Treatment of Heart Failure Secondary to Congenitally Corrected Transposition of the Great Arteries With Heart Transplantation

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Congenitally corrected transposition of the great arteries is an uncommon abnormality, comprising < 1% of all types of congenital heart disease. The patient population is heterogeneous, with the condition not being identified until adulthood in some cases. The majority of patients have one or more related cardiac anomalies that can drastically alter the natural history. Heart transplantation is the preferred treatment for most patients with this condition who have end-stage heart failure. One-year survival rates are higher in patients receiving transplants for cardiomyopathy and coronary artery disease versus congenital heart disease, retransplant, or valvular cardiomyopathy. Congenital heart disease carries a higher 1-year mortality risk factor compared with cardiomyopathy (P < 0.0001). However, long-term survival rates are highest for patients who undergo transplantations for congenital heart disease and cardiomyopathy. For patients surviving the first year post transplant, survival is highest in those undergoing transplantation for congenital heart disease.

Case Presentation
A 54-year-old woman with a past medical history significant for congenitally corrected transposition of the great arteries and chronic atrial fibrillation arrived for evaluation due to repetitive bouts of congestive heart failure. She was a well-developed woman with an otherwise unremarkable lifestyle.

Assessment
Her initial evaluation consisted of a full history and a physical; an echocardiogram; and a right heart catheterization, which showed a severely depressed dilated systemic ventricle, a moderately impaired nonsystemic ventricle, severe aortic regurgitation, moderate nonsystemic atrioventricular valve regurgitation, and pulmonary hypertension. Her hemodynamics improved after Swan-Ganz–tailored therapy; however, she was deemed inotrope dependent. Financial and psychosocial evaluations were also completed and confirmed that the patient had adequate support. After her case was presented to the Heart Transplant Selection Committee, she was placed on the transplant list.

Therapeutic Management
Shortly after the patient was placed on the list, an appropriate donor organ became available, and she was then taken to the operating room to undergo transplantation. Intraoperative images show her congenital heart defect (Figure 1) and her new heart following the transplantation (Figure 2). Her post-
operative course was uncomplicated, and she was discharged home on posttransplantation day 11. At last follow-up, the patient was doing well.

References

The Baylor University Medical Center Heart Transplant Program provides comprehensive medical and surgical evaluation for patients with heart failure secondary to adult heart disease. Patients arriving at our center with advanced heart failure have a number of alternatives to treat their condition.

Transplant Surgeons:
Gonzalo Gonzalez-Stawinski, MD – Chief of Heart Transplantation and Mechanical Circulatory Support and Chief of Cardiac Services for Baylor Dallas
Themistokles Chamogeorgakis, MD – Associate Director of Heart Transplantation & Mechanical Circulatory Support
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Brian Lima, MD – Director of Clinical Research, Heart Transplantation and Mechanical Circulatory Support
Juan MacHannaford, MD – Vice Chairman of CV Surgical Services
Aldo Rafael, MD – cardiothoracic surgeon on the medical staff of Baylor University Medical Center

Transplant Cardiologists:
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For people who have advanced heart disease, but do not live near Baylor Annette C. and Harold C. Simmons Transplant Institute, we bring 30 years of transplant experience to them. Teams of physicians, nurses and assistants travel to outreach clinics throughout Texas, delivering world-renowned transplantation medicine to an expanding network of communities. The clinics are an outreach service of Health Texas Provider Network and are located in Abilene, Amarillo, Lubbock, Odessa and Shreveport.