

# 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.

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## 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

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on drug combination of amiloride and furosemide for the last 1-month. There was no history of any previous surgery or drug allergy. She was scheduled for an elective cesarean section at 34 weeks in view of severe intrauterine growth restriction.

Prophylaxis against aspiration pneumonitis and infective endocarditis (IE) was administered, and the patient shifted to the operation theatre after 8 h of fasting for solids. Right radial artery was cannulated for invasive pressure monitoring in addition to routine monitors; ECG and plethysmography. Baseline heart rate and blood pressures were normal, but saturation was 83%. Venturi mask was attached to  $\text{FiO}_2$  of 0.5 and wedge was placed under right hip. A baseline focus assessed transthoracic echo (FATE) was done [Table 1].<sup>[1,2]</sup>

Combined spinal epidural anesthesia (CSEA) was planned. Co-loading was done with 10 ml/kg of fluid. Epidural catheter was placed in L3-L4 interspace with the patient in the left lateral position. Spinal anesthesia was administered with 1.5 ml of 0.5% hyperbaric bupivacaine and 25  $\mu\text{g}$  of fentanyl with 26 gauge spinal needle. Epidural volume expansion was done with 5 ml of 2% lignocaine, and T4 level of sensory block was achieved. Phenylephrine infusion was initiated at 60  $\mu\text{g}/\text{min}$  and titrated to maintain blood pressures within 20% of baseline. Intraoperative transesophageal echocardiography (TTE) done after CSEA was suggestive of hypovolemia, and a bolus of 500 ml of NS was given, which facilitated discontinuation of phenylephrine infusion.

Baby cried immediately at birth and was born with weight of 1.9 kg. Methylergometrine (0.2 mg) and oxytocin (10 units in 500 ml normal saline at 100 ml/h) were administered. No episode of desaturation or bradycardia was observed during the intraoperative period. TTE was repeated after delivery of child and postoperatively (6 and 24 h postdelivery) to detect regional wall motion and valvular abnormalities, systolic/diastolic dysfunctions and optimize fluid status. Epidural catheter was removed on 2<sup>nd</sup> postoperative day and the patient was administered low molecular weight heparin for deep vein thrombosis prophylaxis.

**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

LVSD = Left ventricular systolic diameter, LVEDD = Left ventricular end-diastolic diameter, FS = Fractional shortening, RWMA = Regional wall motion abnormality, IVS = Inter ventricular septum, PW = Posterior wall, RV = Right ventricle, EF = Ejection fraction, FAC = Fractional area change, IVC = Inferior vena cava, FATE = Focus assessed transthoracic echocardiography

## 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.<sup>[3]</sup> 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.<sup>[4]</sup>

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.<sup>[5]</sup>

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.<sup>[6]</sup> 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.<sup>[7]</sup>

Interest in the application of TTE to anesthesia is now developing, and various training courses for perioperative and critical care physicians have been established.<sup>[8]</sup> Feasibility study conducted by Cowie<sup>[1]</sup> 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.<sup>[9]</sup> This increases the risk of congestive heart failure and pulmonary edema during the peripartum period.<sup>[10]</sup> 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.<sup>[11]</sup>

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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