

Anesthetic management of a parturient with Ebstein's Anomaly

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Abstract

Ebstein's Anomaly (EA) is an uncommon, complex and heterogenous form of cardiac disease. It is characterised by dysplastic abnormalities of tricuspid valve which involves both basal and free attachments of the valve leaflets with downward displacement and elongation of the septal and anterior cusps with resultant tricuspid regurgitation. Proximal part of right ventricle is atrialised becoming thin walled and poorly contractile along with an enlarged right atrium. Supraventricular and ventricular arrhythmias are common, so is congestive cardiac failure, cyanosis, sudden collapse and death. This case is presented because of its rarity and varied clinical presentation and pregnancy further increase demands on the heart. A 28 year old parturient with gravid 2, para1, living1 with post dated pregnancy and leaking membranes was posted for emergency lower segment cesarean section. She was diagnosed with EA. Preoperative antibiotic prophylaxis for infective endocarditis and aspiration prophylaxis was given intravenously. The procedure was carried out under epidural anaesthesia with multi orifice epidural catheter threaded upto 5 centimeters in epidural space. Local anaesthetic agents used was bupivacaine 0.5% 10cc and lignocaine with adrenaline 2% 10 cc in a graded manner. No complications were seen during perioperative period.

Keywords: Ebstein's Anomaly, parturient, epidural anaesthesia

Introduction

Wilhelm Ebstein first described the clinical and anatomical features of an anomaly of the tricuspid valve in 1866^[1] It is a rare congenital heart defect with incidence of less than 1% of patients with cardiac defects (1 in 1,11,000 of general population)^[2] and there is no sex difference. The distinctive features are the downward displacement and elongation of septal and anterior cusps of the tricuspid valve leaflets into the right ventricle (RV) due to abnormal development of the valve leaflets^[3] Thus the proximal part of the RV is 'atrialised', becoming thin walled and poorly contractile with enlarged right atrium^[4] The considerable cardiovascular and hormonal changes during pregnancy carries a greater risk with altered cardiac physiology of Ebstein Anomaly [EA]. EA may have other associated anomalies like patent foramen ovale, with intracardiac shunting, pulmonary hypertension, ventricular or supraventricular arrhythmias and particularly associated with WPW syndrome (up to 20%) (2)(5), and cyanosis. CCF and

sudden cardiac collapse are the most common causes of death^[5].

Case report

A 28 year old multigravida with gravid 2 para 1 and one living male child with diagnosis of Ebstein's anomaly came with post dated pregnancy of 40 weeks and 4 days of gestation with premature rupture of membranes of six hours duration was posted for emergency lower segment cesarean section in view of fetal distress. Preanaesthetic check up was done.

Her general physical examination revealed a blood pressure of 130/84, heart rate of 102/minute, no cyanosis, conjunctiva appeared pale, no pedal oedema and a normal JVP. On palpation thrill was felt in right parasternal area. Liver was not palpable. Oxygen saturation was 97% on room air, On auscultation first and second heart sounds was normal, pansystolic murmur was heard in the tricuspid area. Respiratory system had bilateral vesicular breath sounds. ECG finding revealed a sinus tachycardia with tall and

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broad P waves in lead 2 and V1 because of right atrial enlargement and right bundle branch block (Fig-1). Chest x ray showed enlarged right atrium with cardiomegaly, Echocardiography showed moderately enlarged right atrium with tricuspid regurgitation and downward displacement of the tricuspid valve leaflets, right ventricle was small and a large atrialised right ventricle which confirmed EA (Fig-2). There was no other associated anomalies. Her blood reports were within normal limits. Ultrasonographic finding showed 40 weeks plus 4 days with expected birth weight of 3 kgs.

Crash cart with all emergency drugs and equipments kept at hand. Monitoring was done with ECG, Pulse oximetry, non invasive blood pressure monitoring and urine output. O₂ with face mask at six litres of oxygen was started. Intravenous line was secured with 20G cannula. Inj. ranitidine 150 mgs, inj. metaclopramide 10 mgs was administered for acid aspiration prophylaxis. Inj ceftriaxone and salbactam 1.5 gms was given bolus to prevent infective endocarditis. Central line was avoided to prevent arrhythmias, paradoxical air embolism, and infective endocarditis.

Epidural anesthesia was given with 18g Tuohy needle at L3-4 level with the patient in left lateral position and epidural catheter was threaded upto 5 centimeters inside the epidural space. A test dose of local anaesthetic lignocaine 3 ml of 2% containing 5 micrograms of adrenaline per ml was administered before titrated doses of 0.5% bupivacaine and 2% lignocaine with adrenaline was given to achieve a sensory level upto T6 and until patient could not move the legs. Her vitals well closely monitored. Aortocaval compression was avoided by lateral tilt of the table and displacement of gravid uterus. Epidural catheter was used for providing post-operative analgesia for next 48hours.

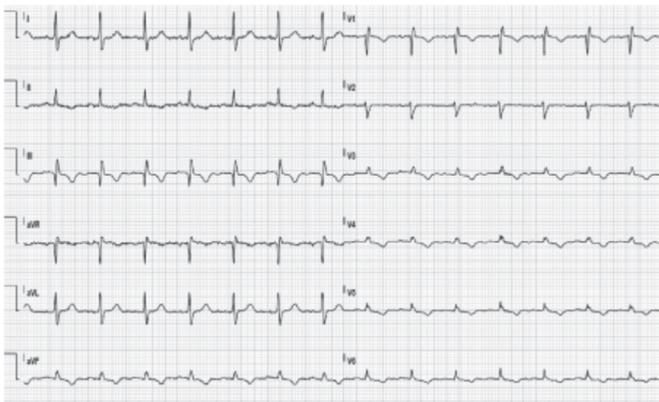


Figure 1: ECG findings

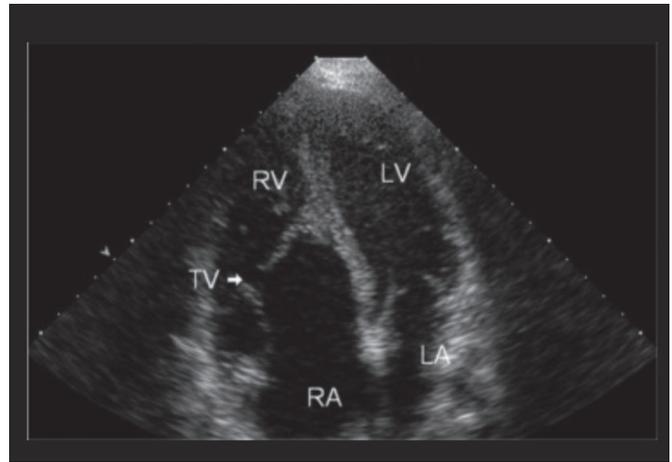


Figure 2: ECHO Findings

The drug used was bupivacaine 0.125% 10ml with inj buprenorphine 90 micrograms once daily for two days and epidural catheter was removed on third post-operative day.

Discussion

The severity of EA can be described anatomically as mild, moderate or severe based on echocardiographic appearance with displacement and tethering of the leaflets and the degree of right ventricular dysfunction^[6].

In EA there is compromised right ventricular size and function which is further impaired by the physiological changes of the pregnancy within the cardiovascular system. Plasma volume increases with a smaller rise in red blood cell mass. Cardiac output is raised by 40% with an increase in both stroke volume and HR and with raised catecholamine levels in pregnancy, further predispose to arrhythmias especially with maternal hypoxia and stress. Circulating catecholamines, maternal hemodynamic instability, hypoxemia with congenital cardiac defects are associated with prematurity, low birth weight, congenital heart disorders (9-14%) and poor neonatal outcomes^[7].

Pregnant patients with EA are also at high risk of major adverse cardiac events like in hospital death, acute myocardial infarction, heart failure, or arrhythmias and postpartum hemorrhage.

Basic principles of anesthetic management are to maintain preload and after load and maintain sinus rhythm to prevent increased right to left shunting, which may occur if systemic vascular resistance is decreased or if pulmonary vascular resistance is increased or with increased intrathoracic pressure^[8]. Finally tachycardia is avoided as it further impair right ventricular filling. The advantages offered by epidural analgesia are minimal intravascular fluid volume

shifts, decreased catecholamine levels, control of maternal hyperventilation and most importantly continuation of analgesia into post-operative period.

Although general anaesthesia offers advantages of better hemodynamics and easily controllable fluid balance, the disadvantages are increased induction time with right to left shunt (increasing aspiration risk), increase in catecholamine levels, increase intrathoracic pressure which increase intracardiac blood shunting^[9].

Spinal anesthesia is not advised as it causes sudden hypotension due to sharp fall in systemic vascular resistance and has major impact on cardiac output. There is dramatic decreases in preload and hence adversely affect contractility of right atrium and RV.

Conclusion: Patients with EA presenting with multitude of problems should be treated as high risk and cared in a tertiary centre under multidisciplinary team of obstetrician, cardiologist and obstetric anesthetist for safe pregnancy and delivery. There is limited literature on the case presented here. A better understanding of pathophysiology of cardiovascular disease, their effect on pregnancy, thorough preparation and close monitoring are the keys for successful outcome. Graded epidural anesthesia provides a stable hemodynamics during perioperative period and may be preferred to a general anaesthesia in these patients.

References

1. Mann RJ, Lie TT. *The life story of wilhelm Ebstein (1836-1912) and his almost overlooked description of a congenital heart disease. Mayo clin proc 1979; 54: 197-203.*
2. Chatterji S, Sengupta I, Mandal R, Sarkar R, Chakarborty P. *Anesthetic Management of CS in a patient with EA. Indian J Anesthesia 2008; 52: 321-3.*
3. Dearani JA, Danielson GK. *Surgical management of Ebstein's anomaly in the adult. Semin thoracic cardiovasc surg 2005; 11: 148-54*
4. Misa VS, Pan PH. *Evidence based case report for analgesic and anesthetic management of a parturient with EA and WPW Syndrome. Int J abstract Anesth 2007; 16; 77-81*
5. Khatib SK, Tadwalkar GV. *Anesthetic implications of EA: A report of two cases Anesthesia pain and intensive care 2012; 16 (1): 60-63*
6. Sushma KS, Shaikh S, Ashwini HR. *Anesthetic management in a parturient with EA. J Obstet Anesthesia crit care. 2013; 3: 101-3.*
7. Khairy P, Oxyang DW, Fernandes SM, Lee-Parritz A, Economy KE, Landzberg MJ. *Pregnancy outcomes in women with cong heart diseases. Circulation 2006;113:157-24.*
8. Ellison N. *Congenital heart disease. In: Stoelting RK, Dierdorf SF (editors). Anaesthesia and co-existing disease. 3rd ed. Churchill Livingstone;1993. p. 37-56.*
9. Groves ER, Groves JB. *Epidural analgesia for labour in a patient with Ebstein's Anomaly. Canadian J Anesth 1995;42:77-9.*

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