We report a very rare case of Primigravida who was 19-year-old female, 50 kg weight, 37 weeks pregnancy with pulmonary hypertension, paramembranous large Ventricular Septal Defect (VSD) (2.5 cm²) bidirectional shunt mainly left to right with severe pulmonary stenosis, peak gradient pressure of 144 mmHg. There was history of breathlessness on exertion otherwise patient was well adjusted to normal life. She was planned for cardiac surgery (VSD closure with valvotomy) after delivery. CVS examination revealed pansystolic murmur.

Patient was taken up for emergency cesarean section. On preoperative examination patient had a heart rate of 107/min, blood pressure 155/106 mmHg and respiratory rate of 28/min. On investigation Hb 11.3gm%, Serum creatinine 0.9 mg/dl, platelet count 1,43,000/cmm, BUn 22.5, InR 0.96. X-ray chest depicted prominence of pulmonary artery and cardiomegaly. Electrocardiogram showed: right ventricular hypertrophy with right bundle branch block.

Echocardiography confirmed pulmonary hypertension with large perimembranous VSD, bidirectional shunt mainly left to right with severe pulmonary stenosis and peak gradient of 144 mmHg. Patient was taken up for caesarean section under general anaesthesia after explaining grave risk to mother and fetus due to severe cardiac disease. Peripheral venous access and radial arterial line were established for continuous blood pressure monitoring antimicrobial prophylaxis was given. Injections ranitidine and metclopramide were given IV stat. About 15° of left lateral tilt was maintained to prevent any supine hypotension. The patient was preoxygenated with 100% oxygen for three minutes and Injection xilocard 60mg was given intravenously followed by rapid sequence induction with injection etomidate 15mg IV and succinylcholine 75mg (1.5mg/kg body weight) IV. The patient was intubated with a 7.5mm size cuffed oral endotracheal tube under direct laryngoscopy and put on volume control mode with a tidal volume 8 ml/kg body weight. The respiratory rate was adjusted to maintain EtCO₂ of around 35cm H₂O. Anaesthesia for the patient was maintained with isoflurane in concentration of 0.8%-1.0% along with oxygen and air mixture at 50:50 concentration. Nitrous gas was avoided. Injection vecuronium 0.1 mg/kg body weight was given as loading dose with top ups as needed. Oxytocin 5 units stat and 15 units in 500 ml ringer lactate was given after delivery of baby. Fentanyl 2.5mcg/kg body weight IV was used as a analgesic and given along with midazolam 0.05mg/kg after delivery of baby. One litre of ringer lactate was infused intraoperatively. Urinary output at the end of surgery was around 400ml. Patient was haemodynamically stable during surgery. An alive male baby weighing 2.4kg body weight with Apgar score of 8 & 10 at 1 and 5 min was delivered. Patient was not extubated in view of cardiac condition. Injection morphine 0.15mg/kg was given intravenously and patient was shifted to ICU for mechanical ventilation and planned gradual extubation. Patient was put on SIMV mode of ventilation and was ventilated overnight. Patient was shifted to continuous positive airway pressure mode in morning and extubated successfully. 5% dextrose at rate of 60 ml/min was administered postoperatively. Patient remained haemodynamically stable throughout the procedure as well as in postoperative period for next 48 hours with successful maternal and fetal outcome.

**DISCUSSION**

The incidence of cardiac disease in pregnant patients in developed countries ranges from 0.2% to 3% and the incidence in developing countries may be still higher. Earlier studies report that 44% women with cardiac disease develop pulmonary oedema in third trimester [1]. As per modified WHO classification of maternal cardiac risk disease, pulmonary hypertension is categorized under class IV [2].

Adults with un repaired large VSD (>1.25 cm²) develop left heart failure and pulmonary hypertension and have a high incidence of bacterial endocarditis [3]. If pulmonary arterial pressure reaches...
systemic levels, there is reversal or bidirectional flow Eisenmenger syndrome [4]. In our case, it was term pregnancy with pulmonary hypertension group I with large paramembranous VSD (2.4 cm²) with bidirectional shunt mainly left to right with severe pulmonary stenosis.

It is associated with significantly high maternal mortality. The woman is at greatest risk during labour and delivery when there is diminished venous return and decreased right ventricular filling both associated with most maternal deaths [4]. There is grave risk of maternal death for 1 month postpartum too.

Proper attention is given to analgesia and to prevent blood loss to avoid hypotension [4]. Women with Group I severe hypertension have delivered successfully while using either inhaled nitric oxide or iloprost [5,6].

The goal of haemodynamic management are maintenance of right ventricular preload and filling pressures, left ventricular afterload and right ventricular contractility [7]. Excessive fluid can precipitate right heart failure. Regional anaesthesia and local anaesthesia were avoided as these can decrease the preload excessively.

The main aim is to avoid hypothermia, hypercarbia, acidosis, hypoxia and high ventricular pressures and reduction of both pulmonary and systemic vascular resistance. General anaesthesia was selected for caesarean section. Inhalational and Narcotic based induction was avoided in view of slow induction, neonatal depression and maternal myocardial depression. Xylocard 2%, 1.5mg/kg body weight IV given to decrease stress response to induction. Systemic vascular resistance was kept low by fentanyl. Respiratory rate was adjusted as per EtCO₂ levels to maintain normocarbia [7].

CONCLUSION
In pulmonary hypertension, WHO risk class IV cardiac disease, pregnancy is contraindicated as it can worsen during labour, delivery and post-partum resulting in high maternal mortality. However, if term pregnancy is encountered, caesarean section may be performed under general anaesthesia. There are very few case reports which describe the anaesthetic management of pregnant women with severe cardiac illness. Larger studies are need of hour to draw a conclusion to manage these challenging cases.

REFERENCES