



ORIGINAL RESEARCH PAPER

Anaesthesiology

PERIOPERATIVE MANAGEMENT OF PATENT DUCTUS ARTERIOSIS WITH EISENMENGER'S SYNDROME WITH SEVERE PULMONARY HYPERTENSION POSTED FOR NONCARDIAC SURGERY.

KEY WORDS: Eisenmenger syndrome, pulmonary hypertension, total abdominal hysterectomy.

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ABSTRACT

Victor Eisenmenger's initial definition of Eisenmenger's syndrome was redefined by Wood as 'the presence of high pulmonary vascular resistance associated with pulmonary artery hypertension at or close to systemic pressure associated with a reversed or bi-directional shunt at the aortopulmonary, interatrial or interventricular level'. Survival beyond 50 years is unusual, but patients may lead a relatively active and productive life in early adulthood and will therefore present from time to time for noncardiac surgery. Though theoretical risks of anaesthesia are considerable patients are known to do well with a variety of techniques, if pathophysiology of the disease is well understood. We describe the anaesthetic management of the patient with Eisenmenger's syndrome with abnormal uterine bleeding with multiple fibroids. She underwent a total abdominal hysterectomy after pre-operative stabilization. Anaesthetizing such a decompensated patient is an anaesthetic challenge.

CASE REPORT

A 35-year-old, 40Kg, Nulliparous lady was admitted with a diagnosis of abnormal uterine bleeding with multiple fibroids. She was a diagnosed case of PDA with a right to left shunt 14 years back, for which she used medication for initial few years. But now stopped using medication and having palpitations and exertional dyspnoea (NYHA class III).

Physical examination revealed clubbing without cyanosis. Vital signs are blood pressure (NIBP): 130/90mm Hg, pulse-90/min, SpO₂-85% on room air. On auscultation, there was loud P2 with pan-systolic murmur over the left sternal border with clear lung fields and bilateral equal air entry. Electrocardiogram (ECG) showed right ventricular hypertrophy with right axis deviation. Her haemoglobin was 9g/dl, and rest of haematological and biochemical investigations were unremarkable. Her echocardiography findings depicted dilated right atrium and right ventricle, moderate T.R., severe PAH (estimated RVSP 165+RAP), PDA with the bi-directional shunt. She was on T.Sildenafil 25mg and T.Ambrisentan 5mg. After 15 days of treatment, her echocardiography showed congenital heart disease, large PDA with the bidirectional shunt, dilated right atrium, right ventricle and pulmonary artery, moderate T.R., severe PAH (estimated RVSP 40+RAP), mild M.R. with an ejection fraction of 63%.

In view of multiple fibroids with abnormal uterine bleeding, she was taken for total abdominal hysterectomy with salpingectomy after explaining a high risk of perioperative cardiopulmonary events including nearly 10% chance of mortality related surgery to anaesthesia. The operative procedure was planned under epidural anaesthesia, and pulmonary vasodilators were continued on the day of surgery with adequate blood reserve. Intraoperative monitoring included ECG, SpO₂, invasive arterial pressure and central venous pressure. Under strict aseptic precautions, 20G epidural catheter was secured in L2 L3 in right lateral position. After administration of test dose, epidural anaesthesia was activated with 7ml of 2% lignocaine with 1:200000 adrenaline along with 1 ml of sodium bicarbonate. T8 sensory level was achieved after 15 minutes with adequate motor block. Epidural anaesthesia was maintained with 0.5% bupivacaine intermittently as per the recession of the sensory block. Adequate oxygenation was given with an O₂ mask along with judicious administration of I.V. fluids for maintenance and to replace the blood loss under CVP guidance. Intraoperative

vitals were maintained well.

During the postoperative period, adequate analgesia was maintained with fentanyl 25mcg/hr continuous infusion, and paracetamol 1gm I.V. TID and pulmonary vasodilators were continued. Patient was observed for three days in the post anaesthesia care unit and 2D Echo was repeated on the 3rd postoperative period, which showed pulmonary artery pressure of 125+RAP mm of Hg. She was shifted towards on 3rd postoperative day where her stay was uneventful and discharged on 21st post operative day.

DISCUSSION

Patients with Eisenmenger syndrome, pose challenge to anesthesiologists due to the inability to adapt to sudden changes in hemodynamics. 1 Maintenance of adequate filling pressure, systemic vascular resistance, optimal analgesia, and timely management of clinical deterioration are the key points. 2 Hematocrit > more than 60%, arterial oxygen saturation less than 80%, right ventricular hypertension, syncopal attack and fixed pulmonary hypertension not responsive to oxygen carries poor prognosis. 3

Sub-arachnoid block is associated with profound systemic hypotension due to the sympatholytic effects and resultant decreases in SVR in these patients with limited ability to augment R.V. stroke volume and cardiac output, and there is also an increased fraction of shunt reversal. 4

G.A. is associated with intubation and extubation response with multiple drug effects and less pain relief (activates sympathetic discharge) compared to central neuraxial block. 5 IPPV with high tidal volumes also augments pulmonary arterial pressure.

Titrated doses of epidurally administered local anaesthetics produce a lesser alteration of hemodynamics and provide adequate pain relief when compared to subarachnoid block. There is lesser manipulation of respiratory parameters when compared to General Anaesthesia. Sudden death in the perioperative period is due to fluid shifts, increased sympathetic tone, increased pulmonary vasoconstriction, and pulmonary thromboembolism that leads to worsening of R.V. failure. In order to minimize sympathetic activation, adequate pain control should be emphasized at all times, and sufficient sedation should be given in order to prevent agitation, especially while the patient is being ventilated.

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CONCLUSION

Our case report emphasizes that adequate and meticulous planning is the cornerstone of successful management of high-risk cases. Graded epidural anaesthesia is a safe technique in such cases, and emphasis should be laid on prevention of hypoxia, hypercarbia, acidosis and hypothermia, which aggravates pulmonary artery pressure.

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