Anesthetic considerations for the anatomical repair of congenitally corrected transposition of the great arteries

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BACKGROUND
Congenitally corrected transposition of the great arteries (ccTGA), also known as L-TGA, is a rare defect representing only 0.5% of all congenital heart disease. The defect is a combination of atrioventricular and ventriculoarterial discordance. L-TGA is commonly repaired using a double switch operation (DSO). Given the complexity of this surgical technique, it may pose several challenges for the anesthesiologist. We present a case of L-TGA and its anesthetic considerations.

Two different approaches are available for L-TGA repair. With conventional repair, L-TGA is repaired while maintaining atrioventricular discordance. Associated lesions are also repaired. The second approach is an anatomic repair, which preserves the morphological LV as the systemic ventricle and the morphologic RV as the pulmonary ventricle. The anatomic repair is more technically challenging with higher morbidity rates but has better long-term outcomes. The coronary arteries in L-TGA are often inverted, making preoperative delineation of the coronary anatomy an important step in preparation for surgical repair. L-TGA causes a predisposition for arrhythmias and postoperative heart block due to inversion of AV bundles and/or presence of a VSD.

The DSO is a complex procedure involving arterial switch with coronary artery transfer, VSD closure if present, and the creation of an inter-atrial baffle by either Senning or Mustard technique. The DSO commonly requires a prolonged CPB time, leading to significant post-CPB coagulopathy. LV dysfunction due to inadequate LV conditioning prior to the DSO may lead to heart failure requiring inotropic support. Patients who are post-DSO are frequently pacer-dependent secondary to the high incidence of AV block. The complex anatomy and surgical repair may lead to difficulty with coronary re-implantation and coronary blood flow, which may lead to LV failure and myocardial ischemia. The anesthesiologist must consider such potential complications when preparing for DSO.

CASE
We present a 22 month old female with L-TGA, ventricular septal defect (VSD), subpulmonary stenosis, bilateral superior vena cava, and coarctation of the aorta who underwent coarctation repair and pulmonary artery banding with subsequent DSO. The repair required a prolonged cardiopulmonary bypass (CPB) time of 194 minutes. The intraoperative course was complicated with significant post-CPB coagulopathy and heart block requiring complete pacer dependence. Despite good left ventricular function on intraoperative transesophageal echocardiogram (TEE), inotropic support with epinephrine and milrinone was required due to persistent hypotension. Four hours postoperatively, the patient progressed to ventricular fibrillation and cardiac arrest, requiring emergent extracorporeal support. After a complicated postoperative course involving cerebrovascular accident, respiratory failure and acute renal failure, the family withdrew support 4 weeks later.

REFERENCES