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# Adult eisenmenger syndrome for emergency exploratory laparotomy: Anaesthesia management without invasive monitoring

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#### Abstract

The Eisenmenger syndrome refers to condition when long standing congenital heart defect with an intracardiac communication leads to pulmonary hypertension that causes reversal of flow and cyanosis. It isn't uncommon for patients with this syndrome live upto adulthood with or without corrective surgery. The requirement of Invasive monitoring or any other invasive procedures should be weighed against usefulness of aquired information and associated complications. The question arrises are Invasive Techniques mandatory for such patients?? This article reviews the anaesthesia considerations and management for a case of adult eisenmengers syndrome came with self inflicted stab wound over abdomen taken in for an emergency exploratory laparotomy.

Keywords: Anaesthesia, eisenmenger, non-cardiac

### Introduction

Pulmonary hypertension with congenital heart disease, is seen in large systemic-topulmonary communications, such as ventricular septal defect (VSD) and patent ductus arteriosus. On progression, it leads to shunt reversal a condition termed Eisenmenger syndrome <sup>[1]</sup>. Victor Eisenmenger in 1897 coined the term Eisenmenger complex, which included large ventricular septal defect (VSD) and pulmonary hypertension <sup>[2]</sup>. Wood redefined this in 1958 as pulmonary hypertension with reversed or bidirectional shunt, associated with septal defects or patent ductus arteriosus <sup>[3]</sup>. Due to advances in medical management eisenmenger syndrome is becoming very rare thus ocasionaly an Anesthesiologist can come across such patients presenting for non-cardiac emergency surgeries and management of such cases are challenging for Anesthesiologist if it is under General anesthesia maintaining hemodynamic stability is an uphill battle.

# **Case Report**

We Report a case of 38year old male came to emergency department in a conscious state, brought by his brother and police Assistant sub-inspector with a history of Self-inflicted knife stab wounds over abdomen. His ultrasonography report was showing no significant abnormality. Primary stiches were taken dressing was done. Patient was shifted for a CT scan; the report suggestive of mild hemoperitoneum collection, patient was then Shifted to OT for an emergency exploratory laparotomy.

Patient was shifted to Operation theatre. Patient was awake conscious and oriented; he came with complains of pain in abdomen and mild cough. he gave history dyspnea on exertion (NYHA grade II) and poor effort tolerance of one floor, as he is a known case of congenital heart disease since childhood. On further evaluation it was found out that 8 years back his 2DEcho and cardiac catheterization had been performed which revealed Large VSD (ventricular septal defect) with eisenmengerization with bidirectional shunt and patent ductus arteriosus. Patient was on medical management and was taking tab.Deriphyllin tab. Amiloride + tab.Hydrochlorothiazide, tab.sildenafil, tab.digoxin 0.25mg since 10 years. Patient gave no other surgical history.

Our patient was second born child of non- consanguineous marriage born with normal fullterm vaginal delivery with low birth weight and history of NICU admission for 1 month, He gave a history of cyanosis since birth during crying, running or climbing stairs. He had delayed milestones. Our patient was thin built. In family history he had a younger sibling who also had congenital heart disease and died at the age of 10yrs. Psychiatry evaluation was done for the patient, he refused on his intension of hurting himself or any suicidal ideation also showed no signs of depression.

Pre operative vitals were showing tachycardia with HR 110/min blood pressure 130/80mmHg. Room air saturation was 75% which increased to 90% with oxygen support. Physical examination revealed grade II clubbing. No peripheral edema or cyanosis was seen.

Cardiac examination revealed Loud P2 with Pansystolic murmur heard over mitral area, Thrill noted over Mitral, tricuspid, aortic area. Palpable P2 with heaving apex was present. Electrocardiogram revealed right axis deviation with T wave inversion in leads V2-V4. 2dimentional echocardiography was not available in an emergency but cardiology opinion was taken.

His haemoglobin was 16.8gm% with platelets 1,96,000/cmm, Renal and liver function tests, serum electrolytes, blood sugars, coagulation profile were within normal limits. Arterial blood gas analysis was done on room air - pH=7.411, pCO2=37.2mmHg, pO2= 77.8mmHg, SaO2=95.5%

In the operation theatre, standard monitors like Pulse oximeter. non-Invasive blood pressure and electrocardiography used. Peripheral were venous cannulations of 20G and 18G were secured. Central venous access lines were kept ready. After 3 min of preoxygenation, SpO<sub>2</sub> increased to 100%. The pre-induction blood pressure (BP) was130/90 mmHg. Intravenous induction was carried out with a low dose of thiopentone sodium 150 mg and fentanyl 30µ gm titrated to anesthetic and hemodynamic effects. The patient was intubated endotracheally with suxamethonium 100mg. Anaesthesia was maintained with 50% oxygen in air and sevoflurane. The neuromuscular blockade was achieved with vecuronium 5 mg. The end tidal carbon dioxide (EtCO<sub>2</sub>) was maintained between 32 and 35 mmHg.

Exploratory laparotomy with primary closure of peritoneal breach with peritoneal wash and deflation of the large intestine with flatus tube was performed. Blood loss was estimated to be about 150-200 mL. The patient remained hemodynamically stable throughout the procedure, Neuromuscular blockade was reversed with neostigmine 3 mg and glycopyrrolate 0.4 mg and the patient was extubated after fully awake and shifted to the post-operative ward with oxygen support for observation.

2-dimentional echocardiography (2D echo) was done postoperatively which revealed large malaligned subaortic ventricular septal defect of 10 mm. Bidirectional shunt across the defect. Right atrium and Right ventricular dilated with severe Pulmonary hypertension measuring 130 mmHg. Post-operatively, the patient was observed for 7days for any complaints or complications but our patient had uneventful post-op. hospital stay and was hemodynamically stable.

# Discussion

Congenital heart diseases can be functionally classified into Acynotic (left to right shunts) and cyanotic (right to left shunt). A left to right shunt exists when oxygenated blood from the left atrium, left ventricle, or aorta transits to the right atrium, right ventricle, or the pulmonary artery Thus the lung receive all the deoxygenated blood from the systemic venous return (effective pulmonary blood flow; amount of deoxygenated blood that is carried to lungs to be oxygenated) plus the volume of fully oxygenated blood that is shunted through the defect. This results in volume overload of one or more cardiovascular chambers or structures depending on the location of the defect. If the defect is large and nonrestrictive, there is both increased flow and transmission of near systemic pressure to the pulmonary vascular bed. Over time, this can lead to irreversible changes in the pulmonary vascular bed, leading to increased pulmonary vascular resistance and associated pulmonary artery hypertension. If the pulmonary artery pressure is at systemic levels, there may be reversed (right to left) or bidirectional shunting at the level of the defect (Eisenmenger syndrome)<sup>[4]</sup>.

In Eisenmenger syndrome the use of invasive monitoring is controversial and the risk of complications must be weighed against the usefulnes of acuired information. The intraarterial catheterization may be associated with a higher incidence of post-cannulation thrombus formation as these patients are polycythemic. Insertion of central venous catheter has probable risk of infection and paradoxical air embolus <sup>[5]</sup>. The complications of pulmonary catheterization are pulmonary arterial rupture in the presence of pulmonary hypertension apart from arrhythmias and systemic embolization <sup>[6, 7]</sup>. As our surgical procedure was short we avoided invasive monitoring.

Adults with CHD are more likely to be living with their parents and to develop a variety of psychosocial issues <sup>[8]</sup>. Consequently, psychological preparation of patients for surgery is important Premedication with anxiolytics and hypnotics must be undertaken very cautiously because hypoventilation and hypercapnia may produce deleterious increases in pulmonary vascular resistance, particularly, in patients with underlying pulmonary hypertension or systemic to pulmonary shunts <sup>[9]</sup>.

In patient with Eisenmenger's syndrome most deaths probably occurred as a result of the surgical procedure and disease and not anesthesia. Although perioperative and peripartum mortalities are high, Both Regional and General anaesthesia techniques have been used with success in literature <sup>[10]</sup>.

Considering the type of surgery, we opted for general anaesthesia, we combined a short-acting i.v. opioid such as fentanyl in addition to a low dose of induction agent such as thiopentone sodium with low dose ketamine and sevoflurane as inhalational agent <sup>[11]</sup> However thiopentone causes a decrease in systemiv vascular resistance (SVR) but the effect is dose-dependent <sup>[1]</sup>. Ketamine has theoretical advantages over barbiturates as an induction agent, as it does not reduce SVR but causes increase in the heart rate which is undesirable <sup>[12]</sup>. We avoided the use of nitrous oxide as it causes pulmonary vasoconstriction<sup>[13]</sup>. Although volatile agents are known to decrease SVR with less effect on the pulmonary system. Sevoflurane was preffered due to less pungency allowing for inhalational induction and patient tolerance. Furthermore, it has known beneficial effects on pulmonary bronchial reactivity, which can exacerbate pulmonary resistance <sup>[14]</sup>. For muscle relaxation we used Vecuronium. Pain control is essential to avoid chatecholamine surge thus we used low dose fentanyl and nonsteroidal anti-inflammatory drugs [14]. Pateint was reversed and extubated when fully awake as early extubation should be avoided in such patients because,

invariably, they may go for worsening of shunt and thromboembolic phenomena.

Post operatively patient was given oxygen support with a

simple face mask and we kept follow up for post- operative complications till 7 days our patient had an uneventful hospital stay.



Fig 1: Original Image: Chest X-ray PA view of the patient showing prominent pulmonary vasculature, Enlarged main pulmonary artery.

# Conclusion

Management of such patient under General Anesthesia is challenging thus for Successful outcome of these patients methodical approach to evaluation, through preparation and meticulous use of anesthetic agents to maintain the cardiovascular stability and post operative analgesia is crucial.The requirement of invasive intervations may not be mandatory for short surgical procedures.

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