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section 15 Gastroenterological disorders 3166 Padda MS, et al. (2011). Drug-induced cholestasis. Hepatology, 53, 1377–87. Ramappa V, Aithal GP (2013). Hepatotoxicity related to anti- tuberculosis drugs: mechanisms and management. J Clin Exp Hepatol, 3, 37–49. Urban, Daly AK, Aithal GP (2014). Genetic basis of drug-induced liver injury: present and future. Semin Liver Dis, 34, 123–33.
15.24.4 Vascular disorders of
the liver Alexander Gimson ESSENTIALS The liver's complex blood supply and high metabolic activity may be affected in a number of clinical situations when there is reduced splanchnic inflow and impaired hepatic venous outflow. Budd–Chiari syndrome is caused by obstruction to hepatic venous outflow, usually by thrombosis within the hepatic veins. Causes in- clude myeloproliferative disease, malignancy, and hypercoagulable states. Presentation may be acute, subacute, or chronic, and the diag- nosis requires consideration in any patient presenting with acute liver failure, acute hepatitis, or chronic liver disease. Diagnosis is made by Doppler ultrasonography of the hepatic veins and confirmed with CT or MRI scanning. Management depends on the presentation, but involves anticoagulation in all cases, and thrombolysis, measures to decompress the liver, and liver transplantation in some. Survival is about 75% at 5 years. Other conditions discussed include congestive hepatopathy, ischaemic hepatopathy, hepatic artery aneurysm, and portal vein thrombosis. Budd–Chiari syndrome Budd–Chiari syndrome is caused by obstruction to hepatic venous outflow. It is described as primary when the obstruction is caused by a process arising within the veins (e.g. thrombosis), and secondary when due to another process (e.g. compression or invasion by ma- lignancy). It is a rare condition (incidence about 2 per million in both men and women) that can present at any age (median 50 years). Most cases develop insidiously over months, but about 20% present acutely. An underlying predisposing cause can be found in most pa- tients, often in combination (Table 15.24.4.1). Clinical features Acute Budd–Chiari syndrome, which develops over a few weeks, typically presents with fever, right upper quadrant pain, and tender hepatomegaly, which may be associated with hepatic encephalop- athy if there is fulminant liver failure (5% of cases). Jaundice and ascites develop rapidly but may not be evident at presentation. Liver blood tests show elevation of bilirubin, aminotransferases (typically 100–500 IU/L), and alkaline phosphatase (typically 300–400 IU/L). Subacute Budd–Chiari syndrome usually

presents with vague epigastric or right upper quadrant pain. Ascites and pedal oedema may be present but are not invariable. The latter is thought to be due to hypertrophy of the caudate lobe of the liver causing com- pression of the inferior vena cava, into which it drains directly, ra- ther than through the hepatic veins. Abnormal venous collaterals may be seen on the abdominal wall. Variceal bleeding can occur. Patients with chronic Budd–Chiari syndrome develop symptoms as a consequence of cirrhosis. In both subacute and chronic cases, Table 15.24.4.1 Causes of the Budd–Chiari syndrome Cause Example Notes Myeloproliferative disease (50% cases of Budd–Chiari syndrome) Polycythaemia rubra vera Essential thrombocythaemia Malignancy (10% cases) Hepatocellular carcinoma Benign liver lesions (5–10% cases) Hepatic cysts and abscesses Hypercoagulable states Oral contraceptive use Clotting factor mutations, e.g. factor V Leiden, factor II (G20210A) deficiency, antithrombin III, protein C and S deficiency, antiphospholipid syndrome Paroxysmal nocturnal haemoglobinuria Nearly 20% of cases occur in women who are taking the oral contraceptive, are pregnant, or have recently given birth Anatomical abnormality Membranous webs of inferior vena cava and/or hepatic veins Miscellaneous conditions Behçet's disease Inflammatory bowel disease Coeliac disease Other autoimmune conditions Idiopathic Some may have undiagnosed myeloproliferative disorder; always check for JAK2 mutations Note: multiple risk factors may contribute in an individual case, for example, a combination of factor V Leiden mutation with a myeloproliferative disorder or use of the oral contraceptive.

 serum bilirubin, aminotransferases, and alkaline phosphatase may be normal or mildly/moderately elevated, but clinically obvious jaundice is rare. Diagnosis The very variable presentations of Budd–Chiari syndrome mean that it should be considered in the differential diagnosis of patients pre- senting with acute liver failure, acute hepatitis, or chronic liver dis- ease. The diagnosis can usually be made by Doppler ultrasonography of the hepatic veins (Fig. 15.24.4.1) and confirmed with CT or MRI scanning (Fig. 15.24.4.2), which provide additional information re- quired for treatment planning. Venography may be required if uncer- tainty remains after these tests, looking for the characteristic 'spider's web' pattern in occluded hepatic veins. Liver biopsy is not generally required. Causes listed in Table 15.24.4.1 should be sought. Management and prognosis Treatment, when possible, should be given for any underlying cause. With regards to the thrombosis within the hepatic veins, the initial priorities are to initiate anticoagulation (usually commencing with low molecular weight heparin), unless there are very strong contra- indications, and to deal with any complications of portal hyperten- sion that are present. Thrombolysis may be given, depending on balance of benefits and risks in each case, if imaging reveals an acute (usually regarded as <4 weeks) and well-defined clot, but thrombolysis should not be administered to patients with chronic Budd–Chiari syndrome. Angioplasty and stenting may be used in symptomatic patients with suitable anatomy, but stenting should only be performed after dis- cussion with a liver transplant centre because it may compromise the prospects for subsequent liver transplantation. Apart from patients with subacute or chronic presentation and well-compensated liver disease, measures to decompress the liver should be considered in most cases with the aim of preventing progression of liver disease. This can be done in many cases by placement of a transjugular intrahepatic portosystemic shunt. Less commonly, it may be possible to surgically create a shunt between the mesenteric or portal venous system and the inferior vena cava, thereby reducing blood flow and pressure within the liver. Liver transplantation may be the only option for those in whom these treatments are not possible or fail, or who have decompensated cirrhosis. Long-term anticoagulation will be necessary, even after transplantation. Before treatments were available, mortality at 3 years was 90%. Recent series

report 5-year survival rates of about 75%. Features associated with poor prognosis include old age, severe liver failure, refractory ascites, chronic disease at presentation, and occlusion of the portal vein (which limits treatment options). Congestive hepatopathy Congestive hepatopathy may occur in the context of reduced car- diac output and high right-sided venous pressure, due to con- strictive pericarditis, mitral stenosis, tricuspid regurgitation, or cardiomyopathy. The hepatic congestion and reduced liver sinus- oidal perfusion is usually asymptomatic as the heart failure dom- inates the clinical scenario. There is an elevated jugular venous pressure, with 'v' waves in the presence of tricuspid regurgitation, hepatomegaly, and a positive hepatojugular reflex. The liver may be pulsatile when tricuspid regurgitation is severe. The consequent reduced sinusoidal blood flow can lead to ischaemia in zone 3 of the hepatic lobule and histological changes are characterized by centrilobular congestion with surrounding fatty change, initially described as the 'nutmeg liver'. If the disorder is long-standing, there may be progressive fibrosis extending peripherally from the centrilobular to the periportal areas although regenerative nodules are usually not present. (a) (b) Fig. 15.24.4.1 Colour Doppler ultrasonographic images in a patient with acute Budd–Chiari syndrome showing (a) absent flow in the middle hepatic vein (MHV), and (b) biphasic flow in the right hepatic vein (RHV). The inferior vena cava is patent (arrows). From Levy AD, Mortele KJ, Yeh BM (eds) (2015). Gastrointestinal imaging. By permission of Oxford University Press.

section 15 Gastroenterological disorders 3168 A modestly elevated serum bilirubin level may be the only ab- normality, with a normal serum alkaline phosphatase and elevated aminotransferases. Although minor changes in liver biochemistry are observed in cases with chronic biventricular heart failure, true cases of cardiac cirrhosis are very uncommon, as are major com- plications from portal hypertension. Occasionally, this situation is complicated by the presence together of an alcohol-induced liver injury and an alcoholic cardiomyopathy. In these circumstances, it may be necessary to undertake histological examination of the liver as well as measuring right-sided cardiac pressures and free and wedged hepatic venous pressure gradients. The treatment for congestive hepatopathy centres on improving cardiac function with standard therapies. Ischaemic hepatopathy This refers to the diffuse hepatic injury that follows an acute re- duction in hepatic blood flow. It may be due to any of the causes of sudden shock (haemorrhage, cardiac arrest, severe septic shock) but a similar syndrome has also been described in hepatic sickle cell crisis, following liver transplantation with hepatic artery throm- bosis, and in severe respiratory failure and hypoxaemia. Histology shows a centrizonal necrosis and blood tests demonstrate marked elevation of transaminases, prolonged prothrombin time, and jaundice. The differential diagnosis includes other cause of sudden hep- atocyte necrosis, including drug hepatotoxicity and viral hepatitis. Rarely, acute liver failure with encephalopathy may develop in the most severe cases of ischaemic hepatopathy, with mortality up to 50% in pooled series. There is no specific treatment for the liver dys- function in this context, management being directed at the cause of the impaired hepatic perfusion. Hepatic artery aneurysm Hepatic artery aneurysms have been found in a number of con- ditions but are most common after surgery. They may also be found in the Osler–Weber–Rendu syndrome, Behçet's syndrome, polyarteritis nodosa, and as part of systemic sepsis with fungal in- fection. It may be an incidental finding on an angiogram or present (a) (b) (c) (d) Fig. 15.24.4.2 Acute Budd–Chiari syndrome. CT images acquired before contrast administration (a) and afterwards—arterial phase (b), portal venous phase (c), and 3-min delayed (d) images. These show diffuse peripheral oedema and hypoattenuation due to congestion (asterisks in (a)), early enhancement of the central portion of the liver ((b) and (c)) with delayed enhancement of the congested periphery (d). The hepatic veins are occluded (arrows in (c) and (d)). From Levy AD,

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