Management Of A Parturient With History Of Ebstein`s Anomaly And Protein S Deficiency

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Citation

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Abstract

Ebstein's anomaly (EA) is a rare congenital cardiac defect associated with both displacement and incompetence of the tricuspid valve. A patient with EA during pregnancy and anesthesia has potential for a variety of hemodynamic disturbances. The condition is commonly complicated by supraventricular tachycardias[1]. We describe the management of a patient with this condition, atrial Septal defect (ASD) and also with history of Protein S deficiency undergoing caesarean section (C-section). This case illustrates the importance of careful attention to the preoperative findings, perioperative hemodynamic parameters and postoperative management.

INTRODUCTION

Ebstein's anomaly (EA) is a rare congenital cardiac defect associated with both displacement and incompetence of the tricuspid valve. A patient with EA during pregnancy and anesthesia has potential for a variety of hemodynamic disturbances. The condition is commonly complicated by supraventricular tachycardias[1]. We describe the management of a patient with this condition, atrial Septal defect (ASD) and also with history of Protein S deficiency undergoing caesarean section (C-section). This case illustrates the importance of careful attention to the preoperative findings, perioperative hemodynamic parameters and postoperative management.

CASE REPORT

The patient is a 22-year-old African-American woman Gravida 3 Para 2 at 29 weeks gestation with past medical history significant for EA, ASD with right to left (R□L) shunting, protein S deficiency, previous history of deep venous thrombosis (DVT) and pulmonary embolism (PE) times two during her previous pregnancies. She was admitted to the OB/GYN service with complaints of chest pain, shortness of breath, and palpitations. She was at that time on 3 liter oxygen nasal cannula at home. She continued to have increased O2 requirements, had to be placed on 100% non-rebreather
and was transferred to the intensive care unit (ICU). She was placed on a heparin drip. A Greenfield IVC filter and right internal jugular CVL were also placed before surgery in the radiology suite. The patient again had oxygen desaturations. It was decided to take her to the operating room for urgent C-section secondary to persistant hypoxia. Pre-op echocardiogram showed severe tricuspid regurgitation, ASD, and RVSP around 57 mmHg. An arterial line was placed in the preoperative area and the patient was intubated after controlled induction maintaining hemodynamic stability through out the procedure. The patient was transported to ICU intubated. The patient was extubated on post-op day (POD) 2. She was re-intubated on POD 3 due progressive hypoxia and respiratory failure. Repeat echocardiogram showed increased RVSP to 123mmHg. She was started on nitric oxide and RVSP decreased to 35-40 mmHg. On POD 10 patient underwent Modified Danielson Procedure and ASD closure and placement of DDD epicardial device complicated by pleural and pericardial effusion requiring chest tube placement. Patient was subsequently discharged home on POD 30 with oxygen saturation in 98-100% on room air.

DISCUSSION

EA is a rare congenital malformation of the tricuspid valve. This malformation, associated with an atrial septal defect, can have an accessory conduction pathway (Wolf-Parkinson White syndrome). Death occurs usually from cardiac arrhythmias.

The importance of echocardiographic evaluation is vital and the probability of maternal and neonatal events may be predicted from the baseline characteristics of the mother.

Pregnancy is well tolerated in the absence of severe maternal cardiomegaly, cyanosis and arrhythmias and in those patients with mild cardiac dysfunction as evaluated at echocardiography and a low NYHA class, but it is associated with an increased risk of abortions and prematurity [2-4]. These patients should be evaluated with serial echocardiogram during the course of their pregnancy. The basic goals in these patients are to maintain both afterload and preload and maintaining sinus rhythm. One should be very cautious about paradoxical air embolism during the peri-operative period as well as with surgical manipulation of uterus itself because of R□L shunt. A high index of suspicion and intraoperative use of precordial doppler or transesophageal echocardiogram will permit earlier diagnosis and timely intervention. In patients with Ebstein's anomaly there is a need for early involvement of subspecialties.

References

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