

Imaging of Budd-Chiari Syndrome

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Budd-Chiari Syndrome (BCS) is a clinical syndrome of ascites, hepatomegaly and abdominal pain secondary to hepatic venous outflow obstruction. The obstruction is usually at the level of inferior vena cava (IVC) or major hepatic veins. Occasionally, the obstruction is at the hepatic venule level, which is called hepatic veno-occlusive disease⁽¹⁾.

Etiologies of BCS are numerous as following⁽²⁾:

1. Congenital obstruction of IVC secondary to web or diaphragm.
2. Tumor invasion of IVC and/or hepatic veins: common tumors include hepatoma, renal cell carcinoma, adrenal carcinoma or IVC leiomyosarcoma.
3. Inflammatory process, such as autoimmune disease, radiation injury, and use of immunosuppressive drugs.

4. Venous thrombosis secondary to many pathological processes, such as polycythemia vera, antiphospholipid syndrome, pregnancy, oral contraceptive pills, paroxysmal nocturnal hemoglobinuria and thrombocytosis.

5. Idiopathic or unknown etiologies.

Imaging findings are as following^(3,4):

1. US shows thrombus within the hepatic veins and/or IVC. Color Doppler may show absent or flat flow within the hepatic veins.
2. CT shows hypertrophy of caudate lobe and atrophy of the right lobe. Caudate lobe tends to spare secondary to its direct venous drainage to IVC. Caudate lobe enhances at the early phase whereas peripheral liver enhances at the later phase (flip-flop sign). Regenerative nodules occur in a chronic form. The

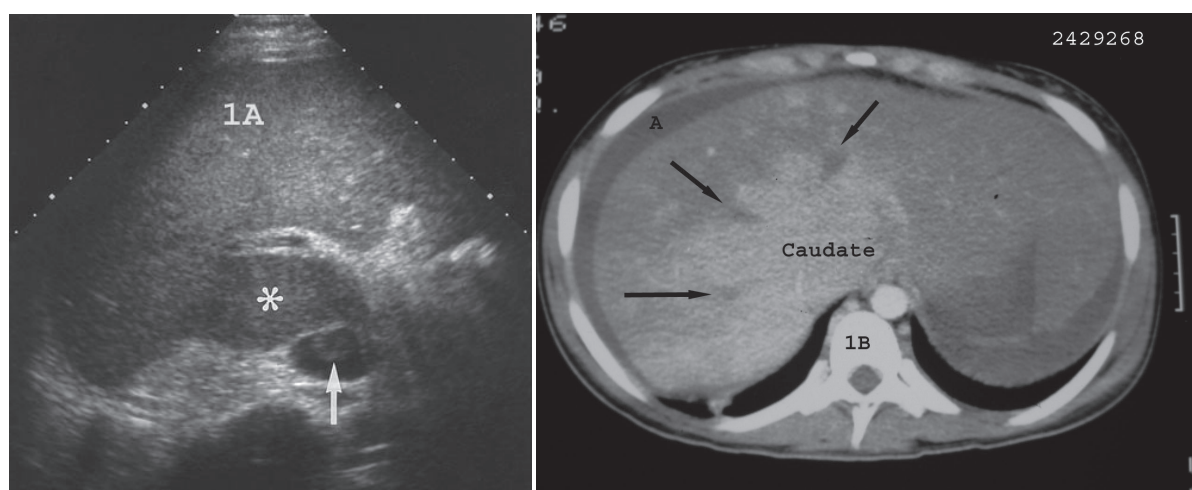


Figure 1 Budd-Chiari Syndrome in a 35-year-old female with a history of thrombocytosis.

A US shows a large thrombus within the inferior vena cava (arrow). Note hypertrophy of the caudate lobe (*).

B CT scan shows caudate lobe hypertrophy with early enhancement of caudate lobe and central part of the liver.

The three hepatic veins are filled with thrombus (arrows). Note the ascites (A) secondary to portal hypertension.

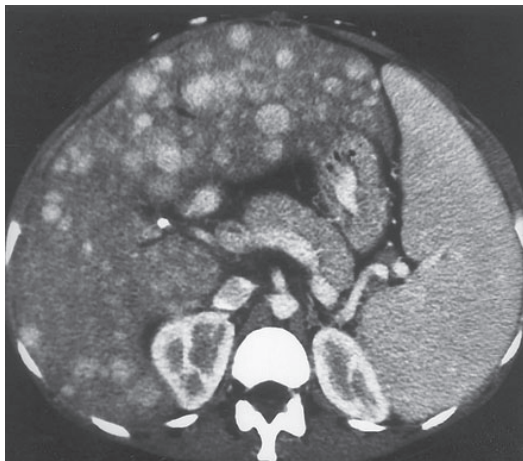


Figure 2 Chronic Budd-Chiari Syndrome in a 42-year-old female with unknown etiologies. Contrast-enhanced CT shows multiple hypervascular regenerating nodules, commonly found in this condition. Splenomegaly is noted secondary to portal hypertension.

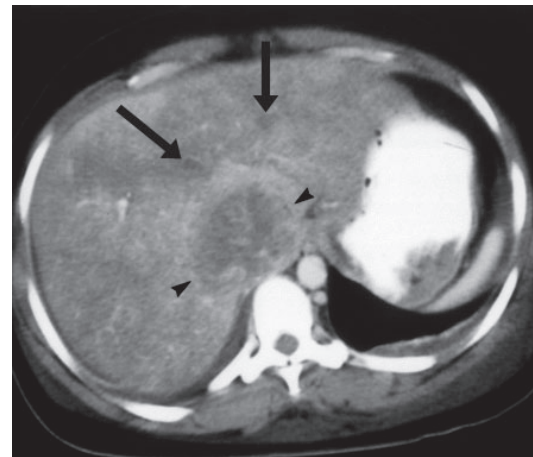


Figure 4 Budd-Chiari Syndrome secondary to IVC leiomyosarcoma. CT scan shows a large tumor occupying and expanding the lumen of IVC (arrow heads). The thrombi within left and right hepatic veins are also noted (arrows).

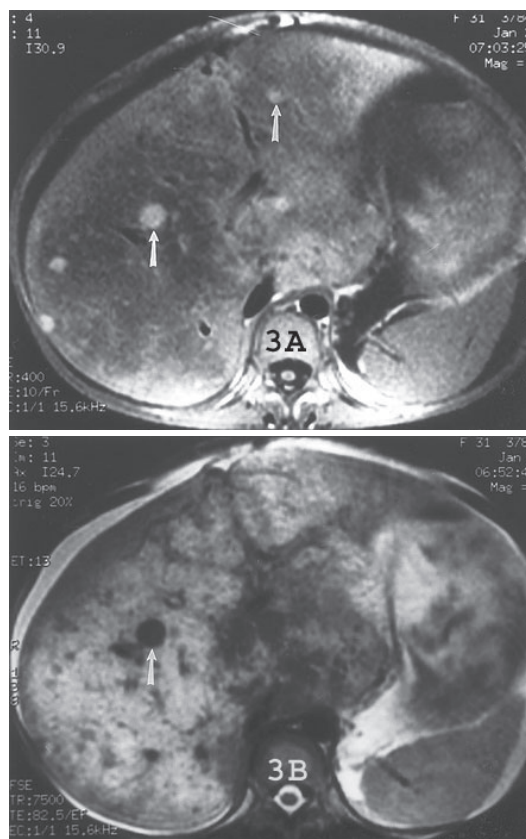


Figure 3 Regenerating nodules in chronic Budd-Chiari Syndrome, possibly secondary to antiphospholipid syndrome.
A Gadolinium enhanced T1W MRI shows multiple hypervascular nodules.
B T2W MRI reveals that these nodules are of low signal intensities.

appearance of regenerative nodules may mimic hepatocellular carcinoma. However, multiple nodules (more than 10) of smaller than 4 cm showing hyperdense at plain CT are suggestive of benignity⁽⁵⁾.

3. MRI shows inhomogeneous, mottled liver and similar findings to CT scan.

4. Inferior venocavography shows spider web pattern of collateral vessels around the hepatic venous obstruction. Web or diaphragm can also be demonstrated.

Some pathological processes that may give similar imaging findings to BCS include cirrhosis, severe right-sided heart failure (cardiac cirrhosis) and infiltrative hepatocellular carcinoma.

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