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Management of Non-Cirrhotic Portal Hypertension during Pregnancy: A Review

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Review Article

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Abstract

Non-Cirrhotic Portal Hypertension is a common cause of Portal Hypertension in India. Pregnancy in these patients poses a challenge to the treating physician owing to hemodynamic changes it brings about. This review discusses the challenges faced and its management strategy during pregnancy.

Methods: A retrospective analysis of 7 pregnancies in 6 patients from 2013 to 2023 at a tertiary teaching hospital was done.

Results: 6 pregnancies in 6 patients were studied. 5 patients were Extra-Hepatic Portal Venous Obstruction (EHPVO) and 1 patient Non-Cirrhotic Portal Fibrosis (NCPF). 5 patients were diagnosed to have Non-Cirrhotic Portal Hypertension (NCPH) at the time of pregnancy. 5 patients had normal vaginal delivery, while one required emergency caesarean section. There was one neonatal death, however no maternal mortality.

Conclusion: Management of Non-Cirrhotic Portal Hypertension patients with pregnancy in a tertiary care with a multidisciplinary approach result in a good outcome of pregnancy.

Keywords: Portal Hypertension; Outcome of Pregnancy

Introduction

Non-Cirrhotic Portal Hypertension (NCPH) includes Extrahepatic Portal Venous obstruction and Non-Cirrhotic Portal Fibrosis (NCPF). Pregnancy in these patients poses a challenge. Though patients of NCPH do not have severe hepatic dysfunction, they have definite risk of thrombosis of collaterals (Porto-Systemic shunt) with consequent hematemesis and or melena. In absence of guidelines for management of pregnancy in these groups of patients, the management approach is not standardised. The present study reviews the current literature with regards to the management of pregnancy in patients of NCPH and its outcome.

Methods

This is a retrospective study of patients of Non-Cirrhotic Portal Hypertension (NCPH) undergoing pregnancy from 2013 to 2023 at a tertiary teaching hospital. Prior Institutional Ethics Committee permission was obtained. 6 pregnancies in 6 women with Non-Cirrhotic Portal Hypertension occurred

during the study period were analysed. The parameters analysed were 1) Age 2) Obstetric details 3) Details of Non-Cirrhotic Portal Hypertension: a) duration of the disease b) treatment received 4) present ongoing treatment obstetric history 5) Laboratory investigations 6) Maternal outcome and 7) Perinatal outcomes. Laboratory investigations included complete blood count, liver function test, including coagulation profile, viral markers, renal function test and Prothrombotic work up. Diagnosis of Non-Cirrhotic Portal Hypertension was confirmed based on Complete blood count, UGI endoscopy and Portal Venous Doppler USG. All the patients were managed along with the GI Medicine and Gynaecology and Obstetrics team. Data was analysed using SPSS software.

Results

A total of 6 pregnancies in 6 patients were studied (Table 1). Their mean age was 25 years (23-29 years). Four patients were primigravida (85.71%). On Doppler USG five patients were diagnosed to have Extra-Hepatic Portal Venous Obstruction (EHPVO) and one patient (patient number 3) to have Non-Cirrhotic Portal Fibrosis (NCPF). Five patients (4 have Extra Hepatic Portal Venous Obstruction & 1 Non-Cirrhotic Portal Fibrosis) were diagnosed to have NCPH at the time of pregnancy. Four patients were asymptomatic for NCPH. One patient was diagnosed to have Extra Hepatic Portal Venous Obstruction, 14 years back at age of 17 years. She had hematemesis on presentation and was treated conservatively with Endoscopic Variceal band ligation. She had delivered one healthy child in past 3 years back through normal uneventful vaginal delivery. At present she was not on active treatment for her Non-Cirrhotic Portal Hypertension. The Liver Profile and Prothrombotic work up of all the patients were normal. None of the patient had symptomatic hypersplenism and portal biliopathy, ascites or jaundice. No patient had pancytopenia (decrease in all three peripheral blood cell line), though one patient (patient number 3) of Non-Cirrhotic Portal Fibrosis had decreased platelet count. This patient had Grade II varices on UGI endoscopy and was banded prophylactically. This patient on antenatal scan was found to have restriction of foetal growth. She had severe per-vaginal bleeding at 32 weeks. She was found to have placenta previa, and was taken up for emergency Caesarean section. Intraoperatively she was transfused 2 units of Single donor Platelet (SDP). She delivered small for gestational age baby, which succumbed on day 3. She also had postpartum haemorrhage, which was controlled successfully with oxytocin drip. Rest all the five deliveries were normal vaginal delivery at full term. There was no maternal mortality. The average weight of the baby was 2.4 kg +/-0.3kg. There was one neonatal death on day three. Except for this baby no other baby required admission in the Neonatal Intensive Care Unit.

Patient	Age	Cause Of Pht	Obstetric History	Tiem Since Diagnosis	Variceal Bleeding	Ascites	Pancytopenia
1	23	EHPVO	G1P0A0	Time of Pregnancy	NO	NO	NO
2	31	EHPVO	G2P1A0	14 years	YES	NO	NO
3	27	NCPF	G1P0A0	Time of Pregnancy	NO	NO	NO, but decreased Platelet count
4	24	EHPVO	G1P0A0	Time of Pregnancy	NO	NO	NO
5	22	EHPVO	G1P0A0	Time of Pregnancy	NO	NO	NO
6	26	EHPVO	G1P0A0	Time of Pregnancy	NO	NO	NO

Table: Patient Profile.

Discussion

Portal hypertension is increased pressure within the portal venous system. It is determined by the increased portal pressure gradient (the difference in pressures between the portal venous pressure and the pressure within the inferior vena cava or the hepatic vein. A pressure gradient of 6 mmHg suggests presence of portal hypertension [1]. The common cause of PHT is Cirrhosis and NCPH. NCPH comprises of EHPVO and NCPF. Hepatic dysfunction in patients of NCPH is comparatively not severe as in Cirrhotic. Nevertheless, pregnancy in these patients poses a risk. In absence of guidelines and not much literature evidence on treatment strategy, its management is challenging. The biggest challenge is the physiological haemodynamic change that occurs following pregnancy, which is superadded on the haemodynamic changes of PHT which increases the risk of bleeding. In patients of PHT following increased Portal Venous pressure there occurs dilatation of portosystemic venous anastomoses. Following pregnancy there occurs increase in plasma volume, which in turn increases stroke volume and heart rate with consequent increase in cardiac output. Up to 30% increase in cardiac output is believed to occur in the first stage of labour. In second stage of labour

the cardiac output further increases up to 50% [2]. The increase in cardiac output, with the effect of progesterone leads to reduction in the vascular resistance. This leads to hyperdynamic circulation and increased flow in collaterals. This increases the risk of variceal bleeding manifesting as either hematemesis or melena [3]. The reported incidence of variceal bleeding is 8-25% [4,5]. This low incidence could be attributed to increased awareness of the risk and preventive and prophylactic measures (starting of NSBB-non selective beta blocker) adopted early during pregnancy. In our study 1 patient (16,66%) presented with hematemesis at 14 weeks of gestation. As per literature reports though variceal bleeding can occur at any stage of pregnancy, it is most likely to occur in second trimester [6]. Though patients of NCPH tolerate variceal bleeding better, severe variceal haemorrhage is ominous. It portends abortion, pre term labour and at times both maternal and neonatal mortality [7]. Hence patients with presence of significant varices need to be subjected to EBVL and NSBB and monitored periodically during pregnancy. In our study two patients were subjected to EBVL at the onset of pregnancy. One patient, known case of EHPVO with oesophageal varices had presented with hematemesis. Another patient was a case of NCPF, who had thrombocytopenia, was detected to have varices and endoscopic variceal band ligation was done. While the first patient had uneventful successful pregnancy, the second patient had complications and neonatal death. Besides variceal haemorrhage, decompensation in form of transient ascites too can occur rarely, with reported incidence being 0.8-10% [8]. Thrombocytopenia due to splenomegaly can occur [9]. Clinically significant thrombocytopenia may necessitate intrapartum infusion of platelet [10]. In our study, one patient of Non-Cirrhotic Portal Fibrosis had thrombocytopenia. She was transfused 2 units of Single Donor platelet.

Maternal prognosis in patients of Non-Cirrhotic Portal Hypertension (NCPH) is comparatively better than in patients with cirrhosis with similar obstetric outcome as that in general population [9]. However, conditions like HELLP syndrome and disseminated intravascular coagulation leading to Post partum Haemorrhage are known to cause maternal mortality [5]. As far as perinatal outcomes are concerned, there are conflicting reports in literature. While Sumana et al reported no preterm delivery or stillbirth, Mandal et al reported a high incidence of preterm delivery, low birth weight, and stillbirth [11,12]. Besides neonatal mortality rate of 16% has been reported by various authors [5,10-13]. In our study one patient of Non-Cirrhotic Portal Fibrosis who had thrombocytopenia and varices which were banded, was detected to have restricted foetal growth on antenatal scan. She had severe per-vaginal bleeding at 32 weeks, was found to have placenta previa, and was taken up for emergency Caesarean section. She delivered small for gestational age baby, which succumbed on day three. However, rest of all the patients had normal vaginal delivery at term with normal baby with their average weight being 2.4 kg +/-0.3kg. This is in accordance with the contemporary published reports where all deliveries are conducted per-vaginally with Caesarean section being done only for obstetric indications [12].

The Key to successful outcome to pregnancy in patients with Non-Cirrhotic Portal Hypertension is to manage these patients as a team including GI Surgeons, GI Medicine and Gynaecology and Obstetrics. All these patients need to be evaluated for Non-Cirrhotic Portal Hypertension with Portal Venous Doppler, UGI endoscopy for varices, Liver function test, Renal function test, and Prothrombotic work up. Endoscopic band ligation should be done in patients detected to have oesophageal varices. They need to be frequently monitored at regular interval.

Conclusion

Pregnancy in patients with Non-Cirrhotic Portal Hypertension calls for a multidisciplinary approach. They should be managed at a tertiary medical care centre.

Statement

The said study has been performed with the approval of an appropriate ethics committee and with appropriate participants' informed consent in compliance with the Helsinki Declaration.

Conflict of Statement

The authors have no conflicts of interest to declare.

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