**Total Cavopulmonary Connection for Functionally Single Ventricle without Cardiopulmonary Bypass Support**

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**ABSTRACT**

A heart with two atriums but one ventricle, an anatomy with a unique physiology, is responsible for many creative surgical and interventional approaches in history. Different surgical techniques have been used to address this strange physiology of parallel circulation. All these attempts met with failure till the concept of Fontan circulation was described. Currently, controversy exists between multistage vs. single stage total cavopulmonary connections. Total cavopulmonary connection is the only definitive procedure performed to provide palliation for patients with complex congenital heart defects which cannot support a biventricular circulation. We report a case with tricuspid atresia with transposition of great arteries and pulmonary stenosis with persistant left-sided superior vena cava and functionally single ventricle. Patient successfully underwent single stage extra-cardiac total cavopulmonary connection. In this case, bilateral Glenn with extra-cardiac inferior vena cava to main pulmonary artery shunt was performed off-pump.

**Key Words:** Congenital heart defects. Univentricular heart. Tricuspid atresia. Total cavopulmonary connections. Fontan procedure.

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**CASE REPORT**

An 13-year boy was referred from the paediatric cardiology department. He had a history of shortness of breath and cyanosis. On examination, he had central cyanosis and grade 2 clubbing. On chest examination, his apex was located in the left fifth intercostal space at midclavicular line with a pan systolic murmur along the left parasteral area. He had hemoglobin of 15 mg/dl with So2 of 75% at room air. He was diagnosed by echocardiogram with transposition of great arteries, tricuspid atresia, pulmonary stenosis, large atrial septal defect (ASD II) of 18mm, large ventricular septal defect (VSD) of 18mm, with normal left ventricular function and mitral valve function. He had adequate sized branch pulmonary arteries (RPA=18 mm, LPA=16 mm), dilated coronary sinus and functionally single ventricle. Cardiac catheterization was done to check suitability for TCPC. Patient had persistent left superior vena cava with intact mitral valve and preserved left ventricular function without left ventricular obstruction. Right and left pulmonary arteries sizes and pressures were 17mm with 17mmHg mean pressure, and 16mm with 14mmHg mean pressure, respectively.

Patient was planned for single stage total cavopulmonary connection.

Median sternotomy was done. Both right and left superior vena cava were dissected free and controlled with silk. Both right pulmonary artery and left pulmonary artery were also dissected free and controlled with silk. Patient was given 300 units/kg heparin to achieve activating clotting time (ACT) of greater than 400 seconds. Right superior vena cava to right atrial shunt was established for passive flow from superior vena cava to right atrium. Right superior vena cava was divided and right atrial end was closed. Right superior vena cava to right pulmonary artery anastomosis was done. Shunt was shifted to left superior vena cava from right superior vena cava. Left superior vena cava was
divided and right atrial end was closed. Left superior vena cava to left pulmonary artery anastomosis was done. After completing bilateral bidirectional Glenn shunts, the passive shunt was shifted to inferior vena cava to drain blood from inferior vena cava to right atrium. However, the passive shunt was not able to drain inferior vena cava, thus requiring a roller pump to establish flow from inferior vena cava to right atrium (Figure 1). An extra-cardiac conduit of 18mm was used to establish inferior vena cava to main pulmonary artery blood flow after dividing the main pulmonary artery from right ventricle (Figure 2). Right atrial end of inferior vena cava was closed. Fenestration was created between conduit and right atrium. After haemostasis, median sternotomy wound was closed and patient shifted to intensive care unit (ICU) on adrenaline and GTN infusion. Patient was extubated in 3 hours and 27 minute. He was maintaining his SO2 at 85% and PO2 57 mmHg at the time of discharge. Patient was discharged at 6th postoperative day.

DISCUSSION

Univentricle physiology has always fascinated pediatric cardiac surgeons. Chemineau was the first who described the univentricular heart in 1699, with an incidence of 54 cases per million live births. Tricuspid atresia occurs less than once for every 10,000 live births and constitutes 1.4% to 2.9% of congenital heart disease at autopsy. Moodie et al. reported the natural history of patients with functionally single ventricle; 70% with left ventricle morphology died before the age of 16 years while 50% patients with right ventricle heart morphology have an average survival of 4 years. These children with univentricular anatomy require a series of interventions from Modified Blalock Taussig shunt or pulmonary artery banding to modified Glenn and finally complete TCPC connection depending on the anatomy and age. In the current era, different modifications of Fontan operation with extra cardiac conduit have been described. One of the major breakthroughs in improving the results of Fonton circulation is the concept of staging the TCPC by a preceding bidirectional Glenn procedure. Currently, there is a strong argument regarding the best strategy, whether single stage vs. two stage Fonton circulation is better. High-risk candidates with low age, significant pulmonary artery hypoplasia, elevated mean pulmonary artery pressure and pulmonary resistance, significant ventricular hypertrophy and impaired ventricular function who had undergone bidirectional Glenn procedure before complete Fonton circulation have shown better outcome. These improved outcomes may be due to ventricular remodelling, secondary to volume reduction associated with bidirectional Glenn procedure prior to TCPC. However, Kostelka et al. in their study have reported a higher mortality (14.2% vs. 7.4%) in the group of two stage TCPC than in the primary TCPC group.

In this case, the suitable anatomy and absence of risk factors, encouraged us to proceed for the single stage Fonton circulation. We performed the whole procedure off-pump except for a short period of roller pump support while performing the anastomosis between inferior vena cava and main pulmonary artery with a conduit. Postoperative course was unremarkable.

REFERENCES


