Original Research Article

Anaesthesia management of patients with Eisenmenger's syndrome: A case series

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Abstract

Background: Eisenmenger's syndrome is a congenital cyanotic heart disease, with septal defects/ patent ductus arteriosus, and advanced pulmonary hypertension with reversed/ bidirectional shunt. **Aims, Settings and Design:** In this non-formal case series, we aim to describe the perioperative and anaesthetic management of four cases of Eisenmenger's syndrome. 1) Emergency lower uterine segment caesarean section of a 30-year-old G_2A_1 female with 30 weeks of pregnancy, large atrial septal defect, dilated right atrium and ventricle, severe pulmonary arterial hypertension with raised right ventricular systolic pressure and reversal of shunt through defect. This patient was operated on later again for 2) secondary suturing. 3) Emergency lower uterine segment caesarean section of a 26-year-old G_2A_1 female with 33 weeks of gestation, pulmonary arterial hypertension, dilated right atrium, ventricle, main, right and left pulmonary arteries, with a patent ductus arteriosus. 4) Ovarian cystectomy of a 25-year-old P2L2 female with a right ovarian cyst, severe pulmonary hypertension, raised RVSP, and a large VSD with a bidirectional flow. **Materials And Methods:** The rationale behind our anaesthetic management was directed towards maintaining systemic vascular resistance, cardiac perfusion, preventing an increase in pulmonary vascular resistance and myocardial oxygen demand, infective endocarditis prophylaxis, multimodal analgesia, and thromboprophylaxis. A plan of management encompassing all of the above was necessary for our successful management of cases. **Conclusion:** Haemodynamic stability and stronger analgesia was the rationale behind choosing epidural and local anaesthesia over general anaesthesia, which was justified in literature. Necessity of adequate preparation for cardiac emergencies is also evident in two cases.

Keywords: Eisenmenger Complex, Phenylephrine, Pulmonary Hypertension, Regional Anaesthesia, Systemic Vascular Resistance.

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INTRODUCTION

Victor Eisenmenger (1864-1932) coined the term "Eisenmenger complex" in 1897, which denoted large ventricular septal defect (VSD) associated with pulmonary hypertension.[1,2] In 1958, it was redefined by Wood as pulmonary hypertension with reversed/bidirectional shunt associated with aberrant pathways like patent ductus arteriosus (PDA) or VSD. Through the aberrant pathways, the degree of right-to-left shunt depends on (a) the size of the communication and severity of pulmonary hypertension, (b) the relation between Pulmonary Vascular Resistance (PVR) and Systemic Vascular Resistance (SVR) and (c) the contractile state of the right ventricle.[3] Mortality risk of non-cardiac surgeries is as high as 30%.[4,5] Vascular resistance and pulmonary hypertension require lifelong conservative management. The prognosis seldom improves with low life expectancy (20–30 years). This non-formal case series describes the perioperative anaesthetic management for four cases of Eisenmenger's syndrome with lower segment caesarean sections (LSCS), secondary suturing of the surgical wound and ovarian cystectomy.

MATERIALS AND METHODS

The high risk of mortality was explained to all the patients and their written consent was taken for anaesthetic

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interventions and publication. All patients received 2 large-bore intravenous cannulas. Bubbles in lines and syringes were avoided meticulously to reduce the risk of paradoxical embolism. Injection pantoprazole 40 mg was given the night before and a 10-hour nil per oral period was followed. Intravenous injection ondansetron 4 mg and Intramuscular injection glycopyrrolate 200 µg was given half an hour before surgery. Inside the operating room, 4 litres per minute (LPM) of oxygen was supplied to all the patients via a Hudson mask. After sensitivity test, intravenous injection Amoxycillin 1 g and Injection Gentamycin 80 mg were given at induction of anaesthesia as a prophylaxis for infective endocarditis. Baseline pulse rate, non-invasive blood pressure, saturation and respiratory rates were recorded on multipara monitors. For epidural anaesthesia, toxic doses of local anaesthetics were precalculated, and the use of bupivacaine was restricted due to its potent systemic vasodilation. Preparations for General Anaesthesia, Cardiac resuscitation, post-operative intensive care and ventilator was kept on standby. Urine output was monitored hourly till post-operative 24 hours.

Case 1 and 2: A 30-year-old pregnant female, 142 cm in height, 37 kg, G_2A_1, belonging to American Society of Anesthesiology (ASA) physical status grade III, was admitted to a tertiary care centre at 29 weeks of gestation. The patient was cyanotic, with grade 4 clubbing of fingers, pedal oedema, pulse rate 110 /minute, blood pressure 110/70 mm Hg, oxygen saturation 90% on room air. On auscultation pansystolic murmur (S2 Split) was heard, air entry was decreased on the right side. The patient was diagnosed with congenital cyanotic heart disease 18 years back during pre-anaesthetic evaluation for ear surgery. 8 months back she had a spontaneous abortion at the 6th week of gestation. 2 months back her two-dimensional echocardiography (2D Echo) with colour doppler showed a large atrial septal defect (ASD) of 26 mm^2, VSD of 4 mm^2 with a right-to-left shunt, dilated right atrium/ right ventricle, severe pulmonary arterial hypertension (PAH), right ventricular systolic pressure (RVSP) 95 mm Hg, left ventricular ejection fraction (LVEF) 60%. She did not take her prescribed medication i.e., tablet Sildenafil 20 mg TDS, tablet Spironolactone 50 mg plus Torsemide 10 mg once daily, tablet Digoxin 0.375 mg once daily. Complete blood investigations revealed haemoglobin 17.4 g dL^{-1}, erythrocyte count 6,33,000/cc, hematocrit 60.6 (indicating erythrocytosis). Preoperative arterial blood gas (ABG) analysis showed pH 7.405, pO_2 46.5, pCO_2 20.9, HCO_3 24.9. The cardiologist advised continuing tablet Sildenafil, tablet Torsemide plus Spironolactone, tablet Digoxin based on a similar repeated 2D Echo report. Fitness for surgery with very high risk (with ~80% mortality) was given. Emergency LSCS was indicated due to uteroplacental insufficiency with reversal of end-diastolic flow. A 950-gram male baby was born with an Apgar score of 1 at birth and 5 at 5 minutes after birth. The baby was intubated and shifted to the neonatal intensive care unit. After delivery of baby injection oxytocin intramuscular 10 units and intravascular 3 unit $hour^{-1}$ infusion was started for our patient. 10 minutes after delivery, with blood pressure dropping to 90/60 mm Hg, Injection Dopamine infusion was started at a rate of 10 µg $kg^{-1}min^{-1}$. Ringer lactate infusion was replaced by titrated infusion of hydroxyethyl starch (a total of 150 ccs ringer lactate and 150 ccs hydroxyethyl starch was given during the procedure). At the end of the procedure, her urine output was 25 ccs. After surgery patient was shifted to the intensive care unit (ICU) for monitoring along with dopamine infusion and oxygen 4 litres min^{-1} by Hudson mask. Her BP was 120/78 mm Hg on injection Dopamine 10 µg$kg^{-1}min^{-1}$. The epidural catheter was removed in ICU and thromboprophylaxis was initiated. During the initial 2 hours of the postoperative period, the patient's oxygen saturation dropped to 75%, and she became tachypnoeic (35-40 breaths/min), bilateral basal crept were present (left> right), wheeze was present on the left side. Chest x-ray revealed left lower lobe haziness. Oxygen therapy was stepped up to 15 litres min^{-1} with a non-rebreathing mask (NRBM), Tab. Digoxin 125 µg was given. Central venous catheterization was done in the right internal jugular vein and initial central venous pressure (CVP) was 12 cm of water. Fluid was restricted and injection Furosemide 20 mg was given intravenously after every 3 hours. Despite this, the patient became increasingly tachypneic, and after 6 hours she was intubated using a cuffed endotracheal tube of 7.5 mm internal diameter, following intravenous injection Etomidate 10 mg and injection succinylcholine 50 mg. The patient was kept sedated, paralyzed and on controlled mechanical ventilation overnight. Injection intravenous Dopamine infusion was continued. Patients ABG analysis showed pH of 7.15, pCO_2 36, pO_2 65, HCO_3^- was 16.2. Injection intravenous $NaHCO_3$ (7.5% w/v) 50 cc was given. Injection intravenous Digoxin 0.5 mg was repeated after intubation, Injection furosemide was continued. After 6 hours her urine output was 1000 ml, bilateral basal crept were resolved. Weaning from controlled mechanical ventilation was initiated, and on spontaneous ventilation, the patient improved clinically with no signs of tachypnea and wheeze, her CVP was 5 cm of water. Serial chest x-ray showed improvement. She was extubated after 20 hours. Injection hydrocortisone 80 mg was given and budesonide nebulization was done. She was taken on oxygen 15 litres min^{-1} by an NRBM. Intravenous Dopamine infusion was stopped and the patient was started on her preoperative medications of tablet Sildenafil 20 mg TDS and tablet Digoxin 0.125 mg once daily. Injection Furosemide was stopped and her routine medication of

tablet Torsemide 10 mg plus Spironolactone 50 mg once daily was re-initiated. Over her stay of 5 postoperative days in the ICU, the patient's oxygen saturation improved to 90% on room air. The patient was shifted to the ward. The baby improved clinically, was extubated on postnatal day 10, started on full oral feeds on postnatal day 13, weaned off from oxygen support on the 15th postnatal day. On the 17th postoperative day, our patient had complaints of purulent discharge from the suture site and gaping of incision site was observed. The culture and sensitivity report confirmed the presence of gram-positive cocci sensitive to Amoxycillin and Clavulanic acid. She received a course of Intravenous injection of Amoxycillin 500 mg and Clavulanic acid 125 mg thrice daily for 5 days. She was posted for secondary suturing of wound gape on a postoperative day 23 after attaining no growth on repeat bacterial culture. Depth of wound gaping was 3 cm at maximum depth and superficial to rectus sheath. This time, the hemogram showed haemoglobin 16.2 g dL^{-1}, erythrocyte count, 5,47,000/cc, hematocrit 54.3. Preoperative ABG analysis showed pH 7.41, pO_2 54.8, pCO_2 19.8, HCO_3^- 25. Intense analgesia utilizing good local anaesthetic infiltration, anxiolysis and sedation was the anaesthetic target this time. Lactated ringer solution was started at 50 ml $hour^{-1}$ rate and intravenous injections of Ondansetron 4 mg, Midazolam 1 mg, Fentanyl 50 µg, Paracetamol 1 gram was given. After all aseptic precautions, 10 ml of Injection 2% Lignocaine with adrenaline and preservatives was infiltrated around the operative site using a 25-gauge spinal needle. Care was taken to avoid accidental intravascular injections. With the loss of sensation confirmed, the Operative procedure was started. The surgery lasted for 25 minutes and intraoperative vitals were stable. Postoperatively the patient was kept under observation for 30 minutes and she was shifted to Post Anaesthesia Care Unit (PACU) after complete arousal. The patient was kept in PACU for 2 hours with 4 litres of oxygen per minute by Hudson mask. Oxygen was stopped after 2 hours and the patient was observed for 15 minutes before shifting to the ward. She showed good recovery and was discharged 7 days later and advised for routine follow up with a cardiologist. She was also advised against further pregnancy.

Case 3: A 26-year-old pregnant female, 162 cm height, 54 kg in weight, a known case of congenital heart disease was admitted to a tertiary care centre at 33 weeks of gestation. Her $Sp0_2$ was 95% in the right upper limb, 92% in the Left upper limb, 90% in both lower limbs. Pansystolic murmur was present, Air entry was equal on both sides. The patient had a childhood history of breathlessness which was diagnosed as a congenital cardiac condition that wasn't evaluated further. 5 years back patient had another bout of breathlessness associated with palpitations which were investigated and evaluated as Severe PAH with RVSP 59 mm Hg. A Computed Tomography (CT) scan of the thorax showed pleural tags in the upper lobe anterior segment of both lungs, 5 mm soft tissue density nodule in the basal segment with mediastinal lymphadenopathy. Spirometry showed a moderate restrictive pattern of lung disease. A twelve-lead electrocardiogram (ECG) showed Right Ventricular hypertrophy (RVH) with strain pattern. The patient was started on tab Sildenafil 5 mg BD, tab Bosentan 62.5 mg OD, which she took for 6 months and stopped. She had a history of incomplete abortion with dilatation and curettage done under Local anaesthesia 1 year ago. She was anaemic at the time and received 350 ml of packed human red blood corpuscle during the surgical procedure. A fresh 2D Echo revealed severe idiopathic PAH, with Pulmonary Arterial Systolic Pressure (PASP) 65 mm Hg, dilated right atrium and ventricle, main, left and right pulmonary arteries dilated, a 6 mm^2 PDA with bidirectional shunt, Pulmonary Arterial End Diastolic Pressure 32 mm of Hg, mild Tricuspid Regurgitation (TR) with Mild mitral and aortic regurgitation. The patient was started on tab Digoxin 0.25 mg OD for 7 days and continued on Tab Sildenafil.

The plan of anaesthesia was epidural anaesthesia with general anaesthesia standby. Her pulse oximetry showed oxygen saturation of 95% on room air. Right internal jugular venous catheterisation was done with a triple lumen central line for central venous pressure (CVP) monitoring. Baseline parameters were recorded as heart rate: 84/min, blood pressure: 98/70 mm Hg, CVP: 6 cm of water, SpO_2: 95% on room air and 100% on 4 litres of oxygen per minute by Hudson mask. After epidural placement, a test dose of Injection of 2% lignocaine with adrenaline (3 ccs) was given to exclude intrathecal or intravascular placement. Hydroxyethyl starch (colloid) preloading was done at a rate of 80 ccs per hour. Incremental dose of Injection Lignocaine 2% without preservatives and Bupivacaine (0.5%) were titrated against anaesthetic and haemodynamic effect over 20 minutes. A total of 8 ml of Lignocaine and 6 ml of Bupivacaine (0.5%) with 25 ug fentanyl were given epidurally to achieve a T8 level of sensory blockade with Bromage scale-3 motor blockade. Surgery was undertaken thereafter. After delivery of baby, 10 units of injection oxytocin was given via intramuscular route and 10 Units via intravenous access (at 3 units $hour^{-1}$ rate). During uterine closure, her blood pressure fell to 90/60 mm Hg with tachycardia, and intravenous injection dopamine infusion was started at 10 µg $Kg^{-1}min^{-1}$ rate, the intravenous colloid infusion was increased up to 100 cc $hour^{-1}$ and injection phenylephrine 50 µg bolus was administered. Her blood pressure improved to 110/80 mm Hg with HR 82/min after the bolus dose of phenylephrine. Intravenous dopamine infusion was

titrated down according to blood pressure and finally tapered off. A total of 150 ml colloid was administered intraoperatively. Epidural analgesia was administered with an 8 ml injection of Bupivacaine (0.125%) plus Tramadol 50 mg at the end of the procedure. Intraoperative CVP was between 8 – 10 cm of H_20. Total urine output was 500 cc, blood loss 700 ccs. Intraoperative capillary blood sugar level noted was 75 mg dL^{-1}, for which intravenous 50 ccs 25% dextrose plus 20 ccs 5% dextrose in normal saline was given. After surgery, the patient was shifted to the intensive care unit for observation. The epidural catheter was removed in ICU and she was started on anticoagulants as per the cardiologist's advice. Postoperatively, vitals were noted in ICU and recorded as Heart rate: 92 beats/min, Blood pressure: 130/80 mm Hg, SpO_2: 100% on Room Air. The patient was monitored in ICU for 2 days and then shifted to the ward. She was discharged after 7 days with advice against further pregnancies and follow up with a cardiologist.

Case 4: A 25 Years old P_2L_2 female, weighing 50 kg, belonging to ASA physical status III, had difficulty passing urine for 10 days. She had suffered from New York Heart Association (NYHA) grade II breathlessness in childhood, with chronic hypoxia and grade II clubbing since childhood. A loud P2, ejection systolic murmur prominent over the pulmonary area, and pansystolic murmur in the tricuspid area were auscultated, and jugular venous pressure (JVP) was found to be raised. Pulse oximetry showed 92% oxygen saturation on room air. Routine biochemical investigations showed: haemoglobin 16.7 g dL^{-1}, haematocrit was 48.9, erythrocyte count of 561000/cc (indicating erythrocytosis), normal serum electrolytes, proteins, renal and liver function tests. High-Resolution CT scan of thorax revealed: Coronavirus disease Reporting And Data System (CORADS) score of 1, RVH and prominent pulmonary arteries. 12 lead ECG portrayed T inversion in V4, V5, V6, ST elevation in aVR, aVL, RV strain pattern in Lead V1. Severe PAH and RVSP 115 mm Hg, with 16 mm^2 peri-membranous VSD having a bidirectional flow was depicted on 2D echo. An 8.3 cm x 7.5 cm x 8.4 cm complex multiloculated single right ovarian cystic lesion with low-level internal echoes and internal septations within, mild free fluid in the pouch of Douglas and a few Nabothian cysts (largest 4 mm) in the vaginal cervix were demonstrated on transvaginal Ultrasonography (USG) and she was posted for right ovarian cystectomy. Preoperative ABG analysis demonstrated a hypoxemic picture with pH 7.36, pCO_2 27.3, pO_2 50.6, sO_2 90.2 and HCO_3^- 23. Four litres of oxygen per minute was initiated by Hudson mask. After the epidural catheter was fixed, a test dose of 3 mL of 2% lignocaine hydrochloride – adrenaline solution (1:1,00,000) was given. Ringer Lactate infusion was restricted at 50 mL $hour^{-1}$. Postoperatively the patient was monitored for 3 hours in the PACU before shifting to ICU. Post-operative analgesia was administered by an epidural dose of 6 ml 0.125% bupivacaine and the epidural catheter was removed. Multimodal analgesia and Anticoagulant therapy with Low Molecular Weight Heparin (LMWH) 5000 IU by subcutaneous route were initiated. One episode of shivering and fall of oxygen saturation occurred 8 hours after surgery. The oxygen flow rate was increased to 8 litres per minute and saturation improved to 96%, with ABGA showing similar improvement in pO_2 (80.3 mm Hg), sO_2 (97.2%). Oxygen was tapered on the 3rd post-operative day. The patient was discharged on the 5th postoperative day and advised routine follow up with a cardiologist.

RESULTS

All 4 cases were successfully managed in the perioperative period with different approaches but goal directed by similar concerns. 2 cases required intraoperative inotropic support. In spite of a tight intraoperative fluid restriction, postoperative ICU management ensued for one patient with intubation and cardiac optimization who became symptomatic for cardiac failure. Management was goal directed to rectify pulmonary edema, maintain SVR, reduce PVR, prevent arrhythmia, decrease cardiac workload along with good analgesia cover to promote faster recovery.

Epidurally given anaesthetic dosages for the three cases are mentioned below:

(Table 1) The achieved level of anaesthesia was adequate for all three procedures. Mild intravenous sedation with Injection Midazolam 1 mg and Injection Fentanyl 0.5 µg kg^{-1} was given.

Table 1: Timing and dosage of aliquots of epidural anesthesia in three out of four cases

Case 1			
Time In minutes (From test dose)	**Drug name**	**Volume**	**Dose**
05:00	Plain Lignocaine (2%)	5ml	100mg
10:00	Bupivacaine(0.5%)	4ml	20mg
20:00	Plain Lignocaine (2%)	3ml	600mg
30:00	Bupivacaine(0.5%)	3ml	15mg
30:00	Fentanyl(50μg/cc)	0.5ml	25μg

Case 3			
Time In minutes (From test dose)	Drug name	Volume	Dose
05:00	Plain Lignocaine (2%)	4ml	80mg
08:00	Plain Lignocaine (2%)	4ml	80mg
15:00	Bupivacaine(0.5%)	4ml	20mg
20:00	Bupivacaine(0.5%)+	2ml	10mg
	Fentanyl (50 μ g/cc)	0.5ml	25ug
45:00	Bupivacaine(0.25%)	4ml	10mg
60:00	Bupivacaine(0.125%)+	8ml	10mg
	Tramadol		50mg
Case 4			
Time In minutes (From test dose)	Drug name	Volume	Dose
05:00	Plain Lignocaine (2%)	4ml	80mg
15:00	Plain Lignocaine (2%)	2ml	40mg
45:00	Bupivacaine(0.5%)	4ml	20mg
60:00	Tramadol (50mg/ml)	1ml	50mg

DISCUSSION

An aberrant connection between pulmonary and systemic circulation, where blood pressure in the pulmonary circulation is similar to or more than that of the systemic circulation, results in a right-to-left/ bidirectional shunt, causing Eisenmenger's Syndrome.[6,7] Raising PVR, or lowering SVR, results in increased right-to-left shunting of deoxygenated blood to enter systemic arterial circulation, causing global hypoxia. Cyanotic episodes due to exacerbation of shunt and chronic hypoxia lead to polycythaemia and subsequent thromboembolic complications. Acute intraoperative hypoxic episodes are promoted by active right-to-Left/ bidirectional shunt.[8] Changes in SVR/PVR ratio, hypoxia, hypercarbia, acidosis, systemic hypotension, hypovolemia, pulmonary hypertension are a few conditions that should be kept in mind while choosing a technique of anaesthesia. Anaesthesiologists should properly maintain SVR, which prohibits the use of intravenous induction agents such as injection Thiopentone or Propofol. PVR can be increased by catecholamines released during or after laryngoscopy or during extubation. Intermittent positive pressure ventilation (IPPV) raises intrathoracic pressure, lowers venous return, elevates pulmonary arterial pressure and right-to-left shunt. Hypercarbia can be exacerbated by spontaneous ventilation, and nitrous oxide can promote hypoxia.[6] Different inhalational agents affect SVR/PVR ratio differently. Although Sevoflurane and Desflurane decrease SVR, they do not affect Cardiac Output (CO) profusely.[9] Systemic opioids provide inferior analgesia, and sympathetic pain response, tachycardia and increased cardiac output in the presence of VSD and pulmonary hypertension depicts increased right-to-left shunt.[10] Based on all these reasons, epidural anaesthesia and local anaesthesia was considered superior to general anaesthesia. The potent systemic vasodilatory effect of bupivacaine rendered its use restricted. Intravenous anxiolysis and sedation were given with inj. Midazolam 1 mg and Inj. Fentanyl 0.5 µg kg^{-1}. There is controversy about the benefit of invasive central venous and arterial catheterisation and monitoring in Eisenmenger syndrome, as the risk of paradoxical emboli is more in these patients, with catastrophic consequences.[11] Hence Central venous catheterisation was kept strictly as a standby measure for 3 out of 4 cases. Arterial line and invasive blood pressure monitoring were also kept on standby for the same reasons. Early thromboprophylaxis with subcutaneous LMWH 5000 IU was started to reduce the risk of thromboembolism during post-operative immobilization.[12] Prophylactic intravenous doses of Inj. Amoxicillin 1 g and Inj. Gentamicin 80 mg were indicated as a measure against infective endocarditis.[13] In maintaining SVR, a significant role can be played by intravenous infusion of 10 µg $kg^{-1}min^{-1}$ Dopamine.[12] Phenylephrine is primarily an α1 adrenergic receptor agonist with minimal β adrenergic activity. It is thus ideal for elevating mean arterial pressure.[14] Intravenous Inj. Phenylephrine causes arterial and venous vasoconstriction and maintains afterload, SVR and preload without any direct effect on cardiac myocytes, avoiding tachycardia. Thus, it can reduce right-to-left shunt without severely increasing myocardial workload and oxygen demand. Moreover, reflex bradycardia maintains the diastolic period and cardiac perfusion, maintaining oxygen supply.[15] So, injection Phenylephrine was preferred for emergencies. It has a short onset of action (1 – 3 minutes) and a short duration of action (15 – 20 minutes). So, it was given in 50 ug bolus aliquots as required. It has also been demonstrated as safe to be used via peripheral venous catheters, as recent studies suggest that it does not increase the chances of tissue necrosis.[16] Although in our patient, we used the right internal jugular central venous catheter access for intravenous boluses of phenylephrine. Spinal anaesthesia should be avoided as it is strongly associated with an intense loss of SVR. Spinal

anaesthesia with intrathecal isobaric 0.75% ropivacaine or isobaric 0.5% levobupivacaine has a less hemodynamic effect, less cardiotoxicity and central nervous system toxicity.[17] However as intravenous fluids were restricted and loss of SVR associated with segmental sympathectomy was prohibited, spinal anaesthesia was avoided. This case series lacks the scope to discuss additives like Dexmedetomidine, Clonidine and Tramadol with intrathecal anaesthesia as adequate literature on the hemodynamic safety of these additives in Eisenmenger patients isn't available.

CONCLUSIONS

Prophylaxis of infective Endocarditis has to be taken. Maintaining SVR is the mainstay of hemodynamic management. Dopamine, Phenylephrine both can be used to maintain SVR in emergency conditions. Epidural anaesthesia is superior to spinal anaesthesia to prevent loss of SVR. Any procedure that exacerbates pulmonary hypertension/ vascular resistance should be avoided. Pulmonary oedema can be precipitated in the presence of pulmonary vascular congestion and PAH. Prompt action should be taken to rectify pulmonary oedema in these patients. Chronic hypoxia leads to erythrocytosis, increased blood viscosity leading to an increased tendency of thromboembolism, so thromboprophylaxis should be a concern. Moreover, extra care should be taken to avoid air embolization as the presence of VSD can result in paradoxical emboli with a catastrophic sequence. Duration of diastole should be maintained, to maintain cardiac perfusion and oxygen supply. And myocardial oxygen demand should not rise. So, tachycardia should be avoided. Using nitrous oxide can exacerbate hypoxia. It is best to maintain the patients on 100% oxygen, in spontaneous ventilation. Hypercarbia, acidosis, hypovolaemia, hypothermia, hypotension should be avoided.

REFERENCES

1. The anaesthetic management of the child with Eisenmenger's syndrome. Lyons B, Motherway C, Casey W, Doherty P. 1995, Can J Anaesth, pp. 42:904-9.
2. Eisenmenger Syndrome- Factors relating to deterioration and death. Daliento L, Somerville J, Presbitero P, Menti L, Brach-prever S, Rizzoli G, et al. 1998, Eur Heart J, pp. 19:1845-55.
3. Shobana Chandrasekhar, Daniel A tolpin, Dennis T Mangano. Anaesthetic Management of the Pregnant Cardiac Patient. [book auth.] B Scott Segal, Roanne Preston, Roshan Fernando, C LaToya Mason Maya S Suresh. Shnider and Levinson's Anaesthesia for Obstetrics. s.l. : Jaypee Medical, 2012, 30, p. 492.
4. Anesthetic management and outcomes for patients with pulmonary hypertension and intracardiac shunts and Eisenmenger syndrome: a review of institutional experience. Bennett JM, Ehrenfeld JM, Markham L, Eagle SS. 2014, J Clin Anesth 26, pp. 286-293.
5. Eisenmenger syndrome: current perspectives. Nashat H, Kempny A, McCabe C, Price LC, Harries C, et al. 2017, Research Reports in Clinical Cardiology, 2 February Volume, pp. 8 Pages 1-12.
6. Incremental spinal anaesthesia for elective Caesarean section in a patient with Eisenmenger's syndrome. Cole PJ, Cross MH, Dresner M. 2001, Br J Anaesth, pp. 86:723-6.
7. The anaesthetic management of the Eisenmenger syndrome. Foster, J. M., and Jones, R. M. 1984, Annals of the Royal College of Surgeons of England, 66(5), pp. 353–355.
8. Anaesthetic management of a patient with Eisenmenger syndrome and β-thalassemia major for splenectomy. Gupta N, Kaur S, Goila A, Pawar M. 2011, Indian J Anaesth, pp. 52:187-9.
9. Pharmacology of anaesthetic agents II: inhalation anaesthetic agents. Khurram Saleem Khan, Ivan Hayes, Donal J Buggy. 2014, Continuing Education in Anaesthesia Critical Care and Pain, Volume 14, Issue 3, June, pp. 106–111.
10. Anaesthetic management of a parturient with Eisenmenger's syndrome. Pollack KL, Chestnut DH, Wenstrom KD. 1990, Anesth Analg, pp. 70(2):212-5.
11. Tetralogy of Fallot repair in a patient with beta-thalassemia major. Venugopal, K., Nair, S. G., and Rao, S. G. 2005, Journal of cardiothoracic and vascular anesthesia, pp. 19(1), 93–96.
12. Anaesthesia for a patient with Eisenmenger's syndrome undergoing caesarean section. Gurumurthy T, Hegde R, Mohandas BS. 2012, Indian J Anaesth, pp. 56:291-4.
13. Guidelines for the prevention of endocarditis: report of the Working Party of the British Society for Antimicrobial Chemotherapy. F. K. Gould, T. S. J. Elliott, J. Foweraker, M. Fulford, J. D. Perry, G. J. Roberts, J. A. T. Sandoe, R. W. Watkin. 6, s.l. : Journal of Antimicrobial Chemotherapy, June 2006, Vols. 57, pages 1035–1042.
14. Flancbaum L, Dick M, Dasta J, Sinha R, Choban P. A Dose-response Study of Phenylephrine in Critically Ill, Septic Surgical Patient. Eur J Clin Pharmacol. 1997, Vol. 51, 6, pp. 461 - 5.
15. Yamazaki T, Shimada Y, Taenaka N, Oshumi H, Takezawa J, Yoshiya I. Circulatory Responses to Afterloading with Phenylephrine in Hyperdynamic Sepsis. Crit Care Med. 1st, Jul 1982, Vol. 10, 7, pp. 432 - 5.
16. Cardenas-Garcia J, Schaub KF, Belchikov YG, Narasimhan M, Koenig SJ, Mayo PH,. Safety of Peripheral Intravenous Administration of Vasoactive Medication. J Hosp Med. Sep 2015, Vol. 10, 9, pp. 581 - 5.
17. Comparison of Intrathecal hyperbaric 0.5% Bupivacaine, isobaric 0.5% Levobupivacaine and isobaric 0.75% Ropivacaine for lower abdominal surgeries. D'Souza AD, Saldanha NM, Monteiro AD et. al. 2014, Int J Health Sci Res, pp. 4(1):22-29.

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