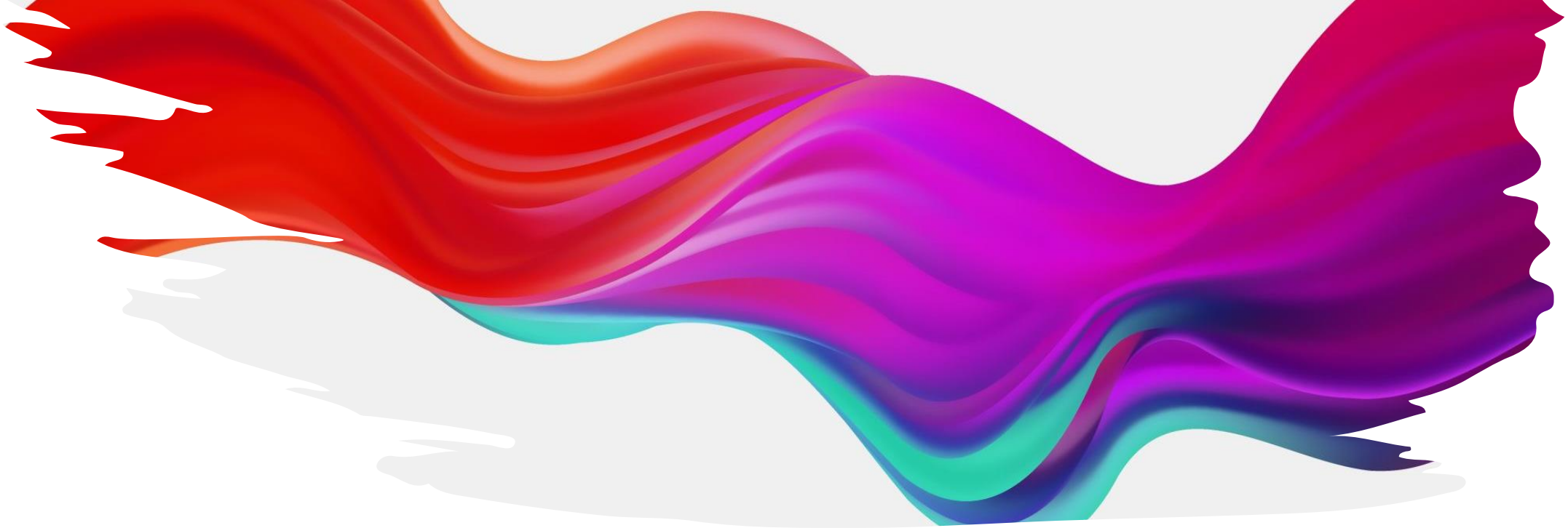
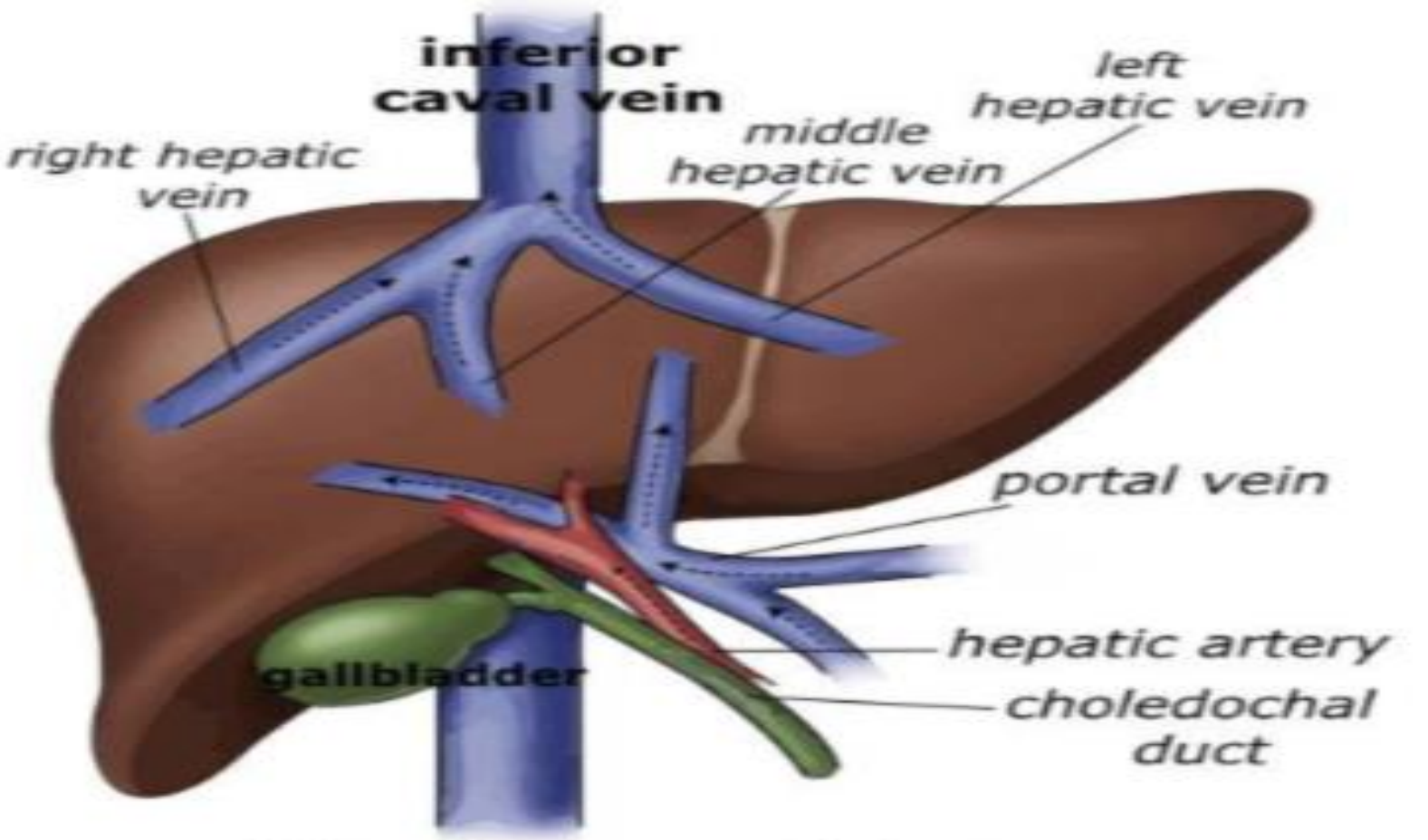


Budd-Chiari syndrome

Hepatic
venous outflow tract obstruction

HEPATIV VEIN



Hepatic vasculature

Primary Budd-Chiari syndrome is present when there is obstruction due to a predominantly venous process (thrombosis or phlebitis).

Secondary Budd-Chiari is present when there is compression or invasion of the hepatic veins and/or the inferior vena cava by a lesion that originates outside of the vein (eg, a malignancy).

EPIDEMIOLOGY

Non-Asian countries, Budd-Chiari syndrome is more common in women and usually presents in the third or fourth decade of life .
Pure hepatic vein blockage is more common
Asia, there is a slight predominance of men, with a median age of 45 years at presentation.
pure inferior vena cava or combined inferior vena cava and hepatic vein blockage predominate

ETIOLOGY

Prothrombotic risk factors for BCS

Acquired thrombophilia :

Myeloproliferative disease

Polycythemia vera

Essential thrombocytosis

Idiopathic myelofibrosis

JAK2 V617F mutation

Paroxysmal nocturnal hemoglobinuria

Behcet disease

Hyperhomocysteinemia

Antiphospholipid syndrome

Inherited thrombophilia

Factor V Leiden

Prothrombin gene G20210A

mutation MTHFR C677T mutation Thalassemia

PC deficiency

Protein S deficiency

Antithrombin deficiency

Systemic factors

Sarcoidosis

Vasculitis

Behcet disease

Connective tissue disease

Inflammatory bowel disease

Hormonal factors

Recent oral contraceptive use

Pregnancy

Myeloproliferative disorders

As many as 50 percent of all cases of the Budd-Chiari syndrome may be due to an underlying chronic myeloproliferative disorder (eg, polycythemia vera, essential thrombocythemia, agnogenic myeloid metaplasia) and an accompanying hypercoagulable state

Infections and benign lesions of the liver

Infections or benign space-occupying lesions of the liver can, like tumors, compress and/or thrombose vascular structures, accounting for nearly 10 percent of cases of Budd-Chiari syndrome. Some of these disorders also may be accompanied by a hypercoagulable state. These lesions include hepatic cysts and abscesses, hepatic adenoma, hepatic mucinous cystic neoplasm (cystadenoma), syphilitic gumma, invasive aspergillosis, zygomycosis (mucormycosis), and aortic aneurysm

Malignancy

Malignancies account for approximately 10 percent of cases of the Budd-Chiari syndrome. Direct compression or invasion of vascular structures and the hypercoagulable state associated with malignancy can result in venous thrombosis and/or obstruction

Hepatocellular carcinomas are found most often, followed by cancer of the adrenal gland or kidney, sarcomas of the right atrium, inferior vena cava or hepatic veins, and cancers of the lung, pancreas, and stomach..

Oral contraceptives and pregnancy

Nearly 20 percent of cases of the Budd-Chiari syndrome occur in women who have been on oral contraceptives (for as little as two weeks), are pregnant, or have delivered a child within the previous two months . It is presumed that the hypercoagulable state in these women is responsible for this association.

Behçet syndrome

Vasculitis is an important feature of Behçet syndrome

Membranous webs

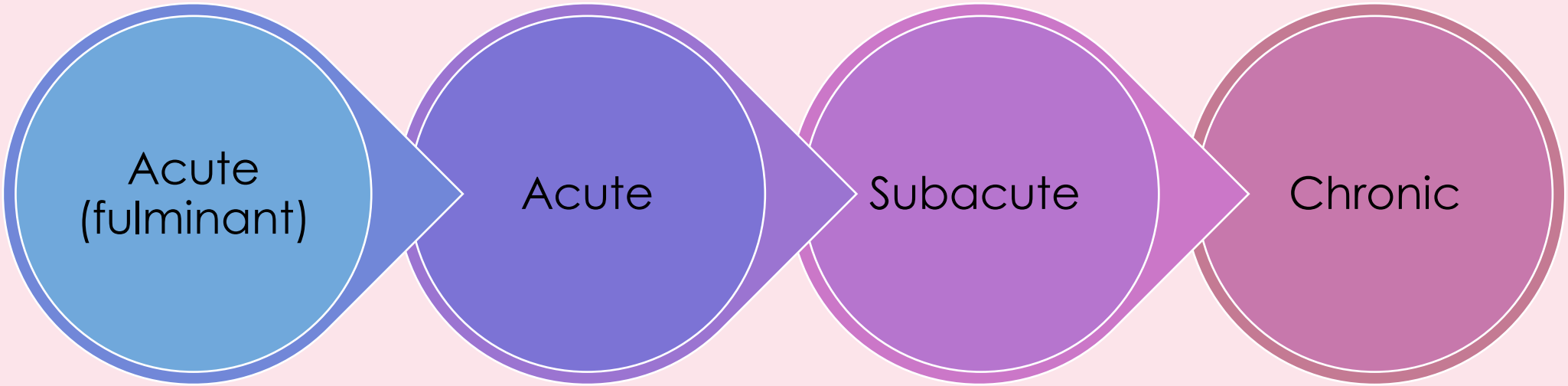
Membranous obstruction (partial or complete) of the inferior vena cava (MOVC) and/or the hepatic veins is an unusual but potentially treatable cause of the Budd-Chiari syndrome in the United States

Idiopathic

Approximately 20 percent of cases of the Budd-Chiari syndrome are listed as idiopathic . As more and more conditions which predispose to Budd-Chiari syndrome, such as occult myeloproliferative disease and factor V Leiden mutation, are recognized, the percentage of cases described as idiopathic will continue to decrease

Underlying disorder can be identified in over 80 percent of patients with the Budd-Chiari syndrome

CLINICAL MANIFESTATIONS


Acute
(fulminant)
Acute
Subacute
Chronic

VENOUS COLLATERAL.

Patients with acute liver failure or acute (non-fulminant) liver disease have not yet developed venous collaterals, whereas venous collaterals are seen in patients with subacute and chronic liver disease.

Typical and acute

ABDOMINAL PAIN, ABDOMINAL DISTENSION FROM ASCITES, HEPATOMEGALY,

Patients with subacute or chronic Budd-Chiari syndrome may be asymptomatic. In such patients, the hepatic venous outflow obstruction is often discovered as part of the evaluation of abnormal liver blood tests or when imaging is obtained for other reasons.

Because the presentation of Budd-Chiari syndrome is highly variable, clinicians should consider it in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease

Acute liver failure

Acute liver failure is characterized by acute liver injury with elevated transaminases, jaundice, hepatic encephalopathy, and an elevated prothrombin time/international normalized ratio; hepatic encephalopathy develops within eight weeks after the development of jaundice

Lab test

Elevation of serum aminotransferases occurs because vascular congestion results in ischemic hepatocellular damage. Serum aminotransferase concentrations can range from 100 to 200 int. units/L to more than 600 int. units/L. The serum alkaline phosphatase in acute Budd-Chiari syndrome is often in the range of 300 to 400 int. units/L and serum bilirubin levels are usually less than 7 mg/dL at the time of presentation, though they may subsequently increase.

SAAG

Ascitic fluid in patients with Budd-Chiari syndrome has a high serum-to-ascites protein gradient (>1.1), reflecting elevated portal pressures

Subacute and chronic Budd-Chiari syndrome

Patients with subacute or chronic Budd-Chiari syndrome may be asymptomatic or minimally symptomatic until the disease progresses . It is estimated that 15 to 20 percent of patients are asymptomatic . Patients who are asymptomatic often have large hepatic vein collaterals

subacute

Patients may report a history of vague discomfort in the mid epigastrium or right upper quadrant. Ascites and hepatic necrosis may be minimal due to decompression of the sinusoids by portal and hepatic venous collaterals.

CHRONIC

Patients who develop cirrhosis may have stigmata of chronic liver disease such as spider angiomata and palmar erythema. They may also have signs of portal hypertension such as ascites (which may be massive) and esophageal varices. Hepatomegaly and abdominal pain are also common, but encephalopathy is not. Hepatopulmonary syndrome has been described in up to 28 percent of patients.

When to consider Budd-Chiari syndrome

Clinicians should consider it in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease, particularly if the patient has known risk factors for Budd-Chiari syndrome . Among patients with acute liver failure, the presence of hepatomegaly, right upper quadrant pain, and ascites should increase the suspicion for Budd-Chiari syndrome.

Establishing the diagnosis

Noninvasively with Doppler ultrasonography

The diagnosis of Budd-Chiari syndrome can usually be established noninvasively with Doppler ultrasonography. The portal and splenic circulation should also be evaluated to exclude concurrent portal or splenic vein thrombosis.

CT AND MRI

Computed tomography (CT) or magnetic resonance imaging (MRI) can be performed to confirm the diagnosis, to aid with treatment planning, or if an experienced Doppler sonographer is not available. In addition, CT or MRI can be performed in patients with an unremarkable ultrasound examination but in whom the suspicion for Budd-Chiari syndrome is high (eg, a patient with a thrombophilic disorder and acute hepatitis whose evaluation for other causes of hepatitis is negative

Venography

Venography should be performed if noninvasive tests are negative or nondiagnostic, but there is strong clinical suspicion for the disease. It can also be used to direct subsequent therapy by clearly defining which vessels are involved. The gold standard for diagnosing Budd-Chiari syndrome is hepatic venography, which is performed by accessing the hepatic venous circulation percutaneously via the internal jugular vein, cephalic vein, or femoral vein.

Management

Correcting underlying disorders that predisposed to the development of Budd-Chiari syndrome (when possible)

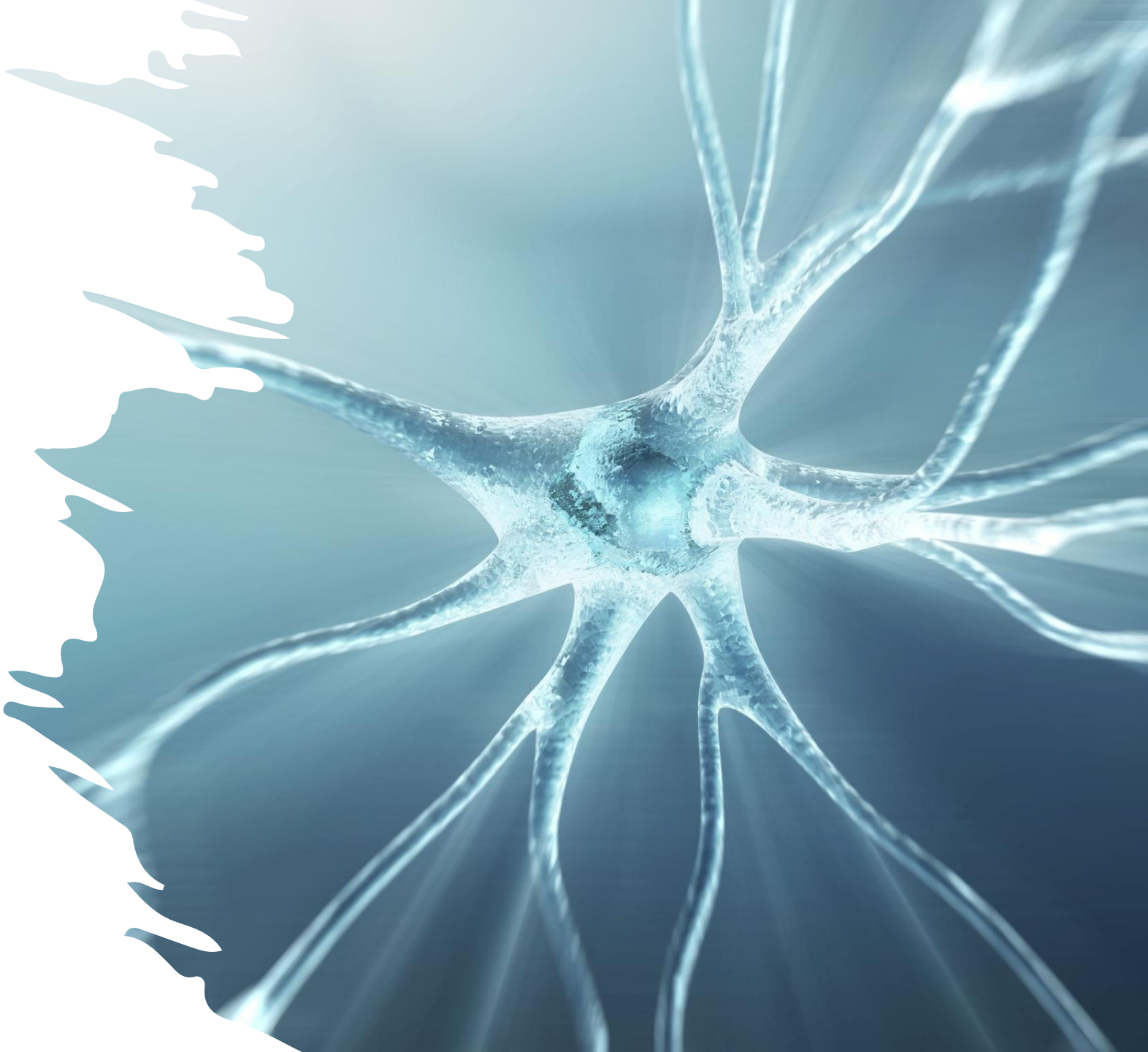
Goals of therapy

Prevent the propagation of the clot

Restore patency of thrombosed veins

Decompress the congested liver

Prevent or manage complications



Prevent propagation of the clot

Anticoagulation should be initiated as soon as possible in most patients to prevent propagation of the clot, provided there are no contraindications. However, the risk of anticoagulation should be considered, especially in patients who present with bleeding complications or who have varices. Prior to initiating anticoagulation, we perform an upper endoscopy to screen for varices.

Treatment

Initiating anticoagulation unless there are contraindications

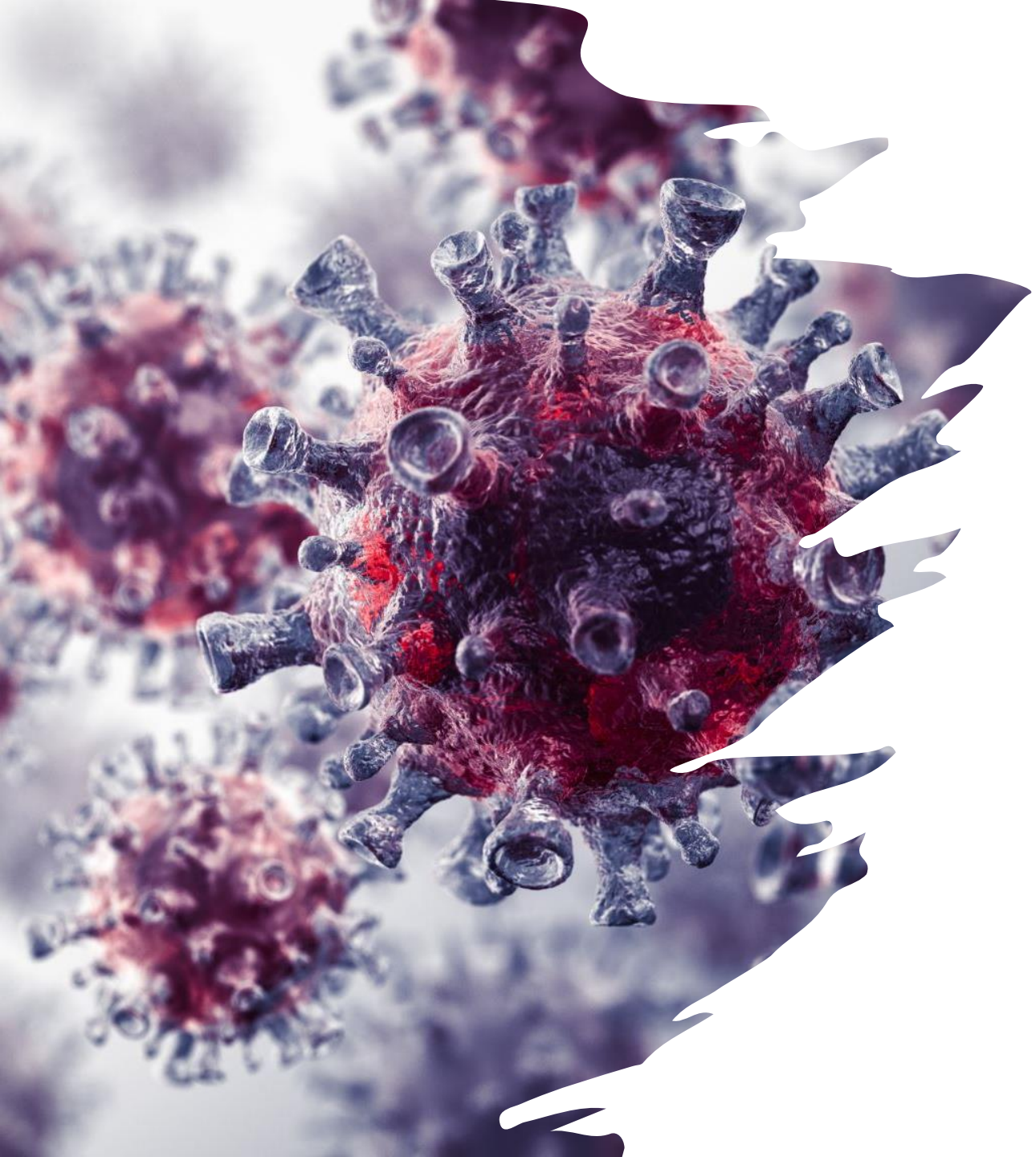
Treating complications of portal hypertension (if present)

Anticoagulant

We prefer to treat initially with low molecular weight heparin. In selected patients, such as those at high risk for bleeding or who have renal impairment, we will check anti-factor Xa activity, targeting it to between 0.5 and 0.8 international unit/mL. We also start patients on an oral vitamin K antagonist (eg, warfarin). Once the international normalized ratio is between two and three, we discontinue the low molecular weight heparin.

THROMBOLYSIS indication

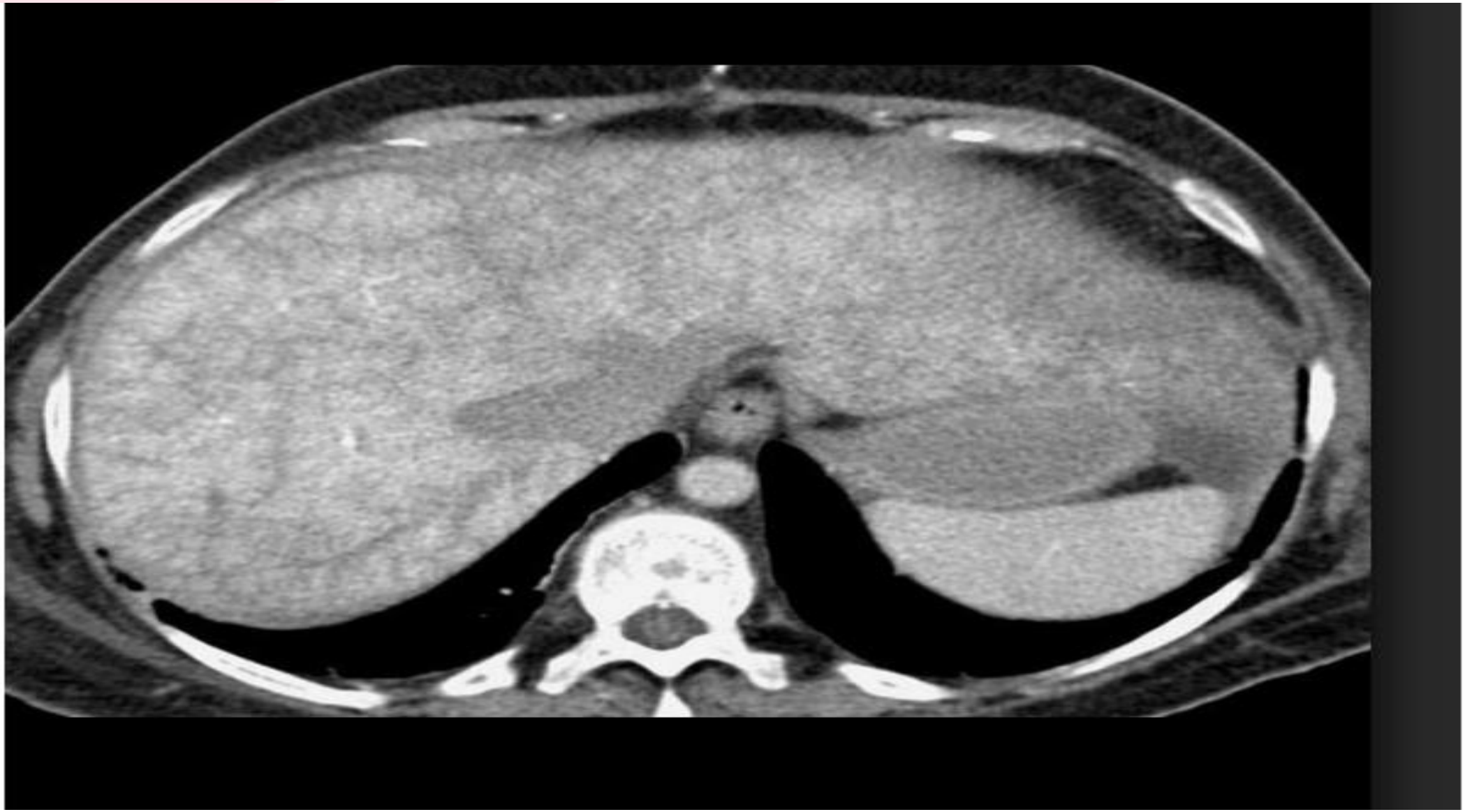
- Well-defined clot is present.
- No contraindication.
- Acute (less than 4 weeks).



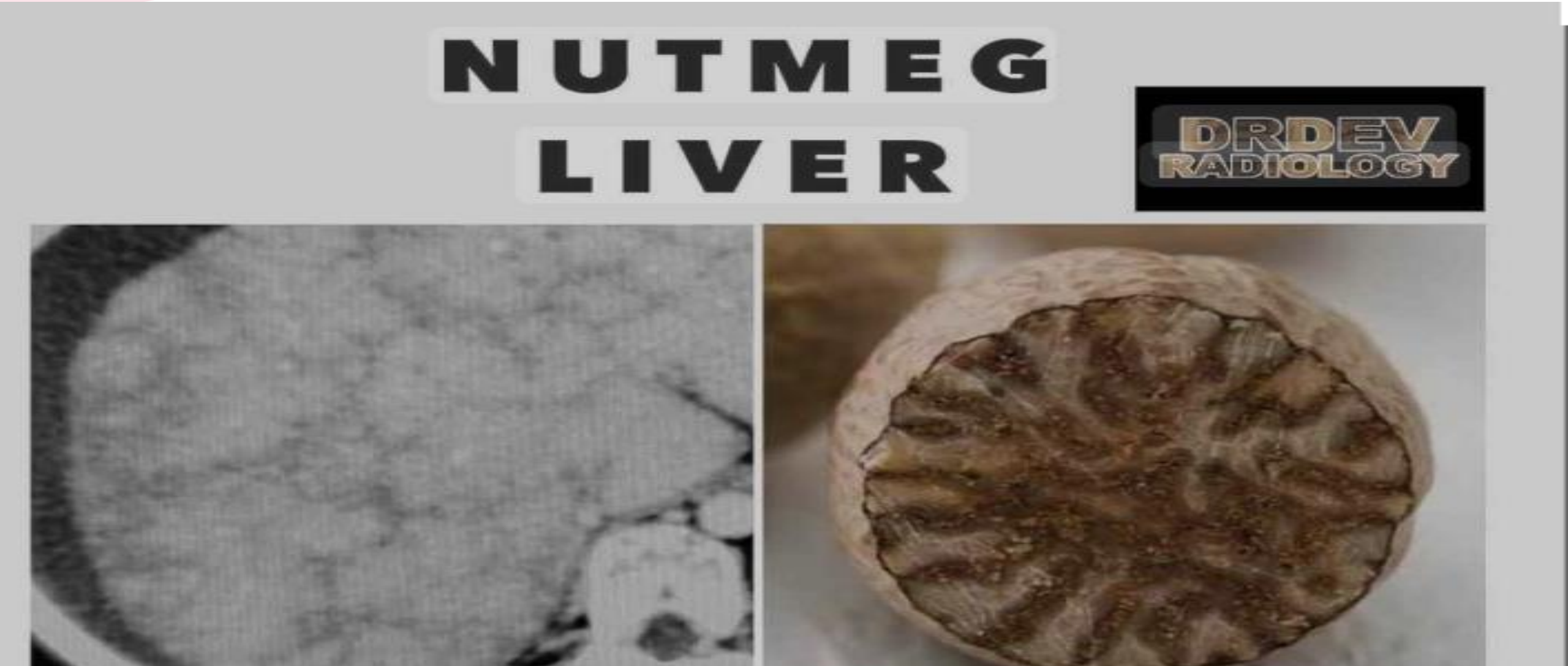
Prognosis

Without treatment, the prognosis is poor (survival rates of approximately 10 percent at three years). However, with treatment, five-year survival rates of approximately 75 percent have been reported

CT imaging



Nutmeg



THANK YOU