

Eisenmenger's Syndrome in Pregnancy

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To the Editor,

Although heart disease approximately complicates 1% of all pregnancies, pregnancy complicated by Eisenmenger physiology having the diagnosis of its condition only after the acute episode of gastroenteritis is rarely seen as indicated in the following case of a 22 year primigravida under regular antenatal checkup from 8-32 weeks period of gestation.

The diagnosis was probably missed as there was no medical history suggestive of heart disease such as chest pain, shortness of breath, palpitation or syncope through childhood to pregnancy until at 34⁺6 weeks when she had episode of acute gastroenteritis to seek admission. At that time, uterine fundal height was corresponding with the gestational age while the abdomen was relaxed. Therefore she was treated with i/v fluid and antibiotics following which vomiting and diarrhea stopped with the return to normal vitals on the second day, therefore i/v fluid was stopped morning. Unfortunately by the afternoon on the same day, patient complained of sudden onset of chest discomfort which worsened by the evening with the rapid development of cyanosis, respiratory rate of 22/min, elevated JVP, pulse rate of 130 bpm, blood pressure 140/100 and SPO2 of 60%, loud P2 and grade IV systolic murmur heard on the precordium with right ventricular lift and crepitations at lung bases.

ECG demonstrated right axis deviation with sinus tachycardia and T inversion in C1-5. Her Echocardiogram revealed marked right atrial and ventricular enlargement, moderate to severe TR, PR and severe PAH with estimated PASP of 120 mm of Hg, and ASD of 2 cm with right to left shunt. The degree of cyanosis increased and SPO2 dropped up to 45%.

As our patient was in right heart failure and had tricuspid regurgitation, we kept her in the left lateral position with 30° head up tilt to keep the right heart pressure lower by decreasing the preload and to maintain uteroplacental circulation.

Quick obstetric palpation verified 2 mild contractions lasting for 10 seconds with engagement of head and fetal tachycardia of 180 bpm corresponding to USG findings. She became more restless on account of labour progression to cervical dilation to 3 cm with 60% cervical effacement and head at – 2 stations.

Taking into consideration of her severe hypoxaemia, respiratory distress and newly diagnosed pulmonary hypertension patient was immediately taken for emergency LSCS. The outcome was alive male baby with Apgar score 3/10, 7/10, 7/10.

We chose not to use oxytocin in the form of a bolus injection because of its direct vasodilatory effect that decreases SVR with a compensatory increase in heart rate and right to left shunt. So uterine massage followed by slow oxytocin infusion was practiced.

Her intra operative period was uneventful with rise of SPO2 from 60 to 99 %. Uterus was well contracted with blood loss of 300 ml. Fluid was restricted throughout. Patient was put on ventilator. After 2 hours of surgery her condition deteriorated, SPO2 dropped down, had sudden onset of per vaginal bleeding. Cardiopulmonary resuscitation was done but mother could not be revived while the baby is alive and well without any congenital cardiac anomaly.

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Discussion

Eisenmenger syndrome is one of the few cardiac conditions for which pregnancy is considered absolutely contraindicated because of high maternal and fetal mortality up to 30-50% and 75%.¹ This is because of the presence of shunt reversal ie left to right shunt, bidirectional shunting progressive pulmonary hypertension. During pregnancy further decrease in the systemic vascular resistance and the degree of right to left shunting decreases pulmonary perfusion that results in hypoxemia thus deteriorating maternal and fetal condition.² Further systemic hypotension as was caused by volume loss on account of diarrhea lead to decrease in right ventricular filling pressure in the presence of fixed pulmonary hypertension such that right heart pressure was insufficient to perfuse the pulmonary arterial bed leading to sudden profound hypoxemia and maternal death. Although the pathophysiology of this conditions is unknown and death usually involves a rebound worsening of pulmonary hypertension.

Therefore, minimization of pulmonary vascular resistance secondary to hypercarbia, hypoxia, acidosis and stress pain is the main goal.

Side by side avoiding haemodynamic changes that might increase the right to left shunt, claimed to increase severity of hypoxaemia is important. Elective caesarean delivery using propofol mixed with ketamine for anaesthesia induction that reduces pulmonary and systemic vascular resistance is in favour.^{1,3} Pulmonary and systemic vascular resistance can be well taken care by epidural anaesthesia, pulmonary artery catheterization during the intrapartum period, avoidance of central hypoxaemia by the use of inhaled nitrous oxide, a pulmonary vasodilator. Medications like sildenafil and

L-arginine, latter which is converted into nitric oxide, are effective in decreasing PVR.⁴ Prostacycline and its analogue such as i/v epoprostenol also have been used. Bosentan, an endothelin receptor antagonist also have been used and it decreases PVR by 25%.¹

In conclusion although pregnancy should be discouraged in Eisenmenger syndrome, careful and meticulous perioperative planning is essential for a successful outcome. The cardiac output should be maintained and SVR must not be allowed to fall.

References

1. Geohas C, McLaughlin VV. Successful management of pregnancy in a patient with Eisenmenger's syndrome with Epoprostenol. *Chest* 2003; 124: 1170-1173 DOI 10.1378/chest.124.3.1170 <http://chestjournal.chestpubs.org/content/124/3/1170.full.html>
2. Makaryus AN, Forouzesh A, Johnson M. Pregnancy in the patient with Eisenmenger's syndrome. *Mount Sinai Journal of Medicine* 2006; 73(7):1033-1036.
3. Pandey R, Garg R, Nath MP, Rajan S, Punj J, Darlong V, Chandalekha. Eisenmenger's syndrome in pregnancy: Use of proseal laryngeal mask airway (PLMA) and epidural analgesia for elective cesarean section. *Acta Anaesthesiol Taiwan* 2009; 47(4): 204-207
4. Lust Km, Boots RJ, Dooris M, Wilson J. Mangement of Labour in Eisenmenger's syndrome with inhaled nitric oxide during pregnancy. *Am J Obstet Gynaecol* 1999; 180:64-67.