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Review article

Congenital dilations of the biliary tract

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A B S T R A C T

Congenital dilations of the biliary tract are a group of rare conditions, commonly associated with the presence of an abnormality at the junction of the common bile duct and pancreatic duct, which may lead to the reflux of pancreatic juice within the biliary tree. The main clinical symptoms are, abdominal pain, cholangitis and acute pancreatitis, and the most serious complication is malignant degeneration. The treatment of choice is cholecystectomy and complete excision of the bile duct from its bifurcation to its intra-pancreatic segment.

Dilataciones congénitas de la vía biliar

Palabras clave:
Dilatación congénita de la vía biliar
Quiste de colédoco
Anomalía de la unión biliopancreática

R E S U M E N

Las dilataciones congénitas de la vía biliar son un grupo de alteraciones poco frecuentes, comúnmente asociadas con la presencia de una anomalía de la unión del conducto biliar con el conducto pancreático que favorece el reflujo de jugo pancreático en el interior del árbol biliar. Las principales manifestaciones clínicas son el dolor abdominal, la colangitis y la pancreatitis aguda, y la complicación más grave es la degeneración maligna. El tratamiento de elección es la colecistectomía y la exéresis completa de la vía biliar, desde su bifurcación hasta su porción intrapancreática.

Introduction

Congenital dilations of the bile duct are infrequent alterations that affect the intrahepatic and extrahepatic bile ducts. Although this condition can be asymptomatic for a long period of time, it tends to start in infancy. In adults, the dilations generally cause pain and intermittent episodes of cholangitis. Less frequently, it can also cause acute pancreatitis and even biliary cirrhosis. However, the main complication that can occur as the condition evolves, and the most common cause for treatment, is malignant degeneration, which can affect both the bile duct and the gallbladder. The surgical treatment of this condition will depend mostly on the type of dilation and generally consists of complete excision of the extrahepatic bile duct.

Pathogeny

The most widely accepted theory is that this condition is related to anomalies in the biliopancreatic junction. In general,

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the bile duct and the pancreatic duct join in the duodenal wall and form a common duct that opens into the major papilla, and has a length of 4.6±2.6 mm.[1] These ducts have their own choledochal sphincter, a common choledocopancreatic sphincter (sphincter of Oddi), and occasionally, a pancreatic duct sphincter. Under normal conditions, the choledochal sphincter is proximal to the common duct and avoids reflux of pancreatic fluid into the bile duct (Figure 1).

The Japanese study group for anomalies of the biliopancreatic junction defines them as an extraduodenal junction of the bile and pancreatic ducts, that occurs proximal to the choledochal sphincter mechanism that impedes reflux of pancreatic juices.[2] Another two anatomical criteria that have been proposed for defining this phenomenon are a length of the common duct greater than 15 mm and a junction angle greater than 30º. When these anomalies occur in the biliopancreatic junction, the absence of a distal choledochal sphincter provokes chronic reflux of pancreatic juices into the bile duct.[3] The activation of pancreatic enzymes inside the bile duct produces a progressive weakening of the duct wall and sustained hyperpressure would produce dilation. The exact mechanism of pancreatic enzyme activation is unknown, but experimental studies have shown that hyperpressure, along with the mix of bile and pancreatic juices, provokes a premature activation of prophospholipase A_2.[4]

Anomalies of the biliopancreatic junction are rare, with frequencies ranging between 0.08% and 3.2% of the population.[3] In Japan, choledochal cystic dilations are almost always associated with anomalies of the biliopancreatic junction, whereas in the West, this correlation only occurs in half of cases.[5-11] Kimura defined two varieties of the anomaly found in the biliopancreatic junction. In variety 1, the pancreatic duct appears to empty into the common bile duct at an acute angle, and the distal common bile duct is not narrowed. In variety 2 (95%), the common bile duct appears to empty into the pancreatic duct and the distal common duct tends to be narrowed, producing severe and early dilations in the bile duct (Figure 2).[5]

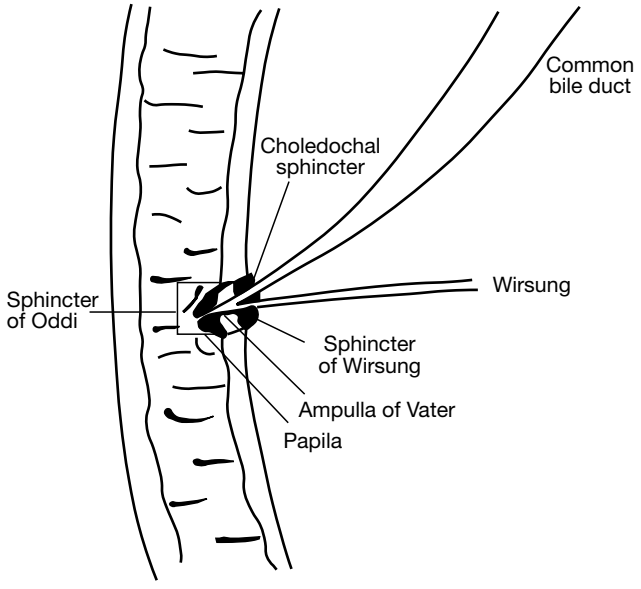

Figure 1 – Normal anatomy of the biliopancreatic junction.

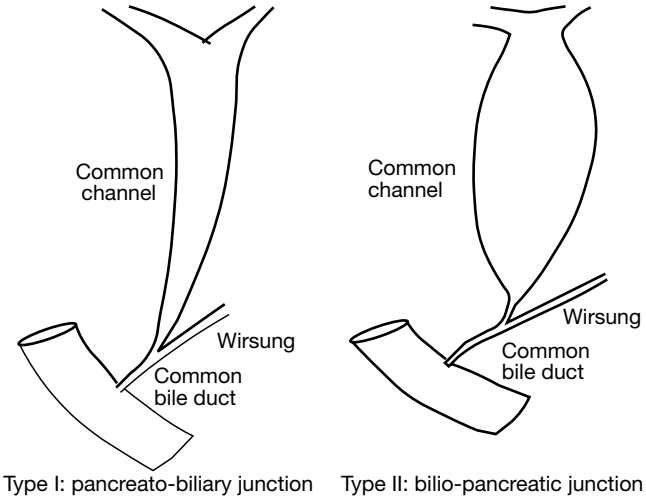

Figure 2 – Kimura classification of anomalies of the biliopancreatic junction.

Although it is widely accepted that congenital dilations of the bile duct are associated with anomalies of the pancreaticobiliary junction, these can occur independently. Indeed, anomalies of the pancreaticobiliary junction without dilation of the bile duct are relatively common, and these cases are frequently complicated with carcinomas of the gallbladder.[12]

Although extremely rare, familial cases have been described, although no chromosomal markers have been discovered, and these cases probably have a multifactorial etiology.[13]

Classification

The most commonly used classification system is the Todani,[14] that distinguishes 5 types (Figure 3). Type I is a dilation of the extrahepatic bile duct. This is further subdivided into subtype Ia, if the dilation is sacciform and affects all or almost all of the hepatic common bile duct; subtype Ib, if the dilation is sacciform but affects only part of the hepatic common bile duct; and Ic, if the dilation is fusiform along the entire duct. Type II is a lateral sacciform dilation of the bile duct, with a short and narrow pedicle. Type III consists of an isolated dilation in the terminal end of the common bile duct in the ampulla of Vater. Type IV is defined as the simultaneous presence of various dilations. This is subdivided into 2 subtypes: IVa is the association of dilations in the intrahepatic and extrahepatic bile duct, and IVb is dilations of the main bile duct and the terminal end of the common bile duct. Type V consists of dilations of the intrahepatic bile duct and corresponds to Caroli's disease.

Epidemiology

In the West, dilations are relatively infrequent, but in Japan and Southeast Asia, the incidence is one in every 1000-1750

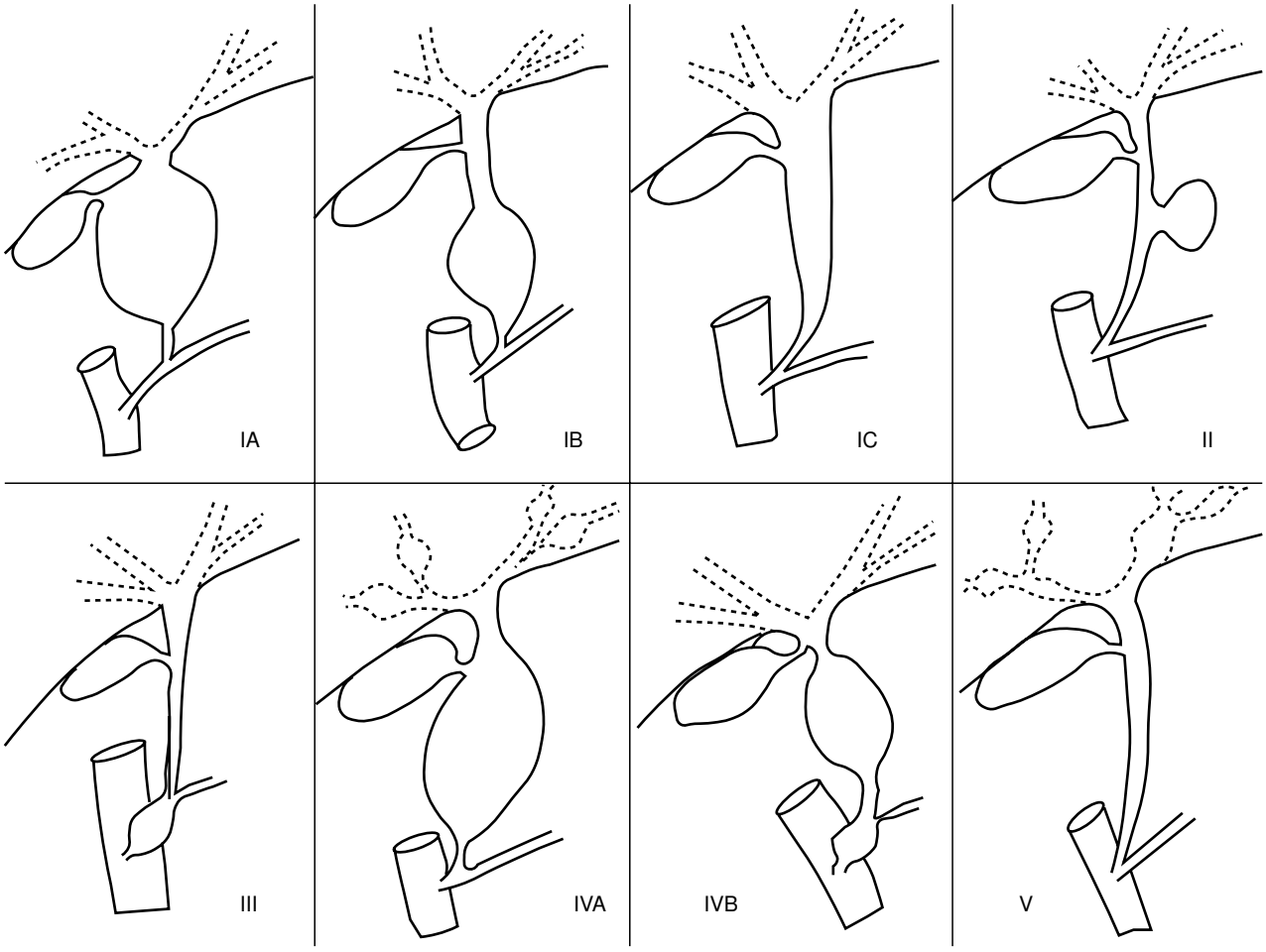

Figure 3 – Todani classification.

births, about 100 times greater than in Western countries. This condition is 4 times more frequent in women than in men, and although it can be diagnosed at any age, two thirds of cases are discovered before the age of 10 years.[3,5-11,14-17] Types I and IV are the most frequent, making up 80% of all cases; none of the other types make up even 5% of the total.[3]

Symptoms

Although patients can remain asymptomatic for long periods, symptoms tend to appear before the age of 16. With respect to the clinical profile of the disease, two different groups can be considered: children younger than one year of age, and those older than one.[18-21] In the first group, jaundice, diminished bile secretion, and a mass in the upper right quadrant of the abdomen are frequent. After one year of age, the most common clinical symptoms are intermittent biliary obstruction with episodes of jaundice and abdominal pain.[8,18] In adults, pain is produced in 60%-90% of cases, whereas jaundice and fever are present in about half. The triad made up of pain, jaundice, and a palpable mass is found in less than 15% of cases. Between 20% and 70% of patients have some type of biliary complication, which in half of cases results in an episode of cholangitis. Close to 30% of patients have episodes of acute pancreatitis, and 15% result in cirrhosis and portal hypertension. Less than 2% of patients have biliary peritonitis secondary to perforation of the cyst.[22] Patients with Kimura variety I, with less severe dilations, tend to be asymptomatic and the diagnosis can be incidental. In contrast, patients with variety 2 tend to have symptoms in childhood.

Imaging tests

Imaging tests have three objectives: to confirm the diagnosis and association of the condition with an anomaly of the biliopancreatic junction, to verify the extent and type of dilation, and to rule out the presence of any possible complications, such as lithiasis and cancer. Ultrasound and CT scans reveal cystic tumours, independent of the gallbladder, that are in the subhepatic space, between the portal confluence and the duodenum.[10,11] The CT scan also provides information on the relationship of the cystic dilation with neighbouring structures, above all with elements of the hepatoduodenal ligament. Even so, the

extent of the dilation tends to be underestimated, and this technique does not offer information on the biliopancreatic junction. For this reason, it is necessary to use methods that provide visualisation of the bile duct. Endoscopic retrograde cholangiopancreatography was traditionally used, but now cholangiography is the technique of choice, with a similar level of sensitivity (90%-100%) and specificity (73%-100%), but without the complications.[23-28] Endoscopic ultrasound may be indicated when an obstruction of the distal common bile duct is suspected, and in order to verify the nature of the biliopancreatic junction.[3] Occasionally, the anatomy of the biliopancreatic junction can only be visualised using invasive methods: endoscopic retrograde cholangiography and percutaneous transhepatic cholangiography.[29]

During pregnancy, ultrasound can be used to diagnose some congenital dilations of the common bile duct and allows for planning of early treatment during the neonatal period.[30]

Moderate dilation of the bile duct

If the bile duct has a diameter less than 20 mm, an anomaly is probably present in the biliopancreatic junction. If unconfirmed, the amylase content of the bile fluid (measured in the gallbladder or common bile duct) must be determined; if this value is less than 10 000 Ul/l, a cholecystectomy is indicated, but if higher, the diagnosis of pancreaticobiliary reflux is confirmed, making the diagnosis of cystic dilation of the common bile duct very probable, and therefore, a complete excision of the bile duct is justified.[3]

Developmental complications

Up to 80% of patients have some type of complication, which can be classified into three groups: mechanical, infectious, and malignant degeneration.

1. Mechanical.
   - Lithiasis. Intracystic lithiasis is the most frequent type, with an incidence of 2%-70%. This tends to be associated with biliary stasis. Intrahepatic calculi occur most frequently in patients with dilations of the intrahepatic bile duct.[31,32]
   - Acute pancreatitis. This condition is especially frequent in adults. Its true incidence is difficult to determine, but it varies between 2% and 70%.[19] It tends to be associated with anomalies of the biliopancreatic junction and cystolithiasis.
   - Ruptured cyst. This condition is rare and is most frequent in adults during pregnancy.[22,33]
   - Biliary cirrhosis. This condition is secondary to chronic biliary obstruction. It can cause portal hypertension, which can also be secondary to thrombosis of the portal vein and congenital hepatic fibrosis associated with Caroli's disease. Cirrhosis can affect up to 15% of adults and is especially frequent in patients that have undergone multiple operations.[19]
2. Infectious.
   Recurrent episodes of cholangitis are relatively frequent. More rare conditions are intrahepatic abscesses secondary to cholangitis or segmental bile duct obstruction by calculi.
3. Malignant Degeneration. In the absence of a sphincter in the distal common bile duct, pancreatic enzymes reflux chronically into the bile duct, inducing the conjugation of bile acids and the transformation of lecithin into lysolecithin. These products are toxic for the biliary mucosa, especially with biliary stasis, and so the malignant degeneration can be produced both in the walls of the cyst and the gallbladder in the absence of biliary dilation.[6,9,34-37] The risk of biliary cancer is 20 times higher than in the rest of the population,[37] and the incidence of this condition ranges between 3% and 40% (mean: 16%).[21,38] The rate is 0.7% during the first ten yeas of life, 14% in patients older than 20 years of age, and approximately 50% in patients older than 50 years.[3,39,40] Patients that undergo cystodigestive bypass have higher incidences of cancer, which appears a mean 10 years earlier than in patients without the bypass.[3,41] Cancer can appear in any kind of dilation, but is most frequent in types I and IV. It is located in the extrahepatic bile duct in 50%-85% of patients, especially in the dilation in the gallbladder in 8%-45% of cases; and in the intrahepatic bile duct or intrapancreatic common bile duct in less than 5% of cases.[3] Patients with anomalies of the biliopancreatic junction that do not have dilation of the bile duct are also at greater risk for biliary cancer, but this is normally located in the gallbladder with an incidence of up to 25%-36%.[12,34,35,37,42-47] Around 80% of tumours are cholangiocarcinomas, but any histological types can occur.[6,38,40,41]

Treatment

The treatment of choice is a complete excision of the bile duct, which definitively suppresses pancreatic reflux and completely removes the wall of the cyst and areas susceptible to malignant degeneration. Biliary transit reconstruction is achieved by performing a Roux-en-Y hepaticojejunostomy.[48-51] The excision is complete when the extrahepatic bile duct is completely resected, including the intrapancreatic common bile duct. In adults, previous surgical procedures, recurrent episodes of cholangitis, and portal hypertension can severely hinder the dissection. In these patients, a complete excision of the extrahepatic bile duct can be made impossible, and at times, an intrawall dissection of the posterior wall of the cyst may be necessary, always eliminating the mucosa layer. The complete resection of the extrahepatic bile duct, along with cholecystectomy, considerably reduces the risk of malignant degeneration, which becomes 0.7%.[38] Recently, these lesions have been successfully treated using a laparoscopic approach.[51-55] If cholangitis is present, wide-spectrum antibiotics must be used for treatment, and endoscopic or percutaneous drainage of the infected bile may also be necessary.

If there are any anomalies in the biliopancreatic junction, cholecystectomy is recommended due to the elevated risk of malignant degeneration of the gallbladder.[41,42,53,56-60]

Treatment according to the type of dilation

- Type I. Cholecystectomy and complete excision of the extrahepatic bile duct, with a Roux-en-Y hepaticojejunostomy.
- Type II. Excision of the cyst and cholecystectomy; complete excision of the bile duct is not necessary. Depending on the size of the neck of the cyst primary closure or a Kehr tube can be used.
- Type III. Cases smaller than 3 cm can be effectively treated by endoscopic sphincterotomy. When greater than 3 cm, a transduodenal excision is needed, and occasionally, reimplantation of the pancreatic duct in the duodenal wall.
- Type IVa. Often the only treatment available is complete excision of the extrahepatic bile duct and reconstruction through a hepaticojejunostomy. If the intrahepatic lesions are limited to one single lobe, a partial hepatectomy is possible. Occasionally, some type of unblocking of the intrahepatic bile duct may be needed. Patients with these types of lesions may require transplantation.
- Type IVb. Complete excision of the extrahepatic bile duct along with a transduodenal excision of the common bile duct or endoscopic sphincterotomy, depending on size.
- Type V. If the disease is limited to one lobe of the liver, partial hepatectomy is recommended, with or without a cholangiojejunostomy. In patients with bilobular disease, a liver transplant may be recommended.

Results of treatment

After a complete excision, the prognosis is excellent, but follow-up must continue for the entirety of the patient's life, as there is still a slight risk for biliary cancer.

Mortality from this type of surgery is practically nil, and morbidity tends to be less than 10%.[3] Immediate postoperative complications include haemorrhage, pancreatitis, pancreatic, biliary, or duodenal fistula, and infections. In the long term, cholangitis occurs in 2%-10% of cases, and between 0%-17% of patients require re-operations, mainly due to stenosis of the biliodigestive anastomosis and lithiasis.[11,21,61-65] The incidence of long-term complications is higher in patients with type IVa than type I dilations, mainly due to cholangitis, particularly when associated with intrahepatic lithiasis.[63,66] Any case of cholangitis following surgery requires ruling out the presence of intrahepatic lithiasis, stenosis of the bilioenteric anastomosis, malignant degeneration of the unresected bile duct, and intrahepatic biliary stenosis that may have gone unnoticed. Intrahepatic lithiasis appears in approximately 10% of patients, and is almost always correlated with areas of biliary stenosis proximal to the bifurcation.[62,66,67]

Conflict of interest

The authors affirm that they have no conflicts of interest.

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