THE JOURNAL OF OBSTETRICS AND GYNECOLOGY OF INDIA

The Journal of Obstetrics and Gynecology of India (November–December 2018) 68(6):447–451
https://doi.org/10.1007/s13224-017-1016-1

ORIGINAL ARTICLE

A Review of Outcomes in Pregnant Women with Portal Hypertension

Anisha Ramniklal Gala[1] · Tarakeswari Surapaneni[1] · Nuzhat Aziz[1] · Sailaja Devi Kallur[1]

Received: 4 April 2017 / Accepted: 23 May 2017 / Published online: 5 June 2017

About the Author

Dr. Anisha Ramniklal Gala is working in Fernandez Hospital since a decade, runs high-risk pregnancy clinic and prepregnancy counseling clinic. She has completed her fellowship in Maternal Medicine from NTR University of Heath Sciences. She has keen interest in teaching and training students. She has served as faculty for workshops conducted by Fernandez Hospital such as Obstetric Emergencies and Life Support Skills, and Fetal Monitoring. She has presented various papers and posters and won awards for posters such as ''Twins—growth profile, HIV in Pregnancy''. She has won a second prize at ''World Obstetric Anaesthesia Congress'' for poster—''Disease Specific Maternal Mortality—Liver Disorders lead the list!'' She was the Principal Investigator for International Observational E-Registry on the use of Dilapan-S.

Abstract

Background The course of pregnancy in a woman with portal hypertension is a difficult one as it is associated with complications like variceal bleeding, splenic artery rupture and coagulopathy. All these pose a threat to a woman's life. Although this condition is rare, every obstetrician should have a high index of suspicion when an antenatal mother presents with splenomegaly, thrombocytopenia or hematemesis. Hence, we aimed to review maternal and fetal outcomes in pregnant women with portal hypertension.

Methods In a retrospective observational study, 41 women and 47 pregnancies were evaluated, from January 2000–December 2015 at Fernandez Hospital, a tertiary referral perinatal center. Maternal outcomes studied were variceal bleed during pregnancy, surgical procedures, morbidity and

Dr. Anisha Ramniklal Gala is consultant obstetrician at Fernandez Hospital, Hyderabad. She has done her fellowship in maternal medicine. Apart from maternal medicine, she has keen interest in teaching and training DNB students. Dr. Tarakeswari S is Head of the Department, Obstetric Medicine at Fernandez Hospital, and her special interest lies in prepregnancy counseling for women with medical disorders. Dr. Nuzhat Aziz is Head of Obstetrics at Fernandez Hospital. Her specializations are Labour Ward Management and Thyroid Disorders in pregnancy. Dr. Sailaja Devi K is consultant obstetrician at Fernandez Hospital and has special interest in maternal medicine.

✉ Anisha Ramniklal Gala
anishagala@gmail.com

[1] Department of Obstetric Medicine, Fernandez Hospital, Unit 2, 3-6-282, opp: Old MLA Quarters, Hyderguda, Hyderabad, Telangana 500029, India

mortality. Neonatal variables were gestational age at delivery, birth weight and morbidities.

Results Mean maternal age was 26.4 years. Average gestational age at delivery was 36.5 weeks. Mean birth weight was 2507.5 g. There were three maternal deaths out of 47 deliveries, the cause of death was massive variceal bleed in one, the second one was due to cardiac arrest on MRI table, and the third death was due to splenic hilar vessel bleed. There was one stillbirth, and no neonatal deaths.

Conclusion A multidisciplinary approach is essential to improve perinatal outcomes in pregnancy complicated by portal hypertension. Surgical measures to reduce portal venous pressure done before pregnancy or beta blockers during pregnancy might help reduce sudden variceal bleeds.

Keywords Portal hypertension · Variceal bleed · Pregnancy · Outcomes

Introduction

Portal hypertension in pregnancy is a rare and potentially lethal condition. Pregnancy is a possibility as long as liver function is well preserved although fertility is compromised in women with cirrhotic portal hypertension [1–3]. Pregnancy is associated with profound hemodynamic changes such as increase in blood volume, increased cardiac output and decrease in systemic vascular resistance [2, 3]. These hemodynamic changes may contribute to rapid deterioration of the underlying portal hypertension [2–5]. There is an increased risk of variceal hemorrhage in pregnancy due to increased portal pressure resulting from increased intra-abdominal pressure, vena caval compression and Valsalva maneuver in labor [3, 4].

Maternal mortality rates in women with portal hypertension range from 2 to 18% [1]. Uncontrolled variceal bleeding, often severe and unpredictable, is a major cause for mortality [1–8]. Other causes of maternal death include splenic artery aneurysm rupture, coagulopathy and liver failure [1–11]. Hematemesis is reported in 20–30% of women with cirrhosis [1]. A mortality rate of 50–60% has been reported in the presence of hematemesis [1]. Perinatal mortality rates may also be as high as 11–18% and are primarily due to preterm birth or fetal growth restriction [1–11].

Aims

The aim of the study was to review maternal and fetal outcomes in pregnant women with portal hypertension managed during a fifteen-year period ending in 2015, and to invite discussion on the development of formal evidence-based guidelines to standardize the safe management of these women in future.

Methods

A retrospective review of pregnancies with portal hypertension was carried out to determine maternal and perinatal outcomes during the fifteen-year period 2000–2015. These women were managed at a tertiary maternal and perinatal care center at Hyderabad, India which conducts approximately 8000 deliveries annually. All pregnant women who book for care at this hospital receive a preliminary assessment using a standardized antenatal care protocol that includes a detailed demographic history, current and past obstetric, medical and surgical history and gestational age, specific ultrasound examination for prenatal genetic screening, structural anomalies and fetal well-being. Any abnormality revealed by this basic assessment is followed up, using appropriate disease specific guidelines of the institute. Guidelines and protocols used at the institute are based on published international standards and have been adapted to suit the needs of the local population.

For review, the records of women with a diagnosis of portal hypertension were extracted from electronic database. Those who did not deliver at the hospital were excluded from the review.

Relevant maternal details such as age and parity were noted. A note was made of the causal factors where possible, and if there were any cirrhotic changes in the liver. Information related to the baseline evaluation of the disease with investigations such as ultrasound, endoscopy and liver function tests was recorded together with interventions to reduce variceal bleeds such as use of beta blockers, sclerotherapy, banding and shunt surgery. In cases where the women suffered complications as a result of portal hypertension or had other preexisting morbidities, these were also recorded. Pregnancy outcomes such as gestational age at delivery, mode of delivery, mean birth weight and whether the outcome was a stillbirth, were noted. Neonatal morbidities such as hypoglycemia, jaundice and bradycardia, were also recorded.

Results

Table 1 summarizes our findings related to these women. There were 41 mothers with portal hypertension who delivered 47 babies during the review period. All the pregnancies were singleton. The total number of deliveries at the hospital for the same period was 76,923. All 41 mothers were referred to the hospital as known cases of portal hypertension or with signs and symptoms such as

Table 1 Characteristics of the 47 pregnancies in 41 women with portal hypertension

Characteristic	*N* (%)
Mean age in years (SD)	26.48 (SD 3.87)
Primigravida	22 (46.8%)
Cirrhotic portal hypertension	18 (38.2%)
Non-cirrhotic portal hypertension	23 (48.9%)
Prepregnancy diagnosis of portal hypertension	30 (63.8%)
Normal liver function tests	43 (91.4%)
Coagulopathy	2 (4.25%)
Platelet count <100,000/mm^3	29 (61.7%)
Variceal bleed	4 (8.5%)
Splenomegaly	47 (100%)
Sclerotherapy	4 (8.5%)
Shunt Surgery	5 (10.6%)
Beta blockers	23 (48.9%)
Variceal banding	7 (14.8%)

Table 2 Interventions for reducing variceal bleed and risk of hematemesis

Intervention	Total	Hematemesis
Beta blocker	23	1 (4.34%)
Sclerotherapy	4	0
Variceal banding	7	1 (14.28%)[a]
No intervention	18	3 (16.66%)

[a] The same woman was on beta blocker and had banding

thrombocytopenia which warranted further investigation. The mean age of the women with portal hypertension was 26.48 (SD = 2.85) years (range 22.00–38.00 years). Twenty-two (46.80%) women were primigravida. Seventeen (36.17%) of the 47 pregnancies were complicated by cirrhotic portal hypertension. Splenomegaly was present in all 47 pregnancies, and hematemesis was present in three (6.38%) of the 47 pregnancies. Endoscopy was performed in 38 (80.85%) pregnancies where the women were booked in the antenatal period, the rest having presented in labor. Esophageal varices of grade 1 severity were present in nine pregnancies (19.14%), and of grade 2 and 3 in 16 pregnancies (34.04%). Interventions for decreasing the risk of variceal bleed are presented in Table 2. Women who did not have any intervention were found to have higher incidence of hematemesis. Preexisting medical conditions seen were Type 2 diabetes in one (2.12%), severe anemia in one (2.12%) and hypothyroidism in three (6.38%) of 47 pregnancies. Maternal complications of pregnancy which occurred were preeclampsia in three (6.38%), gestational diabetes in two (4.25%), acute renal failure in one (2.12%) and premature rupture of membranes in one pregnancy (2.12%).

There were three maternal deaths in this group. The first of these deaths occurred in a woman who was referred late at 36 weeks of gestation. It was at this stage that she was diagnosed to have cirrhotic portal hypertension with abnormal liver function tests, coagulopathy, anuria and acute renal failure. She underwent a cesarean section and required ventilatory support, but subsequently died on the ninth postoperative day following a massive variceal bleed. The second maternal death was of a woman who was referred for evaluation at 20 weeks of gestation with a diagnosis of portal hypertension secondary to portal vein thrombus. She was undergoing an MRI when she suffered a cardiac arrest and could not be resuscitated. The cause of death remains unknown as consent for an autopsy was not given. The third mortality was of a mother referred in labor who suffered a splenic hilar vessel bleed. She had been diagnosed with portal hypertension eight-year prior to her death, but was not on regular follow-up.

Perinatal details of the 47 pregnancies are presented in Table 3. There were no neonatal deaths. There was however one stillbirth resulting in a perinatal mortality rate of 21.27/1000 (1/47 × 1000) births.

Discussion

The diagnosis of portal hypertension in routine antenatal women requires a high index of suspicion. This case series had a good perinatal outcome but had a high maternal mortality rate of 6.38%. Majority of these mothers had a confirmed diagnosis before pregnancy. In this series, non-cirrhotic portal hypertension was more common than cirrhotic disease, probably due to low fertility rates reported in cirrhotic women.

The diagnostic markers which alerted to the high index of suspicion for diagnosis of portal hypertension were

Table 3 Perinatal details of the 47 pregnancies in 41 women with portal hypertension

Characteristic	*N* = 47
Mean (SD) gestational age at delivery in weeks	36.5 (SD 1.66)
Cesarean sections	33 (70.2%)
Stillbirth	1 (Stillbirth Rate = 21.2/1000 births)
Mean (SD) birth weight in g	2507.5 (SD = 498.4)
Small for gestational age babies	8 (17.02%)
Neonatal bradycardia	Nil
Neonatal jaundice	11 (23.4%)
Neonatal respiratory distress	1 (2.12%)
Neonatal hypoglycemia	1 (2.12%)

splenomegaly (100%) and thrombocytopenia (61.7%). By contrast, liver function tests were poor markers of portal hypertension and were found to be abnormal in only four out of 47 pregnancies. It is our view that the presence of splenomegaly and low platelet count should trigger a request for an abdominal ultrasound which is the key to confirm the diagnosis of portal hypertension. The current trend at many clinics in India of not including a basic complete hematological profile through a full blood count or a complete physical examination to make a note of any splenomegaly as part of the first antenatal assessment may be delaying confirmation of the diagnosis and impacting on the pregnancy outcomes.

Pregnancy with portal hypertension has a very high maternal morbidity and mortality, and these women should be referred for delivery at a tertiary care center [1–11]. In this series, mortality was higher for those mothers with hematemesis (one death out of four) as has also been observed by other authors [12, 13]. Furthermore, the incidence of hematemesis was 8.5% in this series, a proportion which falls within the range of 6–33% reported in previous reviews [1, 12, 13]. The incidence of hematemesis was 16.66% (3/18) in women who had no intervention to reduce portal pressures, while hematemesis was seen in 4.34% in women with beta blockers, 14.28% in women who had variceal banding and it was not seen in women who had shunt surgery. The findings of this study suggest that it would be prudent to evaluate all women for the presence of esophageal varices and to offer beta blockers and surgical intervention to avoid variceal bleed. Emergency tamponade measures like a Sengstaken–Blakemore tube should be available along with blood and blood products facilities. Drugs such as octreotide (Pregnancy Category B) should be made available in the emergency crash cart to treat acute variceal bleeding [1, 12, 13].

One of the maternal deaths was due to ruptured splenic artery aneurysm. Splenic artery aneurysm rupture has been reported to occur in 2.6% of all pregnancies with cirrhosis with a high maternal mortality rate of 70% and fetal mortality rate of 90%. Considering the fact that this is associated with high mortality rates, clinicians must consider ruptured splenic artery aneurysm if a woman presents with acute abdomen or hypotensive shock that is unexplainable or there is a sudden maternal collapse [6, 7, 14–16].

There were no neonatal deaths or major neonatal morbidities suggesting that portal hypertension in pregnancy may not be associated with a substantially higher risk of adverse neonatal outcomes. One baby was stillborn and weighed 2.7 kg at term in a woman with non-cirrhotic portal hypertension. The cause of stillbirth was unexplained.

This review suggests a need for a registry to be launched for the purpose of gathering data and information on portal hypertension in pregnancy. The data may help in assessing the effectiveness of some interventions. Such evidence would form the basis of standardized evidence-based guidelines which would enhance the safe management of women with this rare condition and improve their chances of survival.

Conclusion

Although rare, pregnancy with portal hypertension is a potentially life-threatening condition. Uncertainties persist about the optimal management of these women. It is nevertheless evident that their management by a multidisciplinary team at a tertiary center will improve outcomes. Evidence-based guidelines for the management of this condition would further improve the likelihood of uniformly high-quality care across all maternity units.

Compliance with Ethical Standards

Conflict of Interest Anisha Ramniklal Gala, Tarakeswari Surapaneni, Nuzhat Aziz and Sailaja Devi Kallur declare that they have no conflict of interest.

Human and Animal Rights This research does not involve animals.

Informed Consent Informed consent regarding use of data for medical studies/research at the time of admission and antenatal booking is taken from all mothers at Fernandez Hospital. This is an observational study.

References

1. Sandhu GS, Shakti V, Sanjan D. Pregnancy with portal hypertension. J Obstet Gynecol India. 2007;57(3):261–2.
2. Lee NM, Brady CW. Liver disease in pregnancy. World J Gastroenterol. 2009;15(8):897–906.
3. Hay JE. Liver disease in pregnancy. Hepatology. 2008;47(3):1067–76.
4. Joshi D, James A, Quaglia A, et al. Liver disease in pregnancy. The Lancet. 2010;375(9714):594–605.
5. Boregowda G, Shehata HA. Gastrointestinal and liver disease in pregnancy. Best Pract Res Clin Obstet Gynaecol. 2013;27(6):835–53.
6. Aggarwal N, Negi N, Aggarwal A, et al. Pregnancy with portal hypertension. J Clin Exp Hepatol. 2014;4(2):163–71.
7. Tan J, Surti B, Saab S. Pregnancy and cirrhosis. Liver Transpl. 2008;14(8):1081–91.
8. Aggarwal N, Chopra S, Raveendran A, et al. Extra hepatic portal vein obstruction and pregnancy outcome: largest reported experience. J Obstet Gynaecol Res. 2011;37(6):575–80.
9. Shaheen AA, Myers RP. The outcomes of pregnancy in patients with cirrhosis: a population-based study. Liver Int. 2010;30(2):275–83.
10. Mandal D, Dattaray C, Sarkar R, et al. Is pregnancy safe with extrahepatic portal vein obstruction? An analysis. Singap Med J. 2012;53(10):676.
11. Rasheed SM, Monem A, Allam M, et al. Prognosis and determinants of pregnancy outcome among patients with post-hepatitis liver cirrhosis. Int J Gynecol Obstet. 2013;121(3):247–51.

12. Nicolas C, Ferrand E, d'Alteroche L, et al. Pregnancies after transjugular intrahepatic portosystemic shunt for noncirrhotic portal hypertension. Eur J Gastroenterol Hepatol. 2014;26(4):488–90.
13. Lodato F, Cappelli A, Montagnani M, et al. Transjugular intrahepatic portosystemic shunt: a case report of rescue management of unrestrainable variceal bleeding in a pregnant woman. Dig Liver Dis. 2008;40(5):387–90.
14. Crocker A, Girling J, Cotzias C. Massive bleeding in pregnancy from ruptured oesophageal varices complicating portal hypertension: a cautionary tale. Obstet Med. 2011;4(4):169–70.
15. Sadat U, Dar O, Walsh S, et al. Splenic artery aneurysms in pregnancy—a systematic review. Int J Surg. 2008;6(3):261–5.
16. Ha JF, Phillips M, Faulkner K. Splenic artery aneurysm rupture in pregnancy. Eur J Obstet Gynecol Reprod Biol. 2009;146(2):133–7.