The number of adults who have survived with congenital heart disease is increasing rapidly. Although prevalence studies have not been performed, estimates of expected numbers of people reaching adulthood with congenital heart disease (CHD) can now be made because birth prevalence has been studied and mortality rates have begun to stabilize. Birth prevalence for all forms of congenital heart disease detected in the first year of life is estimated at 8.1 per thousand live births on the basis of data from the Centers for Disease Control; estimates for disease of sufficient severity to result in a catheterization, surgery, or death in the first year of life are 2.3 per thousand, using population-based data derived from the Pediatric Cardiac Care Consortium. In 1990, the National Center for Health Statistics reported the number of US live births to be 4,158,212. Age-specific mortality data were obtained from the complete life table for the US population and from population-based death rates during hospital admission using data from 6 states in 1992. Estimates of numbers of individuals reaching adulthood born in 1990 with congenital heart disease of sufficient severity to be diagnosed or treated during the first year of life are shown in the Figure. Disregarding all patients born before 1990 and those not diagnosed in the first year and assuming stable mortality in early adulthood, by 2020, nearly 760,000 individuals will have CHD, with 200,000 in the more severe subgroup.

This striking increase in the estimated number of patients with congenital heart disease who will require competent care within the next 20 years mandates careful consideration of how and by whom this care should be delivered. Pediatric and adult cardiologists who currently treat congenital heart disease in adults (ACHD) have recently begun to grapple with these issues. The Congenital Cardiac Defects Committee of the Section on Cardiovascular Disease in the Young of the American Heart Association has identified defining pathways of excellence for ACHD care as a priority. The purpose of this article is to increase awareness about this important issue and to summarize current concerns. The challenge of providing excellent care to this growing population is important and needs to be placed clearly in the spotlight.

Recurring Themes in Congenital Heart Disease That Will Confront Adult Cardiologists

The recently published 32nd Bethesda Conference on “Care of the Adult With Congenital Heart Disease” and symposia at the World Congress of Pediatric Cardiology and Cardiovascular Surgery attest to the growing international concern that medical expertise may be lacking in the management of patients with repaired and un repaired congenital heart defects who require specialized care when they reach adulthood. At the core of this concern are the issues of which healthcare providers will be most qualified to deliver care, when (if ever) care should be transferred from the pediatric to adult cardiologist, which problems make these patients distinct from those with acquired adult-onset ischemic or valvular heart defects, and how this population of patients will affect financial coverage.

It is important to remember that CHD entail diffuse cardiovascular involvement, and a repaired congenital defect does not imply a “normal” heart. Adaptive postoperative changes associated with patient growth can cause a paradox: Myocardial remodeling may be necessary for survival, but the process itself elicits secondary pathology. Late-onset conduction disorders, arrhythmias, myocardial dysfunction, altered coronary flow, and ischemia can occur in the young adult after many years of an uneventful life after successful CHD repair. Patients at risk include both those with simple defects, such as atrial or ventricular septal defects, and those with more complex ones, including variations of single ventricle, D-transposition of the great arteries, and tetralogy of Fallot. Because the onset of clinical problems can be subtle, early
recognition of adverse changes may enable rapid intervention to slow progressive myocardial deterioration. Physicians must be taught what to look for in these patients and to refer to appropriate centers early.

Any postoperative patient, especially one with artificial valves, carries the risk of embolization, endocarditis, late-onset blood dyscrasias, and/or acquired immune disorders. The possibility of side effects resulting from chronic medications needs continuous monitoring. Re-operation and re-sternotomy raise concerns of direct anatomic issues. Persistently cyanotic patients and those with Eisenmenger syndrome, irrespective of the underlying CHD, carry additional concerns of long-term adverse changes on the joints, bones, kidneys, and brain, as well as alterations in hematological and coagulation parameters. Late-onset aneurysms associated with Marfan’s syndrome or after coarctation repair with aortic root dilations and elastin fragmentation with cystic medical necrosis after aortic valve repair with a pulmonary autograft remain lifelong concerns. Pregnancy-related blood volume alterations in an already altered heart also remain a concern.

Surgery causes incisions, incisions cause scars, and scars cause arrhythmias. Arrhythmias may be viewed as an intrinsic part of the life of any adult patient with CHD, whether they present as sinus or atrioventricular nodal dysfunction, intra-atrial reentry tachycardia, atrial flutter, atrial fibrillation, ventricular tachycardia, or sudden cardiac death. Although seen with any congenital heart defect, arrhythmias may be viewed as an intrinsic part of the life of any adult patient with CHD, whether they present as sinus or atrioventricular nodal dysfunction, intra-atrial reentry tachycardia, atrial flutter, atrial fibrillation, ventricular tachycardia, or sudden cardiac death. Although seen with any congenital heart defect, arrhythmias may be more prevalent among patients with congenitally corrected transposition of the great arteries, ie L-TGA, Ebstein’s anomaly, tetralogy of Fallot, aortic stenosis, or coarctation of the aorta, or after the Mustard/Senning or Fontan procedures. Pacemaker implantation in early childhood, associated with dysynchronous ventricular contraction patterns and chronic bundle-branch block, may intrinsically predispose an individual to myocardial dysfunction by early adulthood.

In children receiving cardiac transplants for nonrepairable CHD, early coronary disease, delayed rejection, and psychological issues with medical compliance beginning in adolescence remain concerns. In addition, lymphoproliferative disease associated with Epstein-Barr virus infection, long-term use of nephrotoxic drugs, and systemic hypertension ultimately may affect the transplant recipient. Early-onset coronary disease also may occur in young adults after coronary artery surgery during infancy, as seen with the arterial switch procedure for D-transposition of the great arteries, or after repair of an anomalous coronary artery.

The adult with CHD carries lifelong emotional, psychological, and financial concerns in addition to cardiac structural, electrical, and mechanical issues. A system of lifelong preventative care by a qualified healthcare team of providers with knowledge both of CHD and age-related systemic health issues will ensure that early warning signs of impending problems are identified and appropriate intervention is initiated. This team, regardless of the patient’s age, should include those with the best working knowledge of structural congenital heart anatomy and physiology. In this regard, a qualified, board-certified pediatric cardiologist, including any of the subspecialties (electrophysiology, catheter intervention, echocardiography), as well as a certified congenital heart surgeon, can provide invaluable service to the management and treatment of a patient of any age with CHD. This combination allows a lifelong working association to optimize care.

Adult Cardiac Issues That Need to Be Addressed in the Adult With CHD

Some portion of the healthcare needs of the adult patient with CHD will inevitably fall to caregivers with little or no specialized training in CHD. Adult patients with CHD have been grouped according to very complex, moderately severe, or more simple conditions. The degree to which the patient’s care is managed by a highly skilled team at a regional center with specialized expertise will vary depending on the severity of the underlying condition and the health issue in question. Although regional centers of excellence will hopefully take a guiding role with the most complex cases, common adult diseases will be experienced by these patients and will be the responsibility of the local physician. It is estimated that 45% of adult CHD patients may not require routine follow-up at a specialized center. The practitioner who functions in the primary care role will be called on to participate in a critically important way in the health maintenance of the patient and the management of coexisting diseases and conditions. These health issues can be primarily related to the underlying CHD or to unrelated comorbidities that can be expected to arise, the treatment of which may or may not be modified by the patient’s cardiac status.

The spectrum of health issues that need to be addressed in the adult patient with CHD expands with age. Early adulthood brings issues of family planning and careful advice regarding contraception, genetic counseling, and the best manner of obtaining care during pregnancy and delivery. Work issues relating to physical limitations may need to be explored. The pediatric caregiver may have addressed many of these issues earlier in the patient’s care. Lifestyle choices also are established in adolescence and early adulthood that will have tremendous impact as the patient ages. Avoidance of recreational drug use, tobacco, inactivity, and obesity in the young adult assists in the optimization of future health. Dietary strategies with regard to heart-healthy diets limited in salt and fat need to be reinforced. Excellent dental hygiene and observance of antibiotic prophylaxis against infectious
endocarditis often need to be reinforced also. Establishing a health maintenance program and encouraging the individual patient to take responsibility for issues such as regular pap smears, mammography, and other routine follow-up are important responsibilities for the primary provider.

Some adult illnesses will not differ significantly between CHD patients and the non-CHD population. Diabetes and obstructive lung disease may not require therapy tailored specifically to the CHD patient, but confounding issues of inactivity and obesity may make management of glucose intolerance and diabetes more difficult. Similarly, chronic lung diseases will complicate preexisting cyanosis and erythrocytosis in patients with highly complex CHD. Renal disease may make volume management more difficult, particularly in patients with underlying congestive heart failure. Hypertension may exacerbate ventricular or valvular dysfunction, and the underlying CHD may determine the choice of therapy. A clear picture of the patient’s hemodynamic reserve is essential to planning an appropriate regimen and target goals for all of these conditions.

Atherosclerosis is the most common disease in American adults, with manifestations throughout the vascular system. Primary risk factor modification starts in young adulthood with diet and exercise strategies. There are no data to suggest that targets for cholesterol levels and triglycerides will be different in this population, and primary cholesterol guidelines should be applied. Advice about maintenance of an active lifestyle and healthy body weight must be clear and consistent. Smoking avoidance or cessation is critical, and other high-risk activities such as illicit drug use should be addressed.

The development of cardiovascular disease in these patients may often compound preexisting abnormalities in ventricular and valvular function, resulting in any amount of vascular disease being magnified in terms of functional impact and prognosis. Early and appropriate recognition of coronary artery disease therefore is even more important to this group of patients. It is uncertain whether the onset or presentation of cardiovascular disease in this population will be different from the general population. Chest pain as a presentation for coronary artery disease is variable among different populations and may be shown to have unique characteristics among adult CHD patients as well. Prior cardiac surgery and symptom awareness may be different in these patients, and a high index of suspicion will need to be maintained with regard to atypical symptoms. In addition, the reliability of commonly used screening and ischemia quantification techniques may need to be defined in this population. Subtle exercise intolerance persists in many adult CHD patients after repair of simple defects and may delay the onset of exertional symptoms. Stress testing may be required to recognize effort intolerance for definition of functional class, as well as for eliciting a diagnostic ischemic response.

Exercise prescription will be a challenging area for the nonexpert clinician. The guidelines most often used in this regard were published in the 26th Bethesda Conference. It remains to be proved whether physical conditioning reduces symptoms and improves exercise tolerance and quality or length of life among adults with CHD. One can only hope that

Choice of therapy for established coronary artery disease may well be impacted by the underlying conditions and surgical history of the patients. Given that prior surgeries may make coronary artery bypass surgery more difficult because of the presence of adhesions and possible alterations in anatomy, these procedures may be performed at a higher than normal risk. The potential for depressed underlying ventricular function, as well as diminished functional class, also increases the risk of surgical and interventional procedures for revascularization.

Among patients with CHD, atrial and ventricular arrhythmias are an increasing problem. They may be the result of underlying anatomic abnormalities, abnormal cardiac chamber dilatation, heart failure, or scars from prior surgeries. Their presence carries an increased risk for morbidity and mortality. Detection and treatment are high priorities. The roles of catheter-based interventions, surgery, and implantable defibrillators are still under study. This specialized issue will often require referral to expert centers.

Poor functional class, pulmonary hypertension, heart failure, and cyanosis increase anesthetic risks. As a result, noncardiac surgery carries increased risk among patients with moderate or complex CHD. In these patients, surgeries optimally would be performed at centers with experienced operative teams.

**Training Issues for Medical Personnel Caring for Adults With Congenital Heart Disease**

The task of training cardiologists to care for adults with congenital heart disease currently presents a difficult challenge. The physiology of a number of repaired or palliated CHD is difficult to understand if there is only minimal training in congenital heart disease. Additionally, medical and cardiac problems unique to adults, as noted above, are challenging for cardiologists whose primary training is in pediatrics.

Comprehensive care for adults with CHD must necessarily come from cardiologists who are trained to address both pediatric and adult cardiac problems, as well as adult medical issues. Historically, because of the small numbers of surviving adults with CHD, ACHD experience has been gleaned primarily from on-the-job training. Now that a significant number of patients with CHD can be expected to reach adulthood, formal training guidelines for these subspecialty cardiologists are becoming imperative. Because of the increasing complexity of cardiac care in both subspecialties, fellowship training time has expanded to a minimum of 3 years post-residency for pediatric and adult cardiologists. The intent to provide the best possible care for adult patients with CHD will require a team approach in the future, both for training specialists in congenital heart disease and for integrated patient care. New training guidelines should be developed within currently existing educational systems or within
new systems designed as part of regional centers for care of ACHD patients. This team approach for training is important not only for learning outpatient management but also for instruction in interventional and surgical procedures. Pediatric cardiologists could be used as consultants to internists and adult cardiologists rather than as primary caregivers for patient management. Pediatric cardiologists, however, would be the primary educators about concepts and management issues related to the anatomy and physiology of repaired and unrepaired CHDs. Efforts to design a comprehensive cardiac training program for caregivers of adults with CHD should be directed toward 3 groups of trainees: those who have completed a residency in pediatrics, those who have completed a residency in medicine, and those who have completed a combined program in medicine and pediatrics. Physicians with initial residency training in both medicine and pediatrics are ideal candidates for competency training in ACHD, though these individuals are rare.

Areas of expertise that are currently adequately addressed in either an adult or pediatric fellowship programs, such as valvular heart disease, arrhythmia diagnosis, treatment of congestive heart failure, and heart transplant management, should be delineated. Subsequently, current deficiencies in CHD training in adult programs, as well as deficiencies in adult cardiac and medical issues in pediatric programs, can be defined. For instance, management of intracardiac shunts and common postoperative congenital heart diseases, such as repaired tetralogy of Fallot, atrioventricular septal defects, conduits, and Fontan palliations, should be emphasized in extensions of adult cardiology training programs, and coronary artery disease and the effects of coexisting medical conditions common in adults, including pregnancy, should be emphasized in pediatric cardiology training programs.

A recent task force effort has been directed toward establishing the specific pathways and length of training that should be required for ACHD subspecialist cardiologists. Clinical Practice Models for Establishment of an ACHD Center

Clinical practice models for the establishment of an ACHD center will vary with individual organizational needs. Ideally, models will maintain strong ties with one or more programs providing care for pediatric patients with congenital cardiac defects and would be constructed in a fashion complementary to, not competitive with, the needs of pediatric programs. Constraints posed by the need for substantial initial investment, the continuing trend toward greater hospital reimbursement and diminished physician reimbursement, and the frequent need for inpatient care will favor hospital over professional group structures as the institutional model for ACHD programs. Proposals satisfying the needs of patients, hospitals, and hospital administrators must define and justify the mission of the program with respect to the local geographic environment and contain elements that describe administrative, financial, clinical, and investigatory structures.

ACHD administrative structures will optimally parallel those of the hospital. A small core group of appropriate clinicians and one or more hospital administrators with policy-making or financial roles should be charged with all facets of ACHD program development, maintenance, and evaluation. Although single hospital models may be capable of incorporating all essential care elements, partnership models (eg, between institutions focusing on care of children with CHD and those providing adult cardiac care) should be considered, especially between hospitals in close proximity, allowing easy transfer of patients.

Because reimbursement for the care of these patients is quite variable (and often low), a successful ACHD unit will incorporate strong financial controls. Financial structures should offer exhaustive data detailing costs and revenues. New cost centers should be established with care to account for inflation and scalability. Revenue streams are estimated by considering sources of patient volume and by analysis of current and future income. Patient volume will depend on the number of patients already in the system, the rate patients are expected to enter the ACHD program from affiliated pediatric cardiovascular programs, the rate of attrition of patients to
other care providers, and the number of patients referred to the program by community physicians. These data often require an exhaustive analysis of patient records from both pediatric and adult components of the hospital system, as well as an analysis of community physician practices that care for the ACHD patients. Estimates of income and even costs often require knowledge of the distribution of diagnoses, the prevalence of major billing codes, and current charges to income ratios for each hospital and professional code.

Clinical structures should account for inpatient and outpatient facilities and personnel requirements separately and should be integrated with the needs of other hospital programs. The complexity of clinical issues to be addressed within ACHD centers points directly toward establishment of clinical care teams as an ideal method to provide shared expertise to patients. Each team must include at least one individual who can be considered a CHD expert, as well as a specialist in adult cardiology. Other personnel elements include surgeons, anesthesiologists, nurses and technicians, social and financial counselors, coordinators, and subspecialty physicians, particularly geneticists and obstetricians/gynecologists. Care should be taken to assign primary responsibility for patient care to those trained in internal medicine, general cardiology, and pediatric cardiology, who satisfy hospital privilege requirements and through whom billing may be generated. Physical facilities must be configured to maintain inpatient and outpatient environments suitable for adolescent and young adult patient care. When sited in an adult hospital, the facility must provide mechanisms for efficient access to the patient by pediatric cardiovascular physicians, surgeons, technicians, and specialized laboratories. With dual institutional models, all team members should have privileges at both hospitals, so that care delivery can be flexibly directed to the most appropriate institution, without regard to billing or other administrative issues.

Because this is a “new” patient population with poorly understood clinical needs and outcomes, clinical practice models should be serially reevaluated in the future to further improve the facilities caring for ACHD patients. Thus, teaching hospitals seem to be an ideal home for such programs. Teaching hospitals are also best capable of training new generations of ACHD experts, the importance of which has been outlined previously. Because funding for these investigative activities is coming under increasing pressure, early attention must be devoted to sustainable funding streams (grants, industrial support, and philanthropy) for this crucial ongoing clinical research.

**Insurability of Young Adult Patients With Congenital Heart Disease**

Health insurance that enables care of adults with CHD is in many cases difficult to obtain or difficult to continue after reaching independence in adulthood. Life insurance also is often not available to these patients. In 2001, the Bethesda conference on the young adult with congenital heart disease published a discussion of the insurance issues facing these young adults, which are discussed in this section.

**Types of Insurance**

**Health Insurance**

Until the age of 21, patients, depending on income levels, will qualify for Medicaid or Crippled Children’s Services (CCS) (some states call this the Bureau for Maternal and Child Health). Many are covered by their parents’ health policies until age 19 unless they are in school. If they continue schooling full-time, various companies’ insurability coverage continues until their 21st or 23rd birthday. If the patient emancipates him or herself (for example, through marriage), he/she usually loses dependent coverage.

Most insured young adults have some form of group coverage, usually purchased by their employer, often through an HMO. Some can pick their physicians; others have care providers chosen for them, and if the cardiac provider chosen by the plan or gatekeeper is unfamiliar with CHD, the result could be extra testing and possibly suboptimal care.

For the young adult with CHD, preexisting conditions sometimes are not covered, or will be covered only after a few years. Often, no coverage is available to the patient, whether an illness is related to the heart or not. If the employer is a large company and subscribes to a health plan that has a guaranteed benefit, there may be no problem. Some employers, however, will not risk a rise in their company’s premiums and the patient is therefore not hired. If they do obtain employment, patients sometimes find themselves in a situation called job lock because they cannot afford to switch companies when loss of insurance is a risk, impeding their future career advancement. The Consolidated Omnibus Budget Reconciliation Act (COBRA) requires employers to continue healthcare coverage to certain employees on termination of work, death, divorce, or cessation of dependency. This extended coverage can last for 18 months. The person to be covered must notify the employer within 60 days.

**Life Insurance**

Although life insurance may be easier to obtain than in the past (at significantly higher premiums), qualifying is still difficult. Qualification requirements vary between companies, with some being reasonable if the patient has essentially no risk. Others, however, will rate someone with an innocent murmur. Data regarding the success of postoperative adults with CHD obtaining life insurance are not yet known.

**What Can Be Done?**

In 1990, the Council on Cardiovascular Disease in the Young of the American Heart Association held a conference on the insurability of young adults who have heart disease. Suggestions from that conference hold true today because little has improved over the last decade. These included care standards developed by cardiologists rather than insurers, definition of appropriate uses of diagnostic and therapeutic technologies, treatment of patients by properly trained cardiologists, a uniform coding mechanism, tort reform, and provision of some form of guaranteed coverage through a national healthcare system or through private providers who would remove the preexisting condition exemption.

Currently, we use individualized strategies in which patients have had to quit work, stay in school, go onto Medicaid
(stopping promising careers), or exaggerate and say their illness is disabling so that Social Security or Medicare covers them. Some seek to avoid clinic visits, cardiac catheterization(s), or operation(s) because of the personal financial consequences. Some have died as a result.

Because CCS covers cystic fibrosis and hemophilia after the age of 21, why not do the same for CHD? These agencies are underfunded, and adults with CHD would be an expensive population, so it has been easiest to ignore them. The same issue applies to insurers, who become more secure financially by not absorbing this group of patients. We must continue to advocate treatment for young adults who have CHD, finding some way to be sure that they are able to continue receiving care for the conditions for which so many resources already have been used, ensuring a state of good health and productivity.

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