8th Current Trends in Aortic and Cardiothoracic Surgery

Adult Congenital Heart Disease

Past, Present, and Future

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In the United States, there are now more people over the age of 20 with congenital heart disease (CHD) than under that age. The population of adult CHD patients is growing at a rate of 5% per year, with 1 to 1.3 million people in the U.S. having CHD as adults—the most common congenital defect in the nation. Survival to adulthood is better because of improved fetal diagnoses, advances in neonatal intensive care, improved surgical techniques, early complete surgical repair, lower perioperative mortality, and increased midterm and late survival. Over 85% of infants with CHD are now expected to reach adulthood: approximately 300,000 of these patients have complex adult CHD, 350,000 have moderately severe defects, and 350,000 have simple congenital cardiac defects.

The most common defects seen in adult patients are atrial septal defects, aortic stenosis, coarctation of the aorta, pulmonary stenosis, Ebstein anomaly, tetralogy of Fallot, and corrected transposition. Other common defects seen in adults are double-outlet right ventricle, postoperative atrioventricular canal, subaortic stenosis, abnormal mitral valve, primum atrial septal defect, and single ventricle.[1]

Overall, the mortality rate for CHD decreased 40% between 1979 and 1997. The annual number of hospitalizations of adults with CHD more than doubled in the U.S. between 1998 and 2005. During that same period of time, hospitalization for adults with complex CHD increased 60%.[2]

We looked at over 84,000 adult CHD hospital admissions in the U.S. during 2007.[3] Seventeen thousand patients, or 20%, were admitted with heart failure, at a mean age of 69 years. Fifty-four percent were women. Atrial and ventricular septal defects and aortic valve abnormalities were the most common defects noted. The mean length of stay among these patients was 7.6 days. These patients had 3 times the risk of death, compared with adult CHD patients admitted during the same time period without heart failure. Comorbidities were common and included arrhythmias, pulmonary hypertension, and renal failure. Independent risk factors for death were respiratory failure, the use of a ventricular assist device, sepsis, acute myocardial infarction, and acute renal failure. In the same year (2007), we looked at these 84,000 adult congenital heart admissions in terms of who had coronary disease. Twenty-seven percent, or 22,493, had the diagnosis of coronary disease upon admission to the hospital. During that time period, there were 332.7 million adult patients admitted without adult CHD, 6.9 million of whom (21%) had coronary disease. In the adult congenital heart group, the mean age at admission was 66 years, and in the nonadult congenital heart group it was age 70. Adult CHD patients had more percutaneous coronary interventions and coronary artery bypass grafting than did nonadult CHD patients, and at a younger age: 64 years versus 66. The mortality rate in adult CHD patients with coronary disease was lower than the rate in patients without adult CHD (2% vs 3%). Accordingly, hospitalized adult congenital heart patients more often have coronary disease than do nonadult CHD patients, and at a younger age. During hospitalization, adult CHD patients are more likely to undergo invasive procedures (coronary artery bypass grafting and percutaneous coronary intervention) and to do so at a younger age, but are more likely to survive. In the addition to the risk of perioperative death in adults with CHD, there are long-term complications: arrhythmias and sudden death, heart failure, and vascular lesions.

The problem in the United States and Canada is that we have nowhere near the number of care facilities or practitioners that is needed to treat the expanding population of adult congenital heart patients. There are 70 self-proclaimed adult congenital

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heart clinics in the U.S. that see a total of about 50,000 patients—only 5% of the 1 to 1.3 million patients who need care. In Canada, which has fewer centers and a longer history of integrated national practice in adult CHD, only 23% of the patients are seen in facilities with expertise in adult CHD. Most patients continue to be seen by internists and occasionally by adult cardiologists, or they have no medical care at all. In the American Deep South (Alabama, Arkansas, Florida, Georgia, Louisiana, Mississippi, South Carolina, Tennessee, and Texas), there are 415 pediatric cardiologists, 59 congenital heart surgeons, and 28 hospitals that perform complex congenital cardiac surgery. Alabama, Arkansas, Georgia, Louisiana, South Carolina, and Tennessee have 1 adult congenital heart center each. There are 4 in Florida and 4 in Texas, for a total of 14. In 2007, these centers saw 6,628 patients and performed 442 surgeries. If one removes Texas and Florida, there were 3,436 patients and 281 surgeries. At Texas Children's Hospital, we had 874 annual adult congenital heart visits in 2007. That number increased to 1,253 in 2008 and 1,542 in 2009. We have averaged approximately 4 adult congenital heart surgical cases per month in 2010, with as many as 11 patients in June of that year.

In addition, the United States and Canada are not training enough physicians to deal with caring for patients with adult CHD. Only 9 of 170 adult cardiology fellowship training programs offer advanced training in CHD for the adult. Fewer than 20 centers in the U.S. provide training in adult CHD. Approximately 60% of these centers see fewer than 10 patients per week in adult CHD clinics and admit fewer than 5 patients per week to their programs.

In addition, we have a major problem with patients' transitions from pediatric to adult care and with the setting in which to do this.[4] In a total population of 360 adult congenital heart patients over the age of 19 in the Canadian study,[4] 47% transferred successfully to adult care. Successful transfer was significantly associated with more pediatric cardiac surgeries, older age (among children) at last visit to the hospital, and documented recommendations in the medical chart for follow-up at an adult congenital heart center.

In a recent summary involving 49 pediatric cardiology training programs, of which 22 (or 45%) respond ed, the following data were noted. Each of these institutions had 11 to 20 faculty members and performed more than 450 CHD surgeries a year. Only 29% had a formal mechanism for transition. Of the 36% that did have such a mechanism, 73% effected the transition when the patient reached 19 years of age. Seventy-three percent of noncardiac consultations were done by specialists in adults. One hundred percent of the catheterizations were performed by pediatric cardiologists, and 77% of the electrophysiologic procedures were performed by pediatric electrophysiologists. Eighty-one percent of cardiac surgeries were performed at a pediatric hospital. Noncardiac surgery was performed at an adult institution in 50% of the cases. Cardiac surgery was performed by a pediatric congenital cardiac surgeon in 95% of the programs.

The medical management of adult patients with CHD will continue to be a challenge, because the numbers of patients increase on a yearly basis while the expertise and personnel remain limited and focused upon a very few centers nationally.

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