Cardiovascular Update

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Adult Congenital Heart Disease

By Joel T. Hardin, MD, FAAP, FACC, FAHA

Overview

In the United States, approximately 1.6 million adults have grown up with congenital heart disease (CHD), and this number is nationally rising by almost 50,000 individuals each year. In 2010, there were approximately 1.47 adult congenital heart disease (ACHD) patients per 1,000 adults in the U.S. Across all Florida counties in BayCare, this currently equates to around 4,600 ACHD patients. By the year 2025, ACHD prevalence will increase to 1.83/1,000 before plateauing around 2050 at 2.31/1,000.

Survival of infants born with CHD remains dependent upon multiple factors, including severity of the heart defect, age at diagnosis and medical/surgical treatment outcomes. Twenty-five percent of those born with CHD have a life-threatening or critical condition requiring surgery or other intervention during the first year of life. Whereas almost all infants born with a non-critical CHD survive past age 18, currently only 80-85 percent of those born with critical CHD will live to become adults.

Even with major treatment successes realized during infancy and childhood, residual cardiac problems are common among ACHD patients, including heart failure, valvular dysfunction, shunts and arrhythmias. Many surviving patients also have significant functional disabilities, medical co-morbidities, and genetic syndromes that complicate their health, ability to work, quality of life and long-term prognosis. Unfortunately, this population is also highly vulnerable to barriers to effective health care, including uninsured status, myriad medical complexities unfamiliar to their primary care and specialty physicians, and a historical shortage of cardiologists specializing in ACHD.

Previously these patients received subspecialty cardiology care from either pediatric cardiologists or general cardiovascular internists, each calling on limited training in ACHD. Now, a growing number of American Board of Internal Medicine-certified subspecialty ACHD cardiologists work within dedicated ACHD centers throughout North America, including locally at the Tampa Bay Adult Congenital Heart Center's three metro-area offices and the inpatient ACHD facility at St. Joseph's Hospital.

Transition from Pediatric Cardiologist to ACHD Cardiologist

The guiding principles for transition into the ACHD clinic revolve more around intellectual and emotional maturity rather than a specific patient age. Transitional discussions between the pediatric cardiologist and the patient often begin as early as age 12, focusing education sequentially on cardiac anatomy, medical management, quality of life and anticipated future health challenges. This transition is facilitated when the pediatric cardiologist ensures that all important active treatment decisions have been addressed, and a complete medical record is transmitted to the ACHD cardiologist. Parents are invited to participate at various stages along this transition, respecting their insight into their son's or daughter's needs, and enlisting them as partners in long-term care planning.

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Joel T. Hardin, MD, FAAP, FACC, FAHA

Consultant Cardiologist

Adult Congenital Heart Disease and Pediatric Cardiology

Tampa Bay Adult Congenital Heart Center

Tampa, St. Petersburg and Land O' Lakes

BayCare

Complexity Risk Stratification Guidelines

ACHD patients can be stratified into low-, moderate- and high-risk categories. Regardless of a patient's CHD complexity, virtually all CHD-related diagnostic or interventional management should be conducted by cardiologists who are specially trained in CHD imaging, CHD catheter intervention and CHD electrophysiology. Diagnostic studies and interventions performed outside these guidelines unfortunately often need to be repeated, increasing both cost and inconvenience for the patient. Additionally, if a cardiac surgical procedure is assessed as indicated, there is ample evidence showing improved outcomes when these surgical procedures are conducted by board-certified congenital heart surgeons within ACHD centers, rather than general cardiovascular surgeons at general heart hospitals or community hospitals.

Low risk conditions include isolated bicuspid aortic valve, small secundum atrial septal defect (ASD), small ventricular septal defect (VSD), small patent ductus arteriosus (PDA), mild pulmonic valve stenosis, or previously repaired ASD, VSD and PDA. These patients may only need a single ACHD clinic encounter to help them understand their CHD, discuss reproductive recurrence risk of CHD, and to educate their primary care physician and general cardiovascular internist regarding long-term management recommendations. If follow up in the ACHD clinic is needed, it is frequently on an every three- to five-year schedule.

Moderate complexity conditions include tetralogy of Fallot, complete atrioventricular septal defects, ostium primum ASD (partial atrioventricular septal defects), sinus venosus ASD, VSD with associated lesions, coarctation of the aorta, fibromuscular subaortic stenosis (excluding hypertrophic cardiomyopathy), Ebstein's anomaly, moderate-severe congenital pulmonic valve stenosis, moderate-severe pulmonic valve regurgitation (typically following earlier intervention for tetralogy of Fallot), and sinus of Valsalva fistula/aneurysms. Patients with moderate complexity CHD may be collaboratively cared for in both the general cardiovascular medicine clinic and in the ACHD clinic, or they may centralize their care exclusively in the ACHD clinic. These patients are typically seen for follow up every 12-24 months, and if hospitalization is required, they are usually admitted to regional ACHD centers or to a community hospital where an ACHD cardiologist has consulting privileges.

High complexity CHD includes unrepaired cyanotic CHD, double-outlet right ventricle, pulmonary atresia with intact ventricular septum, tetralogy of Fallot with pulmonary atresia, transposition of the great arteries, double-outlet right ventricle, heterotaxy syndromes, all variants of univentricular heart disease (e.g., tricuspid atresia and hypoplastic left heart syndrome), CHD treated with surgical conduits (e.g. right ventricle to pulmonary artery homografts), and patients with severe pulmonary arterial hypertension secondary to congenital heart disease (e.g. Eisenmenger syndrome). These patients are seen in an ACHD clinic every 6 to 12 months, and they should be hospitalized for urgent or acute care in ACHD centers or transferred to an ACHD center once medically stabilized.

Conclusions

The emerging ACHD subspecialty builds on the successes of nearly 80 years of innovations in pediatric cardiology, congenital heart surgery, transcatheter treatment for CHD and CHD cardiac electrophysiology. These patients now survive longer, but with more complex health care needs that require coordination of care in specialized ACHD centers for optimal long-term outcomes.

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