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Portal Hypertension in Pregnancy

Dr. Sohini Burman

Junior Resident

Abstract: Even though pregnancy is rare with cirrhosis and advanced liver disease, but it may co-exist in the setting of non-cirrhotic portal hypertension as liver function is preserved but whenever encountered together is a complex clinical dilemma. Pregnancy in a patient with portal hypertension presents a special challenge to the obstetrician as so-called physiological hemodynamic changes associated with pregnancy, needed for meeting demands of the growing foetus, worsen the portal hypertension thereby putting mother at risk of potentially life-threatening complications like variceal haemorrhage. Risks of variceal bleed and hepatic decompensation increase many fold during pregnancy. Optimal management revolves round managing the portal hypertension and its complications. Thus management of such cases requires multi-speciality approach involving obstetricians experienced in dealing with high risk cases, hepatologists, anaesthetists and neonatologists. With advancement in medical field, pregnancy is not contra-indicated in these women, as was previously believed. This article focuses on the different aspects of pregnancy with portal hypertension with special emphasis on specific cause wise treatment options to decrease the variceal bleed and hepatic decompensation. Based on extensive review of literature, management from pre-conceptional period to postpartum is outlined in order to have optimal maternal and perinatal outcomes.

Keywords: Portal Hypertension, cirrhosis, variceal bleed, endoscopic variceal ligation, postpartum haemorrhage

1. Introduction

Pregnancy is a normal physiological state characterized by numerous hemodynamic changes. These hemodynamic changes, although necessary for a normal pregnancy, pose a special problem in the presence of portal hypertension. In developed countries, cirrhosis of liver is the most common cause of portal hypertension. In developing countries, other causes like extrahepatic portal vein obstruction contribute significantly to noncirrhotic portal hypertension (NCPH). In patients with noncirrhotic portal hypertension (NCPH) the hepatic synthetic functions are relatively well preserved and so is fertility. Pregnancy in a patient with portal hypertension is a unique problem that needs specialized care to prevent potentially life-threatening complications gastrointestinal haemorrhage. It is therefore important to understand the effect of pregnancy on portal hypertension and vice versa so that untoward incidents like foetal morbidity, mortality and gastrointestinal haemorrhage can be avoided.

2. Case Report

A 23 years old patient, primigravida with 33 weeks gestational age, came to labour room with c/o 2 to 3 episodes of hematemesis with severe anaemia. Patient's USG was s/o changes of portal hypertension, gross splenomegaly and gross ascites and a single live intrauterine gestation corresponding to the gestational age. In the meantime, pt underwent endoscopy with report s/o large oesophageal varices and mild portal hypertensive gastropathy. In the same sitting, Endoscopic variceal banding was done. In the present pregnancy, there was no history of bleeding tendencies, jaundice. liver function tests, coagulation profile was normal. Markers of infective hepatitis were negative. Pancytopenia was present. Patient was allowed for spontaneous progress of labour, started on antenatal corticosteroids, Tab propranolol 20 mg two times a day (after consultation with the gastroenterologists). High risk consent was taken in view of prematurity, risk of variceal bleeding, pancytopenia, need of blood and blood products. Adequate amount of blood and plasma were arranged. The second stage of labour was cut short by instrumental delivery. Patient delivered a 1.8 kg female baby that cried immediately after birth and was shifted to NICU for observation. Third stage of labour was managed actively. Postpartum recovery was uneventful.

3. Discussion

In pregnant women, alcoholic cirrhosis is uncommon while viral or autoimmune related cirrhosis is more common in developing countries. The non-cirrhotic causes of portal hypertension include extra-hepatic portal vein obstruction, non-cirrhotic portal fibrosis, portal vein thrombosis, Budd-Chiari syndrome, infection or congenital hepatic fibrosis. The pregnant woman has a 20-27% chance of oesophageal bleed which increases markedly in case she has demonstrable varices. Active variceal bleeding may occur at all stages of the pregnancy through second and third trimester, risk of variceal bleeding being the maximum in 2nd stage of labour. Prognosis of portal hypertension during pregnancy depends upon the underlying cause and the extent of derangement of liver function. Maternal prognosis is better with noncirrhotic portal hypertension and poor with cirrhosis of the liver. Maternal mortality ranges between 2% and 18%; being maximum with cirrhosis. The causes of death are generally hematemesis, hepatic coma or postpartum haemorrhage. The mother is also at risk of developing severe anaemia, splenic artery aneurysm rupture, ascites, spontaneous bacterial peritonitis. Perinatal mortality ranges between 11% and 18%, mainly due to preterm delivery or intrauterine growth restriction (IUGR). Extensive and detailed pre-conceptional counselling, evaluation and antenatal and perinatal monitoring is needed in patients with portal hypertension with or without cirrhosis planning pregnancy. Varices should be tackled prior to planning a pregnancy, endoscopic variceal ligation is the preferred therapy and nonresponders should be offered surgery in the form of shunt procedure or splenectomy. Drugs should be reviewed for adverse effects on the foetus and alternative safe drugs to be changed, and also dose needs to be tailored.

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Selective beta-blockers can be continued as their benefits outweigh risks. Medical termination of pregnancy may be advised in case of severe hepatic decompensation like ascites, encephalopathy, and liver failure. Antenatal management requires strict maternal and foetal monitoring by a multi-disciplinary team. The routine antenatal management should be given with special watch out for the potential complications like variceal bleed and liver failure. Liver function and haematological assessment should be done weekly, foetal growth needs to be monitored vigilantly and effects of the drugs need to be watched. Close maternal and foetal monitoring is recommended two weekly. Since variceal bleed is the single important complication linked with poor pregnancy outcome, the basic aim is to prevent it. This can be done by assessment and tackling the varices prior to planning a pregnancy. In cases of unplanned pregnancy, proper risk assessment should be performed. Endoscopy is the gold standard to assess the risk of bleeding in patients with oesophageal varices. Upper gastrointestinal endoscopy is safe during pregnancy, the main risk being foetal hypoxia due to sedation or positioning. Current American Association for the Study of Liver Disease (AASLD) recommendations include screening endoscopy in the second trimester as that is the time of maximum increase in the portal pressure. The treatment options in presence of oesophageal varices are both medical and surgical. Nonselective beta blockers used to reduce portal pressure also reduce the risk of first bleed by half but the principal risk of using them in pregnancy is foetal growth restriction and foetal bradycardia. Endoscopic variceal ligation of the large varices can also be done during pregnancy to prevent variceal bleeding. Current literature recommends EVL for acute oesophageal variceal bleed, although, endoscopic sclerotherapy may be used if banding is technically difficult. Pregnancy can be allowed to go to term if the disease is well compensated. Early termination of pregnancy may be warranted in case of any obstetrical indication or progressive liver failure. In case of planned termination before 34 weeks, antenatal corticosteroids can be administered for foetal lung maturity. There are no recommendations as to the preferred mode of delivery vaginal versus caesarean section in patients with portal hypertension. The management during labour needs to be individualized depending on cause of portal hypertension and the disease status. Adequate amount of blood and plasma should be arranged and measures for balloon tamponade for the variceal haemorrhage must be handy. Second stage of labour may be shortened prophylactically to avoid overstraining by the mother. The third stage should be managed actively. Postpartum haemorrhage should be anticipated and managed vigilantly.

4. Conclusion

Pregnancy with portal hypertension may be associated with adverse maternal and perinatal outcome especially in cirrhotic portal hypertension but pregnancy is not contraindicated as was once believed. The management of pregnancy with portal hypertension should only be done at tertiary care centres by a multidisciplinary team with backup facilities for intensive care and blood transfusion.

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