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 many cases not identified until adulthood. The majority of patients have one or more related cardiac anomalies that can drastically alter the natural history. Heart transplantation is preferred 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 undergoing 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.

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. The following case study demonstrates how a patient with a complex congenital defect was transplanted for a favorable outcome.

**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.

Continued
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 postoperative course was uncomplicated, and she was discharged home on posttransplantation day 11. At last follow-up, the patient was doing well.

References