

CYST OF THE COMMON BILE-DUCT.

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TRUE cysts of the common bile-duct are rare, if we are to judge by the literature on the subject. Up to October, 1917, only thirty-five cases had been reported. As the diagnosis has never been made before operation, and as the operative treatment has not yielded good results, it seems important to place on record every case presenting this condition.

On May 12, 1916, a girl, age 11, was admitted into the Whitworth Hospital on account of progressive distention of the abdomen and abdominal pain. More than a year before admission the child experienced pain in the abdomen of such severity that she lay down and cried during attacks. Six months after the first attack of pain the abdomen began to swell, and steadily increased in size. The child got paler in colour, readily fatigued, and her appetite became poor. Slight transitory jaundice had been noticed on one occasion. With the exception of whooping-cough during infancy, she had been previously healthy. The parents were healthy and had had twelve children, of whom four died while 'teething,' and one was 'poisoned by a dose of bad whiskey.' Physical examination on admission showed the vascular, respiratory, and urinary systems to be normal. There was slight jaundice, and the abdomen was markedly distended, especially in the epigastric and right hypochondriac regions.

On palpation, a hard, irregular mass was felt in these regions. It extended into the right lumbar region, and its edge was palpable from the middle of the epigastrium towards the right anterior superior iliac spine, but its definition ceased at the right mammary plane. The tumour seemed to be continuous with the liver both on palpation and percussion, but respiratory mobility was absent. Blood examinations gave normal results. The

stools were normal; choluria synchronized with icteric attacks, but in the intervals the urine was normal except for the presence of excess of urobilinogen, which was demonstrated with Ehrlich's solution of dimethyl-para-amino-benzaldehyde and hydrochloric acid. The anatomical relations of the mass, the intermittent jaundice, and the indications afforded by the excess of urobilinogen in the urine, suggested that the lesion was intimately related to the liver.

At the request of Dr. Nesbitt I performed an exploratory laparotomy on May 26, 1916. The abdomen was opened by a vertical incision through the right rectus muscle. On opening the peritoneum, the duodenum and

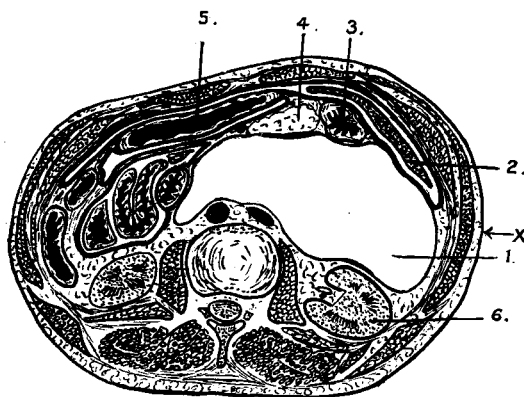


FIG. 431.—Cyst of common bile-duct. 1, Cyst; 2, Free edge of liver; 3, Duodenum; 4, Pancreas; 5, Stomach; 6, Right kidney. X, Needle introduced here. Level = L.V. 2.

hepatic flexure presented in the wound, being pushed forward by a large tense cyst. The gall-bladder was independent of the cyst and was not distended. The liver was cirrhotic.

On further examination the cyst was seen to lie in the position indicated in the diagram (Fig. 431). It extended from the extreme right side of the abdomen to a point

some distance to the left of the middle line, and lay between the aorta, vena cava, and right kidney behind, and the pancreas and duodenum in front. It was obviously a retro-peritoneal cyst, and it was decided to aspirate its contents. To avoid contamination of the peritoneum an exploring needle was introduced at X in *Fig. 431*, and pure normal bile was removed. An incision was then made into the cyst and a finger introduced. The anatomical relations were verified, but it was impossible to reach the left extremity of the cyst, or visually to establish its communication with the proximal or distal portions of the common bile-duct. Over a pint of bile was evacuated, and a small drainage tube inserted. At the end of a week this tube was removed, and within two months the wound had closed. The patient's general condition was much improved, and there was no return of abdominal pain or distention when she left hospital in July, 1916.

In March, 1917, she returned with the following history: A fortnight before, the abdomen began to swell, and in a few days some leakage of bile took place at the original site of drainage. The wound evidently became infected, for when she was brought to hospital she was profoundly toxic, temperature 102°, pulse 120, and the abdomen was greatly distended, tense, and tender. The original wound was immediately opened, and a large quantity of pus mixed with bile evacuated. The whole cavity was washed out with eusol and drained. The patient made a rapid recovery, and the wound closed five weeks after the operation.

I did not see the patient again until October, 1918, when she came to hospital for examination at my request. The wound had remained healed, there was no return of abdominal pain or distention, and nothing definite was palpable in the abdomen, although there was slight fullness in the right hypochondrium. In March, 1919, the patient returned to hospital complaining of loss of appetite and occasional abdominal pain localized in the region of the gall-bladder. The physical signs were similar to those presented six months previously. I decided to perform a laparotomy in order to ascertain the condition of the cyst, and, if necessary, to make an anastomosis between it and the duodenum.

When the abdomen was opened, I was surprised to find that the cyst had shrunk to the size of a walnut, and occupied a position between the second stage of the duodenum and the right kidney. The cirrhotic process in the liver was more pronounced, but no other abnormality was encountered. As the patient had no jaundice, and as the cyst had contracted to such an extent, I did not perform an anastomosis, but closed the abdomen. The patient made an uneventful recovery. I saw her again in October, and her condition showed neither deterioration nor improvement. She still had occasional attacks of pain in the right hypochondrium, and the appetite was poor. She looked rather fat, but the appearance was due to excess of subcutaneous tissue rather than to a deposit of fat therein. She was mentally slow and apathetic. Her general appearance was suggestive of thyroid insufficiency. Thyroid extract was prescribed, and when I saw the patient three months afterwards the change for the better was remarkable. She was bright and alert, had lost the puffy appearance, and felt perfectly well, although her appetite was not keen.

The literature on this subject has been admirably summarized by E. Waller, of Stockholm, in the *Annals of Surgery* of October, 1917. As his paper is so recent it is unnecessary to deal exhaustively with reports of cases in this communication. It is of interest, however, to note that the first case resembling the one described was a patient in the Whitworth Hospital in 1817. It is reported in the *Dublin Hospital Reports* of that year by Dr. Todd. The salient features of the thirty-six cases (including my own) reported may be presented as follows:—

Age.—The average age was 13, the youngest patient being 2 years old, the oldest 25. But Heiliger, according to Waller, reported the condition in a mature male foetus in 1910.

Sex.—Five of the patients were males and the rest females.

Pathology.—The pathological condition found is a large cyst of the upper and middle

portions of the common bile-duct ; the lower part is not affected, nor are the hepatic ducts dilated. Some of the cases described are to be considered as those of excessive dilatation of the bile-passages rather than true cysts of the common duct. The usual size of the cyst when discovered is that of a man's head. A capacity of four to five litres has been recorded. There is considerable uniformity in the anatomical displacements produced by the slowly distending duct. The duodenum is pushed forwards and medially. In two cases, including the one presented, its second stage was flattened out against the anterior abdominal wall. The transverse colon is displaced downwards and towards the left. In advanced cases the gross appearance of the liver suggests biliary cirrhosis.

Etiology.—The fact that a cyst of the common bile-duct, 3 cm. wide and $2\frac{1}{2}$ cm. long, was found in a mature male foetus, suggests that the condition is probably congenital. In this foetus, bile was present in the intestine and there was no icteric discoloration of the tissues. It seems likely that the congenital condition may exist for a variable time before giving rise to symptoms. Whether the onset of symptoms is the inevitable consequence of the congenital abnormality, or whether it depends on some exciting cause such as gall-stones or local inflammation, is at present undecided.

Symptoms.—In only one case was jaundice absent. In two there was slight tingeing of the sclerotic and a small amount of bile in the urine ; but in the remainder jaundice of marked degree was a striking symptom. It is usually intermittent, without complete remissions.

The presence of an abdominal tumour was the prominent physical sign in every case. The tumour was situated in the right hypochondrium, and extended into adjacent regions according to its size. It was sometimes cystic, but often hard ; when the former it simulated a pancreatic cyst, a dilated gall-bladder, or a hydronephrotic kidney ; when the latter, growths of the colon, pancreas, or liver. In most cases the tumour had a very slight degree of mobility. In a few cases which were under observation for a lengthy period, the size and consistency of the tumour were observed to increase some hours after the ingestion of food.

Attacks of colicky pain are usually present, and sometimes exceedingly severe. They may precede the onset of abdominal swelling by some months, as in my case, and are sometimes associated with fever.

Ascites has resulted from pressure on the portal vein in three cases. Intestinal obstruction was a complication in one.

Diagnosis.—The diagnosis in an established case should present few difficulties if the possibility of such a condition is realized. The combination of jaundice, pain, and large tumour in a young individual should indicate the diagnosis.

Attacks of severe pain in the gall-bladder region, with deepening of the jaundice at the end of the paroxysms, may suggest the presence of stone in the common duct, but the presence of a large tumour is not in accordance with the usual symptoms of that condition, nor are gall-stones common in youthful patients. In several cases reported, however, jaundice and pain preceded the appearance of the tumour by a long period, in one case by two and a half years. In such cases the symptoms may resemble those of biliary cirrhosis, especially when the paroxysms of pain are accompanied by fever. The similarity may be accentuated on percussion by finding an increase of liver dullness due to the intimate relation of the liver to the cyst, which is not of itself palpable. There is, however, no enlargement of the spleen, and the pain is of a more acute nature in cysts of the common duct. In one case jaundice was absent ; in this, the paroxysms of pain and the tumour were the only symptoms. When jaundice is present, the involvement of the liver or biliary passages is indicated ; but when pain is the only symptom or tumour the only sign, or when pain and tumour are associated, the relation of the liver and its ducts to the condition is not so clear.

It is in cases of the latter type that I strongly advocate the employment of Ehrlich's test for urobilinogen in the urine, though it should prove of value even when the three cardinal symptoms are present. In every case in which this reaction was positive we have

found definite organic disease in the liver. The reaction depends on the following physiological facts: Hæmoglobin is broken up in the liver into hæmatin and globin; hæmatin undergoes dissociation into iron and bilirubin; the latter is excreted in the bile. When bilirubin reaches the intestine it is reduced by bacteria to urobilinogen, some of which is excreted in the fæces, and some is absorbed by the portal circulation and returns to the liver, where it is converted into bilirubin, thus completing a circulation of this substance. If hepatic activities are modified by disease, the liver fails to transform urobilinogen into bilirubin, and the former is absorbed into the general circulation, to be excreted in the urine.

The test has all the advantages of simplicity. A few drops of Ehrlich's reagent are added to a quarter of a test tube of the patient's urine. In a few seconds a rose-pink colour develops when the reaction is positive. All Dr. Nesbitt's experiments were carefully controlled, and the conclusions which I have given above were reached.

Any urine gives the reaction on boiling. The urine must be fresh; otherwise urobilinogen becomes oxidized to urobilin, the recognition of which is a much more complicated and uncertain matter. This test is, of course, of use in many other conditions than the one under discussion, but its utility is probably greater when a cyst of the common bile-duct is suspected, since in this affection there are so few collateral symptoms. When such a biliary cyst resembles a hydronephrotic kidney on physical examination, the correct diagnosis might be established by a positive reaction to Ehrlich's reagent; but a pyelograph of the right kidney would exclude that organ with greater certainty.

Treatment.—That the successful treatment of the condition is a matter of great difficulty can be seen by the results of operations.

RESULTS OF TREATMENT IN THIRTY-SIX CASES.

1ST OPERATION	2ND OPERATION	NO. OF CASES	DIED	RECOVERIES AND REMARKS
Drainage	—	18	18	1 recovered from operation, lived 3 years with fistula, died of phthisis
Drainage and choledoch-enterostomy	—	1	1	0
Drainage	Drainage	1	0	Living 3 years 8 months; fistula closed 2 years 8 months
Drainage	Attempt at choledoch-enterostomy	3	3	0
Drainage	Choledochenterostomy	3	0	3
Extirpation	—	3	3	0
Choledochenterostomy	—	2	0	2
Unoperated	—	4	—	—
Fœtus	—	1	—	—
	Totals	36	25	6

The fact that each surgeon encountered a condition previously unknown to him must account in some degree for the high mortality; but the prime factor in this mortality was the neglect of the surgical principle that a sterile cavity should not be drained. Infection is practically unavoidable if drainage is established. That aseptic drainage *can* be carried out is beyond dispute, but the probability of infection is in direct proportion to the duration of the external fistula.

Of the 19 cases treated by drainage alone, 18 died, and the single case which survived

had to be operated on a second time for infection of the cyst. This acute inflammation appeared to be curative in the case reported, for there was no subsequent dilatation; but such a result must be regarded as due to an 'act of God' rather than to surgical skill or judgement.

The results of extirpation were uniformly bad, and were in large part due to the fact that the surgeon did not know what he was extirpating. Given such knowledge it might be possible to perform extirpation in spite of its difficulties, and re-form the duct after its ancient image; but if any simpler procedure is feasible it should be adopted.

Of the 6 cases which recovered, an anastomosis between the cyst and the intestine was performed in 5. The difficulties of this operation as a secondary procedure are considerable, as can be judged by the fact that three surgeons have attempted it and failed, their failure leading to a fatal result. Such a primary operation could have been performed in my case between the first stage of the duodenum and the cyst.

The steps of the operation which I suggest are as follows: (1) Make or confirm the diagnosis on opening the abdomen; (2) Further confirm the diagnosis by aspirating the cyst retroperitoneally, as is usually possible; (3) When the cyst wall has collapsed, perform an anastomosis between it and the duodenum as in a cholecystenterostomy.

I have retained the descriptive term 'cyst of the common bile-duct' in order to bring this case into line with similar cases reported. The condition is, however, not a cyst but a diverticulum of the duct, and it would be better to designate it in future under this appellation.