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Case Series

Successful outcome in pregnancies with eisenmenger syndrome for category 1 LSCS: A case series

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ABSTRACT

PAH associated eisemnenger syndrome is a congenital heart disease with bidirectional or reversed shunt. The maternal mortality in this syndrome is very high 30-50%. We present a case series of 3 patients with eisemnenger syndrome with successful materno-faetal outcome of patients under general anaesthesia for category 1 LSCS. The focus of anesthetic management was to preserve SVR (systemic vascular resistance) to PVR (pulmonary vascular resistance) ratio.

1. Introduction

Pulmonary hypertension has incidence of approximately 1.1 in 100,000 pregnancies, defined as a mean PAP greater than 25 mm Hg and pulmonary vascular resistance (PVR) greater than 3 Wood units.[1,2] Congenital heart disease in pregnancy is the major cause of maternal mortality attributing approximately 25 to 30% of total maternal deaths.[3] The left-to-right shunt in the form of septal defects or PDA causes increased pulmonary blood flow leading vascular remodeling and endothelial dysfunction, which increases PVR giving rise to pulmonary hypertension. Over time, this increase in PVR causes physiological & anatomical phenomenom i.e reversal of the shunt (from left-to-right to right-to-left) or bidirectional shunt known as Eisenmenger syndrome, which in pregnancy is very uncommon but notably with high maternal mortality (i.e 30 to 50%), hence classified as high-risk category due to its possible progression to right ventricular failure and death.[2,4] Hence, pregnancy is contraindicated in this condition,[5,6] although outcomes have improved in the past two decades.[7] We have successfully managed three patients of PAH with Eisenmeger syndrome with favorable maternal and fetal outcomes.

2. Case Reports

2.1. Case 1

A 27-year-old primigravida, weighing 55 kg, admitted with bleeding PV was received in emergency OR (For category 1 LSCS) with 7 months amenorrhea, she was dyspnoic and cyanosed. Physical examination revealed Grade II clubbing, HR of 84/min, BP of 128/72 mmHg & SpO_2 of 86%. With pansystolic murmur over apex, ECG showed right axis deviation & right ventricular hypertrophy. Bed side 2D ECHO by cardiologist documented a large VSD, override >50% with biventricular shunting, biventricular hypertrophy (TOF Physiology) and an ejection fraction of 43%. Immediate termination was advised, Patient was induced with intravenous etomidate 10mg, ketamine 60mg and succinylcholine 100mg, Intubated with 7 mm cuffed

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ETT. Anaesthesia was maintained with 100% O_2 plus Isoflurane (0.8 to 1%), vecuronium 4 mg, fentanyl 75μg and paracetamol 1gm.The right radial artery was cannulated for invasive BP monitoring. Intra operative ABG revealed pH 7.34, PO_2 87, PCO_2 29, HCO_3 18.3, Na 135, K 3.3, Hct 39, Lactate 1.4, SO_2 96%. A live baby with Apgar score of 5 at 1 min and 9 at 5 min was delivered. A 3-unit bolus of oxytocin and infusion of 15 units was administered slowly over 2 hours. A volume of 750 RL was administered. Estimated blood loss was 450 mL. Intraoperative hemodynamics were stable with HR of 88 to 93 b/min, SBP 116 to 124, DBP 78 to 82. However, SPO_2 remained 89 to 92% on 100% O_2. Patient was reversed and extubated on table, shifted to the ICU with post op vitals HR 88/ min, BP 132/82, SPO_2 88% on room air. In her first 48 hours of stay in ICU, her serial ABGs showed an increase in PO_2 to 76 mmHg from 60 mmHg, she was discharged from ICU on room air SPO_2 92% with advice for cardiology follow up.

2.2. Case 2

A, 29-year-old, 60 kg, G3A2 (early pregnany losses) at 37 weeks of gestation with AFD was referred from peripheral hospital with ECHO Documented large VSD, biventricular shunting with severe PAH (RSVP>100 mmhg) on metaprolol 25 mg. Despite advised termination in her early pregnancy, she had refused and continued her pregnancy. Patient was received in Emergency OR for category 1 LSCS in view of AFD. Her pre op vitals were HR 113/min, BP 132/82, SPO2 82% on room Air. After preoxygenation, The patient was induced with intravenous etomidate 15 mg and injection succinylcholine 100 mg, intubated with 7.5 cuffed ETT. Anaesthesia was maintained with 70% O2 +30% air + Isoflurane (0.8%), injection fentanyl 100 ug and paracetamol 1gm. left radial artery was cannulated for IBP. Intra operative ABG revealed pH 7.39, PO_2 73, PCO_2 31, HCO_3 17.8, Na 137, K 4.3, Hct 31, Lactate 1.1, SO_2 96%. A live baby with Apgar score of 4 at 1 min and 9 at 5 min was extracted. A 3 units bolus of oxytocin and infusion of 15 units was administered slowly over 100 minutes. Estimated blood loss was 750 mL. Intraoperative hemodynamics were stable with HR 82 to 97b/min, SBP 128 to 136, DBP 71 to 92. SPO_2 remained 90 to 92% on 100% O_2. Patient was reversed and extubated uneventfully on table, shifted to the intensive care unit with post op vitals HR 91/ min, BP 127/80, SPO_2 87% on room air and 91% on facemask @ 6 L/ min. After 6 hours of LSCS, the patient had continuous trickle of bleeding PV, Her Hb dropped to 6.1 g% from baseline Hb of 10.2 g%. She was transferred back to OR for curete and uterine tamonade, induced again with injection etomidate 8mg + ketamine 50 mg and succinylcholine 100mg.Her intra op vitals remained stable with 1 unit of PRBC transfusion, however the SpO2 dropped down to 86%. The patient was not extubated and was shifted to ICU intubated on 100% Oxygen. She was put on SIMV mode, sedated with fentanyl 60mcg /hr over next 12 hours. Her serial ABGs showed a constant improvement in PO_2 from 64 mmhg to 88 mmhg. She was extubated in next morning uneventfully. After 72 hours of her stay in ICU, she was maintaining $SP0_2$ of 90% on room air and she was transferred to ward on oxygen.

2.3. Case 3

A 26-year-old primigravida, 65kg admitted with decreased fetal movements (CTG documented significant dips) at 35 weeks of gestation. With significant history of breathlessness since start of her 8th month of pregnancy and ECG documented right ventricular hypertrophy. Emergency cardiology consultation and Bed side ECHO revealed eissenmenger syndrome secondary to VSD, pulmonic stenosis with PAH (RSVP > 120 mmHg) and EF 65%. Immediate termination of pregnancy was advised by cardiologist. Patient was received in Emergency OR for category 1 LSCS. Her pre op vitals were HR 109/min, BP 102 / 62 and SPO2 88% on room air. Patient was induced with intravenous ketamine 60 mg + Etomidate 12 mg and injection succinylcholine 120mg after 3 minutes of preoxygenation, intubated with cuffed oral ETT (7.0 mm ID). Anaesthesia was maintained with 100% O_2 + Isoflurane (0.8%), injections vecuronium 6 mg, fentanyl 100 ug and paracetamol 1gm. Left radial artery was cannulated for invasive BP monitoring. Intra operative ABG revealed pH 7.41, PO_2 55, PCO_2 26, HCO_3 16.3, Na 134, K 3.8, Hct 27, Lactate 1.8, SO_2 81%. A live baby with Apgar score of 3 at 1 min and 8 at 5 min was extracted. A 3 units bolus of oxytocin and infusion of 15 units was administered slowly over 2 hour. Estimated blood loss was 550 mL. Intraoperative hemodynamics were stable with HR of 89 to 107b/min, SBP 89 to 108, DBP 52 to 67. SPO_2 remained 88 to 94% on 100% O_2. An infusion of dopamine @ 5 mcg/min was started up to end of surgery. Patient was reversed and extubated uneventfully on table, shifted to the intensive care unit with post op vitals HR 112/ min, BP 122/68, SPO_2 85% on room air and 92% on facemask @ 6 L/ min. ABG pH 7.32, PO_2 69, PCO_2 29, HCO_3 19, Na 137, K 3.4, Ca .88. After 56 hours of her stay in ICU, she was maintaining $SP0_2$ of 93% on room air and was trasferred to ward.

3. Discussion

The peripartum period in eisenmenger syndrome is most challenging owing to increased oxygen demand & rapid fluid shifts in this period. In presence of fixed PVR in PAH, the physiological decrease in SVR aggravates the severity of the right-to-left shunt. The pregnancy related decrease in FRC renders parturients unresponsive to increased oxygen demands of pregnancy & predisposes them to hypoxemia and cyanosis. When maternal resting SPO2 is less than 85%, as was seen in our two cases, the maternal risk is extremely

high owing to possibility of development of right ventricular failure or death[4] with the chance of a live birth to only 12%. Therefore, many authors recommend against pregnancy in eisenmenger syndrome. However, late diagnosis or When termination of pregnancy is refused as in one of our cases, such parturients should be managed under the supervision of a multidisciplinary team with facilities of high risk care like cardiac imaging, modern-day safer anesthetic drugs & ICU which have improved the outcome in patients with Eisenmenger syndrome in the last two decades.[7,8]

The primary anaesthetic management goals in obstetric eisenmengers syndrome are: 1) Maintenance of SVR/PVR ratio and intravascular volume 2) Prevention of aortocaval compression, hypoxemia, hypercarbia, pain & acidosis 3) Avoidance of myocardial depression. Early planned delivery at 32 to 34 weeks of gestational age, may contribute to improved outcomes.[9]

Both general anesthesia, epidural or CSE have been safely reported based on multiple case reports and case series studies.[10–13] However, we considered it undesirable to induce a sympathectomy as fall in SVR increases the right to left shunting.[14] Moreover, epidural or CSE Anesthesia are not feasible for type 1 LSCS, when anticoagulation administration in early post-operative period is also anticipated.

We received all our patients in emergency OR for category 1 LSCS. Hence, our choice was restricted to General anaesthesia. We induced either with etomidate (0.3mg/kg) or etomidate (0.2mg/kg) plus ketamine(1mg/kg) combination and succinylcholine. Both etomidate and ketamine maintain SVR - a primary anaesthetic goal in these patients. A study done on 60 peadiatric patients posted for cardiac catheterization has shown that with the use of etomidate and ketamine, there were no hemodynamic changes in the group with a right-to-left shunt.[15]

We didn't resort to RSI as we apprehended a decrease in SVR with rapid induction doses, therefore slow induction dose was preferred. The general anaesthesia and positive-pressure ventilation may cause decrease in venous return and cardiac output. Therefore, we decided to maintain the SVR with prophylactic dopamine infusion. Since all of our patients had poor functional status and hemodynamically significant lesions.[9] The intra arterial cannulation in all patients was secured for hemodynamic monitoring which continued till entire post-operative periods in ICU.

The nitrous oxide in all of our cases was avoided as it is a potent pulmonary vasoconstrictor.[16] The anaesthesia was maintained on an optimal dial concentration of isoflurane with bolus doses of vecuronium, fentanyl and paracetamol so as to avoid systemic hypotension, myocardial depression and vasodilation. After the delivery of the baby in each case, we chose to administer oxytocin in low bolus dose (3mg) and then slow infusion.The study by Cole and colleagues[17] suggested that using uterine massage alongwith a slow oxytocin infusion proved safe and uneventful in these patients,contrary to this a large bolus of oxytocin,causes direct vasodilation,reduces SVR with compensatory increase in the heart rate.[18,19] During the first three to four weeks after delivery, Maternal mortality is high, so prolonged postoperative care in an intensive care unit setting is needed.[19] Our all patients were put on oxygen therapy, monitored in ICU in their post-operative periods and were successfully shifted to the ward after within 72 hours.

4. Conclusions

The eisenmenger syndrome in PAH is anaesthetic challenge, though pregnancy in this syndrome is contraindicated owing to its high mortality rate. However, many case reports have documented successful outcomes. Our case series of three patients with eisenmenger syndrome under general anaesthesia suggests that with meticulous preparation and multidisciplinary care, the successful materno fetal outcome is possible. However, we suggest the multi disciplinary supervision with Early planned termination of pregnancy, which is better rather than unplanned emergency termination.

5. Source of Funding

None.

6. Conflict of Interest

None.

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