

The Growing Number of Adults Surviving with Congenital Heart Disease

Thomas E. MacGillivray, MD; C. Huie Lin, MD, PhD

HOUSTON METHODIST DEBAKEY HEART & VASCULAR CENTER, HOUSTON METHODIST HOSPITAL, HOUSTON, TEXAS

Adults with congenital heart disease (ACHD) are a rapidly growing patient population. Even though congenital heart disease comprises the most common group of birth defects (incidence 1%), historically very few children with complex congenital heart disease survived to adulthood. However, with the tremendous diagnostic and treatment advances made by pediatric cardiologists and cardiac surgeons over the last few decades, the population of adults who have survived congenital heart disease continues to increase. In fact, there are now more adults than children living with congenital heart disease.

Despite the successes of neonatal and childhood treatment of congenital heart disease, there remain previously unforeseen challenges for these long-term survivors and the physicians who care for them. While early surgeries and interventions have altered the natural history of congenital heart disease, these childhood treatments have been palliative or corrective but not curative. As a result, the sequelae of some treatments have created a new natural history. Those patients with ACHD commonly experience arrhythmias, pulmonary hypertension, endocarditis, and heart failure as they live into adulthood. Yet immediate recognition of symptoms and new treatment approaches are providing long-term relief for a majority of patients.

Five years ago, the *Methodist DeBakey Cardiovascular Journal* published its first special issue on adult congenital heart disease, with a focus on transcatheter interventions. This time, we offer more breadth and depth about specific aspects of the disease from some of the country's leading experts.

Drs. Maan Malahfji and Mohammed Chamsi-Pasha kick off this issue with a review on advanced imaging for complex repaired ACHD. Since congenital heart disease is typically diagnosed at a young age, the population of patients with ACHD is growing and is estimated to include roughly 2 million people in the United States alone. Consequently, cardiologists frequently encounter complex patients with multiple prior operations and interventions. Advanced cardiac imaging tests, including echocardiography, cardiovascular magnetic resonance imaging, and computed tomography, collectively provide invaluable information on cardiac anatomy and hemodynamics and are fundamental tools for evaluating and following patients with ACHD. They are also essential in identifying late complications

of complex surgical treatments that are typically performed in the early years of life and for planning interventions. The authors discuss the burgeoning role of commonly used advanced imaging modalities and discuss the pros and cons of each for evaluating patients with ACHD.

Next, cardiothoracic surgeons Carl Backer and Constantine Mavroudis describe their experience performing Fontan conversions at the Ann & Robert H. Lurie Children's Hospital of Chicago. Children with a single functional ventricle who underwent an atriopulmonary Fontan procedure typically develop severe long-term complications, including slow progressive dilatation of the right atrium, thickening of the right atrial wall, and eventual atrial arrhythmias. Studies have shown that atrial arrhythmias in these patients lead to a 6-fold increase in risk for death or need for transplantation. The procedure performed by the authors—which includes a Fontan conversion with arrhythmia surgery and epicardial pacemaker placement—converts the atriopulmonary Fontan to an extracardiac Fontan that improves hemodynamics, controls atrial arrhythmias, and improves both survival and quality of life. The authors provide an overview of Fontan procedures, describe the Fontan surgical technique and patient selection criteria, and present patient outcomes on local, national, and global scales.

The dreaded complication associated with undiagnosed ACHD is sudden cardiac death (SCD), a topic that creates substantial anxiety since most events occur unexpectedly in healthy children or young athletes during or immediately after exercise. Anomalous aortic origin of a coronary artery (AAOCA) is the second leading cause of SCD in young athletes. The clinical manifestations of patients who present with AAOCA are quite variable, ranging from evident myocardial ischemia, such as angina-like chest pain and sudden cardiac arrest (SCA), to complete lack of symptoms. The exact mechanisms leading to SCA or SCD and the specific risks associated with the different varieties of AAOCA are not fully known. Doctors Silvana Molossi, Luis Martinez-Bravo, and Carlos Mery describe the current knowledge of AAOCA, the different modalities used to diagnose and characterize the disease, current management strategies, and an algorithm they use to diagnose and manage these patients.

We then look at one of the most common forms of cyanotic congenital heart disease—tetralogy of Fallot (TOF), which

accounts for 6.7% of all babies born with congenital heart disease. Although the first palliative Blalock-Taussig-Thomas shunt (BTS) was described in 1944, starting around 1960, patients with TOF typically underwent corrective ventricular septal defect closure and relief of right ventricular (RV) outflow tract obstruction (RVOTO) later in childhood after an initial BTS. Since the late 1990s, many children had primary corrective surgery (without BTS) within the first 6 months of life. Yet even after successful surgical repair, RVOT dysfunction is common and often results in pulmonary stenosis or regurgitation, which may lead to RV dilation, dysrhythmias, and ultimately RV failure. In his review of pulmonary valve replacement for TOF, Dr. David Balzer highlights currently available valve technology and its application to patients with dysfunctional RVOTs—not only those with TOF but also those who require an RV-pulmonary artery conduit for any type of congenital heart disease. He also discusses indications for percutaneous pulmonary valve implantation, potential adverse events associated with the procedure or follow-up, and future developments in valve technology.

Dextro-transposition of the great arteries (d-TGA) is another common cyanotic congenital heart defect and one that comprises 5% to 7% of all patients born with ACHD. This once-fatal defect is currently managed with the corrective (previously hoped to be “curative”) arterial switch operation. In fact, neonatal survival for d-TGA is now greater than 90% with aggressive surgical and medical interventions such as the arterial switch operation. However, as more patients successfully treated with the arterial switch grow and enter adulthood, late postoperative complications—including pulmonary stenosis, coronary artery stenosis, and neo-aortic insufficiency—are becoming more evident. Older children and adults who have undergone the procedure in infancy need regular routine follow-up evaluations looking for new symptoms and signs of sequelae. Annual surveillance imaging to assess for declining left ventricular function, increasing pulmonary artery gradients, or dilating neo-aortic roots are necessary to identify and manage these possible complications. In their review, Drs. John Breinholt and Sheba John examine the current guidelines and describe the management strategies of common late complications in patients who have undergone an arterial switch operation.

Doctors Kimberly Holst, Heidi Connolly, and Joseph Dearani follow up with a review of Ebstein’s Anomaly, a rare malformation of the tricuspid valve with myopathy of the RV. Ebstein’s Anomaly presents with variable anatomic and pathophysiologic characteristics that lead to equally variable clinical scenarios, and its natural history demonstrates decreased survival with biventricular failure. Operative interventions include tricuspid

valve repair and surgical arrhythmia ablation, although the RV myopathy inherent to the disease continues to play a role in late morbidity and mortality. The authors discuss the role of echocardiography and magnetic resonance imaging in assessment and operative planning, the evolution of and techniques used in tricuspid valve repair, including the more modern cone approach, strategies to provide RV volume offloading in the setting of severe RV dysfunction, data on late operative outcomes, and the potential of cell-based regenerative therapies.

We close this issue with a review by Dr. Francis Fynn-Thompson on the role of organ transplantation for ACHD. The growing number of children with CHD surviving into adulthood has led to a corresponding increase in advanced heart failure among these now-adult patients. Although an attractive and effective therapy, heart transplantation presents many unique challenges for ACHD patients because they usually tend to have cumulative risk factors that make it difficult to universally apply standard heart failure therapies and transplant paradigms. For this reason, the percentage of patients with ACHD who undergo heart transplantation has remained level over time (about 3%) despite the fact that this population is rapidly increasing. In addition, patients with ACHD who are candidates for heart transplantation are generally labeled as low-urgency status, have longer waiting list times, and are much less likely to have been bridged with mechanical circulatory support. Dr. Fynn-Thompson discusses the scope of transplantation for ACHD, covering indications and contraindications, specific challenges and nuances, and post-transplant outcomes.

We have earnestly attempted to present a balanced overview of topics to enlighten readers about some of the most common issues in ACHD. The burgeoning multidisciplinary subspecialty of ACHD is evolving so rapidly that the Houston Methodist DeBakey Institute for Cardiovascular Education & Training holds an annual ACHD Symposium each fall, providing a unique opportunity for patients, caregivers, and health care professionals to learn more about up-to-date treatment options from leading experts. For a more in-depth look at ACHD, we invite you to join us for the fifth annual Adult Congenital Heart Symposium, which will be held Saturday, October 12, 2019, at the Houston Methodist Research Institute. For further information about CME opportunities and to register, please visit: <https://events.houstonmethodist.org/achsymposium>.

We also invite you to visit the journal’s website at <http://journal.houstonmethodist.org>, where you can enjoy additional content, engage in CME activities, and use the “Dialogue with Authors” link to have an open Q&A with the authors of this issue.