

CONGENITAL CYSTIC DILATATION OF THE COMMON BILE DUCT

FOLLOW-UP ON PREVIOUSLY REPORTED CASE AND REPORT OF ADDITIONAL CASE

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SINCE congenital cystic dilatation of the common bile duct (choledochus cyst) if untreated eventually leads to death, whereas, surgical intervention offers cure or improvement, continued reports of this rare lesion are warranted in order to emphasize its existence, methods of early diagnosis, and proper treatment. For this reason two cases of our own are presented and discussed, one a previously reported case with follow-up study and the other recently observed. In addition the literature on this subject since our previous review¹ is brought to date.

Case 1.—P. W., a white male, age 20, was admitted to the Jefferson Medical College Hospital July 11, 1941, with the chief complaints of pain in the right upper abdomen, jaundice and fever, recurring at varying intervals over a period of eight months. A mass, six inches in diameter, was palpable in the right upper abdominal quadrant.

Laboratory studies revealed a moderate anemia (hemoglobin 65 per cent); grossly bile-stained urine; rapid sedimentation rate (31 mm. within one hour); low prothrombin time (38 per cent of average normal); impaired liver function (15 per cent bromsulfalein dye retention); hyperbilirubinemia (3.5 mg.); nonvisualization of the gallbladder by oral cholecystography; and displacement of the stomach, duodenum, jejunum and colon on roentgenologic examination of the gastro-intestinal tract with barium (Fig. 1, A and C). Peritoneoscopy revealed a large cystic retroperitoneal mass near the right kidney.

The patient was operated upon July 31, 1941, the preoperative diagnosis being retroperitoneal mass, probably choledochus cyst, with obstructive jaundice. A large choledochus cyst together with the gallbladder and cystic duct were excised *en masse*, and the right and left hepatic ducts implanted into the first portion of the duodenum over No. 18 F. catheters. Uneventful recovery ensued, and the patient was discharged on the 37th postoperative day. The catheters were removed through a gastroscope during a second hospital admission 11 weeks postoperatively.

Follow-up study, conducted over a period of three years and eight months, reveals that the patient's general health has remained excellent. He has been working steadily as operator of a metal planer for the past three years and has been happily married for two and one-half years. He is free of pain, jaundice, abdominal masses and fever, and has gained five pounds in weight. Bromsulfalein liver function study performed on March 13, 1945, revealed no dye retention, and the van den Bergh reaction was negative direct with serum bilirubin 1.0 mg. Gastro-intestinal series and barium enema study at this time revealed normal position of the stomach, small intestines and colon (Fig.

i, B and D). There was questionable regurgitation of a very slight amount of barium into the hepatic ducts.

Case 2.—E. K., a white female, age 58, was admitted to the service of Dr. Thomas A. Shallow at the Jefferson Medical College Hospital, July 13, 1944, with the chief complaint of abdominal swelling.

During the year prior to admission the patient noticed that her abdomen was gradually and persistently increasing in size. This enlargement was not associated with any discomfort or pain. Every three or four weeks she also noticed that her urine

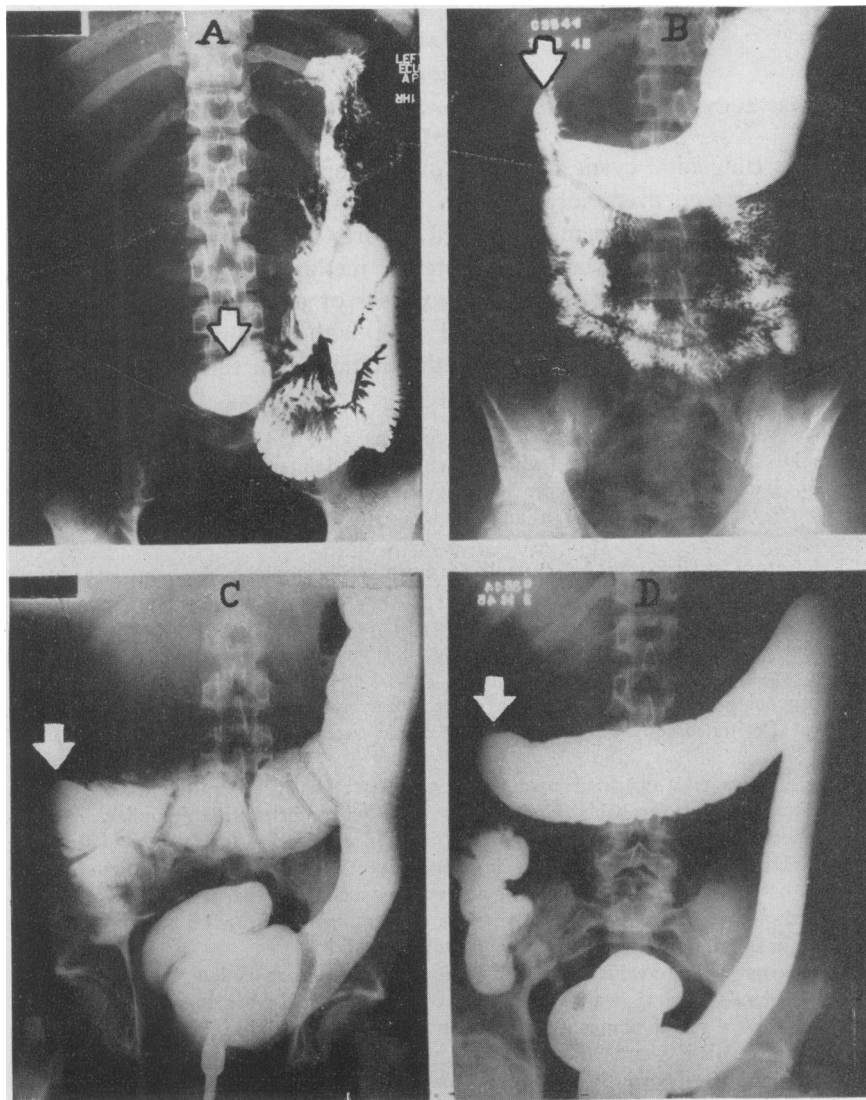


FIG. 1.—Roentgenologic studies in Case 1, in which the cyst was excised. Arrow in (A) shows displaced stomach and duodenum preoperatively, and in (B) the return to normal position postoperatively. Arrow in (C) shows displaced hepatic flexure of colon preoperatively, and in (D) the return to normal position postoperatively.

became darker than usual and her stools lighter in color for several days, but no jaundice was observed. There was no weight loss and her general health had remained excellent.

A review of the remaining systems disclosed a recent episode of senile vaginitis treated successfully by a gynecologist. The past history was otherwise uneventful for serious illnesses or operations. The family history failed to reveal any member ever having suffered from a similar ailment.

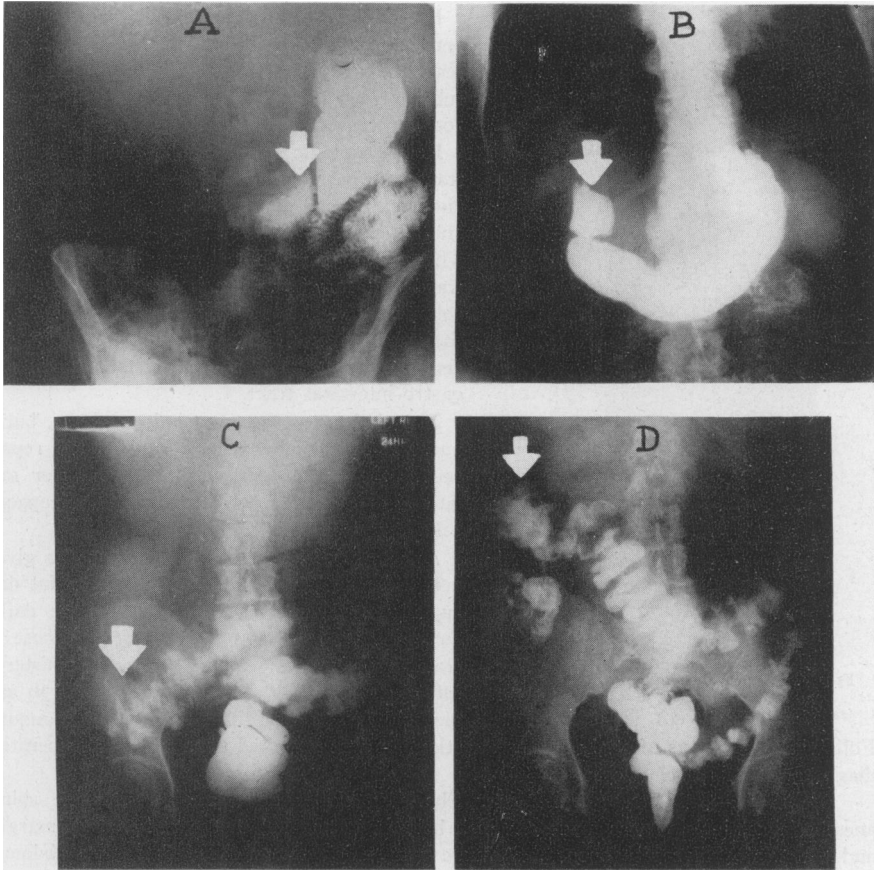


FIG. 2.—Roentgenologic studies in Case 2, in which the cyst was anastomosed to the duodenum. Arrow in (A) shows displaced stomach and duodenum preoperatively, and in (B) the return to normal position postoperatively. Arrow in (C) shows displaced hepatic flexure of colon preoperatively, and in (D) the return to normal position postoperatively.

Physical Examination.—The patient was a well-nourished middle-aged woman, who did not appear ill. The temperature, pulse, and respirations were normal. Blood pressure, 144/94. The scleras were moderately icteric and there was slight jaundice. Examination of the neck and chest was essentially normal. On inspection of the abdomen there was distention resembling that seen in the eighth month of pregnancy; the origin, however, appeared to be upper abdominal. The overlying superficial abdominal veins were slightly more prominent than usual. On palpation, a smooth, elastic, nontender, mobile mass was felt, which filled the entire upper abdomen, and extended to the level of Poupart's ligament on the right side and the lower costal margin on the left. This

mass moved slightly with respiration. Pelvic and rectal examinations as well as the extremities were normal.

Laboratory Studies.—Hemoglobin, 91 per cent; red cells, 4,700,000; white cells, 8,400, with polymorphonuclear cells 64 per cent, eosinophils 1 per cent, lymphocytes 32 per cent, and monocytes 3 per cent. Color index 0.96.

Routine uranalysis on four different days revealed very dark amber color; specific gravity ranging between 1.010 and 1.022; moderate albuminuria; no sugar; no acetone; and many white blood cells.

Bromsulfalein liver function test showed 40 per cent dye retention. The van den Bergh reaction was positive direct with a serum bilirubin of 2.0 mg. The prothrombin time was 100 per cent of average normal.

Roentgenologic study of the gastro-intestinal tract with barium revealed that the stomach and duodenum were displaced markedly to the left and anteriorly by an enormous mass (Fig. 2A). The coils of small bowel were displaced downward and forward. The hepatic flexure of the colon was displaced downward to the level of the pelvic brim (Fig. 2C). There was no evidence of organic disease or obstruction in the gastro-intestinal tract.

The cholecystogram was indeterminate, but a questionable shadow which could have represented the gallbladder was noted anterior and lateral to the mass. No opaque or nonopaque calculi were noted.

Preoperative Course.—The patient was given a high carbohydrate, high protein, low fat diet supplemented by a pint of orange juice daily. In addition, 1,000 cc. of 10 per cent glucose in water with vitamin K mg. 3 were administered intravenously daily, and vitamin C mg. 50 and aminoids, one ounce, three times daily by mouth.

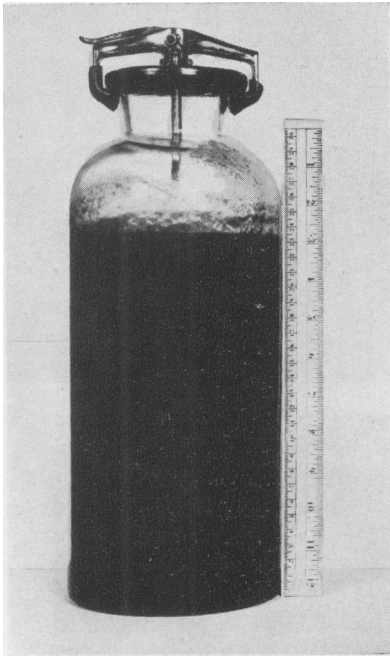


FIG. 3.—Appearance of cyst fluid in Case 2. The jar was filled one and one-half times (5,800 cc.).

Following eight days of study and preparation she was operated upon with a preoperative diagnosis of choledochus cyst.

Operation.—July 20, 1944, Doctor Shallow: Under 20 mg. of pontocaine spinal anesthesia, an incision was made one inch below and parallel to the right costal margin, and the peritoneal cavity opened. A huge cyst which almost filled the entire abdomen was encountered. The duodenum was displaced medially and anteriorly, the transverse colon caudally, and the gallbladder laterally. The gallbladder was normal in size and contained no palpable stones. The liver, likewise, was of normal size and showed only slight cirrhotic change.

With a needle and syringe 10 cc. of thin dark brown fluid were withdrawn from the cyst which on culture yielded no bacterial growth in 48 hours. A suction trochar was then inserted and a gallon collecting jar was filled about one and one-half times (Fig. 3). Subsequent measurement of the fluid disclosed a total quantity of 5,800 cc., and chemical analysis revealed the presence of bile and a moderate amount of amylase.

Because of the age of the patient and the enormous size of the cyst, it was felt that extirpation carried an undue risk. Accordingly, anastomosis was performed between the most dependent portion of the deflated cyst and the first portion of the duodenum. While carrying out this procedure a biopsy specimen of the cyst wall was removed at the anastomosis site. A Penrose drain was inserted into the right kidney pouch and

brought out the lateral pole of the wound which was then closed in layers. The patient remained in good condition throughout the operation which lasted one hour and 15 minutes.

Pathologic Examination of Biopsy Specimen of Cyst Wall.—The cyst wall consisted of fibrous tissue, 5 mm. thick, in which there were areas of recent hemorrhage. An epithelial lining was lacking (Fig. 4).

Postoperative Course.—The immediate response and convalescence were essentially uneventful. Wangensteen suction was instituted for the first three days, during which time the patient received glucose in saline intravenously, supplemented with vitamins C and K and amino-acids. She received 500 cc. of blood shortly after operation and also on the 10th and 18th days postoperatively. The drain was withdrawn during the seventh and eighth days. The wound healed without infection, and the patient was



FIG. 4.—Photomicrograph of wall of cyst in Case 2. ($\times 37.5$)

discharged on August 17, 1944, the 28th day postoperatively. At this time there was no evidence of abdominal enlargement and the serum bilirubin was 0.5 mg.

Subsequent Course.—Follow-up study February 15, 1945, approximately seven months postoperatively, revealed that the patient was entirely well, had a good appetite, and was free of abdominal swelling, jaundice or pain. The wound remained well healed. Roentgenologic study of the gastro-intestinal tract with barium showed the stomach, small intestine and colon in a normal position (Fig. 2 B and D). Cholecystogram failed to reveal a gallbladder shadow and there was no evidence of opaque calculi. Liver function study, March 21, 1945, eight months postoperatively, revealed 5 per cent retention of the bromsulfalein dye. At this time the van den Bergh reaction was negative direct and serum bilirubin 0.7 mg.

DISCUSSION.—Since our previous review of the literature¹ there have appeared case reports by Smith² (two cases), Gray,³ Swartley,⁴ Nitsche,⁵ and Hutchins and Mansdorfer.⁶ These, in addition to our own new case, bring the total authentic cases to 182. The report of Diebold⁷ is not available at present and for that reason is not included. The addition of these few reports adds little of significance to our recent analysis of 175 cases, and, therefore, a further analysis is not warranted at this time.

If the lesion is not suspected preoperatively it is important to recognize it promptly at celiotomy in order to avoid unduly long exploration and improper treatment. The frequent but not constant triad of symptoms and signs, namely, tumor (77 per cent), jaundice (70 per cent) and pain (59 per cent), should suggest the correct diagnosis if the lesion is kept in mind as a diagnostic possibility. Tumor and jaundice, which are the most constant features of the triad, were present in both of the authors' cases, but pain was absent in Case 2. The statement frequently seen in the literature that no relation exists between the size of the cyst and the severity of the pain is well borne out in this case. The importance of preoperative diagnosis is demonstrated by the fact that in 22 cases in which it was correct, or suspected, the mortality was 36 per cent, whereas, in 153 cases in which the diagnosis was incorrect, or none made, the mortality was 62 per cent. In each of our two cases the outcome was favorably influenced by the preoperative suspicion of the lesion, improvement of the patient's condition prior to operation and properly selected surgical treatment.

Anastomosis of the biliary and intestinal tracts is essential to prevent biliary cirrhosis, cholangitis, liver abscess, hemorrhage, or rupture of the cyst. The cyst itself may be anastomosed, as first advocated by Bakes,⁸ in 1907, or excised and the remaining choledochus, common hepatic duct or both hepatic ducts joined to the intestine; or the gallbladder may be anastomosed when a free communication exists between this organ and the cyst. Primary performance of this group of procedures carried a mortality of 27 per cent in 60 cases, whereas, external drainage of the biliary system followed by secondary anastomosis to the gastro-intestinal tract carried a mortality of 29 per cent in 24 cases. The latter procedure entails a slightly higher mortality because of the formation of an external biliary fistula, hemorrhagic tendency due to low prothrombin values, and the extra burden of two operations. It should be performed only in cases in which the operative risk is very great, as by marked debilitation or severe infection.

Although primary anastomosis of the biliary and intestinal tracts has been established as the procedure of choice, it is debatable whether or not the cyst should be excised. Extirpation of the cyst with primary anastomosis of the remaining duct system to the duodenum seems more physiologic, since it creates a more normal union between the liver and duodenum and eliminates the cyst which may harbor infection. This method, however, carries a higher mortality (three deaths in seven reported cases) and should only be considered when the patient is a good operative risk, the cyst large, and infection

minimal or absent. The number of reported cases in which this procedure has been carried out is too few (follow-up reports even fewer) to warrant any conclusions as to the value of the general employment of the method. The authors' Case 1, nevertheless, demonstrates the feasibility of the procedure in a properly selected patient. Anastomosis of the cyst, as in the authors' Case 2, gives satisfactory results, particularly in surgically handicapped patients and is the procedure recommended in the ordinary case. The remaining dilated duct may harbor regurgitated food, however, leading to severe ascending cholangitis (case of Fowler⁹).

The amount of cyst fluid usually varies from 30 cc. to several liters. The aspiration of 5,800 cc. in the authors' Case 2 is most unusual. In the case of Reel and Burrell,¹⁰ disputed by Poate and Wade¹¹ as not being authentic and not included in our previous review, 8,000 cc. are reported. Other authors recording large amounts of cyst fluid are Smith² (Case 1, 5,200 cc.), Yotuyanagi¹² (Case 2, 5,200 cc.), and Fukada¹³ (Case 1, 5,000 cc.).

Little is known about the late results in cases of recovery following operation, as seldom is anything further reported concerning the progress of the patient. McConnell,¹⁴ Berkley,¹⁵ and Gross¹⁶ each reported cases in which the patients were well after four years. Cases reported as well for more than five years are those of Iselin,¹⁷ Hildebrand,¹⁸ McWhorter,¹⁹ Murata,²⁰ Walton,²¹ (second case), Wheeler,²² Sumpster,²³ and Judd and Greene.²⁴ Swartley and Weeder's²⁵ case died ten years later of hemorrhage from esophageal varices due to portal obstruction. The case of McWhorter¹⁹ died 13 years later of uremia with nephrosclerosis, hypertension, and diabetes. Judd and Greene's²⁴ case in a 13-year follow-up is reported by Smith² to have symptoms similar to gallbladder disease. The case of Wheeler²² died 15 years later at the age of 80.

SUMMARY AND CONCLUSIONS

1. Despite the rarity of congenital cystic dilatation of the common bile duct, 182 authentic cases in the world literature, we have personally encountered two cases herein reported.

2. The lesion was suspected preoperatively in both instances. The diagnostic triad—tumor, jaundice, and pain was present in one case, but pain was absent in the other.

3. In Case 2, 5,800 cc. of cyst fluid represent the largest amount recorded in an undisputed case.

4. The safest treatment, and the one recommended for general employment, is primary anastomosis of the cyst to the duodenum. This procedure was successfully performed in Case 2.

5. Extirpation of the cyst followed by primary anastomosis of the remainder of the biliary duct system to the duodenum may be attempted if the lesion is suspected preoperatively, the patient a good operative risk, the cyst large, and infection minimal or absent. The feasibility of this procedure is

demonstrated in Case 1, in which the patient is entirely well three years and eight months postoperatively.

6. A contribution of follow-up studies is needed from authors who report cases of operative recovery, in order to accumulate more conclusive data as to the value of the various surgical procedures and their ultimate prognosis.

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