Adults with Congenital Heart Disease: The Critical Transition from Pediatric to Adult Care

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ABSTRACT

Objective: To review the management of patients with congenital heart disease (CHD) transitioning from pediatric to adult care.

Methods: Review of the literature.

Results: Persons with CHD require close monitoring and evaluation throughout life to address the physiologic consequences of acquired cardiopulmonary, gastrointestinal, and renal disease in the setting of underlying congenital heart lesions. During the transition from pediatric to adult cardiology, a high proportion of patients are lost to follow up or have long gaps in care after leaving pediatric cardiology, which can lead to poor outcomes. Care of the adult with CHD requires close coordination between the patient's primary care physician), cardiologist, adult CHD specialist, and other specialists. The transition process for CHD patients begin at 12 years of age, with a goal of discussing future expectations of the child's education, employment, and independent living. Successful transition programs use a systematic approach to addressing the medical, psychosocial, and educational/vocational needs of the adolescent as he or she moves from the family-centered pediatric to the patient-centered adult health care system.

Conclusion: The transition from pediatric to adult care in ACHD patients is best provided through a comprehensive transition program that begins in early adolescence and enables patients to take charge of their disease process in adulthood, allowing them to maximize their quality of life and societal contributions.

Keywords: adult; congenital heart defects; complications; disease management; patient care team.

The population of adults with congenital heart disease (CHD) in developed countries has grown at an exponential rate in the past 4 decades. With advances in medical care and surgical interventions, the proportion of pediatric patients reaching adulthood has increased from 15% in the 1930s-60s to more than 95% for patients with mild to moderate complexity CHD. The rate of survival to adulthood for patients with severely complex CHD remains lower at around 56%.[1]

There are now more adult than pediatric patients with CHD in the United States. Because adult CHD (ACHD) patients have increased morbidity and mortality in their young adult years, it is imperative for all providers to understand and address the long-terms needs of this population. Unfortunately, adults with CHD do not always receive adequate health care, frequently because they are lost to follow-up, particularly during their adolescent years when they are expected to gain independence in their medical management. As will be discussed, CHD is a chronic illness fraught with numerous expected and unexpected complications that require close monitoring and re-interventions. Effectively anticipating and addressing these complications requires a standardized and comprehensive process of transition from the pediatric to the adult population to ensure maximal quality of life.

Epidemiology

The actual prevalence of ACHD in the United States is unknown, as a national database of persons with CHD has not been established.[2] In contrast, Europe and China have maintained databases that enable ongoing monitoring of the evolving CHD epidemiology in those regions.[3,4] The best estimates of the U.S. incidence and prevalence of ACHD stem from extrapolations from Canadian data.

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According to this data, there were more than 1.2 million adults with ACHD in the United States in 2012, with an anticipated 5% annual increase.[1,5] However, the limitations of such extrapolations must be noted, as the Canadian population does not perfectly mirror that of the United States. Canada has lower infant mortality and adult obesity rates, and the United States has larger African American and Hispanic populations.[6] Also, the juxtaposition of universal access to health care in Canada and the socioeconomic class–dependent access in the United States causes variations in care and outcomes of ACHD between the 2 populations. These differing genetic and social backgrounds may change the incidence of CHD by affecting maternal-fetal health.[7]

The 32nd Bethesda Conference on "Care of the Adult with Congenital Heart Disease" in 2000 was tasked with characterizing the ACHD population in the United States. This project found a prevalence similar to that of the Canadian extrapolation and showed that among persons with ACHD in the United States, 45% have mild disease, 37% moderate disease, and 13% severe disease.[8]

Characterizing the true incidence of CHD in the United States also has proven difficult because of variations in the definitions and methods used to detect lesions across the multiple studies that have looked at this matter. The estimated incidence of CHD, grouped according to severity, is 2.5 to 3 per 1000 live births for severe CHD, and from 3 to 13 per 1000 live births for moderately severe forms.[9] When all forms are considered, including minor CHD (which includes tiny muscular ventricular septal defect [VSDs] present at birth and other trivial lesions), the total incidence of CHD rises to 75 per 1000 live births.[9] CHD is one of the most common chronic illnesses in young adults with special health care needs.

Complications in Adulthood

The ACHD population represents a diverse population in terms of severity of CHD, history of surgical/catheter-based interventions, and socioeconomic status. However, a unifying clinical concern for these patients is their increased risk for morbidity and mortality in the young adult years. Despite the tremendous advances in the field over previous decades, mortality in this population in adulthood is estimated to be up to 7 times higher compared to age-matched peers.[10,11] For many patients, palliative CHD interventions result in a significant drop in early morbidity and mortality but frequently lead to delayed morbidity from secondary multi-organ complications as these patients transition from pediatric to adult care. For example, due to the chronic low flow and low cardiac output state created by Fontan palliations, patients are at risk for diastolic dysfunction, arrhythmias, thrombotic events, protein-losing enteropathy, and cirrhosis/congestive hepatopathy, among other chronic conditions. These patients require frequent follow up and management by a multidisciplinary team including a primary care provider and various specialty groups.

Cardiac Disease

The most common causes of death in ACHD patients are heart failure (27%) and sudden cardiac death (19%), which occur at mean ages of 48 years and 39 years, respectively.[10] The form of heart failure in ACHD patients is related to subsystemic right ventricle (RV) dysfunction, coronary under-perfusion, residual shunts, and residual progressive valve regurgitation. One of the more common examples of this is seen in palliated Tetralogy of Fallot patients who have undergone a transannular patch as a neonate. These patients are frequently left with significant pulmonary regurgitation leading to RV dilation, RV failure, and subsequent left ventricle (LV) failure. Another common example is the patient with dextro-transposition of the great arteries (DTGA) status post atrial switch who has a subsystemic morphologic RV. These patients will often develop significant RV dysfunction related to the chronic high pressures associated with systemic circulation.

Arrhythmias are a major contributor to morbidity and mortality in this population and are the most common reason patients present back into care. Difficult to control, multifocal intra-atrial re-entrant tachycardia is extremely common in ACHD, with an estimated 50% of all patients developing atrial arrhythmia by age 55. A recent study determined that the risk of atrial fibrillation in individuals with CHD was 22 times higher than that in age-matched controls, with the highest risk being seen in patients with conotruncal defects. Furthermore 10% of these patients develop heart failure.[12] The risk for, incidence of, and type of arrhythmia is associated with the severity of the

congenital heart lesions, as well as the type and timing of surgical interventions. Later age of repair has been associated with an increased likelihood of arrhythmias.[13] Tetralogy of Fallot is an example of a moderately complex congenital heart lesion and is the most common cyanotic congenital heart lesion. In these individuals, the risk for atrial tachycardias, ventricular tachycardias, and need for a pacemaker is much higher than in age-matched peers.[14] This includes an increased risk of sudden cardiac death, with many of these patients requiring placement of an implantable cardioverter defibrillator.

Pulmonary Disease

There is a 44% to 56% prevalence of restrictive pulmonary disease in the ACHD population, compared to 9% in the general non-CHD adult population. The incidence of pulmonary hypertension is also significantly higher in the ACHD population. The etiology for development of pulmonary hypertension is multifactorial, including chronic thromboembolic disease, left-sided heart disease, long-standing left to right shunts, and obstructive sleep apnea. These conditions have a significant impact on survival, as moderate/severe lung function impairment is an independent predictor of survival. Patients with shunt lesions are at risk of developing pulmonary arterial hypertension later in life,[1] which quadruples the risk of all-cause mortality and more than triples the risk of cardiovascular mortality.[7]

Liver Disease

Hepatic morbidity associated with palliated CHD is often related to prior surgical interventions. The most common morbidities include chronic hepatitis C and liver failure from chronic under-perfusion and passive congestion, especially following Fontan palliation. Long term, these complications can lead to cirrhosis and hepatocellular carcinoma.[15-18] Unfortunately, hepatic morbidity often precludes patients from having a surgical intervention, complicating the management of a population with baseline significantly increased need for surgical re-intervention.

Renal Disease

Approximately 50% of the ACHD population has some degree of renal dysfunction, with a higher incidence in cyanotic CHD.[19] The American College of Cardiology/American Heart Association (ACC/AHA) recommends routine assessment of renal function in all adults with moderate and severe CHD due to its association with a poor prognosis in the ACHD population.[1] In the immediate cardiac postoperative period, acute kidney injury leads to an eightfold increase in mortality.[20] Over the longer term, there is a fivefold increase in mortality with moderate to severe renal impairment and a twofold increase with mild renal impairment compared to those with normal renal function.[21]

Acquired Cardiovascular Disease

As the ACHD patient ages, acquired cardiovascular disease becomes a significant issue. Approximately 80% of adults with CHD have at least 1 cardiovascular risk factor,[22] though overall there is a relative lack of specific data regarding the U.S. population. Surveillance of the Canadian CHD population older than 65 years shows a 47% prevalence of hypertension,[23] with increased risk in certain conditions such as aortic coarctation and renal disease associated with CHD. Although studies on the increased risk of diabetes mellitus in the ACHD population have yielded conflicting results,[22,24] there is evidence of abnormal glucose metabolism in ACHD patients, which is a predictor of cardiac morbidity and mortality.[25,26] The incidence of hyperlipidemia in U.S. ACHD patients is estimated to be at least as high as that of the general population.[1] These factors combine with abnormalities in the myocardial substrate, hemodynamic abnormalities, arrhythmias, and sequelae of surgical repairs to confer an increased risk of ischemic heart disease and cerebrovascular disease in the ACHD population.[15,27] One large case-control cohort study showed that the risk for ischemic heart disease was 16.5 times higher in patients with CHD as compared with non-CHD patients, with the highest incidence being in those with conotruncal defects and severe non-conotruncal defects. Interestingly, hypertension and diabetes were less common among CHD patients with ischemic heart disease than among non-CHD patients with ischemic heart disease.[28]

Adults with CHD have an increased risk for cerebrovascular disease compared with the general population, and cerebrovascular disease appears to occur at a younger age.[29] The risk of ischemic stroke in individuals

with ACHD younger than 55 years is 9 to 12 times higher than that in the general population. As in the general population, the incidence of ischemic stroke in ACHD patients increases with age, and in those older than 55 years, the incidence remains 2 to 4 times higher than in the general population.[30,31]

Clearly, complications arising from therapeutic interventions in CHD patients contribute significantly to morbidity/mortality in adult life, which underscores the need for life-long follow up and prevention of lapses in care.

The Transition from Pediatric to Adult Care

The monitoring and evaluation of CHD patients throughout life requires close coordination between the patient's primary care physician, cardiologist, ACHD specialist, and other specialists, as appropriate. The timing of routine follow-up appointments is largely dependent on the severity of the congenital heart lesion and clinical status of the individual patient. Routine surveillance often includes cardiac imaging, preconception/genetic counseling, Holter screenings for arrhythmia, laboratory testing, and titration of medication. Unfortunately, only 30% of adults with CHD receive the recommended cardiac care.[32]

Children with chronic conditions transitioning to adulthood frequently experience a drop off in coordinated services as they transition from pediatric to adult medicine. Adult institutions often have less multidisciplinary support staff in the form of social workers and case management.[33] Furthermore, a recent systematic review of articles that outlined the transition process from pediatric to adult cardiology in the CHD population showed that a high proportion of patients were either lost to follow up or had long gaps in care after leaving pediatric cardiology, with the first lapse in care commonly occurring at approximately age 19 years.[28,34] A 2004 study showed that only 48% of adolescents with CHD underwent successful transition.[35] A multicenter study of 922 ACHD patients found a gap in care lasting longer than 3 years in 42%, with 8% having gaps exceeding 10 years.[36] Another study showed that lapses exceeding 2 years occurred in 63% of patients, with a median duration of lapse of medical care of 10 years. The most common reasons for lapse in care were: being told that cardiac follow up was not required (33%); being discharged from a children's hospital without appropriate follow up plans in place (23%); being aware of need for follow up but having no symptoms (19%); lack of insurance (18%); and ignoring follow up recommendations for fear of receiving bad news (7%).[37] Moreover, living independently from one's parents was independently associated with a lapse in care, and patients with moderate complexity defects were more likely to experience a lapse than those with high complexity defects.

In the absence of a structured transition program, there is often delayed or inadequate care, which can result in significant emotional and financial stress on families and increased stress on the health care system.[38] Inadequate, incomplete, or nonexistent transition and transfer for care has been shown to lead to poor health outcomes. Patients who experienced a lapse in care were 3 times more likely to require urgent cardiac intervention and to have an adverse outcome.[37] The urgent interventions required by these patients included pulmonary valve replacement, mitral and tricuspid valve repair/replacement, VSD closure, pulmonary artery stenting, Fontan revision, and pacemaker/defibrillator placement.[37] Clearly, there is significant room for improvement in the transition process of patients with CHD.

Best Practices in Transitioning CHD Patients to Adulthood

The overarching goal of pediatric to adult care CHD transition programs is to empower the patient and their support system to assume ownership of the disease process in order to maximize quality of life, life expectancy, and productivity.[39] This involves ensuring that the patient has a thorough understanding of their diagnosis, heart anatomy, prior cardiac interventions, limitations imposed upon them by their condition, and the frequency of their anticipated follow-up care. The components of a successful transition program include a systematic approach to addressing the medical, psychosocial, and educational/vocational needs of the adolescent as he or she moves from the family-centered pediatric to the patient-centered adult health care system.[40] The visits during the transition period are also an opportunity to discuss reproductive issues and the need for planning pregnancies for women with CHD. The goal is to encourage autonomy and promote ownership of their medical condition to the

best of their social-cognitive ability. Adolescents should be encouraged to speak alone with their doctor to foster independence and self-management in their disease process; this has been shown to be protective against failure in transition.[32] They should be encouraged to start calling their doctors, requesting refills, and making appointments.

The ACC/AHA appropriately recommend that the transition process for CHD patients begin at 12 years of age, with a goal of discussing future expectations of the child's education, employment, and independent living.[41] As part of this process, it is important that the practitioner educate the child and the family of the need for lifelong surveillance. The exact timing of the transition process is heavily influenced by a number of factors, including the degree of dependence of the child on their guardians, the severity of the congenital heart lesion, and the anticipated short- and long-term prognosis. However, regardless of these circumstances a reasonable age of transition into adult services should be established early on so that an expectation remains in place and the family is adequately prepared.

The challenge of learning how to navigate the adult health care system is as daunting for the transitioning patient as the medical consequences of their disease process. It is critical for patients to have easy access to social workers and case managers, ideally in the setting of a medical home, to connect them to community resources as needed. It is incredibly important that patients consider vocational options and training along with planning their insurance and/or disability qualifications as they move into adulthood. Establishing guardianship is also an important consideration for young adults with CHD who have remained dependent on their guardians.

Towards this end, the AHA/ACC has developed a curriculum that outlines the core principles that should be addressed before the patient moves to the ACHD clinic.[27] The transition program should be flexible to accommodate for the patient's degree of development, and the transfer should not occur before the adolescent has demonstrated the ability to independently manage their own health care to the greatest possible extent.

The ideal transition occurs through the auspices of a medical home that can coordinate the multiple subspecialists involved in the patient's care. However, what often occurs is that a patient transitions from the pediatric cardiologist's care before transitioning from pediatric to adult primary care. Prior to transition, the pediatric cardiologist should identify a cardiac destination at an ACHD center. This must be done in conjunction with the pediatrician, who will help identify an internist to take over the patient's primary care and continue the coordination via the medical home. Information regarding the patient's complete medical history, medication lists, exercise prescriptions, dietary restrictions, anesthetic issues, functional status, diagnostic studies, and comorbidities should be compiled in a health summary.[40] To aid the process of transitioning, the ACC has developed several tools that may be used during the transition process, including self-knowledge assessments and medical summary templates.[42]

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The Primary Care Provider's Role and the Medical Home

Ensuring adequate care during the transition period requires close coordination between the patient's various subspecialists. It is vital to avoid multiple subspecialists providing care without knowledge of each other's treatments, as the treatment course for each ACHD patient is dependent on their unique history of prior therapies.[27] The role of the primary care physician in establishing a "medical home" in this setting, as defined by the American Academy of Pediatrics Policy Statement, is exceedingly important.[43] In this structure, the primary care physician maintains an easily accessible, centralized, and comprehensive record of the patient's entire medical history, including surgical and medical treatments of both cardiac and noncardiac issues. Establishing the medical home framework is crucial, as it has been shown to lead to better outcomes in transitioning youth with special health care needs.[44]

With the establishment of this centralized care, the primary care physician must be able to negotiate the various medications prescribed by subspecialists and monitor for drug levels, adverse effects, and drug-drug interactions. ACHD patients also need regular monitoring and care aside from the care related to their chronic disease. Medical issues of particular importance to the ACHD patient include vaccinations, cholesterol and hypertension screening, cancer screening, and nutritional counseling. The primary care physician is responsible for addressing both the cardiac and noncardiac needs of the patient, ensuring that the patient truly receives comprehensive care. Thorough knowledge of a patient's unique medical/surgical history will enable the primary care physician to adequately triage and appropriately refer for the development of a new symptom in an ACHD patient. On the other end of the spectrum, the patient's subspecialists must maintain accurate and up to date information regarding their patient and transmit this to the patient's medical home.

ACHD Centers

ACHD centers are an important part of any ACHD patient's clinical team. Regardless of the complexity of the heart defect, there is tremendous value in the education and anticipatory guidance ACHD centers provide for their patients. The providers at these centers are often board-certified ACHD physicians who will work within a multidisciplinary team that includes mid-level practitioners, electrophysiology physicians, high-risk obstetrics/gynecology physicians, pulmonologists, and hepatologists. Each center differs in terms of their on-site interventional capacity and experience. However, the ACHD provider community is highly capable in directing patients who require interventions to centers of excellence, where there is proven quality in congenital surgical and interventional outcomes. ACHD centers often serve as the portals of reentry into care and are critical for providing and coordinating the complex care of each patient. Regular follow-up at these centers will ensure that patients receive adequate management of complications as they arise and preventive care against acquired heart disease.

The timing of follow-up at ACHD centers varies according to the complexity of heart disease. Individuals with simple CHD should be evaluated at an ACHD center at least once to determine the need for further follow-up. Patients with moderate and complex CHD must be monitored at a minimum of every 12 to 24 months, whereas very complex CHD should be monitored every 6 to 12 months.[23] The frequency with which the young adult population moves may hinder adequate continuity of care and long-term follow up; a searchable directory of ACHD clinics in the United States and Canada can be found at www.achaheart.org/your-heart/clinic-directory/clinic-listings/.

Managing Specific Issues in the Transitioning Patient

Arrhythmias and Heart Failure

As mentioned, arrhythmias in the ACHD population are extremely common, the most frequent being atrial arrhythmias, especially in patients who have undergone single-ventricle repairs. Patients with late repair of an atrial septal defect have a high incidence of supraventricular tachycardia, which can be treated with catheter ablation procedures.[45,46] Pacemaker implantation is another therapeutic option, especially in those who have undergone atrial surgery (ie, Mustard or Senning repairs). In these individuals, particularly in adolescents, abdominal implantation of a pacemaker generator may lessen the psychological impact of the external appearance of the pacemaker. In this population avoiding blunt contact sports (ie, tackle football, wrestling) is also important.[28] It is critical that adult and pediatric electrophysiologists work together in the care and management of these complex, recurrent arrhythmias.

As noted above, many ACHD patients will require surgical or catheter-based interventions (as high as 40% in 1 study),[47] and many encounter late-onset morbidity as a sequela of interventions earlier in life or as a result of failure of these interventions. The key for adult cardiologists and ACHD providers is delineating the reversible causes (eg, residual shunts, progressive valve regurgitation, and recoarctation) through routine intermittent surveillance, including echocardiograms, magnetic resonance imaging, and cardiac catherization, so that heart failure and arrhythmias in these patients can be identified, treated, and even prevented.

Pregnancy

Pregnancy is the most common reason for women to re-enter care. Pregnancy is associated with significant hemodynamic changes, resulting in an increase in cardiac output to up to 150% of pre-pregnancy levels at 32 weeks, and up to 180% during labor. The outcome of pregnancy in patients with CHD is favorable in most instances provided that functional class systemic ventricular function is good. Accordingly, pregnancy is contraindicated in instances of severe pulmonary arterial hypertension (eg, Eisenmenger's physiology), systemic ventricular dysfunction, and severe left-sided obstructions (eg, aortic or mitral stenosis). It is therefore imperative for health care providers to address the risks of pregnancy and the need for contraception with women who have CHD and are of reproductive age. The AHA advises beginning this conversation at 12 years of age and recommends that counseling be provided by health care providers knowledgeable in both CHD and adolescent health.[27] Given the thrombotic potential of estrogen-containing contraception, the selection of contraception for women with ACHD who are seeking birth control requires discussion between the health care provider and patient. Though there have been limited studies performed on the use of contraception in women with CHD, a British working group has developed a consensus statement regarding contraceptive use in women with heart disease based on the World Health Organization format.[48,49]

Surgical Procedures

The need for operative interventions and re-interventions, both cardiac and noncardiac, in many CHD populations is considerable. Regardless of the type of procedure, these patients should receive a comprehensive preoperative risk assessment as well as appropriate intraoperative and postoperative management, ideally at a center equipped to meet their unique needs. Approaching the surgical procedure under the guidance of an interdisciplinary team that includes an ACHD specialist, anesthesiologist, and surgeon ensures that critical issues for appropriate management are not overlooked.

The preoperative risk assessment should be aimed at identifying and minimizing major risk factors. Historical factors to consider include the congenital lesion, outcomes of prior surgeries, history of syncope or arrhythmias, and the presence of pulmonary disease, among others.[27] If the patient has a pacemaker or defibrillator, this should be interrogated prior to the planned procedure to ensure proper functioning. The preoperative evaluation should include consultation with a cardiologist experienced in the care of adolescents with CHD. Cardiac medications should be continued until the time of surgery and restarted as soon after the procedure as possible. Periods without anticoagulation should be minimized if indicated at baseline, and may require substituting warfarin with heparin in the preoperative period. The need for endocarditis prophylaxis must be considered as well; antibacterial prophylaxis prior to dental surgery, respiratory tract procedures, and procedures on infected skin and musculoskeletal structures is recommended in individuals with prosthetic heart valves, previous infective endocarditis, unrepaired CHD, repaired CHD with prosthetic material for the first 6 months after surgery, repaired CHD with residual defects, and valvulopathy after cardiac transplantation.[50]

Fluid management is important intraoperatively and post procedure, particularly in individuals who are preload dependent at baseline (eg, patients who have had Fontan palliation). Mechanical ventilation strategies with high positive end-expiratory pressure and tidal volume may decrease systemic venous return and should be monitored closely. Early mobilization and pulmonary toilet post extubation is advised to avoid pulmonary infection.

Exercise Capacity and Restrictions

The ability to exercise is an important factor in the quality of life of ACHD patients, especially in the adolescent period when participation in school and recreational athletics oftentimes functions as a social institution. Exercise ability is influenced by both real limitations imposed by limited cardiopulmonary reserve as a result of underlying pathology and by misconceptions of and anxiety about their ability to safely participate in these activities. There is evidence of diminished aerobic activity in all groups with CHD. However, symptomatic restrictions account for only approximately 30% of all barriers to exercise,[51] and some studies have shown that exercise training programs can improve functional capacity and some standards of qual-

ity of life in CHD patients, in addition to the general health benefits associated with obesity prevention.[52]

Recommendations regarding exercise capacity are often addressed at primary care visits, and should be reinforced by the patient's cardiologist. In general, most patients with repaired or mild defects can engage in moderate- to high-intensity exercise; those with more complex defects, cyanosis, or arrhythmias should be evaluated by an ACHD specialist to determine an appropriate level of activity.[27] The "exercise prescription" provided to the patient should include type of exercise tolerated as well as heart rate goals and limits. In patients with extremely limited exercise capacity, a cardiac rehabilitation program can be beneficial. The presence of significant pulmonary hypertension, cyanosis or aortic stenosis, symptomatic arrhythmias, or evidence of myocardial dysfunction usually restricts the degree of exercise; full recommendations by activity and lesion type can be found in the guidelines proposed by the 36th Bethesda Conference.[53] The importance of serial and regular evaluations is emphasized in these guidelines due to changing hemodynamic status of the patient over time as their cardiac lesions evolve and new complications arise.

Social and Psychological Impact of Chronic Illness

Living with a chronic disease can have a psychological impact on the child and transitioning adolescent. Frequent hospitalizations, physician visits, medical tests, and management of medical emergencies take a toll on the patient's self-image and self-esteem, particularly during their formative adolescent years. Adolescents with CHD often feel "different" from their peers due to their condition,[54] causing them to withhold disclosures about their heart disease to others out of fear of its impact on personal and professional relationships. Recent studies have shown that children and adolescents with CHD are at risk of internalizing problems and exhibiting behavior problems;[55] they are also more likely to have impaired quality of life secondary to their increased incidence of psychosocial difficulties.[56] The social and physical debility often experienced by patients with ACHD leads to a higher incidence of depression and anxiety in this population.[57] Studies have shown that ACHD patients are interested in psychological treatment and peer support of their mood and anxiety disorders.[58]

At least some degree of the mental health issues ACHD patients experience is thought to have a physiological basis and be related to early cyanosis and neonatal surgical bypass duration. Prolonged duration of deep hypothermic circulatory arrest (DHCA) during corrective surgery is associated with reduced social competence, and has been found to be an independent risk factor for anxiety, depression, aggressive behavior, and attention deficiencies.[59] In other studies, DHCA has been associated with decreased intellectual ability and worse fine motor skills, memory, and visuospatial skills, among other neurodevelopmental outcomes.[60-62] Psychiatric disorders have also been associated with genetic syndromes like DiGeorge syndrome.[63] This impacts executive function, leading to missed appointments, delay in clinical visits, and medication noncompliance. Given the potential for worse outcomes and risk of transition failure, primary care providers should routinely evaluate CHD patients for mood disorders and neurocognitive delay.

Social Determinants of Health and Medical Legal Partnerships

Social determinants of health and workplace discrimination play a large role in determining the ability of individuals with CHD to achieve adequate health care and maintain gainful employment. Individuals with CHD often face significant challenges as they prepare to enter the workforce, including discrimination within the workplace and maintaining employment through medical emergencies. Studies have shown that while educational milestones are similar between patients with and without CHD, those with CHD are much less likely to be employed.[64] Challenges facing adolescents as they enter the workforce include hiring discrimination, physical challenges imposed by functional limitations, and misunderstanding of disease process and actual functional capacity. Career counseling is therefore an integral part of the transitioning process and should be started in early adolescence to allow for full assessment of mental, physical, and social abilities.[65]

Medical-legal partnerships (MLPs) can be extremely beneficial to the CHD population adversely affected by social determinants of health and workplace discrimination. These partnerships integrate lawyers into health

care to address legal problems that create and perpetuate poor health; on a broader scale, these partnerships can advance and support public policy changes that improve population health.[66]

The major social determinants of health addressed by MLPs are income supports/insurance, housing/utilities, employment/education, legal status, and personal/family stability (summarized in the mnemonic I-HELP).[67] Some of the more specific areas in which MLPs may assist in the delivery of care to CHD patients include case management, translation services, health literacy, and legal aid/legal services. ACHD patients also often experience a significant loss of services, including physical, occupational, and speech therapy and nutrition services, as adult clinics may not be prepared to provide these services. While physicians can best address the individual patient's health, members of the legal system can address the systemic ailments that propagate that patient's recurrent hospitalizations and other use of medical resources. Members of the legal system are present onsite in health care settings and participate in clinical meetings, which allows a coordinated and comprehensive screening for social needs that may harm a patient's health.

Loss of insurance coverage is a major issue for transitioning patients; while adolescents with complex medical conditions are eligible for Medicaid to help cover the significant cost of their health care that goes beyond the abilities of private insurance, this eligibility ends when the patient turns 21. Additionally, the Social Security Administration re-determines supplemental security income (SSI) eligibility when the patient turns 18, and about one-third of patients lose their SSI benefits. Without appropriate guidance in navigating the nuances of insurance, many patients are at risk of losing coverage for their health care expenditures as they transition. Uninsured adults with a chronic condition are 8 times more likely to have unmet medical needs and 6 times more likely to have no access to routine care than insured young adults, with a 35% likelihood of the unmet medical need being due to cost.[68] Undoubtedly, linability to pay for health care contributes to the lack of follow-up in the adult population, and MLPs may be a valuable tool to aid in ameliorating this problem.

Studies have shown that when legal services are used to address the social determinants of health, patients with chronic illnesses such as asthma and sickle cell disease have reduced hospital admissions.[69,70] Other studies have shown utilization of MLPs has reduced spending on the care of high-need, high-use patients.[71] According to a 2016 national survey of health care organizations conducted by the National Center for Medical-Legal Partnership, 39% clinicians reported improved compliance with medical treatment and 66% reported improved health outcomes after their patients received MLP services.[72] Families referred to MLPs have shown increased access to health care, food, and income resources, and two-thirds reported improved child health and well-being.[73] Given the numerous challenges faced by patients with CHD, involving MLPs as a part of both the transition process and the patient-centered medical home benefits these patients greatly and allows them to maximize their quality of life.

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Conclusion

As more patients are living to adulthood with CHD, there is an increasing need for long-term care and adequate follow up, especially regarding the need for re-intervention and management of physiologic consequences of acquired cardiopulmonary, gastrointestinal, and renal disease in the setting of underlying congenital heart lesions. Beyond the purely medical aspects of the individual's long-term management, psychosocial issues must be addressed, including preparing the individual for future employment and family counseling. Crucial to this process is the implementation of a comprehensive transition that begins in early adolescence and enables patients to take charge of their disease process in adulthood and ultimately enables them to maximize their quality of life and societal contributions. Towards this end, the role of

MLPs may be important in ensuring that local, state, and federal policies that promote health harming norms are addressed.

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