Case Report

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Anaesthesia Section

Anaesthetic Management of a Case of Eisenmenger Syndrome undergoing Non Cardiac Surgery: A Case Report

NANDITHA PADIKKASU[1], PUSHPARANI ANAND[2], SHALINI THIYAGARAJAN[3]

ABSTRACT

Eisenmenger syndrome is a severe, irreversible complication of congenital heart defects, resulting in large anatomical shunts and leading to Pulmonary Arterial Hypertension (PAH). Patients with Eisenmenger syndrome are complex; when they require anaesthesia for a surgical procedure, they pose a greater risk and present a challenge for the anaesthetist due to compromised haemodynamics. Hereby, authors present a case of a 31-year-old female with Eisenmenger syndrome, characterised by a large Ventricular Septal Defect (VSD) with bidirectional shunting and severe PAH, who is currently symptomatic and underwent an elective open appendicectomy. The authors utilised various modalities, including appropriate haemodynamic monitoring, smooth induction of general anaesthesia, and a regional block to manage postoperative pain. These strategies were employed to maintain the patient's baseline homeostasis throughout the preoperative, perioperative and postoperative periods, ensuring that there was no compromise in the patient's existing condition. Overall, the patient's duration of hospital stay was uneventful and she was discharged shortly thereafter.

Keywords: Eisenmenger complex, Hypertension, Pulmonary arterial hypertension, Rocuronium, Ropivacaine, Sildenafil

CASE REPORT

A 31-year-old female of low socio-economic status, with a known history of Eisenmenger syndrome since infancy, presented with a two-day history of abdominal pain. The pain was dull and aching, initially starting near her belly button and progressively localising to the right lower abdomen. It was aggravated by coughing and movement, associated with two episodes of bilious vomiting and a reduced appetite, and was not relieved by any medication or rest. Following baseline investigations and a Contrast-Enhanced Computed Tomography (CT) of the abdomen, she was diagnosed with acute appendicitis and planned for open appendicectomy.

Routine laboratory investigations indicated a haemoglobin level of 14.9 g/dL (normal range: 12-16 g/dL) and a haematocrit of 54%. Electrolytes, renal and liver function tests, and the coagulation profile were within normal limits. Arterial Blood Gas (ABG) analysis revealed a pH of 7.37, a pCO_2 of 42 mmHg, a pO_2 of 33.8 mmHg, and an HCO_3 of 24.4 mEq/L.

Her clinical history revealed a large VSD with bidirectional shunting and severe PAH, diagnosed at birth, with a progressively worsening trend in severity since. She managed her symptoms conservatively with regular cardiac evaluations. The routine medications taken by the patient included T. Lasilactone (20/50 mg), T. Digoxin (0.2 mg), and T. Sildenafil (20 mg) for pulmonary hypertension. On examination, she appeared thin, with a Body Mass Index (BMI) of 16.2 kg/m^2. She exhibited New York Heart Association (NYHA) Class 3 exertional dyspnoea. Other significant medical history included a previous Intensive Care Unit (ICU) admission eight years ago for chest pain, breathlessness, and cyanosis. During that admission, she was diagnosed with significantly elevated haemoglobin and haematocrit, for which she was managed with phlebotomy; however, medical records from that time were unavailable.

Upon presentation in the emergency room, her vital signs were stable, with a blood pressure of 110/70 mmHg, a heart rate of 90 bpm, and oxygen saturation ranging from 79% to 85% on room air. The primary disadvantage was the unavailability of past medical records to confirm the previous diagnosis. Therefore, a 2D echocardiogram was performed, which confirmed the presence of a large VSD with bidirectional shunting and severe PAH. The right ventricle was hypertrophied and enlarged, with dilation of the right atrium and the main pulmonary artery. Patent Foramen Ovale (PFO) was also noted, but the left ventricle had a normal ejection fraction of 65%.

The CECT of the abdomen further revealed a solitary right kidney with an inflamed appendix. Given the risk of appendiceal perforation, the patient was taken for surgery after a thorough preoperative assessment, classified as American Society of Anaesthesiologists (ASA) III.

Anaesthetic Management

On the day of surgery, the patient was transferred to the operating room. In addition to routine monitors, an arterial line was placed in the left radial artery for invasive blood pressure monitoring. A wide-bore IV catheter (18G) was inserted in the right upper limb.

Premedication included Inj. Ondansetron 4 mg and Inj. Midazolam 1 mg. Due to her underlying pulmonary hypertension and the risk of tachycardia, Inj. Glycopyrrolate was avoided. Opioid induction was chosen to mitigate the intubation response and provide a reversible hypnotic effect. The induction agents included Inj. Fentanyl 160 µg and Inj. Thiopentone 175 mg intravenously. Neuromuscular blockade was achieved with Inj. Rocuronium 30 mg and intubation was performed by an experienced anaesthesiologist to minimise intubation-related haemodynamic fluctuations.

The intraoperative course was uneventful. Postoperatively, prior to extubation, a right-side Transversus Abdominis Plane (TAP) block was performed using Inj. Ropivacaine 0.375% (15 mL) and Inj. Dexamethasone 4 mg to minimise pain and reduce opioid consumption. The patient was successfully extubated after achieving adequate spontaneous breathing and reversing neuromuscular blockade with Inj. Sugammadex 100 mg intravenously. She was then transferred to the Post-Anaesthesia Care Unit (PACU) for haemodynamic monitoring and was discharged two days postoperatively without any complications.

DISCUSSION

Eisenmenger syndrome causes significant perioperative risks due to altered haemodynamics. The main anaesthetic challenge is

S. No.	Author's name and year	Case presentation	Treatment	Outcome
1.	Gupta N et al., 2011 [2]	A patient with known case of Eisenmenger syndrome with Beta Thalassemia was posted for splenectomy	This patient's Ventricular Septal Defect (VSD) was corrected with Pulmonary Artery Banding at 4 months of age. For splenectomy, patient was anaesthetised by inducing the patient with Ketamine	Intraoperative period was uneventful without anticipated complications such as intra-cardiac shunting under anaesthesia.
2.	Raines DE et al., 1996 [6]	A case series was monitored where outcomes of Eisenmenger syndrome patients planned for non cardiac non obstetric surgery were noted	A variety of anaesthetic techniques were used to anaesthetise the patient, i.e., 13 cases were done under general anaesthesia, 6 were done with peripheral nerve blocks, 5 with sedation with or without local anaesthetic infiltration and 1 with Epidural anaesthesia	They concluded that a variety of techniques were successful in the management of a patient with Eisenmenger's physiology undergoing non cardiac surgery, suggesting a lower perioperative mortality rate than those patients that are planned for labour and delivery which has a mortality rate of 10%.
3.	Chaudhuri A et al., 2022 [5]	4 patient outcomes of Eisenmenger syndrome patients who were posted for surgery were monitored	Surgery was done under epidural and local anaesthesia for the patients	2 out of 4 patients required inotropic support to maintain their haemodynamics. Haemodynamic stability and better analgesia were the reasons for choosing epidural anaesthesia. One patient developed symptoms suggestive of cardiac failure requiring postoperative Intensive Care Unit (ICU) and mechanical ventilation which was adequately managed.
4.	Maeda Y et al., 2024 [4]	A patient with known case of Eisenmenger syndrome was posted for laparoscopic hysterectomy	The surgery was done under general anaesthesia with peripheral nerve block	Intraoperative period was uneventful without anticipated complications. Postoperative analgesia was adequate due to the peripheral nerve block that was administered.
5.	Hashimoto K et al., 2024 [1]	A patient with known case of Eisenmenger syndrome was posted for bilateral adnexectomy	The patient was given general anaesthesia along with Remimazolam in the perioperative period	The use of Remimazolam prevented intubation response and maintained haemodynamics intraoperatively without hypotension or desaturation.
6.	Sarma H et al., 2021 [3]	A patient with known case of Eisenmenger syndrome with severe Pulmonary Artery Hypertension was posted for total abdominal hysterectomy	Patient was started on Vasopressin prophylactically in the preoperative period. General anaesthesia was induced with TIVA using Ketamine, Midazolam and Fentanyl	Although the patient had severe Pulmonary Artery Hypertension which was confirmed with right heart catheterisation, the patient was adequately optimised in the ICU before being planned for surgery. The use of TIVA maintained adequate depth of anaesthesia and in result, the intraoperative period was uneventful.
7.	Present study, 2025	A patient with known case of Eisenmenger syndrome was posted for open appendectomy	The surgery was done under general anaesthesia, with an opioid induction, along with TAP block	Opioid induction blunted intubation response, helping maintain haemodynamics and peripheral nerve block provided adequate postoperative analgesia.

[Table/Fig-1]: Table comparing previous literature on how a patient with Eisenmenger Syndrome was managed successfully [1-6].

to maintain preoperative haemodynamic stability, with particular attention to avoiding factors that could increase Pulmonary Vascular Resistance (PVR) or impair cardiac output. Several studies have outlined the benefits and drawbacks of various anaesthetic techniques in the management of such high-risk patients [Table/Fig-1] [1-6]. In present case, authors opted to use both general anaesthesia and a regional block to manage the patient and maintain her as close to her baseline haemodynamics as possible. Neuraxial blockade was avoided to prevent the profound hypotension that would be disastrous for the patient. General anaesthesia offers anaesthesiologists better control over the haemodynamics of the patient.

A case reported by Hashimoto K et al., described the management of an "Eisenmenger patient perioperatively with general anaesthesia, where the patient was induced with remimazolam" [1]. Remimazolam, being a benzodiazepine, "was observed to cause less circulatory depression, which was anticipated to be beneficial for the patient due to their underlying pathology." A study conducted by Gupta N et al., involved the management of an Eisenmenger patient planned for splenectomy under general anaesthesia, where the patient was induced with ketamine [2]. Furthermore, a case study reported by Sarma H et al., detailed the handling of an Eisenmenger patient with severe pulmonary hypertension planned for surgery [3]. This patient was managed with general anaesthesia, induced using Total Intravenous Anaesthesia (TIVA) with ketamine, midazolam and fentanyl.

Opioids are known to mitigate autonomic responses, thereby aiding in the maintenance of baseline haemodynamics for high-risk patients. They also provide a smooth induction, reducing the sympathetic response and preventing significant increases in PVR when used in adequate doses. Present study proceeded with an opioid induction. Fentanyl, a potent opioid with a rapid onset and short duration, was effectively used to induce anaesthesia with minimal cardiovascular alterations. In addition to selecting appropriate induction agents, it is crucial to avoid factors that could exacerbate shunt reversal and worsen oxygenation. Key factors contributing to increased peripheral vascular resistance include hypoxia, hypercarbia, acidosis and hypothermia. Our patient's baseline ABG analysis demonstrated some hypoxaemia (pO2 33.8 mmHg), underscoring the importance of avoiding further hypoxia during the procedure.

A case study conducted by Maeda Y et al., described the management of "an Eisenmenger patient planned for laparoscopic hysterectomy under general anaesthesia with a peripheral nerve block" for adequate analgesia [4]. Similar approach was adopted in present case, intending to provide general anaesthesia for the patient along with a regional block to ensure sufficient analgesia postoperatively after extubation. A study by Niraj G et al., concluded that the use of a TAP block reduced postoperative pain and aided recovery [7]. In line with this, a TAP block was utilised, which provided excellent pain relief with minimal systemic opioid use. Effective pain control is essential to prevent tachycardia, hypertension and increased PVR.

The patient's early extubation and stable postoperative recovery highlight the importance of optimising analgesia while avoiding factors that could exacerbate pulmonary hypertension. A case series discussed by Chaudhuri A et al., emphasised the need for inotropic support to maintain haemodynamics. Fortunately, this was not required in this patient due to minimal haemodynamic variations, which resulted from diligent perioperative management [5].

CONCLUSION(S)

The rarity of such a case presentation instilled extra caution within the anaesthesiology team. A known patient with Eisenmenger syndrome was scheduled for an appendicectomy and was taken up under general anaesthesia, with opioid induction to blunt the intubation response. A peripheral block was administered to provide adequate postoperative analgesia. Thorough perioperative management resulted in an uneventful hospital stay.

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PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Student, Department of Anaesthesiology, SRM Medical College Hospital and Research Centre, Chennai, Tamil Nadu, India.
2. Professor, Department of Anaesthesiology, SRM Medical College Hospital and Research Centre, Chennai, Tamil Nadu, India.
3. Postgraduate Student, Department of Anaesthesiology, SRM Medical College Hospital and Research Centre, Chennai, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Pushparani Anand,
Professor, Department of Anaesthesiology, SRM Medical College Hospital and Research Centre, Kattankulathur, Chennai-603203, Tamil Nadu, India.
E-mail: pushpa82_dr@yahoo.com

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