

Anatomy variant symptomatic of the duplication of the common bile duct Type IV

Variante anatómica sintomática de la duplicación de la vía biliar común tipo IV

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Abstract

Introduction: Duplicity of the common bile duct is an unusual congenital disorder. **Case report:** A 80-year-old woman with duplication of the common bile duct with retrograde endoscopic cholangiopancreatography (ERCP) who did not resolve the symptoms. **Discussion:** Our case is a variant of Type IV to the classification of duplicity of the common bile duct. The magnetic resonance cholangiography and pre-surgical ERCP allows assessment of the bile ducts, their caliber, and assessment of abnormalities. The treatment before duplicity of the common bile duct will depend on the clinic and the type of opening of the accessory common bile duct. **Conclusions:** It is important to perform a pre-surgical study and during surgery with intra-surgical cholangiography.

Keywords: Duplication. Common bile duct. Accessory common bile.

Resumen

Introducción: La duplicidad del conducto biliar común es una alteración congénita insólita. **Caso clínico:** Mujer de 80 años con duplicación de la vía biliar común con colangiopancreatografías retrógradas endoscópicas (CPRE) que no solventan la clínica. **Discusión:** Nuestro caso es una variante del tipo IV de la clasificación de duplicidad del conducto biliar común. La colangiopancreatografía por resonancia magnética y la CPRE prequirúrgica permiten valorar las vías biliares, su calibre y sus posibles anomalías. El tratamiento dependerá de la clínica y del tipo de apertura del conducto biliar común accesorio. **Conclusiones:** Es importante realizar un estudio prequirúrgico y durante la cirugía con colangiografía intraoperatoria.

Palabras clave: Duplicación. Conducto biliar común. Conducto biliar común accesorio.

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Introduction

Biliary surgery is one of the most complex procedures from the structural point of view, since various anatomical variations can be found that can entail highly important complications¹.

Common bile duct duplication is an exceptional congenital alteration². According to Choi et al. classification³, Type IV has two common bile ducts, with one running into the duodenum and the other into different segments of the upper gastrointestinal tract, such as the stomach, duodenum, or main pancreatic duct (which is known as common accessory bile duct)^{2,4}.

This anatomical anomaly is of great importance, since these patients often have some pathology secondary to this alteration, among which ectopic bile duct lithiasis and choledochal cysts stand out⁵. It can also be associated with pancreaticobiliary junction abnormalities and upper gastrointestinal tract neoplasms².

The pre-surgical diagnostic test par excellence for bile duct anatomical assessment is magnetic resonance cholangiography (MRC)⁶. During surgery, the best test for detecting anatomical variations is intraoperative cholangiography (IOC), which in addition allows assessing the type of surgical option to be performed and possible surgical injuries early detection⁷.

We present an unpublished clinical case: a bile duct duplication with a variant not previously described. It is included in a prospective, longitudinal, descriptive study carried out at our hospital for 3 years. To carry out and present the article, a systematic search was carried out in PubMed and after thorough reading about the subject, the presentation was developed.

Clinical case

Eighty-year-old woman presented with a personal history of hypertension, dyslipidemia, Type 2 diabetes, and depressive syndrome under treatment. The patient was referred to the digestive system department for transaminase elevation (glutamate-oxaloacetate transaminase [GOT] 49 IU/L and glutamate-pyruvate transaminase [GPT] 71 IU/L), as well as of other enzymes (gamma glutamyl transferase [GGT] 707 IU/L and alkaline phosphatase [ALP] 384 IU/L), with clinical signs and symptoms of asthenia, anorexia with months of evolution and unquantified weight loss being associated. In addition, she referred uncertain choluria

and pruritic rashes solved with antihistamine treatment. In view of the situation, an abdominal computed tomography (CT) and comprehensive laboratory testing were requested.

Laboratory results indicated bilirubin 0.8 IU/L, GOT 70 IU/L, GGT 1287 IU/L, and ALP 537 IU/L. CA 19.9 marker was within normal range, and antinuclear antibodies were negative, as well as viral serology.

On CT, a hypodense lesion of approximately 12 mm, with cystic morphology, was observed at segment III. The gallbladder had no morphological alterations and a mass with increased uptake was observed at distal common bile duct of about 20 mm in diameter that caused retrograde dilation of the entire intrahepatic and extrahepatic bile duct. The common bile duct, distal to the described lesion, and the Wirsung duct were slightly dilated. The pancreatic head appeared somewhat enlarged, although no lesions were identified at its parenchyma.

Diagnostic workup was completed with MRC, in which a malformation or normality variant of extrahepatic bile ducts was observed, consisting of a duplication of the common bile duct with anomalous debouchment of the duplicated common bile duct. The right hepatic bile duct continued with the common bile duct, while the left one, although it communicated with the right duct at the bifurcation, continued with the duplicated common bile duct, which ran ectopically into the pyloric antrum posterior and cranial margin.

Different endoscopic retrograde cholangiopancreatography (ERCP) procedures were carried out, in which repletion defects were observed in both normal and ectopic common bile ducts, and in the first portions of their right and left ducts and in the bifurcation of both.

During the different ERCP procedures, a wide biliary sphincterotomy and multiple placements and removals of plastic stents of 10 Fr and 7 cm in length were carried out.

Before the last ERCP, a control abdominal CT scan revealed a distended, smooth-walled gallbladder without signs of cholecystitis, as well as central extrahepatic and intrahepatic bile duct ectasia and extensive left aerobilia related to a well-positioned common bile duct plastic stent. Therefore, during the ERCP procedure, removing the stent and extracting biliary sludge with a Fogarty balloon was decided. Subsequently, home treatment with ursodeoxycholic acid was administered every 8 h.

Five months later, the patient developed biliary symptoms again, for which a new MRC was carried out, which disclosed a common bile duct in normal anatomical position, of up to 10 mm in diameter and with clear repletion defects within, as well as a cystic duct, and the posterior hepatic segments that drained through this pathway, and a second system that from two hepatic bile ducts drained the anterior liver segments, which merged into a 12-mm dilated common accessory bile duct that drained into the stomach at the posterosuperior area of the antrum, 1-2 cm from the pylorus, and that showed an image of 8-mm distal lithiasis. The right common bile duct of the accessory system and the left common bile duct of the normal system were in close contact and appeared to be communicated, with an image of a repletion defect being identified at the fusion zone, which was consistent with lithiasis of 14 mm in diameter and that was mainly lodged in the accessory system, but significantly obstructed the passage to the left posterior branches (main system).

After these findings, the patient was referred to general surgery consultation, where exploration of the bile duct and intraoperative cholangiography was proposed.

The procedure was carried out by subcostal laparotomy, with a bile duct in normal position and an accessory bile duct parallel to the debouchment at the pre-pyloric level being observed. Laparoscopic cholecystectomy and intraoperative cholangiography were carried out through the cystic duct (Fig. 1A), with filling of both ducts with repletion defects within being observed, which suggested lithiasis. Since accessory duct distal opening was not clearly observed, performing choledochotomy of this duct and lavage with Fogarty was decided, with multiple calculi and biliary sludge being extracted; in addition, exploration of the bile duct allowed communication with the gastric antrum to be visualized. A T-tube (Fig. 1B) was placed with 3-0 polyglycolic acid suture. Subsequently, a new cholangiography was performed and filling toward the stomach in distal direction and filling of contrast towards the rest of the biliary tree, including the main bile duct in proximal direction (Fig. 2A and B), were verified.

Postoperative period was uneventful and the patient was discharged at seventh day after admission, with good tolerance to the clamped T-tube (Fig. 2C).

Pathological anatomy reported a gallbladder with a chronic cholecystitis component, without any tumor alteration.

The patient is currently asymptomatic.

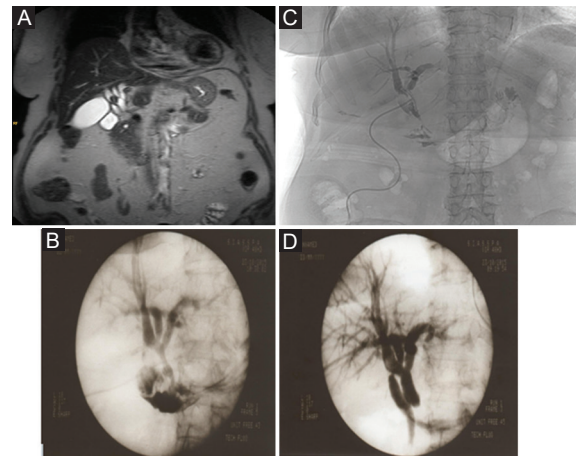


Figure 1. Surgical times. **A:** Cholangiography catheter placement at accessory common bile duct. **B:** T-tube placed at accessory common bile duct fixed with 3-0 polyglycolic acid suture. **C:** Initial cholangiography, in which accessory common bile duct filling defects are identified. **D:** Cholangiography after treatment with Fogarty, prior to Kehr placement, with reperfusion of both ducts being observed.

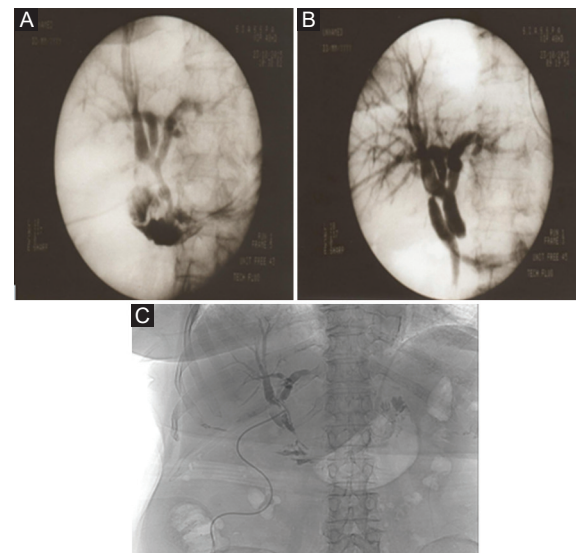


Figure 2. Cholangiography. **A:** Initial, showing accessory common bile duct repletion defects. **B:** After treatment with Fogarty, prior to Kehr placement, reperfusion of both ducts is observed. **C:** After Kehr, during postoperative period, ductal courses without repletion defects are observed.

Discussion

Normal anatomy of the biliary tree is found in 58% of the population, with anomalies occurring in the remaining 42%⁴. The most common abnormalities are observed at intrahepatic bile duct, such as, for example, right posterior bile duct aberrant drainage into

the left hepatic bile duct and simultaneous drainage of the right posterior, right anterior, and left hepatic duct into the common hepatic bile duct^{8,9}.

However, extrahepatic bile ducts duplication is one of the most unusual anatomical variants, with approximately 130 published cases^{3,9}. Its actual incidence is currently unknown, and perhaps, it is much higher than assumed¹⁰.

This anomaly was first described in 1543 by Vesarius^{4,11}. Teilum identified in 1986 a series of 24 cases in the Western world². In 1992, Kanematsu et al.¹² reviewed 56 cases from all over the world. Yamashita et al.¹¹ reviewed the Japanese literature from 1968 to 2002 and collected a series of 47 patients with this anomaly. Gengchen et al. reviewed 24 cases in the Chinese literature⁴.

Embryonic development of the gallbladder, liver and biliary system takes place toward the 3rd week of gestation³, when hepatic diverticulum, which is the result of anterior intestine distal endodermis evolution, begins to grow and its cells penetrate the ventral mesogastrium mesenchyme and divide into a ventral and a dorsal bud. The gallbladder is formed from this ventral bud, and the dorsal bud divides into the right and left lobes. The liver and biliary tree develop inseparably^{2,13}. The bile duct arises from the hepatic primordium stem, and its connection to the ventral bud forms the cystic duct¹⁰. It is important to emphasize that primitive duodenum rotation 90° on its axis makes for the bile duct to rise towards the duodenum^{2,4,9}.

The origin of this anatomical anomaly could be related to random subdivision of the hepatic diverticulum during the first week of embryogenesis^{2,14}. Common bile duct duplicity could be due to hepatic primordium recanalization disturbances¹³.

The accessory common bile duct does not open to the major duodenal papilla; it usually opens into various parts of the upper gastrointestinal tract, such as all portions of the duodenum, including the area just above the major duodenal papilla, into the main pancreatic duct², or be presented by a septum in the common bile duct¹⁰.

There is a classification based on the anatomy of the different types of common bile duct duplicity, developed in 1972 by Goor and Ebert¹⁵, which was modified in 1988 by Saito et al.¹⁶ and later in 2007 by Choi et al.³, according to which there are five types of common bile duct duplication based on morphology (Fig. 3):

- Type I: there is a septum that divides the bile duct.

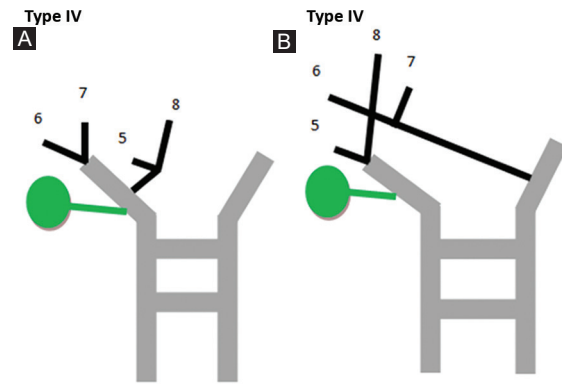


Figure 3. Common bile duct duplication Type IV variation. **A:** Type IV according to Choi et al. classification¹⁵. **B:** Type IV with anatomical variation assessed in our patient.

- Type II: it has a bifurcation of the distal bile duct with independent drainage into the bowel.
- Type III: duplicated extrahepatic bile ducts without (a) or with (b) intrahepatic communicating channels.
- Type IV: duplicated extrahepatic bile ducts.
- Type V: biliary drainage of bile ducts without (a) or with (b) communicating channels^{3,4,9}.

The most common subtypes are III and IV⁹, and the least common are V subtypes, with only one case of each type: Va in Choi et al.³ and Vb in Kim et al.⁹.

According to literature reviews, this anomaly is more common in Asia, especially in Japan⁴; therefore, it could be related to ethnic origin⁴.

The most common complications of accessory common bile duct are usually choledocholithiasis, cholangitis, pancreatitis, gastric, or bile duct cancer, bile duct cysts, and pancreaticobiliary junction anomalies, such as atresia^{2,8,9}.

Gastric cancer is one of the complications that can occur in patients with an accessory common bile duct that drains into the stomach, in type III, as well as gallbladder and ampullary cancers, which occur in patients with opening at second duodenal portion and pancreatic duct^{2,4}.

Choledocholithiasis has an incidence of 33% in these patients, and bile duct cysts, 10%. Approximately one-third of patients with common bile duct duplicity have choledocholithiasis, and 10%, bile duct cysts¹⁷. The association of common bile duct duplicity with abnormal pancreaticobiliary junction is more common in children who present with symptoms of abdominal pain early¹⁸.

Our patient had the accessory common bile duct opening in the stomach, which is why, according to Choi et al.³ classification, our case would be classified as subtype IV. However, the literature does not describe any case of communication between intrahepatic branches (as we found in our patient), although in our case, we could assess a right hepatic duct that continued with a common bile duct regarded as normal, as well as a left hepatic duct that had communication with the right duct and that continued with an accessory common bile duct.

The incidence of this pathology is somewhat higher in women than in men (1.6:1). We can find it at any age (4-80 years), with mean age being 46.1 years¹¹.

The most common symptoms in patients with this anomaly are inflammation of the biliary system, which occurs as pancreatitis, cholangitis or cholecystitis, and in some cases, liver abscesses or malignant tumors appear¹¹.

To diagnose bile ducts anatomical variations before a surgical intervention, MRC and ERCP³ are used, which allow to outline the bile ducts, identify their caliber, and assess for any malformation or abnormality⁶.

IOC in biliary surgery was introduced in 1939 by Pablo Mirizzi^{19,20}. It is an invasive test that allows diagnosing 6-10% of biliary anatomical variants, such as cystic duct opening into right hepatic duct, low junction of both hepatic ducts or right posterior collector low and isolated opening (with an incidence of 20%)¹⁹. Some authors^{21,22} advocate selective use of this test, but others²⁰ prefer its systematic performance to avoid accidental surgical injuries, in addition that it allows lithiasic pathology to be ruled out. In case of ductal duplicity suspicion, IOC use is mandatory, which will prevent bile duct lesions².

Exhaustive pre-surgical workup is highly important, and it is carried out basically with MRC^{4,24}, which will especially indicate the place of right and left hepatic ducts confluence, with a sensitivity of 67% and specificity of 100%²⁰. This way, accidental ductal injuries are prevented³. The accessory common bile duct could be mistaken for a cystic duct and its ligation would cause biliary obstruction⁴.

The treatment that is considered in cases of common bile duct duplicity depends on the clinical signs and symptoms it causes in the patient and the type of accessory common bile duct opening². In patients who do not present with cancer, accessory common bile duct surgical resection is not recommended. In cases without cancer, accessory common bile duct

resection is recommended as surgical treatment.¹¹ Patients with pancreaticobiliary junction abnormalities may require bile and pancreatic duct surgical separation to avoid pancreatic juice reflux⁴. According to Mason and Filipe²⁵, bile reflux alone would not produce tumor pathogenesis, but, in an animal study, pancreatic reflux and the combination of pancreaticobiliary reflux induced the onset of gastric cancer²⁵.

Conclusions

Common bile duct duplication is an extremely rare anatomical variant. It is important to carry out a good pre-surgical workup of the bile ducts with MRC and during surgical intervention with IOC to enable these circumstances to be foreseen and devise the optimal surgical treatment. This will depend on the clinical signs and symptoms the patient presents with and on the presence of tumor pathology.

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Conflicts of interests

There are no conflicts of interest relevant to the development of the article.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained informed consent from the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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