CASE REPORT

Transjugular Intrahepatic Portosystemic Shunt (TIPSS) for Budd Chiari Syndrome

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We report a four year old boy who presented with liver failure secondary to anti-thrombin III deficiency related Budd Chiari syndrome. He was treated with TIPSS (transjugular intrahepatic porto systemic shunt) which reversed the encephalopathy, normalised the liver function and improved growth, pre-empting the need for a liver transplantation. This is the first reported case of TIPSS in a child with a fulminant presentation of Budd-Chiari Syndrome.

Key words: B*udd chiari Syndrome, Hepatic venous outflow obstruction, Transhepatic portosystemic shunt.*

Budd Chiari syndrome (BCS) or hepatic venous outflow obstruction (HVOO) is a disease complex resulting from obstruction to the hepatic venous outflow at the level of the hepatic veins or the inferior vena cava. If left untreated, it results in fatal portal hypertension, cirrhosis and liver failure. Treatment of HVOO involves establishing the patency of the hepatic venous outflow tract.

With the introduction of newer radiological interventions, HVOO has been safely treated with good outcome in adults. Published literature on radiological therapeutic interventions in pediatric BCS is scarce. We report an unusual presentation of a child with BCS presenting with liver failure in whom a radiological intervention reversed the liver dysfunction.

CASE REPORT

A 4 year male child presented with progressive abdominal distension secondary to ascites of two months duration. He was diagnosed to have BCS on ultrasound doppler, which showed occlusion of all three hepatic veins.

He was brought to our hospital in a comatose state, deeply icteric, acidotic with severe abdominal distension (ascites and hepatosplenomegaly; liver 8cm and spleen 3cm palpable). There was a history of fever of three days, prior to hospitalization. Investigations revealed Hb-9 g/dL WBC-17,600/ mm^3, platelets - 50,800/mm^3, serum creatinine – 0.4mg/dL, total bilirubin- 21mg/dL (direct bilirubin-16 mg/dL), aPTT 70/30 seconds, INR-2.2, albumin 2 g/dL. A thrombophilia profile revealed deficient anti thrombin III (39%; range: 80-120%). Factor V Leiden mutation, Protein C, Protein S, Anti-phospholipid antibody and lupus anticoagulant were within normal range. JAK 2 mutation was absent. The blood and urine cultures were negative. On testing, his mother was also anti-thrombin III deficient (3%).

The patient was first stabilized hemodynamically and given broad spectrum antibiotics and anti-coma measures till he regained consciousness and became afebrile. While he improved clinically, the bilirubin levels kept rising to a maximum of 29 mg/dL. On day 6 of hospitalization, under the cover of fresh frozen plasma and cryoprecipitate, IVC and hepatic venogram was performed, which revealed ostial

blocks of all three hepatic veins. The right branch portal vein was cannulated from the IVC, portal venous pressure measured 28 mm Hg. A 10mm × 6 cm 'Viatorr' stent was placed between the main portal vein and IVC **(*Fig.* 1)**. Post procedure, the venogram showed good flow from portal vein to IVC, and the porto-systemic gradient was 4 mm Hg. The liver size reduced rapidly (to just being palpable at time of discharge) and the ascites, which had been therapeutically tapped dry pre-procedure, did not recur. He was off diuretics immediate post procedure and ascites free on day 10. There were no further episodes of hepatic encephalopathy. The INR was 2.8 and total bilirubin was 28 mg/dL; anticoagulation was thus deferred immediate post-procedure. Follow-up showed normalization of liver function over the next six months. Growth monitoring showed a growth spurt and 'catch-up growth' in both weight and height velocity. Follow-up doppler at 10 days and 6 months post procedure showed normal stent flow and no ascites.

DISCUSSION

Transjugular intrahepatic portosystemic shunt (TIPSS) for hepatic venous outflow obstruction, though commonly used in adults(1,2), is rarely used in children(3-5). There is no report of TIPSS for a fulminant presentation of BCS in a child. TIPSS in a setting of such high levels of bilirubin and INR is usually a contraindication, as it may precipitate hepatic encephalopathy in the setting of cirrhosis. However, in our patient, TIPSS was performed with the aim of reducing the portal hypertension and improving the blood flow to the liver. Since the cause (compromised venous outflow) in this child was treated with TIPSS, he actually improved in terms of hepatomegaly, liver function and encephalopathy. TIPSS also averted the need for a liver transplantation. This is unlike performing a TIPSS in a decompensated cirrhotic patient, where encephalopathy is likely to worsen. However, these interventions have to be used selectively. Our recently published data of this intervention in children with BCS shows it to be safe and successful(6).

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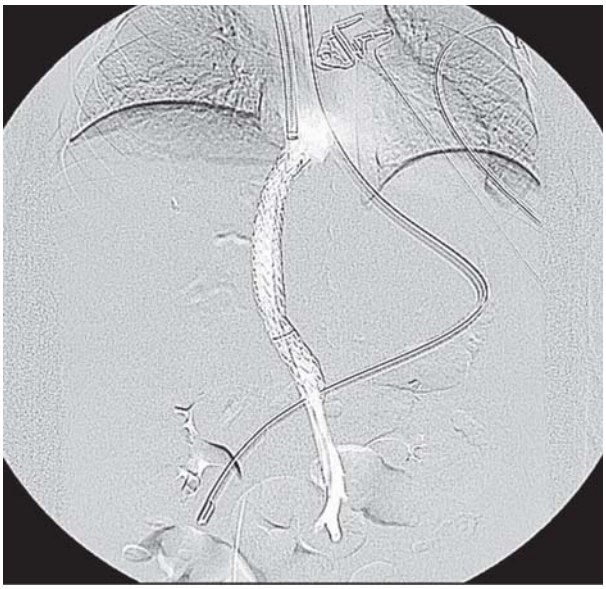

FIG. 1 *Viatorr stent placed between the main portal vein and the inferior vena cava.*

REFERENCES

1. Griffith JF, Mahmoud AE, Cooper S, Elias E, West RJ, Olliff SP. Radiological intervention in Budd Chiari syndrome: techniques and outcome in 18 patients. Clin Radiol 1996; 51: 775-784.
2. Eapen CE, Velissaris D, Hevdtmann M, Gunson B, Olliff S, Elias E. Favourable medium term outcome following hepatic vein recanalisation and/or transjugular intrahepatic portosystemic shunt for Budd Chiari syndrome. Gut 2006; 55: 761-763.
3. Huppert PE, Astfalk W, Brambs HJ, Schweizer P, Schott U, Pereira P, *et al*. Transjugular intrahepatic portosystemic shunt in children. Initial clinical experiences and literature review. Rofo 1998; 168: 595-603.
4. Cauchi J , Oliff S, Baumann U, Mirza D, Kelly D, Hewitson J, *et al*. The Budd-Chiari syndrome in children: the spectrum of management. J Pediatr Surg 2006; 41: 1919 - 1923.
5. Bogdan H, Franchi-Abella S, Aurelie P, Dalila H, Alexis M, Sibert A, *et al*. Budd-Chiari syndrome and essential thrombocythemia in a child: favorable outcome after transjugular intrahepatic portosystemic shunt. J Pediatr Gastroenterol Nutr 2008; 46: 334-337.
6. Nagral A, Hasija RP, Marar S, Nabi F. Budd Chiari syndrome in children: experience with therapeutic radiological intervention. J Pediatr Gastroenterol Nutr 2010; 50: 74-78.