Rare Anomalies of the Extrahepatic Bile Ducts *

JACOB RABINOVITCH, M.D., C.M., PHINEAS RABINOVITCH, M.D., C.M., HARRY J. ZISK, M.D.

Brooklyn, N. Y.

From the Adelphi Hospital, Brooklyn, and the Mount Royal Hospital, Montreal

ALTHOUGH the literature shows a constant increase in the number of reported cases of anomalies of the bile ducts, the condition is still regarded as not too common an occurrence. No doubt many cases are overlooked and others are not reported; it is quite possible, therefore, that the incidence of these anomalies will increase as the condition becomes more widely known. The finding of these anomalies always attracts considerable attention first, because of their extraordinary diversity and mode of development [2, 5] and second, because of the technical difficulties and dangers incidental to their presence.

The vast majority of anomalies of the bile ducts heretofore described in the literature were incidental findings observed during the performance of operations upon the gallbladder, or were discovered accidentally during routine autopsy examination. Beaver [1] made careful dissections of 57 cadavers and found accessory bile ducts in 8.7 per cent of the cases. Flint [3] has made postmortem dissections on 200 consecutive subjects with the purpose of determining the incidence of anomalies of the bile passages among such cases. He discovered 29 cases of accessory bile ducts and 28 instances where there was no supraduodenal common duct at all, the union occuring at a point anywhere from behind the upper border of the duodenum to the part embedded in the pancreas. In three cases the only representative of the common duct was that part which lies in the wall of the duodenum. Flint emphasized the importance of recognizing these anomalies at the time of operation and the ensuing dangers if left undetected. From these considerations and the observations which we have made it is quite clear that anomalies of the biliary tract are of sufficient importance that no surgeon who operates upon the gallbladder and bile ducts can afford to ignore their existence.

While some anomalies of the bile ducts are relatively common, others are unusually rare. The cases brought to our notice recently are of exceptional interest because the anomalies involved the major extrahepatic bile passages and presented anatomic variations which to our knowledge, have not been described before. It is the purpose of this paper to record our findings in three of the rarer and perhaps more important anomalies of the bile ducts which we have observed during the performance of cholecystectomy.

CASE REPORTS

Case 1. L. F., a Cherokee Indian, aged 62 years, was admitted to the Adelphi Hospital on March 12, 1954 with the complaint of severe pain in the right abdomen, nausea and vomiting, of 48 hours' duration. She had had similar but milder attacks on previous occasions; these attacks were said to be due to gallbladder disease. Cholecystographic examination before admission revealed the presence of calculi within the gallbladder. The present attack was the most severe she had had; it was not relieved by the administration of opiates.

On admission there was a deep icterus of the skin and sclerae. The abdomen was soft, not distended and somewhat tender to palpation in the right upper quadrant; the gallbladder could not be felt. The clinical impression was that the patient was suffering from an acute exacerbation of a chronically inflamed gallbladder, probably associated with a block of the common bile duct by a stone.

* Submitted for publication September 23, 1955.

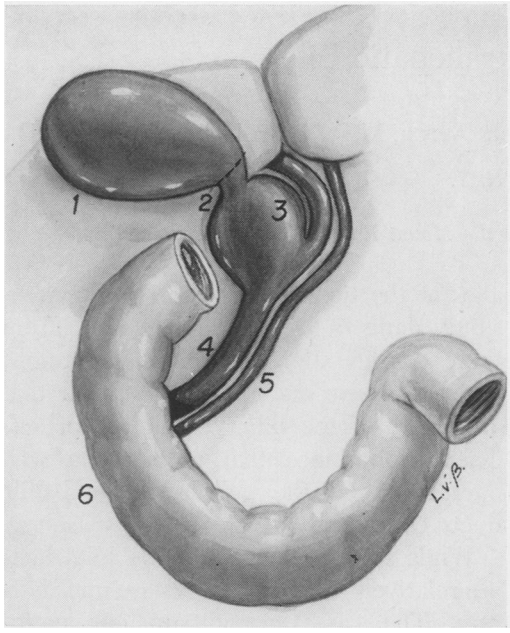

FIG. 1. Drawing showing cystic dilatation of the right hepatic duct. Also, the right and left hepatic ducts in this case do not unite to form a common hepatic duct but pursue an independent course and enter separately into the duodenum. (1) Gallbladder; (2) cystic duct; (3) cystic dilatation of the right hepatic duct; (4) right hepatic duct; (5) left hepatic duct; (6) duodenum.

At operation, the gallbladder appeared chronically inflamed. The wall was thickened and adherent to adjacent structures by well formed bands of adhesions; numerous calculi could be palpated within its cavity; they averaged from 0.5 to 3 cm. in diameter.

Dissection of the gallbladder proceeded from the fundus downwardly towards the cystic duct. The latter was widely dilated and very short; it measured approximately 0.5 cm. in length and 1 cm. in diameter. At the terminal portion of this duct a large cystic swelling was felt and partly seen in the position of the common bile duct. Further dissection revealed that the cystic swelling was a saccular or diverticular dilatation of the right hepatic duct. The latter was very much dilated throughout its length and contained numerous calculi. As the dissection and exposure of the ducts continued it became evident that the common bile duct in this case was the continuation downwards of the right hepatic duct. There was complete failure of union between the two hepatic ducts. The cystic duct joined the right hepatic duct to form a common bile duct; the point of union of these two ducts occurred at the summit of the cystic dilatation (Fig. 1).

It is of interest to note in this case that each hepatic duct maintained its own course from the porta hepatis to the duodenum and each opened through a separate orifice into the duodenum. This was ascertained when the duodenum was opened and two openings were found, one leading into the right hepatic duct and the other into the left hepatic duct. In addition, the right hepatic duct was very much dilated and contained many stones, while the left hepatic duct was of normal caliber and remarkably free from calculi. This unusual anomaly of the ducts is well depicted in Figure 1.

After the gallbladder was mobilized and the related structures identified and clearly exposed, the gallbladder was opened and its contents evacuated; it was then divided at the level of the cystic duct. The common bile duct was thoroughly explored and its contents emptied through the opened and dilated cystic duct. Numerous calculi of various size and of the same appearance and composition as those found in the gallbladder were removed from the duct. An adequate segment of the cyst wall was excised and the duct was reconstructed, care being taken not to impinge on the lumen of the duct. A T tube was then inserted through an opening left in the duct and the abdominal wound was closed in layers.

The postoperative course was unusually smooth and uneventful. The jaundice began to clear after several days of bile drainage and the T tube was removed on the eighth postoperative day; she was discharged from the hospital 2 days later. At the time of writing, almost 2 years since the date of operation, the patient is reported well and free from symptoms.

Case 2. M. S., a 54-year-old man, was admitted to the Adelphi Hospital on February 11, 1955 complaining of severe abdominal pain, nausea, vomiting, chills and fever of 3 days' duration. Nine months previously he had been operated upon in another hospital for gangrenous cholecystitis, at which time the gallbladder was drained but not removed. One month prior to the present admission he was acutely ill with chills and fever and was treated for virus pneumonia which lasted 3 weeks.

On admission a mild icterus was noted in the skin and sclerae. The abdomen was somewhat distended and quite tender and spastic in the gallbladder area. The latter could not be definitely palpated mainly because of the obese abdomen and the resistance offered by the patient. The clinical impression was that the patient was suffering from acute cholecystitis; the possibility of obstruction of the common duct was also entertained because of the existing jaundice and the clinical history of chills and fever.

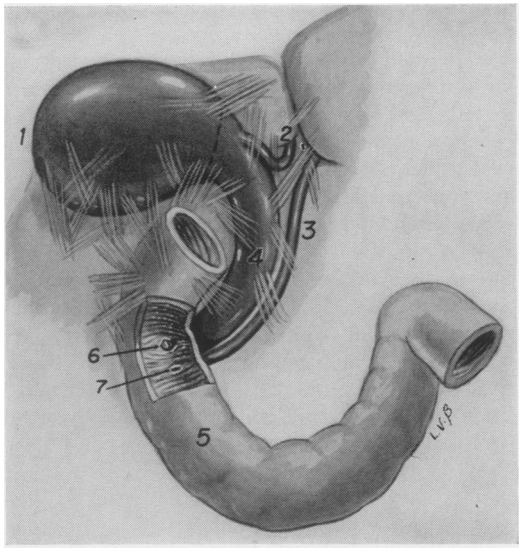

FIG. 2. The right hepatic duct in this case terminates in the gallbladder. Another dilated duct leaves the gallbladder and runs downwards to enter the duodenum independently of the left hepatic duct. (1) Gallbladder; (2) right hepatic duct; (3) left hepatic duct; (4) continuation downward of right hepatic (or cystic) duct; (5) duodenum; (6) orifice of right hepatic duct; (7) orifice of left hepatic duct.

During the first few days of observation in the hospital the jaundice appeared to deepen although the chills, fever, and abdominal pain and tenderness tended to subside. It was decided to operate during this quiescent period so as to prevent a possible recurrence of the attack.

At operation the gallbladder was found hidden from view by a veil of adhesions. When these were cleared and the gallbladder was brought to view, the latter was found to be somewhat thick walled and to contain a number of small calculi. Because of the distorted anatomy created by the numerous adhesions following the first operation, it was thought wise to begin dissection of the gallbladder at the fundus. At about the region of Hartmann's pouch a good sized duct was noted to enter the gallbladder. At first it was taken to be a large accessory hepatic duct. However, when the entire length of the duct was delineated it was noted that the duct was the sole channel draining the right lobe of the liver. In other words, this structure represented the right hepatic duct which began in the porta hepatis and terminated in the gallbladder. Another widely dilated duct was found to leave the gallbladder and run towards the duodenum. It was difficult to say whether the latter duct was the cystic duct or whether it was the continuation downwards of the right hepatic duct with the gallbladder forming an intermediary structure. Unlike the normal anatomical structure of the cystic duct it lacked the spiral configuration and the valves of Heister. We were more inclined to believe that this duct represented the downward continuation of the right hepatic duct; it opened into the duodenum through a separate orifice and did not unite with the left hepatic duct. The latter was of normal size and appearance; it ran parallel to the right hepatic duct and opened separately into the duodenum. In this case, as in the previous one, there was failure of union between the two hepatic ducts, each duct pursuing its own course and each having its own duodenal orifice. This anomaly is well illustrated in Figure 2.

The gallbladder was opened and divided just above the point of entry of the right hepatic duct as indicated by the dotted line in Figure 2. The dilated duct was explored through the opened gallbladder. No stones could be palpated within its lumen, nor were there any to be found after a careful search with the aid of stone forceps and saline irrigations of the duct. However, when a probe was passed into this duct it met with considerable resistance at its distal end, preventing the probe from entering the duodenum. Because of the marked dilatation of the duct, the existing jaundice, and failure of the probe to pass all the way through the duct, it was decided to explore the distal end of the duct through a transduodenal approach. When the duodenum was opened two separate openings were noted adjacent to one another. One of these orifices led into the right hepatic duct and the other into the left hepatic duct. The orifice of the former duct was occluded by a small impacted stone which resisted extraction. The orifice of the left hepatic duct was patent and permitted the passage of a probe along its entire length. A sphincterotomy of the right hepatic orifice was done and the stone was readily removed; the duodenum was then closed along its transverse diameter. A T tube was inserted into the right hepatic duct, the excess of gallbladder removed, and the remaining segment was utilized to reconstruct the opened duct.

The patient made a good postoperative recovery. The jaundice subsided rapidly after several days of bile drainage and he was discharged from the hospital on the tenth postoperative day. The patient has been free from symptoms since the operation.

Case 3. B. R., a 58-year-old woman, was admitted to the Mount Royal Hospital on June 25, 1951, complaining of colicky pain in the right upper abdomen and vomiting. She had had a cholecystogram done several years previously and was told that she had gall stones. In the past she suffered from intermittent attacks of abdominal pain which were relieved by rest and the application of heat

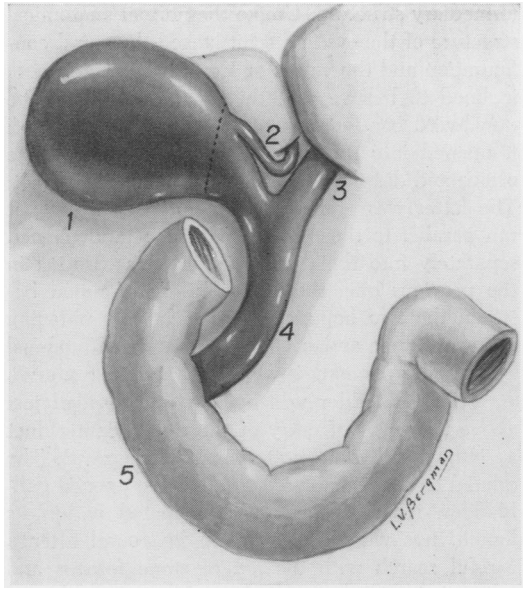

FIG. 3. Drawing showing the right hepatic duct terminating in the gallbladder and the cystic duct joining the left hepatic duct to form a very dilated common bile duct. (1) Gallbladder; (2) right hepatic duct; (3) left hepatic duct; (4) common bile duct; (5) duodenum.

to the abdomen. The present attack was very severe and was not relieved by heat or by the administration of opiates.

On admission to the hospital the patient appeared to have considerable pain. The temperature and pulse were normal. The gallbladder could not be palpated. The abdomen was soft, not distended and only slightly tender in the right subcostal area. There was a noticeable icterus of the skin and the icterus index was elevated to 25. The leukocyte count was within normal limits.

During the first 24 hours stay in the hospital the patient showed very little change in her general condition except that the jaundice appeared to deepen; so operation was advised.

Laparotomy disclosed a very much enlarged gallbladder with a thickened wall and a cavity containing a good number of calculi. Dissection of the gallbladder was begun at the fundus and carried downwards. At about the lower third of the gallbladder a good sized duct was found entering its cavity. This was first taken to be an enlarged accessory hepatic duct. Further dissection revealed, however, that this structure represented the right hepatic duct; it ran from the porta hepatis to the gallbladder where it terminated. The cystic duct was very short, measuring only a few millimeters in length; it joined the left hepatic duct by a wide orifice to form a common bile duct. The latter was markedly dilated and contained several faceted stones. Similar stones were also found within the gallbladder.

The gallbladder was opened and divided just above the point of entry of the right hepatic duct as indicated by the dotted line in Figure 3. The common duct was then explored through the open cystic duct; several stones were found in the duct and were removed. The excess of gallbladder wall was excised and the remainder closed to reconstruct a rather dilated common duct. A T tube was inserted into the common duct and the abdominal wall was closed in layers. Aside from a minor wound infection, which cleared after a few days of drainage, the patient made a good recovery. At the time of writing this paper, more than 4 years since the operation, the patient is reported well and free from symptoms.

EVALUATION

Accessory hepatic ducts are among the most common anomalies of the bile passages. The great majority of these aberrant ducts originate in the right lobe of the liver and drain into the gallbladder. They are frequently of small size and are often inadvertently divided during the performance of cholecystectomy. These ducts, although of little surgical import, may sometimes give rise to serious complications if divided and left to drain freely into the peritoneal cavity. When very small ducts are cut across, generally no ill effects result, except that there may be prolonged drainage of bile through the abdominal wound. The surgeon will act wisely, therefore, by ligating these ducts before dividing them, especially when he chooses to close the abdomen without drainage. When an accessory duct assumes large dimensions it must be well protected against injury or serious trouble may result.

Anomalies of the gallbladder are extremely rare. Gross [4] in reviewing 148 cases of congenital anomalies of the gallbladder found 28 cases of double gallbladder, in each of which there were two separate bladder cavities and two separate cystic ducts. We have observed two anomalous conditions of the gallbladder (to be reported later). In one instance the gallbladder was partitioned off by a median

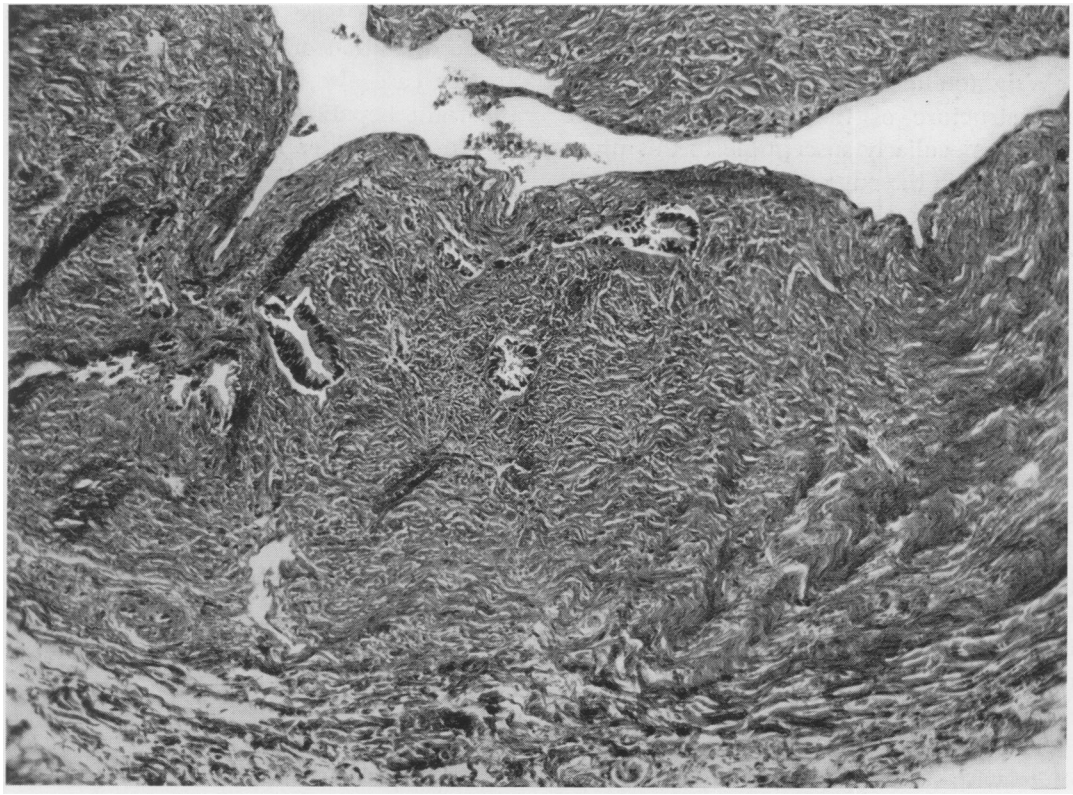

FIG. 4. Normal bile duct of man. This section was taken through the common bile duct. It shows that the wall of the duct is largely composed of connective tissue and very few muscle and elastic fibers. × 150.

septum into two distinct cavities, each cavity having its own cystic duct. The latter joined together to form a common cystic duct which joined the common bile duct. In the second case two separate gallbladders were found adjoining one another, each having its own cystic duct which united separately with the common hepatic duct.

The three cases herein reported illustrate some of the rarer anatomic variations which may occur in the major bile ducts. The cystic dilatation of the right hepatic duct noted in case 1 is of much interest. This anomaly has often been referred to in the literature as congenital cystic dilatation of the duct.

Various hypotheses have been advanced to explain the development of this anomaly. According to Wrighton [7] the anatomic structure of the common bile duct is such that it lends itself readily to the force of stretching and dilatation. Histologic study of the wall of the duct shows that it is composed largely of connective tissue and a small amount of elastic and muscle fibers. This causes the duct to be imperfectly elastic and therefore no great increase in force is necessary to stretch it permanently. If the duct has walls of normal constitution but has attained, as a result of congenital defect, a diameter several times the normal, the material of the wall will be subject then to several times the normal stress and we can expect it to be stretched permanently even by physiologic biliary pressure. If in addition there is a rise of biliary pressure, then the stretching of the wall will be so much greater. In the case under discussion the duct was occluded by stones; this undoubtedly raised the intrabiliary pressure and was responsible, in

part at least, for the cystic dilatation of the duct (Fig. 4).

Wrighton also notes that the situation and structure of the common bile duct make it peculiarly susceptible to this process. It lies in the edge of the lesser omentum unsupported by solid structures and in its walls it contains no elements which can be relied upon under the stress of distension to hypertrophy and resist. It will therefore yield readily to any force which exerts pressure upon its wall.

It may be stated that other factors beside those mentioned may account for this anatomic malformation of the duct. Thus, inflammatory reactions in and about the duct may through traction and distortion produce a similar picture. Such a condition would be analogous to the pulsion or traction diverticulum seen in other hollow organs such as the esophagus, intestine, etc.

The failure of the right hepatic duct to unite with its companion duct in case 1 is of added interest. Anomalies of this kind have rarely been observed and recorded. Also, the presence of stones in the right hepatic duct and their complete absence in the left hepatic duct is quite significant. Since the duct related to the gallbladder contained stones while the one not related to it was empty, it immediately raises the question as to whether all stones found in the bile passages are of gallbladder origin.

Case 2 shows another unusual and rather rare anomaly of the bile ducts. In this case the right hepatic duct ran from the porta hepatis to the gallbladder where it appeared to terminate. The duct leaving the gallbladder was a dilated structure and emptied independently of the left hepatic duct into the duodenum. It was difficult to determine at the time of operation whether the duct leaving the gallbladder represented the cystic duct or whether it was the continuation downward of the right hepatic duct with the gallbladder forming an intermediary structure. If the latter assumption is true, then the gallbladder may be regarded here as a diverticulum or cystic dilatation of the right hepatic duct. In this case also, as in the previous case, the right hepatic duct which was related to the gallbladder had an impacted stone in its duodenal orifice, while the left hepatic duct which was not related to the gallbladder, was remarkably empty. The question as to the origin of biliary stones may be again raised in this case.

In the third case the right hepatic duct pursued a course similar to that seen in case 2; it ran from the porta hepatis to the gallbladder where it terminated. The cystic duct in this case was short and widely dilated; it joined the left hepatic duct to form a common bile duct. A somewhat similar form of anomaly of the ducts was described by Milroy.[6] In our case both the gallbladder and the common bile duct were found to contain stones.

SUMMARY

Three cases are reported with rare anomalies of the major bile ducts. The cases raise some points of interest in the etiology, detection and surgical management of the condition. Damage to any of these ducts in the course of operation if undetected and left unrepaired, may lead to very serious complications.

BIBLIOGRAPHY

1. Beaver, M. G.: Variations in the Extrahepatic Biliary Tract. Arch. Surg., **19**: 321, 1929.
2. Boyden, E. A.: Congenital Variations of the Extra-hepatic Biliary Tract. Minn. Med., **27**: 932, 1944.
3. Flint, E. R.: Abnormalities of the Right Hepatic, Cystic and Gastroduodenal Arteries and of the Bile Ducts. Brit. J. Surg., **10**: 509, 1922–23.
4. Gross, R. E.: Congenital Anomalies of the Gallbladder. Arch. Surg., **32**: 131, 1936.
5. Mentzer, S. H.: Anomalous Bile Ducts in Man. J. A. M. A., **93**: 1273, 1929.
6. Milroy, P.: An Important Anomaly of the Right Hepatic Duct and its Bearing on the Operation of Cholecystectomy. Brit. J. Surg., **35**: 383, 1947.
7. Wrighton, P.: Congenital Cystic Dilatation of the Common Bile Duct. The Australian and New Zealand J. of Surg., **23**: 110, 1953.