DOI: 10.21088/ijaa.2349.8471.12325.7

Indian Journal of Anesthesia and Analgesia
Volume 12 Number 3 July - September 2025

CASE REPORT

Anaesthesia for Posterior Sagittal Anorectoplasty in a Child with Congenitally Corrected Transposition of Great Arteries

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HOW TO CITE THIS ARTICLE:

Sandhya Babu, Swati Chhatrapati, Mano Praveen. Anaesthesia for Posterior Sagittal Anorectoplasty in a Child with Congenitally Corrected Transposition of Great Arteries. Ind J Anesth Analg. 2025; 12(3): 217-221.

ABSTRACT

Congenitally corrected Transposition of the great Arteries (cc TGA) is a rare congenital cardiac anomaly in which the aorta and pulmonary artery are transposed, presenting unique anesthetic challenges. The patient's cardiac physiology, characterized by atrioventricular and ventriculoarterial discordance characterized by abnormal ventricular loading and potential for arrhythmias, required meticulous pre-operative evaluation and careful anesthetic planning. We report the peri-operative anesthetic management of a 3 year old child with ccTGA scheduled for a posterior sagittal anorectoplasty (PSARP) for an anorectal malformation under general anaesthesia in prone position.

KEYWORDS

• Congenital heart disease • Arrhythmia • cc TGA • PSARP

INTRODUCTION

Congenitally corrected transposition of great arteries (cc TGA) is a complex Congenital Heart Disease (CHD) and accounts for approximately 0.5-1% of total CHD population[1]. It involves atrioventricular and ventriculoarterial discordance, resulting in physiologically normal systemic and pulmonary circulation. However, the morphological right ventricle act as the systemic ventricle, predisposing patients to progressive ventricular dysfunction, tricuspid regurgitation, and arrhythmias.

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➢ **Received:** 03-05-2025 ➢ **Accepted:** 16-06-2025

CASE REPORT

We present the case of a 3-year-old male child, weighing 13.5 kg, diagnosed with visceroarterial situs inversus and congenitally corrected transposition of the great arteries (cc TGA) with a history of anorectal malformation (ARM). The child was scheduled for a posterior sagittal anorectoplasty (PSARP). He had previously undergone a sigmoid colostomy on day one of life for the management of ARM. Since birth, the child remained asymptomatic with respect to cardiovascular function and exhibited normal developmental milestones.

On preoperative evaluation, the child was hemodynamically stable. Cardiovascular examination revealed a systolic murmur best heard over the mitral and pulmonary areas.

Transthoracic 2D echocardiography revealed visceroarterial situs inversus with levocardia. A small patent foramen ovale (PFO) with a right-to-left shunt was noted. The study also confirmed the presence of cc TGA with both systemic veins draining into the left-sided morphological right atrium. There was moderate valvular and subvalvular pulmonary stenosis, along with mild right atrioventricular valve regurgitation. Biventricular function was reported to be good.

Electrocardiogram findings included a QS complex in leads I, aVL, and V4–V6, with Q waves in leads II, III, and aVF. The axis was northwest, and poor R wave progression was observed and the findings were consistent with the underlying cardiac anomaly (Figure 1). A chest radiograph appeared normal, with no evidence of cardiomegaly or pulmonary congestion (Figure 2).

Given the complex cardiac anatomy and history of ARM, the child was planned for PSARP under careful multidisciplinary monitoring.

Perioperative Management

The child was admitted prior to the scheduled surgical procedure for preoperative evaluation and optimization. On the day of surgery, preoperative fasting status was confirmed, and a detailed high-risk informed consent was obtained from the guardians.

Peripheral intravenous access was secured using 22G and 24G cannulas, and intravenous fluids were administered to compensate for preoperative fasting losses.

Standard intraoperative monitoring included electrocardiography (ECG), pulse oximetry, non-invasive blood pressure (NIBP), capnography, central venous pressure (CVP) monitoring, temperature monitoring, and perfusion index (PI) monitoring. Given the presence of congenital heart disease, a prophylactic dose of antibiotics for infective endocarditis prophylaxis was administered preoperatively.

The planned anesthetic technique was general anesthesia with a caudal block for intra and post-operative analgesia.

Premedication in the operating room included:

i. Inj. Glycopyrrolate 0.004 mg/kg IV
ii. Inj. Fentanyl 1 mcg/kg IV
iii. Inj. Ketamine 0.5 mg/kg IV

Induction of anesthesia was achieved using:

i. Inj. Etomidate (dose titrated to effect)
ii. Inj. Atracurium 6 mg IV for neuromuscular blockade
iii. Anesthesia was maintained with a mixture of oxygen and air in a 1:1 ratio, sevoflurane at a MAC of 0.9, and intermittent doses of Atracurium as required.

Following induction, ultrasound-guided right internal jugular vein (IJV) cannulation was performed to secure central venous access. A landmark-guided caudal block was administered using 0.5 mL/kg of 0.25% bupivacaine.

The child was then positioned in prone position appropriately, and the planned posterior sagittal anorectoplasty (PSARP) procedure was initiated (Figure 3). All pressure points, including the eyes, were carefully padded. Normothermia was maintained throughout the procedure using a hot air warmer and fluid warmer. Intravenous hydration was guided by CVP monitoring to ensure euvolemia. Special care was taken to prevent the introduction of air bubbles during IV administration, considering the presence of a right-to-left shunt. (Figure 4)

The total duration of the surgery was five hours. During the procedure, 230 ml of intravenous fluids were administered. The estimated blood loss was approximately 60 ml, which was replaced with 60 ml of colloids. The total urine output recorded was 80 ml.

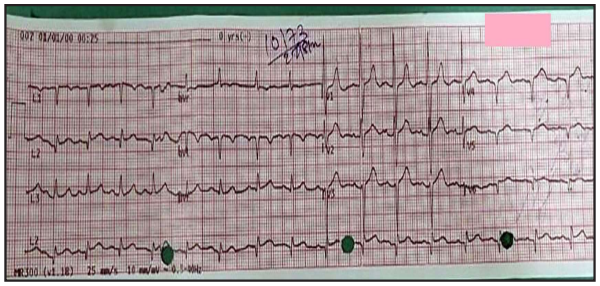

Figure 1: ECG

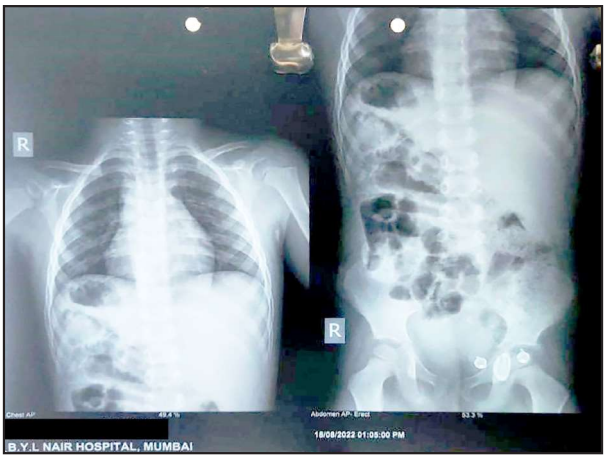

Figure 2: X-ray

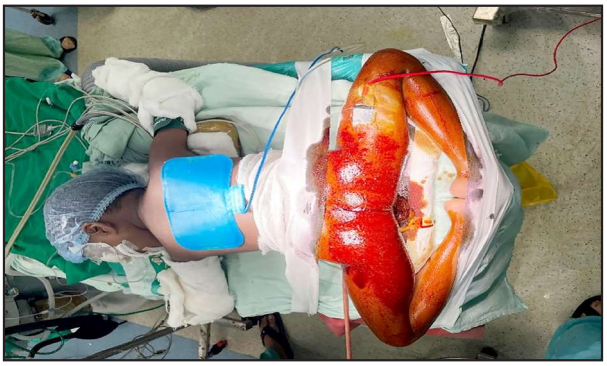

Figure 3:

Figure 4: Monitoring

DISCUSSION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare and complex congenital heart defect characterized by atrioventricular (AV) and ventriculoarterial (VA) discordance (Figure 5). In this anomaly, the morphologic right atrium connects to a morphologic left ventricle, which then connects to the pulmonary artery, while the morphologic left atrium connects to a morphologic right ventricle, which supports the systemic circulation by connecting to the aorta. This double discordance results in physiologically corrected circulation; however, the systemic ventricle being a morphologic right ventricle (RV) poses long-term challenges, especially during periods of stress such as surgery[2] (Figure 6).

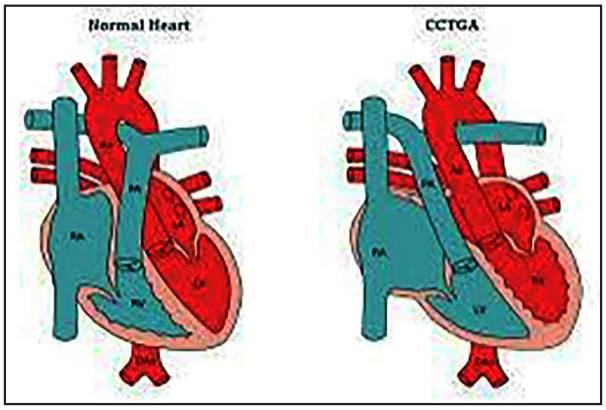

Figure 5

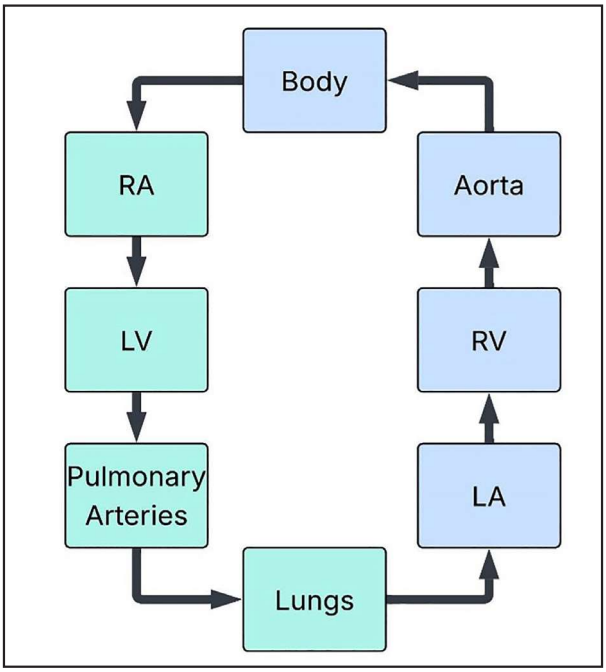

Figure 6:

Anesthetic management in patients with ccTGA is particularly challenging due to the risk of progressive systemic RV dysfunction, tricuspid valve regurgitation, and conduction abnormalities, including complete heart block. More than 30% of patients with ccTGA develop significant conduction defects by adolescence, and approximately 2% develop complete AV block each year.[3] Thus, a thorough preoperative cardiac assessment, including echocardiography and electrocardiography, is essential to evaluate ventricular function, AV valve integrity, and rhythm status.

For procedures such as posterior sagittal anorectoplasty (PSARP), although not directly involving the cardiovascular system, anesthetic management must prioritize the preservation of systemic cardiac output, avoidance of arrhythmias, and meticulous hemodynamic control. The systemic RV is more sensitive to afterload than a morphologic left ventricle; therefore, sudden increases in systemic vascular resistance or decreases in preload can precipitate heart failure.[4]

The choice of anesthetic agents should reflect the need for myocardial stability. Sevoflurane is commonly used due to its favorable cardiovascular profile and minimal myocardial depression when titrated carefully.[6] High-dose opioids (fentanyl or remifentanil) offer profound analgesia with minimal effect on myocardial contractility, making them suitable adjuncts for maintenance anesthesia. Neuromuscular blockade with non-depolarizing agents like rocuronium can be used safely. Avoidance of bradycardia is essential, especially in patients with pre-existing conduction abnormalities; anticholinergics should be readily available to manage vagally mediated bradycardia during induction or surgical stimulation.

Invasive arterial monitoring is not always required for low-risk cardiac patients undergoing non-cardiac surgery, but it may be considered when there is moderate to severe systemic ventricular dysfunction or a high risk of arrhythmias. Central venous access is typically not necessary unless significant fluid shifts or vasoactive drug administration is anticipated.

Intraoperatively, the patient's ventilation must be controlled to maintain normocarbia and normoxia, avoiding hypoxia, hypercapnia, or acidosis, which could increase pulmonary vascular resistance and further strain the right sided systemic circulation. Hemodynamic goals include maintaining adequate preload, avoiding tachycardia, and preserving contractility. Temperature monitoring and warming measures are crucial, as hypothermia can precipitate bradycardia and systemic vasoconstriction. (Table 1)

Table 1:

Anaesthetic Concerns	Goals
• Associated anomaly	• Avoid increase in SVR, increase in Blood pressure, inadequate depth of anesthesi • Avoid Tachycardia, Bradycardia, electrolyte abnormalities, hypothermia, hypercarbia
• Chance of systemic ventricular failure • Conduction abnormalities, Arrhythmia • Prone position	• Avoid increase in pulmonary vascular resistance: i. Acidosis ii. Hypoxia iii. Hypercarbia iv. Hypovolemia

Postoperative care includes continued cardiac monitoring in a high-dependency or intensive care setting due to the ongoing risk of arrhythmias and hemodynamic compromise, especially during the emergence period when sympathetic stimulation and residual anesthetic effects may interact unpredictably with an already compromised cardiac system.[6]

CONCLUSION

Every child with cc TGA has presence of different combinations of cardiac defects. Anesthesia for children with ccTGA undergoing non-cardiac surgery like PSARP requires a comprehensive and multidisciplinary approach. Understanding the pathophysiological implications of the systemic RV, proactive arrhythmia management, and tailoring anesthetic techniques to preserve myocardial function and hemodynamic stability are vital to ensure safe perioperative outcomes.

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