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The opinion on the cystic pathology of the biliary tract, based on their own clinical experience

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Abstract

The experience of studying the diagnosis and the surgical treatment of bile duct cysts by the domestic and foreign researchers is generalized. The authors point out that the clinical picture of this pathology is usually scanty and nonspecific. The main initial diagnostic method is ultrasonic echolocation. In most cases CT, NMR, ERCPG, etc are also necessary. However, the final diagnosis is established after histological examination. A radical method of the treatment is the surgical operation. The nature and extent of the intervention depends on the type of the cysts. Complete removal of the cystic formations with an adequate drainage for decompression of the biliary tract is the standard radical surgical treatment of this category of the patients. All these patients need subsequent periodic examinations to identify possible malignancy.

Key words: bile duct cysts, cystic pathology, biliary tract, choledochal cysts, endoscopic retrograde cholangiograms, malignancies in the cysts.

Cystic dilations of the gallbladder, involving extrahepatic and intrahepatic bile ducts are quite rare anomalies. Although this pathology was first documented by Vater and Ezler in 1723, the first complete clinical description of this defect was published in 1852 by Douglas [1, 2]. At that time, these changes were considered congenital McWhorter G.L.in 1924 [2]. In 1959 Alonso-Lej et al. published a detailed review of 94 cases of common bile duct cysts added two own cases. They classified the mentioned cysts into 3 types [3]. In 1977, Todani et al. divided the bile duct cysts into 5 types, adding 2 new types (types IV and V) [4]. The subtypes found in cholangiography were also described.

Now the classification is as follows:

- Type 1 (Figure 1) - Cystic sac or spindle-shaped dilatation of the common bile duct (most common type, 90-95% of cases).

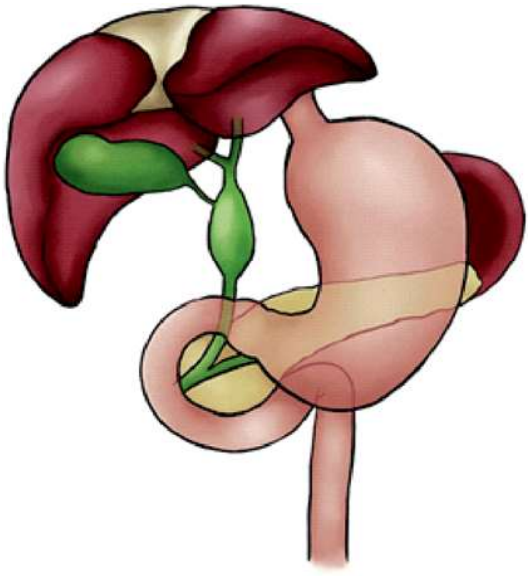

Fig. 1. Cyst of the bile duct of type 1.

- Type 1A is bag-shaped configuration, involves the entire common bile duct or most of it.
- Type 1B is bag-shaped configuration and involves a limited segment of the bile duct.
- Type 1C is more spindly in configuration and involves most or all of the hepatitis choledoch.

Type 2 (Figure 2) - Diverticulum of the common bile duct.

- Type 3 (Figure 3) -Hleedochocele, cystic dilatation of the distal part of the common bile duct.
- Type 4 (Figure 4) - Cystic sac or spindle-shaped dilatation of the common bile duct, associated with a cystic spindle-shaped or saccule dilatation of the intrahepatic bile ducts.
- Type 5 (Figure 5) - Cystic spindle-shaped, or saccule dilatation of intrahepatic bile ducts, associated with the normal common bile duct; can be associated with hepatic fibrosis (the association is referred to as Caroli's disease) [5].

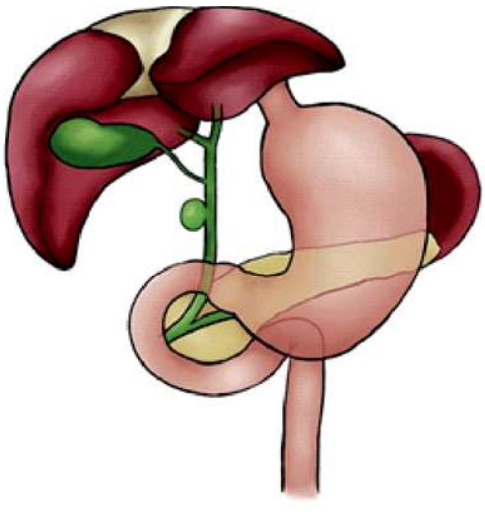

Fig. 2. Cyst of the bile duct of type 2.

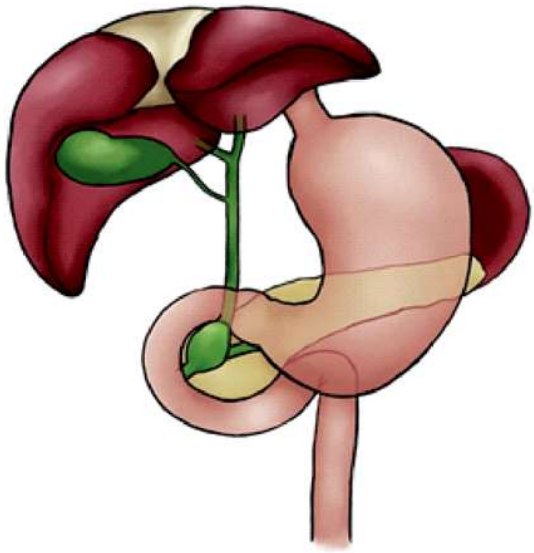

Fig. 3. Cyst of the bile duct of type 3.

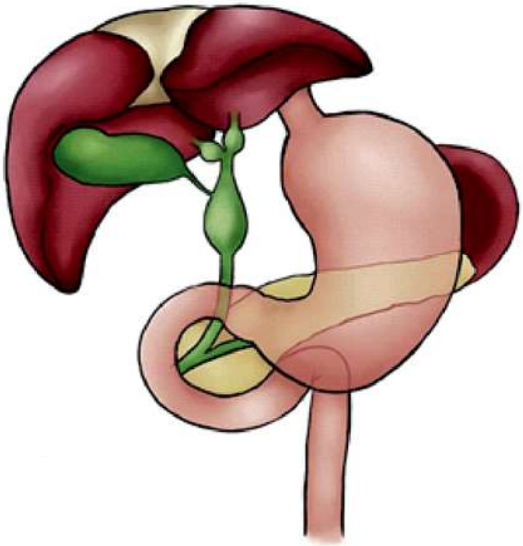

Fig. 4. Cyst of the bile duct of type 4.

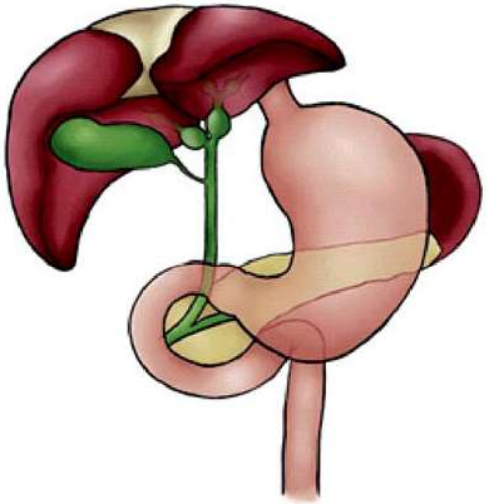

Fig. 5. Cyst of the bile duct of type 5.

Frequency: The prevalence of the bile cysts is from approximately 1 case in 13,000 to 1 in 2,000,000 people. The bile ducts are much more common in Asia than in Western countries [6]. Approximately 33-50% of the reports about this pathology come from Japan, where the frequency of these cysts is close to 1 case per 1000 population [7,8]. In an extensive survey published in 1980, Yamaguchi et al. examined 1,433 cases, 1,204 (more than two-thirds) of which were from Japan [9].

It is known that type 1 is the most frequent. In types 1 and 4, the ratio between women and men is approximately 4: 1, and in types 2, 3 and 5, cysts occur with equal frequency in both sexes.

Cysts of the bile ducts can be found in people of any age. Two-thirds of them are detected in children under 10 years [10,11,12,13,14,15,16]. About 20% of cysts are found in elderly people [17,18,19,20]. In rare cases, cysts of the common bile duct were found in prenatal ultrasonography during pregnancy 15 weeks, shortly after birth, the children underwent a surgical treatment [21].

Pathogenesis

The exact cause of the formation of the bile duct cysts remains unclear. Many authors believe that they are congenital, because most cysts are diagnosed in infants and children. However, due to the fact that approximately 20% of hepatitis choledochal cysts are diagnosed in adults, including elderly patients, allowed to assume several more development mechanisms [14,17,22,23,24]:

- Weakness of the bile duct wall [6,25,26].
- Development of the distal department of choledochus [27].
- A combination of the duct obstruction and weakness of the wall [28].
- Reflux of pancreatic enzymes into the common bile duct, a secondary anomaly of the pancreatobiliary compound [18,29,30,31,32,33].

In 1969, Babbitt and colleagues, after analyzing the cholangiograms of the patients with the common bile duct cysts, found in most of them an anomaly of the pancreatobiliary pancreatobiliary communication in which the pancreatic duct was more proximal. They hypothesized that such an anomaly caused reflux of pancreatic secretion in holedoch, as the pressure in the pancreatic duct was higher than the pressure in the bile duct. It was concluded that the classical triad: fever, abdominal pain, and jaundice occurred due to the recurrent cholangitis attacks. As a result of inflammation, the choledochal wall was damaged, followed by healing and thickening, which led to the obstruction in the distal part of the bile duct [29].

In 1977, Spitz supported the concept of distal obstruction of the common bile duct as the cause of the formation of the choledochal cysts, demonstrating the

dilatation of the bile duct in lambs obtained by ligating the duct near the confluence of the duodenum [27]. The same experiment was performed to simulate the dilatation of the bile duct in mature sheep.

In 1974, Kato et al. were the first researchers who created the cyst of the common bile duct in experimental animals by transpupillary curettage of the bile duct and subsequent, 3-4 days later, ligation of the ampulla of the falcon papilla [28].

In 1984, Todani et al. performed an analysis of the endoscopic retrograde cholangiograms (cholangiopancreatograms) and confirmed this anomaly in the common canal [33]. Cystic lesions were found in most patients. There are other authors' reports of the same research results [18,30,31,34,35]. Reflux pancreatic enzymes in the common bile duct can occur quite early, even in embryonic period, leading to the damage of the duct wall. The distal part of the choledoch is the most vulnerable, and with repeated injuries, its stenosis may occur.

Experimental support of this concept was reported by Kato et al. in 1974. They imposed an anastomosis between the main pancreatic duct and the gallbladder in dogs. Within 9 days after the application of the anastomosis, all the animals studied had different degrees of dilatation of the common bile duct, with edematic changes in its wall. They concluded that proteolytic enzymes were responsible for this damage [28].

Miyano et al. (1981 and 1984) created an experimental model of an abnormal choledochocampanic compound, creating an end-to-side choledochocinchoracic anastomosis in puppies. They successfully reproduced the dilation of choledoch in all experimental animals without exception [31,32].

All these theories are applicable to types 1, 3 and 4, but they cannot be used to explain 2 and 5 types of the common bile duct cysts in which the common bile duct is normal. Perhaps genetic factors play a major role [6,8,23]. Thus, at present there are two most well-founded theories - reflux of pancreatic enzymes into the common bile duct, with an abnormal pancreatobiliary compound and obstruction of the distal part of the choledochus [36].

Pathological features

The size of the cyst of the common bile duct of type 1 is very variable [6,11,12,17,19,34,37,38]. The volume of the cyst can reach up to several hundred milliliters of bile with pancreatic enzymes. The thickness of the cyst wall is also variable.

Intrahepatic cysts can be fusiform or saccular and are connected with the common bile duct. Suspension and stones are sometimes present within the cyst [6,8]. The bile duct, distal to the cyst, is usually stenosed. The liver can have a different level of fibrosis or cirrhosis with portal hypertension. A histological examination of the wall of the cyst of the common bile duct revealed a dense fibrous connective tissue with the inflammation and the formation of mucosal and submucous layers.

The inflammation is much less developed in young patients than in older patients [8,35,39,40]. The cyst is lined with a thin, fragmented cover, but not the normal lining of the biliary tract.

The inflammatory process with the intrahepatic cyst location is more frequent than in the extrahepatic cyst location.

Histological examination: the signs of chronic inflammation are observed in the wall of the cyst. It is thin, fibrous, and often devoid of a true epithelial surface, although it can be lined with a low columnar epithelium. It is noted that the infants can develop a complete obstruction of the distal part of the common bile duct as a result of an acute or chronic inflammation. In the liver, there may be intraductal fibrosis and portal swelling. Cirrhosis like changes may occur in adults with chronical illness. The most unpleasant histological finding is cholangiocarcinoma.

Cysts of common bile duct and malignancy

The possibility of cancer in the wall of the cyst of the common bile duct or in the remaining gall-stone after complete cyst resection is a recognized fact. Malignant development is believed to be the result of a prolonged stagnation of the bile and chronic inflammation with metaplasia. Typical malignant development - adenoskvamozny cancer or small cell cancer.

Malignancies in the cysts of the common bile duct can undergo the distal section of the choledochus, more than half the cases of the cyst wall (even after successful internal drainage), or intrahepatic bile ducts. The complete cyst resection does not prevent the risk of malignant degeneration in the remaining bile ducts. The risk of cancer increases with the age of the patient [4,14,41,42,43,44,45]. So the detection of malignancy in the resected cyst is 0.7% in patients operated up to 10 years of age; 6.8% in patients operated in 11-20 years; and 14.3% in patients operated after 20 years. Malignancy can occur many years after the removal of the cyst and develop in areas of bilious tree distant from the cyst, for example, the gallbladder.

Any type of the cyst can be prone to malignant transformation, but the greatest prevalence of the oncological process is observed in types 1, 4, 5. The increased risk of malignancy of the bile tree, even after radical surgery, requires monitoring of such patients.

Lethality / Complicated course: variants of complicated course associated with the bile duct cysts depend on the age of the patients. Infants and children may develop pancreatitis, cholangitis and signs of hepatocellular injury. In adults with subclinical duct inflamma-

tion and bile congestion that may have been present for many years, one or more severe complications such as pancreatitis, choledocholithiasis, cholangitis, intrahepatic abscesses, portal hypertension, cirrhosis may occur. Cholangiocarcinoma is the most dangerous complication of choledoch cysts. Lethality varies depending on the level of the complication.

Prenatal diagnosis

In connection with the development of prenatal ultrasound, an increasing number of the common bile cysts in the fetus was reported: Hammada, 1998; Lipset, 1994; Marchildon, 1988; Mackenzie, 2001; Shamberger, 1995 [17,21,46,47,48,49]. Incomplete obstruction of a large cyst is one of the typical clinical manifestations in newborns and infants [50]. The earliest cyst of the common bile duct reported is found in the fetus during gestation of 15 weeks, which may correspond to the time of formation of the pancreatic enzymes.

Prenatal detection of the cystic structure in the extrahepatic sections of the bile duct suggests a diagnosis of choledochal cyst, after which the diagnosis must be verified by successive ultrasound examinations. In most surgical clinics, they prefer to remove the cyst shortly after birth. Within a few weeks, it is necessary to stabilize the child's condition and carry out the necessary examinations. The surgical treatment in newborns has shown that it is technically simple and well tolerated by the patients.

Clinical manifestations

Patients with choledochal cysts are divided into two groups according tot their age [12,14,17,39,40].

The first group includes infants younger than 1 year with hepatomegaly or without an obvious increase in liver size, with obstructive jaundice and acholic feces. This clinical picture is indistinguishable from that which occurs with biliary atresia, in the absence of palpation of the formations (cysts) of the right half of the abdominal cavity. The presence of cystic formation may be suspected during clinical examination and is confirmed on ultrasonography, only thus the diagnosis of the cyst choledoch is competent.

In 1995, Todani et al. found that 26 out of 28 infants with a clinical picture younger than 1 year had cysts of choledoch, whereas only 3 out of 8 children aged 13-24 months had that pathology [40]. Other symptoms, such as vomiting, fever, and abdominal pain with hyperamilazemia were not often detected [19]. In infants diagnosed with a common bile duct cyst in the prenatal period, jaundice often didn't not appear until 1-3 weeks after birth [51,52].

In contrast, the second group, infants over 1 year old, with the so-called adult form of the cyst of the common bile duct, generally had 1 or more components of the classical triad: pain, jaundice, and palpation of the cyst. The entire triad is present in less than 30% of patients [14]. Jaundice and intermittent fever were often associated with an undefined abdominal pain. The pain was associated with developing cholangitis and moderate chronic pancreatitis.

Unidentified cysts of the common bile duct can lead to choledocholithiasis, cirrhosis of the liver with portal hypertension, rupture of the cyst, and cancer of the biliary tract.

Laboratory tests that may be useful for diagnosis and preoperative assessment of patients with bile duct cysts include: direct bilirubin, alkaline phosphatase, serum glutamine oxalate transaminase (SGOT), serum glutamine pyruvate transaminase (SGPT), gamma-glutamyl transferase (GGT), a coagulation system, a clinical blood test.

None of these tests is determinative for the diagnosis of the common bile duct cyst.

Instrumental methods of research.

1. Ultrasonography is the best initial study, allowing to detect the changes in the bile duct and liver. In newborns, this may be the only method of investigation. In the antenatal period Sgro and colleagues detected Caroli's disease (2004) only with the help of ultrasound [23].

2. CT and NMR help to outline the anatomy of the lesion and the structures closest to it, and can also determine the presence of an enlargement of the intrahepatic part of the bile ducts. Adult patients may benefit from CT scans in combination with cholangiography.

3. Radiography with cholangiography, when giving contrast per os or intravenous administration, is of limited use and is considered obsolete.

4. Scintigraphy with 99Tc diisopropyl iminodiacetate can show complete obstruction of the distal bile duct [14,17].

5. Endoscopic retrograde cholangiopancreatography (ERCP) is the choice in older patients. In experienced hands, ERCP can be performed with a high degree of success, even in infants. Successfully implemented ERCPH objectively demonstrates the anatomical relationship of the pancreatic-biliary compound [4,11,33,34]. Percutaneous transhepatic cholangiography can also be used for indications.

6. The method of magnetic resonance with cholangiopancreatography has more diagnostic value compare to cholangiography and ERCP in patients with choledochal and biliary ducts in general [31,53,54,55,56].

Treatment

The basis of treatment is the surgical removal of the cyst with subsequent decompression of the biliary tract, except type 5 (with numerous intrahepatic formations). The tactic of the surgical treatment of patients with the

cysts of the common bile duct has developed most during the last three decades. At present, a complete resection of the cyst and the gallbladder has become the main method of choice. Other operations, such as cystoduodenostomy or cystojunostomy, cannot be considered radical, besides they have a high risk of complications and the greatest potential risk of malignant degeneration in the residual cyst. Even after a complete resection of the cyst, sporadic cases of cancer in the remaining gall bladder occur.

In the past, aspiration external drainage was used, which is a simple and quick procedure, although very painful. However, external drainage of the gall bladder was often accompanied by numerous complications, including recurrent cholangitis and biliary fistula. Mortality rates were also high [6,14,17].

Internal drainage: cystoduodenoanastomosis or cystojunoanastomosis, also used in the past. In these variants of operations, there was free reflux of pancreatic enzymes in the cyst through an abnormal pancreatobiliary compound that led to the development of cholangitis, the formation of anastomotic stricture, and the possibility of cancer of the biliary tract. Among the patients undergoing cystoduodenoanastomosis or cystojunoanastomosis, 60% remained on symptomatic treatment, and 40% required re-operation [57,58].

Complete removal of the cyst in types 1, 2 and 4, accompanied by the reconstruction of gall bladder with hepatocojunostomy (method Roux) is widespread, as the preferred method in the treatment of cysts common bile duct. This procedure involves resection of the distal choledochus, which blocks the reflux of pancreatic enzymes and reduces the risk of malignancy of the bile ducts.

Complete removal of the cyst is possible in infants and young children. In adult patients with recurrent cholangitis and inflammation, resection of the anterolateral part of the cyst is indicated, leaving the wall adjacent to the portal vein. This technique is also the most promising in patients who have undergone cystoenterostomy and require a second operation, due to repeated attacks of cholangitis.

Intraoperative cholangiography is performed by puncturing the cyst or gall bladder. It determines the exact anatomy of the cyst of the common bile duct and its relationship to the pancreas. Usually cholecystectomy is performed simultaneously.

Biliary reconstructions can most often be performed with hepatitis-neurastomy on Roux, as high as possible [8,11,39,60]. However, some authors, including Raffensperger and Shamberger, used the jejunum segment to prevent reflux. This idea was not accepted by everyone. Installation of any stands is usually not shown [49,61,62,63].

With type 2 cysts of the common bile duct, a simple removal of the diverticulum with ductoplasty for the reconstruction of the common bile duct is all that is required. Laparoscopic removal was successfully performed with this rather rare type of cyst in 2000 [64].

With type 3 cysts of the common bile duct, duodenotomy with separate drainage of the biliary and pancreatic ducts directly into the duodenum is shown [34,60,65].

In patients with type 4 cysts of the common bile duct with intrahepatic cysts, each case should be evaluated individually, necessarily adequate drainage of the gallbladder. Resection of the enlarged extrahepatic bile ducts should be performed to the liver gates, followed by hepatitis-yoanastomosis at the level of the liver gates, can provide adequate outflow of the bile and effective decompression of the intrahepatic cysts. If the intrahepatic cysts are localized in a limited part of the liver, partial liver resection is indicated [18,66,67].

At type 5 of the cyst of the common bile duct, patients with a limited spread of cysts are shown hepatic lobectomy. If the disease is disseminated, affecting both liver lobes, the treatment is palliative or, in exceptional cases, liver transplantation may be required [14,60,68,69,70].

Complications after the surgical treatment were mainly observed in patients with types 1, 4 and 5 cysts. Mortality and relapse of the disease are low after surgical removal of cystic formations, in comparison with the methods of their internal drainage.

Postoperative complications:

1. Holangite.
2. Formation of stones.
3. Constriction of anastomosis.
4. Dilation of intrahepatic bile ducts, especially with 4 and 5 types of cysts of the common bile duct.
5. Malignancy.

We have been studying this problem for more than 30 years, since the first time we encountered cystic formation of extrahepatic bile ducts. During the planned operation for chronic calculous cholecystitis, a local expansion of hepatiko choledocha was detected, conditionally, in the region of the confluence of the vesicular duct. This education was initially taken for the region of the "neck" of the gallbladder and the surgeon, being sure that he had produced only cholecystectomy, resected part of the extrahepatic bile duct. With intraoperative cholangiography (the terminal part of the supraduodenal department of choledocha was taken for the stump of the vesicular duct), it became clear what happened. In the operating room, Professor E.I. Brekhov. Taking into account what has happened, reconstruction to Roux. The postoperative period proceeded without complications. The patient was discharged on the 12th

day after the operation. Diagnosis at discharge: Cyst of common bile duct, type 1B. With a histological examination, the diagnosis of the cyst is confirmed. Later it was observed for 5 years. Data for the stricture of anastomosis and malignancy were not revealed.

In the following, about 23 cysts of the bile ducts were performed. Of these, with the established diagnosis of the cyst of the bile duct of type 1, 8 patients were routinely operated. All of them were subjected to a resection of the cystic-altered extrahepatic bile ducts and hepatic neuronostomy according to Roux.

Also, the patient with the diagnosis was prepared for the operation in a planned order: Peptic ulcer of duodenal ulcer, complicated by stenosis of the outlet stomach. During the preoperative examination, in addition to the above pathology, the hepaticocholedochus cyst, a rounded shape, 5.0 cm in diameter was detected. A simultaneous operation was performed: 2/3 resection of the stomach according to Balfour. Cholecystectomy. Resection of hepatitis choledoch. Hepatikojannoanastomoz end-to-side. The cystic-altered part of the hepaticocholedochus is resected proximally by 1.0 cm below the bifurcation of the right and left hepatic ducts, and in the distal slightly above the border of the supraduodenal and retro-duodenal parts. The distal part is sewn with a double-suture. Gastroentero and hepatitis yunoanastomoz is divided by a mechanical seam.

In 12 cases, as in our first observation, the cysts of extrahepatic bile ducts are diagnosed intraoperatively. Resections of hepatiko-choledocha with cystic lesions of various volumes were performed. In 4 cases, a hepatopojunostomia was performed according to Roux, and in 8, hepatitis yunoanastomosis with entero-entero anastomosis was applied.

Two cases require a separate mention, due to the fact that the diagnosis of cysts was not performed intraoperatively and the situation is regarded as iatrogenic technical error.

In the first observation, when performing a cholecystectomy, a significant part of the hepatoc choledocha was resected, in the proximal part above the fusion of the right and left hepatic ducts. In connection with the extent of the defeat, a reconstructive-reconstructive operation was impossible. The external drainage of both hepatic ducts has been performed, the safety drainage has been placed in the abdominal cavity. In the future, the safety drainage was removed and the patient was discharged for outpatient treatment. Repeated hospitalization after 4 months. Reconstructive surgery was performed anastomoz in Roux.

In the other case, from the second day after the planned laparoscopic cholecystectomy, the progressive icterus of the skin and the sclera were noted. The RCCP was performed, in which the filling of the terminal part of the common bile duct with filling of the contrast medium was detected during 4.5 cm, there was no further contrasting. Iatrogenic trauma of extrahepatic bile ducts is suspected. Transhepatic drainage of the biliary tract and cholangiography were performed, which showed contrasting intrahepatic, right and left hepatic bile ducts. The filling of the common bile duct with contrast medium is about 1.0 cm. The patient was discharged from the hospital and two months later she was hospitalized again for a reconstructive-reconstructive operation.

Hepatic fusion on the Roux was made. The postoperative period proceeded without complications.

In both cases, morphological examination was used to diagnose cysts (type 1) in the resected parts of the hepaticocholedochus.

Thus, based on the literature and our own research, we can formulate the following conclusions:

1. Patients with suspected cystic changes in the bile duct require a thorough and comprehensive study of the biliopancreato-duodenal region.

2. To resolve the issue of surgical (operative) treatment, it is necessary to determine the nature and type of cystic lesions, clear topographic and anatomical relationships with neighboring organs.

3. The operation of "choice" for cysts of the first type is a resection of hepatitis choledoch with hepatitis-inhurations according to Roux. However, based on our observation with exclusive combined pathology, other variants of reconstructions are possible.

4. In emergency situations, when detecting both the cysts and iatrogenic intraoperatively, it is possible to make simpler decisions that do not complicate the situation, including the choice of anastomosing and external drainage.

5. In spite of the fact that the results of surgical treatment with the pathological changes under consideration are, as a rule, good, the risk of malignancy takes place. Therefore, all patients in this category should be under observation for at least 5 years.

Summarizing the above, it can be stated that the complete removal of the cyst with adequate drainage for decompression of the biliary tract is the standard radical surgical treatment of this category of the patients. All these patients need subsequent periodic examinations for the purpose of early detection of complications and possible malignancy.

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