e-ISSN: 2319-9865
p-ISSN: 2322-0104
Research and Reviews: Journal of Medical and Health Sciences

Anaesthetic Management of a Known Case of Eisenmenger's Syndrome Posted for Elective Caesarean Section.

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Case Report

Received: 12/02/2014
Revised: 03/03/2014
Accepted: 14/03/2014

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Keywords: Eisenmenger's syndrome, caesarean section, general anaesthesia, pulmonary thromboembolism.

ABSTRACT

Eisenmenger's syndrome is a cyanotic congenital heart disease that includes pulmonary hypertension with reversed or bidirectional shunt. Pregnancy induced physiological changes include decrease in systemic vascular resistance and increase in cardiac output.the decreased systemic vascular resistance associated with pregnancy increases the degree of right to left shunt which carries risk to mother and foetus both. Anaesthetic management of such a case requires balance between systemic and pulmonary vascular resistance.we present a case of a 25 years old, primigravida with eisenmanger's syndrome (large vsd and rvsp of 115 mm of hg) at 38 weeks of gestation posted for elective caesarean section.endotracheal anaesthesia with inj. Fentanyl, inj. Ketamine and inj. Vecuronium bromide was given and maintained on oxygen and sevoflurane. Intraoperative period was uneventful. On 3rd post-operative day patient developed tachycardia, tachypnoea, hypotension and decrease in oxygen saturation. She became unconscious and diagnosed as pulmonary thromboembolism. She died on the same day. Thromboembolic phenomena as a complication of eisenmanger's syndrome in pregnancy is a challenging situation to treat in postoperative period.

INTRODUCTION

Physiological changes during pregnancy involving Cardiovascular system are decreased systemic vascular resistance (SVR), increased blood volume, and increased cardiac output (CO) secondary to increased heart rate (HR) and stroke volume (SV). Cardiac disease is a major cause of peripartum maternal death, 25% of which are due to congenital heart disease [1]. Eisenmenger's syndrome is defined as the development of pulmonary hypertension in response to a left to right cardiac shunt with consequent bidirectional or reversal of shunt.

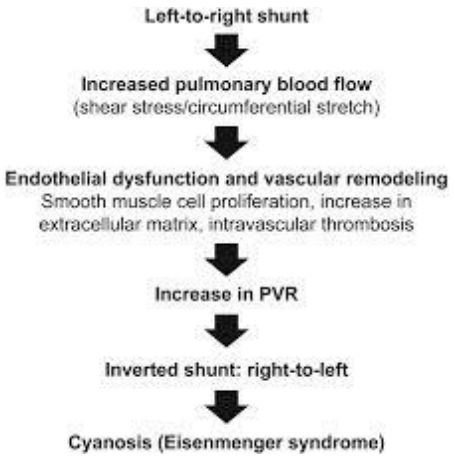

Figure 1: pathophysiology of Eisenmenger's syndrome

e-ISSN: 2319-9865
p-ISSN: 2322-0104

Eisenmenger's syndrome is classified in the high-risk category, with potentially severe cardiac and neonatal complications [2]. Despite advances in medicine, the perioperative maternal mortality remains high (estimated at 30%–50%)

Case Report

A 25 years old primigravida, known case of Eisenmenger's syndrome at 38 weeks of gestation, was scheduled for an elective caesarean section. She was a known case of congenital heart disease since the age of 3 years having large muscular VSD (left to right shunt). Patient was diagnosed as Eisenmenger's syndrome at the age of 23 years with reversal of shunt. Patient was on tab. Frusemide and tab. Sildenafil 25 mg for 2 years. On admission according to cardiologist's advice tab. Sildenafil was stopped because of its side effect of hypotension and patient was continued on tab. Frusemide 20 mg OD, inj. Deriphyline 1 amp 12 hourly and combimist nebulisation. Preoperatively, apart from exertional dyspnoea (grade II), she was asymptomatic. On examination, patient was conscious, following verbal command, oriented to. Temperature was normal by palpation, pulse Rate 106/min, blood pressure 110/64 mm Hg, On air SpO_2 77-79%, Clubbing grade 2 and cyanosis were present with no peripheral oedema. Her lungs were clear to auscultate and cardiac examination showed regular rate and rythem with loud P_2 and pansystolic murmur over the lower left sternal border. On airway examination mouth opening was adequate with Mallampati grade-1. Her haemoglobin was 13.6 gm% with haematocrit of 39.2% and platelet count of 68,600/Cmm. Arterial blood gas (ABG) on room air was PaO_2-39 mmHg, $PaCO_2$-23.5 mmHg, pH- 7.48, HCO_3-17.9, SaO_2 -78.4%. 2D ECHO at the age of 10 years showed acyanotic CHD, large muscular VSD with bidirectional shunt (predominantly left to right) size 11.7 mm, small ostium secondum ASD, severe PAH, RVSP 48 mm Hg, mild MR (15%) with left to right shunt. Whereas 2D ECHO at the age of 25 years showed large mid muscular VSD with bidirectional shunt, predominant right to left shunt, moderate TR with severe PAH with RVSP 115 mm Hg. Cardiologist opinion was taken and patient was advised to undergo elective caesarean section with high cardiac risk. Patient was advised to avoid morning dose of frusemide on the day of surgery.

Informed and written consent for ASA IV was taken. Infective endocarditis prophylaxis was given. In the operation theatre standard monitors including ECG, NIBP, Pulse Oxymetry, ETCO2, Urine output and PNS were applied. The pre-induction pulse rate was 96/ min and blood pressure was 138/86 mm of Hg. She was pre-medicated with Inj ondansetron 4 mg iv and Inj ranitidine 50 mg iv 10 min before induction. Inj glycopyrrolate 0.2 mg iv and Inj fentanyl 70 mcg iv were given 3 min before induction. After 3 min of pre-oxygenation, SpO_2 increased upto 85-86%. Intravenous Induction was carried out with Inj. Ketamine 80 mg iv and Inj. Vecuronium 5 mg iv. The patient was Intubated endotracheally with portex, cuffed endotracheal tube no. 7 mm ID, BLAE checked, cuff inflated and tube fixed. Anaesthesia was maintained with 100% O2 and Sevoflurane and Neuromuscular blockade was achieved with intermittent doses of Inj. Vecuronium. The end tidal carbon dioxide ($EtCO_2$) was maintained between 32 and 35 mm hg.

A live male child with Apgar score of 5 at 1 min and 8 at 5 min, weighing 2.2 kg was extracted after 5 min of induction. Inj oxytocin drip (20 unit in 500 ml NS) was started and inj carboprost 250 mcg was given IM after delivery of baby. Inj diclofenac 75 mg was given iv for analgesia and Inj frusemide 10 mg was given iv. Intraoperative 500 ml of DNS given and urine output was 350 ml. Intraoperatively patient remained stable hemodynamically throughout the procedure, which lasted for 45 min. Intraoperative ABGA was PaO_2 117.8 mmHg, $PaCO_2$ 30.5 mmHg, HCO_3 18.3 and SaO_2 97.5%. Neuromuscular blockade was reversed with Inj. Glycopyrrolate 0.4mg and Inj. Neostigmine 2.5 mg and the patient was extubated when she had sustained spontaneous respiratory effort. Post-extubation, patient was conscious, following verbal commands with respiratory rate of 14 to 16/min, regular with adequate tidal volume and muscle tone, power. SpO_2 was 66-68% on air and 72-74% with ventimask with O2 @ 4 L/min.

The patient was shifted to post op ward with oxygen for observation. Initially patient was stable hemodynamically with SpO_2 80-85%. After 5 hours she developed fever, tachycardia, cyanosis and SpO_2 55-60%. Cardiologist opinion was taken and tab. Digoxin (0.25 mg) 3 tab stat f/b tab Digoxin (0.25 mg) ½ OD (5/7) was started, tab frusemide+spironolactone 1 OD was continued with strict I/O charting. On the second post-operative day, patient developed hypotension with blood pressure of 90/54 mm Hg and SpO_2 50-60%. Inj noradrenaline 2 amp in 500 ml of NS was started @ 10 ml/hr and BP was maintained around 130/78 mm Hg with PR 96/min and SpO_2 64-66% and coagulation profile was advised for clinically suspected pulmonary thromboembolism. Cardiologist opinion was taken for starting heparin. On the third post-operative day, she had tachypnoea and desaturated to 30-40% SpO_2. Patient then deteriorated with loss of consciousness. She was intubated and CPR given but could not be revived.

She was diagnosed as pulmonary thromboembolism with plasma fibrinogen level 4.08, D-Dimer value 7.45 and Fibrinogen Degradation Products >20.

e-ISSN: 2319-9865
p-ISSN: 2322-0104

DISCUSSION

Victor Eisenmenger in 1897 coined the term Eisenmenger complex, which included large ventricular septal defect and pulmonary hypertension [3]. Wood redefined this in 1958 as pulmonary hypertension with reversed or bidirectional shunt, associated with septal defects or patent ductus arteriosus [4,5]. Two major problems in pregnant patient with Eisenmenger's syndrome- a fall in the systemic vascular resistance (SVR) (which could allow a right to left intracardiac shunt) [4]. and thromboembolism (which could fatally interfere with an already compromised pulmonary circulation) [6, 7].

Multisystem/Organ Involvement in Eisenmenger Syndrome

- Haematologic and haemostatic abnormalities
  - Secondary erythrocytosis
  - Iron deficiency
  - Hyperviscosity syndrome
  - Minor bleeding: dental bleeding, epixtasis, menorrhagia
  - Major bleeding: haemoptysis, gastrointestinal bleeding, cerebral haemorrhage
  - Pulmonary arterial thrombosis
- Neurological disorders
  - Stroke
  - Transient ischaemic attack
- Hypertrophic pulmonary osteoarthropathy
- Hyperuricemia and gout
- Renal dysfunction
  - Proteinuria
  - Renal failure
- Gastrointestinal: cholelithiasis
- Dermatological: acne
- Infections
  - Bacterial infectious disease: brain abscess, endocarditis, pneumonia
  - Viral infections
- Arrhythmias
  - Supraventricular tachycardia
  - Ventricular tachycardia
- Right heart failure

Figure 2: maternal complications in Eisenmenger's syndrome

Fetal complications are: IUGR, prematurity and high perinatal mortality rate

Because of high mortality risk during pregnancy and peripartum, women with Eisenmenger's syndrome should be strongly advised against pregnancy as well as terminate pregnancy if at all conceive. If a woman with Eisenmenger's syndrome becomes pregnant, coordinated care should be established early, involving a cardiologist, pulmonary hypertension specialist, obstetrician and anaesthetist. Close cardiovascular monitoring, with specific attention to volume status, is essential throughout pregnancy and peripartum period. Treatment with pulmonary vasodilator therapy should be discussed with a pulmonary hypertension expert. Extended postpartum period of monitoring in hospital is recommended.

The place of invasive monitoring in Eisenmenger's syndrome is controversial, the risk of complications must be weighed against the value of information obtained. 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 [8]. The complications of pulmonary catheterization are pulmonary arterial rupture in the presence of pulmonary hypertension apart from arrhythmias and systemic embolisation. Therefore in our case we avoided invasive monitoring.

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]. In general anaesthesia, positive pressure ventilation may decrease venous return and systemic blood pressure, which can increase right-to-left shunting. The choice of general versus epidural-spinal anaesthesia should be made after considering the patient's unique physiology and with consultation with cardiologists and obstetricians. In our case we opted for general anaesthesia because patient had thrombocytopenia and to avoid decrease in SVR. This was achieved with inj. fentanyl to produce stable hemodynamic without subsequent neonatal depression and inj. Ketamine as an induction agent as it does not reduce SVR [7,8]. For maintenance of anaesthesia, nitrous oxide was avoided because it is a potent pulmonary vasoconstrictor.

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p-ISSN: 2322-0104

The role of anticoagulation in Eisenmenger's syndrome is controversial. Pregnancy represents a hypercoagulable state, and evidence suggests pulmonary thromboembolism as a cause for maternal demise. However, adverse outcome has been associated with prophylactic heparin therapy in parturients with Eisenmenger's syndrome. Judicious use of antithrombotic drugs and early ambulation may increase survival in patients with Eisenmenger's syndrome [10]. In our case anticoagulants were not started in immediate postoperative period by cardiologist because of the high risk of hemorrhage with thrombocytopenia and coagulopathy.

The cause of death was discussed by the multidisciplinary team, and pulmonary embolism was concluded as a cause of death as patient developed tachypnea and desaturated with increased fibrinogen, FDP and D-Dimer value.

CONCLUSION

Pregnancy is the cause of highest mortality in women with Eisenmenger's syndrome due to life threatening complications like thromboembolism and worsening of shunt in perioperative period even at tertiary centre. Therefore such women should be strongly advised against pregnancy. Safe anaesthetic management of patients with eisenmenger's syndrome requires meticulous preparation and multidisciplinary approach to maintain the cardiovascular stability.

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