

Intrahepatic Calculi *

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EARLY in January, 1960, a 57-year-old woman was operated upon by us for recurrent choledocholithiasis. She had several stones removed from her common duct nine months previously. At operation we removed from the common duct 13 faceted yellow-brown calculi, 1 to 2 cm. in diameter. A cholangiogram demonstrated the left hepatic duct to be dilated to 2.5 cm. in diameter. A segment of the left hepatic duct which had become a cistern was the site of formation of the calculi in the common duct. A T-tube was placed in the common duct to maintain decompression; within four months many large calculi were demonstrated to be in the left hepatic cistern and the common duct.

Biliary calculi are frequent in our population, occurring in 15 to 20 per cent of those over 20 years of age. In most instances they form in the gallbladder and cholecystectomy results in relief of symptoms and the interruption of the disease. Choledocholithiasis occurs in from eight to 15 per cent of patients with cholelithiasis; removal of calculi from the common duct is likewise a very satisfactory therapeutic measure. However, "recurrent" calculi occur often enough that most surgeons with some experience in the biliary tract anticipate being confronted with patients, in whom they have exercised thoroughness in removing "all" the calculi from the common duct, who require additional operations. (Occasionally calculi may be found in the ductal system and none in the gall-

bladder.) The question most frequently raised is, were stones left behind at the previous operation, or did they form thereafter. Hepatic calculi and intrahepatic ductal calculi were recorded by pathologists and surgeons two centuries ago. It may be well to ever keep in mind that calculi have been described within the liver both in patients who have had their gallbladder removed and in patients whose gallbladders remain *in situ*, with or without cholelithiasis. From a practical viewpoint it may be considered that complete evaluation of the intrahepatic ductal system is too cumbersome a procedure to impose upon each patient undergoing cholecystectomy and choledocholithotomy. Be that as it may, such evaluation becomes obligatory when common duct calculi recur.

In searching the literature for reports on this subject a number of excellent articles have been reviewed describing a wide range of calcareous deposition and stone formation within the intrahepatic ducts. That there are several instances reported of "liver" calculi associated with abscess, fistula formation and peritonitis does not invalidate the common concept that the site of stone formation was within the ductal system. One of the most comprehensive articles "Liver Stones" by Ivan Rufanov,⁸ from Moscow, was published in the ANNALS OF SURGERY, in 1936. Best² and others have emphasized that there were certain differences between intrahepatic stones and common duct stones commonly associated with cholelithiasis. First, the intrahepatic calculi were more

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often than not relegated to one part of the ductal system that did not freely communicate with other branches. Second, gravel and paste-like material often occluded the small branches of the segment containing the large calculi. Third, abscess and fistulae formation as well as suppurative cholangitis was more frequent than is observed in "common duct stones." Papers dealing with single case reports, special aspects of the subject and reviews of the world's reported experience have been appearing at relatively infrequent intervals in the United States since Vachell and Stevens' ⁹ publication "The Report of a Case of Intrahepatic Calculi." in 1906.

They described a patient, aged 52 who had recurrent attacks of gallstone colic for 29 years. Only in the final attack did he become jaundiced. The jaundice became intense, the liver enlarged and he died. At necropsy the liver weighed 2,750 Gm., the gallbladder contained no calculi but the intrahepatic ducts were greatly dilated and contained 520 calculi ranging in diameter from a millimeter or two up to 4.38 cm. There was a suppurative cholangitis and an abscess extending from the liver into the diaphragm.

In 1916, Lewisohn ⁶ of New York, reported a man of 31, who recalled typhoid at 15 and jaundice at 16, complaining of severe abdominal pain of 24-hour duration with some pain in the right shoulder. He had had a chill but was not jaundiced. The preoperative diagnosis rested between gangrenous cholecystitis and perforated peptic ulcer. Free bile was found draining from the diaphragmatic surface of the liver where a small opening led into an abscess cavity containing small black "gallstones." The gallbladder contained 20 calculi that were yellow in color. It was removed. The site of the biliary fistula of the liver was drained, it discharged bile for 6 weeks and then closed. Five months later the patient was well.

Judd *et al.*⁵ of the Mayo Clinic, reported

a woman of 50 with a 10-year history of recurring attacks of biliary colic. She was operated upon, a greatly thickened gallbladder without stones was removed as well as four large stones from the common duct. It was then found that there were three large calculi in the hepatic duct; these were crushed and removed in fragments. After being relatively free of symptoms for slightly more than a year she again had attacks of abdominal pain and in addition evidence of pyloric and intestinal obstruction, requiring two operations. At postmortem examination, 12 years after her first operation the liver weighed 1,549 Gm., the common and hepatic ducts were dilated and contained 13 stones ranging from 2 mm. to over a centimeter in diameter.

In order to portray in proper perspective the current clinical significance of intrahepatic calculi there follows a description of their pathogenesis and most frequent sequelae. The incidence of intrahepatic calculi in relation to calcareous biliary tract disease is estimated. Emphasis is placed upon the signs and symptoms suggestive of this condition as well as the means now available for establishing the diagnosis. Four clinical cases are reported illustrating diagnostic and therapeutic procedures (palliative and definitive) employed in their management.

Pathogenesis and Incidence

The pathogenesis of intrahepatic calculi is not known. Neither, of course, is that of calculi elsewhere in the biliary tract. There are many theories that afford a reasonable and logical explanation. These in principle rest mainly on 1) metabolic disturbances; 2) obstruction and stasis of bile; and 3) infection. Viewing the problem as a matter of comparative incidence or occurrences it is commonly accepted that stones in the gallbladder are very common in our adult population (10-20%).

That stones are found in the gallbladder in 99 per cent of patients with calculi in the biliary tract is strong evidence that they are formed there. The reason for this would seem to rest in the difference between the gallbladder and the remainder of the biliary tract in its contact with the bile. In the transportation of bile from its source in the liver to its destination in the duodenum, stasis is most prolonged in the gallbladder where it is concentrated by loss of water. Among the patients collected by Rufanov with intrahepatic calculi 20 per cent had no stones elsewhere in the biliary tract, i.e. gallbladder or common duct, and 40 per cent had none in the gallbladder. It is thus logical to consider that stasis, infection and metabolic disturbances may play an important role in the development of calculi within the hepatic ducts as within the gallbladder. Obstruction in the intrahepatic ducts may be due to congenital anomalies as described by Norman.⁷ We too have observed these. Postoperative stricture of the common duct at the site of exploration or injury and neoplasia arising any place from within the ductal system may cause obstruction.

Calculi in the intrahepatic ducts proximal to these various types of obstruction have been reported but they are by no means a frequent finding. Furthermore, there are examples of intrahepatic calculi with neither evident congenital anomalies, strictures or tumors of the intrahepatic ducts. The exact role of obstruction in pathogenesis of intrahepatic calculi would be interesting to ascertain. That small calcareous fragments pass from the gallbladder into the common duct where they increase in size by deposition of solid material derived from the bile is generally accepted. It would be within the realm of reasonable possibility for a small calculus to migrate into an intrahepatic duct and become lodged there by one mechanism or another causing obstruction and at the same time increase in size. Of those who have calculi

in the gallbladder or have had a cholecystectomy for cholelithiasis eight to 15 per cent may be expected to have stones in their common duct. There is much clinical evidence to support the contention that the longer the duration of calculi in the gallbladder, the greater is the incidence of choledocholithiasis. In the aged, those 65 years old and over, the incidence rises to almost 50 per cent. Calculi within the liver or intrahepatic ductal system at any age is commonly looked upon as being rare. Data from postmortem examinations and surgical operations in our opinion is too meagre to indicate the accurate incidence. Beer,¹ Rufanov, Best and others have reported intrahepatic calculi present in from 5-8 per cent of all patients with calculi in the extrahepatic biliary system, including the gallbladder.

In reviewing our own experience and the reports of others in this country we are inclined to contend that the actual incidence is higher, perhaps at least twice the above figures. We have failed repeatedly to correctly diagnose their presence at the primary operation. As at autopsy material it should be emphasized that unless they are specially searched for they may be readily overlooked. With an ever-increasing number of patients being operated upon for cholelithiasis each year in this country it is incumbent upon the surgeons who perform operations upon the biliary tract to be alert for this condition.

In contrast, in the Far East the incidence of intrahepatic calculi is high as reported by Cook,⁴ Chih-Ch'iang³ and Rufanov who are in agreement that this is particularly true among the Chinese and Japanese. Huang Chih-Ch'iang in his personal experience with 110 patients with primary bile duct stones reports that 30 per cent had calculi in the intrahepatic ducts. He also states that 40 to 71 per cent of patients operated upon for cholelithiasis in various areas in China have calculi in the

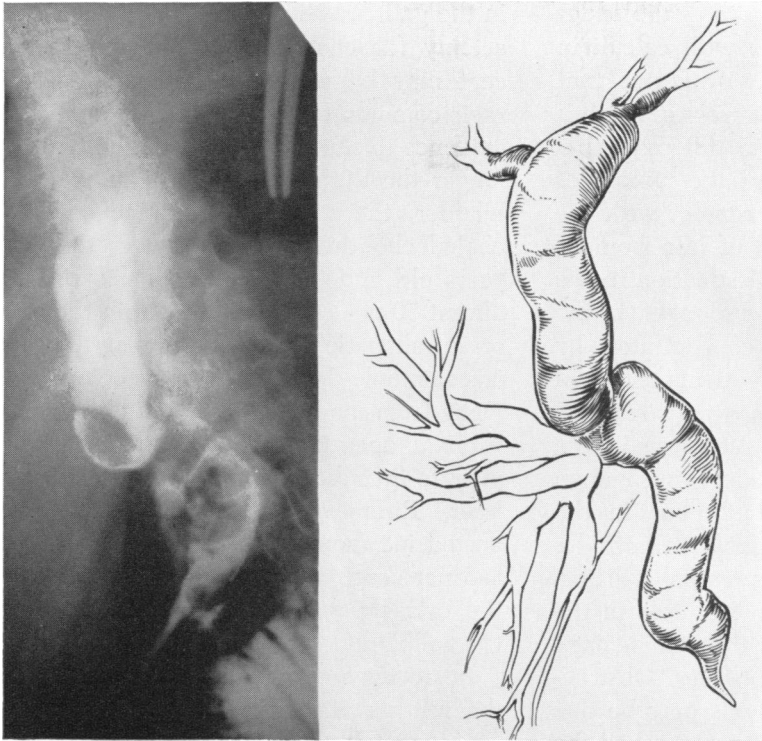


FIG. 1. Case 1. Cholangiogram and line drawing demonstrating a greatly dilated common duct and left hepatic duct containing several calculi and gravel-like material.

common duct. Our experience with biliary tract disease in those of Chinese and Japanese descent in this country indicates no difference from the population in general. For this reason we believe parasites are in some way implicated. No mention was made in the papers we have reviewed as to the comparative incidence of acalculous gallbladders occurring with stones elsewhere in the biliary tract. Such information might be significant.

Case Reports

Case 1. (F. K. NYH 844,643.) Sex: F.; Age: 57. This patient had her initial attacks of biliary colic during the last trimester of her first pregnancy at 30 years of age. She entered another hospital with severe mid-epigastric pain and jaundice in 1938, at 36 years of age. Her jaundice subsided spontaneously and a cholecystectomy was performed without complication. The ensuing ten years were symptom free.

In the early part of 1948, she had the first of several severe attacks of mid-epigastric pain, belching, flatulence, chills and fever. None of these attacks were accompanied by jaundice and

all responded to conservative management. An intravenous cholangiogram in March of 1959 revealed two common duct stones and a choledocholithotomy was performed. A large cystic duct remnant was excised and a sphincterotomy carried out to insure more adequate drainage of a remarkably thickened and dilated common duct. She remained symptom free for a period of three months when again she began to have recurrent episodes of mid-epigastric pain. An intravenous cholangiogram again revealed a dilated common duct filled with large stones. There was no history of jaundice, light stools, or dark urine.

On her first admission to this hospital in January of 1960 she was found to be moderately obese, healthy and nonicteric. The total bilirubin was 0.8 mg. per cent and the alkaline phosphatase 4.7 Bodansky units. At exploration on 2/4/60 the common duct was found to be markedly thickened and dilated to 2.5 cm. Thirteen large faceted yellow-brown stones were removed from its lumen which extended as a cast up into the left hepatic duct. Probes could easily be passed through the sphincter of Oddi. A T-tube was placed into the common duct with a long arm extending into the duodenum. The postoperative period was smooth except for a wound infection

which responded readily to conservative therapy. Culture of the bile revealed *E. coli*, *Cl. welchii* and *Ps. pyocyaneus*. Postoperative cholangiograms revealed a dilated common and left hepatic duct. She was discharged with her T-tube in place draining only small amounts of cloudy amber bile.

During the ensuing three months she complained of occasional attacks of right upper quadrant pain. A cholangiogram revealed a residual calculus within the mid-aspect of the common duct. On 5/31/60 a choledocholithotomy with transhepatic drainage of the left hepatic duct was performed. An area of constriction was palpated within the left hepatic duct just proximal to its junction with the common hepatic duct. The left hepatic duct was markedly dilated beyond this level, and it was in this area that a mushroom catheter was placed. The left lobe of the liver was remarkably atrophic, with its parenchyma being only 1 cm. thick about the hugely dilated ductal system. Biopsy of the common duct revealed a thick fibrous stroma infiltrated by numerous neutrophiles and containing nests of bilirubin crystals. Her postoperative period was characterized by decreasing amounts of drainage from both the T-tube draining the common duct and the catheter within the left hepatic duct. She was discharged with both catheters in place and attached to straight drainage.

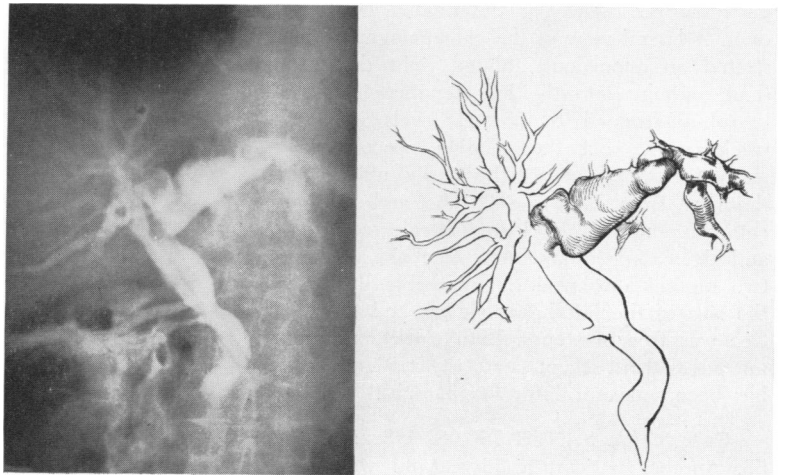
Case 2. (M. O. NYH 78,937.) Sex: F.; Age: 69. A 69-year-old housewife gave a history of initial symptoms of chronic cholecystitis following her first pregnancy at the age of 18. In 1914, at 22 years of age, she underwent cholecystectomy which was not accompanied by common duct exploration as far as can be determined. She

was admitted to this hospital in 1934 for toxemia of pregnancy, and in 1949 and 1957 for ventral hernia repairs. Throughout this period and on all these occasions she was free of gastro-intestinal symptoms.

Her present illness began in May of 1958 with an 18-hour history of unremitting epigastric pain accompanied by nausea, vomiting and fever. She was acutely ill and demonstrated scleral icterus. The total bilirubin was 3.6 mg. per cent and alkaline phosphatase 6.7 units. At exploration she was found to have a cystic duct packed with stones, and a common duct 2 cm. in diameter containing three stones distally as well as a stone in the major right hepatic duct. The sphincter of Oddi accommodated a No. 22 F. bougie and T-tube drainage was carried out. Operative cholangiograms revealed a dilated left hepatic duct with otherwise poor filling of the left biliary radicles. Postoperative cholangiograms demonstrated a stone within the distal common duct and an inability to visualize the left ductal system. At re-exploration a stone was removed from the common duct; the left hepatic duct probed and irrigated freely. Postoperative cholangiograms, however, again demonstrated a markedly dilated left hepatic duct behind a stone just proximal to the junction of the left hepatic and common hepatic ducts. She remained on T-tube drainage for two months. Her course was then symptom free for a period of 18 months.

Re-admission was required on 2/17/60 for a 24-hour history of cramping epigastric pain, chills, fever, light stools and dark urine. The total bilirubin was 4.8 mg. per cent and the alkaline phosphatase 8.6 units. At exploration on 3/1/60 she was found to have a dilated common and left hepatic ducts which contained numerous stones.

FIG. 2. Case 2. Postoperative cholangiogram and line drawing following removal of calculi from common duct revealing a markedly enlarged left hepatic duct proximal to an obstructing stone at the junction of the left hepatic and common hepatic ducts.



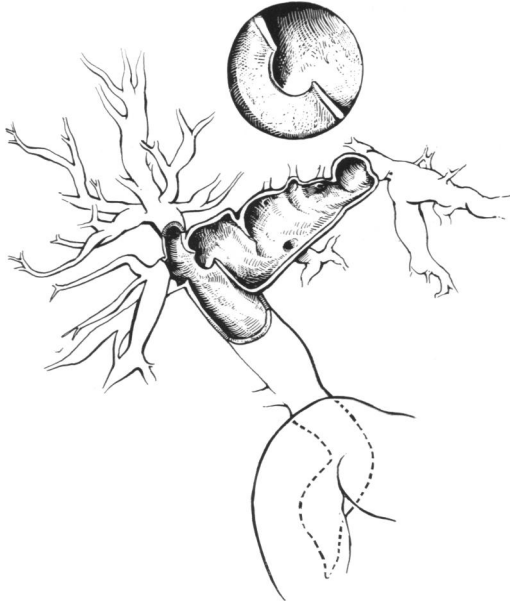


FIG. 3. Case 2. Artist's sketch of a diaphragm-like obstruction at junction of left hepatic duct with common duct found during transhepatic exploration.

Transhepatic drainage on the left hepatic duct was performed as well as T-tube drainage of the common duct. A liver biopsy revealed early cirrhosis, whereas biopsy eighteen months earlier revealed fatty infiltration of the liver. Bile culture grew out *E. coli*, *Aerobacter aerogenes*, and *Ps. pyocaneus*. The stones contained calcium phosphate and bilirubinate.

Her immediate postoperative course was characterized by an *E. coli* septicemia which responded promptly to massive antibiotic therapy. A postoperative cholangiogram on 3/23/60 demonstrated persistence of the dilated left hepatic duct. A lateral view of the cholangiogram demonstrated an anomalous, dilated right dorsocaudal duct draining into the left hepatic cistern. The bilirubin dropped to normal levels in a two-week period, but the alkaline phosphatase remained elevated at 8.2 Bodansky units. Biliary drainage from both the T-tube and mushroom catheter progressively cleared of calcareous debris, and decreased in amounts. A cholangiogram at two months displayed a remarkable decrease in the size of the left hepatic duct, and the biliary tree was free of stones. Both catheters were removed without event, and the patient's course has been uneventful for five months.

Case 3. (A. S. NYH 756,062.) Sex: F.; Age: 53. Another patient, a 53-year-old woman was

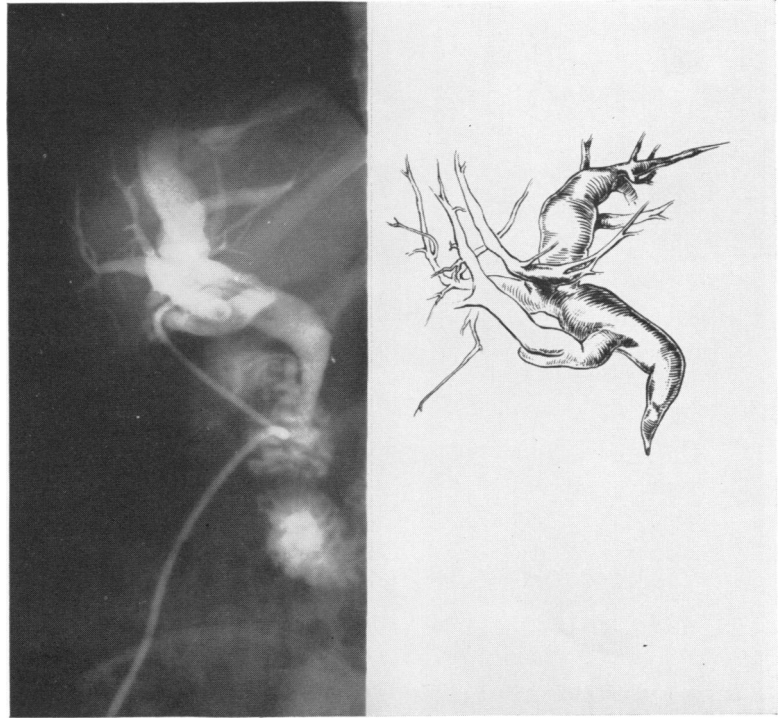
admitted for the first time on 11/1/58 with a 30-year history of intermittent episodes of pain in her right upper quadrant associated with fatty foods. Four years previous she had had an episode of pain, chills, fever, and jaundice which subsided spontaneously. These symptoms recurred four days prior to her present admission.

Examination on admission revealed her to be acutely ill with mild icterus, right upper quadrant tenderness and a liver percussible one finger breadth below the right costal margin. The total bilirubin was 6.0 mg. per cent and alkaline phosphatase 7.9 Bodansky units. Blood culture demonstrated an *E. coli* septicemia. This responded to massive chemotherapy which included Chloromycetin, penicillin, streptomycin and tetracycline. At exploration on 11/6/58 the common duct was thick-walled and dilated to 3 cm. in diameter. It was packed with a cast of brown and yellow-green stones of various sizes which extended up into the hepatic radicles. As many as could be reached were removed digitally. A No. 22 bougie could easily be passed through the ampulla of Vater. The gallbladder, which was chronically inflamed and contained stones, was removed. Liver biopsy revealed chronic cholangitis; bile cultured *E. coli*. The postoperative course was prolonged by depression of hepatic and renal function. A delayed cholangiogram revealed numerous stones throughout the extra and intra-hepatic biliary tree. An anomalous right hepatic duct entering a large cystic duct remnant also



FIG. 4. Case 2. Artist's sketch indicating transhepatic drainage of enlarged left intrahepatic duct with a No. 12 (A) mushroom catheter following removal of calcareous material. Decompression of common duct is maintained by T-tube.

FIG. 5. Case 3. A delayed postoperative cholangiogram and line drawing revealing enlargement of the left hepatic and common ducts together with an anomalous hepatic duct from the right lobe entering a cystic duct remnant. Calculi had previously been removed from the left intrahepatic, common and cystic duct remnant.



contained stones. She was discharged on her 42nd postoperative day with T-tube in place.

The ensuing four months were uneventful. She was reexplored on 4/23/59, and numerous brown and yellow-green calculi were removed from the common duct and biliary radicles. A sphincterotomy was performed and the operation terminated after a most diligent attempt to clear her biliary tree of calculi. The bile cultured *Ps. pyocyaneus*. A delayed cholangiogram again demonstrated retained stones and the anomalous duct as previously described. She was discharged with T-tube in place to be further observed in the clinic.

Her subsequent course was again unremarkable for several months. The T-tube was removed on 12/4/59 and she continued well until 7/19/60, when admission was required for severe right upper quadrant pain, chills, fever, and jaundice. Blood culture demonstrated a *Ps. pyocyaneus* septicemia. An emergency decompression of the common duct was carried out. The duct was found to be markedly thickened and enlarged to three centimeters in diameter. A large stone was removed from its mid-aspect releasing frank purulent material admixed with yellow-green friable calcareous material. Approximately 25 bilirubinate stones were removed and T-tube drainage carried out. A delayed cholangiogram

continued to show retained calculi. She had an uneventful recovery. The T-tube was removed on 9/9/60 and she was symptom free and in apparent good health on her last clinic visit on 10/14/60.

Case 4. (N. S. NYH 460,537.) Sex: M.; Age: 42. This man reported that he first experienced attacks of epigastric pain at 27 years of age, which over a two-year period became accompanied by jaundice. Exploration statedly revealed a "cyst" of the biliary tree; a cholecystostomy was performed. This was followed by a symptom-free period of three years. Intermittent episodes of cholangitis recurred and became associated with bile drainage from the previous cholecystostomy site which led to his first New York Hospital admission on 11/9/46. Physical examination was unremarkable except for bile drainage from his cholecystostomy site. Injection of Lipiodol into this demonstrated a gallbladder with multiple radiolucencies, but an otherwise normal extrahepatic ductal system. A cholecystectomy was carried out for an acutely inflamed gallbladder which contained a cast of inspissated bile. The liver and extrahepatic biliary tree were normal to inspection. He made an uneventful recovery.

An asymptomatic period of six months was interrupted by a recurrence of epigastric pain

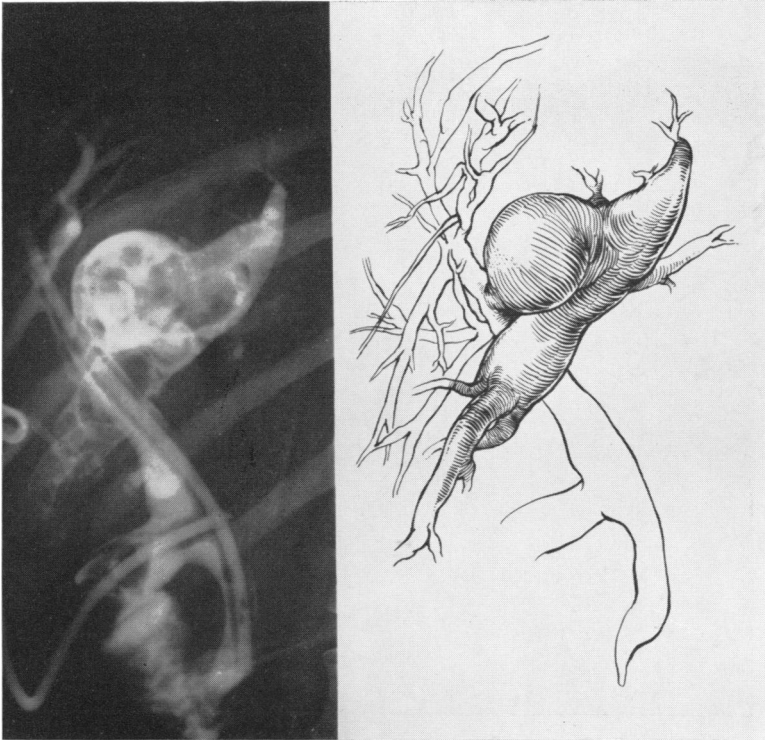


FIG. 6. Case 4. Post-operative cholangiogram and line drawing showing an enormous multilocular cavity in the right hepatic lobe containing numerous radiolucent stones.

accompanied by progressive jaundice leading to his second New York Hospital admission on 6/30/47. Examination revealed an acutely ill, febrile, icteric man. Exploration revealed the common duct to be thick-walled and dilated to 2 cm. There were numerous small, soft, black faceted small stones within its lumen. The ampullary sphincter allowed easy probing. T-tube drainage of the common duct was carried out. A delayed cholangiogram revealed an enormous multilocular cavity with numerous radiolucent stones lying within the liver. The T-tube was removed on his 25th postoperative day.

The ensuing four years were characterized by long symptom free periods interrupted in July, 1948 and June, 1949 by episodes of cholangitis which required hospitalization but responded to conservative medical management. Between these hospitalizations he was free of pain, chills, and jaundice.

Four days prior to his fifth admission on 5/11/51 he developed severe cramping mid-epigastric pain accompanied by chills, fever, and jaundice. On examination he was deeply jaundiced and acutely ill. The total bilirubin was 18.1 mg. per cent and the alkaline phosphatase 15 units. Exploration revealed the liver to be enlarged but otherwise normal in appearance. The common

duct was thickened and measured 1 cm. in diameter. It was filled with clotted blood, but was free of stones or bile. A large catheter could be passed into the duodenum with ease with no evidence of distal obstruction. The left hepatic duct could not be probed. A silk woven catheter was forced into the right hepatic duct and 400 cc. of purulent material was removed with fragmented calcareous material. T-tube drainage was again carried out and an uneventful recovery ensued. A delayed cholangiogram demonstrated a large cystic area filled with stones. At discharge his bilirubin was 9.4 mg. per cent and the alkaline phosphatase 29.3 units. His T-tube was left in place.

The ensuing three years were remarkable in that he remained free of symptoms referable to his biliary tree. In April and June, 1953 he was admitted for back pain of obscure etiology. His bilirubin determinations were 0.1 and 0.3 and his alkaline phosphatase 25.3 and 21 units, respectively, on these occasions. Examination revealed the liver to be down three finger breadths below the right costal margin but the remaining liver chemistries were normal.

His eighth and last New York Hospital admission was preceded by a ten-week history of progressive right upper quadrant pain. On ex-

amination he appeared in good health, without icterus. The alkaline phosphatase was 24 units, and the bilirubin 0.7 mg. per cent. BSP determination was 34.6 per cent retention. Papaverine was administered on the eight post-hospital day with mild relief of his pain. This, however, was followed by chills, fever, and progressive icterus which led to exploration. The liver was three finger breadths below the right costal margin and contained a cystic area in the medial aspect of the right lobe. The common duct was 3 cm. in diameter and contained 400 cc. of purulent material within its lumen. A finger placed proximally into the right hepatic duct revealed a large cavity within the liver which contained numerous large stones. Many were removed, but some were known to be left behind. His subsequent course was one of unrelieved common duct obstruction accompanied by a right subphrenic abscess, progressive cachexia, and death 102 days after his initial exploration on this final hospital admission.

Autopsy: The liver weighed 2,800 Gm. The left hepatic duct was completely occluded by a tumor mass which completely replaced the left lobe of the liver and extended into the right lobe as well as into the pericholedochal nodes and the head of the pancreas. The right lobe of the liver contained a large cystic cavity, 15 cm. in diameter, which was filled with purulent material which cultured *E. coli*. There were in addition numerous black stones which measured 0.5 cm. to 2.5 cm. in diameter. The common bile duct measured 1.0 cm. and the right hepatic duct 0.5 cm. in diameter. Microscopic section revealed a well defined carcinoma of the bile ducts.

If calculi form in the intrahepatic ducts they are prone to be propelled by the flow of bile into the common duct and dependent upon their size may or may not be passed on into the duodenum. If they are retained in the intrahepatic ductal system they may completely occlude the same radicle within which they had their origin. There is then bile obstruction to that segment of the liver and one of several processes may take place: 1) The unit of liver involved may atrophy with scar formation. 2) The obstructed bile and bacterial growth may produce a localized cholangitis and even progress to abscess formation. This may subside or the abscess may rupture with a discharge of the calculus into

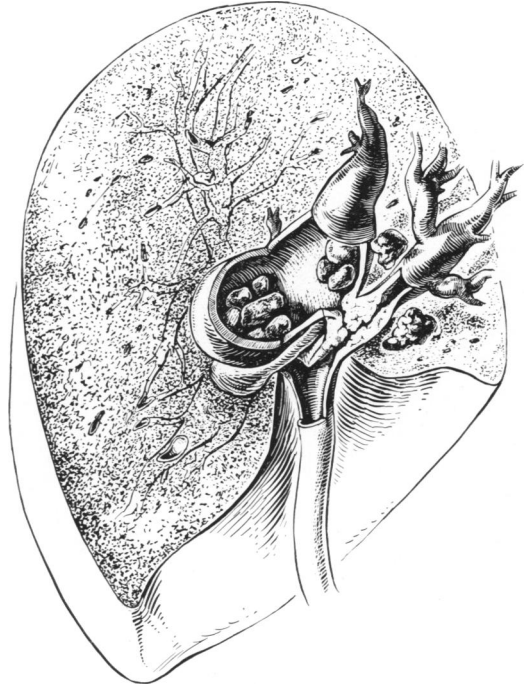


FIG. 7. Case 4. Drawing from postmortem specimen through section of medial portion of the right lobe. The right lobe of the liver contained a large cystic cavity measuring 15 cm. in diameter filled with purulent material and numerous black stones measuring from 0.5 cm. to 2.5 cm. in diameter. The left hepatic duct was completely occluded by a yellow tumor mass invading the adjacent structures. Microscopic section proved this to be a well-defined carcinoma of the bile duct.

the peritoneal cavity or an adjacent structure such as the intestine or the diaphragm. 3) The occluded duct may cause a dilatation of its tributaries proximal to it and the stone become dislodged so that the dammed up material passes into the larger ductal system. The calculus then increases in size as solid particles adhere to it and obstruction occurs again with the same sequence of events. Gradually the site of obstruction moves toward the junction of the main hepatic ducts. Perhaps some may correct themselves with the passage of the calculus or calculi into the common duct.

If calculi have existed previously or developed concomitantly in the gallbladder,

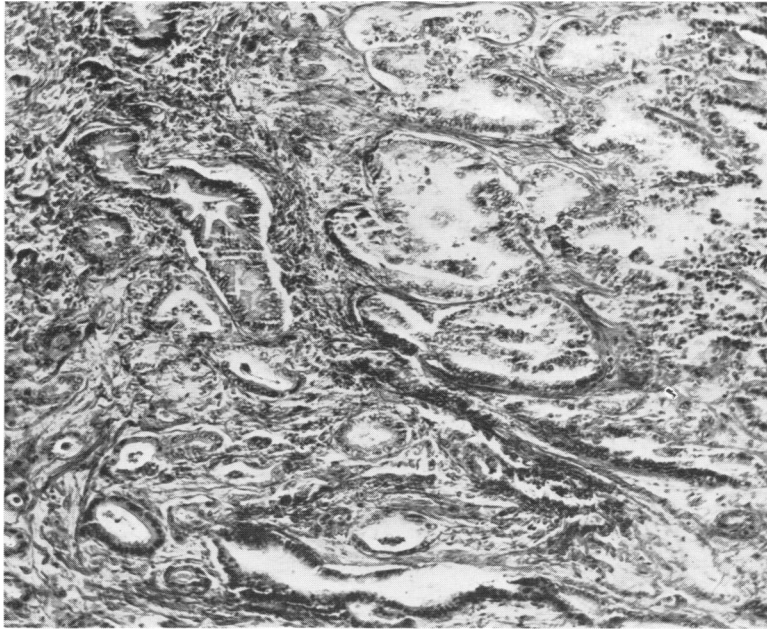


FIG. 8. Case 4. Photomicrograph ($\times 100$) manifesting carcinoma of the bile ducts. The tumor which consists of well formed ductal structures with a pleomorphic columnar epithelium is embedded in a fibrous matrix.

then the symptoms and signs that lead to operation may be ascribed to the more obvious and the gallbladder removed. The underlying pathology remains. From our experience we believe that in the usual course of events there are calculi retained in a cistern-like dilatation of an intrahepatic duct, and that particles of calcareous material are constantly being passed into the common duct. Here these fragments act as a nidus for large stone formation. These in turn may cause intermittent or complete common duct obstruction with its attending jaundice and other clinical manifestations. Operation is performed and the calculi in the common duct removed. Numerous large calculi may form again in the common duct within a few weeks or months causing obstruction. It is on the basis of this reasoning that it is proposed that any patient with a history of "recurrent" common duct calculi be evaluated as possibly having an intrahepatic cistern with calculi.

Dr. Olof Norman has described an anomaly of the left hepatic duct that we believe plays a role in intrahepatic cistern

formation. This consists of the left hepatic duct, within 2 cm. of its junction with the right, extending dorsally where it receives an anomalous communication from the right hepatic duct. Beyond this junction it curves slightly caudal and then turns anteriorly to give off its numerous branches in the left lobe. The role that this plays in stone formation is uncertain; it would seem to favor stagnation or incomplete emptying and is not readily accessible to exploration. Dr. Norman's description and cholangiograms of his case are almost duplicated by those of our patient (Case 4) who had a cistern type of dilatation of the left hepatic duct filled with calculi which had an anomalous duct communication with the right hepatic duct. In addition there was a carcinoma arising from the left hepatic duct just distal to its dilated portion. The deviation of the left hepatic duct to the right may readily be overlooked as an intrahepatic cistern appearing to be located in the right lobe. Thus it may be regarded as either an involvement of the right hepatic ductal system or a true intrahepatic accumulation separate and distinct

from either the right or left main hepatic ducts.

One of us (F. G. M.) reviewed with Dr. Kenneth Jensen, a member of the Department of Radiology, a series of 169 consecutive patients undergoing common duct explorations. Many of these had operative cholangiograms. In the course of their illnesses 20 patients were demonstrated to have an abnormality of the left hepatic duct and only two had changes in the right.

The findings believed to be pathologic were as follows:

| | |
|--|----|
| Poor filling of the left hepatic duct | 8 |
| Radiolucent defect in cholangiogram | 5 |
| Dilated left hepatic duct | 4 |
| Stones removed at exploration, left duct | 2 |
| Stenosis of left hepatic duct | 1 |
| Stones in right hepatic duct at autopsy | 1 |
| Dilated right hepatic duct | 1 |
| | — |
| | 22 |

In the 57 patients with intrahepatic stones collected from the literature by Rufanov 40 per cent had no calculi in the gallbladder and 60 per cent did.

| | |
|------------------------|----|
| Operation and autopsy | 25 |
| Operation only | 16 |
| Autopsy (no operation) | 16 |
| | — |
| Total | 57 |

Rufanov as we understand his report, believes that since 60 per cent of calculi in the intrahepatic ducts are associated with calculi in the gallbladder that they are secondary to cholelithiasis. He seems to adhere to the sequence of events being, stone formation in the gallbladder followed by appearance of calculi in the common duct, and then because of obstruction and ascending infection calculi form in the intrahepatic ducts. These intrahepatic calculi may cause obstruction of the ducts proximal to where they lodge or form. He also recognizes that up to 40 per cent of patients with intrahepatic calculi have acalculous gallbladders and therefore other mechanisms must be involved.

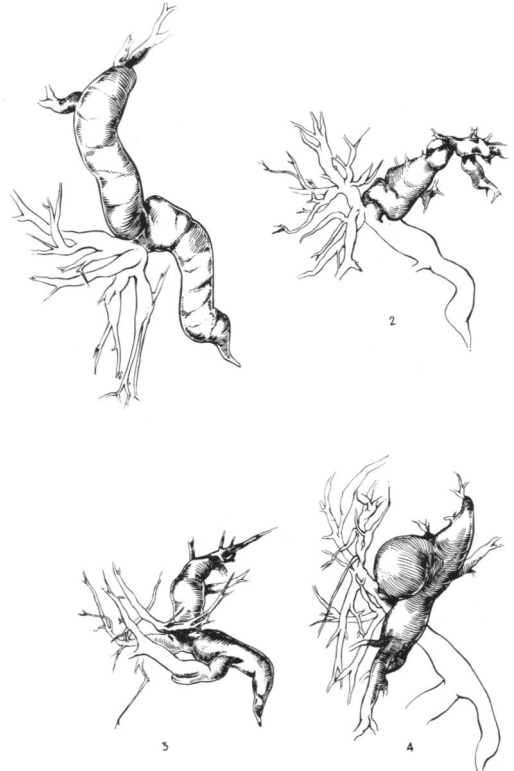


FIG. 9. Artist's sketch of ductal system of four patients (Case 1, 2, 3, 4) with intrahepatic calculi.

Intravenous cholangiography is in our opinion indicated whenever choledocholithiasis is suspected. In those patients who have had a previous cholecystectomy, with symptoms (persistent or recurrent) compatible with common duct calculi, it is one of the most important examinations in the over all evaluation. Direct and indirect evidence of calculi in the ductal system is usually obtained if stones are present, and sometimes their location and number can be demonstrated. By this examination also information suggestive of intrahepatic calculi may be gained. There are three items as bits of evidence to be looked for on the films: 1) The localized accumulation of the dye within the intrahepatic ductal system. 2) The visualization of the extrahepatic ductal system and the intrahepatic ductal system in one lobe and not the other. 3) Calcium deposition in liver parenchyma, alone or with 1) and 2), above.

At operation the liver should be meticulously scrutinized and evaluated. Because the left lobe is probably more frequently the site of intrahepatic calculi we prefer to examine it first, noting its comparative size, color and consistency. Division of the gastrohepatic ligament and attachment to the diaphragm will often facilitate such an examination. The palpation of the entire surface of the liver which is usually smooth may at times reveal an irregularity with changes in texture of the parenchyma adjacent to an abnormal process. In many instances the gallbladder has been previously removed and there may be scarring with distortion of structures in the subhepatic area. If there has been a previous choledochotomy and common duct drainage, then the scarring is greater and it will require considerable dissection to expose the hepatic hilum, the common duct at its bifurcation, and the adjacent liver. Here frequently are to be found indications of intrahepatic abnormalities.

It is desirable to visualize well the extrahepatic ductal system beginning proximal to the junction of the two hepatic ducts as they form the common duct. If there are calculi only in the common duct, then the entire ductal system may be distended. If there are intrahepatic calculi then there is usually a difference between the two main hepatic ducts, the calibre being less where the duct drains an intrahepatic cistern.

Gross differences between the lobes of the liver if present are evidence in favor of intrahepatic calculi. These vary from one patient to another and seem to depend upon the degree of obstruction, amount of infection and the duration of these processes. The uninvolved lobe appears normal, the lobe harboring an obstructed ductal cistern containing calculi may be enlarged, darker in color and edematous. These changes commonly associated with cholangitis may be combined with those of bile obstruction causing increased pig-

mentation. Bimanual palpation of the lobe may reveal an intrahepatic mass, a tumor, a ductal system containing calculi, or an obscure deep-seated area of induration. Atrophy of the involved lobe may have taken place if the obstructive process has been marked and is of long-standing. This is readily recognized in the left lobe on inspection and confirmed by palpation if calculi are present within the occluded ductal portion.

Before exploration of the common duct an operative cholangiogram provides important information. A small catheter inserted into the common duct is used to introduce 15 cc. of 30-per cent Urokon. The head of the table is lowered 20 degrees and the dye introduced by gravity. If the first roentgenogram shows poor visualization of the intrahepatic ductal system and escape of the dye into the duodenum, then the common duct distal to the site of introducing the catheter is temporarily occluded and another 15 cc. of the dye introduced by gravity. Views are taken in the A-P, lateral and $\frac{3}{4}$ or oblique position to further demonstrate the intrahepatic ducts.

The common duct should be incised longitudinally within 1 or 2 cm. of the bifurcation when intrahepatic calculi have been demonstrated. The nature and number of the calculi in the common duct should be noted and removed in an orderly and planned manner so that none will be overlooked. Digital examination of the intrahepatic duct that is involved should be attempted. Scarring or tumor may be encountered. A narrowed segment may be dilated to facilitate the removal of the calculi. When tumor is suspected then this should not be attempted but rather following pathological confirmation resection of the lobe considered. On the left this can be accomplished with little risk. When the tumor is present on the right additional factors require evaluation before reaching a decision. These include: 1) Right lobe

resection carries with it a high mortality rate. 2) Autopsy material suggests that tumor in the right lobe is often associated with widespread metastasis. 3) Unmolested the lesion has a grave prognosis; resection for such tumors have been reported so seldom that the results are statistically unpredictable.

In either lobe, if tumor can be reasonably excluded, and resection is not embarked upon, transhepatic exploration and drainage may be done. To accomplish this an incision is made through the liver parenchyma of sufficient length to permit direct vision into the dilated duct or cistern. This should facilitate the removal of any calculi with minimal additional trauma. Irrigation will sometimes flush gravel-like material that is present from the adjacent biliary radicles. Repeated palpation of the lobe, seeking variations in consistency that may be caused by blocked radicles and visualization of the ductal system by cholangiography should again be done to reduce the likelihood of residual calculi being overlooked.

With all calculi and debris removed from the liver ducts, a No. 12 (A) mushroom catheter is placed through the liver incision into the central area of the dilated segment and mattress sutures placed on each side to approximate the hepatic parenchyma. A T-tube is placed in the common duct in the usual manner. There should be a free flow of saline between T-tube to catheter without any leakage from the common duct and minimal leakage from the liver incision.

The T-tube may be removed after two weeks. This should be preceded by a post-operative cholangiogram to demonstrate the presence or absence of calculi and to determine if the intrahepatic cistern has decreased in size. The transhepatic mushroom catheter is left in place for an additional two weeks or longer. Before it is finally removed opaque material is intro-

duced through this again to provide a tangible record. The period of drainage should be governed by the decrease in the calibre of the ductal system, as well as the amount of calcareous and bacterial debris present in the bile.

Rufanov concluded from his review that because the operative results upon intrahepatic calculi were unsatisfactory, and because they were a cause of death, early operation upon calculous disease of the extrahepatic biliary system would be the most effectual therapy—actually one of prevention. He stressed that neglected biliary tract disease of prolonged standing in the aged was far less rewarding. In the general approach to the therapy of calculous disease of the biliary tract we are in agreement. However, we contend that intrahepatic calculi amongst our population who are relatively free from parasitic disease that involves the liver are more often due to anomalous anatomical variations and distortions of the intrahepatic ductal system. Studies that will provide information as to their incidence and extent are much needed. Such information would be of particular value when operating upon patients with biliary tract disease who presented certain unusual findings. For example, a patient who has a convincing history of biliary tract disease, a faintly visualized gallbladder and who at operation has no calculi in the extrahepatic biliary system but an enlarged left lobe merits cholangiography.

Huang Chih-Ch'iang recently reported four patients with intrahepatic stones associated with stricture of the hepatic ducts treated by hepatic lobectomy. Because intrahepatic stones are difficult to completely remove when there is a stricture of the intrahepatic duct distal to the calculi and because they tend to recur, he advocates hepatic lobectomy in selected patients. His criteria are the presence of a stricture of the intrahepatic duct containing calculi; atrophy and fibrosis of the

lobe; absence of calculi elsewhere in the liver.

It would appear to us that if the lobe is fibrotic and atrophic that bile production would be reduced and that the likelihood of new stone formation would be minimal. Hence we favor removal of calculi by transhepatic incision followed by a reasonable period of drainage and decompression by transhepatic catheter and a T-tube in the common duct supplemented by chemotherapy of penicillin and streptomycin. However, we believe that left hepatic lobectomy may be a better definitive procedure in selected patients and intend to employ it in the future.

Comment

Calculi within the intrahepatic ductal system may cause symptoms that closely mimic those produced by calculi elsewhere in the biliary tract. Where calculi are retained in a cysternal dilation of an intrahepatic duct they may be the source of "recurrent" common duct stones. The incidence of intrahepatic calculi is quite low in the United States when one considers the large number of patients treated surgically for calculous disease of the biliary tract. It is seen more frequently throughout the Far East, specifically in China and Japan. That there have been so few reports from America and Europe probably indicates that it is not only rare but it escapes recognition by both clinician and pathologist. Each must exercise extra effort to establish the diagnosis. The surgeon informed of this condition and suspecting it either because of a history of repeated operations for removal of common duct calculi, or because of findings at operation may establish the diagnosis by cholangiography. The pathologist at post-mortem examination, of course, has the optimum opportunity to see in detail the enlarged ducts, containing calculi and associated changes.

Summary

Intrahepatic calculi occur in patients with and without cholelithiasis. We have no reason to believe that the factors in stone formation are not the same in both. In America and Europe intrahepatic calculi are so rare that few surgeons have more than a limited experience in meeting the difficult problems presented by this disease. In the Far East the incidence appears greater. A brief review of the literature indicates that diagnosis is established late and often in the course of the surgical treatment of the complications that have occurred. It further indicates the limitations of the approaches most commonly reported. Four patients with intrahepatic calculi illustrating variations in the clinical course, findings at operation, and surgical procedures employed in their treatment are presented. An awareness of the condition should result in earlier recognition and more effectual management.

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