The study introduces a case of a 51-year-old patient with Caroli's disease of left liver lobe. In 2011 the patient was admitted to Clinic of General, Vascular and Transplantation Surgery. She was after first in her life incident of an acute pancreatitis and subsequent ERCP procedure with left hepatic biliary tract drainage. The lady was qualified to left-hemihepatectomy, which was successfully conducted in our clinic. The only complication of the procedure was surgical site infection which was properly treated with typical antibiotics. One-year observation occurred no other complications and liver function was fine.

Key words: Caroli’s disease, segmental cystic dilatations, hemihepatectomy

Congenital cystoid dilation of intrahepatic biliary ducts was reported for the first time by Caroli and Couinaud in 1958. The literature reports list two types of this disease: simple type and type with periportal fibrosis (congenital hepatic fibrosis) – both disease entities are autosomal recessively inherited, however a gene responsible for these diseases has not been described so far. If widening of biliary ducts is accompanied by the periportal fibrosis and developmental renal disturbances (polycystic kidney, sponge kidney), we deal with Caroli syndrome (1-5).

Frequent complications of both the Caroli disease and the syndrome are recurrent cholangitis, intrahepatic abscesses, cholelithiasis and persistent abdominal pain. Infections of biliary ducts may lead to a generalized infection, becoming in many cases the indirect cause of death. In patients with periportal fibrosis, development of portal hypertension is observed, manifesting by bleeding varicose veins of the oesophagus. Less frequently observed consequences of the disease are amyloidosis and biliary ducts cancer (7, 8).

Widely available ultrasonography and computer tomography with vascular option belong to the basic imaging used in diagnostics of Caroli disease and syndrome. Accurate visualization of biliary ducts is possible due to transcutaneous or retrograde cholangiography and cholangio-MR (9), and hepatic metabolism and possible atrophy of the organ part affected by disease process is evaluated with the use of scintigraphy. Some centres use intraoperative cholangiography, due to which planning of the appropriate range of excision of hepatic parenchyma is possible. Among laboratory tests used in Caroli disease and syndrome diagnostics, routine enzymatic indexes of cholestasis and impairment of hepatic cells should be mentioned (1-4).

In cases of single cysts or formation of cysts, the treatment of choice is a surgical resection
of the hepatic parenchyma affected by disease process and, if needed – with use of choledoodenostomy of Roux-en-Y type, and less frequently choleduodenal junction (6). In the qualification, typical limitations concerning the extent of the operations should be considered with evaluation of the volume of the left hepatic parenchyma. The less effective and the only direct treatment methods of lower efficacy are endoscopic draining procedures with evacuation of deposits or external drainage of „T” type of common biliary duct (10, 11, 12).

In the Clinic of General, Vascular and Transplantation Surgery in 2011, hemihepatectomy was successfully carried out in a female patient with Caroli’s disease limited to the left hepatic lobe. In December 2010, 51-year-old female patient was hospitalized in the Clinic of Gastroenterology and Hepatology of Silesian Medical University in Katowice due to biliary-derived acute pancreatitis (AP). It was the first episode in her life, earlier, since about 10 years ago, epigastric pain was observed periodically, enhanced after meals. In family history – mother died at age of 45 years due to post-operative infective complications after liver resection carried out due to calculosis. Besides, no cases of Caroli disease were observed in the family. Within 24 hours after occurrence of jaundice and biochemical exponents of AP, retrograde endoscopic cholangiopancreatography was carried out, in which cystic widening of intrahepatic biliary ducts was visualized, limited to the left hepatic lobe. Deposits were visible in the cysts. In the right hepatic lobe, the tree image was normal. In not widened common biliary duct, biliary deposits were also seen, not exceeding 6-8 mm in diameter, which were removed after sphincterotomy (SE). Because free flow of contrast from biliary ducts of the left hepatic lobe was not achieved – a prosthesis was inserted to the left hepatic duct with a satisfactory effect (bile flow to duodenum). During the following days, pain disappeared, and biochemical parameters of cholestasis and AP were gradually normalized. Control cholangio-MR examination was carried out on 15th day after endoscopic intervention – showing calculosis of the gall-bladder, numerous widening of intrahepatic biliary ducts of the left hepatic lobe with the presence of deposits in them. The picture of extrahepatic and intrahepatic biliary ducts in the right lobe was normal (fig. 1).

In angio-CT, no anatomical anomalies were found in the arterial and venous vasculature of the liver.

After surgical consultation, the patient was qualified for left-sided hemihepatectomy. The patient was admitted to the Clinic on the 60th day after endoscopic retrograde cholancreatography and SE. At admission, she reported pain in the right space under ribs and middle epigastrium, enhancing after meals. In the physical examination of the abdominal cavity, apart from palpable enlarged, painful left hepatic lobe, other abnormalities were not fund. The results of biochemical tests were within the range of normal. Ultrasonographic examination of the abdominal cavity confirmed widening of biliary ducts of the left hepatic lobe with ductal and vesicular calculosis. Free fluid in peritoneal cavity was not visualized. Elective left-sided hemihepatectomy was carried out (segment 2, 3 and 4) leaving morphologically healthy hepatic parenchyma. On the 8th postoperative day, subfebrile states were observed with an increase of CRP concentration in blood serum up to 163 mg/l without leucocytosis. Purulence of postoperative wound was found – treated locally by typical method, including also wide-spectrum antibiotic therapy with a good result. The patient was discharged from hosp-
tal on 25th day after surgery in a good general condition, without pain, without clinical and laboratory symptoms of infection and cholestasis. The next control examinations (history, physical examination, CT and ultrasonography) – in 14 days after discharge and after 3 and 6-month observation did not show any abnormalities (fig. 2).

In summary, the significant role of imaging diagnostics in form of resonance and computed tomography of the rare disease which is Caroli disease, should be stressed.

It seems that the best current method of treatment is resection of diseased hepatic parenchyma, which, in the long term, offers full recovery.

REFERENCES