ANAESTHETIC MANAGEMENT OF CASE OF EINSENMENGER SYNDROME POSTED FOR EMERGENCY CAESAREAN SECTION

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ABSTRACT

Eisenmenger's syndrome is a cyanotic congenital heart disease that includes pulmonary hypertension with reversed shunt associated with septal defects or patent ductus arteriosus. The decreased systemic vascular resistance associated with pregnancy increases the degree of right to left shunting, thereby carrying substantial risk to both the mother and the foetus. The maternal mortality rate of pregnancy in the presence of Eisenmenger's syndrome is reported to be as high as 30–70%. We present a case of a 31‑year‑old tertigravida with Eisenmenger's syndrome who gave birth at 34 weeks of gestation via caesarean section to a live female baby under general anaesthesia. We describe the anaesthetic management for caesarean section in a patient with Eisenmenger's syndrome. Although pregnancy should be discouraged in women with Eisenmenger's syndrome, it can be successful.

KEYWORDS: Eisenmenger's syndrome, ductus arteriosus, anaesthesia, foetus.

INTRODUCTION

Cardiac disease complicates 1% to 4% of all pregnancies. Concern for significant cardiac instability arises when there is an inability of the heart to meet the dynamic physiologic changes required during pregnancy, parturition and puerperium. Whatever the cause, pulmonary hypertension carries a very poor prognosis during pregnancy; maternal mortality is 30% to 50% during labour and in the peripartum period.[1] Pulmonary hypertension with congenital heart disease is seen in large systemic to pulmonary communications like ventricular septal defect, patent ductus arteriosus. If this is allowed to progress, it leads to irreversible changes in the vessel wall with reversal of shunt & this condition is known as Eisenmenger syndrome.[2,3,4] Hence, patients with eisenmenger syndrome are advised to avoid pregnancy and to consider termination of pregnancy, if it occurs. In some cases the patients wish to become pregnant and refuse therapeutic abortion. Management of pregnancy, labour and delivery in women with Eisenmenger syndrome is a challenge to the whole health care team requiring multi-disciplinary approach & meticulous preparation. We report successful management of such a case with general anaesthesia.

CASE REPORT

A thirty-one year old tertigravida with history of congenital cardiac disease was admitted to our hospital in active labour with 34 weeks of pregnancy. The patient was diagnosed as a case of congenital heart disease with mild ventricular septal defect at the age of 9years & was on the following medications:- Tab.Lasilactone 50mg twice daily & Tab. Sildenafil 50mg half tablet twice daily. She was advised surgery at the age of 9 years. Inspite of strict medical advice to avoid pregnancy she became pregnant for the 1st time at the age of 28 years but the pregnancy was terminated at 6 weeks of amenorrhoea. At the age of 30 years she became pregnant for the 2nd time. This time she gave birth to a still born child at 30 weeks of amenorrhoea. In this third pregnancy she was asymptomatic until 30 weeks of amenorrhoea, then she developed shortness of breath, palpitation and cough. Symptomatic treatment was given. She was advised to terminate the pregnancy but she got discharged against medical advice. She didn't turn up for further antenatal visits. As pregnancy progressed, dyspnoea which was present on exertion worsened & she had dyspnoea at rest. She again presented to labour room with labour pain at 34 weeks of pregnancy. Her 2D ECHO report done 1 month ago revealed large VSD of 18mm, severe PAH with right to

left shunt and severe tricuspid regurgitation (TR) & moderate pulmonary stenosis. Her right ventricular systolic pressure (RVSP) was 75 mm Hg. PR Gmax was 73mmHg & TR Gmax was 125mm Hg. On examination, the patient was moderately built with height of 148cm and weight 51 Kgs, lying in left lateral position with pulse rate of 98 per minute, blood pressure of 100/70 mm Hg, respiratory rate 24 per minute; oxygen saturation was 82% without oxygen & 94% with oxygen flow of 6 liters per minute by face mask. Patient had NYHA grade IV dyspnoea. Patient was afebrile. On examination, there was cyanosis, grade III clubbing and respiratory auscultation no added sounds were found & bilateral air entry was present. On auscultation soft 2/6 pansystolic murmur at left lateral sternal border with a loud single P2 with no splitting was heard with parasternal heave, suggestive of early shunt reversal. Electrocardiogram showed right axis deviation with right ventricular hypertrophy. Chest x-ray obtained after abdominal shielding showed biventricular cardiomegaly with prominent pulmonary conus with perihilar haze. Her haemoglobin was 10.8g/dl with haematocrit of 35% & platelet count was 1,68,000/mm^3. Arterial blood gas (ABG) on room air was pH‑7.28, PaCO2‑38 mmHg, PaO2‑55 mmHg and SaO2‑85%. Cardiologist advised to avoid exertion during labour. Obstetrician detected the development of late decelerations in fetal heart monitor & diagnosed it as a case of fetal distress hence it was decided to deliver the baby by caesarean section & we decided to give general anaesthesia as it was a dire emergency. Cardiologist advised to give fluids intraoperatively at the rate of 50 ml/hour. Written & informed high risk consent of ASA grade V with death on table consent was taken. Anaesthesia work station was checked and difficult airway cart was kept ready. In the operating room the patient continued to receive oxygen by facemask; ECG lead II and pulse oximeter were attached for continuous monitoring. Intravenous access was secured in peripheral vein. The baseline parameters were HR 90 beats/ min, BP 100/70 mm Hg and SpO2 was 99% on 100% oxygen 6liters/min during pre-oxygenation. Ampicillin and gentamicin were administered for prophylaxis against bacterial endocarditis. She was premedicated with Inj Ondensetron 4mg iv & Inj Glycopyrrolate 0,2mg iv. Inj. Midazolam 2mg iv & Inj. Fentanyl 150 mcg iv was given & patient was intubated with 7.0 size ET tube. Bilateral air entry checked, cuff inflated & tube was fixed. Anaesthesia was maintained with 100% oxygen and 0.8% sevoflurane and neuromuscular blockade was achieved with Inj. vecuronium 4 mg i.v. The end tidal carbon dioxide (EtCO2) was maintained between 32 and 36 mmHg. A live female baby with Apgar score of 5 at 1 min and 8 at 5 min, weighing 1.9 kg, was extracted. Intravenous infusion of oxytocin 10 units was administered slowly over 30 min. Blood loss was estimated to be about 500 mL. The patient remained haemodynamically stable throughout the procedure, which lasted for 45 min. Under aseptic & antiseptic precautions right internal jugular vein was cannulated.

CVP was found to be 12cm H_2O. IV fluids were titrated so that CVP of 8-9 cm H_2O was maintained & volume overload was avoided as it could easily precipitate right ventricular failure in these cases. For post-operative analgesia Epidural catheter was inserted at L2-L3 level in left lateral position after completion of surgery. Epidural space was located at 4cms & catheter was fixed at 10cms from skin. Patient was not reversed nor extubated & was shifted to ICU for continous monitoring & post-operative care. Inj.Tramadol 60mg was given 8 hourly through epidural catheter. 6 hours after delivery anticoagulation therapy was started with subcutaneous heparin 5000 IU subcutaneously given once daily. A 2D ECHO study performed 24 hours post partum, showed subaortic VSD with bi-directional shunt with RVSP of 45 mmHg and Qp: Qs:: 1.7:1 with TR. Patient was extubated after 2 days in ICU. Dose of heparin was withheld for 12hours & Epidural catheter was removed. Patient was later shifted to cardiology institute where cardiac catheterization was done. 8days after catheterization patient was discharged. A 2D ECHO study performed 3 days prior to discharge revealed subaortic VSD with bi-directional shunt with RVSP of 36 mmHg & Qp:Qs: 1.4:1 with TR.

DISCUSSION
Pulmonary hypertension in pregnant women is uncommon. It is associated with high mortality and death may occur regardless of the severity of disease. Hence it has been suggested that pregnancy is contraindicated in the patient with pulmonary hypertension. Pregnancy in secondary vascular pulmonary hypertension carries more than 50% mortality rate as compared to 30% risk of maternal mortality in idiopathic pulmonary hypertension and 30%‑40% in Eisenmengers syndrome.[1] Patients with Eisenmenger syndrome develop cyanosis & also present with polycythemia secondary to persistent hypoxia which in turn leads to thromboembolic complications. The two important factors which decide safe anaesthetic management of a pregnant patient with Eisenmenger's syndrome are, firstly, a fall in the SVR (which could allow a right to left intracardiac shunt) and, secondly, thromboembolism (which could fatally interfere with an already embarrassed pulmonary circulation). The resting cardiac output increases 30 to 40% with the maximum increase reached by the end of the second trimester. The goal of management of a patient with Eisenmenger's syndrome is the maintenance of systemic vascular resistance to prevent the increase in right to left shunt. Poor prognostic signs in maternal congenital heart disease include maternal hematocrit greater than 60%, arterial oxygen saturation lower than 80%, right ventricular hypertension, and syncopal attacks. A fixed pulmonary hypertension not responsive to oxygen also carries a grave prognosis and may be an absolute indication to terminate the pregnancy. The presence of fixed pulmonary hypertension not responding to oxygen therapy may perhaps be an absolute indication for termination of pregnancy.[8] As advised by the cardiologist, to avoid the stress of labour

and the late pregnancy, caesarean section was done. There are reports regarding elective caesarean section to optimize fetal development and to minimize the maternal risks at term in these types of patients.[3] But in our case the patient had fetal distress hence we couldn't wait for optimization. These patients, however, are high‑risk candidates for anaesthesia.[7,8] The main anaesthetic goal is to avoid a fall in the arterial blood pressure by maintaining both the cardiac output and the systemic vascular resistance.[4,6,7] The goal of monitoring is to detect sudden changes in the haemodynamics early so as to initiate appropriate treatment and prevent further complications. The need for invasive monitoring in Eisenmenger's syndrome is controversial. These patients are polycythemic and intraarterial catheterization may be associated with a higher incidence of post‑canulation thrombus formation. Insertion of central venous catheter has a potential risk of infection and paradoxical air embolus.[7] The complications of pulmonary catheterization are pulmonary arterial rupture in the presence of pulmonary hypertension apart from arrhythmias and systemic embolisation.[8,9] Weighing the risks & benefits we inserted a central venous catheter to optimize the pre‑load & detect right heart failure as the right heart is ejecting against high pulmonary vascular resistance. If the patient requires an operation, either regional or general anaesthesia are suitable. Several authors have recently suggested that it is safe to administer epidural anaesthesia to a patient with Eisenmenger's syndrome.[7,9] In our case as it was dire emergency we gave general anaesthesia but inserted epidural catheter post-operatively for analgesia. However, in each of these reports, it appeared that the patient's pulmonary vasculature dilated in response to oxygen. In Eisenmenger's syndrome, the amount of right‑to‑left shunt depends in part on the ratio of SVR to pulmonary vascular resistance (PVR). Epidural anaesthesia causes sympathetic blockade that reduces SVR. If SVR decreases without a concomitant decrease in PVR, the amount of right‑to‑left shunt increases.[9,10] Therefore, in our case, we considered it undesirable to induce a sympathetic blockade that might have resulted in an increased right‑to‑left shunt. We opted for general anaesthesia. This may be achieved by combining a short‑acting i.v. narcotic such as fentanyl in addition to Although thiopentone causes a decrease in SVR, the effect is dose‑dependent.[2] Hence, we used fentanyl & midazolam as inducing agent for intubation. Although ketamine has theoretical advantages over barbiturates as an induction agent, in that it does not reduce SVR but causes increase in the heart rate, which is undesirable. Traditional and contemporary teaching considers all obstetric patients to be at increased risk for pulmonary aspiration compared with patients scheduled for non‑obstetric elective surgical procedures, mandating antacid prophylaxis as well as rapid sequence induction with cricoid pressure. However, the requirements of a patient at risk for aspiration are difficult to reconcile with a judicious, titrated induction of anaesthesia that is ideal for a patient with severely compromised cardiac function. Thus, rapid sequence induction was avoided in our case. The problems of general anaesthesia and positive‑pressure ventilation are decrease in venous return and cardiac output. We avoided nitrous oxide because it is a potent pulmonary vasoconstrictor.[8] After the extraction of the baby, we chose to administer oxytocin as slow infusion. Cole and colleagues[6] have mentioned that uterine massage along with a slow oxytocin infusion proved safe and helpful in these patients. Oxytocin as a bolus causes direct vasodilation and reduces SVR with compensatory increase in the heart rate and cardiac output.[6,9] Because of the risk of pulmonary thromboembolism and paradoxical systemic embolisation, heparin was given in a low dose,[10] as many case reports suggest death in post operative period due to pulmonary thromboembolism.

CONCLUSION
In conclusion this article suggests that the pregnancy in a patient with eisenmenger's syndrome can remotely be successful but, must be discouraged. Anaesthetic management of these patients requires meticulous preparation, multi-disciplinary approach, pros & cons of all the anaesthetic agents to maintain adequate cardiovascular stability. Early extubation should be avoided in such patients because, invariably, they may go for worsening of shunt & heart failure. Adequate measures should be taken to prevent thromboembolism. Thus, we recommend a general anaesthetic technique with maintenance of haemodynamics as close to normal as possible, with adequate control of pain and early initiation of thromboprophylaxis for successful management of similar cases.

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