Diagnosis and Management of Primary Sclerosing Cholangitis

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Algorithmic Approach

A. The first step in the evaluation of a patient with primary sclerosing cholangitis (PSC) is a thorough history and physical. Patients may experience varying degrees of symptoms, often long-standing, from 12 to 24 months that can include fatigue, jaundice with or without pruritus, choluria, steatorrhea, and acute cholangitis.

B. Evaluation of these symptoms includes routine laboratory studies including a liver function panel. Marked elevation in bilirubin, gamma-glutamyl transferase (GGT), and/or aspartate aminotransferase (AST)/liver function tests (LFTs) will generally prompt an ultrasound of the right upper quadrant that may or may not show dilation of intrahepatic or extrahepatic bile ducts. If PSC is suspected, autoantibodies such as antinuclear, antismooth muscle, anticardiolipin, and IgG4 may be obtained, although they are of uncertain significance. Intrahepatic saccular dilations and signs of portal hypertension may be visualized in long-standing disease or end-stage PSC [1].

C. If no abnormalities are observed on ultrasound, further investigation of the hyperbilirubinemia is warranted, as ultrasound is not the most sensitive diagnostic modality for intrahepatic biliary pathology and is very operator dependent. MRCP or ERCP should be performed at this point; however, ERCP will yield better results from both diagnostic and therapeutic perspectives [2].

D. Liver biopsy is not routinely performed in the diagnosis of PSC. It is indicated when visualization of the biliary ducts by MRCP has not yielded any abnormal findings and other causes of cholestatic liver disease need to be investigated. Liver biopsy tends to show an "onion-skin" appearance of concentric periductal fibrosis in PSC. As the disease progresses, the periductal fibrosis advances to necrosis, periportal fibrosis, and eventually biliary cirrhosis [3].

E. ERCP is the preferred route for cholangiography and can demonstrate the characteristic appearance of PSC: multifocal, diffusely distributed dilations, and strictures of the intrahepatic and extrahepatic biliary system. The classic pattern is described as "beading," resembling the arrangement of beads on a string. Cholangioscopy is currently being investigated and may play a role in directed tissue biopsy; however, it is not considered to be a standard of care in PSC [4].

F. Medical treatment of PSC has shown little to no lasting effects. Dominant intrahepatic strictures are best managed endoscopically

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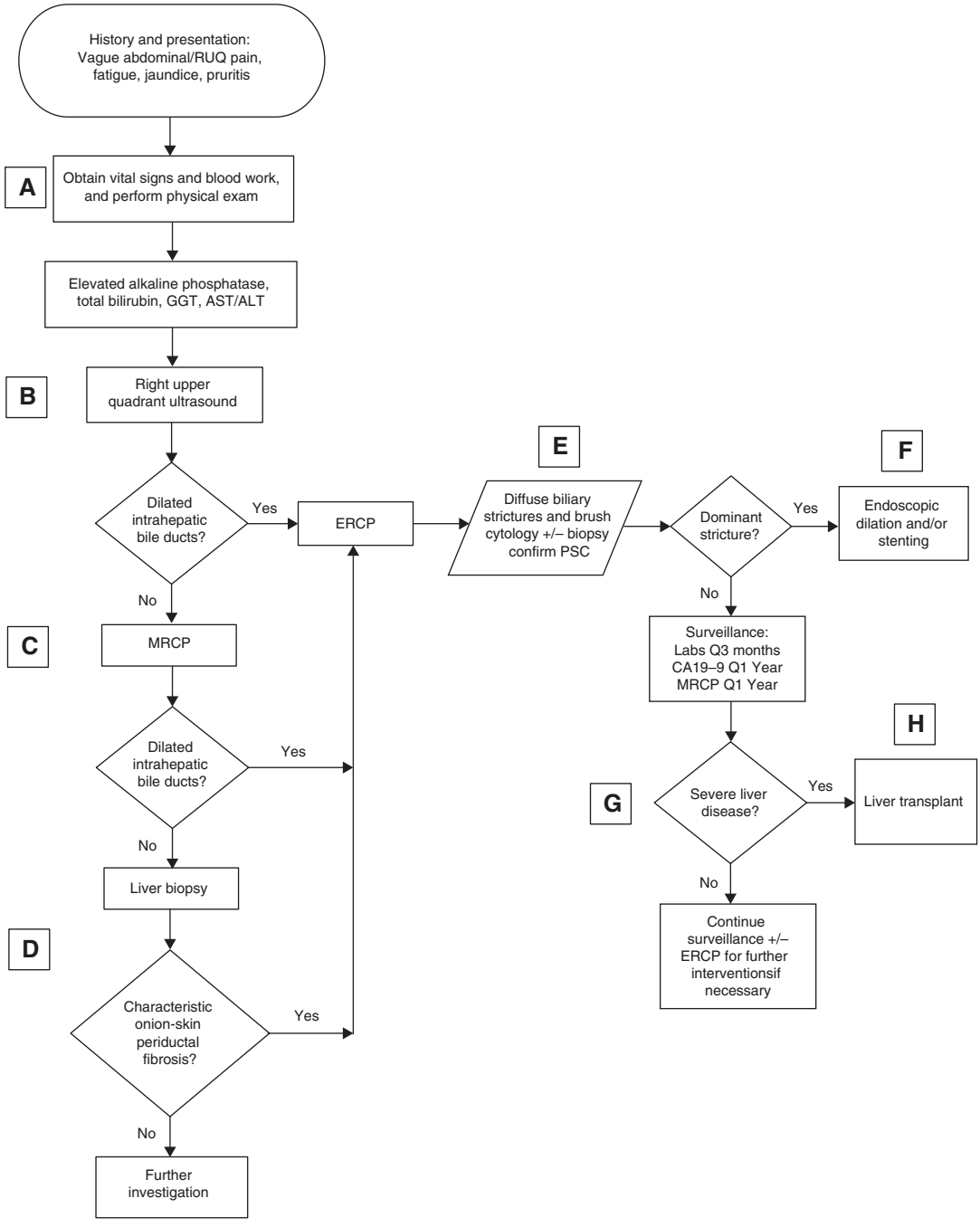
with balloon dilation and/or stenting. Biliary reconstruction is an option for a select group of patients with dominant extrahepatic strictures only and minimal intrahepatic disease. With the high risk of developing cholangiocarcinoma in PSC, as well as increased success of orthotopic liver transplantation (OLTx), the use of biliary reconstruction procedures has decreased.

G. If no dominant strictures are identified during ERCP and no endoscopic interventions performed, the patient must continue to be surveilled. Both the American Association for the Study of Liver Disease (AASLD) and the European Association for the Study of the Liver (EASL) recommend having quarterly blood work evaluation and yearly MRCP and CA19-9 tumor markers [5].

H. OLTx is the only curative option for patients with progressive liver disease due to PSC. The 5- and 10-year survival rates for PSC after OLTx have been reported between 75% and 87%.



Algorithm 82.1

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