Surgery for adults with congenital heart disease should be performed by congenital heart surgeons

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Patients with congenital heart disease require lifelong medical care. The frequency of congenital heart disease in the population is approximately 1%. The population of the United States is approximately 295 million. Consequently, the number of citizens in the United States with congenital heart disease is estimated to be 3 million.

The introduction of cardiopulmonary bypass in the 1950s revolutionized the surgical treatment of intracardiac congenital heart malformations such as ventricular septal defect, tetralogy of Fallot, and atrioventricular septal defect. Treatment of univentricular circulations with the Fontan procedure and its modifications was introduced in the 1970s, and by the 1980s, success was achieved in complex operations performed in the neonatal period—the arterial switch procedure and the first-stage Norwood palliation for hypoplastic left heart syndrome complexes. Because 85% to 90% of infants with congenital heart disease survive now into the adult years, there is a new specialty in the care of adults with congenital heart disease. Interestingly, at the present time in North America, there are more adults than children with congenital heart disease.

The increasing population of adults with congenital heart disease is occurring with limited medical and surgical expertise to care for this unique patient population. The adult with congenital heart disease can be a patient previously operated on during infancy or childhood or a patient with congenital heart disease that has gone unrecognized until later in life.

Although many controversies and unsolved issues surround the care of adults with congenital heart disease (eg, location of care in a children's hospital vs an adult hospital), the focus of this article will be on the issue of which cardiac surgeon, adult or congenital, is best suited to deal with these operations.

WHAT IS A CONGENITAL CARDIAC SURGEON?

The congenital heart surgeon is defined as one who has completed a congenital cardiac surgery fellowship. Most practicing congenital surgeons in North America have trained in postgraduate congenital cardiac surgery fellowship programs in the setting of a children's hospital in the United States, Canada, Europe, and Australia. The length

of time spent in these fellowship positions has ranged from 6 months to 2 years, depending upon the trainee and the program. In general, congenital subspecialty training has followed completion of an Accreditation Council for Graduate Medical Education (ACGME)-approved Thoracic Surgery Residency training program with subsequent American Board of Thoracic Surgery (ABTS) certification.

The practice of the staff congenital surgeon is >75% congenital or pediatric cardiovascular cases and includes performing >75 major operations annually.¹ For the majority of congenital surgeons, this practice setting occurs most commonly in a children's hospital.

The vast majority of published literature on all subjects of congenital heart disease, in both children and adults, has been authored by congenital heart surgeons or congenital heart groups (congenital cardiologists, surgeons, intensivists, etc). In addition, the vast majority of national and international presentations on all topics of surgery for congenital heart disease have similarly been done by congenital heart surgeons.

SPECTRUM OF CASES

The range of congenital heart disease in adults includes patients previously operated on during infancy or childhood and patients with congenital heart disease that has gone unrecognized until later on in life. At Mayo Clinic, the primary operation (ie, the first operation performed on a patient $\geq$ 18 years of age) may occur often and for a very wide spectrum of congenital malformations.² Diagnostic examples include atrial and ventricular septal defects, partial anomalous pulmonary venous connection, partial atrioventricular septal defect, Ebstein's anomaly, congenitally corrected transposition, aortic coarctation, anomalous coronary artery anomalies, and many others.

Monumental advances in the surgical management and perioperative care of neonates and infants with complex congenital heart disease have resulted in the vast majority of children affected by these malformations living well into adult years. Although many of these operations were initially viewed as "curative or corrective" (eg, repair of tetralogy of Fallot or atrioventricular septal defect), many patients will require reoperation for progressive valvular dysfunction, arrhythmias, or other causes.² There are other "corrective" operations (eg, truncus repair, Rastelli procedure) that also require reoperation because of conduit/valvar degeneration or the eventual development of patient-prosthetic mismatch because of somatic growth (eg, valve replacement for Shone's complex, Ebstein's anomaly).

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Finally, the lifelong need for additional procedures following palliative operations, such as after the Fontan procedure, is well documented. Numerous reoperations for some anomalies may be required over the course of a lifetime, and the number of previous operations performed when a given reoperation is being considered as an adult is increasing.

Finally, the need for simultaneous treatment of acquired abnormalities (eg, coronary artery bypass grafting for atherosclerotic coronary artery disease or the Maze procedure for ablation of atrial tachyarrhythmias) are also common.

OUTCOME DATA

A national practice pattern for management of adults with congenital heart disease was presented by Karamlou and colleagues at the Southern Thoracic Surgical Association meeting in November 2007.³ A nationwide inpatient sample (ie, a cross-sectional database that contains 20% of hospital discharges in the United States) identified ~30,000 congenital cases, yielding a national estimate of ~150,000 cases. Of these, ~40,000 were adult congenital cases. Importantly, although congenital surgeons performed >80% of cases done among pediatric patients, they performed only 5% of the adult cases of congenital heart disease. In other words, >95% of adults with congenital heart disease had operations by noncongenital heart surgeons (the adult cardiac surgeon). Most procedures included the simple lesions of atrioventricular septal defect and ventricular septal defect, but more complex lesions, which often represented reoperations in the diagnostic categories of tetralogy of Fallot, double outlet right ventricle, transposition of the great arteries, and single ventricle diagnoses, were also included. Early mortality was significantly greater if the operation was performed by a noncongenital heart surgeon (5% vs 2.5%; $P < .0001$). Independent risk factors for early mortality included the operation being performed by a noncongenital heart surgeon (odds ratio [OR], 3.3), and the risk of early mortality was reduced if the surgeon performed a higher percentage of congenital cases (ie, the congenital heart surgeon; OR, 0.9).

Finally, analysis of resource utilization in this review demonstrated a decreased hospital length of stay and lower total hospital charges if operations were performed by surgeons who performed a greater percentage of pediatric cases (ie, the congenital heart surgeon).

Other data addressing outcome following operation in patients with congenital heart disease have been published elsewhere.⁴⁻⁹

GUIDELINES FOR MANAGEMENT OF ADULTS WITH CONGENITAL HEART DISEASE

The 32nd Bethesda Conference addressed the “care of the adult with congenital heart disease.” Task Force 4 focused on the “Organization of Delivery Systems for Adults with Congenital Heart Disease” and indicated that designated

regional centers for adults with congenital heart disease require full-time, expert congenital heart surgeons who perform a critical volume of pediatric and adult cases of congenital heart disease.¹⁰ Similarly, the consensus document of the Canadian Cardiovascular Society guidelines for the management of adults with congenital heart disease also outlined that at least two full-time surgeons with training and experience in congenital heart disease be responsible for the care of adult patients with congenital heart disease.¹¹⁻¹³ Finally, the American College of Cardiology/American Heart Association Guidelines for the Management of Adults with Congenital Heart Disease indicate for each diagnosis that operations should be performed by surgeons with training and expertise in congenital heart disease.¹⁴

TRAINING AND EDUCATION

Defining the competent congenital surgeon has become increasingly difficult at the present time. The number of skills and knowledge necessary to be successful have increased. The trainee must be prepared to perform open as well as endovascular procedures, understand complex congenital anatomy and imaging techniques, interpret complex hemodynamic data provided by cardiac catheterization, be knowledgeable of critical care medicine, and be comfortable with the medical and surgical management of transplantation issues.

The number of congenital cases required by the ABTS has decreased over the last decade. The 2008 ABTS requirements indicate a minimum of 10 cases as surgeon *or* first assistant for the general thoracic pathway, and 20 cases for the cardiac pathway (10 surgeon, 10 first assistant). Details can be reviewed on the ABTS website at: <http://www.abts.org/>. The numbers of cases submitted to the ABTS over the last 5 years (2003 to 2007) demonstrated the mean number of congenital cases performed as surgeon to be 24 and first assistant to be 39. Because there has been a shift to early repair, in addition to the impact of interventional cardiology, there are fewer straightforward congenital cases for the resident in thoracic surgery to obtain experience in.

In the fall of 2005, the ABTS established the need for formal subspecialty training in congenital heart surgery because the “unique skills and education necessary for the care of these patients” was not being provided in the standard thoracic surgery residency.¹ The requirement for matriculation into a congenital fellowship is successful completion and ABTS certification of Thoracic Surgery in an ACGME-approved program. The congenital fellowship is 1 year in length in a U.S. program and must include a minimum of 75 major cases. The minimum distribution of cases must also include ventricular septal defect (5), atrioventricular septal defect (4), tetralogy of Fallot (4), aortic arch reconstruction/coarctation (4), arterial switch (4), and Glenn/Fontan (5). ABTS certification for practicing congenital

heart surgery is obtained after passing a written and oral board examination.¹ This specialty certificate in congenital heart surgery was officially approved by the ACGME. The ABTS has also proposed a “grandfather” provision to allow those currently engaged in the practice of congenital heart surgery to obtain a subspecialty certificate.

The importance of establishing standards in the care of patients with congenital heart disease is to improve overall quality of care for these patients in the United States—both children and adults. The certificate in congenital heart surgery will recognize and document a standard of education, operative experience, and cognitive knowledge for individuals practicing in this field.

THE IDEAL MODEL

The ideal model for the care of adults with congenital heart disease does not exist at the present time. Previously published guidelines referenced above address many controversial and unsolved issues that confront adult patients with congenital heart disease and their families, as well as all of the medical personnel involved with their care.

As a practitioner who has a focus in the care of adult congenital patients and has reflected on prior guidelines and participated with the development of the most recent ACC/AHA guidelines, it is my belief that the most important requirement for successful care and optimum outcome is with appropriately trained personnel.² In the majority of circumstances and in most settings, the proper personnel are those trained in congenital heart disease. The nuances and complexities of congenital cardiac anatomy coupled with the high probability of previous operation during childhood make the trained congenital cardiovascular surgeon best suited to deal with this growing patient population.

The core multidisciplinary team of specialists caring for adults with congenital heart disease should be professionals with additional training and expertise in congenital heart disease in the specialties of cardiology, cardiac surgery, cardiac anesthesia, cardiac perfusion, cardiac imaging, and critical care. In addition, there should be access to other specialists who have knowledge of congenital heart disease in the areas of hepatology, nephrology, hematology, pulmonary hypertension, maternal/fetal medicine, heart failure, and transplantation. Finally, there should be allied medical professionals—nurse specialists and physician assistants, respiratory therapists and pharmacists—who are also knowledgeable of the various aspects and issues of congenital heart disease in the adult.

At the present time, the best location of the procedure—children’s hospital versus adult hospital—should be individ-

ualized for each surgeon and practice depending upon the comfort level and availability of the trained personnel who will care for any given patient.

During the next decade, we need to remedy some of the chronic structural problems that adults with congenital heart disease have increasingly encountered in the United States. There is evidence of a growing cadre of health care professionals dedicated to improving the care of these patients. More information has become available about their care, which will be improved upon in the next decade. The care of these patients should become more secure in the next decade with the support of the pediatric and pediatric cardiology communities, the Adult Congenital Heart Association, and the persistence and growth of trained adult congenital cardiac health care providers nationwide.

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