ISSN 2394-4781(Print)
e-ISSN 2394-4994(Online)

Case Study

POSTOPERATIVE MORTALITY IN A PATIENT WITH EISENMENGER`S SYNDROME UNDERGOING EMERGENCY CAESAREAN SECTION: FOREWARNED BUT YET TO BE FOREARMED

Dr. Priyam Saikia[1], Dr. Mridu Paban Nath[2,*], Dr. Marie Ninu[3], Dr. Ruma Thakuria[4], Dr. Dipika Choudhury[5]

[1]Dr. Priyam Saikia M.D, Assistant Professor, Department of Anaesthesiology and Critical Care, Gauhati Medical College and Hospital, Guwahati, Assam, India, PIN-781032
[2]Dr. Mridu Paban Nath M.D., Assistant Professor (Cardiac Anesthesia Division), Department of Anaesthesiology and Critical Care, Gauhati Medical College and Hospital, Guwahati, Assam, India, PIN-781032,
[3]Dr. Marie Ninu M.D, Senior Resident, Department of Anaesthesiology and Critical Care, Northeastern Indira Gandhi Institute of Health and Medical Science, Shillong, Meghalaya, PIN 793018
[4]Dr. Ruma Thakuria M.D, Senior Resident, Department of Anaesthesiology and Critical Care, GNRC Hospitals Limited, North Guwahati, Sila Grant, Assam, India.
[5]Dr. Dipika Choudhury M.D., Professor and Head of the Department, Department of Anaesthesiology and Critical Care, Gauhati Medical College and Hospital, Guwahati, Assam, India, PIN-781032.

***Corresponding Author:**
E-mail: drmridupaban@yahoo.com

Abstract: *Eisenmenger's syndrome includes pulmonary artery hypertension, reversed or bidirectional shunt associated with an atrial septal defect, ventricular septal defect and patent ductus arteriosus. Death can occur at any time during pregnancy as well in the puerperium. We present here a known case of Ventricular septal defect in Eisenmenger syndrome who presented in 29 weeks of gestation with orthopnea. Elective caesarean section was done with post-operative intensive care under general anesthesia uneventfully. The anesthetic management is discussed here.*

Key words: *Eisenmenger syndrome, Echocardiography, Lower segment caesarean section*

Introduction

In 1958, Paul Wood coined the term Eisenmenger syndrome (ES) to describe pulmonary hypertension at systemic level with reversed or bidirectional shunt through a large ventricular septal defect (VSD). [1]Pregnancy influences the pathophysiology in this group of patients and increases the risk of a poor maternal outcome. [2]Although patients with ES present considerable risks during perioperative period, successful outcomes in different surgeries with varied anesthetic technique have been described. [3-6] Nonetheless perioperative mortality has remained an enigma.

Case Report

25-year-old lady weighing 42kg, a known case of VSD with episodes of transient ischemic attack conceived against medical advice and presented with gradually progressive dyspnea at 29 weeks of gestation. She was cyanotic with clubbed fingers, raised jugular venous pressure, dyspnea of New York Heart Association (NYHA) grade IV, respiratory rate 26 breaths/ min, noninvasive blood pressure (NIBP) 104/80 mmHg and pulse rate 105 beats/min(low volume and regular). On auscultation, a loud S2 and grade-III/IV systolic murmur was heard over the precordium with right ventricular lift along with fine crepitation over bilateral basal lung region. Peripheral Pulse oximerty (SpO_2) reading at room air was 60%, which increased to 70% with 100% oxygen (O_2) via simple facemask at 10Lmin^{-1} flow rate. The patient was transferred to the intensive care unit (ICU) and treated by a multidisciplinary team of obstetricians, cardiologists, and anesthesiologists. She was started with diuretic, digoxin and antibiotic. A transthoracic echocardiogram done 2 months prior to presentation to our hospital revealed biventricular enlargement, an 18mm peri-membranous VSD with right to left shunt, a dilated pulmonary

artery with severe pulmonary hypertension with ejection fraction of 50%. Her attending cardiologist advised urgent termination of pregnancy. Preoperative investigations including a hemogram, biochemical tests and coagulation screen (PT, aPTT) revealed no abnormalities except polycythemia (Hb-16.7Gm/dL), leukocytosis (18,020/cumm), thrombocytopenia ($40X10^3$/cumm), and hypokalemia (2.7mmol/L). Arterial blood gas analysis (ABG) at room air revealed pH 7.34, $PCO_2$27.6mmHg, PO_2 30.2 mmHg, base excess -7 mmol/L, HCO_3^- 15.7 mmol/L and SaO_2 62%. Electrocardiography (ECG) showed normal sinus rhythm, right atrial abnormality, right axis deviation. Portable X-ray did not reveal any lung pathology. She was given 2 units of platelet concentrate (PLC) transfusion and the post transfusion platelet count increased to $70X10^3$/cumm. It was decided to deliver the baby by Lower Segment Caesarean Section under general anesthesia and to monitor the patient in ICU. All risks were explained and written informed consent was gained. Preoperatively, prophylaxis for infective endocarditis and aspiration were administered. The need for use of opioid before clamping of umbilical artery was discussed with the patient's attendants and the attending pediatrician. Care to prevent introduction of air bubble via the infusion devices was taken and continued in the postoperative period. She was conveyed to the operating room in a 30^0 head-up tilt with left lateral position with oxygen supplementation by facemask. Standard monitoring including SpO_2, NIBP, ECG (5lead) and skin temperature was started. She was given 40µgm fentanyl intravenously (IV) and under local anesthesia the left radial artery was cannulated with 20G catheter and her arterial blood pressure (ABP) was 118/88mmHg with SpO_2 of 70%. Pre-oxygenation with 100% O_2 increased SpO_2 to 80%. Anesthesia was induced with etomidate 8mg and neuromuscular blockade was achieved with rocuronium 40mg. Her lungs were ventilated with volume control mode (tidal volume of 320ml, respiratory rate of 14/min, I: E ratio of 1:2, without PEEP) with isoflurane (end tidal 0.5-0.8%) in O_2 and air (80:20 mixture) delivered via a size 3 Proseal Laryngeal Mask Airway. Then a triple lumen 18G central venous catheter was placed in right internal jugular vein and her initial central venous pressure (CVP) was 4 mmHg. Her ABP, CVP, End tidal CO_2, ABG, nasopharyngeal temperature, urine output, inspiratory and expiratory gas concentration, peak airway pressure, compliance, airway resistance, inspired and expired tidal volume and minute ventilation with intermittent monitoring of pressure volume (PV) loop was done. A male baby weighing 910 gm was delivered and was shifted to neonatal ICU after resuscitation. Uterine massage and a slow oxytocin infusion were started. Another bolus of 40µgm fentanyl was given IV after the delivery of the baby. Estimated blood loss was ~400ml and urine output was 70ml in 90min; she was infused with 300ml Ringers Lactate and 200ml of 6% HES (VOLULYTE) in balanced salt solution to keep her CVP at around 8-10mmHg. 800 µgm misoprostol was administered per rectally at the end of the procedure. Throughout the intraoperative period her hemodynamic remained stable except for one episode of hypotension responded immediately to bolus injection of 40 µgm phenylephrine. As the patient remained hemodynamically stable throughout surgery, residual neuromuscular block was reversed with neostigmine (2.0 mg) and glycopyrrolate (0.4 mg) and the supraglottic airway device was removed. Immediate post-operative ABP-102/57mmHg, SpO_2 80%, CVP 8mmHg, sustained head lift >5 sec present and Richmond Agitation and Sedation Scale was 0. The patient was sent to the ICU with oxygen (10 L/min) supplementation by facemask. For maintenance of fluid, IV crystalloids were given at 2 mL/kg/hr. Analgesia was provided by trans- dermal fentanyl patch (release rate of 25µgm /hr).Her preoperative drugs were continued. Her platelet count improved to 1, 00,000/cumm with normal PT and aPTT values in the first postoperative day. Intravenous infusion of Paracetamol 1gm every 8 hourly was added as an analgesic and she was pain free in the postoperative period. She started to take feed orally, invasive vascular catheters removed and mobilized in a wheelchair on the 2^{nd}postoperative day. After an uneventful 2-day stay in the ICU, she suddenly developed generalized tonic clonic seizure with stable hemodynamic parameters, which was initially successfully managed with IV Midazolam. ABG and blood sugar estimation revealed no gross alteration compared to preoperative values. In the postictal period left sided hemiparesis with deviation of the corner of mouth to the right side was noted. Though management protocol for stroke was initiated was started, we lost her to a spiraling deterioration in her hemodynamic status state even before imaging studies could be carried out and autopsy was refused by the patient's attendants.

Discussion

Although risks of anesthesia are considerable in patients with ES, the anesthetic concerns and their managements are very well described in literature. [3-6, 7] Effective interdepartmental communication, meticulous planning and their execution has led to an uneventful intra-operative course in this patient. Although because of reasons cited above, we are not able to determine the exact cause, considering the clinical features we strongly believe that a cerebrovascular accident (CVA) caused her death. Patients with ES are inherently at high risk of CVA. Apart from the usual risk factors for CVA, abnormality of haemostasis, paradoxical emboli, altered rheology of blood, abnormality of vascular regulatory mechanism, inappropriate phlebotomies, and infectious complications are believed to contribute to this increased risk. [7, 8] Moreover, the multifactorial etiology along with inaccuracy of laboratory tests of coagulation and therapeutic dilemma surrounding drugs acting on coagulation system further complicates the scene.[7] The use of drugs with known effects on coagulation system might have further increased the risk of CVA in this patient. Although appropriate imaging study of brain could have helped in defining the probable etiology, the clinical events did not allow that. Although CVA is a well described phenomenon, the exact etiology of mortality and their predictors in patients with ES undergoing obstetric surgery is largely unknown, as cases that are unsuccessful are often not reported in the literature. [4] Historically, management options for patients with ES have been limited to palliative measures or heart-lung transplantation, the latter of which is suitable only for a small subgroup of patients. [8]Mebus*et al* has recently reviewed various therapeutic medical options for patients with ES, however none of these approaches significantly modified survival or risk of deterioration and opined that further study is needed so that customized treatment can be tailored to patient with ES. [9] But Dimopoulos reported that use of advance therapy is associated with a lower risk of death. [10] Even in this era there is high perioperative mortality in patients with ES with reported maternal mortality rate of 30-60 %, which has remained nearly same compared to data from 2 decades ago. [7, 8] Though most deaths occur in the immediate perioperative period, it has been reported even 14 days after cesarean section and it has been recommended to hospitalize patients after 20 wks of gestation and continue post-operative surveillance. [7, 8] Major causes of death are worsening functional status, pulmonary and cerebral complication, arrhythmia, endocarditis, renal dysfunction, thromboembolism, hypovolemia, hemoptysis or preeclampsia. [7, 8] Not only mortality, the high incidence of morbidity in this group of patients reflects the risk of pulmonary vascular disease in addition to the risk of abnormal hemostasis in cyanotic patients. [11] No data is available regarding predictors of perioperative mortality but laboratory and ECG-related parameters associated with chronic heart failure and arrhythmia (low serum albumin, low potassium levels, and longer QRS duration and QTc interval) are predictive of mortality in general .[12] An experienced cardiac anesthesiologist, skilled surgeon, preoperative phlebotomy , avoidance of percutaneous flotation catheters, use of an air/particle filter in intravenous lines, intraoperative and postoperative monitoring of systemic vascular resistance, use of pulse oximeter and prevention of hypotension have been proposed as strategies for reduction of perioperative risk . [3-6, 7, 11] Although successful anesthetic technique has been described, various events e.g. hypoxemia, hemodynamic perturbations, CVA, seizure, bleeding with or without anticoagulant therapy, venous thrombosis and concomitant peripartal cardiomyopathy adversely influence postpartal recovery. [13]It is evident that not only the acute changes in pathophysiology of ES by anesthetic and surgical factors, many other factors inherent to the disease influences the perioperative outcome. As much new therapy has been recently utilized for overall management in these patients with encouraging results, it would be worthwhile to see if their use positively affects the outcome in patients with ES undergoing surgery. [9, 10]

This report has some limitations. First, we could not use pulmonary vasodilator therapy as it was not available at that time. Second, we could not perform brain imaging study which could have provided insight into the pathophysiology (ischemic *vs* hemorrhagic) surrounding her death.

Conclusion

We have presented the peripartum management of a patient with ES who had a fatal outcome in the postoperative period. This report highlights the potential for complication that may mar an uneventful intraoperative course. We hope that such disappointments will be less with individually tailored treatment during pregnancy and medical therapy, once the role of laboratory investigation and medical therapies for patients with ES will become less ambiguous and development of intensive care support focused on the postpartal period.

References

1. Wood P. The Eisenmenger syndrome or pulmonary hypertension with reversed central shunt. Br Med J 1958; 2:701-9.
2. Branko M. Weiss, Lea Zemp, Burkhardt Seifert, Otto M. Hess. Outcome of Pulmonary Vascular Disease in Pregnancy: A Systematic Overview from 1978 through 1996. J Am Coll Cardiol1998; 31(7):1650–7.
3. Sammut MS, PaesML. Anesthesia for laparoscopic cholecystectomy in a patient with Eisenmenger`s syndrome. Br J Anaesth 1997; 79:810-12.
4. Cole PJ, Cross MH, Dresner M. Incremental spinal anaesthesia for elective caesarean section in a patient with Eisenmenger'ssyndrome. Br J Anaesth 2001; 86:723–6.
5. Ravindra Pandey, RakeshGarg, MriduPaban Nath, ShakthiRajan,JyotsnaPunj, VanlalDarlong, Chandralekha. Eisenmenger'sSyndrome in Pregnancy: Use ofProseal® Laryngeal Mask Airway (PLMA) and Epidural Analgesia for Elective Cesarean Section. Acta Anaesthesiol Taiwan 2009; 47(4):204–07.
6. B.Ghai, V. Mohan, M. Khetarpal, N. Malhotra. Epidural anesthesia for cesarean section in a patient with Eisenmenger'ssyndrome. Int J ObstetAnesth 2002; 11(1):44-47.
7. Oechslin E, Mebus S, Schulze-Neick I, Niwa K, Trindade PT, EickenA, Hager A, Lang I, Hess J, Kaemmerer H. The Adult Patient with Eisenmenger Syndrome: A Medical Update after Dana Point Part III: Specific Management and Surgical Aspects. Curr Cardiol Rev. 2010 Nov; 6(4):363-72.
8. Daliento L, Somerville J, PresbiteroP, Menti L, Brach-Prever S, Rizzoli G, Stone S. Eisenmenger syndrome. Factors relating to deterioration and death .Eur Heart J. 1998 Dec; 19(12):1845-55.
9. Mebus S, Schulze-Neick I, Oechslin E, Niwa K, Trindade PT, Hager A, Hess J, Kaemmerer H. The Adult Patient with EisenmengerSyndrome: A Medical Update after Dana Point Part II: Medical Treatment - Study Results. Curr Cardiol Rev. 2010 Nov; 6(4):356-62.
10. Dimopoulos K, Inuzuka R, Goletto S, Giannakoulas G, Swan L, Wort SJ, Gatzoulis MA.Improved survival among patients with Eisenmenger syndrome receiving advanced therapy for pulmonary arterial hypertension. Circulation. 2010; 121(1):20-5.
11.KoichiroNiwa, Joseph K. Perloff, Samuel Kaplan, John S. Child, Pamela D. Miner.Eisenmenger syndrome in adults Ventricular septal defect, truncusarteriosus, univentricular heart. Am Coll Cardiol, 1999; 34:223-232?
11. Diller GP, Dimopoulos K, Broberg CS, Kaya MG, Naghotra US, Uebing A, Harries C, Goktekin O, Gibbs JS, Gatzoulis MA. Presentation, survival prospects, and predictors of death in Eisenmenger syndrome: a combined retrospective and case-control study. Eur Heart J. 2006 Jul; 27(14):1737-42.
12. Weiss BM, Zemp L, Seifert B, Hess OM. Outcome of pulmonary vascular disease in pregnancy: a systematic overview from 1978 through 1996. J Am Coll Cardiol. 1998 Jun; 31(7):1650-7.