The adult with congenital heart disease: medical and surgical considerations for management

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Current Opinion in Pediatrics 2009, 21:561–564

Purpose of review

The purpose of the present review is to outline some of the challenges of surgical and medical management in the adult with congenital heart disease (CHD).

Recent findings

The number of adult patients with CHD continues to grow. These patients require specialized care and there are few cardiologists and surgeons, as well as other subspecialists (e.g., anesthesia, hepatology, nephrology, etc.) with training who are comfortable in the management of this patient population. When operations on adults with CHD are performed by surgeons trained in congenital cardiac surgery, mortality rates and hospital costs are significantly lower than when performed by adult cardiac surgeons. Reoperations are frequent; peripheral vascular access may be compromised and sternal reentry is more likely to result in cardiac injury. End organ dysfunction, particularly liver and kidney, is not uncommon, further complicating perioperative care. Finally, adults with CHD may have complex psychosocial issues. A comprehensive multidisciplinary team approach can best address all of these issues.

Summary

Adults with CHDs present difficult challenges in the preoperative, intraoperative, and postoperative setting. Regional centers of excellence with congenitally-trained cardiac surgeons, cardiologists, and other medical subspecialists are required to optimize outcome.

Keywords

adult, cardiac surgery, congenital

Curr Opin Pediatr 21:561–564

1040-8703

Introduction

The population of the United States is estimated at over 300 million, and the prevalence of congenital heart disease (CHD) is around 1%. Therefore, there are approximately 3 million American citizens who have CHD. The number of adults with CHD continues to grow for two reasons. First, there is a process of natural selection in which children with previously undetected CHD or children with inoperable CHD survive into adulthood with uncorrected lesions. Second, there is a process of 'unnatural' selection, in which survival of children with CHD has been enhanced because of surgical therapy. Improvements in the perioperative care of neonates and infants in the past 3 decades have led to a major increase in survival of children with CHD. Patients with CHD are rarely cured, whether the initial therapy was corrective or palliative, and thus the prevalence of adult patients living with CHD continues to increase.

Care of the adult with congenital heart disease

The American College of Cardiology created guidelines to address the needs of adults with CHD [1••]. In 2000, the 32nd Bethesda Conference recognized that adults with CHD have specialized requirements for care that were not being met by the healthcare system. These consensus documents called for the establishment of adult CHD centers, with one for every 5–10 million people [2,3]. It was advised that all patients with CHD, even 'simple' lesions, should be seen at least once at a regional center. However, patients with more complex defects should have regular follow-up at a specialized regional center. There should be a team of medical specialists led by congenital cardiologists and congenital heart surgeons. These regionalized centers should provide patient counseling, medical personnel with expertise in adults with CHD, at least two congenital heart surgeons with 24 h coverage, cardiac anesthesia, intensive care, in-patient service, transplantation, congenitally-trained staff in catheterization, noninvasive imaging, and electrophysiology, expertise in high-risk obstetrics and maternal-fetal medicine, and cardiac pathology.

The preferred surgical expertise required for adults with CHD continues to be a subject of debate. A recent analysis of the United States administrative database, the *Nationwide Inpatient Sample*, found that early mortality

was greater for adults with CHD when the operations were performed by adult cardiac surgeons, compared with pediatric (congenitally-trained) heart surgeons (4.8% versus 1.9%, $P < 0.001$) [4••]. Furthermore, this survival advantage increased with increasing pediatric volume. Another study from Atlanta also found that operations performed by an adult heart surgeon had a greater operative mortality than those performed by a congenital heart surgeon (15.2% versus 2.7%, $P < 0.001$) [5•]. Risk factors identified for mortality included older age, surgery performed at a children's hospital, and surgery performed by an adult heart surgeon. Although adult operations may be more easily performed in centers with adult expertise, formal congenital training and an established adult congenital infrastructure are necessary to have the best surgical outcomes in adult patients with CHD.

Patient population

Since 1955, we have performed over 6000 operations on adults with CHD. The number of adult patients with CHD who undergo operation at Mayo Clinic has grown to approximately 300–400 patients per year. Patients present at varied ages with a bell-shaped distribution, peaking during early adulthood (Fig. 1).

Adult patients with previously unrecognized defects who presented at Mayo Clinic for their initial operation included Ebstein anomaly, anomalous coronary arteries, coronary artery fistulas, sinus venosus atrial septal defects, aortic coarctation, subvalvular aortic stenosis, and ventricular septal defects, and others. However, the vast majority of adult patients with CHD have undergone one or more operations during childhood. Over the last decade, there has been a steady increase in the number of patients who have had more than one prior operation, and every year, there are patients who have had over five previous operations (Fig. 2). Multiple repeat sternotomies are not benign; as the number of previous operations increases, the operative mortality increases. Between 1993 and 2007, the operative mortality for all adult CHD patients with one previous operation was 1.8%; however, in patients with four previous operations the operative mortality increased to 8%. Detailed analysis of risk factors for early and late mortality will be forthcoming in a separate publication.

Figure 1 The age of adults with congenital heart disease presenting at time of operation at Mayo Clinic from 1993 to 2008

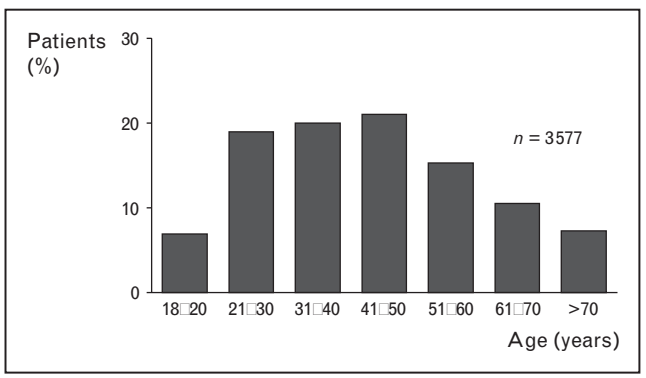
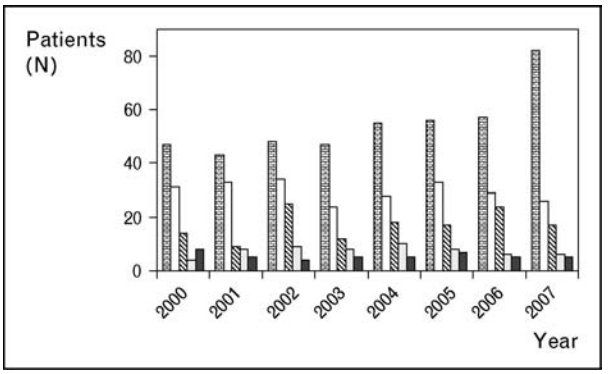


Figure 2 Numbers of previous operations in adults (>18 years) with congenital heart disease undergoing cardiac reoperation at Mayo Clinic



▨, 1; □, 2; ▧, 3; □, 4; ■, ≥5.

A review of a single center's operative experience in the Netherlands of adults with CHD found that 49% were reoperative procedures [6]. The 30-day mortality was 1.5% (14 of 830 patients). Major complications included tamponade in 3.2%, bleeding requiring re-exploration in 7.1%, and renal failure requiring dialysis in 0.4%. Pulmonary hypertension was associated with early postoperative mortality, and ventricular dysfunction and cyanosis were predictors of late mortality.

Surgical considerations

Adults with CHD must be approached cautiously as there are many potential pitfalls during their preoperative, operative, and postoperative care. Both adult acquired surgeons and pediatric congenital surgeons must recognize that this subgroup of patients has special needs that must be addressed.

Vascular access

Peripheral vascular access may be compromised and limited in adult patients with CHD. These patients have typically had multiple prior cardiac procedures including open-heart surgery with perioperative invasive monitoring, cardiac catheterization(s), electrophysiology procedure(s), and/or pacemaker insertion. This may lead to scarring, stenosis, and obstruction of peripheral arterial

and venous structures in the neck and groin. When the repeat sternotomy is deemed to be hazardous, potential access for cardiopulmonary bypass includes the femoral or iliac artery and vein, the axillary artery, the carotid or innominate artery, the internal jugular vein, the right atrium or ascending aorta (via right thoracotomy), the apex of the left ventricle (via small left thoracotomy), and rarely the abdominal aorta. Surgeons must be aware of these potential challenges and know vascular access options when planning the repeat sternotomy in adults with CHD. Preoperative ultrasound imaging of the neck and groin vessels are necessary if peripheral cannulation is anticipated. Preoperative computed tomography (CT) and/or MR imaging of the chest can outline intrathoracic cardiac anatomy and establish relationships of the cardiac chambers and great vessels to the sternum.

The repeat sternotomy and intraoperative strategies

Most adult patients with CHD will require a repeat sternotomy. The type of CHD is important. The position of an extracardiac conduit in patients with tetralogy of Fallot or pulmonary atresia-ventricular septal defect is typically leftward of the midline. The aorta in these two diagnostic groups is frequently enlarged and is in the midline, immediately beneath the sternum. Conversely, in patients who have previously undergone the Rastelli operation for transposition, or truncus repair, the conduit is a midline structure with frequent erosion into the posterior table of the sternum. The autograft in patients who have undergone the Ross procedure is commonly aneurysmal and at risk with sternal reentry. In other situations, for example, Ebstein anomaly, a cardiac chamber (right atrium or right ventricle) may be severely enlarged and thinned and are at risk during reoperation.

The procedure to be performed must also be considered. For mitral or tricuspid repair or replacement, a right thoracotomy is an alternative when there have been multiple previous sternotomies.

Caution must be taken to be aware of the presence of any intracardiac shunts (atrial-septal defect or ventricular-septal defect) in the adult with CHD, particularly if bypass is required before the mediastinal dissection has been performed in order to avoid the catastrophic complication of air embolism. Preoperative transthoracic and intraoperative transesophageal echocardiography is routinely performed. Inadvertent entry into the right atrium or right ventricle, although not common, is a significant issue in some patients. Right ventricular enlargement is common due to the frequent presence of right-sided pathology (e.g., pulmonary and/or tricuspid regurgitation). To avoid complications with sternal reentry, a preoperative decision should be made as to the likelihood of right heart injury. If the risk is deemed high, a decision should be made regarding exposure and cannulation of the femoral vessels. Opening and dissection may be performed on cardiopulmonary bypass; however, a positive central venous pressure of more than 5 mmHg should be maintained in the presence of an intracardiac shunt to avoid inadvertent entry of air into the right heart and systemic air embolism.

Significant ascending aortic dilatation with aortic regurgitation should also be identified preoperatively because left ventricular venting will be required to avoid significant ventricular distension during hypothermic cardiopulmonary bypass.

Pregnancy and parenthood

In these young adults with CHD, consideration of both pregnancy and parenthood is required. Young women must be counseled regarding the risk related to the type of valvular prosthesis and expected reoperation(s) and anticoagulation. Desire for pregnancy may result in requirement for repeated replacement of biological valves that must be considered in patients who will require multiple prosthetic replacements over a lifetime. In addition, adult patients with CHD should be counseled regarding life expectancy. Although late outcomes have improved tremendously in the last few decades, some patients, such as those who have only undergone palliation (systemic-to-pulmonary artery shunt) or those with univentricular physiology (Fontan procedure), will likely have a shortened life span.

Psychosocial issues

There are psychosocial issues to consider when taking care of the adult patient with CHD. Transition of care is frequently an issue as patients outgrow pediatric care and move toward adult cardiologists and potentially adult cardiac surgeons. It is during this transition period that patients are most likely to be lost to follow-up. It has been estimated that up to 80% patients may be lost to follow-up at 5 years [7]. Exercise and athletics are also a concern in young adults that must be considered. Patients may want to pursue competitive sports and must have appropriate counseling and guidance on these issues. Employment and insurability can also be a significant challenge in these patients. It was reported that at the adult congenital center in Tampa Bay, 21% of patients were uninsured and 28% were on Medicare or Medicaid [7]. Another survey of 335 adults who had undergone isolated repair of Tetralogy of Fallot, transposition of the great arteries, or ventricular septal defect found that 76% had a fulltime job but that 18% received a full or partial disability pension [8].

It has also been reported that up to one third of adult patients with CHD have mood or anxiety disorders [9]. In a survey of 155 patients, 40% patients had previous mental health treatment and one third of patients were interested in receiving assistance with stress management and coping with heart disease.

Team approach

Although the team approach is often emphasized in many areas of medicine, the multidisciplinary care of the adult CHD patient is incredibly important. Expertise in high-risk pregnancy and reproductive issues, heart failure and transplantation, pulmonary hypertension, hepatology, nephrology, and hematology are necessary to provide for the varied needs of these patients. Many patients will not have seen a physician for an extended period of time prior to their operation as an adult. In addition, they may have been provided with outdated or inaccurate information by other physicians who do not have experience with patients with adult CHD.

These patients are also susceptible to smoking, diabetes, and hypertension, which are also risk factors for coronary artery disease, as well as potentially exacerbating their existing congenital cardiac disease. In a review of 250 coronary angiograms in adult patients with CHD (mean age 51 ± 15 years), significant coronary artery disease was found in 9.2% [10]. Coronary artery disease was predicted by the presence of hypertension and hyperlipidemia, but had no relation to chest pain. Thus, adult patients with CHD must be considered for primary prevention strategies. In patients with risk factors for coronary artery disease or patients greater than 40 years of age, preoperative diagnostic imaging of the coronary arteries with coronary angiography or CT angiography is advised to rule out concomitant coronary artery disease prior to cardiac operation. The postoperative management in the ICU also requires professionals experienced in the care of patients with CHD.

Conclusion

The numbers of adults with congenital heart disease will continue to grow. The goal must address all of the needs of these complex patients preoperatively, intraoperatively, and postoperatively. Regionalized centers with larger volumes of patients will be required to provide educational opportunities and enable the development of this subspecialty. Surgeons, cardiologists, anesthesiologists, intensivists and others, should pursue specialized training to optimize care for this unique and growing patient population.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- • of special interest
- •• of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (pp. 678–679).

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