Emergency Cesarean Delivery in Primigravida With Portal Hypertension, Esophageal Varices, and Preeclampsia

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The incidence of cirrhosis and advanced portal hypertension during pregnancy is very low, and the literature is scarce with regard to the anesthetic management of a parturient with this coexisting disease. We report the successful perioperative management of a parturient with a history of cirrhosis and portal hypertension with esophageal varices and mild preeclampsia who presented at 38 weeks’ gestation in active labor with a breech presentation requiring emergency cesarean delivery. She required endoscopic esophageal varices banding during the second trimester of pregnancy. After correction of her coagulopathy, she was administered subarachnoid block and cesarean delivery, which was conducted uneventfully. Anesthetic management of these patients depends on understanding and avoiding variceal hemorrhage, encephalopathy, renal failure, and careful fluid and electrolyte management.

Keywords: Anesthesia, cesarean delivery, cirrhosis, portal hypertension, preeclampsia.

Pregnancy with portal hypertension is an uncommon condition. The exact incidence is unknown.1,2 The prevalence of cirrhosis in the reproductive age group is 0.45/1,000 persons, with a maternal mortality rate of 10% to 61%.1,3 Although it is difficult to distinguish the effects of pregnancy from the natural history of cirrhosis, maternal complications have been described in nearly 30% to 50% of pregnancies affected by cirrhosis and portal hypertension, largely as a result of variceal hemorrhage and liver failure.2,4,5 Advanced cirrhosis increases the risk of maternal and fetal morbidity and mortality.1 Pregnant women with portal hypertension have a high variable incidence of fetal wastage ranging from 10% to 66% and also have a spontaneous abortion rate of 20% to 40%.2,4,6 In those pregnancies that do result in live births, the risk of prematurity is significantly increased, with a rate up to 25%.7 Preeclampsia is not more frequent in the presence of portal hypertension syndrome, but such a combination of this comorbidity and the implications for anesthetic management has not been previously reported. We report the perioperative management of a parturient with a history of portal hypertension secondary to hepatic cirrhosis and mild preeclampsia who presented with a breech presentation in active labor necessitating emergency cesarean delivery.

Case Report
A 20-year-old, ASA physical status 3E, 65-kg, 154-cm, parturient presented at 38 weeks with cephalopelvic disproportion and breech presentation in active labor. Because she was in active labor the patient was prepared for an emergency cesarean delivery. Her medical history was significant for pain and fullness in the abdomen for the past 3 years. Pain was mild, dull aching and present in right upper abdomen. A gastroenterologist evaluated the patient and examination revealed massive splenomegaly. Endoscopy revealed grade 4 esophageal varices. A diagnosis of portal hypertension due to hepatic cirrhosis or noncirrhotic portal fibrosis was made. The patient was kept on conservative management and was advised for regular follow-up but she did not report for her regular check-ups.

During this pregnancy her prenatal course was complicated by 2 episodes of hematemesis during the second trimester. Endoscopic evaluation by the gastroenterologist during the second trimester revealed grade 4 esophageal varices at the gastroesophageal junction, which required endoscopic banding on 2 separate occasions under monitored anesthesia care. In addition, during the second trimester, the patient was diagnosed with mild preeclampsia (hypertension, proteinuria, and edema). She was started on propranolol (10 mg, 3 times a day), spironolactone (200 mg daily), and a salt-restricted diet.

Presently, clinical examination revealed a blood pressure of 150/86 mm Hg and bilateral pitting pedal edema. There was no clinical evidence of portosys-
temic encephalopathy. Her respiratory, cardiovascular, and neurological examinations were unremarkable. Airway examination revealed Mallampati class 2, mouth opening of 3 fingers, thyromental distance of 3 fingers, and normal neck movements. On examination of the patient’s back, the presacral area was edematous and spinous processes of the vertebrae could be palpated by applying gentle pressure. Patient was given nothing by mouth for 3 hours. The laboratory results are listed in Table 1. Urinary protein was ++. Ultrasonography revealed a grossly enlarged spleen with no splenic aneurysm, enlarged liver, intra-abdominal ascites, and fetus with footling breech.

After obtaining informed consent and notifying the neonatologist, the primigravida was premedicated with intravenous ranitidine (50 mg) and metoclopramide (10 mg) and transported to the operating room. Vital signs were heart rate, 92/min; blood pressure, 150/88 mm Hg; and oxygen saturation of 96%. Fetal heart rate was 144/min. Two 16-gauge intravenous cannulae were inserted. To correct coagulopathy, 4 U of fresh frozen plasma were transfused. The repeat prothrombin time was 18 s (vs a control of 12 s). Subarachnoid block was given using a 27-gauge Sprotte needle. A dose of 9 mg of 0.5% hyperbaric bupivacaine and 25 μg of fentanyl was injected into the subarachnoid space. The patient was put back in the supine position with left uterine displacement. The heart rate was 78/min and blood pressure was 118/76 mm Hg. After an adequate block was verified (sensory level T5 by pinprick method using a blunt short beveled needle), cesarean delivery commenced.

During the surgery, about 600 mL of ascitic fluid mixed with amniotic fluid was drained. A male infant with Apgar scores of 7 at 1 minute and 9 at 5 minutes was delivered. No oxytocin was administered. Uterine massage was given by the obstetrician and the uterus was well contracted. Estimated blood loss was 400 mL. The urine output was 350 mL intraoperatively. Hemodynamics were maintained with normal saline solution (2 L). During the surgery, the heart rate ranged 68 to 84/min. The systolic blood pressure remained 110 to 130 mm Hg. The oxygen saturation was 98% to 99%. The patient was transported to the intensive care unit after surgery for observation. On arrival, vital signs were heart rate, 78/min; blood pressure, 122/78 mm Hg; and oxygen saturation, 98%. The patient course was uneventful with no further need for transfusion of blood products. The patient remained hemodynamically stable and was shifted to a high dependency unit the next day. This period was uneventful, and the child and mother were discharged on postoperative day 4 and advised follow-up in the gastroenterology and obstetric clinics.

**Discussion**

The concerns in our patient were presence of portal hypertension with esophageal varices and cirrhosis, pre-eclampsia, emergency cesarean delivery, coagulopathy and its correction, need of blood typing and cross match-
ing, need of rapid transfusion, and possibility of post-partum hemorrhage. Also, anesthetic management of a patient with these comorbidities depends on understanding and avoiding variceal hemorrhage, encephalopathy, and renal failure (hepatorenal syndrome) (Table 2).

Maternal complications occur in 30% to 50% of patients with preexisting portal hypertension. These include variceal hemorrhage, hepatic failure, postpartum hemorrhage, rupture of splenic artery aneurysm, rupture of splenorenal shunts, spontaneous bacterial peritonitis, and maternal death. Hypersplenism may cause anemia and its related complications. It also poses an added risk of bleeding due to thrombocytopenia during this type of pregnancy in such patients. The presence of underlying liver disease does not affect the risk of preeclampsia. The adaptation of the mother to the hemodynamic and other changes during the pregnancy have effects on preexisting portal hypertension. Abnormal renal sodium retention often accompanies hepatic disease and contributes to ascites formation. Renal sodium and water retention in pregnancy can contribute to several complications such as pulmonary edema and generalized convulsive seizures. This could create confusion in the management of eclampsia in such patients. Preeclampsia will be differentiated by presence of hypertension with proteinuria after the 20th week of gestation (Table 3). The overall risk of variceal bleeding in pregnant women with portal hypertension is almost 400 times greater than in pregnant women without portal hypertension. Esophageal variceal bleeding has been reported in 18% to 32% of pregnant women with cirrhosis and in up to 50% of those with known portal hypertension and increases to 62% to 78% if there are endoscopically visible varices. Among those with preexisting varices, up to 78% will have gastrointestinal bleeding during pregnancy, with a mortality rate of 18% to 50%. Variceal bleeding during pregnancy is associated with a higher incidence of abortion (29.4%) and perinatal death (33.3%). The factors contributing to bleeding from gastroesophageal varices are not entirely understood but include the degree of portal hypertension (>12 mm Hg) and the size of the varices. Estrogens are believed to make capillaries fragile, and progesterone is responsible for the enlargement of the venous system. Hemodynamic and hormonal changes may be the cause of vascular alterations, which can lead to weakening of the vein wall. During the first trimester, variceal bleed can be precipitated with violent retching associated with morning

### Table 2. Portal Hypertension: Anesthetic Implications

<table>
<thead>
<tr>
<th>Feature</th>
<th>Anesthetic implication</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastroesophageal varices</td>
<td>Variceal rupture</td>
<td>Endoscopy and banding, β-blocker drug therapy, blood pressure monitoring, prepare for possible hemorrhage</td>
</tr>
<tr>
<td>Ascites</td>
<td>Fluid and electrolyte imbalance, spontaneous bacterial peritonitis</td>
<td>Fluid management, monitor electrolytes closely, aldosterone antagonist drug therapy</td>
</tr>
<tr>
<td>Hypersplenism</td>
<td>Pancytopenia, coagulopathy</td>
<td>Blood product transfusion, type and cross matching, arterial line blood pressure monitoring</td>
</tr>
<tr>
<td>Hepatorenal failure</td>
<td>Acute renal failure</td>
<td>May require liver transplant</td>
</tr>
<tr>
<td>Hepatic encephalopathy</td>
<td>Impaired mentation</td>
<td>Avoid precipitating factors, monitor electrolytes and urine output</td>
</tr>
</tbody>
</table>

### Table 3. Characteristics of Liver Disorders of Pregnancy

<table>
<thead>
<tr>
<th>Portal hypertension</th>
<th>Preeclampsia</th>
<th>HELLP syndrome</th>
<th>Acute fatty liver of pregnancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastroesophageal varices</td>
<td>Hypertension</td>
<td>Hemolysis (lactate dehydrogenase level &gt;600 U/L)</td>
<td>Hypoglycemia</td>
</tr>
<tr>
<td>Ascites</td>
<td>Proteinuria, after 20 weeks of gestation</td>
<td>Elevated aspartate aminotransferase level (&gt;70 U/L)</td>
<td>Elevated urate level</td>
</tr>
<tr>
<td>Hypersplenism</td>
<td>Elevated liver enzyme or bilirubin level</td>
<td>Low platelet count (&lt;100,000/mm³)</td>
<td>Leukocytosis</td>
</tr>
<tr>
<td>Hepatorenal failure</td>
<td></td>
<td></td>
<td>Elevated transaminase levels</td>
</tr>
<tr>
<td>Hepatic encephalopathy</td>
<td></td>
<td></td>
<td>Elevated ammonia level</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Renal impairment</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Coagulopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Hepatic encephalopathy</td>
</tr>
</tbody>
</table>

Abbreviation: HELLP, hemolysis, elevated liver enzymes, and low platelet count.
sickness.7 The presence of reflux esophagitis that commonly occurs in pregnancy can also increase the chances of variceal bleeding. Variceal bleeding is most common during the second trimester, occurring in approximately 20% to 45% of women with portal hypertension when maternal blood volume is maximally expanded and the larger fetus causes increased compression of the inferior vena cava and collateral vasculature.2 This results in a larger percentage of venous return being routed through the azygos vein. Variceal bleeding may also occur during labor, leading to 20% maternal death.11 This happens due to straining and repetitive Valsalva maneuvers with pushing during the second stage of labor that can result in increased intra-abdominal pressure. Contraction of the diaphragm can elevate the portal pressure 3-fold, which can precipitate variceal bleeding.6,12 As a result, elective cesarean delivery is usually recommended. If vaginal delivery is attempted, the use of vacuum- or forceps-assisted delivery is recommended under epidural anesthesia.6,12

Therapeutic measures that have been advocated to reduce the incidence of variceal bleeding include endoscopic variceal banding, sclerotherapy, portosystemic shunting, esophageal transaction, and lowering of portal pressure using β-blockers and vasodilators.1,3,6 Various methods have been used to treat variceal hemorrhage such as the balloon tamponade (Sengstaken-Blakemore or Minnesota tube), shunt procedures, endoscopic sclerotherapy, and banding.6 At present, surgery and transjugular intrahepatic portosystemic shunt (TIPS) are only recommended as rescue therapies in patients with failure in endoscopic or pharmacologic treatments. Although TIPS placement is generally contraindicated during pregnancy because of the risk of radiation exposure to the fetus, it may be an appropriate rescue therapy for failed attempts to control variceal bleeding with band ligation or sclerotherapy.2 Surgical shunt procedures are generally performed only in the setting of a life-threatening hemorrhage that is refractory to medical and endoscopic treatments.2 Drug-induced diuresis with aldosterone antagonist is an effective treatment for removing ascitic fluid. Treatment for active bleeding, including invasive monitoring, large gauge canulae, cross matched blood, fresh frozen plasma, platelets, a Sengstaken-Blakemore tube, and vasopressin infusion must be available.6 The use of vasopressin and its analogues to control active bleed should be avoided if at all possible due to the risk of inducing labor.

Our patient was managed by endoscopic ligation of esophageal varices, and portal pressure was controlled with propranolol and spironolactone. Infants born to mothers on continuous therapy with propranolol should be considered at risk for developing complications such as small placenta, intrauterine growth retardation, fetal depression at birth, and postnatal hypoglycemia and bradycardia.7 Therefore, such infants need to be monitored for at least 24 to 48 hours for any such symptoms. These drugs have been found to be compatible with breast feeding as these are not concentrated in breast milk. Diuretics (eg, furosemide, hydrochlorothiazide, spironolactone) may reduce milk volume and thus adequate hydration needs to maintained in the perioperative period. Up to 24% of pregnant patients with cirrhosis will also experience hepatic decompensation, which can lead to rapid clinical deterioration. This has been described in all stages of pregnancy but often occurs after episodes of variceal bleeding.2 Hepatic encephalopathy is due to rising ammonia levels in the blood and may be precipitated by hypotension, gastrointestinal bleeding, hypoxia, hypokalemia, hyponatremia, alkalosis, sedatives, diuretics, or by the stress of surgery.13 Renal failure, or hepatorenal failure, is characterized by worsening azotemia, hyponatremia, progressive oliguria, and hypotension. This syndrome may be precipitated by severe gastrointestinal bleeding or altered renal hemodynamics as in severe hypotension, and it may also occur without an obvious cause.14 The anesthetic considerations would include avoidance of hypotension or hypertension, straining, hypoxia, use of sedative drugs, or electrolyte imbalances. The deranged hepatic function may lead to abnormal coagulopathy and metabolism of various drugs required in the perioperative period in such patients. Acute and chronic parenchymal liver disease results in impaired synthesis of clotting factors I, II, V, VII, and X. The presence of thrombocytopenia may be caused not only by hypersplenism but also concurrently by the occurrence of HELLP (hemolysis, elevated liver enzymes, and low platelet count) syndrome. This mandates perioperative monitoring of the coagulation profile and correction by transfusing suitable blood products such as platelet concentrate and fresh frozen plasma. The deranged coagulation profile may also precipitate postpartum hemorrhage and thus suitable availability of blood products; a need for rapid transfusion should be kept in mind while managing such patients. Surgical and obstetric patients with microvascular bleeding usually require platelet transfusion if the platelet count is less than 100,000/mm3 and rarely require therapy if it is greater than 50,000/mm3. With intermediate platelet counts, the determination should be based on the patient’s risk for more significant bleeding. Although the minimum platelet count at which the risk of hemorrhagic complications from neuraxial techniques becomes prohibitively high remains undefined, similar ranges for platelet counts have been suggested to avoid any neurologic complications.14,15 The drugs with active metabolites such as morphine need to be used cautiously in these patients. Similarly, other sedative drugs such as benzodiazepines also require cautious administration because of risk of encephalopathy. At times the occurrence of hepatorenal syndrome may
lead not only to abnormal metabolism of the drugs but also its excretion.

The perinatal management of a pregnant portal hypertensive mother has to be individualized according to the status of the liver disease and size of esophageal varices. The specific complications contributing to perinatal mortality include variceal hemorrhage, progressive liver failure, preeclampsia, severe anemia, and worsening renal function. Though the fetal outcome was good in our patient, it is prudent to emphasize that perinatal mortality caused by variceal hemorrhage can be effectively reduced in patients who are diagnosed to have portal hypertension before pregnancy by adoption of effective means for variceal eradication.

Anesthetic management of patients with portal hypertension secondary to hepatic cirrhosis and mild preeclampsia requiring emergency cesarean delivery for breech presentation can be challenging. It requires the anesthesiologist, hepatologist, neonatologist, and obstetrician to work as a team in a closely coordinated manner. Another area of controversy in pregnant patients with portal hypertension is how to approach delivery. Generally, vaginal delivery with early analgesics for the mother assisted by an extraction device should be preferred to cesarean delivery, which must be reserved for obstetrical indications. Women with cirrhosis generally tolerate laparotomy poorly; therefore, the option for cesarean delivery should be availed with care and caution.

We preferred regional to general anesthesia as the patient had a very high risk of aspiration because of a full stomach, term pregnancy, and further increase in intraabdominal pressure due to ascites and splenomegaly. The hypertensive response to intubation and straining during extubation would increase the risk of variceal hemorrhage. The mother would receive a number of sedatives and narcotics and there are chances of hypoxia, which can be avoided in regional anesthesia. One more advantage of regional anesthesia is that it minimizes the requirements of oxytocics, while inhalational anesthetics cause tocolysis and increase oxytocin consumption. However, the performance of regional anesthesia in these patients is not without problems either. In our case, the problems faced were deranged hepatic function leading to decreased clotting factors synthesized in liver, with an international normalized ratio (INR) level of 1.5, and thrombocytopenia due to hypersplenism.

Since the patient had increased INR level, 4 U of fresh frozen plasma were transfused in our patient preoperatively. In portal hypertension with esophageal varices, the already engorged extradal veins may be engorged further. There is increased risk of bloody tap when sitting, so we preferred a left lateral decubitus position with a slight head down tilt, which corrected immediately after administration of the intrathecal drug. We preloaded the patient adequately to avoid hypotension resulting from sympatholysis because of spinal anesthesia and during decompression of the abdomen and drainage of the ascitic fluid. Fentanyl was added as adjuvant to bupivacaine to provide prolonged analgesia with less hypotension. Oxygen was supplemented by face mask to avoid hypoxemia. Fluid administration was used as the first line of therapy to treat hypotension rather than vasoconstrictors such as ephedrine because they cause a sudden rise in blood pressure that may precipitate variceal bleed.

Use of oxytocics should be minimized and ergometrine should be avoided, as they may cause forceful uterine contractions squeezing large blood volumes to the inferior vena cava and then to esophageal varices. The gentle massage of the uterus by the surgeons can enhance the contractility of the uterus. Pregnant patients with portal hypertension have a high likelihood of postpartum hemorrhage, ranging from 7% to 26%, as a result of coagulopathy and thrombocytopenia because of hypersplenism. The coagulopathy is mainly caused by a deficiency of factors V and VII, with occasional true prothrombin deficiency. Rupture of a splenic artery aneurysm is another potentially life-threatening condition during pregnancy in a patient with portal hypertension and carries a considerably high fetal, as well as maternal, mortality. So, special efforts must be made to look for a splenic artery aneurysm during ultrasound and Doppler examination in such patients.

We conclude that the patient with portal hypertension and pregnancy induced hypertension requires not only intensive care monitoring in the perioperative period but also possible use of a tertiary care center with a multidisciplinary team (ie, anesthesiologist, obstetrician, hepatologist, neonatologist) for the most comprehensive care for this rare presenting case. During perioperative management, the changes associated with portal hypertension, pregnancy induced hypertension, and pregnancy itself needs to be cautiously interpreted for an uneventful anesthetic management. The esophageal varices should be prophylactically banded and measures should be taken to prevent rupture of esophageal varices. The presence of coagulopathy and its correction, need of blood typing and cross matching, need of rapid transfusion, and possibility of postpartum hemorrhage should be kept in mind while managing such patients.

REFERENCES


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