



ORIGINAL RESEARCH PAPER

Anaesthesiology

GENERAL ANESTHESIA IN POST-PARTUM BUDD CHIARI SYNDROME DURING EMERGENCY TRANSJUGULAR INTRAHEPATIC-PORTO-SYSTEMIC SHUNT (TIPS) PROCEDURE – A RARE CASE REPORT

KEY WORDS: Postpartum Budd Chiari Syndrome, TIPS Procedure, Anaesthesia management

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ABSTRACT

Budd-Chiari syndrome (BCS) is associated with hepatic venous outflow obstruction, leading to acute or chronic symptomatology, classically treated with liver transplantation or portosystemic shunts. Prevalence in pregnancy is about 6.7% in world and 7% in India. But BCS in postpartum period is a rare condition and can be fatal & very little knowledge is revealed about anaesthesia management during shunts (TIPS) of such cases. Here we describe anaesthesia considerations during interventional radiological procedure (TIPS) done under general anaesthesia in a primipara female with chronic presentation of Budd-Chiari syndrome in postpartum period.

INTRODUCTION

Budd-Chiari Syndrome (BCS) is defined as obstruction of the hepatic venous outflow located anywhere between small hepatic veins to supra-hepatic inferior vena cava. BCS can occur due to various aetiologies presenting in an acute fulminant form or chronic ascitic presentation. Acute form is treated with surgical shunt or liver transplantation. The transjugular intrahepatic portosystemic shunt (TIPS) is performed to create an alternative venous outflow tract for preventing parenchymal injury from hepatic congestion. There have been reports of anaesthesia management of BCS cases for various surgeries including caesarean sections. There have been reports of successful treatment of the chronic form with TIPS in BCS, but scarce is reported about BCS in postpartum period. Here, we describe anaesthesia management of a case of chronic Budd-Chiari syndrome developed in female in postpartum period, undergoing emergency TIPS procedure [7].

CASE REPORT: A 22 year old primipara was admitted for severe pain in abdomen in emergency. Past History- Normal vaginal delivery 5 months back followed by mild abdominal pain, since 5 months of postdelivery, pain severity gradually increasing. She was investigated and diagnosed of having Budd Chiari Syndrome since then. Oesophageal variceal banding was done 2 months back for episodes of hematemesis and tablet Propranolol 20 mg bid. No history of oral contraceptive pills use.

Clinical examination—thin built, weight-38kg, afebrile, severe pallor, PR- 90/min regular, BP-80/60mmHg, Abdominal examination-enlarged liver with irregular margins, splenomegaly and ascites.

Investigations- severe anaemia (Hb-6.2gm %), slightly elevated liver enzymes, normal blood coagulation profile. Markers for hepatitis A, B, C and E viruses were negative. Contrast enhanced computed tomography-Thrombosis of right hepatic vein. Cirrhotic changes of liver, moderate ascites and splenomegaly. Hepato-Portal Doppler study-suggestive of liver cirrhosis, moderate splenomegaly and perisplenic varices Dilated Hepatic veins showing reversal of flow which confirmed diagnosis of Budd-Chiari Syndrome (fig1).

Emergency Transjugular intrahepatic portosystemic shunt insertion was planned as being minimally invasive procedure to lower the raised portal pressure and provide symptomatic

relief.

Preoperative Preparation- 3units blood transfusion done for anaemia correction, haemoglobin rose to 11.2gm%. General anaesthesia was planned as patient had risk of gastric regurgitation and disrupted respiratory mechanics owing to raised abdominal pressure and expected longer duration of intervention. Controlled ventilation was also useful since a motionless patient and the ability to provide apnoea would aid radiologist to position the shunt.

Consent for general anaesthesia/ICU care was obtained. Intravenous access secured on forearm with two 18G intracath. Extension catheters for IV line attached to prevent stretching by C arm. Monitors (ECG, NIBP, SpO2) attached. Premedication - IV inj. Glycopyrrrolate (5mcg/kg), ondansetron (0.1 mg/kg), midazolam (0.03 mg/kg) Fentanyl (1mcg/kg) was given. Antibiotic inj. Cefotaxim 1gm IV. As the procedure involved injection of contrast, IV inj. Hydrocortisone 100mg and Dexamethasone 8mg prophylactically. Rapid sequence induction with Propofol 100mg and intubated using Succinyl Choline 80mg with 6.5 cuffed Portex ETT. Anaesthesia Maintained with Isoflurane and O2:N2O-50:50, Atracurium.

Intra-operative- Urinary catheterization, judicious fluid administration in order to avoid fluid overload after insertion of shunt, stable hemodynamics and negligible blood loss. Anaesthesia reversed with IV inj. Neostigmine 2mg and Glycopyrrrolate 0.4mg.

Postoperative- Patient was shifted to ICU for observation as there was risk of postprocedural sepsis, cardiac failure, encephalopathy and/or occlusion of shunt. Broad spectrum antibiotics and intravenous heparin 5000 IU QID for the next 24 hours were given.

Post procedure hepato-portal Doppler showed widely patent TIPS shunt with good antegrade flow. The procedure was a success without any postoperative complications. The patient was relieved of her symptoms and discharged home on 5th postoperative day.

DISCUSSION

Budd-Chiari syndrome is a congestive hepatopathy caused by blockage of hepatic veins. Prevalence is 1:1 million in general population. BCS is primary or secondary. If

obstruction is the result of endoluminal venous lesion-like thrombosis, primary BCS is considered. In secondary BCS, cause originates from neighbouring structures like extrinsic compression or tumour invasion [10].

Genetic tendencies for BCS include protein C deficiency, protein S deficiency, Factor V Leiden mutation, hereditary anti-thrombin deficiency and prothrombin mutation G20210A. The use of oral contraceptives is also a risk factor for BCS (particularly high-estrogen-content pills) [1]. Hepatic vein thrombosis has been described both in pregnancy and in immediate postpartum period. Many patients in whom BCS develops in association with the use of oral contraceptives or pregnancy may also have an underlying thrombophilia, either inherited or acquired [11].

The increased portal pressure due to obstruction and sinusoidal dilatation causes increased filtration of vascular fluid leading to ascites and collateral venous flow through alternative veins leading to oesophageal, gastric and rectal varices. Obstruction also causes centrilobular necrosis and peripheral lobule fatty change due to ischemia.

BCS may present in acute or chronic form. Acute onset, present with jaundice, abdominal pain, nausea, vomiting, fatigue, minimal ascites etc. Patients with chronic disease present with cirrhosis and portal hypertension. Our case presented with the chronic form following mostly due to postpartum hepatic vein thrombosis.

Hepato-portal Doppler, with sensitivity and specificity of 85%, is the technique of choice for initial investigation when BCS is suspected [8]. Supportive investigations -contrast enhanced computed tomography, liver biopsy, haemogram, liver function tests and coagulation profile.

A subset of patients having membranous/short segment occlusion of IVC and/or Hepatic Vein (HV) require opening of occluded segment followed by balloon dilatation and stenting. The role of radiological intervention in acute BCS is to thrombolysise involved HV done either by using local/direct pharmacological agents or by mechanical thrombolysis to restore deteriorating liver function. Chronic BCS patients having native HV replaced by intrahepatic collaterals form other subgroup of patients where restoration of normal hepatic venous outflow is not possible. A possible radiological intervention in these patients consists of creating a shunt across portal vein and IVC. With a relatively good midterm outcome, currently TIPS is the preferred intervention for patients having chronic BCS [2]. Since our patient had hepatic vein thrombosis with development of collaterals along with symptoms of cirrhosis and portal hypertension, TIPS was the best option for intervention.

Patients undergoing TIPS are medically complex due to chronic liver disease causing altered multisystem physiology. Patients exhibit hyperdynamic circulation with low-normal arterial pressure due to persistent splanchnic vasodilatation [5]. Cardiac output will increase after TIPS insertion as pooled venous blood returns to the systemic circulation; hence, any degree of heart failure must be assessed before shunt insertion as this is likely to deteriorate with the effective fluid challenge post-procedure [3].

Thrombocytopenia and coagulopathy are common in cirrhotic patients and needs to be corrected before shunt insertion. Cross-matched blood should be requested as patients have often had atypical antibodies due to multiple transfusions in past after repeated variceal haemorrhages. Complexities of remote location anaesthesia should be

considered. The choice of drugs demands consideration of physiological/pharmacokinetic changes seen in chronic liver disease patients [9].

Hemodynamic instability may occur after blood loss due to vessel rupture, so preprocedure correction of anaemia and coagulopathy needed. Increased venous return can precipitate heart failure, which may require medical stabilization followed by diuresis. Haemolytic anaemia may develop between 7 and 14 days post-procedure, due to mechanical shear stress on blood cells after passing through shunt.

There may be TIPS shunt size reduction or occlusion. If iodine-based contrast is used, there is a risk of contrast nephropathy and exacerbation of hepato-renal syndrome if this was present.

There is risk of post-procedure sepsis. Early identification and administration of antibiotics, fluids and vasopressors may be required. Adequate attention to anticoagulation and asepsis was given in our case [4].

Al Falaki et al (1999) stated use of sedation and general anaesthesia during TIPS and complications that developed during and post procedure were mainly due to the pre-existing medical condition and the procedure itself [6]. Adequate stabilization and facilities for advanced monitoring and resuscitation are essential during critical incidents.

CONCLUSION:

Planning for general anaesthesia in remote locations during TIPS and vigilance for postoperative complications during management of postpartum Budd Chiari syndrome is necessary. Preoperative evaluation should be directed towards the best intervention to prevent fulminant hepatic failure and post procedural complications thereby preventing any single maternal mortality.

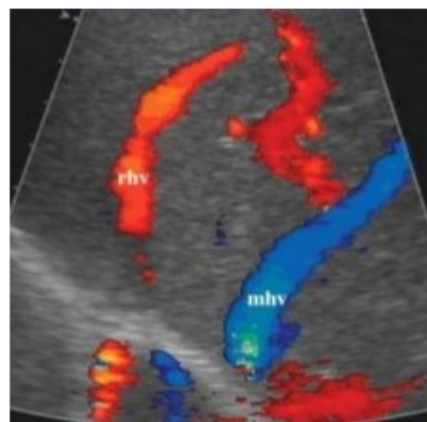


Fig 1- Hepato-Portal Doppler showing reversal of flow in right hepatic vein and normal flow in middle hepatic vein

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