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CARDIOLOGY (W LAI AND W ZUCKERMAN, SECTION EDITORS)

Transitioning the Pediatric Patient to the Adult Congenital Heart Disease Service

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Abstract Advances in the management of pediatric patients with congenital heart disease (CHD) have led to an increased number of adults with CHD (ACHD). While many have undergone cardiac surgery, ACHD patients have premature morbidity and mortality and often die from cardiovascular events. In order to ensure appropriate follow-up for these patients, transition to a practice with experience managing ACHD patient is crucial. Ideally, these adolescents with CHD are placed on a seamless pathway for transition, but there are multiple barriers to this at the patient, provider, and health system levels. Repeated patient and family education is one intervention that has been shown to improve follow-up and cardiovascular outcomes.

Keywords Cardiology · Pediatric · Heart disease · Congenital heart disease

Introduction

Over the last several decades, advances in the management of infants and children with congenital heart disease (CHD) have led to decreased morbidity and mortality with subsequent growth in the population of adults with CHD (ACHD). Currently, there are more adults with CHD than children [1, 2•], and it is estimated that 88.6 % of children born with CHD will survive to adulthood [3]. In addition to improved survival in general, an increase in the prevalence of patients with severe CHD has been shown in Canada between 2000 and 2010; adults made up 60 % of patients alive with severe CHD at the end of that study [2•]. While it was previously felt that much of CHD was "cured" by cardiac surgery, we now know that ACHD patients suffer premature morbidity and mortality and often die of cardiovascular events [1, 3]. Given the rising number of ACHD patients, it is not surprising that there has also been a significant increase in the number of ACHD admissions to the hospital and the amount of healthcare resource utilization between 2003 and 2012, based on United States (US) data [4]. Admissions for simple ACHD-related issues increased by 101 % over the time interval studied, while complex ACHD-related admissions increased by 53 %. Additionally, the proportion of complex ACHD-related emergency admissions also increased from 43 to 48 %.

In order to provide ongoing, effective care for these patients, it is essential that they have lifelong tailored cardiac care, preferably in an ACHD center [5]. During adolescence, patients typically transition from a pediatric health care provider to an adult health care system. This presents several potential difficulties that are offset by significant opportunities once a successful transition is completed. This transition can be daunting to young individuals with CHD because they must balance transferring from a known provider to an unknown provider while developing personal responsibility for their health care needs and medical decision making. While it would be ideal to have uninterrupted healthcare with a seamless transition between pediatric and adult providers, this is difficult to do consistently, even in the most ideal situation—this difficulty is exacerbated in

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patients who have complex CHD and multiple other medical problems. It is important that this period of transition of care be optimized to provide the best medical care possible for each patient.

Despite the known long-term risks associated with CHD, many adults have lapses in cardiac follow-up. In a multicenter US study of 922 patients, 42 % of patients with CHD had a gap in care of at least 3 years and 8 % of patients had a gap of at least 10 years [6••]. This study found that patients with increasing complexity of CHD were less likely to experience gaps in care, with 59 % of patients with mild CHD reporting a gap versus 26 % of patients with severe CHD. Reported factors most commonly associated with a gap in care included a lack of cardiovascular symptoms and the subjective notion that the patient was healthy and doing well, as well as financial factors such as unemployment or a change in health insurance. Despite the increasing prevalence of ACHD patients and well-published guidelines for their transition, a significant portion of ACHD patients are still cared for in pediatric cardiology practices [7]. Furthermore, a significant portion of ACHD patients fail to follow-up with an adult medical provider. Studies indicate that up to 76 % of ACHD patients who would be expected to follow-up at an ACHD center are lost to follow-up, which has resulted in a "virtual lost generation" [8, 9]. There is a significant amount of variation in the success of transition of care between centers, which is dependent on multiple factors, including the patient, the health care providers themselves, and the availability of health insurance. In Belgium, almost 90 % of patients were successfully transferred to ACHD providers [10]. This is a higher rate than has typically been seen in Western countries and the reasons for this are multifactorial: easier access to health care and insurance, a high population density, co-location of pediatric and adult cardiology programs, and ease of transfer of medical data due to shared medical record systems. Conversely, in Canada, 47 % of 19–21-year old were successfully transitioned to an ACHD center [11], and, in the US, at a single center with a seemingly ideal model of transition (a pediatric hospital in adult hospital setting with a well-established ACHD program), only 34 % of patients successfully transitioned to ACHD care [12].

Benefits and Successes of Transition

Given these difficulties, it is postulated that successful transition of care for patients is not merely a physical transfer from one medical facility to another, but it also includes the individual patient's development of increased responsibility and self-management, and adolescents and young adults with CHD must be empowered to take on this task [14]. There is a wide variability in the amount of ACHD follow-up needed depending on the complexity of the underlying cardiac diagnoses and the overall stability of the patient, ranging from some patients who have one visit to establish care and come back only if needed to those who are followed multiple times a year and are frequently admitted to the hospital; thus, education of the adolescent with CHD or young ACHD patient, along with their families or other individuals who provide support, is paramount [5]. The onus therefore lies with the pediatric cardiologist to introduce the concept of transition and follow through on the conversation. As such, pediatric cardiologists are encouraged to use the tools provided in the American Heart Association/American College of Cardiology consensus guidelines for ACHD care, which has been associated with improved outcomes for ACHD patients [5]. In a population-based Canadian study using a time-series analysis, an increase in referral to specialized ACHD centers was noted after the publication of national consensus guidelines for ACHD care [13••]. Concurrent with this increase in referral to ACHD centers, there was a significant decrease in expected ACHD mortality over the ensuing years. In an exploratory analysis, care in a specialized ACHD center was independently associated with decreased mortality and much of this difference appeared to be mediated by better outcomes for patients with severe CHD [13••].

With the benefits of transition to ACHD care in mind, multiple studies have evaluated factors associated with both successful transfer and gaps in care. In one US study with a transition success rate of 68 %, characteristics of patients more likely to have a successful transfer included prior cardiac surgery, cardiac medication use, and a follow-up frequency of $\leq$1 year [14]. In a Canadian trial, a 1-h nurse led teaching intervention in 15–17-year-old patients with either moderate-severe CHD or cardiomyopathy led to improved knowledge of their cardiac disease and improved scores on self-management assessments at 6 months [15•]. In a prospective, multicenter US study, an intervention consisting of the creation of a health information passport and an introduction to web resources led to improvement in the ability of patients to name their underlying cardiac disease, identify concerning symptoms and appropriate exercise, and understand birth control options and pregnancy safety [16]. Overall, effective transition of care requires a concerted effort by patients, families, physicians, nurses, and support staff with a focus on educating young patients about their cardiac issues and offering support as issues arise.

Ideal Transition Scenario

As noted previously, transition encompasses a complex process with multiple stakeholders. Successful transition relies on effective education and fostering self-

management skills, such as those described in Table 1. Given the relative infrequency of patient visits in their adolescent years, it is important that the process starts early and involves education and engagement that is ongoing throughout adolescence. The transition process should ideally start around 12 years of age, and involvement of both the patient and the parent is encouraged. Initial introductory conversations should lead to more in depth discussions, which should encompass disease knowledge, risk avoidance, and anticipatory guidance [17]. Young CHD patients should be addressed directly and are expected to have a growing role in their health care decisions. Given the varied developmental ages of patients in adolescence, it is also important to individualize the transition process to the developmental age of the patient instead of directing all patients to go down the same rigid transition pathway [17]. Additionally, patients and families may have different learning styles. It is important to present this information in a variety of ways, individualized to the patient's own learning style, while ensuring that there is an adequate opportunity for questions. An important component of transition may be the creation of a health passport for the young person that can provide health information and serve as background in both the outpatient as well as emergency room and inpatient settings [6••]. Contraception and pregnancy counseling in an integral component of the visit with female adolescents and should start early in an attempt to provide education regarding possible risks of pregnancy and to prevent miscommunication should there be a later lapse in care [18].

Despite the best efforts of both pediatric and ACHD cardiologists, lapses in care are inevitable in some patients. However, the following factors have been associated with patient's avoidance of lapses in care: poorer overall health status, attendance of pediatric appointments without parents, the belief that specialized adult care was necessary, and pediatric referral to an ACHD center [11, 19]. Although a large proportion of patients with congenital heart abnormalities show a poor level of knowledge about their heart condition [20], children more knowledgeable about their diagnosis demonstrated an improved understanding regarding the transition to adult care and were more likely to communicate directly with their providers than those who were less or not knowledgeable. This further underscores the importance of health education through the transition process [21].

Table 1 Characteristics of patients who undergo successful transition [17, 29–31]

Independent interactions with health care providers
Compliance with medication regimen and follow-up
Clear communication of health concerns
Understanding when and how to access emergency services
Understanding of underlying disease process and need for long-term follow-up
Empowerment by provider and family to "own" the transition process
Effective insurance transition
Avoidance of risky behavior
Development of coping skills

Barriers to Transition

Given the difficulties noted above with inconsistent transition to ACHD care, several specific transition barriers have been identified. These barriers involve the patient and family as well as both pediatric and adult providers and affect both the provider and the patient. For patients, the potential problems include an incomplete understanding of cardiac disease and ongoing risks, difficulty with communication, and changing insurance coverage [19, 22, 23]. In addition to these issues, many cardiac defects are associated with developmental delay due to both underlying genetic abnormalities and neurotoxic influences [24], which may limit an individual's understanding of information important for transition. In these situations, the patient's parents remain an active participant in their child's health care and may have legal authorization as a medical decision maker. ACHD programs, in general, must be understanding of, and accommodating to, this complex relationship.

For the provider, barriers include the desire to continue an established relationship and not abandon the patient as well as the misconception that complex heart conditions can only being understood by pediatric physicians. In a survey of US pediatric cardiologists [25], emotional attachment of both parents and patients to the cardiologist was identified as a barrier transition in more than 85 % of responses, and cardiologist attachment to the patient/family was identified as a barrier in 70 % of responses. Of the cardiologists surveyed, 76 % also stated that there was a lack of qualified ACHD providers, while providers affiliated with an academic institution were less likely to identify this as a barrier.

In order to ensure follow-up and help prevent long-term complications, it is important for patients to have a well-developed understanding of their specific disease process. Methods to consistently increase this understanding are not universal. While most parents have a better understanding of the CHD than the patient, this understanding is often still not ideal. In a study of 116 dyads of adolescents with CHD and their parents [26], the only determinant for parental knowledge was educational level. However, the determinants of knowledge in adolescents included both age as well

as parental knowledge. Given this, it is important to address both patients and parents with educational interventions. Many adolescents are not concerned about the process of transition, and they continue to lack sufficient knowledge about their CHD to optimally facilitate transition. The most common reasons for this include a lack of cardiovascular symptoms, being unaware that follow-up was required, as well as a complete absence from medical care [6••, 19]. One potential way to improve the education provided is to utilize alternative communication strategies such as text messaging, social media, or web-based interventions. In a cross-sectional study from Texas, all of the adolescents studied had access to the Internet via a mobile phone, but just over half had looked for information regarding CHD online. These adolescents also expressed interest in mentorship with other adolescents (90 %) and adults (60 %) with CHD either in person or via protected social media [27•]. Additionally, a small exploratory study showed that teens could effectively use text messaging for the health passport as well as having conversations regarding CHD [28].

In addition to previously mentioned potential issues, there are also coexistent financial issues. Patients over 17 years of age and those without private insurance are more likely to be admitted to the hospital via the emergency department. Additionally, the adult hospitalizations were also dispersed among a larger number of hospitals; 55 % of admissions of patients aged 21–44 years were to hospitals with fewer than 12 ACHD hospitalizations per year [22].

Summary

While it is clear that the number of adults with congenital heart disease is rising, there are significant limitations in our current ability to provide them with adequate follow-up over the long term. Lapses of care adversely affect patient outcomes and predispose these individuals to inappropriate follow-up with medical providers who may not be familiar with congenital heart disease. One opportunity for focused intervention to improve the follow-up and decrease the frequency of lapses in care in adults with CHD is repeated education targeting the patient and the patient's family while optimizing the transition to appropriate ACHD medical providers. More research is needed to evaluate the most effective way to do this, and it is likely that different methods will be effective for different patients and families.

Compliance with Ethics Guidelines

Disclosure Stephen J. Dolgner, Jason F. Deen, and Karen K. Stout declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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