THE SURGICAL ASPECT OF CONGENITAL ABSENCE OF THE GALLBLADDER*

REPORT OF TWO CASES

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RARE CONDITIONS do not always attract the general interest of the medical profession as do the more common ones. There are some conditions which are thought of as rarities only because few cases have been reported. And yet we know that although many surgeons come upon very remarkable and rare cases, the information concerning these cases never gets beyond their office files. When an anomaly, hitherto considered extremely rare, is encountered almost simultaneously by two surgeons working separately in the same city, one begins to doubt if that anomaly is so rare after all. The purpose of this paper is to bring to the attention of surgeons two cases of congenital absence of the gallbladder in adults, and to consider certain practical surgical aspects of the anomaly.

There have been many theories of the causation of congenital absence of the gallbladder. Mauro,[1] in a recent paper, offers what we feel to be a reasonable explanation. As early as the second week of fetal life the hepatic diverticulum develops a ventrocaudal extension, the cavity of which is soon obliterated by proliferation of the epithelium. The extension from which the gallbladder develops is located in the mesenchymatous tissue and remains connected with the hepatic diverticulum by a pedicle from which the cystic duct later develops. The hepatic duct originates from the cranial portion of the hepatic diverticulum and the common bile duct from the caudal portion. During the third fetal week, the liver becomes more separated from the intestine and the bile ducts are thus lengthened and at the same time occluded. The common bile duct reopens during the fourth week and the hepatic duct during the fifth week. Sometime during the sixth week of fetal life, the cavity of the gallbladder reappears. It is almost impossible to determine at the operating table whether absence of a certain portion of the biliary system is due to agenesia or to atresia. This fact is especially true of the gallbladder, which represents the terminal extension and, therefore, may never have existed, or may have undergone secondary involution; on the other hand, if the terminal portion is present, the absence of an intermediate portion can be due only to atresia.

It has been generally accepted that congenital anomalies are apt to be multiple. There are numerous reports of congenital absence of the gallbladder in infants associated with atresia of the bile ducts, according to Bruchsaler[2] and Ladd;[3] this paper is not concerned with such cases. Since

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there is a possibility that one of our cases (A. O. L.) represents an intrahepatic situation of the gallbladder, we must not neglect that condition. Kehr, quoted by Walter,[4] reported three such cases, in all of which he resorted to exploratory incision of the liver and found the gallbladder, which contained calculi. In one of his cases, there was profuse hemorrhage which required packing; the patient died on the twelfth postoperative day of diffuse cholangitis and peritonitis. At operation, however, one cannot discover the presence of an intrahepatic gallbladder by any less formidable procedure, since aspiration is done blindly and the bile obtained might well come from one of the bile ducts. Mauro[1] warns against all attempts to perform cholecystectomy in such cases because of the danger of uncontrollable hemorrhage. It is interesting to note that the great majority of intrahepatic gallbladders reported have contained one or more calculi.

Gross[5] records from the literature 38 cases of congenital absence of the gallbladder, in which there was no other anomaly of the liver or biliary system. He does not include two cases observed by Whipple,[6] quoted by Sarma,[7] or the report by Danzis[8] of one case. Since the appearance of Gross's paper, five other cases have been reported (Mauro,[1] Sarma,[7] Melville,[9] Robertson[10]), bringing the total to 46.

Case reports of the anomaly have been notably lacking in the essential data. A review of the 11 complete cases reported, including our own, shows that calculi were found in the common bile duct in all, and that the duct was dilated in ten and within the limits of normal in only one. These facts agree with the finding of a proportionately large common bile duct in animals possessing no gallbladder, and also with the common observation that the biliary ducts undergo dilatation following cholecystectomy. The gallbladder fossa may be absent and the quadrate lobe indistinct in outline. Corresponding with the greater incidence of cholelithiasis in females, we find that congenital absence of the gallbladder is twice as common in women as in men. Symptoms usually take the form of a gaseous type of indigestion, with intolerance of fatty foods, and sometimes episodes of sharp lancinating pain in the right upper quadrant referred through the chest to the inferior angle of the right scapula, and occasional jaundice. Thus far, to the best of our knowledge, the diagnosis has never been made preoperatively.

The first case to be presented is that of the senior author (G. G. F.):

Case A. O. L.—A male, age 68, referred by Dr. Louis P. Hamburger, was admitted to the Union Memorial Hospital, March 26, 1941, with the complaint of recurrent attacks of upper right quadrant pain, together with fever, nausea, vomiting, and jaundice of a little more than a year's duration.

Past History.—This contains a number of interesting and important facts. At age 17 the patient had a rather severe case of typhoid fever, and at age 32 he had an attack of what was called pleurisy. At age 50 the patient had a siege of dysentery, with blood and mucus in the stools. For the 23-year period from 1910 through 1933, he had suffered with frequent prolonged attacks of hyperacidity, regurgitation, and

sharp epigastric pain, coming on three to four hours after meals. The pain would frequently awaken him at night. During this period he was put on a restricted diet on numerous occasions, and was hospitalized for treatment three separate times. The last occasion was in December, 1933, at which time he spent some three weeks in the Union Memorial Hospital under strict dietary care, with the diagnosis of duodenal ulcer. From this time, the patient had been completely relieved of all symptoms referable to his gastro-intestinal tract until the beginning of his present illness.

Present Illness.—In January, 1940, the patient first had an attack of severe right upper quadrant pain which radiated through to the back. This was associated with nausea and vomiting and a moderate degree of fever. The pain was so severe that a hypodermic was required before relief could be obtained. The following day clay-

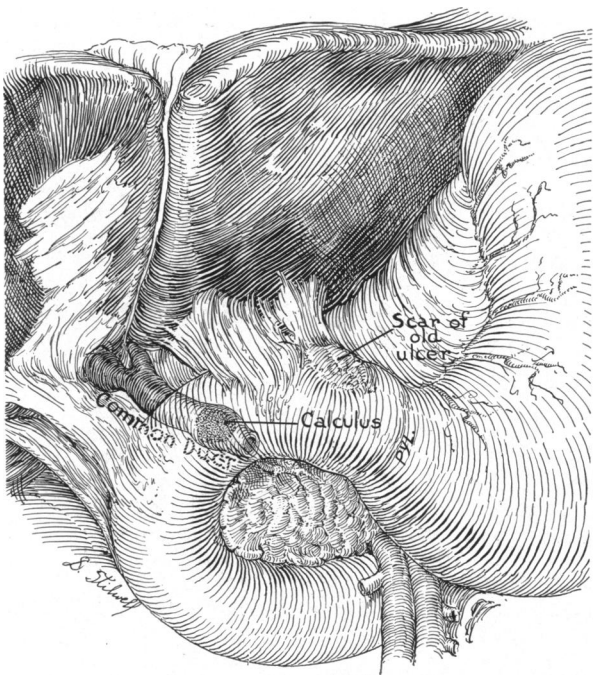

FIG. 1.—Appearance at operation (Case No. 1).

colored stool was noted and also jaundice. Since this first attack, the patient had had four or five similar episodes, the last one a few days before admission to the hospital.

Physical Examination.—The patient was in no apparent distress, but evidenced a moderate icteric tinge to the skin and sclerae. The temperature, pulse, and respirations were within normal limits. Examination of the heart and lungs was normal, and the blood pressure was 144/86. On palpating the abdomen, which was rather large, no mass could be felt and no abnormal resistance or spasm. The liver edge could be felt just below the right costal margin and there was no irregularity or abnormal firmness. Deep pressure in the whole right hypochondrium caused moderate discomfort. In other respects, the examination was normal.

Gallbladder visualization studies were carried out the second day after admission, and the radiologist reported that the gallbladder was not visualized, indicating gallbladder disease probably with cystic duct obstruction. Other laboratory tests showed a relatively normal blood picture—R.B.C., 4,360,000; W.B.C., 8,950; Hb. 98 per cent; polymorphonuclears, 83 per cent; sedimentation rate 25, and the coagulation time nine and one-half minutes. The prothrombin time was 18 seconds with the control 17

seconds. The nonprotein nitrogen was 27, blood sugar 99, and the blood cholesterol 218 mg. per cent. The icteric index was 30.0, and the blood Wassermann was negative. The urine examination was normal except for the presence of some bile and a few white blood cells in the centrifuged specimen. *Preoperative Diagnosis:* Cholecystitis with cholelithiasis and possible common duct stone.

Operation.—March 31, 1940: The abdomen was opened through an upper right rectus incision. On entering the peritoneal cavity, the omentum was found lying over the region where the gallbladder should be located. On exposing the under surface of the liver, the groove for the gallbladder was seen, but when the entire upper right quadrant was examined carefully, no gallbladder could be found. The stomach was normal, but the duodenum showed the scar of an old ulcer on the anterior superior portion. There was apparently no obstruction to the pylorus. The common duct was then visualized. Its identity was ascertained by aspiration from it of a small amount of bile. The common duct itself was not unusually distended, being at the upper limits of normal in size. When the duct was opened, it was possible to pass a uterine sound up in the hepatic ducts both to the right and to the left. When an attempt was made to pass the sound down toward the duodenum, an obstruction was met 3.5 mg. below the opening in the duct. This obstruction was caused by one small stone and a larger one, 1x1.5 cm. in diameter, somewhat irregular and black in color. This latter stone was lodged in the ampulla. After its removal, a sound could be passed into the duodenum with ease. The duct was thoroughly washed out with nomal salt solution and a No. 16 F. catheter was placed in the duct for drainage and the duct closed around this with sutures of No. 000 chromic catgut. The wound was closed in layers with catgut, except for continuous silk sutures in the skin (Fig. 1).

Postoperative Course.—Convalescence was very satisfactory until the fourth postoperative day when a hacking cough developed. This became very bothersome, and in spite of medication there were very severe paroxysms even though the chest examination remained negative. On the eighth postoperative day, after a hard cough, the patient suffered an almost complete disruption. Under pentothal sodium anesthesia, the wound was again closed, through-and-through interrupted silver wire sutures being used. Following this procedure the patient's cough for some reason stopped, and the rest of his convalescence was uneventful except for minor difficulties in voiding at times. He was discharged from the hospital May 12, 1940, with his wound healed, and ate a normal diet without any discomfort. He was last seen a few weeks ago, and showed only a small weakness at the upper end of his incision as a result of his disruption, but asymptomatic.

The second case is that of the junior author (J. K. O.):

Case C. A. G.—White, female, age 69, was admitted to the Hospital for the Women of Maryland, February 5, 1941, complaining of a dull ache in the right upper quadrant following exertion. The pain was steady and was accompanied by dyspnea; usually, but not always, the pain was relieved by resting or by the administration of spirits of niter. The pain often extended to the substernal region and sometimes radiated through the chest to the inferior angle of the right scapula. Attacks lasted from a few minutes to several hours and were often associated with nausea and vomiting and sometimes chills but never jaundice. She also noted ankle edema in the evening. The pain had never radiated to the left shoulder or down the left arm. At the time of an attack she complained of palpitation. She avoided greasy foods because of postprandial distress in the form of gaseous eructations and heartburn. All the foregoing symptoms had been recurring for two years prior to admission. She was moderately constipated and thought she had passed acholic stools. There has been no weight-loss. There was no history of typhoid fever.

Physical Examination.—Blood pressure varied between 186/80 and 150/40. The patient was orthopneic and required three pillows. There was pitting edema of both lower legs. The lungs were clear to percussion and auscultation. The heart was not

enlarged to percussion and sounds were clear, without murmurs. An occasional ventricular extrasystole was heard. There was moderate tenderness over the entire right upper abdominal quadrant, but no spasm.

Special Studies.—A gallbladder roentgenologic series was done twice, without visualization of the gallbladder. Electrocardiograms showed evidence of myocardial disease. Urine negative. Hemoglobin was 82 per cent (Sahli); fasting blood sugar 85 mg. per cent. Blood nonprotein nitrogen 30 mg. per cent. Blood Wassermann negative. The van den Bergh test showed delayed reaction.

Preoperative Course.—It was felt that the patient, unquestionably, had heart disease, and biliary disease as well. The plan was to treat her biliary disease conservatively and to concentrate on her cardiac pathology, but, on February 9, 1941, she had an attack of severe pain in the right upper quadrant; there was marked tenderness and

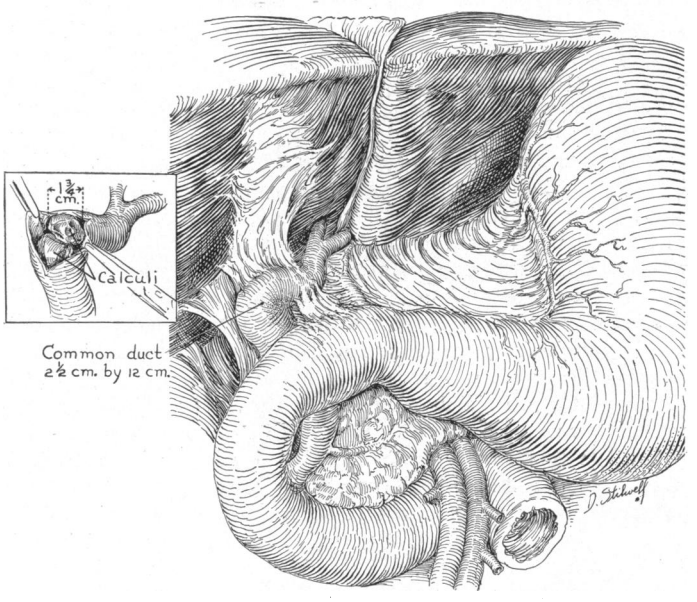

FIG. 2.—Appearance at operation (Case No. 2).

moderate spasm over the region, and the temperature rose to 103.2° F. by mouth. She pleaded for some form of relief, and we all agreed that operative intervention was indicated, even though it involved considerable risk in the face of obvious heart disease. Therefore, on February 10, 1941, a course of aminophylline gr. 1.5 t.i.d. and coramine gtt. 15 t.i.d. was begun. She had still another attack on February 12, 1941.

Operation.—February 12, 1941: The abdomen was opened through a right subcostal approach, and the liver was found enlarged to three fingers breath below the costal arch; it had the classic appearance of chronic passive congestion. Lying in a very shallow groove of the liver, where, ordinarily, the gallbladder fossa is located, was a mass of tissue surrounded by light adhesions and containing two calculi. The adherent structure was dissected free and was found to be a considerably dilated common bile duct. There was no evidence anywhere of a vestige of the gallbladder or cystic duct. The common bile duct was incised and two calculi, measuring 1.5 cm. in diameter, were removed. Thorough exploration of the dilated hepatic ducts and common bile duct was carried out with sounds and a catheter, and no other calculi were encountered. The quadrate lobe was indefinite in outline. The pancreas felt normal throughout. The stomach, pylorus, and duodenum all appeared normal. The incision into the common bile duct was closed about rubber tubing, interrupted sutures of medium silk being used. One cigarette drain was placed through the foramen of Winslow into the

lesser peritoneal cavity and another to the site of the choledochostomy. The operation was prolonged by efforts at exploration, and the patient left the operating room in only fair condition.

Postoperative Course.—Hypodermoclysis in both thighs was begun the evening of the operation. The next day the pulse became weak and rapid and the patient became slightly cyanotic. She was given oxygen, which did not greatly improve her color. Her temperature rose during the day and at noon was 103.4° F. by rectum. She was given the usual stimulants and even artificial respiration, but she died that afternoon, some 24 hours following operation.

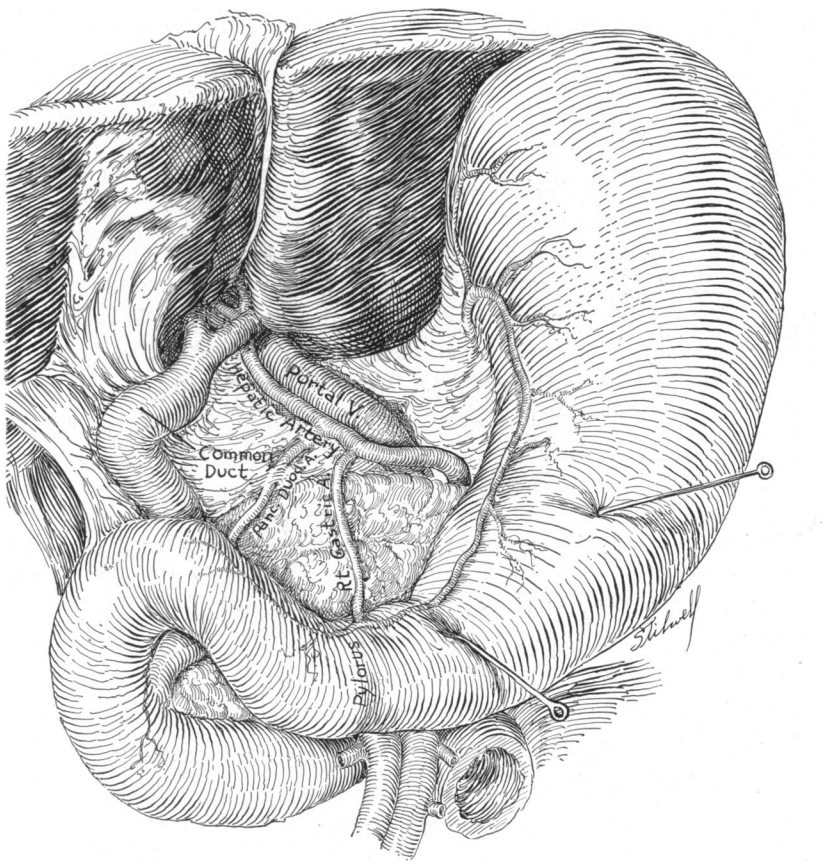

FIG. 3.—Appearance at autopsy after the stomach had been rolled downward. Note unusual relative position of hepatic artery and portal vein.

Partial Autopsy.—Rigor mortis had not set in. There was pitting edema of both lower legs. The intestine was moderately distended. There was no evidence of bile leakage. The liver still showed chronic passive congestion. The common bile duct measured 2 cm. in its greatest diameter. The liver was sectioned in every plane, and there was no sign of an intrahepatic gallbladder. The biliary duct system was carefully dissected and no evidence was found of a vestige of the cystic duct or gallbladder. The coronary vessels were sclerotic but there was no infarction. There was considerable calcification of the aortic arch. Pancreas, stomach, pylorus, and duodenum were grossly normal. Histologic study showed midzone necrosis of liver lobules with shadow-type of cell, together with many leukocytes. The large veins were engorged and contained many leukocytes. Pancreas and myocardium were normal. *Pathologic Diag-*

noses: Congenital absence of the gallbladder. Cholangitis, acute, with cholelithiasis. Arteriosclerotic heart disease, with hypertension.

There are a number of interesting surgical considerations that arise when we study the two cases herewith reported, and other similar ones collected from the literature. In the first place, when abdominal exploration is carried out in a patient with suspected gallbladder disease and no gallbladder can be found, there is the possibility of an intrahepatic gallbladder, as well as calculi in the biliary ducts. Exploration of the ducts should then be carried out, and if no calculi are found, in spite of a definite history of biliary disease, one is justified in making a reasonable attempt to find whether there is an intrahepatic gallbladder. In one of our cases, we are sure there was no intrahepatic gallbladder since the patient did not survive because of her heart condition and complete section of the liver was possible. In the other case, however, because of the fact that, on thorough exploration, there was no evidence of an intrahepatic gallbladder, as far as could be determined, and also because the patient has been completely relieved of his symptoms following the removal of the stones from the common duct, it seems quite presumptive that there was congenital absence of the gallbladder. Unfortunately, in many case reports of congenital absence of the gallbladder, the data are not given in enough detail to permit definite conclusions to be drawn. It is true, however, that in practically all the cases where the common duct was much larger than normal, stones were present.

SUMMARY

We have presented two cases of congenital absence of the gallbladder together with 46 similar cases collected from the literature after a rather exhaustive search. We have included only those cases in which no other pathologic condition was present, and have, therefore, left out a large number of cases where there was either complete or partial absence of the common and hepatic ducts as well. Some consideration has been given to the cause of absence of the gallbladder from the embryologic standpoint, and a few of the surgical aspects are discussed.

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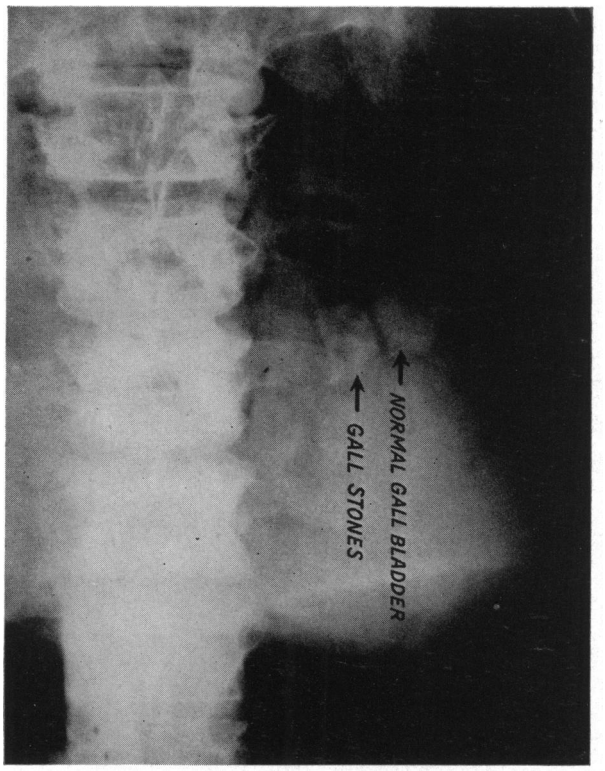

FIG. 1.—Roentgenogram showing a duplicate of the gallbladder—one containing calculi, the other apparently containing dye only.

DISCUSSION.—DR. HUGH TROUT (Roanoke, Va.): In 1933, we* reported a case of duplication of the gallbladder. At that time we looked up the literature and found there had been 34 cases reported to date. A subsequent review shows that since then 18 additional cases have been reported. So far as I know the literature does not reveal a case where the diagnosis was made roentgenologically prior to operation. We were fortunate enough to have one such case. (Figs. 1 and 2).

At operation, the two gallbladders were removed. Each was complete, with cystic duct, the cystic ducts joined just before entrance into the common duct. Boyden† demonstrated that this was the most frequent type

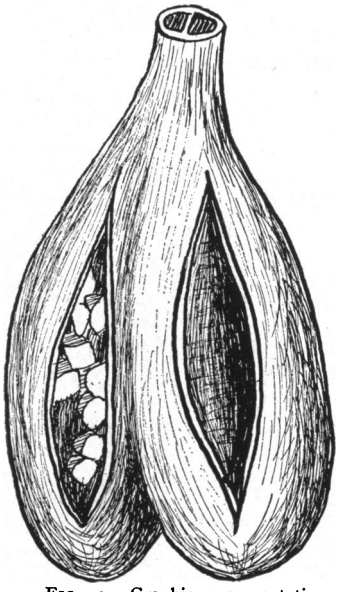

FIG. 2.—Graphic representation of roentgenographic interpretation of duplication of the gallbladder. (The artist has transposed the roentgenographic shadows.)

* Slaughter and Trout: Amer. Jour. Surg., 19, 124–125, January, 1933.

† Boyden: Amer. Jour. Anat., 38, 177, November, 1926.

of cystic duct relationship in the duplication of the gallbladder. Boyden, also, after a study of 19,000 autopsies done in several anatomic laboratories, showed five cases of duplication of the gallbladder, an incidence of one in 4,000. I think this condition perhaps is more frequent than has been supposed. A number of lower animals, especially cats, have been found to have duplication and accessory gallbladders. In fact, as many as four gallbladders have been found in one cat.