

## Clinical decision making in acute Budd Chiari Syndrome

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### Dear Editor.

A 51-year-old male firefighter with a remote history of deep vein thrombosis following ankle fracture was hospitalized at our center for a 2 week history of jaundice, ascites and right upper quadrant abdominal pain. His bloodwork on admission was significant for ALT and AST 100 times the upper limit of normal, bilirubin 30 mg/dL (normal 3-22), hemoglobin 16.8 mg/dL (normal 13.5-16), platelets  $570 \times 10^9$  cells/mL (normal  $150-400 \times 10^9$ ) and INR 1.5 (normal 0.9-1.1). CT scan revealed caudate lobe hypertrophy, compression of the inferior vena cava (ICV) and opacification of the hepatic veins (Figure 1), consistent with a diagnosis of Budd Chiari Syndrome (BCS). Further work-up revealed Janus kinase 2 mutation, confirming polycythemia rubra vera.

He was started on anticoagulation and diuretics, but had no improvement after 5 days. A hepatic venogram showed total obstruction of the 3 hepatic veins, and severe IVC stenosis; thus, the portal vein could not be accessed to perform a transjugular intrahepatic portosystemic shunt (TIPS). Liver biopsy showed zone 3 occlusion and cell dropout, consistent with venous outflow obstruction. A surgical side-to-side portocaval shunt was performed on day 7 of admission. Due to marked hypertrophy of the caudate lobe, the distance from the portal vein to IVC was excessive at 4.1 cm, necessitating a prosthetic shunt. However, postoperatively, he had worsening liver function with new onset of progressive hepatic

encephalopathy and acute renal injury. He was therefore promptly listed for transplantation for fulminant liver failure.

After 6 days, he underwent ABO incompatible liver transplantation (ILT); a veno-venous bypass was required before the hepatectomy. The patient received plasmapheresis and rituximab following the ILT. He gradually recovered, and remains stable at 12 months post-ILT, maintained on coumadin and tacrolimus.

BCS is a rare condition defined as hepatic venous outflow obstruction originating anywhere from the small hepatic veins to the right atrium. The classic presentation is a triad of abdominal pain, ascites and hepatomegaly, but there is a wide spectrum of clinical manifestations from asymptomatic to acute liver failure. Most patients with BCS have an underlying thrombophilic disorder, namely myeloproliferative syndrome.<sup>1</sup>

Five-year survival rates from diagnosis of Budd Chiari range from 50 to 80%.<sup>1</sup> Age, response of ascites to diuretics, Child score and serum creatinine

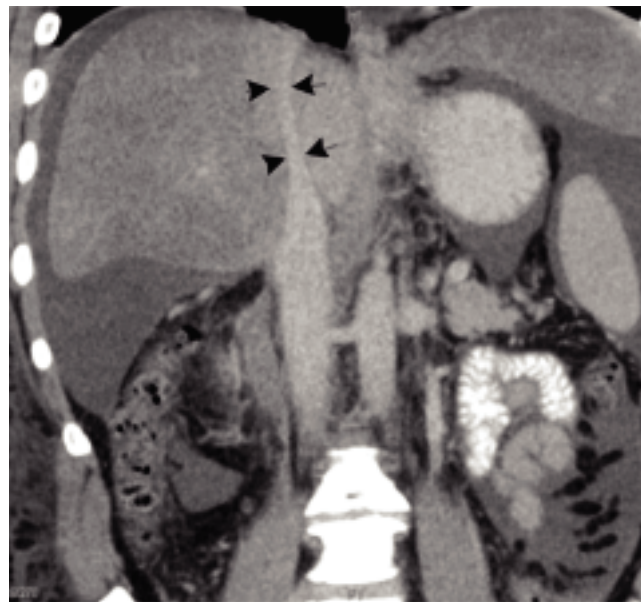


Figure 1. Coronal CT image of acute Budd Chiari Syndrome.

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independently predict survival.<sup>2</sup> Half of patients with BCS will improve with anticoagulation and diuretics alone.<sup>1</sup> The remaining half, like our patient, require invasive treatment, including TIPS, surgical portosystemic shunt placement, liver transplantation, or other procedures such as thrombolysis or percutaneous hepatic vein balloon angioplasty. Patients with significant liver dysfunction may have improved survival with surgical portosystemic shunt placement compared to medical management alone; for this reason, a surgical shunt was attempted in our patient.<sup>3</sup> In experienced centers, TIPS placement is a less invasive, and therefore a preferred alternative to surgical portosystemic shunts when they are technically feasible.<sup>4</sup> In the era of the model for end-stage liver disease, patients transplanted for BCS have comparable survival rates for patients transplanted for alcoholic liver disease or hepatitis C.<sup>5</sup> Rituximab, an anti-CD20 monoclonal antibody, is routinely given for ILT to minimize the risk of humoral rejection. Recipients of ABO-incompatible hepatic grafts have similar survival to recipients of ABO-compatible grafts beyond 3 months.<sup>6</sup>

Our case highlights the complex and multidisciplinary clinical decision making involved in the management of severe acute BCS. Patients who fail to improve with TIPS or surgical shunt require expeditious liver transplantation with ABO-incompatible grafts if need be.

## ABBREVIATIONS

- **ALT.** Alanine transaminase.
- **AST.** Aspartate aminotransferase.
- **CT.** Computed tomography.
- **ILT.** Incompatible liver transplantation.
- **IVC.** Inferior vena cava.
- **JAK2.** Janus kinase 2.
- **MELD.** Model for end-stage liver disease.
- **TIPS.** Transjugular intrahepatic portosystemic shunt.

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