Perioperative use of transthoracic echocardiography in a patient with congenitally corrected transposition of great arteries, atrial septal defect and severe pulmonary stenosis for lower segment cesarean section

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Abstract

A 25-year-old female with congenitally corrected transposition of great arteries (CCTGAs), atrial septal defect, and severe pulmonary stenosis underwent lower segment cesarean section at 34 weeks of gestation using combined spinal epidural anesthesia (CSEA). We used transthoracic echocardiography (TTE) for intraoperative monitoring of the cardiovascular system because these patients are reported to have a high prevalence of myocardial perfusion defects, regional wall motion abnormalities, and impaired ventricular contractility. Scanning was done at four different time intervals; preoperatively, after initiation of CSEA, after delivery of child and postoperatively (6 and 24 h postdelivery) to detect regional wall motion and valvular abnormalities, calculate ejection fractions and optimize fluid administration. In this case report, we thus discuss the anatomical defects of CCTGA, physiologic concerns and emphasize on the use of TTE for perioperative management of such cases.

Key words: Anesthesia, arteries, cesarean section, congenitally corrected transposition of the great arteries, combined spinal epidural anesthesia, complications, labor, obstetric, pregnancy

Introduction

In congenitally corrected transposition of the great arteries (CCTGAs) the stronger left ventricle (LV) with the mitral valve pumps blood to the lungs and the weaker right ventricle (RV) with the tricuspid valve pumps blood to the entire body. The arteries coming off the ventricles are also inverted and so the blood flow pathway is normal. The associated lesions of the valve, septum, and electrical conduction pathway complicate the patient’s cardiovascular physiology and anesthetic management.

Case Report

A 25-year-old primigravida presented to the hospital at 33 weeks of gestation with history of dyspnea on exertion (NYHA II), frequent chest infections and palpitations for the last 4-5 years. On examination, she had peripheral cyanosis and clubbing. Ejection systolic murmur was present on cardiac auscultation. Airway and spine were normal on examination. Biochemical and hematological investigations, chest X-ray and electrocardiogram (ECG) were within normal limits. Echocardiography reported an ostium secundum ASD (25 mm) with bidirectional shunt, atrioventricular/ventriculoarterial discordance, transposition of great arteries, thickened pulmonary valve, PS with a peak gradient across pulmonary valve = 93 mmHg. Patient was...
Discussion

Congenitally corrected transposition of the great artery is a rare disorder with an incidence of 1/33,000 live births (0.05% of all congenital cardiac malformations). It is associated with ventricular septal defect in 78%, PS in 50% and ASD in 20% of the cases. It is due to discordant ventriculoarterial and atrioventricular alignments during embryogenesis. The double discordance physiologically corrects the discordance intrinsic to each malalignment and thus systemic venous blood from the right atrium enters the pulmonary artery via LV and pulmonary venous blood enters the aorta from the left atrium via RV. Thus, the patient will be asymptomatic if there is no other associated anomaly. Cyanosis will be present if there is PS and a ventricular septal defect while heart failure will develop in patients with hemodynamically significant ventricular septal defect.

Our patient had CCTGA with PS and ASD; the co-existence of PS increased the LV work and decreased the volume of blood circulated to the pulmonary vasculature. Right to left shunt through the ASD led to cyanosis. PS in a patient with an anatomically oriented heart is poorly tolerated as the RV is not accustomed to pumping at high pressures. This is in contrast to patients with CCTGA in which the PS is relatively better tolerated as the LV is accustomed to work against high pressures. However, still the risk of contractile dysfunction is very high, and this necessitates the need of peri-operative TTE.

Parturients with cyanotic heart disease have been reported to have a high incidence of cardiovascular complications (32%). The likelihood of a live birth is reported to be higher in patients with arterial oxygen saturation >85% at rest and hemoglobin <20 g/dL at the start of pregnancy.

Goals of anesthesia in our patient were maintenance of preload and prevention of the increase in pulmonary vascular resistance, systemic vascular resistance (SVR), heart rate and myocardial contractility. This was accomplished by preventing hypoxia, hypercarbia, hypothermia, acidosis and high peak airway pressures.

In our patient, the administration of CSEA led to a decrease in preload and SVR and this was clinically evident by the development of hypotension. It was initially managed by phenylephrine infusion, but TTE helped in identifying the etiology, which is hypovolemia and facilitated discontinuation of the vasopressor after appropriate volume expansion. Static and dynamic parameters of fluid responsiveness may not be accurate in a parturient with CCTGA. Echocardiography is an ideal noninvasive tool for optimum fluid management in such cases.

Table 1: FATE protocol

| Views                        | Measurements taken                                      |
|------------------------------|--------------------------------------------------------|
| Parasternal long axis        | LVEDD, LVSD, FS, RWMA, IVS, PW, RV                     |
| Parasternal LV short axis    | FAC, RWMA                                              |
| Apical four chamber          | EF, RWMA                                               |
| Subcostal four chamber       | To confirm finding of apical four chamber              |
| Subcostal vena cava          | IVC diameter, phasic respiratory collapse of IVC       |

LVEDD = Left ventricular systolic diameter, LVSD = Left ventricular end-diastolic diameter, FS = Fractional shortening, RWMA = Regional wall motion abnormality, IVS = Interventricular septum, PW = Posterior wall, RV = Right ventricle, EF = Ejection fraction, FAC = Fractional area change, IVC = Inferior vena cava, FATE = Focus assessed transthoracic echocardiography
Interest in the application of TTE to anesthesia is now developing, and various training courses for perioperative and critical care physicians have been established.\textsuperscript{8} Feasibility study conducted by Cowie\textsuperscript{11} on the perioperative use of FATE by anesthesiologists has reported a change in management in 84\% of the patients. Reliability of the echocardiographic examinations has been reported to be 87\% and this favors the perioperative use of TTE examinations for hemodynamic stabilization of patients with congenital cardiac lesions.

It is of paramount importance to assess the systemic (morphologic right) ventricle for systolic and diastolic dysfunction in patients with CCTGA. End diastolic volume, stroke volume and cardiac output (reaches approximately 140\% of pre-pregnancy level by 20-28 weeks of gestation) increase significantly during pregnancy and thus the systemic (morphologic right) ventricle in patients with CCTGA may dilate and decrease in function.\textsuperscript{9} This increases the risk of congestive heart failure and pulmonary edema during the peripartum period.\textsuperscript{10} TTE is the best modality for cardiovascular assessment of such patients; pulmonary artery catheter placement may result in cardiac dysrhythmias and is thus avoided. Its use is further limited in patients with severe sub pulmonary and pulmonary valve stenosis.

We administered antibiotic prophylaxis against IE in our patient because of an institutional protocol. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease recommend antibiotic prophylaxis (before vaginal delivery and at the time of membrane rupture) in only selected patients with CCTGA:\textsuperscript{11}

1. Patients with prosthetic cardiac valve or a prosthetic material used for cardiac valve repair.
2. With unrepaired and palliated cyanotic congenital heart disease, including surgically constructed palliative shunts and conduits.

Prophylaxis against IE is not recommended for non-dental procedures in the absence of active infection.

Thus to conclude anesthetic management in patients with CCTGA should be tailored on an individual basis depending on the type and severity of associated intracardiac lesion, systemic ventricular dysfunction and rhythm disturbance. Patients with isolated CCTGA and good functional capacity are at low risk, but patients with symptoms of heart failure, cyanosis or arrhythmia are at high risk for peri-operative adverse cardiovascular events. These patients are extremely difficult to resuscitate if cardiac arrest occurs as external chest compression is not effective in forcing blood across a stenotic pulmonary valve. Decreases in systemic blood pressure need to be promptly treated with sympathomimetic drugs. Cardiac dysrhythmias leading to hemodynamic compromise need to be rapidly corrected with antiarrhythmics; An electrical defibrillator should be available when anesthesia is administered to patients with PS. Patients with severe PS can be administered either general or regional anesthesia. CSEA is advantageous as it obviates the risk of aspiration, difficult tracheal intubation and prevents pressor response to laryngoscopy and intubation. However, it decreases SVR and may lead to hypotension, which can be minimized by optimization of volume status of the parturient. Prompt detection and management of a cardiogenic cause of hemodynamic compromise is paramount for safety of the mother and the fetus. We thus advocate the peri-operative use of TTE in parturients with complex congenital cardiac lesions.

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Conflicts of interest
There are no conflicts of interest.

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