Case report

Anesthesia for an elderly female with a rare congenital heart disease – A case report

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Abstract The long term survival of patients with Eisenmenger syndrome will make challenge for anesthetics; the risk during anesthesia of Eisenmenger syndrome may be increased. Combination of anesthetic considerations of Eisenmenger syndrome and anesthetic management of elderly patients must be considered.

We present a case of female patient 78 year old with long standing Eisenmenger syndrome. Her ECHO cardiography showed ejection fraction 57%, dextrocardia, large VSD 1.4 cm with right to left shunt with systolic pressure gradient 70 mmHg, moderate to severe TR, moderate MR, dilated and hypertrophied RV with preserved systolic function, hugely dilated right atrium, and severe pulmonary hypertension with pulmonary artery systolic pressure 125 mmHg.

The patient had future neck femur, arthroplasty under hemi spinal anesthesia. After one month she presented to us with dislocated joint, and hemiarthroplasty under hemispinal anesthesia was done again.

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1. Introduction

Only 15–25% of congenital heart disease patients survive into adulthood [1]. Approximately 90% of these children survive to adulthood due to Advances in prenatal diagnosis, and interventional cardiology; pediatric cardiac surgery, anesthesia, and critical care have resulted in survival of patients.

Eisenmenger syndrome (or **ES**, **Eisenmenger's reaction** or **tardive cyanosis**) is defined as the process in which a left to right shunt caused by a congenital heart defect in the fetal heart causes increased flow through the pulmonary vasculature, causing pulmonary hypertension [2] which in turn causes increased pressures in the right side of the heart and reversal of the shunt into a right. Eisenmenger syndrome is a cyanotic heart defect characterized by a long-standing intracardiac shunt caused by ventricular septal defect, atrial septal defect.

The long term survival of patients with **Eisenmenger** syndrome will make challenge for anesthetics; the risk during anesthesia of **Eisenmenger** syndrome may be increased. Combination of anesthetic considerations of Eisenmenger syndrome and anesthetic management of elderly patients must be considered.

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Common causes of heart failure in Eisenmenger syndrome are coronary artery disease including a previous myocardial infarction (heart attack), high blood pressure, atrial fibrillation, valvular heart disease, excess alcohol use, infection, and cardiomyopathy of an unknown cause [3,4].

Geriatric patients have decreased beta-adrenergic responsiveness and they experience an increased incidence of conduction abnormalities, bradyarrhythmias and hypertension. Fibrotic infiltration of cardiac conduction pathways makes the elderly patient vulnerable to conduction delay and to atrial and ventricular ectopy [5].

Complications of Eisenmenger syndrome are fainting spell, thromboembolism, and hypovolemia. Elderly patients also have an increased reliance on Frank-Starling mechanism for cardiac output. It is therefore important to consider fluid administration carefully. In the non compliant older heart, small changes in venous return will produce large changes in ventricular preload and cardiac output. Due to diastolic dysfunction and decreased vascular compliance, the elderly patient compensates poorly for hypovolemia. Similarly, exaggerated transfusion is also poorly tolerated [6].

In Eisenmenger syndrome there is pulmonary hypertension lung congestion and increased risk of pneumonia.

In elderly non cardiac patients there are increased risk of COPD, pneumonia, and sleep apnea which are very common among the elderly. Closing volume increases with age, and FEV1 declines 8–10% per decade due to reduced pulmonary compliance. PaO2 decreases progressively with age because of V/Q mismatch and anatomical shunt. All these pathophysiological changes may increase respiratory complications of Eisenmenger syndrome [7,8].

As regards Pathophysiology of the renal function due to aging, renal blood flow and kidney mass decrease with age. Serum creatinine level remains stable due to a reduction in muscle tissue. Impairment of sodium handling, concentrating ability and diluting capacity predisposes elderly patients to dehydration and fluid overload. Reduced renal blood flow and decreased nephron mass increase the risk of acute renal failure in the postoperative period. Eisenmenger syndrome has renal problems, which may increase risk of renal impairment when accompanied with the pathophysiological changes in elderly patients [7,8].

Dosage requirements for local and general anesthetics are reduced. Administration of a given volume of epidural anesthetic results in a more cephalic spread, having though a shorter duration of sensory and motor block. Elderly patients take more time to recover from general anesthesia especially if they were disoriented perioperatively; however, there is a risk of thromboembolism and stroke in Eisenmenger syndrome and Geriatric patients experience varying degrees of delirium. They are sensitive to centrally acting anticholinergic agents. The incidence of delirium is less with regional anesthesia, provided that there is no additional sedation [9–11].

The circulating level of albumin which is the main plasma binding protein for acidic drugs decreases with age. On the other hand, the level of α-1 acid glycoprotein the binding protein for basic drugs increases. The effect of aging on pharmacokinetic depends upon the drug used.

The decrease in total body water leads to a reduction in the central compartment and increased serum concentrations after a bolus administration of a drug. On the other hand, the increase in body fat results in a greater volume of distribution, thus prolonging drug action.

Drug metabolism could probably be altered by the aging effect on hepatic or renal function.

The elderly are more sensitive to anesthetic agents and generally require smaller doses for the same clinical effect, and drug action is usually prolonged. A person with Eisenmenger syndrome is paradoxically subject to the possibility of both uncontrolled bleeding due to damaged capillaries and high pressure, and random clots due to hyper viscosity and stasis of blood. Coughing hemoptysis and bleeding may lead to iron deficiency, anemia, coagulation defects which may interfere with regional anesthesia [9–11].

Anesthetic management of an elderly **Eisenmenger** syndrome should include participation of anesthesiologists, cardiologists, intensivists, and surgeons.

Aim of the anesthetic technique should include avoidance of increase of pulmonary pressure, avoidance of volume over load, light anesthesia and patchy regional anesthesia, which can lead to increase the systemic vascular resistance, maintenance of euvolemia and care of the renal function, management of tachyarrhythmia, or bradyarrhythmia, venous line care of any air bubbles, bleeding tendency and intraoperative bleeding, coagulopathy and regional block, infection, and the endocarditis antibiotics prophylaxis, positioning (prone, Trendelenburg), oxygenation and prevention of hypoxia in these already cyanotic patients [11].

When an elderly patient with long standing Eisenmenger syndrome needs an emergency anesthesia for hemiarthroplasty the anesthetic management becomes a challenge. Anesthetic consideration must include anesthesia for pulmonary hypertension, heart failure, ischemic heart disease, dysrhythmias, hypertension, endocarditis, cardiomyopathy, bleeding, thromboembolism, chest infection, and kidney problems. It is more complicated when failure of operation occurred and another operation was needed within one month as in our present case [12].

Case of female patient 78 year old with long standing Eisenmenger syndrome had future neck femur, arthroplasty under hemi spinal anesthesia after one month recurrence of fractures, and hemiarthroplasty under hemi spinal anesthesia again.

1.1. Preoperative preparation

1.1.1. History

A 78 year old female was admitted with fracture left neck of femur for hemiarthroplasty operation. The patient is known case of congenital heart disease. No history of dyspnea, orthopnea, paroxysmal nocturnal dyspnea, syncope, chest pain or stroke. There is history of lower limb edema treated with furosemide 20 mg once daily and tablet digoxin 0.25 µg daily.

1.1.2. Physical examination

On examination blood pressure was 120/60, heart rate was 100 irregular, and congested neck veins were up to the angle of the mandible. There are central cyanosis, and bilateral clubbing of upper and lower limb with no lower limb edema. Chest examination is free. Cardiac examination revealed Dextrocardia, and harsh pansystolic murmur over the right parasternal area. Another pansystolic murmur over the apex of the heart propagating to the right axilla was osculated.

1.1.3. Investigations before the first operation

Electrocardiogram showed atrial fibrillation rate 105 per minute, right bundle branch block and poor progression of 'R' wave in V1-V leads. Chest X-ray was obtained unremarkable.

ECHO cardiography showed ejection fraction 57%, dextrocardia, large VSD 1.4 cm with right to left shunt with systolic pressure gradient 70 mmHg, moderate to severe TR, moderate MR, dilated and hypertrophied RV with preserved systolic function, hugely dilated right atrium, and severe pulmonary hypertension with pulmonary artery systolic pressure 125 mmHg.

Laboratory investigations: Complete blood picture, renal and liver function tests, coagulation profile, and abdominal ultrasonography were within normal limits, and Na and k, exclude hypokalemia and digoxin toxicity.

1.2. Preoperative preparation

The patient started Sildenafil therapy 25 mg tablets every 12 h for three days preoperative.

Furosemide 20 mg was given once daily and tablet digoxin 0.25 µg treatment was taken before operation.

Ampicillin and gentamicin were administered for prophylaxis against bacterial endocarditis.

An 18G intravenous cannula was inserted and secured in left hand vein and left external jugular vein and 16G cannula was inserted.

DC shock was prepared for uneventful dysrhythmias.

1.3. Intraoperative management

1.3.1. Monitoring

ECG leads, non invasive blood pressure, and pulse oximeter were attached for continuous monitoring. Right internal jugular vein triple lumen central venous line was inserted under local anesthesia.

20G Arterial cannula was inserted and secured in left radial artery after modified Allen's test for invasive blood pressure monitoring.

The baseline parameters were pulse rate: 100/min and BP: 110/70 mmHg SpO2 87% on oxygen by face mask. central venus pressure was 10 cm H_2O.

1.3.2. Anesthetic management

After preloading with 250 ml of Ringer solution, patient was positioned in left lateral position. Spinal anesthesia was done, and spinal needle 22G was inserted in L4-5 interspace under complete aseptic precautions after local infiltration of skin with 2% lignocaine. We injected 2 ml of 0.5% bupivacaine heavy and 25 µg fentanyl.

1.4. Intraoperative events

Patient developed hypotension (90/50) (a fall of more than 20% of the baseline level) at 5 min after institution of spinal anesthesia, and it was successfully treated with bolus doses of injection 6 mg ephedrine each and titrated intravenous Ringer's solution. Pressure was maintained in same range throughout the procedure. Central venous pressure was maintained in 10 cm water with ringer solution. The surgical procedure was done in lateral position and was completed in 50 min; the total fluid output was 600 cc urine and 500 cc blood.

500 cc crystalloids, 250 ml of blood and two units of fresh frozen plasma were infused to the patient.

After surgery, the patient was transported to intensive care unit for continuous hemodynamic and SpO_2 monitoring with oxygen via a face mask. Postoperative ECHO cardiology was as before surgery.

On the third day post operative the patient suffered from an attack of convulsion which lasted for 1 min followed by disturbed level of consciousness; the patient was put on mechanical ventilator; and CT brain study was done with no abnormalities. 12 h later the patient regained consciousness. Subsequent recovery was uneventful and satisfactory. The patient was discharged from the hospital on 10th day postoperative.

After one month recurrence of fracture occurred again repeating investigation revealed the same as first time, and Preoperative and intraoperative management was the same as the first operation.

Intraoperative events were as follows:

Hypoxia developed with oxygen saturation 87% and was rapidly managed by oxygen face mask.

Another attack of hypotension was developed (90/50) after institution of spinal anesthesia, and it was successfully treated with bolus doses of injection 6 mg ephedrine each and titrated intravenous Ringer's solution. Postoperative recovery was satisfactory. The patient was discharged from the hospital on 10th day postoperative.

2. Discussion

A primary goal of anesthetic management in this case is to minimize increases in pulmonary vascular resistance and to maintain systemic vascular resistance. Abrupt increases in pulmonary vascular resistance may precipitate either acute right ventricular failure or oxygen desaturation followed by decreased cardiac output. Severe bradycardia may occur with progression to cardiac arrest.

Prevention and treatment of pulmonary hypertensive crisis include hyperventilation, correction of acidosis, avoidance of sympathetic nervous system stimulation, maintenance of normothermia, minimization of intrathoracic pressure, and use of inotropic support.

Regional anesthesia may be an acceptable alternative to general anesthesia. However, spinal may produce unacceptable decreases in systemic vascular resistance in patients, and this action could exacerbate right to left shunting.

We chose to do low dose spinal anesthesia to decrease afterload, prevented myocardial depression. Fluid management is also critical and preloading the patient in the preoperative period is not desirable because it may precipitate a congestive heart failure. In our case fluid overloading was prevented by titrating the fluids to maintain a CVP of 10 cm H_2O. Invasive blood pressure monitoring and central venous pressure monitoring were used to facilitate early recognition of blood pressure changes and to guide fluid therapy and maintain normovolemia.

The major risks during the intraoperative and postoperative period are similar to the risks described above. These risks include bleeding, dysrhythmias, and thromboembolic events,

and oral pulmonary vasodilators such as sildenafil may be beneficial [12].

Similar case study was done by B. Ghai, V. Mohan, M. Khetarpal andN. Malhotra [13]. They reported the anesthetic management for cesarean section of a 27-year-old multigravid female at 35 weeks' gestation with Eisenmenger's syndrome. Titrated epidural anesthesia was administered with incremental doses of 2% lidocaine. Intraoperative course was uneventful except for an episode of hypotension immediately after delivery of the baby, which was managed successfully. They conclude that carefully titrated epidural anesthesia may be effective for patients with Eisenmenger's syndrome for cesarean section.

Robert S. Holzman, Charles D. Nargozian, Richard Marnach, and Curtis O. McMillan [14] presented two cases of patients with palliated cyanotic congenital heart disease received Epidural anesthesia. The first case is a 17 year-old, 54kgs boy with a d-transposition of the great arteries, hypoplastic right ventricle, ventricular septal defect (atrioventricular canal type), atrial septal defect, small patent ductus arteriosus, and an overriding tricuspid valve. The patient took digoxin and furosemide and he presented for a right inguinal herniorrhaphy. The second one is a 30-year-old, 50-kgs woman with a history of tetralogy of Fallot with pulmonary atresia, an interatrial communication, and large aortopulmonary collaterals presented for exploratory laparotomy for excision of an ovarian cyst. The patient took tetracycline, digoxin, and furosemide. Prior surgical procedures included a central shunt, left pleurectomy, odontectomy, and bilateral tubal ligation. Surgery proceeded uneventfully once anesthesia was established; Surgery lasted for 75 min in patient 1, and 50 min in patient 2.

In a study of Martin et al. [15], they had a literature identified 57 articles describing 103 anesthetics in patients with Eisenmenger's syndrome. An additional 21 anesthetics were identified in patients receiving regional anesthesia for labor, and they found that Overall perioperative mortality was 14%; patients receiving regional anesthesia had a mortality of 5%, whereas those receiving general anesthesia had a mortality of 18%. This trend favored the use of regional anesthesia but was not statistically significant. A better predictor of outcome was the nature of the surgery (and presumably the surgical disease). Patients requiring major surgery had mortality of 24%, whereas those requiring minor surgery had mortality of 5% (P < .05). Patients in labor receiving regional anesthesia had a mortality rate of 24%, and most of these occurred several hours after delivery. They concluded that most deaths probably occurred as a result of the surgical procedure and disease and not anesthesia. Although perioperative and peripartum mortalities are high, many anesthetic agents and techniques have been used with success.

In another study of Gurumurthy et al. [16], they published a case of 22-year-old primigravida weighing 46 kgs, known case of Eisenmenger's syndrome at 37 weeks of gestation, who was scheduled for an elective cesarean section. At the age of 15 years, a cardiac catheterization was performed, which revealed a large mid-muscular VSD with severe pulmonary arterial hypertension (120/50 mmHg). She was explained about the existing cardiac condition and advised to avoid strenuous work. They concluded that although pregnancy must be discouraged in women with Eisenmenger's syndrome, and it can be successful. Safe anesthetic management of these patients requires meticulous preparation and familiarity with all the anesthetic agents to maintain the cardiovascular stability. Then, early extubation should be avoided in such patients because, invariably, they may go for worsening of shunt and thromboembolic phenomena as these complications can occur as late as the third post-operative day, as seen in our patient and other reports. They recommended a general anesthetic technique with maintenance of hemodynamics as close to normal as possible, with adequate control of pain and early initiation of thromboprophylaxis for successful management of similar cases.

In another case study for Lipi Mishra et al. [17], they described a case of a pregnant patient with a large ventricular septal defect (VSD) and pulmonary artery hypertension, presented to the hospital and underwent elective cesarean section under epidural anesthesia and postoperative analgesia. The procedure was uneventful till the patient was discharged on 10th day.

3. Conclusion

When an elderly patient with long standing Eisenmenger syndrome needs an emergency anesthesia for hemiarthroplasty the anesthetic management becomes a challenge. Proper understanding of the pathophysiology of the disorder and Careful anesthesia planning, pre-operative assessment, intraoperative and postoperative management can help in reducing the mortality. Low-dose intrathecal bupivacaine and fentanyl provided the advantages of spinal and should be under invasive cardiovascular monitoring.

Conflict of interest

There is no conflict of interest.

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