

Diseases of the bile duct

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13 March 2024

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1 Anatomy

Bile is a digestive fluid produced and secreted by the liver, and it is transported by a series of branching bile ducts known collectively as the biliary tree. At the end of the biliary tree (extrahepatic ducts), the interlobular ducts merge to form the two main bile ducts of the liver: the right hepatic duct and the left hepatic duct. Extrahepatically, these ducts coalesce to form the common hepatic duct until coming into contact with the cystic duct, the bile duct, which connects to the gallbladder. The common hepatic duct and cystic duct merge to form the common bile duct. Bile can travel into the gallbladder for storage via the cystic duct. Also, bile travels to the duodenum to be released through it.

The primary function of the biliary tract is to transport and release the bile. The gallbladder stores and concentrates bile. Bile assists with the absorption and digestion of fats and fat-soluble vitamins in addition to the removal of lipid-soluble waste products. The main components of bile are cholesterol, bile salts, and bilirubin. Bilirubin, responsible for the signature yellow-green color of bile, is also excreted in bile and is responsible for the pigmentation of feces (i.e., stools).

2 Main symptom of biliary disease: Jaundice

Jaundice, also known as hyperbilirubinemia, is defined as a yellow discoloration of the body tissue resulting from the accumulation of excess bilirubin. Deposition of bilirubin

happens only when there is an excess of bilirubin, and this indicates increased production or impaired excretion. Bilirubin has two components: unconjugated (or indirect) and conjugated (or direct), and hence elevation of any of these can result in jaundice.

The pathophysiology of jaundice is best explained by the metabolism of bilirubin.

Production Bilirubin is released by senescent or defective red blood cells. A red blood cell dies (usually in the spleen) and releases bilirubin.

Hepatic uptake Bilirubin in its unconjugated form gets transported to the liver.

Conjugation of bilirubin This unconjugated bilirubin undergoes conjugation to glucuronic acid resulting in the formation of conjugated bilirubin, which is soluble in the bile. That is, bilirubin binds to glucuronic acid.

Bile secretion from liver Conjugated bilirubin is now released into the bile duct, stored in the gallbladder, reaching the small bowel and finally enters the colon.

Dysfunction in the **first two phases** results in elevated serum levels of unconjugated bilirubin. Dysfunction in the **last two phases** results in elevated conjugated bilirubin.

Unconjugated hyperbilirubinemia is caused by increased production of bilirubin, such as increased destruction of erythrocytes (red blood cells) or altered hepatic uptake, such as drugs or diseases that interfere with the mechanism of uptake of bilirubin.

Conjugated hyperbilirubinemia is also called cholestasis. It is caused by a marked reduction in bile secretion and flow. Cholestasis can be due to a functional impairment of the hepatocytes in the secretion of bile and/or due to an obstruction at any level of the excretory pathway of bile. Cholestatic jaundice can be classified into intrahepatic or extrahepatic cholestasis, depending upon the level of obstruction to bile flow.

3 Clinical Significance

Biliary obstruction refers to the impairment of bile flow from the liver to the small intestine due to blockage of the biliary duct system. Bile obstruction affects a large portion of the population around the world, with significant morbidity and mortality. The most common etiology of biliary obstruction is gallstones causing common bile duct obstruction, which manifests as pain, nausea/vomiting, and jaundice. If infection is present, the patient will present with fever.

The biliary tract provides an outlet to transport bile into the gastrointestinal tract, which facilitates the absorption of fat-soluble nutrients. In general, obstruction of the biliary tract can lead to steatorrhea, and fat-soluble vitamin deficiency (A, D, E, and K). Proximal to the site of obstruction also harbors an opportunity for bacterial infection, as in the case of ascending cholangitis.

Obstruction of bile duct causes jaundice and acholic (pale) stools. This is called cholestasis, and the liver function test often confers to cholestasis pattern (bilirubin, alkaline phosphatase, GGT, AST and ALT increases in serum).

Cholelithiasis, commonly known as gallstones, is a common disease. Most of people with gallstones are asymptomatic, but biliary pathology is often associated with gallstones. For example, acute cholecystitis is most commonly caused by gallstone obstruction of the cystic duct. This obstruction leads to inflammation of the gallbladder. Classically, the patient will present with right upper quadrant abdominal pain, fever, nausea, vomiting, and an elevated white blood cell count.

4 Clinical Vocabulary

- Cholelithiasis means gallstones present within the gallbladder, which can cause...
- Biliary colic is the intermittent, dull pain due to transient blockage of bile flow, which can cause...
- Choledocholithiasis means gallstones situate within the common bile duct, which can cause...
- Gallstone pancreatitis is pancreatic inflammation from gallstone blockage of the pancreatic duct
- Cholecystitis is inflammation of the gallbladder due to prolonged obstruction of the cystic duct
- Cholecystectomy is the surgical removal of the gallbladder
- Cholangitis is an infection of the bile ducts
- Cholangiocarcinoma is an aggressive cancer of the bile ducts

Bile flow is obstructed by stones within the common bile duct, which leads to obstructive jaundice and possibly hepatitis and/or pancreatitis. The stagnant bile can also lead to the presence of bacteria in the bile fluid, causing ascending cholangitis. Cholangitis and sepsis are more common in patients with choledocholithiasis than other sources of bile duct obstruction because a bacterial biofilm typically covers common bile duct stones. The pancreatic duct joins the common bile duct near the duodenum, and therefore, the pancreas may also become inflamed by the obstruction of pancreatic enzymes. This is termed gallstone pancreatitis.

4.1 Cholelithiasis

Cholelithiasis or gallstones are hardened deposits of bile that can form in the gallbladder. The gallbladder is a small organ located just beneath the liver. Asymptomatic gallbladder stones found in a normal gallbladder and normal biliary tree do not need treatment unless they develop symptoms. These gallstones may go on further to develop complications such as cholecystitis, cholangitis, choledocholithiasis, or gallstone pancreatitis. Stones are composed of a mix of bilirubin, bile lipids, and cholesterol. Cholelithiasis is more common in female patients, pregnant patients, older patients, those with high serum lipid levels, and in obese patients

4.2 Biliary colic

Biliary colic is defined as pain in the abdomen, due to obstruction usually by stones in the cystic duct or common bile duct of the biliary tree. It typically occurs after eating a large, fatty meal that causes contraction of the gallbladder, pushing the stones out of it towards the bile duct. Treatment of this disease is primarily surgical, involving removal of the gallbladder.

4.3 Choledocholithiasis

Choledocholithiasis is the presence of stones within the common bile duct. The treatment of bile duct stones is endoscopic retrograde cholangiopancreatography (ERCP) or in some cases a laparoscopic cholecystectomy with bile duct exploration.

4.4 Acute cholecystitis

Acute cholecystitis is inflammation of the gallbladder that occurs due to occlusion of the cystic duct or impaired emptying of the gallbladder. The pathophysiologic mechanism of acute cholecystitis is blockage of the cystic duct, which causes inflammation. Cholecystitis is a condition best treated with surgery; however, it can be treated conservatively if necessary (antibiotics). This condition can be associated with or without the presence of gallstones. Regardless of the cause of the blockage, the gallbladder wall edema will eventually cause wall ischemia and become gangrenous. The gangrenous gallbladder can become infected by gas-forming organisms, causing acute emphysematous cholecystitis; all of these conditions can quickly become life-threatening, and rupture has a high rate of mortality.

4.5 Acute cholangitis

Acute cholangitis, also known as ascending cholangitis, is a life-threatening condition that is caused by an ascending bacterial infection of the biliary tree. Delay in diagnosis and treatment can lead to septic shock. Choledocholithiasis is the most common cause, with infection-causing stones in the common bile duct leading to partial or complete obstruction of the biliary system.

5 Diagnosis

The diagnosis of **biliary obstruction** is made by clinical presentation, abnormal laboratory results, and imaging studies implying biliary obstruction, and sometimes, infection. Cases of biliary colic present with progressing right upper quadrant abdominal pain with bloating, and food intolerances (nausea and vomiting). If there is bacterial infection (either cholecystitis or cholangitis), the patient may present with fever.

White blood cell count may be elevated. Liver enzymes may also be elevated. If there is a high bilirubin level, then it should be considered a possible common bile duct stone. Even in the presence of severe gallbladder disease, laboratory values may be normal. Amylase and lipase must also be checked to rule out pancreatitis. Often a CT scan is ordered in the emergency department as the first imaging test in the workup. Findings of cholecystitis and gallstones can often be seen in this imaging. A **gallbladder ultrasound** is the best test (actually, it is the **diagnostic test of choice**) to evaluate gallbladder disease initially. A thickened gallbladder wall and gallstones are common findings with this condition.

6 Treatment and Management

The first approach in case of acute infection is antibiotic therapy. The most appropriate management of biliary obstruction is laparoscopic cholecystectomy. In severely ill patients with sepsis, treatment is immediate or emergent biliary drainage (temporizing percutaneous drainage of the gallbladder). Those who are clinically improving after medical therapy may be candidates for decompression. Biliary decompression or drainage can be achieved by ERCP. RCP is the gold standard and treatment of choice for biliary decompression, as it is effective in 94 to 98% of cases.