Adults with congenital heart disease for non-cardiac surgery: approach to their care.

Nina A. Guzzetta, M.D.
Emory University School of Medicine
Children’s Healthcare of Atlanta

Introduction:

Congenital heart disease (CHD) occurs in approximately eight out of 1000 live births. Today as many as 95% of these children will survive into adulthood (1). It is has been estimated that, for the first time in history, there are currently more adults than children living with CHD (2). Also, based on estimates of the growing population of adults with CHD, it has been suggested that there will be approximately 400,000 adults with moderate or complex CHD living in the United States by the year 2010 (2). A significant number of these patients will require additional cardiac and non-cardiac surgical procedures during their lifetime. While there is broad agreement that the medical and surgical care of adult patients with CHD will expand in the coming years, it is unclear exactly where these services will be provided.

Learning Objectives:

1. To recognize the changing population of adults with CHD and their associated morbidity and mortality.
2. To assess the risk of morbidity and mortality of adult patients with CHD undergoing non-cardiac surgery.
3. To review the practice guidelines provided by the American Heart Association in conjunction with the American College of Cardiology concerning the operative management of adult patients with CHD.

The changing population of adults with CHD and their associated morbidity and mortality:

The hemodynamic and cardiac problems of adults living with CHD are shaped by trends in cardiac surgery such as the decision to perform definitive repairs at an ever earlier age and continuously changing operative procedures. For example, adult survivors of transposition of the great arteries are now more likely have undergone an arterial switch procedure than the older atrial baffle procedure. The cardiac manifestations in adult life of patients who have undergone an arterial switch procedure are quite different from those of the older Mustard or Senning procedures. The long-term concerns after an arterial switch include stenosis of the great vessels at their anastomotic sites, coronary insufficiency and myocardial ischemia as opposed to the systemic ventricular dysfunction and dysrhythmias of the Mustard or Senning procedures. Patients with complex single-ventricular physiology and various modifications of the Fontan procedure will also continue to increase in number. Conversely, because of improvements in noninvasive diagnosis and earlier definitive repairs, one might anticipate a decrease in the prevalence of pulmonary vascular disease and Eisenmenger’s syndrome.

The profile of adult patients with CHD is changing over time not only because these adults are living longer, but also because of improved survival of patients with complex anomalies. Whereas in the past, most adults with CHD had simple defects, we are now seeing an increasing number of adults with moderate and complex forms of CHD. A recent study from Denmark (3) followed 225 adult patients with CHD over a 15 year period from January 1998 through December 2005. Anatomical diagnoses of CHD were classified into simple, moderate or complex according to criteria developed by the 32nd Bethesda Conference of the American College of Cardiology on adults with CHD (4). Over the course of the 15 year study period, investigators found that the number of adult patients with simple congenital defects steadily declined while patients with moderate and complex defects steadily increased. The largest
diagnostic groups, accounting for approximately 50% of the patients, consisted of three categories: adults with a history of a secundum atrial septal defect (ASD); adults with a history of tetralogy of Fallot (TOF); and adults with a history of an aortic coarctation. The overall mortality of patients during the study period was 1.3% (3 patients out of 225) and all three deaths occurred in patients over 60 years of age.

Another large European study (5) retrospectively followed a cohort of 4,110 adults with CHD from 79 participating centers in 26 countries. The median follow up was 5.1 years. Again, the largest diagnostic categories of adult patients included those with a history of a secundum ASD, a ventricular septal defect (VSD) or TOF. The overall mortality during the study period was 2.8% (115 patients out of 4,110). Mortality was lowest in those patients with simple defects (ASD, VSD and aortic coarctation) and was highest in those with complex disease (cyanotic heart disease and Fontan circulation). Despite disease complexity, a considerable proportion of patients experienced significant morbidity including endocarditis, dysrrhythmias and stroke.

The increasing severity of CHD in adult patients puts a very different face on the education needed to maintain optimal health in this growing patient population. Both of the above large cohort studies confirm that morbidity is high among adult patients with all severities of CHD and emphasize the importance of specialized medical care for those with complex disease.

Risk factors in adult patients with CHD undergoing noncardiac surgery:

A diagnosis of congenital heart disease adds significant risk to both pediatric and adult patients undergoing non-cardiac surgery (6,7). Knowing which patients have a high risk for perioperative complications is critical information when deciding where and how to care for them. Several studies have attempted to identify factors associated with an increased risk of morbidity for adult patients with CHD who undergo non-cardiac surgery. Warner et al. (8) identified three major risk factors associated with a significant increase in complications after non-cardiac surgery: pulmonary hypertension (pulmonary artery systolic pressure $\geq 35$ mm Hg or mean pulmonary artery pressure $\geq 25$ mm Hg); cyanosis (arterial oxygen saturation $\leq 90\%$); and treatment for congestive heart failure (use of both digoxin and a diuretic). Patients with pulmonary hypertension had a markedly higher complication rate (15% for patients with pulmonary hypertension vs. 4.7% for patients without pulmonary hypertension). Additionally, procedures performed on the respiratory and nervous systems were associated with the highest frequency of complications. However, the authors did support the idea that certain outpatient surgical procedures could be performed safely in select patients without the above risk factors. Thirty-six percent of the non-cardiac procedures were performed on an outpatient basis, and only three patients (1.7%) had a complication necessitating hospital admission. This admission rate was similar to their unanticipated admission rate for all outpatient surgical procedures.

Another interesting risk factor associated with adverse outcome in adults with CHD is a lapse of medical care (9). American Heart Association guidelines dictate that adults with moderate or complex cardiac lesions should be evaluated every two years in an Adult Congenital Heart Disease center. In a recent study of 148 adults with CHD, a lapse in medical care greater than two years was present in 63% of the patients, and the median duration of the lapse was 10 years. The most common reason cited for the lapse of care was that the patient was told that there was no need for further follow-up (32%). Nonetheless, patients with a lapse in medical care were 3.1 times ($p = 0.003$) more likely to require urgent cardiac intervention.

Finally, it should be noted that dysrrhythmias are extremely common in adult patients with congenital heart disease. In a recent study evaluating various types of surgical procedures performed on adults with CHD, the authors found that implantation or revision of a pacemaker or defibrillator was one of the most common procedures being performed in this patient population (10). Another study cited dysrrhythmias as the most frequent cause of hospitalization in their population of adult patients with CHD. In this study,
supraventricular arrhythmias occurred in at least one of every five patients and ventricular arrhythmias occurred in one of every 20 (5).

Guidelines for clinical care of adults with congenital heart disease:

The transition from pediatric to adult care requires a carefully coordinated transfer process with input from both the pediatric and adult teams. Structured plans need to be developed early and discussed with patients and their families. Most policies recommend transfer to adult care at age 18 or upon leaving high school (11). Patients and their families should also be educated regarding individual diagnoses, specific health issues and the importance of continuing expert medical care (11).

The American College of Cardiology 32nd Bethesda Conference on adults with CHD recommends that the medical care of adult patients be coordinated by regional adult centers. Approximately 30 to 50 such centers are being developed and supported across the United States. These facilities and their personnel need to be equipped and trained to offer intraoperative and postoperative care to adult patients with CHD and to provide care for any complications that may occur during their lifetime. The 32nd Bethesda Conference also recommends that all pediatric cardiology programs, adult cardiology programs and emergency care facilities have a referral relationship with a regional adult CHD center and that these centers be appropriately equipped. They suggest at least one pediatric cardiologist trained in adult CHD, at least one adult cardiologist trained in adult CHD, two mid-level practitioners, two congenital cardiac surgeons, cardiac anesthesia providers with experience in CHD, echocardiography (including transesophageal echocardiography), diagnostic and interventional cardiac catheterization services, electrophysiology service, transplantation service, cardiac pathology and the technology to perform data collection, quality assessment reviews and interface with local practitioners (12).

Regarding adult patients with CHD undergoing non-cardiac surgery, the 32nd Bethesda Conference recommends the following: “ideally, non-cardiac operations on patients with moderate and complex CHD should be performed at regional adult CHD centers with the consultation of an anesthesiologist with experience in CHD” (11). This applies particularly for patients with adverse risk factors, including poor functional class, pulmonary hypertension, congestive heart failure and cyanosis, or when complex surgery is indicated.

Summary:

Continuing advances in cardiac care have resulted in a new population of adults living with CHD. While the number of adults living with simple CHD has been decreasing, there is now an increasing population of adults living with complex CHD, such as those lesions associated with single-ventricle physiology. Many of these patients experience cardiac sequelae that increase in frequency over time. Anesthesiologists confronted with these patients face numerous challenges. Identification of risk factors associated with adverse outcome is crucial in determining where these patients can receive optimal care. Adult CHD centers are now being developed across the country to identify and follow adults living with complex CHD, provide life-long diagnostic and treatment services, and increase the number of appropriately trained individuals to care for these high risk patients.

References:
