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Anura Deshmukh Date

LOSS TO FOLLOW-UP IN PATIENTS WITH CONGENITAL HEART DEFECTS: FAILED TRANSITION FROM PEDIATRIC TO ADULT CARE OR A RETENTION ISSUE IN ADOLESCENCE?

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Bachelor of Science

Rutgers University

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Abstract

LOSS TO FOLLOW-UP IN PATIENTS WITH CONGENITAL HEART DEFECTS: FAILED TRANSITION FROM PEDIATRIC TO ADULT CARE OR A RETENTION ISSUE IN ADOLESCENCE?

By Anura Deshmukh

Purpose: To determine 1) how many Georgia adolescents with a Congenital Heart Defect (CHD) received continuous care from 2008-2010; 2) how many of them successfully transitioned into adult congenital care; and 3) a predictive model of risk factors for loss to follow-up and successful transition.

Method: Data from an ongoing pilot CHD surveillance project were used to identify a cohort of adolescent patients, 16-21 years old seen at Sibley Heart Center, Pediatric Cardiology Services (PCS), or Children's Healthcare of Atlanta (CHOA) during 2008 or 2009. Evidence of transitioning into adult care was searched for in Emory Healthcare, St. Joseph's Hospital, Grady Health, or Georgia Medicaid data during 2008-2010. Odds ratios were calculated using multivariable logistic regression.

*Results***:** After controlling for age, sex, insurance, proximity, CHD severity, number of procedures, and comorbidities, more than half (53.6%) of the adolescents were lost to follow-up and only about 20% successfully transitioned into adult congenital care. Being older and female predicted loss to follow-up, while severity, procedure history and having a comorbidity were protective. Being older, and having public insurance, a severe CHD, a non-CHD birth defect, and a respiratory/pulmonary comorbidity predicted successful transitioning.

Conclusion: As adolescent patients age, follow-up care and proper transitioning into an adult congenital heart defect practice must be reinforced. Implementing a national CHD surveillance program and continuing research on the factors affecting loss to follow-up and successful transitioning can help increase specialized healthcare utilization for those living with a CHD.

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List of Abbreviations:

AA	African American
ACHD	Adult Congenital Heart Disease
ASD	Atrial Septal Defects
CDC	Centers for Disease Control and Prevention
CHD	Congenital Heart Defects
CHOA	Children's Healthcare of Atlanta
ECHO	Echocardiography
EKG	Electrocardiogram
MACDP	Metropolitan Atlanta Congenital Defects Program
PCS	Pediatric Cardiology Services
PDA	Patent Ductus Arteriosus
PHI	Protected Health Information
TOF	Tetralogy of Fallot
VSD	Ventricular Septal Defects

CHAPTER I

BACKGROUND

Congenital Heart Defects

The heart is a muscle that is responsible for circulating blood and oxygen throughout the body. It is comprised of four chambers, two atria and two ventricles, four valves to prevent a backwards flow of blood, arteries that carry blood out of the heart to the rest of the body, and veins to carry blood back to the heart (1). Congenital Heart Defects (CHDs) are malformations of the heart's structure that are present at birth (2). A CHD develops when the heart or blood vessels near the heart do not develop normally in utero and result in irregular blood and oxygen circulation throughout the body (3). CHDs can range from simple problems that are easily fixed to more complex problems that are life threatening and often require immediate surgery (2). There are many different types and variations of CHDs, but some examples of common simple CHDs are: a) Atrial Septal Defects (ASD): a hole of varying sizes in the structure that usually separates the two atria; b) Ventricular Septal Defects (VSD): a hole of varying sizes in the structure that usually separates the two ventricles; c) Patent Ductus Arteriosus (PDA): irregular blood flow between the aorta and pulmonary artery; and d) Narrowed Valves: irregular flow of blood through the heart's valves. Complex CHDs are usually comprised of a combination of the CHDs mentioned above or some other more severe complication (2, 4, 5). Examples of common severe CHDs, classified as severe by Marelli et al. and Warnes et al. (see Appendix B and Appendix C), are: a) Tetralogy of Fallot (TOF): a tetrad of (i) ventricular septal defect with (ii) over-riding of the aorta, (iii) right ventricular outflow obstruction, and (iv) right ventricular hypertrophy; b) Endocardial

Cushion Defects: the walls separating all four chambers of the heart are poorly formed or absent defined as a combination of ASD and/or VSD; c) Univentricular Heart: only one ventricle forms instead of two; d) Truncus Arteriosus: only one great blood vessel or trunk leaving the heart instead of the normal two; and e) Transposition Complexes: the reversal of the normal connection of the ventricles to the great arteries (6-11).

CHDs are detected using specific diagnostic tests either in utero or postnatally. Echocardiography (ECHO) is a test that uses ultrasound waves to create a moving picture of the heart that allows the cardiologist to see the heart's structure and any problems it may have. An electrocardiogram (EKG), a test that accounts for the heart's electrical activity, is an important tool in discovering irregular heart rhythms, electrical signals passing through the heart and enlarged heart chambers. Another test is a cardiac catheterization which injects a dye into a catheter which is then inserted into the body so the clinician can determine blood flow through the heart and vessels. Other tests to help diagnose CHDs include transvaginal ultrasonography, pulse oximetry, and chest X rays (2, 3, 12).

CHDs are the most common type of birth defect (13, 14). Prevalence at birth has been estimated to be between 3.7 and 50.0 per 1,000 births in various studies (6, 15-19). Prevalence estimates can have a wide range depending on the type of CHD, the severity of the defect, and how the CHDs are ascertained or captured (18, 19). Prevalence at birth is also affected by prenatal diagnosis and treatment or voluntary pregnancy termination (20, 21). According to a study that focused specifically on the Atlanta metropolitan area, the CHD prevalence at birth for infants born between the years 1998 and 2005 was estimated to be 8.14 per 1,000 live births (17). The Metropolitan Atlanta Congenital

Defects Program (MACDP) is a population-based tracking system for birth defects that was the first of its kind in the US. Its primary purpose is to register all babies born with a congenital birth defect within the metro Atlanta area (defined as Fulton, DeKalb, Cobb, Gwinnett, and Clayton counties); however, the MACDP does not follow these babies throughout their life (22). While most clinical diagnoses of CHDs are detectable early in pregnancy, some may develop later on in utero or are diagnosed postnatally (23). An estimated 2 million people of all ages are living with a CHD in the United States as of 2010. There are slightly more females compared to males, and more adults compared to children living with a CHD (24).

Follow-up Care

Before the arrival of cardiac surgery, less than 20% of CHD patients survived to adult life (25). Today, while cardiac surgeries may still result in death (26), the current expected survival for individuals with CHDs has increased due to better outcomes following cardiac surgery and intensive care techniques. This has allowed more severely affected CHD patients to live well into adulthood (4). Some infants who have survived cardiac repair may never experience a complication, and cardiac surgery often gives the patient the perception of being "cured". Despite having surgery, however, many patients continue to experience problems such as arrhythmias, ventricular dysfunction and require more surgery (27). A 2010 study in Belgium reported that for CHD patients born between 1990 and 1992, survival to 18 years of age was 88.6%, which was significantly greater than CHD patients born in previous decades (28). Due to this increase in survival, the need for follow-up in adult care is expected to increase linearly (29). The United States and European Task Forces for CHD agree that it is important for all adolescents with a

CHD to be seen at an adult congenital heart disease (ACHD) center at least once to determine the degree and regularity of required follow-up care (30, 31); an ACDH center is a full care facility capable of providing specialized and comprehensive services to adult patients with CHDs (32, 33).

Residua, meaning "that which remains, a residue, what is left over," and sequelae, meaning "what follows or arises out of an earlier event," are terms that describe the problems that may emerge or follow a CHD surgery. Residua and sequelae could involve post-operative "electrophysiological, valvular, non-cardiovascular, ventricular and vascular" problems (34). Another serious issue resulting from CHDs is bacterial endocarditis, a condition in which the inner surface of the heart becomes inflamed or infected. Knowledge of this condition and antibiotic prophylaxis, a precautionary antibiotic treatment given to patients before dental or medical procedures who are at high risk for infection and bacterial endocarditis, is recommended for patients with CHDs (35, 36).

Follow-up care for CHD patients is important to maximize health and minimize overall health care costs. Current guidelines for follow-up recommend examination every 3-5 years for patients with simple CHDs. Patients with moderate or severe CHDs are recommended to see a specialized provider for follow-up appointments every 12-24 months (4, 32). Continuity of care is essential to avoid or treat the complications (27, 32). On average, CHD patients use more health care resources than the general population (37). Therefore, it is important to be in continuous care rather than have the patient return only when the problem has become more complex and expensive to treat.

Transition

A vulnerable time in which continuity of care is particularly challenging is when adolescents with CHDs become adults and therefore should transition from pediatric to adult cardiac care. This transition is generally considered to be necessary before the patient reaches his or her 21st birthday. (30, 32, 38-43). Transition is a multi-dimensional concept that is a "purposeful, planned process that addresses the medical, psychosocial, educational, and vocational needs of adolescents and young adults as they move from child-centered to adult-oriented health care systems" (44). In Toronto, only 47% of CHD patients successfully transferred from adolescent to adult care, and 27% of patients aged 19 to 21 reported having had no cardiac appointments at all since the age of 18 (40). In this Toronto study, diagnostic codes of eligible participants were extracted, and participants were asked to fill out a questionnaire and be interviewed. The correlates of successful transfer included living closer to an adult cardiology center, having undergone more pediatric cardiovascular operations, older age at last pediatric visit, and having been recommended to see an adult cardiologist by the pediatric provider. The investigation concluded that discussion about transition to adult care in the final pediatric session is not adequate to ensure that the adolescent transitions successfully into adult care, and that conversations should be initiated in early adolescence (40). The Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology recommends informing patients and families of the transition process as early as age 12, with an adaptable strategy of transition between ages 14-16 (43). A Report of the American College of Cardiology/American Heart Association Task Force also recommends that the transition process should start at 12 years of age, but the process should be individualized based on the patient's maturity level to prepare the patient for

transfer to adult care (30). However, it is important to note that while studies agree there is no set age when adolescents must transition to adult care, flexibility is essential for the transition to occur gradually depending on individual circumstances and the readiness of each CHD patient (30, 38, 43). After conducting a systematic review of the literature, Heery et al. report that both adolescents and health professionals appreciated modifiable timing of transfer, which supports recommendations that transfer should happen when the patient is medically stable and has the required skills to cope in an adult care facility (45, 46).

According to Higgins and Tong, transition for adolescent CHD patients must be understood from four viewpoints: the patient, the parents, the pediatric cardiologist, and the adult cardiologist (41). Heery et al. reported that adolescents who still depend on their parents for daily care and assistance valued their parent's support throughout the transition period (46). Examples of the challenges of transition include the lack of quality health care and different approaches to care between the adult care and pediatric settings (39, 42). Another problem is that 30% of all persons ages 18-24 do not have the means to pay for their health care (47). A study that evaluated California hospitalization patterns for CHD patients found that the proportion of CHD patients admitted to hospital emergency departments approximately doubled during the time of transition to adulthood. This study noted that although general hospital admissions declined for CHD patients ages 17-23, a higher proportion of emergency department admissions were seen regardless of private or public insurance status (48). Marelli et al. note that this "indicates a dispersion of care during the transition years" (33). Adolescents who do not have access to or cannot pay for adequate healthcare may resort to emergency room services during

the transition period. Other issues contributing to not transitioning include the patient's uncertainty of adult providers, the patient's lack of knowledge, migration to new cities and healthcare systems, and timing of the transfer (39, 41, 42, 47).

Loss to Follow-up

An understanding of why adolescent CHD patients drop out of care before successfully transferring into adult cardiac care is essential to increasing the likelihood that the CHD teen successfully transitions (39, 41, 42, 46-48). Adolescents and adults are frequently out of care for more than recommended periods of time (49-54). For a Canadian cohort of 643 CHD patients identified from insurance claims, 28%, 47%, and 61% of them, after their 6^{th}, 13^{th}, and 18^{th} birthdays, respectively, were reported to not have received proper follow-up care from a cardiologist (52). In another study conducted in the Netherlands, one third of adult CHD patients were lost to follow-up care (50). Differences between the two studies include dissimilarity in CHD birth prevalence of the two populations, differences in geography between Quebec and The Netherlands, variable resource availability to identify the lost CHD patients, and a difference in methodology between the two studies (50, 52, 53). In the Health Access and Research Trial Study (2013), 26% of adult patients with a severe CHD from 12 ACHD centers in the U.S. had a greater than three year gap in follow-up care (54). A study that looked at a specific severe congenital heart defect repair, Tetralogy of Fallot (TOF), from an institution in the UK between 1964-2009, found that 24% of patients were not in follow-up care (49). In a 2003 German Heart Center registry study, there were over 10,500 diagnosed or treated adult CHD patients, and of this population, 8,028 patients or over 76% had failed to return for follow-up care for more than five years (51).

Risk Factors / Predictors for Loss to Follow-up

A few studies have looked at risk factors for loss to follow-up among CHD patients (55-58). Mackie and colleagues (2012) reviewed 74 records of children or adolescents with CHDs who had previously been seen for a follow-up appointment in either a pediatric or adult center, but who had not been seen for at least three years. These 74 cases were then compared to 222 records of control patients who had been seen within three years. They found that CHD cases who were lost to follow-up were 13 times as likely to have a history of one or more missed cardiology appointments compared to controls (57). In telephone interviews conducted by trained nurse interviewers, cases lacked the knowledge of the importance of follow-up appointments and commonly saw a general practitioner instead of a specialized cardiac provider (57).

Lack of knowledge / education about follow-up or CHD continuous care seems to be a recurring theme for loss to follow-up (40, 49, 52, 56, 58). Young CHD patients may have a basic knowledge, but may lack an in depth understanding of their condition (46). Some CHD patients also expressed fear that adult CHD providers might not understand how to manage their condition (59, 60). Males were 1.5 times as likely as females to be lost to follow-up in Quebec (52). Additional risk factors included having a simple lesion versus having a severe lesion, no cardiac hospitalization before the age of six, cardiac hospitalization before the age of six, but no invasive procedure, and fewer visits to both a cardiologist and non-cardiologist (52).

CHD severity has also shown to be predictive in having a greater than three year gap in care. In a study done at 12 ACHD centers of patients 18 years and older, 59% of simple, 42% of moderate, and 26% of severe disease patients reported gaps in care (54).

Patients with more complex defects more commonly gave “changing or losing insurance” or having financial problems as reasons for discontinuity in cardiology care, whereas those with more simple CHDs gave reasons such as “lost track of time” or “decreased parental involvement” (54). Several other studies reported that patients with simple CHDs tended to believe they were no longer at risk for cardiac complications after initial treatment (27, 56, 58) or reported being told that a cardiac follow-up was not required (55). Another common reason for lapse in care was that although the patient was aware for the need of follow-up, he/she felt healthy (54, 58).

Protective Factors for Loss to Follow-up / Returning to Care

Variables protective against the loss to follow-up have also been identified in various studies (40, 49, 54, 57). These include a higher family income, cardiac catheterization within the last five years, and documentation of the need for follow-up appointments with a cardiologist (57). Those who had follow-up care in the past three years also had a better personal system of remembering cardiology appointments when compared to cases who had not had follow-up care for the past three years (57). Having had more cardiovascular surgeries in childhood and consistent compliance with dental-related antibiotic prophylaxis use were protective in another study (40). Common reasons for returning to cardiac care after a gap include the wish to avoid future health complications, a recommendation from another health care provider, and emerging symptoms or health issues (54). In a qualitative analysis, Wray proposed two major themes for attending follow-up care: 1) the fact that patients felt responsible for themselves and others to remain in cardiac care; and 2) the reassurance of well-maintained health (49).

In summary, as more adolescents are surviving to adulthood with their CHDs, transitioning to adult cardiac care is an increasingly relevant public health issue. This study aims to examine a population of adolescents with a CHD who successfully transfer into adult cardiac healthcare and those who do not. Multiple factors play a key part in whether an adolescent transfers, such as insurance status, proximity to care, and disease severity. Based on prior literature and available information, this study seeks to determine whether any of these and other factors or a combination of factors plays a significant role in whether or not the adolescents transition.

CHAPTER II

Loss to Follow-up in Patients with Congenital Heart Defects: Failed Transition from Pediatric to Adult Care or a Retention Issue in Adolescence?

Anura Deshmukh

INTRODUCTION

Congenital Heart Defects (CHDs) are the most common birth defect with approximately 2 million people in the United States living with a CHD (13, 14, 24). Prevalence at birth has been estimated to be between 3.7 and 50.0 per 1,000 births in various studies (6, 15-19). According to a study that focused specifically on the Atlanta metropolitan area, the CHD prevalence for infants born between 1998 and 2005 was estimated to be 8.14 per 1,000 live births (17). Prevalence estimates vary depending on the severity and type of CHD and method of CHD surveillance (18, 19) as well as prenatal diagnosis and voluntary pregnancy termination (20, 21). Due to better surgery, medicine, and care in recent years, approximately 90% of CHD patients are expected to live well into their adult lives (4, 28). This increase in survival has led to the need for proper follow-up cardiac care for adolescents and adults to remain in good health (29).

Transition from pediatric to adult care for CHD patients is important to the continuity of care. Transition is a multi-dimensional concept that is a "purposeful, planned process that addresses the medical, psychosocial, educational, and vocational needs of adolescents and young adults as they move from child-centered to adult-oriented health care systems" (44). Adolescents aged less than 21 are often lost during this transition because of lack of awareness of the need for follow-up care, uncertainty about the adult provider, doubt regarding the quality of care from the adult provider, or inability

to pay for follow-up care (39, 41, 42, 47). In a study by Reid et al., only 47% of CHD patients successfully transferred from adolescent to adult care with 27% of patients aged 19 to 21 reporting no cardiac appointments at all since the age of 18 (40). The U.S. and European Task Forces for Cardiology both suggest that it is important to begin having conversations about transitioning into adult follow-up care at an early age (30, 43).

A Canadian study by Mackie et al. reported that 61% of CHD patients were lost to follow-up by their 18th birthday (52). Another study reported that risk factors for loss to follow-up included a history of missed cardiology appointments, lack of education about follow-up care, and fewer visits to a cardiologist or non-cardiologist (40, 49, 52, 56-58). Additionally, patients with more simple CHDs and those who had corrective surgeries tended to think of themselves as "cured" and in no need of follow-up care (27, 58). Factors found to be protective against the loss to follow-up include increased number of cardiovascular surgeries in childhood and clinical documentation for the need of follow-up (40, 57). While gaps in care have been documented, reasons for these gaps have not been well studied, especially in the U.S. (49-54).

This retrospective cohort study design used data from 2008-2010 from Georgia-based CHD centers or congenital cardiac healthcare practices and was part of a larger ongoing pilot CHD surveillance project between collaborators from Emory University and the Centers of Disease Control and Prevention (CDC). The research aims for this study are to determine: 1) the number of adolescents living with a CHD who were lost to follow-up care between 2008, 2009 and 2010; 2) the rate of successful transition to an adult CHD provider from a pediatric CHD provider; and 3) risk factors contributing to CHD patients not transitioning from adolescent to adult care. Understanding the reasons

and risk factors for the lack of CHD follow-up amongst adolescent patients during the transition phase of care will help cardiology providers improve compliance with continuous cardiac healthcare and increase CHD healthcare utilization.

Hypotheses

1. Based on the literature, about 50% of adolescent CHD patients will be lost to follow-up during the time of transition from pediatric to adult care.
2. Having a comorbidity will result in an odds ratio of less than one, and will not be protective against loss to follow-up.
3. Adolescents with a severe CHD and with more than five procedures will be more likely to successfully transition into adult cardiac care compared to their adolescent counterparts with a minor to moderate CHD and with less than five procedures.

METHODS

Study Design and Population

This study was a secondary analysis of a retrospective surveillance of adolescents living with a CHD who were ready to transition into adult cardiology care based on recommended age guidelines. The data were obtained from a pilot CHD surveillance project at Emory University in collaboration with the Centers for Disease Control and Prevention (CDC). The study cohort consisted of adolescent patients, ages 16-21 years, who were seen at Sibley Heart Center, Pediatric Cardiology Services (PCS), or Children's Healthcare of Atlanta (CHOA) during 2008 or 2009. Evidence of their transitioning to an adult cardiologist was searched for in Emory Healthcare, St. Joseph's Hospital, Grady Health, and Georgia Medicaid claims data in 2008-2010.

Data Management and IRB

The parent study had approval from Emory University's Institutional Review Board (IRB). Prior to the start of this study, the IRB approved an amendment for this analysis (#IRB0000064051). To ensure data confidentiality, data were stored on a secure, private drive at the Emory University, Rollins School of Public Health, IT Department server system, and were accessible only by the researchers. Protected Health Information (PHI) was excluded from this dataset to maintain confidentiality and replaced with a unique identifier for each patient.

Inclusion/Exclusion Criteria

Patients included in the study were between 16 years of age and less than 22 years of age as of January 1, 2010. They could have any of the 71 ICD-9-CM CHD diagnostic codes which were collapsed as either a severe or not severe CHD by Marelli et al (6) (see Appendix B.). Patients with a history of a heart transplantation were excluded from this study.

Measures

CHD conditions defined by ICD-9-CM diagnoses codes were reported by the three adolescent provider sites (see Appendix A for case definitions by ICD-9-CM codes).

Outcome Variables

This study's primary interest was whether adolescents who had received care, defined as at least one appointment in a pediatric facility (Sibley, PCS, or CHOA) during 2008 or 2009, sought follow-up healthcare service through 2010. Those with no

evidence of care through 2010 were classified as "lost to follow-up" (primary outcome). The reference group for this outcome included all patients who did receive care sometime in 2010. Adult care was defined as at least one appointment in Emory Healthcare, St. Joseph's Hospital, Grady Health, or Georgia Medicaid claims data in 2010. Patients who transitioned to adult care (secondary outcome) were classified as "successfully transitioned." The patients who received care in 2010 but who did not have evidence of transitioning to adult care were classified as "retained in pediatric" (tertiary outcome). Since the "retained in pediatric care" group was used as the referent group for the "successful transition" group during modeling, the crude and multivariable analyses for this outcome are referenced only in Appendix D.

Predictor Variables

The following demographic information for the adolescent cohort were examined: age, sex, insurance status, and proximity to care. An attempt to examine race, weight, height, body mass index, and primary language was conducted, but these variables were sparsely reported, and were not included in the analysis.

Age

Age was computed as of 01/01/2010. All patients in the adolescent cohort were between the ages of 16 and 21 years old.

Gender

Gender was categorized as '0' Male and '1' Female.

Insurance

Insurance status was categorized as '0' Private and '1' Public Insurance. Public insurance was defined as Medicare or Medicaid. The additional insurance categories:

self-insurance, uninsured, and unknown insurance status, had extremely small sample sizes and were therefore not included in analysis.

Proximity to Care

Proximity to care was defined as ‘0’ residing outside the metro Atlanta catchment area or ‘1’ residing within the five county metro Atlanta catchment area. The five metro Atlanta counties include Fulton, DeKalb, Cobb, Gwinnett, and Clayton. The proximity to care variable served as a proxy for access to care / distance to comprehensive congenital cardiac care.

We also captured the following clinical characterizations: CHD diagnosis, severity status, procedure history, and comorbidities.

Severity

Severity status of CHD diagnoses was computed using Marelli’s classification (6) and collapsed into a bivariate variable; severity status was classified as ‘1’ severe and ‘0’ not severe based on ICD-9 codes (See Appendix B).

Procedure History

Procedure history was categorized as ‘1’ having greater than 5 distinct procedures or ‘0’ having less than or equal to 5 distinct procedures during the study period

Comorbidities

The comorbidities investigated by the parent Emory-CDC pilot surveillance study were: Diabetes Mellitus, Hypertension, Hyperlipidemia, Coronary Artery Disease, Other Cardiovascular, Other Endocrine (Non-Diabetes), Hematologic, Neoplasms / Cancer, Gastrointestinal, Renal / Other Genitourinary, Neurologic, Immunologic / Rheumatologic

/ Allergy, Musculoskeletal, Respiratory / Pulmonary, Central Nervous System, Injury / Trauma, Infectious Disease, Mental Health, and Non-CHD Birth Defects. This analysis examined the three comorbidities with the most robust sample sizes: Other Cardiovascular, Non-CHD Birth Defects, and Respiratory / Pulmonary.

A deduplication ID was initially constructed which included first name and last name, gender and date of birth, and was used to link patients across datasets and determine whether patients found in any of the pediatric service provider databases were also found in any of the adult care databases or whether the patient stayed in pediatric care (or in other words, had an encounter in one of the pediatric databases). If the unique identifier of an adolescent CHD patient was found in any of the adult care databases, that patient was considered to have successfully transitioned into adult care. Once deduplication and linking occurred, another unique identifier which retained a combination of year of birth, gender and an additional 6 encrypted digits was generated.

Data Analysis

SAS 9.4 was used for all descriptive and statistical analyses. For analysis, adolescent patients were classified into three groups: 1) those who were lost to follow-up; 2) those who successfully transitioned to adult care; and 3) those who remained in pediatric care. Chi square analyses were used for comparing proportions of these three groups and bivariate logistic regression analyses were conducted to assess the likelihood of the three outcomes associated with various predictor variables. Confounding was assessed for the three models using the crude and adjusted odds ratios. A final multivariable logistic regression model predicting the three outcome measures with all associated covariates was fit, controlling for all variables, and the backwards elimination

option was applied, constrained to a .05 p-value cut off, to identify a final logistic regression model controlling for significant variables.

RESULTS

Univariate and Bivariate Analysis

From the three adolescent data sources, 1,424 adolescents between the ages of 16-21 were included in the analysis. Table 1 displays the distribution of demographic characteristics for the adolescent cohort as well as the three outcomes: lost to follow-up, successful transition, and retained in pediatric care. Tables 2 and 3 display the crude odds ratios (ORs) and 95% confidence intervals (CIs) for the lost to follow-up and successful transition outcomes, respectively. The majority of adolescents [n=764, 53.65%] were lost to follow-up, while only 19.59% [n=279] of adolescents successfully transitioned to adult care, and 26.76% [n=381] of adolescents remained in pediatric care, [X^2= 275.5, p<0.0001].

The mean age for the entire cohort and lost to follow-up cohort was about 19 years of age, while those adolescents who successfully transitioned were slightly older approaching 20 years of age, and those who remained in pediatric care were slightly younger, closer to 18 years of age (Table 1). Older adolescents were both more likely to be lost to follow-up [OR=1.11, 95% CI (1.05, 1.17)] or to transition to adult care [OR=1.57, 95% CI (1.42, 1.72)] (Tables 2 and 3). Females were more likely to be lost to follow-up than males [OR=1.32, 95% CI (1.07, 1.63)] (Table2), but gender was not significantly associated with successful transition to adult care. The majority of adolescent patients in the cohort as well as those who were lost to follow-up, who transitioned successfully, and those who remained in pediatric care had private insurance

[75.90%, 80.90%, 56.36%, and 80.48%], respectively (Table 1). Public insurance had a 3.19 OR of successful transition [95% CI (2.25, 4.53)] and a 0.56 OR of loss to follow-up [95% CI (0.44, 0.72)] (Table 2 & 3). Additionally, while most of the cohort resided outside of the metro Atlanta area [n=818, 60.24%], proximity was not significantly associated with either of the two outcomes.

A majority of the adolescent cohort [n=866, 60.81%] was classified as having a non-severe CHD. While most of the adolescents in the lost to follow-up group had non-severe CHDs [n=597, 78.14%], almost all of the patients who successfully transitioned to adult care were classified as having a severe CHD [n=264, 94.62%]. Severe disease status was significantly associated with successful transition [OR=35.2, 95% CI (20.06, 61.76)] (Table 3). Similarly, while the majority of adolescent patients who were lost to follow-up had five or fewer distinct procedures [n=596, 78.01%], adolescent patients who successfully transitioned into adult care were those who had more than five distinct procedures [n=218, 78.14%]. There were 331 patients with missing values for comorbidities, but the comorbidity variables remained in the analysis due to their importance in other studies. A large portion of all adolescent patients had a cardiovascular comorbidity [n=922, 84.35], and similarly, the majority in all three outcome groups had a cardiovascular comorbidity. The non-CHD birth defect and respiratory/ pulmonary comorbidities were not as prevalent [36.87% and 22.05%], respectively, as the cardiovascular comorbidity. However, the comorbidity crude ORs for lost to follow-up were all significant, and less than one (Table 2). For successful transition, the crude ORs were significant for the birth defect and respiratory/pulmonary comorbidity, but not significant for the cardiovascular comorbidity (Table 3). An

ANOVA assessing differences for mean age between groups was statistically significant [p<.0001] (Table 1). A post hoc analysis using the Tukey procedure revealed that each outcome group was statistically significantly different from each other.

Confounding

Confounding is considered present if the adjusted OR is $\pm$ 10% of the crude OR. For the loss to follow-up model, sex, insurance, severity status, procedure history, birth defect comorbidity and respiratory/pulmonary comorbidity were revealed to be confounders as they each changed the crude OR by more than 10%. For the successful transition model, age, sex, insurance, severity, procedure history, and all three comorbidities were revealed to be confounders as they each changed the crude OR by more than 10%.

Multivariable Logistic Regression Modelling

Two models were constructed to assess the relationship between the predictor variables and: 1) the loss to follow-up (Table 4); and 2) the successful transition (Table 5). The logistic regression full models assessed whether each outcome could be predicted from a set of individual variables including age, gender, insurance status, proximity, severity, procedure history, and three separate comorbidities. After controlling for all variables in the full model, backwards elimination was used to reduce the number of predictor variables necessary to account for nearly as much variance as the full model.

Full Model Lost to Follow-up Outcome

$$\text{Logit P (Lost)} = \alpha + \beta_1 (\text{Age}) + \beta_2 (\text{Sex}) + \beta_3 (\text{Insurance}) + \beta_4 (\text{Proximity}) + \beta_5 (\text{Severity}) + \beta_6 (\text{Procedure History}) + \beta_7 (\text{CVD Comorbidity}) + \beta_8 (\text{BD Comorbidity}) + \beta_9 (\text{RP Comorbidity})$$

When backwards elimination was applied, insurance and proximity were dropped from the final model.

Final Model Lost to Follow-up Outcome

$$\text{Logit P (Lost)} = \alpha + \beta_1 (\text{Age}) + \beta_2 (\text{Sex}) + \beta_3 (\text{Severity}) + \beta_4 (\text{Procedure History}) + \beta_5 (\text{CVD Comorbidity}) + \beta_6 (\text{BD Comorbidity}) + \beta_7 (\text{RP Comorbidity})$$

For every one year increase in age, the odds of being lost to follow-up increased 1.18 times [95% CI (1.08, 1.28)]. Gender had a significant OR of 1.51 [95% CI (1.13, 2.01)] for loss to follow-up when adjusted for the remaining variables in the model. Severe disease status and a procedure history of greater than 5 procedures both had significant ORs of less than one. Additionally, the presence of any of the comorbidities significantly decreased the likelihood of being lost to follow-up (Table 4).

Full Model Successful Transition Outcome

$$\text{Logit P (Successful Transition)} = \alpha + \beta_1 (\text{Age}) + \beta_2 (\text{Sex}) + \beta_3 (\text{Insurance}) + \beta_4 (\text{Proximity}) + \beta_5 (\text{Severity}) + \beta_6 (\text{Procedure History}) + \beta_7 (\text{CVD Comorbidity}) + \beta_8 (\text{BD Comorbidity}) + \beta_9 (\text{RP Comorbidity})$$

When backwards elimination was applied gender, proximity, procedure history, and CVD comorbidity were dropped from the final model.

Final Model Successful Transition Outcome

$$\text{Logit P (Successful Transition)} = \alpha + \beta_1 (\text{Age}) + \beta_2 (\text{Insurance}) + \beta_3 (\text{Severity}) + \beta_4 (\text{BD Comorbidity}) + \beta_5 (\text{RP Comorbidity})$$

Adjusting for all variables in the final model, for every one year increase in age, the odds of successful transition increased by 1.66 [95% CI (1.42, 1.95)]. The odds of successful transition for public insurance was 2.12 times that of private insurance holders. The odds

of successful transition for severe disease status was 43.43 [95% CI (20.20, 93.34)] times that of those with non-severe disease status. The presence of a birth defect [OR=5.39, 95% CI (3.12, 9.32)] and a respiratory/pulmonary [OR=11.40, 95% CI (5.99, 21.68)] comorbidity showed significantly greater likelihood of successful transition (Table 5).

The logistic regression full and final model for the "retained in pediatric" outcome are found in Appendix D.

DISCUSSION

This study sought to determine which adolescent CHD patients would continue to receive congenital heart care and which would not. About half of the adolescent patients failed to receive care, and therefore, were determined to be lost to follow-up, which is consistent with the initial study hypothesis. The lost to follow-up results of the final model demonstrate that older female patients are more likely to be lost to follow-up than younger male patients; the gender finding in the current analysis contradicted the literature (52). Data also revealed that having a severe CHD, greater than five previous procedures, and the three comorbidities were protective against being lost from continuous care. This is a reasonable finding as patients with more serious and severe health problems would be more likely to transition and seek continuous healthcare for their CHD. The majority of patients who were lost to follow-up had a non-severe CHD [78.14%] and these results support prior findings that suggest people with simple CHDs may not require follow-up care every year. Therefore, it is possible that a portion of patients classified as 'lost' were not actually expected to have a follow-up or care appointment.

Of those adolescent patients who did receive continuous care, 42% transitioned to adult congenital care, while the remaining patients were retained in pediatric care. Interestingly, only about 20% of all adolescent patients successfully transitioned to adult care. The results of the final model revealed that disease severity was the strongest predictor for a successful transition. The odds of transitioning was 43 times as likely for patients with a severe CHD as for patients with a non-severe CHD. This finding is widely supported within the literature and it is known that patients with a severe CHD generally require much more care than patients with a non-severe, or simple, CHD. Patients who were older and had a non-CHD birth defect comorbidity and/or a respiratory/pulmonary comorbidity were also more likely to transition successfully to adult care. Since increased age was indicative in both the loss to follow-up and successful transition models, it may be important to urge clinicians and cardiac centers to educate their adolescent CHD patients of the importance of continuous care as they get older. Also, adolescent patients with public insurance had a higher odds of successfully transitioning into adult care than patients with private insurance. This association could be directly attributed to the Medicaid claims data or there could be a true health insurance association. A study looking at the survivorship of infants with different types of insurance found that "publicly insured infants had a 30% reduced mortality risk than that of privately insured infants during the neonatal period, but had a 30% increased risk in the post-neonatal period" (61). Also, Medicaid may provide better benefits for continuous care than many private insurances. However, information on public versus private insurance for adolescents is sparse, and this topic should be investigated further in future studies.

Strengths and Limitations

A major strength of this study was that the data were readily available due to the established on-going parent project between Emory University and the CDC. Also, the sample of adolescent CHD patients that met the inclusion criteria was robust and allowed for appropriate analyses to be conducted. The study design, a retrospective cohort, easily allowed for assessment of multiple outcomes for CHD adolescents.

In terms of limitations, this study relied on exposure variables already present due to the retrospective cohort design. Also, many of the variables were sparsely reported such as race, height, weight, body mass index, and primary language and so, they were excluded from the analysis. The number of years of data also limited results to three years and lapses in care have been reported to be as long as ten years, and so, a wider range of data years could have painted a more explanatory picture of lapses in care. Also, given the flexible recommendations and inconsistent advice regarding exactly when adolescents should transition to an adult cardiac provider, it is possible that there could be misclassification in this study. Patients who were not ready to transition yet or were not recommended to have follow-up appointments within the next year may not have been classified correctly. Also, without knowing family dynamics and other specifics of each patient, it is impossible to know if a patient was ready to transition or not, regardless of their age. While the current study looked at a number of predictor variables, other risk factors mentioned in the literature like parental involvement, patients' education/knowledge of their CHD, and recommendation from pediatric providers were not available in the data.

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TABLES

Table 1: Demographic Characteristics of Adolescent CHD Patients Seen in Pediatric Care in 2008-2010

	All Patients n (%)	**Lost to Follow-up** n (%)	**Transitioned into Adult Care** n (%)	**Retained in Pediatric Care** n (%)	**P-value**
N	1424 (100.0)	764 (53.65)	279 (19.59)	381 (26.76)	**<.0001**
Age, mean (SD)	19.12 (1.81)	19.27 (1.75)	19.76 (2.00)	18.34 (1.49)	**<.0001**
Sex					**0.0147**
Male	717 (50.35)	360 (47.12)	143 (51.25)	214 (56.17)	
Female	707 (49.65)	404 (52.88)	136 (48.75)	167 (43.83)	
Insurance Status (N=1,382)					**<.0001**
Private	1049 (75.90)	593 (80.90)	155 (56.36)	301 (80.48)	
Public	333 (24.10)	140 (19.10)	120 (43.64)	73 (19.52)	
Proximity (N=1,358)					0.1565
Outside metro Atlanta	818 (60.24)	421 (58.23)	179 (64.86)	218 (60.72)	
Within metro Atlanta	540 (39.76)	302 (41.77)	97 (35.14)	141 (39.28)	
Severity					**<.0001**
Not Severe	866 (60.81)	597 (78.14)	15 (5.38)	254 (66.67)	
Severe	558 (39.19)	167 (21.86)	264 (94.62)	127 (33.33)	
Procedure History					**<.0001**
Less than or equal to 5	785 (55.13)	596 (78.01)	61 (21.86)	128 (33.60)	
Greater than 5	639 (44.87)	168 (21.99)	218 (78.14)	253 (66.40)	
CVD comorbidity (N=1,093)					**<.0001**
Not present	171 (15.65)	115 (23.96)	22 (7.94)	34 (10.12)	
Present	922 (84.35)	365 (76.04)	255 (92.06)	302 (89.88)	
BD comorbidity (N=1,093)					**<.0001**
Not present	690 (63.13)	359 (74.79)	83 (29.96)	248 (73.81)	
Present	403 (36.87)	121 (25.21)	194 (70.04)	88 (26.19)	
RP comorbidity (N=1,093)					**<.0001**
Not present	852 (77.95)	440 (91.67)	114 (41.16)	298 (88.69)	
Present	241 (22.05)	40 (8.33)	163 (58.84)	38 (11.31)	

Note: p-values were generated from chi-square test of proportions except for age which was generated from the ANOVA. Significant p-values are bolded

Table 2: Crude ORs for the Odds of Lost to Follow-up Cohort (N=1,424)

	Estimate	SE	P-value	OR	95% CI	
Age	0.1021	0.0298	0.0006	**1.107**	**1.045**	**1.174**
Sex						
Male	---	---	---	1.00	---	---
Female	0.2793	0.1066	0.0088	**1.322**	**1.073**	**1.629**
Insurance Status *						
Private	---	---	---	1.00	---	---
Public	-0.5837	0.1273	<.0001	**0.558**	**0.435**	**0.716**
Proximity *						
Outside metro Atlanta	---	---	---	1.00	---	---
Within metro Atlanta	0.1795	0.1114	0.1072	1.197	0.962	1.489
Severity						
Not Severe	---	---	---	1.00	---	---
Severe	-1.6479	0.1181	<.0001	**0.192**	**0.153**	**0.243**
Procedure History						
Less than or equal to 5	---	---	---	1.00	---	---
Greater than 5	-2.1794	0.1227	<.0001	**0.113**	**0.089**	**0.144**
CVD comorbidity *						
Not present	---	---	---	1.00	---	---
Present	-1.1422	0.1763	<.0001	**0.319**	**0.226**	**0.451**
BD comorbidity *						
Not present	---	---	---	1.00	---	---
Present	-0.9270	0.1327	<.0001	**0.396**	**0.305**	**0.513**
RP comorbidity *						
Not present	---	---	---	1.00	---	---
Present	-1.6800	0.1862	<.0001	**0.186**	**0.129**	**0.268**

Note: Significant Odds Ratios are bolded

* n=1,382 for insurance status, n=1,358 for proximity, and n=1,093 for comorbidities

Table 3: Crude ORs for the Odds of Successful Transition Cohort (N=660)

	Estimate	SE	P-value	OR	95% CI	
Age	0.4477	0.0491	<.0001	**1.565**	**1.421**	**1.723**
Sex						
Male	---	---	---	1.00	---	---
Female	0.1978	0.1581	0.2110	1.219	0.894	1.662
Insurance Status *						
Private	---	---	---	1.00	---	---
Public	1.1607	0.1783	<.0001	**3.192**	**2.250**	**4.528**
Proximity *						
Outside metro Atlanta	---	---	---	1.00	---	---
Within metro Atlanta	-0.1769	0.1661	0.2866	0.838	0.605	1.160
Severity						
Not Severe	---	---	---	1.00	---	---
Severe	3.5610	0.2868	<.0001	**35.200**	**20.063**	**61.757**
Procedure History						
Less than or equal to 5	---	---	---	1.00	---	---
Greater than 5	0.5922	0.1810	0.0011	**1.808**	**1.268**	**2.578**
CVD comorbidity *						
Not present	---	---	---	1.00	---	---
Present	0.2662	0.2865	0.3529	1.305	0.744	2.288
BD comorbidity *						
Not present	---	---	---	1.00	---	---
Present	1.8851	0.1806	<.0001	**6.587**	**4.624**	**9.384**
RP comorbidity *						
Not present	---	---	---	1.00	---	---
Present	2.4170	0.2111	<.0001	**11.212**	**7.413**	**16.96**

Note: Significant Odds Ratios are bolded

* n=1,382 for insurance status, n=1,358 for proximity, and n=1,093 for comorbidities

Table 4: Full and Final Multivariable Logistic Model Adjusting for All Variables in Lost to Follow-up Cohort (N=1,424)

Parameter	B	SE	P-value	OR	95% CI	
Full						
Intercept	-0.7768	0.8789	0.3768	---	---	---
Age	0.1514	0.0449	0.0007	**1.163**	**1.065**	**1.270**
Female	0.4640	0.1548	0.0027	**1.590**	**1.174**	**2.154**
Public Insurance	0.2793	0.1820	0.1248	1.322	0.926	1.889
Within metro Atlanta	0.1558	0.1569	0.3209	1.169	0.859	1.589
Severe	-1.1562	0.1619	<.0001	**0.315**	**0.229**	**0.432**
Procedures	-1.2974	0.1576	<.0001	**0.273**	**0.201**	**0.372**
CVD	-1.1955	0.2363	<.0001	**0.303**	**0.190**	**0.481**
BD	-1.1647	0.1994	<.0001	**0.312**	**0.211**	**0.461**
RP	-1.0310	0.2383	<.0001	**0.357**	**0.224**	**0.569**
Final						
Intercept	-0.7642	0.8333	0.3591	---	---	---
Age	0.1619	0.0426	0.0001	**1.176**	**1.081**	**1.278**
Female	0.4088	0.1485	0.0059	**1.505**	**1.125**	**2.014**
Severe	-1.1021	0.1544	<.0001	**0.332**	**0.245**	**0.450**
Procedures	-1.3224	0.1502	<.0001	**0.266**	**0.199**	**0.358**
CVD	-1.2775	0.2309	<.0001	**0.279**	**0.177**	**0.438**
BD	-1.1173	0.1897	<.0001	**0.327**	**0.226**	**0.475**
RP	-0.8626	0.2231	0.0001	**0.422**	**0.273**	**0.653**

Note: Significant Odds Ratios are bolded

Table 5: Full and Final Multivariable Logistic Model Adjusting for All Variables in Successful Transition Cohort (N=660)

Parameter	B	SE	P-value	OR	95% CI	
Full						
Intercept	-15.8836	1.9965	<.0001	---	---	---
Age	0.6229	0.0927	<.0001	**1.864**	**1.555**	**2.236**
Female	-0.3257	0.2836	0.2507	0.722	0.414	1.259
Public Insurance	0.7395	0.3070	0.016	**2.095**	**1.148**	**3.824**
Within metro Atlanta	-0.2399	0.2873	0.4038	0.787	0.448	1.382
Severe	4.1652	0.4415	<.0001	**64.405**	**27.110**	**153.006**
Procedures	0.4397	0.3412	0.1976	1.552	0.795	3.030
CVD	-1.0984	0.5128	0.0322	**0.333**	**0.122**	**0.911**
BD	1.5200	0.3014	<.0001	**4.572**	**2.533**	**8.254**
RP	2.6419	0.3644	<.0001	**14.040**	**6.873**	**28.677**
Final						
Intercept	-14.3996	1.6849	<.0001	---	---	---
Age	0.5080	0.0805	<.0001	**1.662**	**1.419**	**1.946**
Public Insurance	0.7529	0.2914	0.0098	**2.123**	**1.199**	**3.759**
Severe	3.7710	0.3904	<.0001	**43.425**	**20.203**	**93.342**
BD	1.6851	0.2793	<.0001	**5.393**	**3.119**	**9.323**
RP	2.4339	0.3278	<.0001	**11.403**	**5.998**	**21.678**

Note: Significant Odds Ratios are bolded

FIGURES

Figure 1: Directed Acyclic Graph for Loss to Follow-up Cohort with All Considered Variables

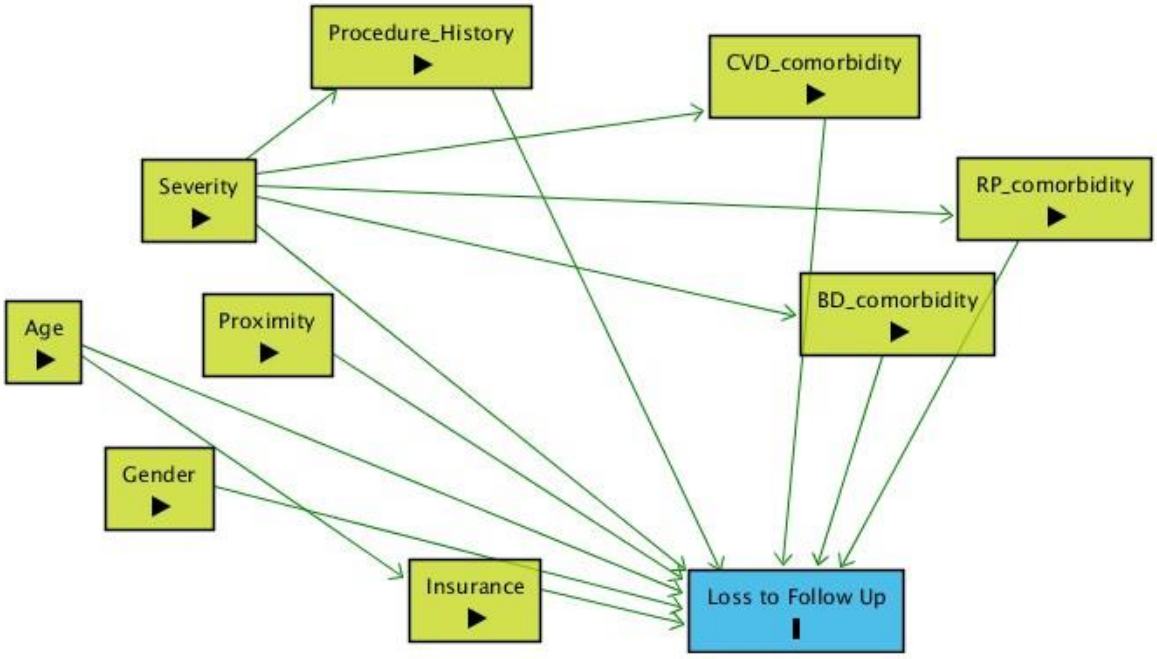
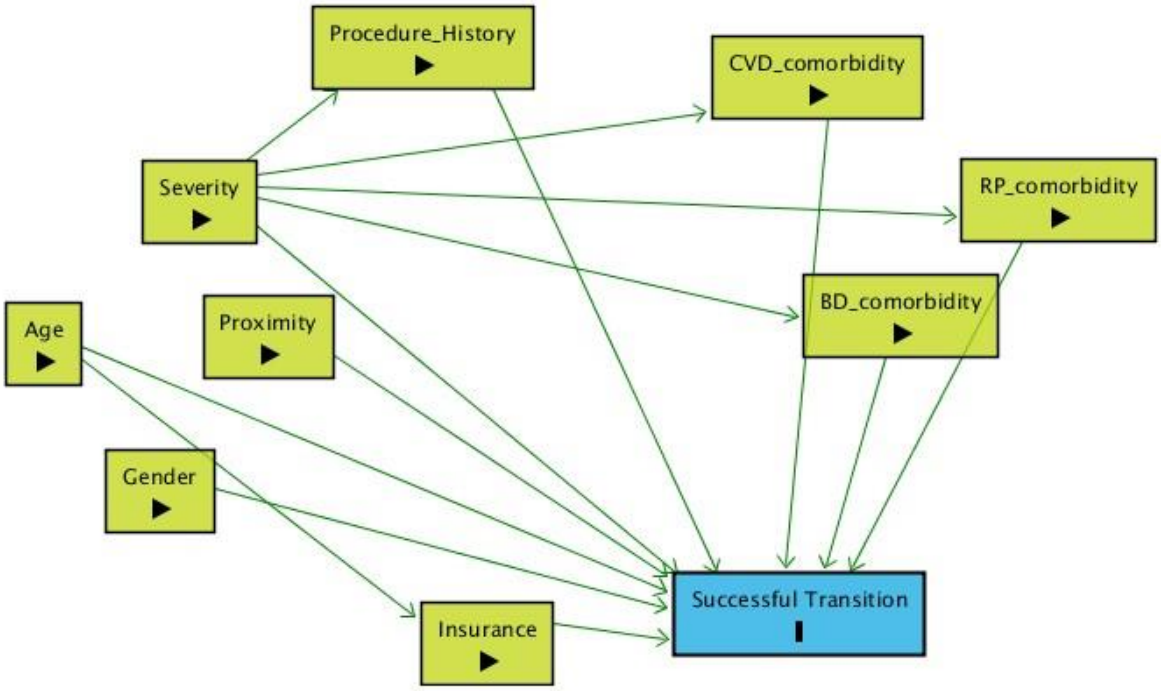


Figure 2: Directed Acyclic Graph for Successful Transition Cohort with All Considered Variables



CHAPTER III

Public Health Implication & Future Directions

This study provides a more detailed picture of adolescent patients who do and do not receive continuous care for their CHD in Georgia between 2008 and 2010. Adolescent CHD patients face a number of challenges during the transition period and their outcome can vary depending on a myriad of factors. These adolescents and young adults may successfully transfer to adult care, stay in pediatric care, or be lost to follow-up. This study uncovers some of the risk factors, like age and gender, which increase the risk of a patient being lost to follow-up. Protective factors from being lost to follow-up included presence of a comorbidity and a greater history of procedures. This study also uncovers some of the variables that contribute to a successful transfer to adult care, such as public insurance and severity. Understanding these risk factors, as well as improving factors that are out of the scope of this study, can help lead to a better continuity of cardiac care for adolescents with a CHD.

In the U.S. in 2005, due to their condition, all people living with a CHD under the age of 55 had a total of "192,000 total years not lived in good health" (62). These good health years lost are comparable to the years of good health lost due to diseases such as leukemia, prostate cancer, and Alzheimer's disease combined (62). Additionally, a healthcare cost utilization study that captured about 80% of hospital discharges found that hospital costs for children with a CHD aged 18 and under were nearly 1.5 billion dollars in 2009. Adults who were primarily treated for a CHD in 2009 had health care costs of about 280 million dollars. These outstanding costs do not include outpatient care, medications, inpatient doctor care, discharges that were not primarily for a CHD

diagnosis, and nonmedical costs to families such as transportation (63). Furthermore, it is illegal to deny health care to a child with a CHD under the age of 18, but it is estimated that 10% to 22% of adults with a CHD do not have health insurance (64). The patients who are lost to follow-up sometimes come back into the system, often in the emergency room, with comorbid conditions and complications which increase healthcare costs and decrease the number of good health years. Therefore, more studies that look at loss to follow-up and successful transition to adult care are needed to improve the generalizability of the current study and to examine other covariates that may play key parts in these outcomes so that healthcare of all CHD patients can be improved.

There is a need for more data on transition rates and loss to follow-up. Although there are some studies that document these numbers, there is still room for more studies and analysis on adolescents, especially in the United States (30, 32, 38-43, 49-52). At the Congenital Heart Public Health Consortium in 2012, it was stated that, "Because there is no population-based surveillance of CHD across the lifespan in the United States, no prevalence data are available on children, adolescents, and adults living with CHD" (65). The current study was part of a larger parent CHD surveillance pilot project between collaborators from Emory University and the Centers of Disease Control and Prevention. However, this is one of the few and very new surveillance projects that the U.S. has for CHD. It is pertinent to public health and in ensuring the continuity of care for all CHD patients who require it, for the U.S. and countries around the world to have a robust CHD surveillance program in place.

Guidelines published in 2001 recommended the ratio of regional ACHD centers to the U.S. population to be from 1 in 3 million to 1 in 5 million (32). It is estimated that

only 10% of adults who may find adult congenital health services advantageous are in those types of programs (66). There is very limited data on regional ACHD centers around the world. Additionally, there is only one comprehensive survey of ACHD centers which looked at the 6 largest centers in the world: 4 in the U.S., 1 in Canada, and 1 in Europe. Of all the registered patients at these 6 centers, 52-81% had undergone one or more reparative cardiac surgeries. The study reported that the centers all enjoyed close collaboration between both pediatric and adult cardiologists and other multidisciplinary cardiac specialists. Although there are more ACHD centers emerging, this study reports that there is a shortage of ACHDs that provide this level of comprehensive care (67). There are over one hundred ACHD centers listed on the American Congenital Heart Association website, however, they are self-identified, have not been independently verified and many of these are very new and do not offer complete, comprehensive care (66). Marelli et al. used an "epidemiologic approach to provide a framework for examining patients with CHD and the ACHD health services required to care for them" (33). After reviewing frequencies and the distribution of CHDs, Marelli et al. demonstrate that in order to improve access to specialized care for adult CHD patients, 1 regional ACHD center per 2 million adults seems to be closer to what is necessary in the U.S. (33).

The public health implications of CHD are a clear burden on the health of individuals with a CHD and the health care costs of our nation. With our current changing system for U.S. health care and insurance, it will be important to study how these changes affect adolescents with a CHD. In order to relieve some of this strain in the future, it is important that the U.S. takes some steps towards better CHD care.

Surveillance systems for CHDs that follow patients throughout their lifespan, comprehensive ACHD centers, and the recommendation for adolescents to continue care as they get older are all needed to ensure adequate congenital heart care for people of every age. Additionally, more studies examining the transition period for adolescents with a CHD are needed with special attention to certain risk factors such as insurance type.

APPENDICES

Appendix A: Congenital Heart Defects Case Definition

For an adolescent or adult with a CHD to be included, the following criteria must be met: must have at least one of the following CHD ICD-9 codes within 745-747, 648.5, 648.6, V42.1, 996.83; must have been seen in at least one of the eight healthcare facilities from which we are receiving data between 2008-2010; must be at least 11 years of age as of 1/1/2010; and must live in the state of Georgia.

Birth Defects	**ICD-9-CM Codes**
Pregnancy associated with cardiac conditions	648.5
Pregnancy associated with cardiac conditions	648.6
Bulbus cordis anomalies & anomalies of cardiac septal closure	745
Compl transposition of great vessels	745.10
Double outlet right ventricle, Dextratransposition aorta, Incomp	745.11
Corrected transposit great vessels	745.12
Transposition great vessels; other	745.19
Tetralogy of Fallot, Fallot's pentalogy	745.22
Common ventricle, Cor triloculare biatriatum, Single ventricle	745.3
Ventricular septal defect, Left ventricular-right atrial communic	745.43
Ostium secundum type atrial septal defect, Defect: atrium secundum	745.54
Atrioventricular septal defect (endocardial cushion defect)	745.6
	746.61
Endocardial cushion defects; other	745.69
Cor biloculare, Absence of atrial and ventricular septa	745.7
Bulbus cordis anomalies & cardiac septal closure; other	745.8
Other congenital anomalies heart; Pulmonary valve anomaly, unspec	746
Atresia, congenital, Congenital absence of pulmonary valve	746.01
Stenosis, congenital	746.02
Anomal pulmon valve; othr, Congen insufficiency pulmon valve, Fallot's	746.09
Tricuspid valve atresia & stenosis	746.15
Ebstein's anomaly	746.2
Congenital stenosis of aortic valve, Congenital aortic stenosis	746.3
Congenital insufficiency of aortic valve, Bicuspid aortic valve, Congenital aortic insufficiency	746.4
Congen mitral stenosis, Fused commissure mitral valve, Parachute deform mitral valve, Supernum cusps	746.5
Congenital mitral insufficiency	746.6

Hypoplastic left heart syndrome, Atresia, or hypoplasia aortic orifice/valve, hypoplasia ascend aorta & defective develop left ventricle (w mitral valve atresia)	746.75
Other specified anomalies of heart	746.85
Subaortic stenosis	746.81
Cor triatriatum	746.82
Infundibular pulmonic stenosis, Subvalvular pulmonic stenosis	746.83
Obstructive anomalies heart, NEC, Uhl's disease	746.84
Coronary artery anomal, Anomalous origin/commun coronary artery, Arteriovenous malform coronary artery: absence, aorta or pulmon	746.85
Congen heart block, Compl or incompl atrioventri [AV] block	746.86
Malposition of heart and cardiac apex, Abdominal heart, Dextrocardia, Ectopia cordis, Levocardia (isolated), Mesocardia,	746.87
Spec anomal heart; other, Atresia cardiac vein, Hypoplasia cardiac vein, Congen: cardiomegaly, divert, left ventr, pericardial defect	746.895
Unspec anomaly heart, Congen: anomaly heart NOS, heart disease NOS	746.9
Other congen anomalies circ sys	747
Patent ductus arteriosus, Patent ductus Botalli, Persist ductus arteriosus	747
Coarctation of aorta	747.1
Coarct of aorta (preductal) (postduct), Hypoplasia aortic arch	747.106
Interruption of aortic arch	747.11
Other anomalies of aorta	747.2
Anomaly of aorta, unspecified	747.2
Anomaly aortic arch, Anomal orig	747.21
Atresia & stenosis aorta, Absence or Aplasia aorta	747.22
Anomalies aorta; other, Aneurysm sinus Valsalva	747.29
Anomalies of pulmonary artery	747.3
Pulmonary artery coarct & atresia	747.31
Pulmonary arteriovenous malform	747.32
Other anomal pulmon artery & pulmon circ	747.39
Anomalies of great veins	747.4
Anomaly great veins, unspec, Anomaly NOS pulmon veins, vena cava	747.4
Total anomalous pulmon venous connection, Total anomalous pulmonvenous return [TAPVR]: subdiaphragm, supradiaphragm	747.41
Partial anomal pulmon venous connection, Part anomal pulmon venous return	747.42
Other anomalies great veins, Absence vena cava (inferior) (superior), Congen stenosis vena cava (inferior/superior), Persist: left post cardinal vein, left super	747.49

Absence/hypoplasia umbilical artery, Single umbilical artery	747.5
Other anomalies of peripheral vascular system	747.6
Other spec anomalies circulatory sys	747.8
Anomalies cerebrovascular sys, Arteriovenous malformation brain	747.81
Spinal vessel anomaly, Arteriovenous malform spinal vessel	747.82
Persistent fetal circ, Persistent pulmon hyperten, Primary pulmon hyperten newborn	747.83
Specified anomalies circ sys; other, Aneurysm, congen, spec site not elsewhere classified	747.89
Unspec anomaly circulatory sys	747.9
Heart transplant codes	V 42.1
Heart transplant codes	996.83

Appendix B. Marelli Classification Scheme

Classification adapted from Marelli et al (6).

Marelli AJ, Mackie AS, Ionescu-Ittu R, et al. Congenital heart disease in the general population changing prevalence and age distribution. *Circulation* 2007;115(2):163-72.

1.Severe

Atrioventricular Canal Defects

745.6 Endocardial cushion defects

745.60 Endocardial cushion defect, unspecified type

745.61 Ostium primum defect

745.69 Other

Absence of atrial septum

Atrioventricular canal type ventricular septal defect

Common atrioventricular canal

Common atrium

Tetralogy of Fallot

745.2 Tetralogy of Fallot

Fallot's pentalogy

Ventricular septal defect with pulmonary stenosis or atresia, dextraposition of aorta, and hypertrophy of right ventricle

Excludes: Fallot's triad (746.09)

Transposition Complex

745.1 Transposition of great vessels

745.10 Complete transposition of great vessels

Transposition of great vessels, NOS or classical

745.11 Double outlet right ventricle

Dextratransposition of aorta

Incomplete transposition of great vessels

Origin of both great vessels from right ventricle

Taussig-Bing syndrome or defect

745.12 Corrected transposition of great vessels

745.19 Other

Truncus

745.0 Common truncus

Absent septum between aorta and pulmonary artery

Communication (abnormal) between aorta and pulmonary artery

Aortic septal defect

Common aortopulmonary trunk

Persistent truncus arteriosus

Hypoplastic Left Heart Syndrome

746.7 Hypoplastic left heart syndrome: Atresia, or marked hypoplasia, of aortic orifice or valve, with hypoplasia of ascending aorta and defective development of left ventricle (with mitral valve atresia)

Univentricular Heart

745.3 Common/single ventricle or Cor triloculare biatriatum

New Additions:

747.11 Interruption of aortic arch

746.01 Congenital atresia or absence of pulmonary valve

746.1 Tricuspid atresia and stenosis, congenital, includes absence of tricuspid valve

747.41 Total anomalous pulmonary venous connection

Total anomalous pulmonary venous return [TAPVR]:
subdiaphragmatic
supradiaphragmatic

2. Shunts

Atrial Septal Defect

745.5 Ostium secundum type atrial septal defect

Defect in atrium secundum or fossa ovalis
Lutembacher's syndrome
Patent or persistent foramen ovale

Ventricular Septal Defect

745.4 Ventricular septal defect

Eisenmenger's defect or complex
Gerbode defect
Left ventricular-right atrial communication
Roger's disease
Excludes: common atrioventricular canal type (745.69) and single ventricle (745.3)

PDA

747.0 Patent/persistent ductus arteriosus

Coarctation

747.1 Coarctation of aorta

747.10 Coarctation of aorta (preductal) (postductal) hypoplasia of aortic arch

Other/unspecified defects of septal closure

745.8 Other Specified defect of septal closure

745.9 Unspecified defect of septal closure, septal defect NOS

3. Shunt + Valvar: any combination of codes in block #3 and #4

4. Valvar

Pulmonary artery

747.3 Anomalies of pulmonary artery
747.31 Pulmonary artery coarctation and atresia
Agenesis of pulmonary artery
Atresia of pulmonary artery
Coarctation of pulmonary artery
Hypoplasia of pulmonary artery
Stenosis of pulmonary artery
747.39 Other anomalies of pulmonary artery and pulmonary circulation

Pulmonary Valve
746.0 Anomalies of pulmonary valve
Excludes: infundibular or subvalvular pulmonic stenosis (746.83), tetralogy of Fallot (745.2)
746.00 Pulmonary valve anomaly, unspecified
746.02 Stenosis, congenital
746.09 Other
Congenital insufficiency of pulmonary valve
Fallot's triad or trilogy

Aortic Stenosis
746.3 Congenital stenosis of aortic valve
Excludes: congenital subaortic stenosis (746.81) or supravalvular aortic stenosis (747.22)

Aortic Insufficiency
746.4 Congenital insufficiency of aortic valve
Bicuspid aortic valve

Mitral Stenosis
746.5 Congenital mitral stenosis
Fused commissure of mitral valve
Parachute deformity of mitral valve
Supernumerary cusps of mitral valve

Mitral Insufficiency
746.6 Congenital mitral insufficiency

Ebstein Anomaly
746.2 Ebstein's anomaly

5. Other

Other unspecified anomaly of heart

745.7 Cor biloculare
Absence of atrial and ventricular septa
746.8 Other specified anomalies of heart
746.81 Subaortic stenosis
746.82 Cor triatriatum
746.83 Infundibular/subvalvar pulmonic stenosis
746.84 Obstructive anomalies of heart, NEC
Shone's syndrome

Uhl's disease
Use additional code: for associated anomalies, such as:
coarctation of aorta (747.10)
congenital mitral stenosis (746.5)
subaortic stenosis (746.81)

746.85 Coronary artery anomaly
Anomalous origin or communication of coronary artery
Arteriovenous malformation of coronary artery
Coronary artery absence, single, or arising from aortic/pulmonary trunk

746.87 Malposition of heart and cardiac apex
Abdominal heart
Dextrocardia
Ectopia cordis
Levocardia (isolated)
Mesocardia
Excludes: dextrocardia with complete transposition of viscera (759.3)

746.89 Other
Atresia of cardiac vein
Hypoplasia of cardiac vein
Congenital:
cardiomegaly
diverticulum, left ventricle
pericardial defect

746.9 Unspecified anomaly of heart
Congenital anomaly of heart NOS or congenital heart disease NOS

Other unspecified anomaly of aorta

747.2 Other anomalies of aorta

747.20 Anomaly of aorta, unspecified

747.21 Anomalies of aortic arch
Anomalous origin, right subclavian artery
Dextraposition of aorta
Double aortic arch
Kommerell's diverticulum
Overriding aorta
Persistent right aortic arch
Persistent convolutions, aortic arch
Vascular ring
Excludes: hypoplasia of aortic arch (747.10)

747.22 Atresia and stenosis of aorta
Absence of aorta
Aplasia of aorta
Hypoplasia of aorta
Stricture of aorta
Supra (valvular)-aortic stenosis

Excludes: congenital aortic (valvular) stenosis or stricture, so stated (746.3)
hypoplasia of aorta in hypoplastic left heart syndrome (746.7)

747.29 Other
Aneurysm of sinus of Valsalva
Congenital aneurysm of aorta or congenital dilation of aorta

Other anomaly of great veins

747.4 Anomalies of great veins

747.40 Anomaly of great veins, unspecified
Anomaly NOS of:
pulmonary veins
vena cava

747.42 Partial anomalous pulmonary venous connection
Partial anomalous pulmonary venous return

747.49 Other anomalies of great veins
Absence of vena cava (inferior) (superior)
Congenital stenosis of vena cava (inferior) (superior)
Persistent:
left posterior cardinal vein
left superior vena cava
Scimitar syndrome
Transposition of pulmonary veins NOS

Other unspecified anomaly of circulation

747.9 Unspecified anomaly of circulatory system

Appendix C. Warnes et al. Severity Classification of Congenital Heart Disease (4). Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. *Journal of the American College of Cardiology* 2001;37(5):1170-5.

Simple: Types of Adult Patients with Simple CHD*

Native disease
Isolated congenital aortic valve disease
Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
Isolated patent foramen ovale or small atrial septal defect
Isolated small ventricular septal defect (no associated lesions)
Mild pulmonic stenosis
Repaired conditions
Previously ligated or occluded ductus arteriosus
Repaired secundum or sinus venosus atrial septal defect without residua
Repaired ventricular septal defect without residua

∗Those patients can usually be cared for in the general medical community.

Moderate: Types of Adult Patients with CHD of Moderate Severity*

Aorto-left ventricular fistulae
Anomalous pulmonary venous drainage, partial or total
Atrioventricular canal defects (partial or complete)
Coarctation of the aorta
Ebstein's anomaly
Infundibular right ventricular outflow obstruction of significance
Ostium primum atrial septal defect
Patent ductus arteriosus (not closed)
Pulmonary valve regurgitation (moderate to severe)
Pulmonic valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm

Sinus venosus atrial septal defect
Subvalvar or supravalvar aortic stenosis (except HOCM)
Tetralogy of Fallot
Ventricular septal defect with
Absent valve or valves
Aortic regurgitation
Coarctation of the aorta
Mitral disease
Right ventricular outflow tract obstruction
Straddling tricuspid/mitral valve
Subaortic stenosis

∗These patients should be seen periodically at regional adult congenital heart disease centers.

Severe: Types of Adult Patients with CHD of Great Complexity*

Conduits, valved or nonvalved
Cyanotic congenital heart (all forms)
Double-outlet ventricle
Eisenmenger syndrome
Fontan procedure
Mitral atresia
Single ventricle (also called double inlet or outlet, common or primitive)
Pulmonary atresia (all forms)
Pulmonary vascular obstructive diseases
Transposition of the great arteries
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of atrioventricular or ventriculoarterial connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

∗These patients should be seen regularly at adult congenital heart disease centers.

Appendix D. Crude and Multivariable Analyses: Retained in Pediatric Care

Table 6: Crude ORs for the Odds of Retained in Pediatric Cohort (Compared with Successful Transition Cohort) (N=660)

	Estimate	SE	P-value	OR	95% CI	
Age	-0.4477	0.0491	<.0001	**0.639**	**0.580**	**0.704**
Sex						
Male	---	---	---	1	---	---
Female	-0.1978	0.1581	0.2110	0.821	0.602	1.119
Insurance Status *						
Private	---	---	---	1	---	---
Public	-1.1607	0.1783	<.0001	**0.313**	**0.221**	**0.444**
Proximity *						
Outside metro Atlanta	---	---	---	1	---	---
Within metro Atlanta	0.1769	0.1661	0.2866	1.194	0.862	1.653
Severity						
Not Severe	---	---	---	1	---	---
Severe	-3.5610	0.2868	<.0001	**0.028**	**0.016**	**0.05**
Procedure History						
Less than or equal to 5	---	---	---	1	---	---
Greater than 5	-0.5922	0.1810	0.0011	**0.553**	**0.388**	**0.789**
CVD comorbidity *						
Not present				1		
Present	-0.2662	0.2865	0.3529	0.766	0.437	1.344
BD comorbidity *						
Not present				1		
Present	-1.8851	0.1806	<.0001	**0.152**	**0.107**	**0.216**
RP comorbidity *						
Not present				1		
Present	-2.4170	0.2111	<.0001	**0.089**	**0.059**	**0.135**

Note: Significant Odds Ratios are bolded

* n=1,382 for insurance status, n=1,358 for proximity, and n=1,093 for comorbidities

Table 7: Full and Final multivariable logistic model adjusting for all variables in Retained in Pediatric cohort (Referent group is Successful Transition Cohort, N in model=660)

Parameter	B	SE	P-value	OR	95% CI	
Initial						
Intercept	15.8836	1.9965	<.0001	---	---	---
Age	-0.6229	0.0927	<.0001	**0.536**	**0.447**	**0.643**
Female	0.3257	0.2836	0.2507	1.385	0.795	2.414
Public Insurance	-0.7395	0.307	0.016	**0.477**	**0.262**	**0.871**
Within metro Atlanta	0.2399	0.2873	0.4038	1.271	0.724	2.232
Severe	-4.1652	0.4415	<.0001	**0.016**	**0.007**	**0.037**
Procedures	-0.4397	0.3412	0.1976	0.644	0.33	1.258
CVD	1.0984	0.5128	0.0322	**2.999**	**1.098**	**8.195**
BD	-1.52	0.3014	<.0001	**0.219**	**0.121**	**0.395**
RP	-2.6419	0.3644	<.0001	**0.071**	**0.035**	**0.145**
Final						
Intercept	14.3996	1.6849	<.0001	---	---	---
Age	-0.508	0.0805	<.0001	**0.602**	**0.514**	**0.704**
Public Insurance	-0.7529	0.2914	0.0098	**0.471**	**0.266**	**0.834**
Severe	-3.771	0.3904	<.0001	**0.023**	**0.011**	**0.049**
BD	-1.6851	0.2793	<.0001	**0.185**	**0.107**	**0.321**
RP	-2.4339	0.3278	<.0001	**0.088**	**0.046**	**0.167**

Note: Significant Odds Ratios are bolded