CONGENITAL ABSENCE OF THE GALLBLADDER WITH CASE REPORT

LEOPOLDO VILLAREAL, M.D.
EL PASO, TEXAS

CONGENITAL ABSENCE of the gallbladder is a rare anomaly. Gross[1] in a review of the literature in 1936 was able to find only 38 cases reported since 1905. Since 1936, 13 additional cases[2-7] have been reported. Dixon and Lichtman[8] in 1945 added ten cases from both operative and necropsy records of the Mayo Clinic. This anomaly is probably not as rare as these reports would indicate. Undoubtedly many cases are encountered, which are simply not recorded. A truer estimate of the incidence of congenital absence of the gall bladder will be reached only if all cases are reported. This will also help in focusing attention on certain surgical aspects of this condition; as well as aid in the study of the patho-physiologic changes which take place in the post cholecystectomy state.

Only the 60 cases reported since 1900, which are available to us, will be considered in this review, since many of the cases reported previous to that time are lacking in details and found unsatisfactory for analytical study. Cases of congenital absence of the gallbladder, associated with atresia of the extrahepatic ducts will not be considered since they concern an entirely different problem. A case encountered by us will be presented.

The causes listed for this anomaly are many. Two, however, stand out. Both of these deal with the embryologic development of the liver and bile ducts. The two theories are as follows: (1) The hepatic diverticulum from the foregut forms the liver, gallbladder and extrahepatic bile ducts. The gallbladder and cystic duct form an outpocketing from this diverticulum. Failure of development of this outpocketing would cause an absence of gallbladder and cystic duct.

(2) The gallbladder, hepatic, cystic and common ducts in their early embryologic development are hollow structures. In the so-called solid phase, their lumina become obliterated. Failure of the gallbladder and cystic duct portions to recanalize, would cause an absence of these structures.

The condition is more common in women than in men, with 38 cases found in women and 23 in men. One report did not mention sex. This falls in line with other hepato-biliary diseases. The average age of the patients was 46 years.

In 26 of the 60 cases, the condition was found at necropsy. In not one of these cases was the cause of death hepatic or cholecystic disease. These patients apparently had no symptoms referable to this anomaly. Symptoms suggestive of cholecystic disease were present in 37 cases. Jaundice was present in 30 cases.

In 17 cases cholecystographic studies were carried out and failed to reveal a gallbladder. The diagnosis of nonfunctioning gallbladder and cystic duct obstruction was frequently made. In no case, to our knowledge, was this anomaly diagnosed preoperatively.

The gallbladder fossa in the liver was present in seven cases and absent in 27. In the others it was not mentioned. The common duct was dilated in about 50 per cent of cases in which the size of the duct was mentioned. Gall stones were found in the common or hepatic ducts in 18 cases, not found in 19 cases and not mentioned in 25 cases.

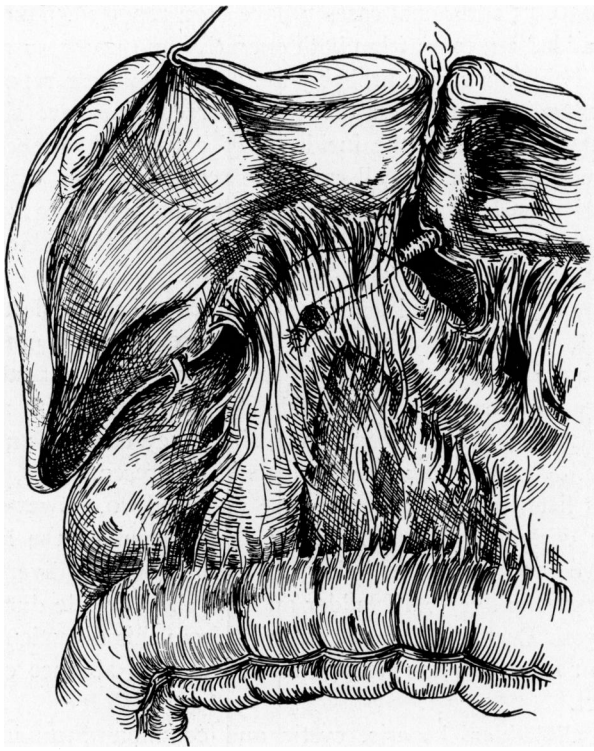

FIG. 1.—Artist's drawing of findings at operation. Wide fibrous band, holding duodenum up to hilar region of liver. Hepatic and common ducts readily visualized, with stone in the latter. No cystic duct or gallbladder outpouching to be seen.

The pancreas should be examined carefully, for Bower[5] lays stress on the frequency of associated pancreatic disease. The pancreas was mentioned in 12 cases, pancreatitis existing in 11 of these.

Case Report.—Hotel Dieu Hosp. Case No. G. 9207. Mrs. J. C., 41 years old, female. Admitted to hospital on 4-3-46, with a history that for 4 months she had had severe pain in right upper quadrant, requiring morphine for relief. The pain radiated to the right scapula and right shoulder. Associated with the pain, she had fever and chills, nausea and vomiting. For 2 months she had been jaundiced, on and off, with the

jaundice varying in intensity. She had lost 25 lbs. in weight since the beginning of her illness.

Her past history was essentially irrelevant.

Physical examination showed a middle aged, moderately obese woman who was markedly jaundiced. Pulse, temperature and respiration were within normal limits.

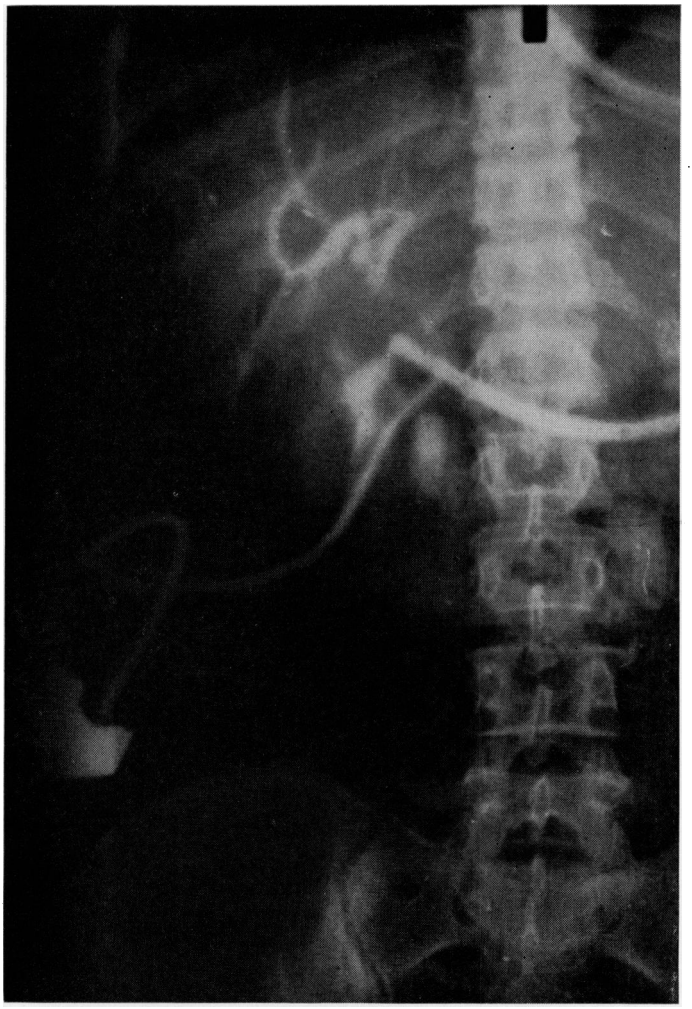

FIG. 2.—Cholangiogram, taken on operating table. All ducts are readily visualized but no vestige of gallbladder or cystic duct is seen.

Sclerae were yellow. Examination of the heart and lungs was normal. The abdomen was soft, with tenderness in right upper quadrant. There were no palpable masses. The liver was moderately enlarged, with edge two fingers breadth below costal margin. Spleen was not palpable.

Gallbladder visualization studies showed no gallbladder shadow and no stones were seen on the plain film. The radiologist made a diagnosis of gallbladder disease with

cystic duct obstruction. Her laboratory studies showed RBC 4,770,000 Hgb. 96.8 per cent WBC 8250 62 per cent polymorphonuclears. Negative Kahn and Eagle. Urine showed 2+ bile pigment and trace of albumin. Prothrombin time was 30 sec. (normal 15 sec.) All other laboratory studies were normal. A preoperative diagnosis of cholelithiasis, and cholecystitis with common duct stone was made. The patient was placed on the usual preoperative preparation which included vitamin K and when the prothrombin time was 15 sec. operation was carried out.

Operation was performed on April 6th, 1946, with nupercaine (1–1500) spinal anesthesia. The abdomen was opened through a right upper oblique incision. The undersurface of the liver was exposed and the region explored. No gall bladder was found. The outer border of the duodenum was found attached by a short wide fibrous band to the hilar region of the liver (Fig. 1). This was divided and the duodenum mobilized, exposing the common duct, which was about three times enlarged. No gallbladder fossa was seen. The common duct was explored upwards, to the point of junction with the two hepatic ducts. These were followed to their point of exit in the liver. No cystic duct, either fully developed or in the form of a stump was to be found. No cystic artery was seen. A large stone was palpated in the retroduodenal portion of the duct. The pancreas was hard and suggestive of pancreatitis. The common duct was incised, and the stone removed. A catheter was passed through the ampulla of Vater to demonstrate patency. The duct was irrigated with normal saline. A T-tube was placed in the duct and the duct closed with interrupted silk sutures. The appendix was removed. One Penrose drain was placed at the foramen of Winslow and brought out with T-tube through the stab wound. The wound was closed in layers with interrupted cotton sutures. With the patient still on the operating table, a cholangiogram was performed and both hepatic ducts, common duct and smaller intrahepatic ducts were well visualized (Fig. 2). No cystic duct or gallbladder was seen. The dye was seen passing readily through the duodenum into the upper jejunum.

Postoperative course was entirely uneventful. The patient was discharged on her 18th hospital day, with the jaundice rapidly receding and the T-tube still in place. The tube was removed on May 17th, 1946 and the stab wound rapidly healed. The patient has been seen at frequent intervals and she has remained entirely relieved of all her symptoms, with no further reappearance of her jaundice.

The possibility of an intrahepatic gallbladder was considered. It is felt that this possibility was eliminated, as surely as one can with a live patient, by the search at operation, the immediate postoperative cholangiogram which failed to show any cystic duct or gallbladder, and finally by the complete relief of the patient's symptoms during the period of over a year since her operation.

The possibility of an intrahepatic gallbladder must always be considered, when the gallbladder is not to be found in its usual position. There are a few of these cases reported, many of them containing calculi. One can with a fair degree of certainty, eliminate the possibility of this condition at the operating table by doing a cholangiogram, which should show all or part of the cystic duct and gallbladder, if they are present.

SUMMARY

A review of some of the findings in congenital absence of the gallbladder is presented. The necessity of considering the possibility of an intrahepatic gallbladder is brought out. The value of a cholangiogram at the operating table in eliminating this possibility is presented.

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605 Caples Bldg.
El Paso, Texas