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# PRIMARY SCLEROSING CHOLANGITIS, ETIOLOGY, CLINICAL PICTURE AND TREATMENT.

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**Annotation:** This article is devoted to the etiology, clinical picture, and treatment of primary sclerosing cholangitis.

**Key words:** benign, perinuclear antineutrophil antibodies, alkaline phosphatase level, suprastenotic dilatation, ursodeoxycholic acid.

Primary sclerosing cholangitis (Delbe's disease) is a disease with a not fully understood etiopathogenesis, characterized by intense inflammatory fibrosis of the extrahepatic bile ducts involving (or not) the intrahepatic ducts, leading to a narrowing of their lumen. This is a benign, slow-onset disease leading to cholestasis, biliary cirrhosis, portal hypertension and, ultimately, death from liver failure. It occurs more often in men aged 40–43 years, in women less often and at a younger age.

Although the cause of the disease is unknown, primary sclerosing cholangitis is often associated with inflammatory bowel disease (about 70% of patients). 5% of patients with ulcerative colitis and 1% of patients with Crohn's disease develop primary sclerosing cholangitis. This association, as well as the presence of certain antibodies (for example, antinuclear antibodies and perinuclear antineutrophil antibodies, suggests an autoimmune mechanism for the development of the disease. It appears that T cells are involved in the process of damage to the bile ducts, indicating altered cellular immunity. Heredity also plays a role.

Diagnosis based on clinical manifestations - skin itching, jaundice, asthenic syndrome.

•Diagnostic criteria: increased alkaline phosphatase, changes in the biliary tract during cholangiography in the form of a rosary, biopsy data, exclusion of secondary sclerous cholangitis.

•Morphology: simultaneously detected - focal necrosis, interlobular inflammatory changes, pronounced proliferation of the bile ducts. Fibrosis of the portal tracts. Intralobular cholestasis. Damage to the interlobular bile ducts up to obstruction of the ducts.

•Determination of antimitochondrial antibodies sensitivity is close to 100%. Usually combined with antinuclear factor in 70-80% of patients.

Clinical picture: cholangitis with fever and chills is one of the early manifestations, but the main symptom of the disease is obstructive jaundice, which has a persistent progressive intermittent nature with the duration of jaundice episodes from 1 month to 1 year; the appearance of jaundice is sometimes preceded by persistent skin itching. The frequency of symptoms, according to various authors, is as follows: jaundice ~50–60%, itching ~30%, pain in the right hypochondrium ~20–25%, fever ~15%, without clinical symptoms ~30%.

A constant laboratory sign is a significant increase in the level of serum alkaline phosphatase compared to relatively low bilirubin levels (the direct fraction predominates); a significant increase in the content of aromatic and sulfur-containing amino acids; A diagnostic and prognostic sign is an increased copper content in the liver tissue and its excretion in the urine.

Among additional research methods for primary sclerosing cholangitis, the most informative are radiocontrast research methods (variants of direct cholangiography - ERCP,

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percutaneous transhepatic cholangiography). Cholangiograms clearly show limited or extended strictures of the bile ducts with dilations in the form of "rosary beads"; the absence of suprastenotic dilation is characteristic.

Laparoscopy reveals a moderate enlargement of the liver with a smooth, greenish surface and pronounced fibrosis in the form of a "spider web." Liver biopsy - proliferation of connective tissue around the intrahepatic bile ducts, proliferation of ductules, infiltration of portal tracts with lymphocytes, plasma cells, neutrophils, signs of cholestasis.

Treatment A distinction is made between intrahepatic (therapeutic) and extrahepatic (surgical) cholestasis. Corticosteroids, immunosuppressants, antihistamines, also D-penicillamine ℘ and colchicine are used.

Ursodeoxycholic acid preparations are used (expected to have a cytoprotective effect when its molecule is embedded in cell membranes, an immunosuppressive effect - a decrease in the expression of HLA genes and the prevention of autoimmune reactions; a decrease in alkaline phosphatase activity). In case of bacterial cholangitis, broad-spectrum bactericidal antibiotics are used in short courses. Surgical treatment is indicated in cases of significant strictures of the extrahepatic bile ducts.

The full range of minimally invasive and open interventions draining the ductal system is used. Internal drainage (biliodigestive anastomoses) is considered the most physiological, but it is not always technically possible to perform them. External drainage of the common bile duct is widespread, but the negative aspect of such an operation is the long-term leaving of drainage (several months or years), and this is a real threat of secondary infection.

In case of local forms of damage, resection of the ducts with biliodigestive shunting, their dilatation, and nasobiliary drainage are possible. In recent years, orthotopic liver transplantation has been increasingly used. The prognosis is doubtful. In case of disease progression, death occurs within a period of six months to 17 years (on average 3–6 years) from liver failure or bleeding from varicose veins of the esophagus and stomach (a manifestation of progressive secondary biliary cirrhosis of the liver).

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