

Anaesthetic Management of a Parturient with Incidental Diagnosis of Ebstein Anomaly in COVID 19 Era-A Rare Case Report

Neha Yadav¹, Khushbu Rani², BK Sandhya², Simar Anand²

¹Assistant Professor, Department of Anaesthesiology, MMIMSR, MMDU, Mullana, Ambala, Haryana, India, ²PG Student, Department of Anaesthesiology, MMIMSR, MMDU, Mullana, Ambala, Haryana, Fax: India.

Abstract

Ebstein anomaly is a rare congenital disorder comprising less than 1% of patients with congenital heart diseases. Although it is rare, but this congenital cardiac anomaly is one of the most diverse in presentation, severity and management. Average life expectancy in the patients born with this anomaly is around 25-30 years. However, pregnancy in these patients is well tolerated but they are at increased risk of developing severe complications especially in the current scenario of covid 19. Here we present a case of 26-year-old, unbooked, primigravida patient with severe preeclampsia at period of gestation 32 weeks for emergency cesarean section with unknown covid status. Patient was diagnosed with ebstein anomaly after presentation to hospital.

Keywords: Ebstein anomaly, Parturient, combined spinal epidural anaesthesia, Covid 19

Corresponding Author: Neha Yadav, Assistant Professor, Department of Anaesthesiology, MMIMSR, MMDU, Mullana, Ambala, Haryana, India.

E-mail: nehayadav05@gmail.com

Received: 02 January 2021

Revised: 10 February 2021

Accepted: 18 February 2021

Published: 16 May 2021

Introduction

Ebstein anomaly is a rare congenital heart disorder with an incidence of 0.3% to 0.5%. Wilhelm Ebstein first described the anomaly in 1866. It is usually associated with malformation of the right side of heart involving primarily tricuspid valve and right ventricle with or without atrial septal defect, patent foramen ovale or supraventricular arrhythmias. It has a wide spectrum of presentation ranging from asymptomatic patients to patients with severe disease with congestive heart failure and sudden collapse as the major cause of death. Although literature suggests that pregnancy in women with Ebstein's anomaly is well-tolerated but presence of arrhythmias, cyanosis and preeclampsia are associated with increased fetal and maternal risk. Maternal mortality varies from less than 1% in asymptomatic patients to as high as 5-15%, if supraventricular arrhythmia, WPW syndrome or atrial fibrillation.^[1]

As it is a rare clinical entity with very short life expectancy at birth with very few cases reported till date, ours become an extremely rare case in view of presentation in the covid 19 pandemic times with specific anaesthetic management taking in consideration the protocols established in covid 19

pandemic.

Case Report

A 26 year old unbooked parturient G2A1 with period of gestation 32+3 weeks presented to emergency room with chief complaints of amenorrhoea for 8 months with severe headache, shortness of breath and swelling bilateral lower limbs for 4 days and vertigo for 1 day with no significant past history. Patient's covid 19 status was not known. On general examination, patient was afebrile, generalized pitting edema present all over body, mild pallor present, JVP was normal. Vitals recorded were PR- 84/min, regular, BP – 170/110 mmHg, SPO₂ -98% on room air. On auscultation, bilateral air entry was equal with no adventitious sounds. In cardiovascular examination, S1S2 were normally heard with pansystolic murmur over tricuspid area. Electrocardiography was suggestive of RBBB with p pulmonale. Chest Xray showed cardiomegaly with prominent right heart border [Figure 1]. Echocardiography findings were suggestive of dilated right atrium and atrialised right ventricle with true RV-16cm², atrialised RV-21 cm², RA-16cm², severe tricuspid regurgitation with apical displacement of tricuspid leaflet of more than 34 mm, no PS or PAH, no ASD/PFO with normal

LV and EF of 55-60%. On the basis of these findings, diagnosis of Ebstein's anomaly was made by the consulting cardiologist. Patient's blood investigations were within normal limits.

Patient was planned for emergency caesarean section in view of severe preeclampsia. We planned it to be done under combined spinal epidural anaesthesia with low dose spinal and epidural drug supplements. All emergency drugs were kept ready and proper arrangements for personal protective equipments were made with everyone wearing kits, N95 masks and face shields. After attaching standard ASA recommended monitoring, we put an arterial line for beat to beat monitoring of blood pressure and to roughly estimate pulse pressure variation during intraoperative period. SAB was given at L3-L4 space with 1 ml of 0.5% bupivacaine (heavy) with 25 mcg of fentanyl with supplementation of 10 ml (6+4ml) of 0.5% plain bupivacaine in epidural catheter. Complete sensory blockade till T6 was achieved within 12 minutes of epidural drug injection. No significant hemodynamic changes were recorded and PPV was maintained. Injection oxytocin 5 mg was given in slow iv infusion after cord clamping and the procedure got completed within 40 minutes. A 2.0 kg male baby was delivered with APGAR score of 9. Postoperative pain was managed with intermittent epidural aliquots of 6 ml of 0.125% plain bupivacaine and epidural catheter was removed at POD₂.



Figure 1: Chest X ray PA view showing prominent right heart border and cardiomegaly

Discussion

Approximately 50% of patients with Ebstein's anomaly present in neonates and infants with cyanosis and congestive

cardiac failure and those who survive till adulthood may remain asymptomatic or their condition can get deteriorated with the presence of arrhythmias or pregnancy. In presence of Ebstein's anomaly, there is impairment of right ventricular size and function with atrialisation of right ventricle. It can further get deteriorated by the increased blood volume and cardiac output during pregnancy.^[2] Increased right atrial pressure and volume both worsen tricuspid regurgitation.

Due to raised catecholamine levels in pregnancy there is further predisposition for development of arrhythmias, particularly with maternal hypoxaemia and stress. So, basic principles for anaesthetizing such patients is to maintain both afterload and preload along with a sinus rhythm.^[3]

We chose regional anaesthesia over general anaesthesia due to many reasons. As we all know by now, maximum aerosol generation takes place during intubation under general anaesthesia. So for prevention of aerosol generation, regional anaesthesia is the best alternative considering the patient's clinical status, in the present covid 19 era. And also, there is a 20% incidence of supraventricular arrhythmias during general anaesthesia induction due to maternal stress response in patients with ebstein anomaly.^[4] Factors which are known to precipitate arrhythmias like light plane of anaesthesia, acid base imbalance, hypoxia and hypercapnia which are more common under general anaesthesia were avoided under SAB. Defibrillator was kept ready to terminate any possible arrhythmias which may occur intraoperatively. Fluid status and patient's haemodynamic parameters were maintained by giving titrated fluids and appropriate drugs under IABP guidance.^[5] Good postoperative analgesia, oxygen supplementation and strict monitoring of vital parameters in the ICU ensured minimal complications.

Advantages of epidural anaesthesia are minimal intravascular volume shift, decreased catecholamine levels, control of maternal hyperventilation and most importantly, postoperative analgesia.^[1,6] Intrathecal anaesthesia may complicate a right to left cardiac shunt due to sudden decrease in sympathetic vascular resistance. As our patient did not have any intracardiac shunt, we preferred to give combined spinal epidural anaesthesia. Also, large doses of intrathecal drug can precipitate sympathetic block, decreasing right ventricular and subsequently compromised left ventricular output. So, we preferred low dose of intrathecal agent. Large doses of oxytocin have marked vasodilating effects and subsequent hypotension can be detrimental, so we used slow iv infusion. Methylethylamine and prostaglandins increase pulmonary vascular resistance which can further compromise right ventricular output, are generally avoided. Pricking of iv fluid bottles should be avoided in patients with atrial septal defect or patent foramen ovale due to the fear of paradoxical air emboli.

Our patient was lucky that she had no problem throughout the pregnancy and all throughout intrapartum and postpartum period. Connolly and Warnes analyzed the outcome of 111

pregnancies in 44 women with EA.^[7] In this report, 16 patients were cyanotic, and 20 had an interatrial communication (ASD/PFO). Majority (76%) of pregnancies resulted in live birth, 89% were delivered vaginally, and 11% by cesarean section.

Conclusion

Pregnant women with Ebstein's anomaly are high risk patients and are better cared by multidisciplinary team approach. Knowledge of pathophysiology, hemodynamic derangements, thorough preparation and diligent care results in successful outcome of such rare congenital cardiac anomalies. Regional anaesthesia may be preferred over general anaesthesia after proper patient selection which can help prevent the spread of Corona virus infection.

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How to cite this article: Yadav N, Rani K, Sandhya BK, Anand S. Anaesthetic Management of a Parturient with Incidental Diagnosis of Ebstein Anomaly in COVID 19 Era-A Rare Case Report. *Acad. Anesthesiol. Int.* 2021;6(1):32-34.

DOI: dx.doi.org/10.21276/aaan.2021.6.1.7

Source of Support: Nil, **Conflict of Interest:** None declared.