MANAGING THE TRANSITION TO ADULTHOOD BEGINS IN CHILDHOOD

Transition into the adult health care system is crucial for patients with congenital heart disease (CHD), as well as for adolescents with many other chronic conditions. Indeed, “arranging efficient and caring transfer for adolescents from pediatric to adult care (is) . . .one of the great challenges facing pediatrics—and indeed the health care services—in the coming century” (1). Centers that care for adolescents and young adults with CHD need to develop structured plans for the transfer of care from the department of pediatric cardiology to that of adult cardiology. A comprehensive program taking a developmental approach beginning in childhood and adolescence should achieve better results than programs that focus only on the transfer to adult care at a specified age.

Currently, sufficient empirical data are not available to support the identification of “best practices” regarding transition in this patient group. However, descriptive and qualitative studies have indicated that the key elements of an effective transition program include:

1. A policy on timing of transfer to adult care (age 18 or upon leaving school is recommended by many, with some flexibility);
2. A preparation period and education program that focus on a set of skills that enables young people and their families to function in an adult clinic (e.g., understanding the disease, treatment rationale, and source of symptoms; recognizing deterioration and taking appropriate action; learning how to seek help from health professionals and how to operate within the medical system);
3. A coordinated transfer process (including a detailed written plan and pretransfer visit to the adult clinic, with an introduction to the adult provider and with a designated coordinator such as a clinic nurse);
4. An interested and capable ACHD regional center that is at least equivalent in quality to that of the pediatric source the patient is leaving (see subsequent discussion);
5. Administrative support; and
6. Primary care involvement.

The published data also suggest that transitions are more successful in health care settings where:

1. The preparation for transition begins before adolescence, and transition is seen as an essential component of high-quality health care.
2. There is a formal transition program.
3. Young people are not transferred to adult services until they have the necessary skills to function in an adult service and have finished growth and puberty.
4. There is an identified person on both the pediatric and adult teams who has responsibility for transition arrangements (usually nurse specialists).
5. Management links are developed between the pediatric and adult systems, and financial and contracting issues are worked out in detail and put in writing.
6. The evaluation of transition arrangements is undertaken as part of a continuous quality improvement process.
7. Transfer is planned and carried out during a period of medical stability; and
8. After transfer, there should be ongoing consultation with the referring pediatric cardiologist.

Sometimes parents need counseling and support to let go of their adult offspring. Adolescents need support and encouragement to begin making decisions, maximizing their strengths and abilities, and taking control of their lives. Support groups for patients and/or parents may provide a reality-based interchange of shared issues. Large referral institutions can provide direct support group access, but patients in other areas may be best served by Internet support groups (e.g., CACHnet [www.cachnet.org], Children’s Health Information Network [http://tchin.org]) and newsletters.

Factors such as a lack of symptoms and strong attachment to a pediatric cardiology program may result in the failure to transfer successfully to ACHD services. Many adults with CHD have the impression that the operation performed in childhood was a “cure.” They may be unaware of their prognosis and believe that regular cardiac follow-up is unnecessary. Comprehensive, individualized education regarding their cardiac condition and health care needs, as well as a “road map” regarding life-style implications, can help patients to achieve independence. In addition to routine health care recommendations, the health education objectives should be specific for young adults with CHD and should include specific information on diagnoses and operations, medications and their side effects, endocarditis prophylaxis, exercise prescription, contraception and family
planning, career planning and resources, insurance, guidelines for frequency of medical follow-up, and dental care. Education regarding symptoms that could be serious, such as arrhythmias, is essential. This information should be summarized in a “health passport” that can be held in the patient’s possession.

The timing and manner of communicating with patients is very important. Presenting an overwhelming amount of information in a single session should be avoided, especially during early transition visits. The potential to throw the patient into despair or denial exists and could lead to avoidance of much-needed visits to a medical center in the future. Discussions should proceed at a rate commensurate with the psychosocial development and circumstances of the patient. Traditionally, many pediatricians have continued to care for adults with developmental disabilities, but provision for this group needs to be part of the planning for adult care.

Children with CHD should be given the expectation that they will grow up to be healthy and able to work. Questions like “what do you want to be when you grow up?” should be asked by pediatric primary and specialty care providers, starting in early childhood (~age 4). In their transition to adulthood, adolescents should prepare for economic independence and vocational competence. Although the majority of patients with postoperative CHD are functionally normal, the label of heart disease or a mild disability can heighten these developmental challenges. The possibility of further surgery can also impede long-term planning. Adolescents should be encouraged to achieve higher education and skills necessary for employment in occupations that are reasonable in relation to their work capacity. Vocational guidance should be emphasized in early adolescence (ages 13–15), as training and acquisition of special knowledge and skills are important to the young adult if he or she is going to compete with other applicants.

**UNIQUE MEDICAL ISSUES**

**Cyanotic patients.** Cyanotic patients with CHD should be seen regularly at a regional ACHD center. Their physicians should be aware of special issues in their care. There are medical problems extrinsic to the cardiovascular system, which can cause significant morbidity and mortality in such patients. Severe cyanosis leads to marked erythrocytosis and, frequently, to low platelet counts (>100,000) (2), which, fortunately, seldom lead to significant bleeding. The absence of erythrocytosis (e.g., hemoglobin >17.0 g/dl) in such patients should raise concern about a “relative anemia” and its cause and implications. Excessive erythrocytosis adversely affects whole blood viscosity, but this problem is normally not associated with symptoms until the hematocrit levels are at least >60% to 65%. The only valid indication for therapeutic phlebotomy is to alleviate symptoms of hyperviscosity. Many patients undergo phlebotomy by primary care physicians to reduce their hematocrit. This practice should be discouraged; phlebotomy should be undertaken only in a center that manages cyanotic patients. When blood is removed, volume replacement with normal saline is recommended. Failure to follow this procedure can be associated with hypotension, increased right to left shunting, stroke, seizures, and death, especially in patients with pulmonary vascular disease. Multiple phlebotomies result in iron depletion, which is disadvantageous and has an independent negative effect on exercise performance. Iron deficiency is associated with impaired small-vessel blood flow and an increase in the risk of reversible ischemic neurologic deficits and stroke (3). When iron deficiency does occur, it should be treated.

Cyanotic patients frequently have hyperuricemia with arthralgia, gouty arthritis, and overt tophaceous deposits of urate. These abnormalities are secondary to low uric acid fractional excretion, not to urate overproduction (4). Therefore, hyperuricemia is a marker of abnormal renal function, which can be a problem in patients with long-standing cyanotic heart disease. Acute gouty arthritis responds to colchicine; special care should be taken to avoid dehydration that could occur with vomiting and diarrhea, which would require cessation of drug therapy. Allopurinol, with or without a uricosuric agent, is considered for symptomatic hyperuricemia refractory to other medications (2).

Pulmonary vasodilator therapy, specifically prostacyclin, has been used successfully in a small number of patients with Eisenmenger syndrome, serving as a bridge to repair in a few. The role for pulmonary vasodilators is being studied intensively.

**Noncardiac surgery.** Adults with moderate and complex CHD (see Tables 4 and 5 of Task Force #1) who require noncardiac surgery have special needs to be addressed by the surgical and anesthesia team. Ideally, operations in patients with complex CHD should be performed at a regional ACHD center with physicians experienced in the care of these individuals and with the consultation of cardiologists trained in this discipline (5). Frequently encountered management issues include cessation of anticoagulant agents and use of antibiotics for endocarditis prophylaxis (6). Important considerations for anesthetic management include the functional class of the patient, ventricular function, persistent shunts, valvular disease, arrhythmias, erythrocytosis, pulmonary disease, and pulmonary vascular disease.

Risk factors that help predict the possibility of perioperative risk include cyanosis (p = 0.002), treatment for congestive heart failure (p < 0.001), poor general health (p < 0.001), and younger age (p = 0.03) (7). Patients with pulmonary hypertension probably have a higher complication rate (15%) than patients without pulmonary hypertension (4.7%; p = 0.08). Procedures performed on the respiratory and nervous systems seem to be associated with the most complications.

The extent of preoperative evaluation varies depending on the complexity of the heart disease. A complete understanding of the patient’s underlying anatomy is necessary. A preoperative echocardiogram and, rarely, cardiac catheter-
ization may be indicated when recent data are not available. Stress studies may be indicated to exclude coexistent coronary artery disease in older adults. Estimates of pulmonary hypertension are useful, because some patients are at increased risk for Eisenmenger syndrome (8). If pulmonary disease exists, preoperative pulmonary function tests may be necessary to determine its severity and to estimate the need for postoperative ventilation. Preoperative laboratory testing in cyanotic patients should include an evaluation of the hematologic system (5), including coagulation and platelet abnormalities (9). Isovolumetric phlebotomy to a hematocrit <65% has been recommended to improve hemostasis. Practices vary among centers, and further study is warranted.

In patients receiving long-term anticoagulation, protocols for stopping warfarin, by using perioperative heparin, and restarting warfarin should be developed and coordinated with the surgical and dental team to minimize blood loss and prevent complications. The decision for invasive monitoring, such as intra-arterial catheters and/or central venous catheters, should be based on the magnitude of the operation and the specific nature of the cardiac defect. The decision to monitor invasively should be weighed against the risk of complications. In all cyanotic patients, meticulous attention should be paid to all intravenous lines to ensure freedom from air bubbles, which may cause systemic air embolism. Intraoperative transesophageal echocardiography may be useful for continuous monitoring of ventricular function and for estimating preload conditions. The choice of anesthetic agent depends on the severity of the cardiac disease and other co-morbidities and must be tailored to the operation. Avoidance of myocardial depression and hypovolemia is emphasized. Epidural anesthesia can provide excellent operative and postoperative analgesia, with minimal cardiovascular side effects in select patients.

CONTRACEPTION AND PREGNANCY IN WOMEN

Contraception. All patients must be well informed of the risks of pregnancy associated with their condition and the available options to avoid pregnancy when desired. The risks of pregnancy vary widely among the specific types of CHD (see subsequent discussion). There are no systematic outcome data on the safety of contraceptive methods in women with CHD. The choice of contraceptive method is usually made by the patient. However, it is the responsibility of the physician to provide thorough counseling about the risk of unplanned pregnancy in the case of non-compliance, poor acceptance or failure of the contraceptive technique, and any risk associated with the specific method including infective and thromboembolic complications. In considering surgical sterilization because of high risk, the patient should be fully informed of the potential for medical advances that may permit future pregnancy at lower risk.

Pregnancy risk. Pregnancy in women with CHD not complicated by Eisenmenger syndrome is associated with a low mortality (10–12). However, potential risk factors for maternal morbidity include poor maternal functional class, poorly controlled arrhythmias, heart failure, cyanosis, significant left heart obstruction, and a history of cerebral ischemia (10–14). Cyanosis is a risk factor for fetal and neonatal complications (10–12,14). On the basis of these risk factors, patients can be stratified into low-, intermediate-, or high-risk categories (12). An absence of these risk factors would generally place patients into a low-risk category. The highest risk is associated with Eisenmenger syndrome, in which postnatal maternal mortality can exceed 50%. Because much of the current data are based on retrospective case series from tertiary care institutions, one should exercise caution in risk stratification of pregnant women with uncommon conditions such as Mustard/Senning or in those who have had a Fontan procedure (15–17). Patients with these lesions or procedures should be placed in the intermediate-risk category until additional data become available. The risk of in vitro fertilization for surrogate pregnancy in high-risk women with CHD has not been defined. Medical or surgical termination of pregnancy in intermediate or high-risk patients requires careful monitoring, and preferably it should be done in a regional ACHD center.

Special needs of pregnancy. Women with heart disease who are at intermediate or high risk or an uncertain level of risk for complications should be managed in a high-risk perinatal unit by a multidisciplinary team including an obstetrician, cardiologist, anesthesiologist, and pediatrician. The team should meet early in the patient’s pregnancy to review the cardiac lesion, anticipated effects of pregnancy, and potential problems and to develop a management plan. Specific issues that need to be considered include the timing and mode of delivery, the type of anesthesia to be used, the need for hemodynamic monitoring before and after delivery and the use of antibiotic prophylaxis. Women with heart disease in the low-risk group can usually be managed in a community hospital setting.

Risk of recurrence of CHD in offspring. Genetic counseling regarding etiology, inheritance, recurrence risk, and prenatal diagnosis options should be made available to all patients with CHD. It is important to obtain the patient’s prenatal and postnatal history, including maternal exposure to teratogens, as well as a detailed family history, and to perform a thorough examination looking for congenital abnormalities (18).

In all women contemplating pregnancy, exposure to teratogens should be investigated; in some cases, finding an alternative medication will be necessary. Angiotensin-converting enzyme inhibitors and angiotensin II receptor antagonists should not be used during pregnancy. Medications for which substitution should be considered include warfarin and amiodarone. No medications, including over-the-counter preparations, should be taken during pregnancy without physician approval. Preconception consumption of
multivitamins including folic acid decreases the incidence of CHD (19).

Knowledge of the genetic basis of CHD is expanding rapidly. The role of genetic testing is evolving, and genetic counseling should be made available. The recurrence rate of CHD in offspring is variable, ranging from 3% to 50%. A higher recurrence risk when the mother rather than the father is affected has raised the possibility of mitochondrial inheritance in some patients (20). Diseases with a single gene disorder and/or chromosomal abnormalities are associated with a high recurrence rate. In Marfan, Noonan, and Holt-Oram syndromes, there is a 50% risk of recurrence.

Fetal echocardiography at 16 to 18 weeks gestation should be available to all patients with CHD. Chorionic villus sampling or amniocentesis may be useful after discussion of the potential risks and benefits.

**EXERCISE TOLERANCE AND REHABILITATION**

Exercise data. The ability to exercise is one measure of quality of life, and it is used to assess the effect of disease, the results of treatment, and the ability to tolerate the stress associated with pregnancy or needed surgery. There have been numerous studies of exercise tolerance in children and adolescents with CHD but very few studies in adults.

Adults with pulmonary stenosis have well-preserved but still subnormal exercise tolerance. Exercise tolerance for adults with aortic stenosis or a ventricular septal defect (VSD) is subnormal and even less than that for adults with pulmonary stenosis. Rather surprisingly, two investigators have reported that exercise tolerance is subnormal for patients who had repair of an atrial septal defect (ASD). Reybrouck et al. (21) have demonstrated that the age when closure of an ASD is performed influences postoperative exercise tolerance. Adults with complex conditions, such as Ebstein’s anomaly or a single ventricle, have a markedly abnormal exercise tolerance. There are few studies of exercise tolerance of adults with transposition of the great arteries, pulmonary atresia with or without VSD, and other complex conditions. However, studies of children and adolescents with these defects show subnormal exercise tolerance, and it is assumed that exercise tolerance would be no greater in adults with similar defects.

Recommendations for athletic participation for patients with CHD were published in the 26th Bethesda Conference (22) and are the best consensus data available.

Exercise training and rehabilitation. Because adults with CHD have subnormal exercise tolerance, an obvious question is whether physical conditioning reduces symptoms and improves exercise tolerance and quality and/or length of life. These issues have not been studied. There are numerous studies validating the benefits of exercise for healthy adults and those with coronary artery disease (23). There have been several studies of the utility of exercise programs for children with CHD (24,25). Despite major design problems, these studies demonstrate that a structured rehabilitation program can increase exercise efficiency. Improved exercise performance (i.e., maximal oxygen consumption) was not demonstrated in most studies. Because all of these studies were small, a survival benefit could not be demonstrated. An alternative to a costly structured rehabilitation program is a simple home program of physical rehabilitation, which was successful in one study (26). The efficacy and safety of a structured exercise rehabilitation program for adults with CHD are unknown. Issues that require further study in adult patients include the efficacy of such a program in improving fitness and aerobic capacity, the safety of such programs, and the interaction between congenital and acquired heart disease.

**PSYCHOSOCIAL ISSUES**

Only recently have patients with complex CHD survived into adult life in large numbers. Their survival creates hope that continuing advances will help them maintain both quality of life and longevity. However, patients may experience despair due to their awareness of residual morbidities and the knowledge of possible or probable early mortality, or limitations in their social lives and educational or occupational attainment. Healthy psychosocial functioning depends on their ability to balance hope and despair. Adults with CHD must also confront both CHD-specific and general developmental tasks. Psychosocial issues may be affected by lesion severity (simple vs. complex), visibility (e.g., cyanosis), and functional disability (27).

Life-span development and CHD. Table 1 is a proposed model outlining developmental tasks faced by individuals with CHD, beginning in adolescence; this model could be tested in future studies.

Physical development. Adults with CHD may struggle with physical appearance (e.g., scars, smaller body size, cyanosis, clubbing), physical limitations, and acute or gradual decreases in physical functioning (28–30). Physical decline may be difficult to deal with, as peers are often less able to empathize with these changes.

Social and family relations. Adults with CHD are less likely to be married or cohabiting or to have children and are more likely to live with their parents, as compared with healthy peers (31,32). These differences may reflect life-style decisions made based on beliefs or knowledge regarding shortened life-expectancy, concerns about pregnancy risks, economic constraints, or the need for social support (33,34). Patients limiting themselves due to misinformation need counseling. Difficulty discussing CHD issues with family or friends is common among adolescents and young adults with CHD, especially among patients whose parents rarely discussed their own disease (35). Patients and their families may need assistance in finding a balance between independence and interdependence that optimizes the psychosocial and physical health of the patient with CHD. Adolescent and young adult patients, in particular, may need assistance...
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*Life-expectancy varies with lesion severity and is increasing with improved medical care.
in ways to educate their peers regarding CHD and to maintain a peer network.

**Emotional health.** Although most adolescents and adults with CHD appear to be free of psychopathology, there are too few studies to draw conclusions about the emotional health of this group of patients. Results of comparisons of emotional adjustment between patients with CHD and healthy peers have been mixed. One study found that when items likely reflecting CHD symptoms (e.g., dizziness) were removed from analyses, group differences were no longer significant (36).

In patients with acquired heart disease, depression and anxiety are linked to an increased risk of cardiac and all-cause mortality and sudden cardiac death (37–40). Given the high prevalence of arrhythmias with complex CHD, this potential relationship should be examined. Twenty percent of all adults have a psychiatric disorder (41). Even if adults with CHD were not at increased risk of psychopathologic conditions, one in five patients would be expected to have a significant mental health problem. Surgery, hospital admission, invasive medical procedures, and even routine appointments may trigger emotional distress, especially in individuals with a pre-existing emotional disorder.

**Medical issues.** Only one half to three fourths of adults with CHD can correctly state or describe their diagnosis (42–44). Given complex anatures and surgical repairs, it may not be possible for all patients with CHD to have precise anatomic knowledge, but aids such as a “health passport” may be helpful in providing the patient with the most important information.

Adults with CHD may have difficulty coping with repeated hospital admissions, operations, and other painful medical procedures. They underwent cardiac surgery during an era of inadequate pain control (45), which may result in centrally mediated pain sensitization for them (46). Patients with CHD should receive education about pain and its management and receive optimal medication and management of procedure-related pain. Health care personnel must also be informed of the special issues related to pain management in this group.

**Health behaviors.** Poor knowledge of behaviors related to endocarditis and its prevention are common (42–44,47). Unrealistic fears (e.g., fear of damaging the heart or having a cardiac arrest) may be a factor in their lack of activity. However, some patients who have been advised against heavy exercise will engage in it anyway (e.g., by engaging in contact sports) (30). Although consensus (27) and common sense suggest that regular medical follow-up, as compared with nonattendance, should result in better outcomes, proof is lacking. Patients’ beliefs regarding treatment effectiveness (e.g., medication) or health behaviors (e.g., regular medical appointments) are a significant predictor of compliance and should be assessed.

**Personality and identity.** Establishing one’s identity, balancing independence with interdependence, and accepting death are tasks of normal development. In addition, adults with CHD must incorporate their condition into their identity, deal with a lack of control over changes in physical functioning, resolve the loss or disruption of typical developmental achievements (e.g., surgery may result in loss of an academic year), and face the prospect of premature mortality. These issues must be faced repeatedly throughout adulthood and may necessitate counseling.

**Screening and prevention.** Routine screening for psychosocial or physical problems is not without risk (48–50) and should be undertaken only if there are accurate measures, appropriate mechanisms to provide feedback, and appropriate resources for treatment. Because validated CHD-specific measures do not exist, measures of perceived risk of CHD complications and health behaviors should be developed.

**Treatment issues.** Although there are validated psychosocial and pharmacologic treatments for many psychological disorders (41), treatment effectiveness could be enhanced if interventions are adapted to deal with CHD-specific issues. Treatments specific to CHD should focus on enhancing knowledge, modifying maladaptive beliefs, and dealing with periods of transition and acute stress.

The level of treatment intensity could be tailored to the severity of the problem, ranging from self-help materials for those with mild or moderate problems to individual or group therapy for those with severe problems. Creative solutions that offer individual counseling for patients at a distance from a regional ACHD center are needed. As interventions are developed, it is crucial to document procedures and evaluate effectiveness so centers can share and build on each other’s experiences.

**RECOMMENDATIONS**

**Transition to Adult Life**

- Structured plans should be developed to transition patients from pediatric to adult CHD care. Transition to a regional ACHD center can be difficult for patients, and the presentation of an overwhelming amount of information in a single session should be avoided. Discussions should proceed at a rate commensurate with the psychosocial development and circumstances of the patient.
- Individual patient education regarding his/her diagnosis and specific health behaviors should be a priority.
- Important historic information, including comprehensive diagnostic data, procedures, operations, and medications, should be kept by the patient as a summary of past and present important health issues. The American College of Cardiology should support development of a health care “passport,” which would be useful for all patients with CHD and their health care providers.
- A continuum of vocational assessment beginning in childhood should be available for patients with CHD and should be continued during the developmental, adolescent, and young adult years.
Noncardiac Surgery

- Ideally, noncardiac operations on patients with complex CHD should be performed at a regional ACHD center with the consultation of an anesthesiologist with experience in CHD, particularly for more complex surgery or for patients with adverse risk factors that include poor functional class, pulmonary hypertension, CHF, and cyanosis.

Reproductive Issues

- Contraceptive counseling must be available, when appropriate, to all patients with CHD.
- A multidisciplinary team at a regional ACHD center is needed for pregnancy and delivery, as well as for the management of indeterminate-, intermediate-, or high-risk patients.
- The recurrence risk of CHD is highly variable and should be discussed prospectively with all patients. Genetic counseling should be made available through regional ACHD centers.

Exercise and Rehabilitation

- Guidance for athletic participation for patients with CHD should be in accordance with the published recommendations of the 26th Bethesda Conference report, which represents the best consensus data available.
- The efficacy and safety of exercise rehabilitation programs in adults with CHD have not been studied, and research in this area should be supported.

Psychosocial Issues

- The emotional health of adults with CHD should be a priority in the overall care of this patient population. Appropriate screening and referral sources for treatment should be available at all regional ACHD centers.
- Tools for screening of psychosocial problems in this population should be developed and tested.
- Data should be developed to assess the impact of regular follow-up care on the long-term physical and psychosocial health of adults with CHD.
- Available professionals and facilities for the treatment of psychological disorders are scarce, and creative solutions for counseling patients in groups and/or those at a distance from home should be developed.

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Task Force 3: Workforce Description & Educational Requirements for the Care of Adults With Congenital Heart Disease

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INTRODUCTION

The expansion of the population of adults with congenital heart disease (CHD) and the increasing survival of patients with complex disease into adulthood have heightened the need for specifically trained individuals who can provide comprehensive outpatient and in-patient care and consultative services to these patients (1–3). In addition, such individuals should have the educational background necessary for successful academic careers in order to advance knowledge and educate other providers. At present, only a few specialists in the U.S. have been specifically trained for this role. Most adult congenital heart disease (ACHD) patients are followed by adult cardiologists who have not had much training in the diagnosis or management of CHD or by pediatric cardiologists who have had little or no experience or training in comprehensive adult care. Adult cardiologists often unofficially consult with pediatric cardiologists to plan management, but uncompensated time and medico-legal risks have made this practice increasingly difficult for pediatric cardiologists. Pediatric cardiologists may effectively co-manage adult patients with an internist or family medicine practitioner, but they cannot provide the full complement of in-patient or invasive services that may be needed. In some cases, adult and pediatric cardiologists follow ACHD patients in a joint clinic. These practices vary considerably depending on patient volume, institutional resources, and physician interest.

The routes by which adult and pediatric cardiologists in this field arrived at their level of expertise are varied. Many, if not most, adult cardiologists have had on-the-job training which provided them with an opportunity to learn, in an environment of collaboration, from pediatric cardiologists and cardiac surgeons. Many pediatric cardiologists have become increasingly involved with adults with CHD as their pediatric patients have aged. Although ACHD patients will continue to rely on these traditionally trained