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Anaesthesia for Caesarean Section in a Patient with Eisenmenger's Syndrome

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ABSTRACT

Eisenmenger's syndrome was originally described in 1897 and redefined by Wood in l958. This syndrome includes pulmonary hypertension with reversed or bi-directional shunt associated with septal defects or a patent ductus arteriosus. A 27-year-old G2 PO with Eisenmenger's syndrome presented to the hospital for management at 17 weeks of pregnancy. She was advised termination of pregnancy but she refused. An elective caesarean section was performed successfully under general anaesthesia uneventfully at 29 weeks due to severe intrauterine growth retardation (IUGR). Patient's postoperative complications like pulmonary thromboembolism, the advantages and disadvantages of anti-coagulation are discussed. Pregnancy carries substantial maternal and fetal risk for patients with pulmonary hypertension and Eisenmenger s syndrome. Although pregnancy should be discouraged in women with Eisenmenger's syndrome it can be successful.

Keywords: Eisenmenger's Syndrome, Pulmonary hypertension, anaesthesia, Caesarean Section, Complications

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Eisenmenger's syndrome was originally described in 1897 and redefined by Wood in 1958[1]. This syndrome includes pulmonary hypertension with reversed or bi-directional shunt associated with septal defects or a patent ductus arteriosus. Maternal mortality is high in pregnant patients with a cumulative risk of 30-70%[2]. Death can occur anytime during the pregnancy or the puerperium. We describe the successful anaesthetic management for Caesarean section in a patient with Eisenmenger's syndrome.

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CASE REPORT

A 27-year-old primigravida was admitted to our hospital at 17 weeks of gestation for the management of her high-risk pregnancy. She was advised termination of her pregnancy due to the high maternal mortality but she refused. After admission, she was treated with bed rest, oxygen therapy and anticoagulation (subcutaneous heparin) to minimize intrauterine growth retardation (IUGR). She was scheduled for an elective Caesarean section at 32 weeks of pregnancy but it was carried out at 29 weeks due to severe IUGR.

She was first detected to have a cardiac murmur at the age of seven but she declined further investigation. A cardiac catheterisation was finally performed when she was sixteen years old. She was found to have situs inversus, secundum atrial septal defect (ASD) and severe pulmonary hypertension (118/56 mm Hg). Since then, she had been on regular follow-up by the cardiologist. Preoperatively, apart from cyanosis, she was asymptomatic and the effort tolerance was good.

On physical examination, she had cyanosis and clubbing. Her weight was 44 kg. The jugular venous pressure, blood pressure and heart rate were within normal limits. A grade 3/6 ejection systolic murmur was heard over the precordium with a loud second heart sound. The lungs were clear. Preoperative electrocardiography showed right axis deviation with right ventricular hypertrophy. Two-dimensional echocardiography revealed an enlarged right atrium and ventricle, ASD with right to left shunt and right ventricular hypokinesia. Arterial blood gas on room air was as follows: pH 7.46, PaO_2 49 mmHg, $PaCO_2$ 34 mmHg, and SaO_2 of 87%. The preoperative chest X-ray was normal. Her full blood count showed a haemoglobin of 14.1 gm%, platelets 110 000/mm^3. Serum urea, creatinine and electrolytes were within normal limits.

She was premedicated with ranitidine 150 mg and 30 ml of sodium citrate prior to the scheduled operation. Prophylactic antibiotic ampicillin and sulbactum 1500 mg given intravenously. Patient was on subcutaneous heparin 3000 IU bd. and received the morning dose.

When the patient arrived in the operating theatre, standard ECG, non-invasive blood pressure and a pulse oximeter were attached. Initial oxygen saturation was 84% on room air but this was improved to 99% on breathing oxygen 5L min^{-1} via facemask. An invasive

arterial line and a central venous catheter were inserted under sedation with midazolam 1 mg and fentanyl 50 µg. The preinduction blood pressure and central venous pressure were 135/78 and 15 mm hg respectively. Intravenous induction was carried out with etomidate 10mg slow bolus, fentanyl 50 µg and suxamethonium 100mg with cricoid pressure. Following induction of anaesthesia, the trachea was intubated and anaesthesia was maintained with 50% oxygen and sevoflurane (end-tidal concentration 1-2%) in air. The neuromuscular block was achieved with 15 mg of atracurium. The end tidal carbon dioxide was maintained between 32 and 35 mm hg. A live baby with the Apgar score of five at 1 and 5 minutes, weighing 800gm was delivered. Intravenous syntocinon 10 unit was administered over a period of 5 minutes to facilitate uterine contraction. The patient remained hemodynamically stable with systolic pressures between 130 and 140mmHg, central venous pressure around 10 mmHg and oxygen saturation above 98%. Incremental dose of morphine was given up to 7mg for analgesia intra-operatively. Blood loss was about 200 ml. At the end of the operation, neuromuscular block was reversed with 0.6 mg atropine and 1.25 mg of neostigmine, and the trachea was extubated. The patient was transferred to the intensive care unit for observation. She remained stable and maintained her saturation around 95% while breathing 5Lmin^{-1} of oxygen via facemask. She was given intravenous morphine for postoperative pain relief. She was discharged to the labour ward on the second postoperative day. In the labour ward, she became hypotensive (95/58 mmHg) after a few hours and oxygen saturation decreased to 64% despite supplementary oxygen. She was then readmitted to the coronary care unit. She had tachycardia, tachypnoea and her temperature raised to 38ºC. At this time her arterial blood gas while breathing 8 L of oxygen via face mask showed pH 7.36, $PaCO_2$ 33.6 mmHg, PaO_2 32.1 mmHg and SaO_2 57.9%. Her haemoglobin level decreased to 8.5gm%, but there was no obvious source of bleeding. The haemoglobin level was raised to 11 gm% after 3 units of red blood cell transfusion. The blood pressure was maintained above 100 mmHg systolic with the aid of l0 µg/kg/min of dopamine infusion. The oxygen saturation remained between 75 and 80% despite oxygen therapy. She improved gradually and dopamine was weaned off over the next few days. The patient was finally discharged home on the 16th postoperative day.

DISCUSSION

Pregnancy involves remarkable physiological changes. Many of these involve cardiovascular system directly or indirectly, increasing the demand on the heart, arterial and venous circulation. The resting cardiac output increases 30 to 40% with the maximum increase reached by the end of the second trimester[2]. 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[2].

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[3]. A fixed pulmonary hypertension not responsive to oxygen also carries a grave prognosis and may be an absolute indication to terminate the pregnancy[4]. When supplementary oxygen was given to our patient during induction, her oxygen saturation improved from 84 to 99% implying a reversible shunt. Oxygen is a pulmonary vasodilator which resulted in reducing the flow across the right to left shunt thus improved her oxygen saturation[5,6].

We obtained continuous measurement of blood pressure via an intra-arterial line but did not insert a pulmonary artery catheter to avoid catheter-induced arrhythmias, misdirection of catheter, thrombus formation, embolization and pulmonary artery rupture in the presence of pulmonary hypertension[7]. Instead, we inserted a central venous line, which will detect right heart failure as the right heart is ejecting against high pulmonary resistance[7] and to optimize the preload.

Various anaesthetic techniques for Caesarean section have been described, both regional and general anaesthesia. The risk of fall in systemic vascular resistance cannot be ruled out with regional technique[8]. The problems of general anaesthesia and positive pressure ventilation are decreased venous return and cardiac output due to raised intra thoracic pressure. Both can lead to an increase in the right to left shunt with resultant arterial hypoxemia. In our patient, we chose a general anaesthetic technique because the patient was on subcutaneous heparin. We minimised the risk of fall in systemic vascular resistance by using etomidate and atracurium as a slow bolus to minimize histamine release. Vasoconstrictors were not started prophylactically, as we wanted to avoid sudden hypertension and bradycardia, which can lead to a decrease in oxygen saturation[9].

Sevoflurane and isoflurane cause similar changes to systemic vascular resistance[10]. However, sevoflurane reaches steady state faster than isoflurane. Its onset and offset is also rapid because of low blood gas solubility. We chose to use sevoflurane because the effect of sevoflurane on systemic vascular resistance may be reversed more quickly than isoflurane or halothane. We avoided nitrous oxide because it is a potent pulmonary vasoconstrictor.

Our patient's general condition deteriorated on the second postoperative day. Her haemoglobin decreased from 11.5 gm% to 8.5 gm%. No obvious source of bleeding was found. We are not able to explain this drop in haemoglobin. This was accompanied by a rapid fall in oxygen saturation despite increasing inspired oxygen concentration. It was possible that the patient may have developed multiple pulmonary thrombosis or emboli. The thromboembolic phenomena have been associated with up to 43% of all maternal deaths in patients with Eisenmenger's syndrome. Naeye et al. found that in post-mortem studies of maternal congenital heart disease, there was widespread pulmonary thrombosis due to platelet aggregation. This would lead to rapid increase in pulmonary vascular resistance and worsening of the shunt[11]. There was no conclusive evidence for an thrombo-embolic phenomenon in our patient, (as no ventilation/perfusion scan was done) but this could explain the lack of improvement in the oxygen saturation with oxygen therapy in our patient. The use of heparin to prevent the thrombo-embolic phenomenon also carries a substantial risk of bleeding in the immediate postpartum period[13]. In our patient the evening dose of subcutaneous heparin was omitted and restarted on the first postoperative day. This was converted to warfarin from the third post-operative day. The complication of pulmonary thromboembolism was probably not prevented despite anti coagulation in our patient.

Pregnancy carries substantial maternal and fetal risks in patients with pulmonary hypertension and Eisenmenger's syndrome. Although pregnancy should be discouraged in women with Eisenmenger's syndrome, it can be a successful[12]. Prolonged bed rest, use of heparin, and oxygen therapy can produce satisfactory maternal and fetal outcome. If the patient requires an operation, either regional or general anaesthesia can be suitable. However these patients should be monitored closely in the postoperative period for up to a week, for worsening of the shunt and thrombo-embolic phenomenon as these complications can occur as late as third postoperative day, as seen in our patient and other reports[7,13].

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