Anatomic Repair of Congenitally Corrected Transposition of the Great Arteries: Single Center Intermediate Term Experience

Authors: Shagun Sachdeva, Roni M. Jacobsen, Ronald K. Woods, Michael E. Mitchell, Joseph R. Cava, Nancy S. Ghanayem, Peter C. Frommelt, Peter J Bartz, James S. Tweddell.

Author information: Medical College of Wisconsin, Milwaukee, Wisconsin

Background: Congenitally corrected transposition of the great arteries (CCTGA) is a rare cardiac malformation characterized by atrioventricular and ventriculoarterial discordance, resulting from abnormal looping of the embryonic cardiac tube. Surgical options include a physiologic repair or an anatomic repair. A physiologic repair involves correction of the associated cardiac defects, leaving the morphologic right ventricle as the systemic ventricle. An anatomic repair consists of either a double switch operation or an atrial switch operation with Rastelli procedure, redirecting inflow and outflow so that the morphologic left ventricle is in the systemic position. Midterm outcomes for an anatomic repair are encouraging with a 70-80% survival rate at 10 years. We present the 14-year experience for patients who have undergone an anatomic repair for CCTGA at the Medical College of Wisconsin.

Methods: A retrospective chart review patients who underwent anatomic repair of CCTGA from 2001-2015 was performed. Each patient's demographic data, surgical procedure, need for reoperation, functional status, and survival were evaluated.

Results: Of the 15 patients, 74% were male. Median age of anatomic repair was 1.33 yrs (range 4.5- 45.6 months). Twenty percent of the patients had heterotaxy syndrome. Three patients had a bidirectional Glenn (BDG) prior to anatomic repair in the setting of mild right ventricular hypoplasia. At the time of anatomic repair, 60% underwent a Senning/Rastelli procedure, 26% had a double switch operation, and 13% underwent only a Senning with a ventricular septal defect closure.. A ventricular septal defect was present in 93% of patients at time of anatomic repair. One patient underwent BDG at time of anatomic repair. Another patient required BDG in the immediate post operative period due to superior vena cava baffle obstruction. In addition, 1 patient required revision of the pulmonary venous baffle and 1 patient required closure of a residual ventricular septal defect prior to discharge. Twenty six percent developed transient complete heart block (CHB) post operatively. Of these, 2 patients required pacemaker placement prior to discharge. One patient underwent pacemaker placement for CHB 4 years after surgery.

Two patients were lost to follow up with no access to records after their discharge from surgical repair. Of the 13 remaining patients, the median duration of follow up was 4.5 years (0.05 -14 yrs). Three patients required late operations, including repair of left ventricular outflow tract obstruction, conduit replacement, and melody valve placement. No patients have heart failure symptoms. Ninety-two percent have normal biventricular systolic function and 1 patient had mildly diminished LV function at most recent follow up. Mild mitral regurgitation was noted in 46%, moderate MR in 15%, and severe MR in 7% of the patients. Mild tricuspid regurgitation was noted in 38% patients, moderate in 15%, and no patients had severe tricuspid regurgitation. Seven percent of patients had greater than mild aortic insufficiency. There was a 100% survival rate at the time of the last follow up.

Conclusions: Patients with CCTGA who have undergone anatomic repair have excellent functional status and survival, although reoperations are common. Further studies are needed to determine long-term outcome.