

# Non-Cardiac Surgery in Eisenmenger Syndrome: A Case Report and Review of Literature

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## 1. Introduction

Eisenmenger Syndrome (ES) is an uncommon complication of many congenital heart diseases, such as ventricular and atrial septal defect, and single ventricle [1]. With the increase in modern technologies in medical science, more and more patients with congenital heart diseases are surviving into adulthood and some even into the sixth and seventh decade [2]. As a result, many of them will undergo non-cardiac surgery in their lifetime [3]. Such patients of Eisenmenger syndrome are known to be at very high risk when undergoing Non-Cardiac Surgery (NCS) [4]. These patients are usually comparatively younger and are at lower risk of atherosclerotic coronary artery disease, but are at greater risk of arrhythmias, heart failure, paradoxical embolism or defect-specific complications [5].

This high risk of NCS is due to extent of the surgical procedure, severity of pulmonary hypertension, severity of Tricuspid regurgitation, right ventricular dysfunction, and additional acquired cardiac & systemic diseases such as systemic heart diseases, and renal dysfunction. These patients are very vulnerable to alteration in hemodynamic status induced by anaesthetics or surgery. This includes a minor fall in systolic blood pressure that can increase right-to-left shunting and possibly potentiate cardiovascular collapse. Additional surgical risks include increased risk of bleeding due to platelet dysfunction, arterial and venous thrombosis secondary to increased blood viscosity, paradoxical embolus and arrhythmia.

## 2. Case Report

A 51-year-old patient presented in the OPD with the complaints of swelling in B/L Inguinal region for last 6 months (L>R). He was a known case of Ventricular septal defect, Eisenmenger syndrome, laevocardia and pulmonary hypertension.

On examination he was found to be having B/L Inguinal hernia (L>R).

- He was admitted for evaluation and management (Inguinal hernioplasty).
- At the time of admission his pulse rate was 72/min, BP was 120/80 and SPO<sub>2</sub> was 95%. His ultrasonography scrotum showed B/L Inguinal hernia (L>R).
- His ECG showed Sinus Rhythm, Borderline PR interval, Rt ventricular hypertrophy and Rt ventricular strain pattern.
- His 2D Echo revealed mildly dilated size of Rt Atrium and Rt ventricle with normal Rt ventricular function, Dilated Pulmonary artery, Mod TR and severe PAH. Normal LA size.
- Normal LV size and fair LV systolic function. LVEF 50%. No diastolic relaxation abnormality. No MR and No AR. No visible clot/mass/ PE/ vegetation.
- His INR was 1.24 and Hb was 21. Physician and a Cardiologist reference was taken for his fitness for surgery. Pre-anaesthetic check-up was done by our senior anaesthetist.
- High risk consent (Informed consent) for the surgery was taken. It was decided to go for surgery on left side only considering his overall clinical condition.
- Pt was then taken for surgery under spinal anaesthesia, all the parameters were being monitored during the surgery and surgery was completed uneventfully.
- Pt was then shifted to ICU for postoperative care. At that time his SPO<sub>2</sub> was 99% on Nasal oxygen 3 litres/min.

On 1<sup>st</sup> postoperative day in the morning his vitals were stable. But in evening at about 4 pm, patient suddenly developed blurring of vision, slurring of speech, Ptosis left eye and slight weakness on right side. His MRI Brain was done which showed acute ischemic infarcts in mid brain and left thalamus with multiple chronic infarcts. Neurologist and Cardiologist opinion taken and advise followed. Hydration was increased and phlebotomy (two units) done.

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On 2<sup>nd</sup> postop day blurring of vision was bit clear but Ptosis and slurring of speech was same. Again, one-unit phlebotomy done and after that in evening patient was feeling better. There was no blurring of vision, no weakness but there was slight Ptosis. His Hb came to 17.9 from 21, and Hct came to 56.7 from 65.

On 3<sup>rd</sup> postop day, his vitals were stable and after stabilisation on 4<sup>th</sup> postoperative day he was discharged. He was called again for follow up on 8<sup>th</sup> postop day. His stitches were removed, and he was fit and fine.

### 3. Discussion

Eisenmenger syndrome is one of the leading causes of perioperative death (up to 19%) in patients undergoing Non-Cardiac Surgery (NCS) [6]. Some studies have reported the mortality rate even up to 30% [4]. Perioperative risk and outcome depend on urgency, duration of surgery, anaesthesia used and underlying pathology. Profound hemodynamic variability such as heart rate, blood pressure, volume status, oxygenation, and neurohormonal activation adds an extra stress on an already abnormal cardiopulmonary system [7]. Haematocrit >60%, arterial oxygen saturation <80%, right ventricular hypertension, syncopal attack and a fixed pulmonary hypertension not responsive to oxygen carries poor prognosis [8].

The Eisenmenger Syndrome (ES) heart is highly preload dependent; fluid shifts can lead to a significant fall in cardiac output. Decreases in Systemic Vascular Resistance (SVR) can enhance right-to-left shunt, worsening cyanosis and precipitating cardiovascular collapse. Increases in SVR can lead to a decrease in ventricular function, especially if associated with a sudden rise in PVR (PH crisis) [9]. This precarious physiology requires meticulous peri-operative management, with maintenance of SVR and ventricular contractility and avoidance of excessive blood loss and intravascular volume depletion. In cases of primary inguinal hernia repair, intraoperative fluid shifts are limited with experienced anaesthetic guidance, and pre-operative dehydration was avoided by allowing the patient to drink clear fluids until 2 hours prior to anaesthesia.

Only very few generalized guidelines exist for the preoperative evaluation of patients of Eisenmenger syndrome undergoing NCS [1]. The guidelines for preoperative cardiovascular evaluation have been recently reviewed by Eagle et al., however, these guidelines were intended for adults with acquired rather than congenital heart disease [10]. Preoperative evaluation of Eisenmenger syndrome patients should be performed by physicians experienced in managing these complex cases. Both a detailed history and a physical examination are necessary in addition to appropriate preoperative testing. Perloff and others suggested that preoperative therapeutic phlebotomy with fluid replacement is indicated in patients with hyper viscosity syndrome and a haematocrit of more than 65% as this will improve the haemostatic defect and reduce the risk of intraoperative bleeding [2]. Both general and regional anaesthesia technique have been used [11].

Numerous studies have suggested that there is no one best myocardial protective anaesthetic technique in these complex patients [12]. So, the choice of anaesthetics and intraoperative monitors is best left to the discretion of an experienced anaesthesia care team [13]. Eisenmenger pts pose a difficult challenge as they have lost the ability to adapt to sudden changes in hemodynamic status because of fixed pulmonary vascular disease [14]. As a result, a minor fall in systemic blood pressure can increase right to left shunting and potentiate cardiovascular collapse [15]. So, the basic principle of any anaesthetic technique chosen needs to maintain both cardiac output and systemic vascular resistance [16]. Although epidural anaesthesia has been successfully employed for minor surgeries such as tubal ligation and caesarean section, the resulting sympathetic blockade and decrease in both preload and afterload may be very hazardous, so general anaesthesia is preferable for these pts [16,17]. Periodic arterial blood gas determinations facilitate assessment of acidosis, hypercarbia and hypoxia, which can increase the pulmonary vascular resistance and increase right to left shunting.

Volume depletion and prolonged fasting should be avoided before surgery and all intravenous lines should be equipped with a device to filter air bubbles to prevent paradoxical air embolism. Alpha adrenergic agonist should be used if there is systemic arterial hypotension, and hypovolemia should be treated with intravenous volume replacement. Blood loss should be minimized. Prevention of thromboembolism should be encouraged by early ambulation and if prolonged immobilization is anticipated subcutaneous administration of heparin should be given.

The postoperative care of cardiac pts undergoing NCS is as important as the preoperative evaluation and intraoperative management. Eisenmenger pts should be observed on a monitored bed in ICU because of their predisposition to develop ventricular and supraventricular tachycardia. In addition, depending on the co-morbid conditions, they might be prone to bradyarrhythmia and myocardial ischemia [3]. Precautions must be taken to prevent venous stasis by early ambulation and by applying effective elastic stocking or periodic pneumatic compression. In addition, every attempt should be made to prevent hypovolemia; thus, meticulous attention to fluid balance is essential. Furthermore, adequate pain management is crucial because the stress of postoperative pain can result in adverse hemodynamic state and possibly hypercoagulable state [10].

### 4. Conclusion

In conclusion adult patients with Eisenmenger syndrome require special care while undergoing NCS, directed toward ameliorating their preoperative status and avoiding complications related to Eisenmenger syndrome. A thorough, multidisciplinary pre-assessment and meticulous perioperative management by an experienced anaesthetist should be presumed to be the standard of care [18]. Such patients should always be treated in hospitals having full cardiac backup. Finally, the patient, family and the treating teams should be prepared for an Intensive Care Unit stay

and prolonged post-operative ventilation [19].

### Authors Contribution

Corresponding author is the main author and 2nd author contributed in surgery and taking care of patient.

### Ethics Approval

Proper approval has been taken from the Ethical committee of the hospital.

### Consent for Publication

Written consent has been taken from the patient for publication.

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