

## **CASE REPORT**

# **Perioperative management of patients with tricuspid atresia and univentricular congenital cardiac defect**

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## **ABSTRACT**

Complex congenital heart diseases like univentricular cardiac defects are relatively common. Tricuspid atresia (TA) with hypoplastic right ventricle is one such univentricular congenital defect which if not corrected surgically leads to fatality. Palliative bidirectional Glenn procedure is usually done as initial measure for such patients. We present this case report based discussion to enumerate the challenges faced by the anesthesiologists while managing such cases for bidirectional Glenn shunt surgery (BDGS) during perioperative period.

**Keywords:** Tricuspid atresia; Hypoplastic right heart syndrome; Bidirectional Glenn procedure; Univentricular congenital cardiac defect; Perioperative Management

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## **INTRODUCTION**

Univentricular hearts are a heterogeneous group of cardiac malformations characterized by a single functional ventricle of either left or right ventricular morphology and usually bear a fatal course in the neonatal period or in early infancy. Tricuspid atresia, the second most common subtype of univentricular heart has an incidence of one in every 10,000 live births and has been found to be present in 2.9% and 1.4% of congenital heart disease autopsy and clinical series, respectively.<sup>1</sup> Fontan and Baudet<sup>2</sup> in 1971 introduced the principle of bypassing the right heart by diverting the systemic venous blood directly to the pulmonary arteries with the functional single ventricle supporting the systemic circulation. Since then significant modification has occurred into it and it became a standard of treatment for almost all univentricular congenital heart defects. Palliation for such defects usually involves different modifications of Fontan operation. This can be done in one-stage procedure or two-stage procedure with initial bidirectional Glenn shunt (BDGS),<sup>3</sup> where the upper body

venous returns are directly diverted to the lung without any ventricular support.

Perioperative management of such patient is challenging and needs good knowledge of patient's cardiac status in terms of both physiology and pathology.

## **CASE REPORT 1**

A 7 year old boy was brought to cardiothoracic and vascular surgery (CTVS) outpatient department with complains of extreme tiredness, and palpitations which severely restricted his physical activity. There was a history of bluish discoloration of the body since birth, exaggerated by feeding, crying or any kind of exertion and relieved by squatting. Parents also complained that the boy was not gaining weight as compared to other children of same age group. There was no history of loss of consciousness, seizure and wheeze.

The boy was poorly built weighing 15 kg, pallor was absent while clubbing was present. Both central and peripheral

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cyanosis was present. Heart rate was 89 /min, blood pressure 90 / 70 mm Hg , respiratory rate 23/min and SpO<sub>2</sub> 70% on room air.

Apex beat was found at left 5th intercostal space around the midcalvicular line. Parasternal heave and pansystolic murmur were present.

Clinical diagnosis of cyanotic congenital heart disease was made and echocardiography was advised for confirmation of the diagnosis.

### CASE REPORT 2

The mother of a three years old boy brought him to us with history of failure to gain weight, easy fatigability even while eating or sucking breast milk, and bluish discoloration of the peripheries since infancy which was aggravated by exertion or crying etc. There was a history of brief episodes of loss of consciousness on many occasions especially on exertion.

Baby was poorly built and weighed only 8 kg. Clubbing and both central and peripheral cyanosis were present. Blood pressure was towards higher side (94/68 mmHg) while heart rate and respiratory rates were 92/min and 25/min respectively. SpO<sub>2</sub> on room air was 74%.

On palpation there was parasternal heave found to be present with apex beat diffuse and maximum thrust at left 5th intercostal space in the midcalvicular line. A pansystolic murmur was also present on auscultation.

A diagnosis of cyanotic congenital heart disease was made clinically and echocardiography was advised for confirmation of the diagnosis.

Echocardiographic findings in both the cases were similar. Both patients had tricuspid atresia with ventricular septal defect with pulmonary stenosis (TA+VSD+PS). They had rudimentary hypoplastic right ventricles. Cardiac catheterization was done for further details including pulmonary vascular resistance. Case I had pulmonary gradient of around 42 mmHg and case II had pulmonary gradient of 40 mmHg.

PO<sub>2</sub> was 50.5 mmHg on room air, PCO<sub>2</sub> was 35.5 mmHg and hemoglobin was 22.7 gm%.

Other blood investigations including total count, differential leukocyte count, coagulation profile, electrolytes, blood urea, and serum creatinine were within normal range.

Patients were examined and assessed for fitness for anesthesia, labeled as ASA physical status IV and advised to be nil per orum status. Both the patients were premedicated with 2 puffs of midazolam nasal spray and IV line was secured. Injection fentanyl 1.5 µg/kg was given IV under observation and patients were taken to operating room.

Standard monitoring was applied. After pre-oxygenation

with 100% oxygen, anesthesia was induced with ketamine 50 mg, vecuronium bromide 2.5 mg and intubated with 5.5 mm size endotracheal tube. ST analysis, central venous pressure, invasive blood pressure, core temperature and anesthetic gas concentration were also monitored.

Anesthesia was maintained with isoflurane in 100% oxygen under controlled ventilation with a closed circuit; vecuronium and fentanyl were given as required.

Hemodynamic stability was maintained with vasoactive drugs. Surgery was performed through median sternotomy route and under cardiopulmonary bypass under heparin anticoagulation, maintaining ACT above 400 sec in both cases. Bidirectional Glenn shunt (BDGS) was performed. The patients remained on heart lung bypass machine for 45 min and 48 min respectively. Post-bypass period was uneventful. Intraoperative metabolic acidosis was corrected with soda bicarbonate.

Patients were extubated 3-4 hours after surgery. Post-extubation hemodynamic parameters were within normal limits. An oxygen saturation of 88% and 90% were achieved respectively with 2 lit/min oxygen by face mask. Postoperative analgesia was maintained with NSAIDs and opioids. After 48 hrs observation in CTVS-ICU, the patients were shifted to the ward without any complication.

### DISCUSSION

A single ventricle or univentricular heart is characterized by a lack of two well-developed ventricles.<sup>4</sup> Tricuspid atresia is the second most common subtype of univentricular heart and is characterized by the absence of the right atrioventricular valve and hypoplasia of the right ventricle. The left ventricle pumps blood directly to the aorta and indirectly to the pulmonary artery via a ventricular septal defect (VSD) to a rudimentary right ventricle serving as a conduit and thereby doing nearly twice the expected amount of work. Blood flow depends upon the ratio of pulmonary vascular resistance (PVR) to systemic vascular resistance (SVR). Thus these patients present to the anesthesiologist in a delicate balance of ventricular performance and PVR to SVR ratio.

The definitive palliation for single ventricular morphologic heart is Fontan procedure or its modification, in which systemic venous return is directed to the pulmonary arteries without an intervening pumping chamber.<sup>2,3</sup> Low pulmonary vascular resistance is essential for blood flow into the pulmonary arteries passively at an acceptable venous pressure. Preoperative assessment should evaluate pulmonary pressure; moderate to severe pulmonary hypertension is regarded as a contraindication to BDGS or such procedures.<sup>5</sup> Cardiac catheterization can provide a detailed assessment of anatomic and functional features of univentricular heart.<sup>6</sup>

The goal of anesthesiologist is to maintain the homeostasis of the PVR to SVR ratio and ventricular performance.<sup>7</sup> Ventricular performance is dependent on sinus rhythm, heart rate, preload, afterload, and contractility. Increased PVR associated with hypoxemia, hypercarbia, atelectasis, acidosis should be avoided to maintain good pulmonary flow. On the other hand increased pulmonary blood flow will increase oxygenation but will also decrease systemic blood pressure leaving anesthesiologist with another challenge. Anxiety and pain also can change PVR to SVR ratio. Invasive hemodynamics, vasopressor and inotropic agents thus become an integral part of management of such patient.

Therefore, anesthesiologist should lead a well-planned anesthetic management, starting from judicious use of premedication to smooth induction, to maintenance, reversal and up to postoperative period including adequate pain relief to facilitate maintaining all these factors.

Patients should be extubated early as positive pressure ventilation increases intrathoracic pressure leading to decreased venous return which in turn decreases the pulmonary blood flow via shunt.

BDGS can also be performed off-pump,<sup>8,9</sup> which is associated with significant elevation of the proximal superior vena cava (SVC) pressure that may lead to neurological injury and anesthesiologist should be vigilant on this.

## CONCLUSION

Good perioperative anesthetic management is an indispensable part of management for univentricular heart patients. Ventricular performance and PVR to SVR ratio should be tightly maintained throughout the procedure for successful management of such patients.

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