CASE REPORT
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ANAESTHETIC MANAGEMENT OF EXTRAHEPATIC PORTAL VENOUS OBSTRUCTION

Nishith Govil,[1*] Nilesh Chandra,[1] Surjyendu Ghosh,[1]
Mridul Dhar,[1] Intezar Ahmed[2]

ABSTRACT

Extrahepatic portal venous obstruction (EHPVO) is the commonest cause of portal hypertension and variceal bleeding in children. Major concerns are growth retardation, decrease lean body mass and loss of muscle that may lead to postoperative respiratory failure. Upper abdominal surgery with severe pain may compound the problem of pulmonary atelectasis. Poor preoperative nutritional status and anemia may lead to delayed wound healing, delayed ambulation, and respiratory complications. To avoid postoperative respiratory complications and surgical site infection we inserted epidural catheter despite the possibility of intraepidural space bleed due to low platelet counts. The patient had an excellent recovery profile in term of analgesia, decrease postoperative nausea and vomiting due to decrease consumption of opioids, increase sedation-free period, early ambulation and parent's satisfaction. The intraoperative epidural also provides better abdominal muscle relaxation, wider surgical bloodless field and optimal pressure in anastomosis vessels. Regional techniques are avoided in thrombocytopenia, but risk and benefits must be assessed in each case.

Keywords: extrahepatic portal venous obstruction, epidural anesthesia, thrombocytopenia, enhanced recovery
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[1]Departments of Anaesthesiology and
[2]Pediatric Surgery
All India Institution of Medical Science (AIIMS), Rishikesh, India

[*]Correspondence to:
Nishith Govil, Department of Anesthesiology, AIIMS Rishikesh, Virbhadra Rd, Near Barrage, Sturida Colony, Rishikesh, Uttarakhand 249203, India
nishithgovil@rediffmail.com

INTRODUCTION

Extrahepatic portal venous obstruction (EHPVO) characteristically refers to obstruction in the trunk of the portal vein, its branches and even extend to splanchnic veins. Etiology of EHPVO can be infection and/or inflammation (omphalitis, neonatal umbilical sepsis and intraabdominal infection post umbilical catheterization), portal vein injury, developmental anomaly, prothrombotic causes, and idiopathic.[1] Pathogenesis of EHPVO includes presinusoidal blockage of the portal vein, normal intrahepatic venous pressure, raised intrasplenic pressure, hypersplenism and the opening of portosystemic collaterals. Patients present with a massive upper gastrointestinal (GI) bleed, ectopic varicose, and pancytopenia.

Anesthesiologists should be aware of the anesthetic management of EHPVO due to its epidemiology and the challenges it posed during surgery. Variceal bleeding due to EHPVO is not a rare condition. In developing countries, EHPVO is the cause of 35-40% of portal hypertension. In the Indian subcontinent, 20-30% of all variceal bleeds are due to portal vein thrombosis. In children, 70% of all variceal bleeds are due to EHPVO.[2] Major concerns in such cases are hypersplenism, variceal bleed, growth retardation, decreased lean body mass and loss of muscle, that may lead to postoperative respiratory failure.

CASE REPORT

After taking informed written consent from both parents of the patient for possible publication of the case in medical literature, we describe perioperative management of a 10-year-old boy diagnosed with extrahepatic portal vein thrombosis with the aid of epidural anesthesia along with general anesthesia. The patient had a recurrent history of hematemesis for the last 3 years due to esophageal varicose. He already underwent band ligation twice. At the perioperative period, he was on vasopressors and broad-spectrum antibiotics.

The patient complained of progressive abdominal distension associated with pain and recurrent infections. History revealed preterm vaginal delivery, admission to the Neonatal Intensive Care Unit for 7 days and delayed developmental milestones after 5 years of age. Patient operated for distal splenorenal or "Warren" shunts surgery and splenectomy at All India Institution of Medical Science Rishikesh, India.

Preoperatively patient's vitals were stable except for tachycardia (pulse rate 118 x/min), the systemic examination was within normal limits and airway assessment was significant only for grade 4 bilateral enlarged tonsils, prominent upper incisor, and malar eminence. Blood investigations reveal pancytopenia due to splenomegaly: hemoglobin (Hb) 7.5 g/dL, total leucocyte count of 2,180/mm^3

and platelet count 72,000/mm^3. Except for an increased indirect bilirubin level, liver function test, prothrombin time and the international normalized ratio (INR) were within normal limit.

Essential monitoring along with invasive blood pressure monitoring (22G cannula in right radial artery) and induction of anesthesia with propofol, fentanyl, and atracurium was done as per departmental protocol. Atraumatic video-assisted intubation done with 6 mm ID endotracheal tube. Anesthesia was maintained with nitrous oxide and oxygen mixture in a ratio of 1:1 and sevoflurane.

For better perioperative pain management and avoidance of postoperative pulmonary complications, a 19G epidural catheter inserted in T9-T10 intervertebral space in left lateral position. Epidural infusion with 0.25% bupivacaine at 2 mL/hour started after giving bolus dose of 0.25% bupivacaine 10 mL. Porto-renal shunting with splenectomy performed uneventfully in 6 hours. Blood loss was 500 ml (allowable blood loss was estimated at 330 ml) and urine output was adequate (1.1 ml/kg/hour).

Intraoperatively, the patient received two pediatric unit of packed red blood cells (250 ml), fresh frozen plasma, and platelet concentrate. At the end of a surgery, the patient was extubated after complete reversal of anesthesia effects and muscle paralysis. Patient shifted to post anesthesia care unit for monitoring for 24 hours before moving to the ward. Patient's postoperative stay was uneventful and he was then discharged on the eighth postoperative day after documenting good blood flow in the shunt.

DISCUSSION

The diagnosis of portal hypertension is clinically confirmed with features of cirrhosis, supportive hematology (pancytopenia), or diagnostic radiological findings (recanalization of the umbilical vein, or reversal of blood flow in the portal vein). Hepatic venous pressure gradient (HVPG), is the gold standard for assessing portal hypertension and is measured by hepatic venous catheterization. Normal HVPG is up to 5 mmHg and portal hypertension is defined as an HVPG ≥ 6 mm Hg along with clinical manifestations.

EHPVO poses serious challenges for perioperative anesthetic management like hypersplenism, hepatopulmonary syndrome (HPS), portopulmonary hypertension (PPH), portal biliopathy (PB) bone marrow hyperplasia leading to skeletal changes and growth retardation. HPS occurs due to intrapulmonary vascular shunts in liver disease, but HPS has also been seen in the absence of intrinsic liver disease with prehepatic portal obstruction especially in children as a complication of congenital portosystemic shunts, polysplenia syndrome with interruption of the inferior vena cava and absence of portal vein.[3] HPS is diagnosed by hypoxemia, confirmation of intrapulmonary vascular shunts by saline-contrast echocardiography or Tc99m-macroaggregated albumin scintigraphy, and portal hypertension with or without cirrhosis.

PPH is a complication results from the obliteration of the pulmonary artery and pulmonary artery pressure >25 mmHg with normal pulmonary capillary wedge pressure (PCWP) <15 mmHg in the presence of portal hypertension. PPH has been documented in children of extrahepatic portal hypertension due to congenital anomalies of the portal vein.[4] HPS and PPH in milder form can be managed with supplemental oxygen, avoidance of further hypoxia, hypercarbia, and acidosis, though our patients do not have any of the features of HPS and PPH.

PB is a condition that refers to cholangitis and biliary stasis in the extrahepatic and intrahepatic bile ducts due to external compression by the portal cavernoma or collaterals due to long-term portal obstruction. Seen in almost every adult patients of EHPVO but few cases are reported in children also.[5]

Growth retardation due to growth hormone resistance is common in children with EHPVO. Diminished portal blood flow results in decreased insulin delivery to the liver and thereby decreased the production of insulin-like growth factor-1 and insulin-like growth factor binding protein-3. Upper abdominal surgery with severe pain may compound the problem of pulmonary atelectasis but not easily recognizable clinically in the immediate postoperative period. Poor preoperative nutritional status and anemia may lead to delayed wound healing, delayed ambulation, and respiratory complications. Surgical site infection is very common if simple techniques like the adoption of hand hygiene and procedural follow up of aseptic working is not followed.

To avoid postoperative respiratory complications we inserted epidural catheter despite being fully aware of possible complication of intra epidural space bleed due to low platelet counts. 4 hour before surgery, two unit of platelet concentrates transfused and two units transfused after stump of spleen ligated to decrease the risk of bleeding. The patient had an excellent recovery profile in term of analgesia, decrease postoperative nausea and vomiting due to decrease consumption of opioids, increase sedation-free period, early ambulation and parent's satisfaction. The intraoperative epidural also provides better abdominal muscle relaxation, wider surgical bloodless field and optimal pressure in anastomosis vessels.

Regional techniques are avoided in thrombocytopenia, but risk and benefits must be assessed in each case. A number of studies reported no cases of epidural hematoma when epidural given in patients with less than 100,000 platelets mm^{-3}. It is prudent to do Thromboelastography and platelet transfusion before the puncture, although there is no strong evidence to support it.[6] It is important to maintain anesthesia and analgesia depth at the time laryngoscopy and surgical incision to prevent hypertension and tachycardia, which could cause variceal bleeding. An endoscopy is beneficial to treat potential esophageal varicose bleed especially in the presence of thrombocytopenia, and associated anemia.[7]

Massive splenomegaly causes the destruction of the pooled blood cell components leading to thrombocytopenia, leucopenia, and anemia making the child susceptible to recurrent infections, which may be the cause of the grossly enlarged tonsils. Due to multiple transfusions in the past patients may develop atypical antibodies and therefore require extended cross matching.

The airway was challenging in this patient as low platelet count may have led to epistaxis during airway manipulation. The bilaterally enlarged tonsils and prominent upper incisors prompt us to be ready with alternative techniques to secure the airway. EHPVO is also associated with temporomandibular joint ankylosis possibly due to protein C deficiency induced delayed lysis of fibrin leading to ankylosis of the joint.[8] We proceeded with video-assisted intubation as direct visualization avoided any trauma of airway, however, even it is not devoid of complications. Thrombophilia reported as a causative risk factor and involved in the pathogenesis of EHPVO in both adults and children. The most commonly studied thrombophilia risk factors include factor V Leiden mutation. Some studies suggest that the deficiencies in protein C, protein S and Antithrombin in EHPVO may be due to altered hepatic blood flow. Regardless of the cause, splanchnic vasoconstriction and thrombosis leading to intestinal ischemia must be avoided with adequate cardiac output, oxygen delivery and decreasing release of stress hormones.[6] It is recommended that thrombophilia be ruled out in cases of EHPVO and anticoagulation therapy should be started early in the prothrombotic state especially to maintain the patency of shunt.[9]

CONCLUSIONS

In EHPVO though mortality related to variceal bleeding is uncommon, morbidity due to massive splenomegaly with hypersplenism, growth failure, and ectopic varicose is significant. The conventional treatment for EHPVO has been medical management with beta-blockers and sclerotherapy, with or without banding procedures. Portosystemic shunts were the mainstay of surgical management and were reserved for children in whom medical management failed to control bleeding symptoms. Surgery requires meticulous vigilance of recognition and management of the anesthetic concern. Benefits of a successfully managed case of EHPVO include resolution of portal hypertension, correction of the coagulation abnormalities, and improved growth and neurocognitive function associated with EHPVO.

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The authors report no conflicts of interest.

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