Placement of a left ventricular assist device in a patient with congenitally corrected transposition of the great arteries

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A 41 year old man with congenitally-corrected (L) transposition of the great arteries (ccTGA) and congenital heart block presented with left heart failure of the morphologic right ventricle for left ventricular assist device (LVAD) placement. His morphologic tricuspid valve was replaced mechanically 20 years prior.

His complex medical history further includes severe pulmonary hypertension, atrial fibrillation, history of CVA with residual weakness, and an episode of ventricular fibrillation with resultant mild anoxic encephalopathy. Other history includes seizure disorder, asthma, SVC thrombosis, and tracheal stenosis with prior tracheostomy and tracheal reconstruction.

An intraaortic balloon pump was inserted on admission. Two days later he underwent placement of a Heartmate II LVAD. His intraoperative course was complicated by right iliac vein injury due to femoral vein cannulation for CPB requiring laparatomy. A surgical VSD was inadvertently created and repaired, but he was successfully separated from CPB. Later, it was noted that his PA catheter had been sewn into the VSD repair. This was removed in the cath lab on POD 1. His sternum was closed on POD 2; however, his hospital course was complicated by ventilatory failure, C. difficile infection, and persistent fevers. He developed shock liver and renal failure. A duodenal perforation was noted on POD 16 and required urgent laparotomy. He continued to decompensate. Ultimately he developed thrombocytopenia and an intracranial hemorrhage, and care was withdrawn on POD 24.

ccTGA accounts for approximately 0.05% of congenital heart disease. These patients have a double discordance of both atrioventricular and ventriculoarterial connections, such that venous return drains to the right atrium, traverses the morphologic mitral valve into the left ventricle which pumps to the pulmonary arterial system. Oxygenated blood returns to the left atrium, passes through the morphologic tricuspid valve, into the right ventricle which then pumps systemically to the aorta. More than 2/3 of ccTGA patients also have associated cardiac anomalies such as VSD, which dictate the natural history of the disease. Many will experience systemic AV valve regurgitation and ultimate myocardial failure due to the systemic load on the right ventricle over time. The most common causes of mortality are sudden death, ventricular failure, and perioperative death. Currently, there exists scant data regarding successful LVAD placement as bridge-to-transplant in ccTGA patients. However, when surgery is to be undertaken, special considerations must include intimate knowledge of their unique physiology and anatomic variants. Nuances the surgeon must be aware of are ventricular orientation and presence of a VSD for proper cannulation. Given the advances in surgical palliation, the expanding longevity of these patients necessitates an increased command of heart failure management. Though our patient experienced a poor outcome, VAD placement in the face of cardiac failure in ccTGA patients is a feasible therapy as bridge-to-transplant.

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