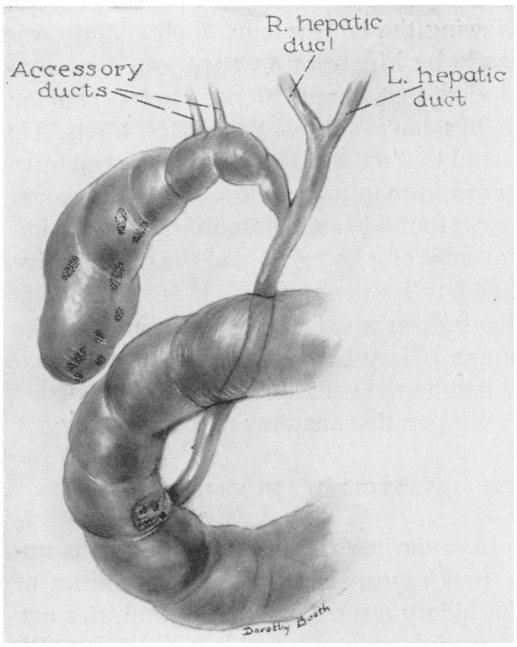

FIG. 2. Two accessory ducts from right lobe of liver entering gallbladder.

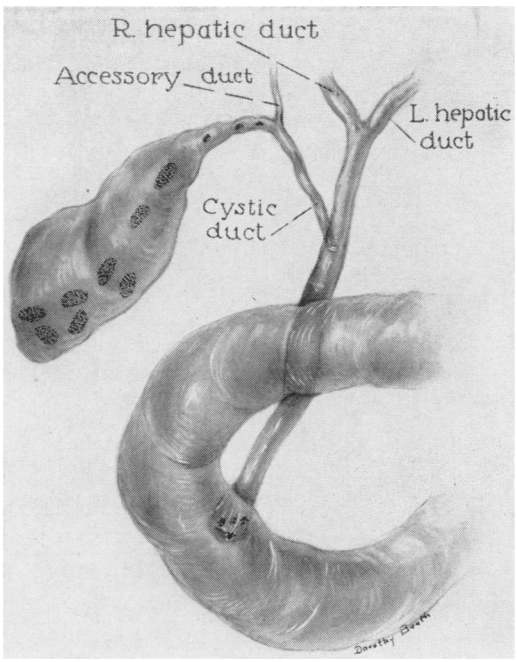

FIG. 3. Accessory duct from right lobe of liver entering cystic duct.

were found entering the cyst on its right side.

Abnormalities of the hepatic ducts are undoubtedly often present and not recognized. Milroy[23] reported two interesting anomalies of the right hepatic duct. In one case this duct opened into the gallbladder, which then emptied into the left hepatic duct through two cystic ducts to form the common duct. In the other the right hepatic joined the gallbladder at the beginning of the cystic duct, and this large joint duct entered the left hepatic to form the common duct. His cases prompted the familiar debate over removing the gallbladder from "fundus down" or "cystic duct upwards." Meade[18] reported a case of a small diverticulum arising from the common hepatic duct.

Accessory hepatic ducts are also present more often than one would expect. In 1922 Flint[10] reviewed 29 cases of accessory bile ducts varying in size from a bristle to the size of the right hepatic duct. Continuous drainage of bile after cholecystectomy is attributed in many cases to accidentally and unknowingly cutting accessory ducts. Beaver[1] in 1929 described variations in the extrahepatic biliary tract in 57 cadavers, and one case in which an accessory duct entered on the left side. All of those previously reported were on the right side.

More recent reports of accessory bile ducts were made by Neuhof and Bloomfield[21] (1945) and Macpherson[16] (1951). The latter reported an autopsied case of congenital hypoplasia of the gallbladder with a "cyst-hepatic duct" entering the fundus of the gallbladder. The former reported two cases of "cholecystohepatic duct," and concluded that ligation of small accessory ducts is safe in most cases, and is attended by less morbidity than leaving them open to drain. The latter alternative was felt to be possibly indicated when there is stasis and infection in the liver segment drained by the duct.

In 1916 Schachner[25] reviewed anomalies of the gallbladder and bile passages and predicted that refinements in operative and diagnostic technic would demand detailed knowledge of anomalies.

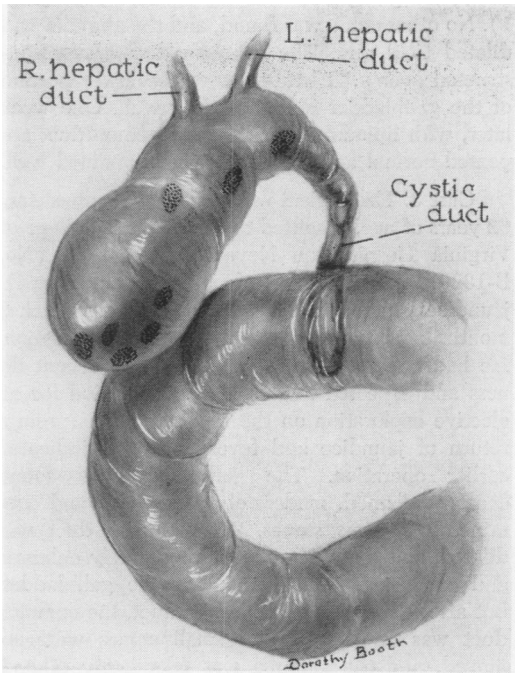

FIG. 4. Right and left hepatic ducts entering gallbladder.

The cystic duct abnormalities, as reported in the literature, consist mainly of modes of union with the hepatic ducts. In 100 autopsy specimens Eisendrath[9] found a normal angular union in 75 per cent. Variations included the short and long parallel, and the spiral types with relation to the common hepatic duct and junction with the cystic duct to form the common bile duct. Rarer variations in junction of the cystic duct have been reported recently by Leiter[15] (1947), Hager[12] (1952), and Rachlin[24] (1951), all of whom reported cases in which the cystic duct entered the right hepatic duct.

ABNORMAL DUCTS AT OPERATION

The most frequent of the abnormalities, and fortunately the least serious, are the accessory ducts. Practically all of these ducts arise from the right lobe of the liver, and most of them empty into the gallbladder, with a few entering the cystic duct. These ducts vary from minute structures to considerable size and may, as noted above, be the entire right hepatic duct. These accessory ducts can be easily divided when dissecting the gallbladder from the liver whether the dissection is made from the fundus downward, or from the cystic duct upward. The division of small ducts can be readily overlooked, and may not be suspected until there is drainage of bile through the wound. Such an extravasation of bile has caused trouble when the wound was closed without drainage. When the divided duct is recognized it should be ligated, provided it is small and the operator is sure that its output is not important. If the duct is large, however, it should be anastomosed, when possible, to the cystic duct, the common duct, or the duodenum. Such an occurrence was reported by Wapshaw[28] where he was able to anastomose a large accessory hepatic duct, which had been divided, to the remnant of the cystic duct.

The abnormalities of the major ducts can lead to much more serious trouble. The right hepatic duct may open into the gallbladder or the cystic duct in such a fashion that careful and adequate dissection alone will prevent the division of the only outlet from the right lobe of the liver. Emptying of the cystic duct into the right hepatic has been reported by Daseler[7] in three of 500 cases, and by Thompson[27] in one of 100 cases, both at autopsy. This abnormality should not cause trouble.

The cystic duct may be entirely absent, as will be reported in two of our cases. Each of these had a large stone bulging from the gallbladder into the common duct, and this stone probably gradually obliterated the duct (Fig. 1) rather than formed where the duct was congenitally absent.

Great dilatation of the common duct may result from stone or stricture in its lower end and appear as a cyst.

CASE REPORTS

Case 1.* This patient was a 33-year-old colored female admitted to St. Philip Hospital on De-

* Case 1 was a patient of Dr. Carrington Williams, Jr., and was included with his permission.

cember 7, 1948 (No. B-57641), complaining of severe, acute upper right abdominal pain. She had had previous episodes of such pain. On admission her temperature was 101.6°, and there was tenderness and muscle spasm in the epigastrium. The leukocyte count was 19,700; serum bilirubin, 0.6 mg. direct and 1.2 mg. total. Her temperature rose shortly to 103°, and then to 105° following a chill. She was explored with the diagnosis of acute cholecystitis. The gallbladder was small, buried in the fissure of the liver, and involved in acute inflammation which appeared to have developed on an old process. There was such a dense inflammatory reaction about the common duct that the dissection of the gallbladder from the liver was made from above downward. What appeared to be the cystic duct was clamped, divided and ligated, and the gallbladder removed. Bile was then discovered coming from the hilum of the liver, and dissection revealed a large duct opening, evidently the common hepatic duct. The gallbladder was opened and the two large stones present were located, one in the small fundus and the other in the common duct, with complete absence of the cystic duct (Fig. 1). It was then evident that in removing the gallbladder the common bile duct had been divided below, and the common hepatic duct above. Fortunately, a large stump remained. Probing revealed that this was the common hepatic, for both the right and left hepatics were identified. An anastomosis was made between the proximal divided duct and the duodenum. The patient made a good recovery. She was examined recently and found to be in excellent condition, having had no symptoms related to the biliary tract during the 6 years since operation.

A case similar to this was reported by Stetten.[26]

Case 2. This patient was a 58-year-old female admitted to Stuart Circle Hospital on March 17, 1954 (No. 27723). She complained of severe, upper right abdominal pain, nausea, vomiting and fever. She had previously had similar, but milder attacks. On admission mild icterus was noted, and the diagnosis was cholelithiasis and choledocholithiasis. During several days of observation the jaundice increased and the fever continued, so exploration was advised. At operation the gallbladder was found to be thick walled, small, densely adherent to the duodenum, and molded over a stone 4.5 x 2.5 cm. in size. The adherence to the duodenum was evidently the prelude to extrusion of the stone into the bowel. The gallbladder was separated from the duodenum, and a mass of adhesions found about the ducts. The gallbladder was opened, the large stone removed, and the ducts explored through this opening. It was found that there was no cystic duct, so that the stone occupied at the same time the small contracted gallbladder and the common duct (Fig. 1). No other stone was found, and the ampulla was dilated to 9 mm. The common duct was reconstructed over a "T" tube, using the lower portion of the gallbladder for its anterior wall. One week later, with lipiodol injection, the common duct appeared normal and the patient has remained well.

Case 3. This patient was a white male physician, 62 years of age, admitted to the Medical College of Virginia Hospital on November 11, 1953 (No. B-132073), complaining of upper abdominal pain, jaundice and fever. He had had a similar attack 6 months before, and again 3 weeks before admission. He had apparently recovered from this recent illness and expected to return to the hospital for an elective exploration on the bile tract. This prompt return of jaundice and fever, however, indicated earlier operation. The gallbladder was found acutely inflamed, moderately contracted, and contained numerous stones. The common duct was dilated, and the pancreas was moderately enlarged and congested. Before removing the gallbladder, but after dissection of the cystic duct, the common duct was opened, several small stones were removed, and the ampulla was adequately dilated. The cystic duct was dissected upward to the gallbladder and the vessels were identified but not clamped when it was discovered that two large accessory ducts opened into the gallbladder. These were considered too large for ligation (Fig. 2). The patient was quite fat, and in as much as preservation of the accessory ducts seemed necessary, no dissection along the common hepatic duct was done. The condition of the pancreas demanded a free flow of bile, so in spite of removal of stones from the common duct and dilatation of the ampulla it seemed important to avoid leaving any of the gallbladder to drain through the common duct. It was therefore decided to open the gallbladder, remove the stones, and anastomose the fundus of the gallbladder to the duodenum. The opening in the common duct was sutured and the anastomosis made. The patient has been very well since the operation.

Case 4. This patient was a 57-year-old male admitted to Stuart Circle Hospital on February 22, 1954 (No. 42514), complaining of acute severe upper abdominal pain, nausea, vomiting, fever and rigidity of the abdomen. The diagnosis was acute pancreatitis, and this was supported by serum amylase of 145 units. He recovered promptly from this illness. Eighteen years previously he had phlebitis in the lower extremities, which caused pulmonary emboli and infarctions. Eight years before he had roentgenologic evidence of multiple gallstones, and had had several mild attacks of colic. He had then remained well until 8 weeks before

admission, when he had a moderately severe anterior myocardial infarction from which he made a good recovery.

He was treated with epidural injection of novocain and supportive measures, and made a prompt recovery. He had had no jaundice, but was advised to return for elective cholecystectomy and exploration of the common duct. He returned to the hospital for this procedure on April 4, 1954. At operation the gallbladder was found to be considerably thickened but not contracted, and it contained numerous small and large stones. The ampulla of the gallbladder and the cystic duct were dissected. The duct was quite long and had a spiral course, so that it entered the common duct on its posterior surface and rather low down. Near the gallbladder a stone was impacted in the duct and just below this an aberrant hepatic duct of small caliber entered the cystic duct (Fig. 3). It was thought wise to preserve this duct; the cystic duct was therefore divided and ligated proximal to this point. The common duct was then opened, several small stones were removed from the ampulla, the entrance of the cystic duct on the posterior wall was identified, and the ampulla was dilated. The caliber of the common duct was rather small, and the ampulla could be dilated only to 5 mm. The postoperative cholangiogram suggested a stone remaining in the ampulla, but this deformity disappeared after three irrigations with chloroform and ether (Best[2]). He has had no complaint since leaving the hospital.

Case 5. This patient was a 40-year-old male, admitted to Stuart Circle Hospital on October 14, 1945 (No. 03993), with the diagnosis of carcinoma of the stomach. This was evidenced by an annular deformity at the pylorus demonstrated by roentgenologic examination. He had been in the hospital one year previously because of upper right abdominal pain thought to be due to gallstones. At exploration the deformity of the stomach was found to be due to an inflammatory mass about the gallbladder, to which the stomach was densely adherent. The gallbladder was dissected out with great difficulty and what appeared to be a normal cystic duct was clamped, divided and ligated. On account of the dense adhesions the common duct was not visualized. The gallbladder was dissected upward and removed. A very small duct, thought to be accessory, was encountered; it was so small that it was divided and ligated. Bile was then found coming from the hilum of the liver, but only a very small duct could be found. This was not ligated. It was hoped that the common duct lay hidden in the scar tissue, and the patient's condition had deteriorated so that termination of the operation was imperative. His convalescence was stormy; bile drained through the wound and the stools were acholic. Gradually the external drainage of bile decreased; it returned to the stools and he was finally discharged without bile drainage but with mild jaundice. During the next year he had several periods of jaundice, fever and acholic stools. He was again explored, and the duodenum was found densely adherent to the liver. The duodenum was opened and exploration revealed one small opening discharging bile. This was dilated, a catheter inserted with end free in the duodenum, and the wound closed. He remained well and free of jaundice for several months, but during the next 8 years had recurring attacks of pain and jaundice. During this time the liver became markedly enlarged and nodular. Because of the increasing frequency and severity of these episodes he was again explored on March 9, 1954. The liver showed marked cirrhosis. The duodenum was mobilized and a long search was made for the common duct, which might have been overlooked previously; there was no duct present. The duodenum was opened and a small duct was found draining bile. This was dilated and the duodenum and the wound closed. There has been no jaundice since but the liver is still large. It would seem then that the two small ducts discovered entering the gallbladder at the first operation were actually the hepatic ducts, and that the cystic duct carrying all the bile entered the duodenum (Fig. 4). The combination of this unusual anomaly and the severe inflammatory reaction about the area resulted in disconnecting the small abnormal ducts between the liver and the duodenum. The small channel later established has been inadequate, so that he has had repeated periods of jaundice and has suffered severe liver damage.

COMMENT

Four of these patients with abnormal ducts had acute inflammation involving the biliary tract in addition to chronic inflammatory lesions. The fifth (Case 4) had pancreatitis. It would seem, therefore, that the abnormal arrangements were factors in the development of inflammation and stones. In the presence of dense adhesions the discovery of the abnormality is difficult, and serious damage to the continuity of the biliary tract can result in spite of great care. Under such circumstances it would seem wise to limit dissection to the parts involved because further exploration might result in damage to the ducts or to vital blood vessels. For example, in Case 3 it would have been interesting to see the hepatic ducts, but no

good for the patient would have been accomplished by dissecting them, and damage to the cystic or hepatic vessels might have resulted.

In Case 1 the divided common hepatic duct was successfully anastomosed to the duodenum and the patient has been well for six years. This is the ideal outcome for such a catastrophe. Case 2 was similar, but the duct arrangement was found by exploration inside the gallbladder.

In Case 3 the anastomosis of the gallbladder to the duodenum seemed the only way to preserve the accessory ducts, and to avoid the danger of further formation of stones in the gallbladder which would probably pass into the common duct and cause further damage to the pancreas.

In Case 4 the accessory duct was small and might have been ligated safely, but its preservation with the cystic duct was simple. We cannot agree with those who attribute residual symptoms to remnants of cystic ducts, and routinely prefer to leave a small portion of the cystic duct as advocated by Coller.[6]

The tragic result in Case 5 might have been avoided by dissection of the gallbladder from the fundus downward but the ducts were so small and the adhesions so great that it is likely they would have been damaged in spite of this.

SUMMARY

Abnormalities of the bile ducts are rare but occur with sufficient frequency to be of importance and should be kept in mind by the surgeon. Damage to abnormal ducts should be repaired immediately. When they are discovered, they should be preserved unless they are very small accessory ducts, and before these are ligated other adequate ducts must be demonstrated. Abnormal ducts and acute inflammation are often present together. This combination may easily result in serious damage to the bile ducts at operation, and accounts for a considerable number of damaged ducts.

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DISCUSSION.—DR. CHERYL HART, Durham, N. C.: I would like to make one comment in support of Dr. Carrington Williams' observation on the stone in the cystic duct protruding into the common duct. I have had experience with two such cases that I remember quite well. Both patients were jaundiced. One was the mother of one of our house staff who had had for years a large radiopaque silent stone, 5 to 5 cm. in its greatest diameter. She then developed gradually progressing painless jaundice, and was thought to have a carcinoma developing around the stone. At operation we could not explore in the region of the common duct because of the inflammatory reaction. It seemed almost equally hopeless to approach it from the fundus. As we dissected down the gallbladder we came to a vascular pattern that suggested the blood vessels running along the common duct and supplying the duodenum. We then opened the fundus, removed the stone, and found a structure of equal size extending from the fundus of the gallbladder to the pancreas. The blood supply over the lower end of this structure was the blood supply to the duodenum. The stone had extended from the fundus of the gallbladder through the cystic duct area and down the common duct to the head of the pancreas. This explained why we could not explore between the duodenum and the gallbladder and find the common duct. With the gallbladder open and the stone removed we found an inverted T-shaped space, each branch being 3 to 5 cm. in diameter. The upper branch, the common hepatic duct, the lower branch and the common duct were separated by at least 4 to 5 cm. where they merged with the dilated cystic duct and the gallbladder which formed the upright of the T. A probe could be passed upward into the right and left hepatic ducts and downward into the duodenum. We then cut off the excess part of the gallbladder and closed the resultant opening to construct a much dilated duct. She made a satisfactory recovery.

The other patient also had a large stone, 2 to 3 cm. in diameter, lying in a greatly dilated cystic duct and protruding into the common duct to produce a common duct obstruction. The result was again a roughly T-shaped ductal system composed of the common hepatic duct, the common duct, and the dilated cystic duct and gallbladder forming the upright part of the inverted T.

I should like to ask Dr. Mahorner if he has had any experience with chronic or acute pancreatitis as a complication. The only patient I have lost from an acute fulminating pancreatitis following common duct exploration was one who had had his gallbladder removed previously. On common duct exploration in the presence of jaundice, nothing could be found on transduodenal exploration. A probe in the common duct could be seen with only a thin layer of mucous membrane over it, but no ductal opening could be found. A simple incision was made through the mucous membrane from within the duodenum, and was enlarged to give an adequate opening. The patient died within 36 hours of a fulminatingly acute pancreatitis. I do not run into such cases as commonly as he does, but I wondered if he had had experience with acute fulminating pancreatitis from common duct manipulation.

DR. ROBERT M. MOORE, Galveston, Texas: I have enjoyed these papers and wish to speak briefly in relation to Dr. Williams' paper dealing with congenital common duct cyst. This is a relatively rare condition, and I am sure I have seen more than my share of cases since personally or in association with others I have had care of six patients with this anomaly. In one patient the cyst was discovered only when the child suffered a rupture of the cyst in a playground injury.

The usual advice is that these immense cystic dilatations of the common duct are best treated by direct anastomosis of the cyst to the duodenum, and without doubt many surgeons have had excellent results using this method. I have seen two instances, however, in which for some unknown reason such an anastomosis seemed to result in a