BUDD-CHIARI SYNDROME – A REVIEW ARTICLE

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ABSTRACT

Budd-Chiari syndrome is defined as obstruction of hepatic venous outflow from the small hepatic veins inside the liver to the inferior vena cava and right atrium. Patient may have no symptoms to fulminant liver failure. The diagnostic studies are Doppler ultrasonography. Management includes supportive medical therapy and measures to establish and maintain venous patency, such as thrombolysis, decompression with shunts, and long-term anticoagulation to remove obstruction.

Key words : Budd Chiari syndrome, liver failure, Thrombolysis, anticoagulation.

INTRODUCTION

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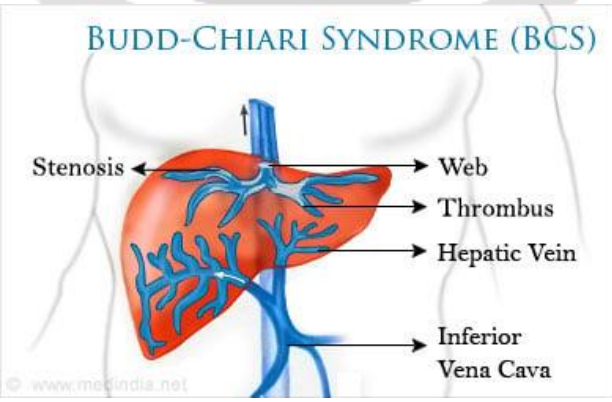
DEFINITION

Budd-Chiari syndrome is defined as obstruction of hepatic venous outflow from the small hepatic veins inside the liver to the inferior vena cava and right atrium.

STENOSIS AND OBSTRUCTION IN HEPATIC VEIN



CAUSES

The main cause is a clot obstructing the hepatic veins and the adjacent inferior vena cava. Clots commonly result from the following:

- Thrombotic conditions like protein C deficiency, protein S deficiency, antiphospholipid syndrome, antithrombin III deficiency, factor V Leiden mutation,during pregnancy and oral contraceptive use)
- Hematologic disorders like myeloproliferative disorders such as polycythemia and paroxysmal nocturnal hemoglobinopathy)
- Inflammatory bowel disease
- Systemic rheumatic heart diseases
- Trauma or severe injury
- Infection example hydatid cyst, amebiasis)
- Tumor that invasion into the hepatic vein (eg, hepatocellular carcinoma or renal cell carcinoma)
- Sometimes Budd-Chiari syndrome begins during pregnancy .

Budd-Chiari syndrome develops over weeks or months to show signs and symptoms

TYPES OF BUDD-CHIARI SYNDROME

Acute Budd-Chiari syndrome with acute liver failure: Symptoms develop quickly (within days or weeks), and liver function rapidly declines.
Acute Budd-Chiari syndrome without liver failure: Symptoms develop quickly but without loss of liver function.
Subacute Budd-Chiari syndrome: Symptoms set in more slowly, usually over several months. This is the most common type of Budd-Chiari syndrome.
Chronic Budd-Chiari syndrome: Symptoms usually aren't noticeable until the blockage has already caused irreversible scarring (cirrhosis) in liver.
Budd-Chiari syndrome may be primary or secondary:
Primary Budd-Chiari syndrome occurs because of a narrowing or blockage within blood vessels. Causes include a blood clot (bland thrombus) or a blockage from a collection of tumor cells. Medical conditions that cause inflammation of hepatic veins may also contribute.
Secondary Budd-Chiari syndrome occurs because of an issue outside of blood vessels. Causes include a malignant (cancerous) or benign (noncancerous) tumor placing pressure on one of hepatic veins or inferior vena cava (IVC).

DIAGNOSTIC EVALUATION

- Vascular imaging
- History collection in patients with hepatomegaly, ascites, liver failure, or cirrhosis when there is no obvious cause (eg, alcohol abuse,alcoholic hepatitis) or when the cause is unexplained.
- Liver tests are usually abnormal like liver enzymes, bilirubin level.
- Imaging usually begins with Doppler ultrasonography in abdomen.
- CT , MRI, PETCT of abdomen
- Liver biopsy is done occasionally to identify the acute stages and identify whether cirrhosis has developed.

SIGNS AND SYMPTOMS

The signs and symptoms of Budd-Chiari syndrome include:

- Abdominal pain (particularly in right upper region of abdomen).
- Ascites (accumulation by excess fluid in peritoneum).
- Jaundice (skin, and eyes and tongue turns yellow in colour).
- Hepatic encephalopathy (confusion or coma caused by hepatic disease).
- Hepatomegaly (enlarged liver).
- Splenomegaly (enlarged spleen).
- Bleeding from esophagus, stomach or rectum.
- Edema (swelling) in both legs.
- Fatigue (extreme tiredness).
- Liver failure (liver have no functions adequately)

TREATMENT

Treatments for Budd-Chiari syndrome dissolve clots or improve narrowing in hepatic veins or IVC to decrease swelling and scar formation in liver. Whenever possible, treatments involve managing the underlying condition that caused to develop clots. Also, managing complications, like portal hypertension (high blood pressure in portal vein), is essential.

Medications

Doctor may prescribe blood thinners (anticoagulants), such as heparin or warfarin, to dissolve the blood clots.

Nonsurgical procedures

Patient may need a procedure to help redirect blood flow and relieve pressure in blood vessels. Procedures include:

Transjugular intrahepatic portosystemic shunt (TIPS): A healthcare provider makes a tunnel through liver with a needle. Then, they connect the portal vein (the vein that carries blood to liver) to one of hepatic veins (the three veins that carry blood from liver). They'll use X-ray imaging to guide them as they place a stent (tiny metal tube) into this tunnel to keep the track open.

Percutaneous transluminal angioplasty: A healthcare provider inserts a catheter (a thin, hollow tube with a balloon at the tip) through the skin and into a blood vessel. They'll guide the catheter to the clot or narrowed area. When the catheter reaches the affected area, inflate the balloon to widen the vein. They may place a stent at the site to keep the vein open.

Liver transplant

Patient may need a liver transplant if other treatments haven't helped or aren't an option. For example, patient likely need a liver transplant if he have liver failure, cirrhosis of the liver, bleeding from varices or ascites that can't be treated with medications.

Supportive care

- Restoration and maintenance of adequate venous outflow into the liver veins
- Treatment varies according to onset (acute vs chronic) and severity (fulminant liver failure vs decompensated cirrhosis or stable/asymptomatic).
- Giving supportive therapy according to the complications (eg, ascites, liver failure, esophageal varices)
- Preventing propagation of the clot and prevention of complications

COMPLICATIONS

- Hepatic encephalopathy.
- Variceal hemorrhage.
- Hepatorenal syndrome.
- Portal hypertension.
- Bacterial peritonitis in case of ascites.
- Hepatocellular cancer.

CONCLUSION

Budd Chiari syndrome is termed as obstruction in the hepatic and portal system in the liver. This shows clinical manifestations like abdominal pain, vomiting. the management includes thrombolytic agents , trans hepatic porto systemic shunt.

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