Clinical Aspects of Liver Diseases

32 Cholangitis and cholangiodysplasia

32 Cholangitis

1 Definition

The term cholangitis subsumes localized or diffuse inflammatory changes of diverse aetiology, i.e. between the canal of Hering and the ampulla of Vater, affecting the intrahepatic and extrahepatic bile ducts. Cholangitis can be acute or chronic; it may originate as a primary disease in the bile ducts or develop as secondary concomitant cholangitis in the course of another underlying disease. Forms of cholangitis which exclusively affect the intrahepatic bile ducts generally give rise to the clinical picture of liver disease.

2 Systematics and aetiology

► *The bile is sterile under physiological conditions.* • Pathophysiological events can cause **asymptomatic bacteriocholia,** which is of no clinical importance. Microorganisms are verifiable in bile in 75–100% of patients with obstruction of the large bile ducts – whereas this applies only to 0–10% of patients with obstruction due to pancreatic carcinoma. Diagnostic or therapeutic endoscopic interventions in the bile ducts are often followed by **bacterial cholangitis**, attributable to the importation of microorganisms, particularly as a result of a (usually temporary) hindrance of bile flow. • *Cholangitis caused by infection is not a separate entity.*

Initially, **ascending cholangitis** has to be considered on account of its pathogenetic development. This condition originates in the gall bladder, duodenum or pancreas. Moreover, the bile ducts are liable to infection by bacteria or parasites as a consequence of cholestasis and/or achylia. • **Descending cholangitis** is considered to be less frequent, with the infection descending from a chronically infected gall bladder or from a primary infection of the liver, for example in the case of salmonellosis. • An infection of the bile ducts may cause **pyogenic cholangitis,** which can take an acute, relapsing or chronic course, the latter mainly being caused by a hindrance of bile flow. • Depending on the time taken for an obstruction to develop, **obstructive cholangitis** manifests as either acute or chronic disease. In the case of obstruction, the increase in intraductal pressure (>15–20 cm H_2O) causes a cholangiovenous or cholangiolymphatic *reflux* of bacteria or endotoxins into the blood circulation. As a result, signs of systemic and, in severe cases, septic disease appear. • **Toxic cholangitis** may be triggered by chemicals, medicaments or toxins. • Furthermore, there is also the clinical picture of **immunological cholangitis.** This form includes (*1.*) *primary biliary cholangitis*, (*2.*) *primary sclerosing cholangitis*, (*3.*) *autoimmune cholangitis*, and (*4.*) *overlap syndromes.* (s. tab. 32.1)

1. **Aetiology**	
• **Infections**	
– bacteria	– parasites
– mycoses	– viruses
• **Obstruction**	
– benign stenoses (stenosis of the papilla of Vater, Mirizzi's syndrome, postoperative strictures, chronic pancreatitis, juxtapapillary diverticula, *etc.*)	
– blood clots	
– mycoses	
– gallstones	
– malignant stenoses (histiocytosis X, Hodgkin's disease, CCC, *etc.*)	
– oriental cholangitis	
– parasites	
– suture material, clips, sponges	
– highly viscous mucus (e.g. mucoviscidosis)	
• **Immunological causes**	
– primary biliary cholangitis	
– primary sclerosing cholangitis	
– autoimmune cholangitis	
– graft-versus-host disease	
– rejection reaction	
– sarcoidosis	
– pharmacons	
• **Toxic causes**	
– burn injury	
– cytostatics	
– pharmacons	
• **Caroli's disease**	
2. **Clinical forms**	
• acute	• non-suppurative
• chronic	• suppurative
• relapsing	
► asymptomatic	
► symptomatic	
3. **Pathogenesis**	
• **primary development**	
– genetic/congenital	
– immunological	
– toxic	
• **secondary development**	
– ascending	– periductular lymphogenic
– descending	– septicaemic via hepatic artery

Tab. 32.1: Classification, causes and pathogenesis of cholangitis