Congestive hepatopathy

AUTHOR: Lawrence S Friedman, MD
SECTION EDITOR: Sanjiv Chopra, MD, MACP
DEPUTY EDITOR: Kristen M Robson, MD, MBA, FACG

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INTRODUCTION

The liver's complex vascular supply and high metabolic activity make it particularly vulnerable to circulatory disturbances. The severity and characteristics of hepatic injury depend upon the blood vessels that are involved and the degree to which injury is related to passive congestion or diminished perfusion [1-3].

There are several well-recognized forms of vascular injury to the liver, including Budd-Chiari syndrome, hepatic sinusoidal obstruction syndrome, passive congestion due to heart failure, hepatic infarction, and ischemic hepatitis. Congestive hepatopathy refers to hepatic manifestations attributable to passive hepatic congestion, as occurs in patients with right-sided heart failure. Passive congestion often coexists with reduced cardiac output, making their relative contributions to hepatic injury intertwined. (See "Pathogenesis of liver injury in circulatory failure".) Moreover, cardiac dysfunction may occur in the setting of cirrhosis, and cardiohepatic interactions are an area of active investigation [4].

This topic review will focus on passive congestion with a brief discussion of constrictive pericarditis, while discussions on Budd-Chiari syndrome, hepatic sinusoidal obstruction syndrome, ischemic hepatitis, and hepatic infarction are presented separately.

- (See "Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis".)
- (See "Hepatic sinusoidal obstruction syndrome (veno-occlusive disease) in adults".)
- (See "Ischemic hepatitis, hepatic infarction, and ischemic cholangiopathy".)

ETIOLOGY

Any cause of right-sided heart failure can result in hepatic congestion, including constrictive pericarditis, mitral stenosis, tricuspid regurgitation, cor pulmonale, and cardiomyopathy. Tricuspid regurgitation in particular can be associated with severe hepatic congestion because of the transmission of right ventricular pressure directly into the hepatic veins. Liver dysfunction and passive congestion are common in patients with congenital heart disease and single-ventricle physiology who have undergone the Fontan procedure, which directs systemic venous return to the pulmonary artery with bypass of the right ventricle [5,6].

CLINICAL FEATURES

Clinical manifestations — Patients with hepatic congestion are usually asymptomatic. In such patients, hepatic congestion may be suggested only by abnormal liver biochemical tests during routine evaluation.

Symptomatic patients may present with jaundice, which may be mistaken for biliary obstruction [7]. Patients with acute cardiac decompensation may have jaundice and a significant increase in serum aminotransferases, simulating acute viral hepatitis [8,9]. Right upper quadrant discomfort (due to stretching of the liver capsule) and ascites may also be present.

Several cases of acute liver failure with coma, some resulting in death, have been reported in patients with congestive heart failure [7,9-12]. Most have been in the setting of superimposed shock and hepatic ischemia (ischemic hepatitis) rather than passive congestion alone. The term "acute cardiogenic liver injury" has been used when ischemic hepatitis occurs in the setting of congestive hepatopathy [13].

Physical examination — A number of physical findings may be present depending in part upon the severity of the congestion and its chronicity. In addition to jaundice, patients may have hepatomegaly, which can be massive (normal liver span on physical examination as measured by the liver dullness is 10 to 12 cm for men and 8 to 11 cm for women; by ultrasound, a normal liver is typically less than 16 cm in the midclavicular line). The liver edge in congestive hepatopathy is typically firm, smooth, and somewhat tender. Ascites may be detected. Splenomegaly is uncommon, but like cardiac ascites, is attributable to transmitted central venous hypertension rather than liver disease.

Hepatojugular reflux is generally present and is useful in differentiating hepatic congestion from either primary intrahepatic liver disease or Budd-Chiari syndrome. In patients with

significant tricuspid regurgitation, the liver may be pulsatile, with palpable presystolic pulsations corresponding to prominent "v waves" on a right atrial pressure tracing. A loss of hepatic pulsatility in a patient with long-standing congestive hepatopathy suggests progression to cardiac cirrhosis.

Laboratory testing — The most common liver biochemical abnormality is a mild elevation in the serum bilirubin level, which occurs in up to 70 percent of patients [14]. The total serum bilirubin is usually less than 3 mg/dL, most of which is unconjugated [14,15]. Patients with severe, usually acute, right-sided heart failure can develop striking hyperbilirubinemia [16]. The total serum bilirubin level was a predictor of mortality and morbidity in one report [15].

The precise cause of the hyperbilirubinemia is uncertain. Contributing factors may include hepatocellular dysfunction, hemolysis, pulmonary infarction, canalicular obstruction due to distended hepatic veins, medications, and superimposed sepsis [16]. Serum bilirubin levels correlate with right atrial pressures but not with the cardiac output [16].

Other liver biochemical tests are usually only mildly increased. The serum alkaline phosphatase level is usually normal or only minimally elevated in acute heart failure even in the presence of jaundice, helping distinguish jaundice due to congestion from biliary obstruction. However, the serum alkaline phosphatase level may be elevated in chronic severe heart failure [17-19]. The serum gamma-glutamyl transpeptidase (GGT) may also be increased in patients with an acute exacerbation of heart failure [20]. Serum aminotransferase levels are elevated in about one-third of patients, but typically no more than two to three times the upper limit of normal. Occasional patients have higher levels, probably because of coexisting ischemic hepatitis due to decreased cardiac output [8,10]. The degree of elevation in aminotransferases in such patients correlates with the extent of zone 3 necrosis seen on liver biopsy specimens [21]. Striking elevations in aminotransferases (exceeding 1000 international units/L or 50 times the upper limit of normal) may be seen in patients with hypotension due to heart failure and may cause diagnostic confusion with viral hepatitis [22].

Serum albumin levels are decreased in 30 to 50 percent of patients, but are rarely less than 2.5 g/dL. The degree of hypoalbuminemia does not correlate with the degree of histologic liver damage [23], but is an independent predictor of death in patients with acute or chronic heart failure [13]. Hypoalbuminemia is more likely to be caused by malnutrition and protein-losing gastroenteropathy due to increased intestinal lymphatic pressure. (See "Protein-losing gastroenteropathy".)

The prothrombin time is mildly abnormal in many patients. The cause is incompletely understood. It may in part be due to impaired hepatic synthesis of coagulation factors II, V, VII,

IX, and X. However, the prothrombin time may not correct completely with administration of vitamin K, suggesting that other coagulation defects (such as disseminated intravascular coagulation) may also contribute [24].

Serum ammonia levels may occasionally be elevated. Even more rarely, patients may have hepatic encephalopathy in the absence of any other evidence of liver dysfunction. The mechanism leading to hyperammonemia in heart failure is uncertain, but in most cases it appears to be related to heart failure itself rather than to liver disease [25]. Limited data suggested that hyperammonemic encephalopathy due to heart failure improved with lactulose therapy [26].

DIAGNOSIS

Hepatic congestion should be suspected in patients with the above clinical and laboratory features in the setting of congestive heart failure or other cardiac condition associated with elevated central venous pressure. Other causes of liver dysfunction should also be considered, including biliary obstruction, acute or chronic viral hepatitis, hepatic infiltrative disorders, and drug or herbal medication toxicity, depending upon the clinical setting.

In addition to a careful history (ie, risk factors for liver disease and medications that might contribute to hepatic injury) and a physical examination, a reasonable evaluation might include the following:

- **Laboratory testing** – Serologic testing for viral hepatitis, hemochromatosis, Wilson disease (especially in a young person), autoimmune hepatitis, alpha-1 antitrypsin deficiency, celiac disease, and thyroid dysfunction can be obtained.

  A markedly elevated serum N-terminal-proBNP level distinguishes ascites due to heart failure from ascites due to cirrhosis [27]. In one study, a serum BNP level >364 pg/mL had a sensitivity of 98 percent for heart failure-related ascites, and a level <182 pg/mL excluded heart failure-related ascites [28]. (See "Evaluation of adults with ascites".)

- **Imaging** – Imaging studies include right upper quadrant ultrasonography with Doppler studies of the portal and hepatic veins and hepatic artery, electrocardiogram, and echocardiography. Characteristic findings on conventional imaging studies include dilatation of the inferior vena cava and hepatic veins, retrograde hepatic venous opacification shortly after intravenous contrast injection, and heterogeneous hepatic enhancement due to stagnant blood flow [29,30]. Other imaging approaches to identify hepatic congestion and assess fibrosis, including ultrasound elastography [31], diffusion-

weighted magnetic resonance imaging, and magnetic resonance elastography, are under study [29,32-35].

- **Diagnostic paracentesis** – Patients with ascites should undergo a diagnostic paracentesis, which typically reveals a high ascitic protein content, usually greater than 2.5 g/dL, reflecting the relatively well-preserved serum albumin levels and the contribution of "hepatic lymph" to the ascites. The increased protein is presumed to be due to the rupture of hepatic lymphatics, which are rich in protein. The serum-to-ascites albumin gradient is ≥1.1, reflecting portal hypertension [36].

Improvement in liver biochemical tests with treatment of the underlying cardiac condition provides support for the diagnosis. A liver biopsy can help confirm the diagnosis of congestive hepatopathy in equivocal cases (particularly in patients with coexisting liver disease) and establish the severity of histologic injury, but it is rarely required. Moreover, the distribution and pattern of fibrosis is heterogenous throughout the liver, and sampling error can occur with percutaneous liver biopsy [37]. Before considering a percutaneous liver biopsy, careful attention should be given to coagulation parameters, and a percutaneous biopsy should not be performed in patients with ascites. (See "Approach to liver biopsy".)

PATHOLOGY

The term "nutmeg liver" has been used classically to describe the gross appearance of a congested liver. This description reflects reddish central areas (representing sinusoidal congestion and bleeding into atrophic regions surrounding enlarged hepatic veins) with contrasting yellowish areas that represent either normal liver tissue or fatty liver. The liver tends to be enlarged with a purple hue and prominent hepatic veins.

Microscopic examination shows sinusoidal engorgement, hepatocyte atrophy, and variable degrees of hemorrhagic necrosis in zone 3 of the hepatic acinus, fatty change, and variable degrees of cholestasis with occasional bile thrombi in the canaliculi [38]. Bile thrombi are more commonly seen in patients with severe jaundice [16].

In patients with chronic or recurrent heart failure, reticulin and collagen accumulate in zone 3, eventually causing fibrous bands to extend outward from the central veins, occasionally linking with portal tracts to produce a lesion called cardiac sclerosis that resembles micronodular cirrhosis. Progressive fibrosis may lead to bridging between adjacent hepatic venules, producing rings of fibrosis around the spared portal regions that characterizes "cardiac cirrhosis" (or, more accurately, cardiac fibrosis). The histologic pattern is distinct from other

forms of cirrhosis where fibrous bands tend to link adjacent portal areas and is often referred to as "reverse lobulation" [39]. The presence of portal fibrosis in the setting of congestive hepatopathy correlates with high right atrial pressure, right atrial dilatation, and right ventricular dilatation [40]. A histologic congestive hepatic fibrosis scoring system has been proposed [41].

Fibrosis can also involve terminal hepatic venules, causing phlebosclerosis. The regeneration of periportal hepatocytes may result in discrete nodules, producing a pattern known as focal nodular hyperplasia. (See "Focal nodular hyperplasia".)

MANAGEMENT

The cornerstone of management for congestive hepatopathy is treatment of the underlying heart disease. Hepatic congestion and its clinical features, including jaundice and ascites, may respond dramatically to diuretics. Care should be given to avoid excess diuresis, which could impair hepatic perfusion and precipitate zone 3 necrosis [12]. Of paramount importance is maintenance of cardiac output. (See "Overview of the management of heart failure with reduced ejection fraction in adults".)

As noted above, patients may have an underlying coagulopathy, making them particularly sensitive to warfarin anticoagulation. Therefore, coagulation status should be monitored closely if warfarin anticoagulation is being considered. Similar concerns relate to other drugs that require hepatic metabolism.

PROGNOSIS

The prognosis in patients with congestive hepatopathy is predicted mostly by the severity of the underlying heart disease. Liver disease per se rarely contributes significantly to morbidity or mortality. However, increasing serum aminotransferases on day 3 and decreasing serum albumin on day 4 of a hospital admission for acute heart failure have been associated with adverse outcomes at six-month follow up [42].

Early histologic changes associated with passive congestion may resolve with medical therapy to treat the underlying cardiac condition. By contrast, prolonged hepatic congestion over months to years can lead to cirrhosis. Most of these patients have either constrictive pericarditis or mitral valve disease and secondary tricuspid regurgitation.

The presence of cirrhosis does not necessarily signal a poor prognosis. Such patients uncommonly develop serious sequelae of chronic liver disease such as variceal bleeding [16,43]. However, emerging data suggested that liver elastography and a higher score on a noninvasive measure of liver stiffness (eg, Fibrosis-4 index, which reflects liver fibrosis and correlates with right atrial pressure) were associated with higher rates of all-cause mortality in patients with heart failure [44-47]. (See "Noninvasive assessment of hepatic fibrosis: Overview of serologic tests and imaging examinations", section on 'Indirect markers of fibrosis'.)

The Model for End-Stage Liver Disease (MELD) score excluding the international normalized ratio predicts outcomes in patients with congestive hepatopathy, who are frequently taking an anticoagulant [34,48].

Hepatocellular carcinoma may occur in patients with cardiac cirrhosis, particularly those who have had a Fontan procedure in childhood [49-51]. (See "Management of complications in patients with Fontan circulation", section on 'Liver disease'.)

CONSTRICTIVE PERICARDITIS

Constrictive pericarditis is associated with clinical and pathologic changes in the liver similar to those seen in the Budd-Chiari syndrome [52]. Hepatic vein pressures are generally higher than those seen in patients with right-sided heart failure, making patients with constrictive pericarditis relatively more likely to develop hepatic necrosis and ultimately cirrhosis.

Patients with pericardial constriction may present with two types of complaints: those related to fluid overload, ranging from peripheral edema to anasarca; and those related to diminished cardiac output in response to exertion, such as fatigability and dyspnea on exertion. Constriction should be considered in any patient with an unexplained elevation in jugular venous pressure, particularly if there is a history of a predisposing condition. (See "Constrictive pericarditis: Diagnostic evaluation".)

Hepatomegaly, a pulsatile liver, massive ascites, and peripheral edema are common. By contrast, jaundice is characteristically absent, for unclear reasons [53]. The clinical features described are relatively nonspecific, making it easy to mistake the underlying cause for primary hepatic cirrhosis or Budd-Chiari syndrome. Thus, a high index of suspicion for the diagnosis must be maintained. Important clues include elevated jugular venous pressure, Kussmaul's sign (a rise in the jugular venous pressure on inspiration), a pericardial knock, and pericardial calcification on chest radiograph. A correct diagnosis is critical since pericardiectomy is curative if performed early. (See "Constrictive pericarditis: Diagnostic evaluation".)

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Heart failure in adults".)

SUMMARY AND RECOMMENDATIONS

- **Etiology** – Hepatic congestion can result from any cause of right-sided heart failure, including constrictive pericarditis, mitral stenosis, tricuspid regurgitation, cor pulmonale, and cardiomyopathy. (See 'Etiology' above.)
- **Clinical features** – Hepatic congestion should be suspected in patients with clinical and laboratory features suggestive of congestive heart failure or other cardiac conditions associated with elevated central venous pressure. (See 'Clinical features' above.)
- **Diagnosis** – In addition to a careful history (especially for risk factors for liver disease and medications that might contribute to hepatic injury) and physical examination, a reasonable evaluation might include serologic testing for viral hepatitis, hemochromatosis, Wilson disease (especially in a younger person), alpha-1 antitrypsin deficiency, celiac disease, autoimmune hepatitis, thyroid dysfunction, right upper quadrant ultrasonography with Doppler studies of the portal and hepatic veins and hepatic artery, electrocardiogram, echocardiography, and, if indicated, diagnostic paracentesis. (See 'Diagnosis' above.)
- **Management** – The cornerstone of management is treatment of the underlying heart disease. Care should be given to avoid excess diuresis, which could impair hepatic perfusion. Patients who require warfarin anticoagulation for their cardiac condition should be monitored closely, since patients with congestive hepatopathy are at increased risk for excessive anticoagulation. (See 'Management' above.)

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Contributor Disclosures

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