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Case Report

Epidural Anesthesia for Cesarean Section in a Patient with Severe Familial Restrictive Cardiomyopathy -Case Report and Review of the Literature

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Abstract

Restrictive familial cardiomyopathy (RCM) is a rare cardiac disease which imposes serious risks on the parturient, especially during labor and delivery. I describe a case of a patient with severe RCM who successfully underwent emergency cesarean section utilizing epidural anesthesia. Other similar cases in the literature were reviewed and discussed. To the best of my knowledge this is the first case report describe the anesthetic management of the restrictive type of this disease.

INTRODUCTION

For patients with severe cardiac disease, unique risks are posed when they undergo the physiologic changes of pregnancy, labor, and delivery. Patients with severe cardiac disease have traditionally been strongly discouraged from pregnancy due to high risk of mortality but has changed since major advances in medical care. Here, I describe the first case of a parturient with severe restrictive familial cardiomyopathy admitted at 29 weeks gestation with rapid atrial fibrillation requiring an emergency cesarean section with epidural anesthesia.

CASE REPORT

A 31-year-old G_1P_0 parturient was admitted to my hospital at 29 weeks of gestation for management of symptomatic recurrent atrial fibrillation with rapid ventricular response without symptoms of cardiac failure exacerbation. Past medical history included familial restrictive cardiomyopathy (RCM) requiring hospital admissions for cardiac failure exacerbations, insertion of an automatic implantable cardioverter defibrillator (AICD), recurrent atrial fibrillation and deep venous thrombosis. Regular medications included sotalol, potassium chloride, furosemide, subcutaneous enoxaparin 80 mg twice a day, digoxin, and prenatal vitamins. During this admission, amiodarone was commenced and digoxin dose reduced. Due to nonreassuring fetal well-being (episodes of fetal bradycardia and decreased long term variability), the patient was scheduled for urgent cesarean section. Preoperative monitoring included central venous and pulmonary arterial catheter. Her preoperative vital signs were: BP 100/70 Atrial fibrillation with rate of 89. SaO_2 98% on room air. Right ventricular pressure 35/15. Right atrial pressure 15. Pulmonary arterial pressure 45/22. Pulmonary artery occlusive pressure 22. Her complete blood count and coagulation studies were within normal limits. Slow onset epidural anesthesia was chosen over general anesthesia and spinal anesthesia because epidural anesthesia has a better safety profile for parturients than general anesthesia and more hemodynamically stable than spinal anesthesia. Lumbar epidural was easily placed at L3-4 level using 18 gauges Touhy needle in a sitting position. The patient was placed in supine position with left uterine displacement to avoid aortocaval compression and minimize the risk of obstruction of venous return. Test dose of 2 mL of ropivacaine 0.5% was negative. The lumbar epidural anesthesia slowly titrated, total of 10 ml of ropivacaine 0.5% and 5 ml of lidocaine 2% was required to establish a T6-8 block to ice. The epidural took 30 minutes to set up, and during that time a radial arterial line was inserted. Her intraoperative hemodynamics were stable (maximum HR was 90 and lower BP 90/60). Baby girl (1300 gm) was delivered with an Apgar score of $5^1,7^5$. Total crystalloid fluid was given 1200 mL, most of the fluids was given in the first hour on the anesthetic. Estimated blood loss was 600 mL. After anticoagulation and cardioversion, the patient was discharged without sequalae.

DISCUSSION

Restrictive cardiomyopathy is a rare disease. It is accounting for 5% of all primary myocardial diseases [1]. It characterized by increased stiffness of ventricles, which leads to left ventricular

diastolic dysfunction. Systolic function usually remains normal, at least early in the disease. Increased stiffness of the myocardium causes pressure within the ventricle to rise precipitously with only small increases in volume, thereby leading to a hemodynamic profile similar to that of constrictive pericardial disease [2]. RCM may develop in association with inflammatory, infiltrative, or storage disease. It may also be idiopathic. A significant number of those who manifest RCM without systemic disease may be caused by mutations in sarcomeric disease genes (*TTNI3*) that may be associated with hypertrophic, dilated, and noncompaction cardiomyopathy [3]. Adult patients will often present with dyspnea, fatigue and limited exercise capacity. Patients frequently exhibit biatrial enlargement with resultant atrial arrhythmias and thromboembolic complications [2]. The prognosis of this disease is very poor, such 32-44% of adults who are diagnosed with this disease die of cardiovascular related death within five years, often of heart failure or embolic stroke [3,4].

Anesthetic management of patients with RCM for non-cardiac, non-obstetric procedure has been reported. For example, Nishida et al described the anesthetic management for a patient underwent general anesthesia for gastrectomy. He used a combination of dopamine, nitroglycerin infusions and prostaglandin E infusions [5]. There is little, if any, medical literature specifically addressing the peripartum management of RCM during. Pryn et al described two cases with peripartum cardiomyopathy and the two with hypertrophic obstructive cardiomyopathy. Three of those cases underwent cesarean section with neuraxial (combined spinal-epidural, epidural) and one case with general anesthesia. The neuraxial anesthesia group did well without significant complications. The general anesthesia patient decompensated at the end of the surgery and was kept in the intensive care unit till she received heart transplantation almost six weeks later [6]. Thaman et al followed up 127 consecutively referred women with hypertrophic cardiomyopathy, patients had uncomplicated general and epidural anesthesia for their cesarean section [7]. Toda et al described a case of combined spinal-epidural anesthesia for cesarean section in a patient with dilated cardiomyopathy. They used 5 mg of bupivacaine mixed with 10 mcg of Fentanyl for the spinal anesthesia. The epidural anesthesia was titrated slowly. The patient had good anesthesia and analgesia without complications [8]. Frost et al reported a case of slowly titrated epidural anesthesia with different concentration of bupivacaine for a patient with familial cardiomyopathy and cardioveter-defibilator [9]. Logically; one may aim for an anesthetic plan similar to that for a patient with constrictive pericarditis, since they have similar hemodynamic profiles. Thus, it may be wise to keep the patient "fast, full, and strong" [3], because stroke volume is relatively fixed, it is important to maintain sinus rhythm and to avoid any significant decrease in heart rate. A slightly fast heart rate may be beneficial. Fluid management involves maintaining a delicate balance. On one hand, preload must be maintained in order to preserve cardiac output. However, fluid overload is detrimental, since there is a pressure limitation in diastolic filling [10]. Echocardiography may be useful to guide fluid management and use of pressors [11]. As echocardiography more accurately estimates left ventricle volume, as opposed to pulmonary artery catheter which estimates it indirectly using pressure which can be affected by the left ventricle compliance. Sachs et al reported successful vaginal delivery with epidural anesthesia in a patient with constrictive pericarditis [12]. In conclusion; anesthetic management for cesarean section in patient with severe restrictive cardiomyopathy requires clear understanding the pathophysiology of this disease and the cardiovascular changes during pregnancy and labor. We believe that a slowly dosed epidural may avoid major changes in preload, heart rate, and contractility and maintain hemodynamic stability in this group of very high-risk obstetric patients. When neuroaxial anesthesia is not an option, general anesthesia with close invasive monitoring including pre-induction arterial line, trans-esophageal echocardiography or pulmonary artery catheter can be utilized.

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