MedicalXpress

Researchers report successful cardiac transplant outcomes in adult patients with congenital heart disease

April 4 2016

Congenital heart disease, which includes various conditions involving defects in the structure of the heart or blood vessels that develop before birth, affects nearly 1 percent of children - about 40,000 babies each year. Though many of these conditions were once fatal in infancy and childhood, surgeries and other treatments have evolved during the past few decades, and some 800,000 adults have now grown into adulthood with these conditions. In time, however, these patients commonly develop heart failure, which is the leading cause of death in those with adult congenital heart disease.

While there is little data to inform best practices for treating patients with adult congenital heart disease (ACHD), a team of researchers from the Perelman School of Medicine at the University of Pennsylvania found that heart transplants can be performed in adult patients with prior corrective surgery for congenital heart disease with excellent outcomes. These findings were presented today at the American College of Cardiology 65th Annual Scientific Session in Chicago.

Most patients being evaluated for a heart transplant are assessed using specific criteria from the United Organ Sharing Network (UNOS), to determine whether they should be listed, and under which classification they are categorized based on their condition. For heart transplantation, these criteria encompass whether the patient is on mechanical support, such as a ventricular assist device (VAD), a pump used to support heart

function and blood flow, if they are on any medications helping to improve heart function, and an evaluation of ejection fraction, the measure of how well the heart is pumping, among other benchmarks.

"A congenital heart looks, acts and presents very differently than a normal heart," said Jonathan Menachem, MD, a fellow in Penn's division of Cardiovascular Medicine. "Due to the complexities of ACHD, the current standards may not assess the needs of this patient population adequately, and therefore ACHD patients in heart failure are listed, and transplanted, much less frequently than non-CHD patients with heart failure stemming from other causes."

Menachem explains that in these cases ACHD patients can be listed "by exception," which allows for additional information to be presented during evaluation, however this is done on an individual basis. Unfortunately, there is not have enough data to identify the right time for transplantation, and historical data indicates that evaluation often occurs too late in this patient population.

"We are constantly in search of ways to improve the length and quality of life for those with ACHD and our research indicates that greater consideration should be given to the potential for these patients to receive heart transplants," Menachem said. "The multi-disciplinary approach we've taken to achieve these results could serve as the model for providing ACHD patients with the best chance of a successful transplant."

Menachem and a team of researchers examined data from 17 consecutive patients with ACHD who underwent heart transplantation between March 2010 and July 2015. Patients were evaluated by a multi-disciplinary team of adult and pediatric subspecialists, including heart failure and transplant, ACHD cardiology, cardiac surgery, and in some cases, hepatology and pulmonary teams.

The transplanted patients ranged in age from 23 to 57, and of the 17 reviewed, five were male, 12 were Caucasian, and three were African American. Seven patients required pre-transplant inotropes - a drug therapy that impacts the heart's muscle contractions - and two required pre-transplant mechanical support. Eight underwent a heart and liver transplant, one underwent a heart and lung transplant, and none of the patients required post-operative mechanical support. Of the eight heart and liver transplants, seven were in patients with single ventricles - as opposed to two ventricles in normal hearts - who previously underwent a Fontan, a pediatric surgical procedure used to reconfigure the heart's circulation to maximize the efficiency of the single ventricle without overworking it. These patients are among the highest risk transplants undertaken. Researchers found that with an average follow-up of 35 months, as of September 2015, there was 100 percent survival at both 30 days and one year.

"While this study looked at only a small sample of patients, and much more research is needed to fully understand the outcomes of heart transplant for ACHD patients, this integrated team-based approach presents a promising start for understanding successful treatment outcomes in patients with this unique pathophysiology and anatomy," Menachem said.

Provided by University of Pennsylvania School of Medicine